



INTERNATIONAL SOCIETY OF CRANIOFACIAL SURGERY **Three Decades of Innovation**

15th International Congress
September 10-14, 2013 • Jackson Hole, Wyoming

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Official Program

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15th International Congress **Three Decades of Innovation**

INTERNATIONAL SOCIETY OF CRANIOFACIAL SURGERY
September 10-14, 2013 • Jackson Hole, Wyoming

OFFICIAL PROGRAM

2013 Meeting Management Office
International Society of Craniofacial Surgery
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MATTHEW H. MEAD
GOVERNOR



STATE CAPITOL
CHEYENNE, WY 82002

Office of the Governor

July 15, 2013

International Society of Craniofacial Surgery (ISCFS)
15th Biennial Congress
Jackson, Wyoming

Dear ISCFS Members and Biennial Congress Participants,

Welcome to Wyoming and the town of Jackson. You have chosen a great place to hold your 15th Biennial Congress, and I know the week of September 9th will be both pleasant and productive for you.

You have come from around the globe to participate in an impressive program related to your health care specialty. Truly, it is an honor for our state to play host to you, as I understand the United States is the location for an international congress only once every eight years.

In whatever free time you may have, I hope you will explore all that Jackson and the surrounding areas have to offer – and they offer a lot. I say this from personal experience because Jackson is my home town and my family has ranched in Teton County for four generations. So if I were to start naming the attractions, including cultural and outdoor opportunities, available in the area, I would not be able to stop. Besides, I know the folks from the Wyoming Tourism Office and the Jackson Chamber of Commerce will provide the information you need to make the most of your stay.

My wife, Carol, and I send our best wishes to you. We appreciate the commitment it takes to attain a specialty in craniofacial surgery. We appreciate the work you do in countries around the world to make a difference in people's lives.

Have a great congress and please come back again!

Sincerely,

Matthew H. Mead
Governor

MHM:mh



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ISCFS PRESIDENTIAL WELCOME

I welcome you to attend the 15th Congress of the International Society of Craniofacial Surgery in Jackson Hole, Wyoming in September. Our record submission of abstracts for this event promises a program combining the best new work in research and clinical areas related to craniofacial surgery. Our Pre-Congress Craniofacial/Neurosurgery Symposium, a number of special panels, the award of the Tessier Medal and award for the best paper presented, our Keynote Speaker, and more than 300 free papers and posters will ensure that this congress will offer cutting edge work with innovative new thinking. Social activities to be enjoyed by the whole family - all in the midst of spectacular scenery in the Grand Tetons - will make this the best meeting you will attend this year.



The 15th Congress will be held in the Grand Teton National Park, largely recognized as the most scenic mountain range in the lower 48 states. Jackson Hole in early September combines warm days with cool evenings, and generally sunny weather. Many exciting outdoor activities are waiting for you to explore and enjoy. Our dates coincide with the 29th Annual Jackson Hole Fall Arts Festival, widely recognized as one of the premier cultural events in the Rocky Mountain West. Thousands of art enthusiasts are drawn each year to experience the diverse artwork and breathtaking natural surroundings that make Jackson Hole a leading cultural center. We encourage you to consider an extended stay in the region to visit Yellowstone National Park, just 60 miles away by car.

Getting to Jackson Hole is easy on five airlines through seven major US cities. Five marvelous hotels with a broad range of accommodation types and prices will make your stay with us most comfortable. Our venue will be Teton Village, situated 11 miles from Jackson, with easy public access to town - an authentic western community with a flavor all its own. All hotels are no more than a five minute walk from our general session meeting hall.

I hope you will make plans early to travel to the Wild West and join us for a superb educational and social experience - in a casual setting. Please, no suits or ties or evening wear!

Scott P. Bartlett, MD
ISCFS President





Welcome to Tokyo for ISCFS 2015

The 16th Biennial Congress of the International Society of Craniofacial Surgery (ISCFS) will be held in Tokyo on September 14th to 18th in 2015. I am privileged to host the next Congress in Japan.

This society has been changing for over three decades. Dr. Tessier and the founding members in the world established this particular field as an identity in plastic surgery during the first decade. In the next decade, the application of distraction osteogenesis in the craniofacial skeleton resulted in the enrollment of oral maxillo-facial surgeons. In the third decade, the society needed to increase membership giving it much more power. The very particular field of orthodontics has been clarified and the membership category of Craniofacial Orthodontics was founded in 2007. In craniofacial surgery, which is basically the realm of plastic surgeons, this trans-cranial surgery could not have grown without the cooperation of neurosurgeons and membership from neurosurgery was first encouraged in 2009. In the same year, a craniofacial research membership category was founded because of the importance of translational research between clinical craniofacial surgery and basic research. Active and associate membership has recently increased a lot indeed.

This society, as it was founded by Dr. Tessier, has been primarily led by western surgeons and it is true that there are very few Oriental members. Compared with western countries, craniosynostosis is rather rare in Asia. However, we have many cleft lip and palate, microtia and related craniofacial malformations, and skull base reconstruction cases. In 2015, Japan will be the second country in Asia to welcome the ISCFS Congress. Dr. Yu-Ray Chen held this congress in Taiwan in 1999. Japan has hosted many congresses including plastic surgery. It is our Japanese colleagues' earnest wish to provide a memorable event for ISCFS members and we will make every effort to provide a successful ISCFS congress in two years.

We invite you to come and join us. Tokyo is one of the biggest cities in the world. The congress site of Maihama Tokyo Bay is a very special place located between Tokyo and Chiba - where Chiba University is located. The venue is in a coastal area with much sunshine. Tokyo station is 16 minutes by train. We have many beautiful places in Japan to take a short trip, before or after the congress, such as Yokohama, Kamakura, Nikko, Mt. Fuji, and even Kyoto. We are sure you will enjoy our beautiful country and are very proud of the hospitality shown to foreigners by our people. You will certainly enjoy in particular the old and modern Far East Asian Japanese culture that is so different from yours.

Welcome to Tokyo, Japan in September in 2015.



Kaneshige Satoh, MD, PhD
Vice President of ISCFS
Department of Plastic, Reconstructive, and Aesthetic Surgery
Chiba University, Chiba, Japan



ISCFS Program Committee

Nivaldo Alonso, MD – Brazil
Eric Arnaud, MD – France
Stephen B. Baker, MD, DDS – United States
Scott P. Bartlett, MD – United States
Stephen Beals, MD – United States
James Bradley, MD – United States
Steven R. Buchman, MD – United States
Juan Martin Chavanne, MD – Argentina
Yu-Ray Chen, MD – Taiwan
Michael Cunningham, MD, PhD – United States
Jeffrey Fearon, MD – United States
Christopher R. Forrest, MD – Canada
Mirko S. Gilardino, MD – Canada
Dov C. Goldenberg, MD, PhD – Brazil
Joseph S. Gruss, MD – United States
Craig Hobar, MD – United States
William Y. Hoffman, MD – United States
Larry Hollier, MD – United States
Richard A. Hopper, MD – United States

Joseph Losee, MD – United States
Anil Madaree, MD – South Africa
Xiongzhen Mu, MD – China
Frank Papay, MD, FACS – United States
John A. Persing, MD – United States
Juan Carlos Rodriguez, MD – Argentina
Gary F. Rogers, MD – United States
Kaneshige Satoh, MD – Japan
Ramesh Sharma, MD – India
Davinder J. Singh, MD – United States
Henry Spinelli, MD – United States
Sherard A. Tatum, MD – United States
Jesse A. Taylor, MD – United States
Mark M. Urata, MD – United States
John van Aalst, MD – United States
Steven Wall, MD – United Kingdom
S. Anthony Wolfe, MD – United States
Vincent Yeow, MD – Singapore

ISCFS Officers: 2011 - 2013

President

Scott P. Bartlett, MD - United States

Vice President

Kaneshige Satoh, MD - Japan

Secretary-Treasurer

Eric Arnaud, MD - France

Immediate Past President

Anil Madaree, MD, FRCS - South Africa

Past President

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Council Members

Nivaldo Alonso, MD - Brazil

Stephen Beals, MD - United States

Xiongzhen Mu, MD - China

Juan M. Chavanne, MD - Argentina

Congress President

Scott P. Bartlett, MD - United States



GENERAL INFORMATION



GENERAL INFORMATION

Registration

The Congress Registration desk hours are:

Tuesday, 10 September 2013	7:00 am - 5:00 pm
Wednesday, 11 September 2013	7:00 am - 5:00 pm
Thursday, 12 September 2012	7:00 am - 5:00 pm
Friday, 13 September 2012	7:00 am - 12:00 pm
Saturday, 14 September 2012	7:00 am - 5:00 pm

Speaker Ready Room

The main Speaker Ready Room is located in the Walk Festival Hall. A secondary Speaker Ready area will be at the back of the ballroom at the Four Seasons Hotel.

Hours are:

Tuesday, 10 September 2013	7:00 am - 5:00 pm
Wednesday, 11 September 2013	7:00 am - 5:00 pm
Thursday, 12 September 2012	7:00 am - 5:00 pm
Friday, 13 September 2012	7:00 am - 12:00 pm
Saturday, 14 September 2012	7:00 am - 5:00 pm

Registration Entitlements

- Entry to all scientific sessions and exhibit hall
- Daily Lunch
- Morning and Afternoon Refreshments
- Congress Bag
- Copy of Program/Abstract Book
- Welcome Reception

Exhibit Hall Hours

All registrants are invited to visit the exhibits located in the Hotel Terra Ballroom. The exhibits will be open:

Wednesday, 11 September 2013	7:00 am - 5:00 pm
Thursday, 12 September 2012	7:00 am - 5:00 pm
Friday, 13 September 2012	7:00 am - 10:00 am
Saturday, 14 September 2012	7:00 am - 12:00 pm

Banks

Banks in Jackson Hole are open from 9:00 am - 5:00 pm Monday to Friday. Most banks are open on Saturday from 9:00 am - 12:00 pm. Automatic Teller Machines are open 24 hours and are located at the airport, all hotels and in town. Facilities for exchange of currency are not available at the Jackson Hole airport. We recommend changing funds at a major airport while you are in transit or before leaving home.

Electricity

Electricity in the US is 120 Volts, alternating at 60 cycles per second. If you travel to the US with a device that does not accept 120 Volts at 60 Hertz, you will need a voltage converter.

Travel and Program Disclaimers

In the event of any travel disruptions, Congress Organizers will not be held responsible for any losses incurred by registrants en route to or from the Congress. The program is correct at the time of printing. Congress Organizers reserve the right to alter the program as necessary.

Liability

Congress Organizers are not liable for personal accidents, losses or damage to the private property of registered attendees or any accompanying persons during the Congress. Please make your own arrangements with respect to personal insurance.

Congress Attire

Congress Sessions: Casual

Welcome Reception: Casual

Council Dinner (*by invitation only*): Casual

Farwell Dinner: Casual Western Wear – Cowboy Boots are a good idea!

Mobile Phones and Pagers

As a courtesy to speakers and other registrants, we request that all mobile phones and pagers are switched off or muted before entering sessions.

No Smoking Policy

Registrants and guests should be aware that smoking is not permitted in public buildings and many hotels and restaurants including the Congress venue. Smoking is prohibited in the meeting and exhibit rooms at all times.

Program Content Disclaimer

The content of this program is presented solely for educational purposes for use by medical practitioners. This material is intended to express the opinions, techniques or approaches of the presenters. Sponsorship of this program and/or advertising by any company or organization are not to be construed, in any fashion, as an endorsement of the materials or products presented.

Topic and Abstract Disclaimer

All presentation titles and abstracts are printed as submitted by the speakers. ISCFS cannot be responsible for errors in spelling or content.

Recording Policy

Recording (photographic, video and audio) of the sessions, entertainers and exhibit hall during the ISCFS Congress is strictly prohibited.

PROGRAM AT-A-GLANCE



Tuesday, 10 September

7:00 am - 5:00 pm	Speaker Ready Room	Walk Festival Hall & Four Seasons Ballroom
7:00 am - 5:00 pm	Registration	Bridger Center
7:00 - 8:00 am	Continental Breakfast	All Hotels
8:30 am - 5:30 pm	Pre-Congress Symposium: Controversy and Consensus in Craniosynostosis and Cranial Vault Surgery	Four Seasons Ballroom
11:00 - 11:30 am	Coffee Break	Four Seasons Ballroom
12:30 - 1:30 pm	Lunch	Four Seasons Ballroom
3:30 - 4:00 pm	Coffee Break	Four Seasons Ballroom
5:30 - 7:00 pm	Symposium Reception	Four Seasons Ballroom
8:00 - 10:00 pm	ISCFS Council Meeting	Four Seasons Ballroom

Wednesday, 11 September

7:00 am - 5:00 pm	Speaker Ready Room	Walk Festival Hall & Four Seasons Ballroom
7:00 am - 5:00 pm	Registration	Bridger Center
7:00 - 8:00 am	Continental Breakfast	All Hotels
8:00 - 8:15 am	Opening Remarks	Walk Festival Hall
8:15 - 9:00 am	Eulogies	Walk Festival Hall
9:00 - 9:45 am	Keynote Address	Walk Festival Hall
9:45 - 10:15 am	Coffee Break	Hotel Terra/Commons Tent
10:15 - 11:30 am	Founders Session	Walk Festival Hall
11:30 - 11:45 am	Presentation of the Tessier Medal	Walk Festival Hall
11:45 am - 12:00 pm	Earthfire Institute Presentation	Walk Festival Hall
12:00 - 1:00 pm	ASCFS Business Meeting	Walk Festival Hall
12:00 - 1:00 pm	Lunch	Hotel Terra/Commons Tent
1:00 - 3:00 pm	General Session 1 Hypertelorism, Craniofacial Tumors & Encephaloceles, Rare Craniofacial Clefts	Walk Festival Hall
3:00 - 3:30 pm	Coffee Break	Hotel Terra/Commons Tent
3:30 - 5:00 pm	General Session 2 Non-Syndromic Synostosis Part I	Walk Festival Hall
5:30 - 9:00 pm	Welcome Reception <i>There will be a 15 minute gondola ride up to the restaurant</i>	Bridger Restaurant - Couloir



Thursday, 12 September

7:00 am - 5:00 pm	Speaker Ready Room	Walk Festival Hall & Four Seasons Ballroom
7:00 am - 5:00 pm	Registration	Bridger Center
7:00 - 8:00 am	Continental Breakfast	All Hotels
8:00 - 8:45 am	Ortiz Monasterio Memorial Lecture	Walk Festival Hall
9:00 - 10:00 am	Concurrent Session 1A Basic Science Research	Walk Festival Hall
9:00 - 10:00 am	Concurrent Session 1B Craniofacial Orthodontics & Orthognathic Surgery	Four Seasons Ballroom
9:00 - 10:00 am	Concurrent Session 1C Panel: Craniofacial Microsomia: Ethics, Embryos, Management and Outcomes	Four Seasons Ballroom
10:00 - 10:30 am	Coffee Break	Hotel Terra/Commons Tent
10:30 am - 12:00 pm	Concurrent Session 2A Basic Science Research	Walk Festival Hall
10:30 am - 12:00 pm	Concurrent Session 2B Craniofacial Orthodontics & Orthognathic Surgery	Four Seasons Ballroom
10:30 am - 12:00 pm	Concurrent Session 2C Genetic and Non-Surgical Aspects of Craniofacial Conditions	Four Seasons Ballroom
12:00 - 1:30 pm	Lunch	Hotel Terra/Commons Tent
12:00 - 1:30 pm	ISCFS Biennial Business Meeting	Walk Festival Hall
1:30 - 3:30 pm	Concurrent Session 3A Aesthetic Surgery of the Craniofacial Skeleton & Craniofacial Trauma	Walk Festival Hall
1:30 - 3:30 pm	Concurrent Session 3B Craniofacial Microsomia/Related Conditions	Four Seasons Ballroom
1:30 - 3:30 pm	Concurrent Session 3C Craniofacial Imaging	Four Seasons Ballroom
4:00 - 5:30 pm	Poster Session	Hotel Terra
6:30 - 7:30 pm	ASCFS Reception	Four Seasons Ballroom
7:30 - 10:00 pm	ISCFS Council Dinner (by invitation)	Four Seasons
8:00 - 10:00 pm	Seattle Children’s Craniofacial Fellowship Alumni Dinner	Alpenhof Lodge



Friday, 13 September

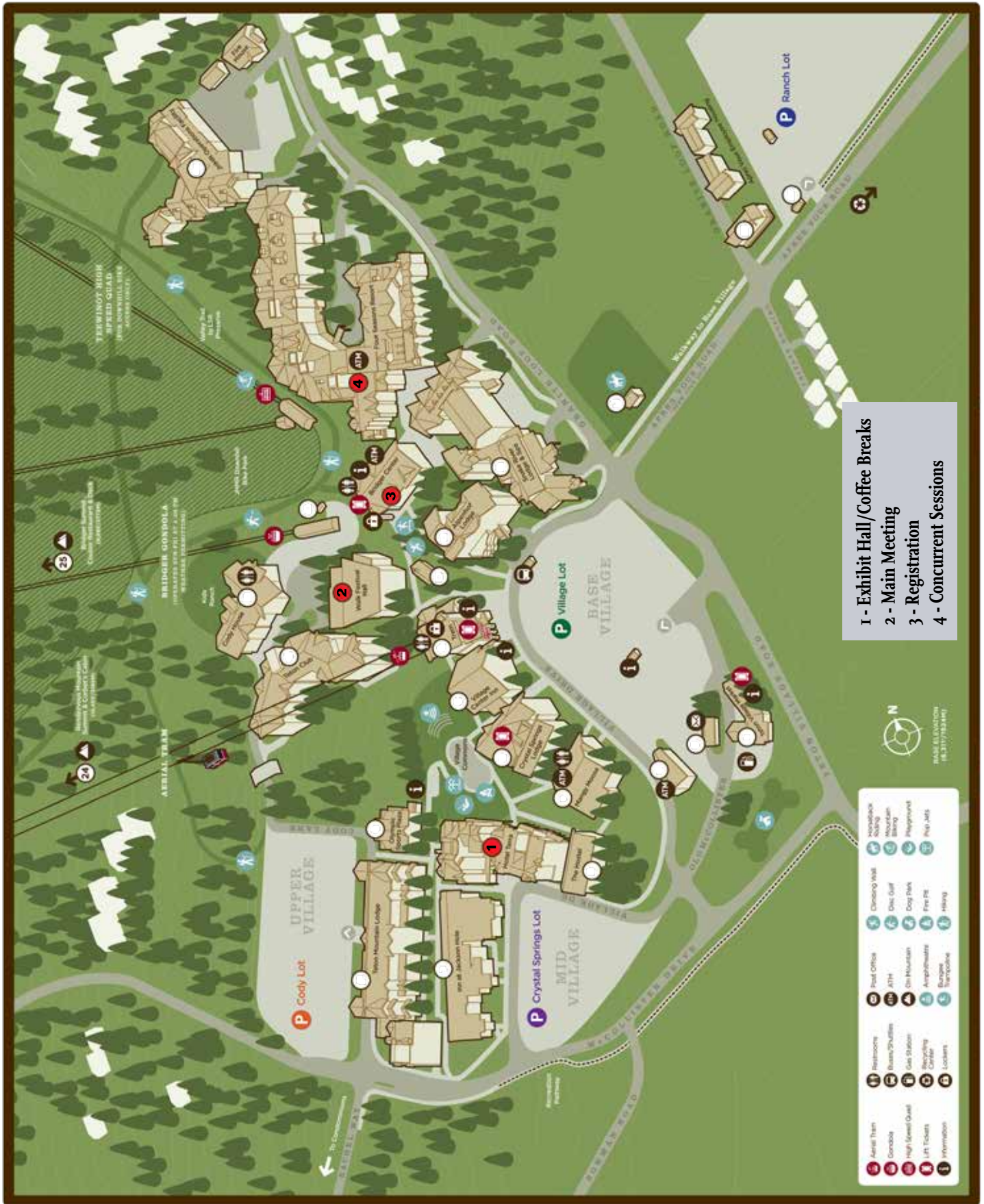
7:00 am - 5:00 pm	Speaker Ready Room	Walk Festival Hall & Four Seasons Ballroom
7:00 am - 12:00 pm	Registration	Bridger Center
7:00 - 8:00 am	Continental Breakfast	All Hotels
7:30 - 9:30 am	Concurrent Session 4A Basic Science Research	Walk Festival Hall
7:30 - 9:30 am	Concurrent Session 4B Non-Syndromic Craniosynostosis II - A	Four Seasons Ballroom
7:30 - 9:30 am	Concurrent Session 4C Non-Syndromic Craniosynostosis II - B	Four Seasons Ballroom
10:00 am - 12:00 pm	Innovation and Invention in Craniofacial Surgery - Open Optional Seminar	Walk Festival Hall
7:00 - 9:00 pm	Penn Alumni Dinner	Mangy Moose
7:00 - 9:00 pm	WCF Partner Dinner (by invitation)	Alpenhof Lodge

Saturday, 14 September

7:00 am - 5:00 pm	Speaker Ready Room	Walk Festival Hall
7:00 am - 5:00 pm	Registration	Bridger Center
7:00 - 8:00 am	Continental Breakfast	All Hotels
8:00 - 10:00 am	General Session 3 Craniofacial Distraction	Walk Festival Hall
10:00 - 10:30 am	Coffee Break	Hotel Terra/Commons Tent
10:30 am - 12:00 pm	General Session 4 Craniofacial Distraction	Walk Festival Hall
12:00 - 1:00 pm	Lunch	Hotel Terra/Commons Tent
1:00 - 3:00 pm	General Session 5 Syndromic Craniosynostosis	Walk Festival Hall
3:00 - 3:30 pm	Coffee Break	Hotel Terra/Commons Tent
3:30 - 5:00 pm	General Session 6 Syndromic Craniosynostosis	Walk Festival Hall
6:00 - 9:00 pm	Farewell Dinner - Tickets Required	Commons Tent



Photo credit: Robert Trueman Photography



1 - Exhibit Hall/Coffee Breaks
2 - Main Meeting
3 - Registration
4 - Concurrent Sessions

- 1 Restrooms
- 2 Bus/Charters
- 3 Gas Station
- 4 Recycling Center
- 5 Lockers
- 6 Information
- 7 Kiosk/Thru
- 8 Concessions
- 9 High Speed Quad
- 10 Lift Tickets
- 11 Post Office
- 12 ATM
- 13 Gas Station
- 14 Amphitheater
- 15 Bungee
- 16 Zip Line
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The Master of Advanced Surgery in Cranio-maxillofacial Surgery

The Master of Advanced Surgery in Cranio-maxillofacial Surgery is a one-year program that is ideally suited for those who have completed a surgical training program. In conjunction with the Australian Craniofacial Unit in Adelaide, ASAM offers the degree to Fellows of the Royal Australasian College of Surgeons (RACS) (or equivalent for overseas applicants).

Graduates will demonstrate mastery of surgical competence in cranio-maxillofacial surgery, acquisition of life-long learning skills and a capacity for leadership.

Modules in the degree include:

- advanced management of cleft lip and palate
- advanced management of craniosynostoses
- advanced management of rare craniofacial clefts
- advanced multidisciplinary care of craniofacial defects
- craniofacial tumours
- cranio-maxillofacial trauma
- degenerations of the craniofacial skeleton
- dento-facial deformities



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Australian Craniofacial Unit

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CONGRESS PROGRAM



PROGRAM SCHEDULE

The scientific program is correct at the time of printing; however, the Program Committee reserves the right to alter the schedule as necessary.

Tuesday, 10 September - Four Seasons Ballroom

7:00 am - 5:00 pm Registration

7:00 - 8:00 am Continental Breakfast

PRE-CONGRESS SYMPOSIUM: CONTROVERSY AND CONSENSUS IN CRANIOSYNOSTOSIS AND CRANIAL VAULT SURGERY

CO-CHAIRS: *Scott Bartlett, MD & Stephen Beals, MD*

8:30 - 9:45 am

How is Intracranial Pressure Defined?

Moderators: *Hal Rekate, MD (USA) & John Persing, MD (USA)*

Panelists: *Brent Collett, PhD (USA)*

Steven Wall, MD (United Kingdom)

Steven Newman, MD (USA)

This session will focus on objective assessment measures to document neural function in children with craniosynostosis, and possible warning signs to indicate need for treatment. For instance, what is elevated intracranial pressure in young children? What is its significance? What is the clinical implication of a beaten copper appearance on a skull radiograph? What is the best method to measure cognitive neurologic function that predicts cognition in school years? What is the significance of protrusive skull deformities seen following previous cranioplasty surgery for craniosynostosis? What is the sensitivity/reliability of papilledema in the neurologic assessment of a child with possible increased ICP and craniosynostosis?

9:45 - 11:00 am

How is Intracranial Pressure Managed?

Moderators: *Stephen Dover, FDSRCS, FRCS (United Kingdom) & Jay Storm, MD (USA)*

Panelists: *Richard Hayward, MD (United Kingdom)*

Gerry Grant, MD (USA)

Jayaratnam Jayamohan, BSc, FRCS (United Kingdom)

Panelists will discuss different management techniques for patients diagnosed with elevated intracranial pressure and whether the increased pressure is uniform throughout the vault. They will discuss timing (shunt v. vault expansion) as it applies to age of the patient and severity of the symptoms, and the pros and cons of the different techniques. They will also explore the potential role of vault expansion in craniofacial patients who have developed slit ventricle syndrome. The session will also include the management of recurrent raised intra-cranial pressure despite previous interventions. It is hoped that this session will promote debate with colleagues in the audience.

11:00 - 11:30 am

Coffee Break

11:30 am - 12:30 pm

How Do We Improve Quality and Reduce Risk in Craniosynostosis Surgery?

Moderators: *Jeffrey Fearon, MD (USA) & Paul Stricker, MD (USA)*

Panelists: *Jack Yu, MD (USA)*

Marcin Czervinski, MD (USA)

Franklyn Cladis, MD (USA)

Kevin Cook, MD (USA)

Rachel Ruotolo, MD (USA)

Jennifer Rhodes, MD (USA)

This multidisciplinary panel will examine perioperative risk and quality improvement in craniosynostosis surgery. Significant perioperative morbidities in the modern era will be discussed, as well as strategies to minimize these risks, with a focus on blood loss, transfusion, and the optimal use of diagnostic imaging modalities. These discussions will be put in the broader context of how quality is defined in craniosynostosis surgery and identification of targeted outcomes for improvement efforts.



12:30 - 1:30 pm	Lunch
1:30 - 2:30 pm	<p>What are the Best Treatment Choices for Skull Defects? Moderators: <i>Davinder Singh, MD (USA) & Federico Di Rocco, MD, PhD (France)</i> Panelists: <i>Matthieu Vinchon, MD, PhD (France)</i> <i>Stephen Dover, FDSRCS, FRCS (United Kingdom)</i> <i>David J. David, MD, FRACS (Australia)</i></p> <p>International experts in craniofacial and neurosurgery will address the management of congenital and iatrogenic skull defects in both pediatric and adult patients. Surgical indications and techniques including autologous and alloplastic reconstruction, as well as long term results and complications, will be discussed in depth. The session will conclude with challenging case presentations and guidelines for skull defect reconstruction.</p>
2:30 - 3:30 pm	<p>What is the Role of Minimally Invasive Surgery in the Management of Craniosynostosis Surgery? Moderators: <i>Mark Proctor, MD (USA) & Irene Mathijssen, MD, PhD (Netherlands)</i> Panelists: <i>Eric Arnaud, MD (France)</i> <i>MarieLise van Veelen, MD (Netherlands)</i> <i>Gray Rogers, MD (USA)</i></p> <p>Looking at new trends in craniosynostosis surgery, this panel will focus on minimally invasive surgery including endoscopic techniques and the use of springs in both isolated and syndromic craniosynostosis. The discussion will present the pros and cons of this development, concentrating on associated complications, blood loss, and functional and aesthetic outcome parameters. This session leaves plenty of room for an in depth debate among the experts.</p>
3:30 - 4:00 pm	Coffee Break
4:00 - 5:00 pm	<p>What are New Perspectives in Craniosynostosis Surgery? Moderators: <i>Richard Hopper, MD, MS (USA) & Richard Hayward, MD (United Kingdom)</i> Panelists: <i>Irene Mathijssen, MD (Netherlands)</i> <i>Christopher Forrest, MD (Canada)</i> <i>MarieLise van Veelen, MD (Netherlands)</i> <i>Jeffrey Fearon, MD (USA)</i> <i>David Dunaway, MD (United Kingdom)</i></p> <p>In this concluding session, an expert panel will present different perspectives on three topics: cranial expansion in the very young, treatment of Chiari malformation with craniosynostosis, and approach to the Apert midface. The presentations will be focused and brief to allow time for audience discussion and panel debate.</p>
5:00 - 5:30 pm	Open Forum Discussion
5:30 - 7:00 pm	Symposium Reception
8:00 - 10:00 pm	ISCFs Council Meeting



Wednesday, 11 September

7:00 am - 5:00 pm Registration

7:00 - 8:00 am Continental Breakfast

8:00 - 8:15 am **Opening Remarks**
Scott Bartlett, MD - ISCFS President (USA)

8:15 - 9:00 am **Eulogies**
Joseph Murray, MD, FACS (USA)
Speaker: Mutaz Habal, MD, FACS, FRCSC (USA)
Alexander Stratoudakis, MD (Greece)
Speaker: Steven Wall, MD (United Kingdom)
Fernando Ortiz Monasterio, MD (Mexico)
Speaker: Fernando Molina, MD (Mexico)
Daniel Marchac, MD (France)
Speakers: Eric Arnaud, MD (France) & Bryant Toth, MD (USA)

9:00 - 9:45 am **Keynote Address**
Some Favorite Rocky Mountain Stories
Speaker: Dr. Harold J. Stearns (USA)

9:45 - 10:15 am Coffee Break

10:15 - 11:30 am **Founders Session**
Introduction: Scott Bartlett, MD (USA)
What is the Future of Craniofacial Surgery?
Speaker: Kenneth E. Salyer, MD (USA)
Craniofacial Surgery: From Spectacular Innovation to Sound Healthcare Delivery
Speaker: David J. David, AC, MD, FRCS, FRACS (Australia)
Craniofacial Distraction: How It Started and Where It's Going
Speaker: Joseph G. McCarthy, MD (USA)
Tessier's Classification of Facial Clefts and the Unfinished Symphony
Speaker: S. Anthony Wolfe, MD, FACS, FAAP (USA)
Beyond the Surgery
Speaker: Linton Whitaker, MD (USA)

11:30 - 11:45 am **Presentation of the Tessier Medal**
Speaker: Scott Bartlett, MD - ISCFS President (USA)

11:45 am - 12:00 pm **Earthfire Institute Introduction**
Speaker: Susan Eirich, PhD

12:00 - 1:00 pm Lunch

12:00 - 1:00 pm ASCFS Business Meeting - *Walk Festival Hall*

1:00 - 3:00 pm **General Session 1**
Hypertelorism, Craniofacial Tumors & Encephaloceles, Rare Craniofacial Clefts
Moderators: *Dov Goldenberg, MD, PhD (Brazil) & Robert Havlik, MD (USA)*

1:00 pm **I**
FIBROUS DYSPLASIA OF THE ZYGOMATICOMAXILLARY REGION: OUTCOMES OF SURGICAL INTERVENTION
Presenter: Phuong D. Nguyen, MD
Affiliation: University of California Los Angeles
Authors: Andrews BT, Gabbay JS, Yuan JT, Kawamoto HK, Bradley JP



- 1:07 pm 2
CRANIOFACIAL AND HEAD, AND NECK NEUROFIBROMATOSIS: CLINICAL CLASSIFICATION AS AN AID TO SURGICAL TREATMENT
 Presenter: Joseph S. Gruss, MD
 Affiliation: Seattle Children’s Hospital
 Authors: Gruss JS, Latham KL, Buchanan EB, Suver DS
- 1:14 pm 3
IMPROVEMENT OF TREATMENT OF FACIAL ARTERIO-VEINUS VASCULAR MALFORMATIONS BY MULTIPLE EMBOLIZATIONS: OUTCOMES IN A SERIES OF 31 PATIENTS
 Presenter: Dov C. Goldenberg, MD, PhD
 Affiliation: Hospital das Clinicas University of Sao Paulo Medical School
 Authors: Goldenberg DC, Hiraki PY, Puglia P, Caldas JG, Ferreira MC
- ~~1:21 pm 4~~
~~**THE CHOICE OF SKULL BASE RECONSTRUCTION VARIANT IN THE TREATMENT OF TUMORS WITH CRANIOFACIAL LOCALIZATION**~~
NOT PRESENTED
~~Presenter: Igor Reshetov, MD~~
~~Affiliation: Moscow PA Hertenzen Cancer Research Institute~~
~~Authors: Reshetov I, Chissov VI, Cherekaev VA, Zaitzev AM, Belov AI, Polyakov AP, Matorin OV, Ratushniy MV~~
- 1:28 pm 5
TREATMENT OF CRANIOMAXILLOFACIAL FIBROUS DYSPLASIA IN OUR DEPARTMENT
 Presenter: Eiji Miyamoto, MD
 Affiliation: Fujita Health University School of Medicine
 Authors: Miyamoto E, Okumoto T, Yoshimura Y
- 1:35 - 1:45 pm Discussion
- 1:45 pm 6
MODIFIED FACIAL TRANSLOCATION APPROACH IN ONE MONTH OLD WITH SKULLBASE TUMOR
 Presenter: Sherard A. Tatum, MD
 Affiliation: State University of New York
 Author: Tatum SA
- 1:49 pm 7
REVIEW OF VARIOUS SURGICAL PROCEDURES IN RADIATED ORBIT RECONSTRUCTION
 Presenter: Junyi Yang, MD
 Affiliation: Huashan Hospital Fudan University
 Authors: Mu X, Yang JY, Guo ZL, Yu ZY, Yang XX
- 1:53 pm 8
MIDLINE FRONTO NASAL DERMIDS - A REVIEW OF 55 CASES AND A PROTOCOL FOR TREATMENT
 Presenter: Mike A. Moses, BM, BCh, BSc, FRCSEd(Plast)
 Affiliation: Great Ormond Street Hospital
 Authors: Moses MA, Green BC, Cugno S, Jeelani NO, Bulsrode NW, Dunaway DJ
- 1:57 pm 9
LANGERHANS CELL HISTIOCYTOSIS IN THE PEDIATRIC POPULATION: HOW SHOULD WE TREAT ISOLATED CRANIOFACIAL LESIONS?
 Presenter: Francesco Gargano, MD, PhD
 Affiliation: The Warren Alpert Medical School of Brown University Providence RI
 Authors: Gargano F, Klinge PM, Welch J, Sullivan SR, Taylor HO



- 2:01 pm **I0**
COMPLICATIONS IN MEDIAL FACIOTOMY
Presenter: Sarah L. Versnel, MD, PhD
Affiliation: Erasmus Medical Center
Authors: Versnel SL, Mathijssen IM
- 2:05 pm **II**
OPTIMIZING THE TIMING AND TECHNIQUE OF TREACHER COLLINS ORBITAL MALAR RECONSTRUCTION
Presenter: Justine C. Lee, MD, PhD
Affiliation: University of California Los Angeles
Authors: Fan KL, Federico C, Kawamoto HK, Bradley JP
- 2:09 pm **I2**
TESSIER 0-14 CLEFTS FROM BIRTH TO MATURITY: A REVIEW
Presenter: Walter J. Flapper, FRACS(Plast), MASurg(Craniofacial)
Affiliation: Australian Craniofacial Unit
Authors: Flapper WJ, Pidgeon T, David DJ, Anderson PJ
- 2:13 pm **I3**
RARE CRANIOFACIAL CLEFTS AND AGENESES
Presenter: S. Anthony Wolfe, MD, FACS, FAAP
Affiliation: Miami Children's Hospital
Author: Wolfe SA
- 2:17 pm **I4**
SECONDARY CORRECTION OF HTO AVOIDING PROBLEMS DURING SKELETAL MOBILIZATION
Presenter: Juan M. Chavanne, MD
Affiliation: WCF Bs As Craniofacial Center
Authors: Chavanne JM, Schauvinhold C, Steinberg D, Pringles D
- 2:21 - 2:30 pm Discussion
- 2:30 pm **I5**
TOWARDS PREDICTABLE AESTHETIC CHANGE IN HYPERTELORISM SURGERY - A RADIOLOGIC STUDY IN 18 PATIENTS AND 30 CONTROLS
Presenter: Jonathan A. Britto, MB, MD, FRCS(Plast)
Affiliation: Great Ormond Street Hospital for Children
Authors: Syme Grant J, Karunakaran T, Abela C, Dunaway DJ, Evans RD, Britto JA
- 2:37 pm **I6**
OUR THERAPEUTIC EXPERIENCE OF ORBITAL HYPERTELORISM: A STUDY OF 23 CHINESE CASES
Presenter: Min Wei, MD
Affiliation: Shanghai 9th Peoples Hospital/Shanghai Jiao Tong University School of Medicine
Authors: Wei M, Yuan J, Yu ZY, Xu L
- 2:44 pm **I7**
PROTOCOL MANAGEMENT AND LONG TERM OUTCOME OF TESSIER CLEFTS 3, 4, 5, 6 & 7
Presenter: David J. David, AC, MD, FRCS, FRACS
Affiliation: Womens and Childrens Hospital
Authors: David DJ, Flapper WJ
- 2:51 - 3:00 pm Discussion
- 3:00 - 3:30 pm Coffee Break



3:30 - 5:00 pm

General Session 2

Non-Syndromic Synostosis Part I

Moderators: *Kant Lin, MD (USA) & Juan C. Rodriguez, MD (Argentina)*

3:30 pm

18

SHUNT-INDUCED CRANIOSYNOSTOSIS: DIAGNOSIS AND SURGICAL MANAGEMENT

Presenter: Matthieu Vinchon, MD, PhD

Affiliation: Lille University Hospital

Authors: Vinchon M, Guerreschi P, Wolber A, Baroncini M, Pellerin P

3:37 pm

19

LONG TERM OUTCOMES OF EARLY CORRECTION OF SAGITTAL SYNOSTOSIS VIA ENDOSCOPICALLY ASSISTED CALVARIAL SYNOSTECTOMY AND POST SURGICAL HELMETING

Presenter: Davinder J. Singh, MD

Affiliation: Barrow Childrens Cleft and Craniofacial Center/Phoenix Childrens Hospital

Authors: Singh DJ, Beals SP, Joganic EF, Manwaring K, Bristol R

3:44 pm

20

RAISED INTRACRANIAL PRESSURE IN NON SYNDROMIC SAGITTAL CRANIOSYNOSTOSIS FOLLOWING CORRECTIVE SURGERY

Presenter: Greg Thomas, PhD, FRCS (Plast)

Affiliation: Oxford Craniofacial Unit

Authors: Thomas G, Wall SA, Jayamohan J, Johnson D, Magdum S, Richards PG

3:51 pm

21

AGE-BASED APPROACH TO SAGITTAL SYNOSTOSIS: APPLICATION OF EXTENDED STRIP CRANIECTOMY (ESC) AND TOTAL CRANIAL VAULT RESHAPING (TCVR) TECHNIQUES

Presenter: Christopher Forrest, MD, MSc, FRCS, FACS

Affiliation: The Hospital for Sick Children

Author: Forrest CR

3:58 pm

22

CAN COGNITIVE FUNCTIONING BE PREDICTED BY PRE-OPERATIVE (CT-SCAN) ANOMALIES IN PATIENTS WITH TRIGONOCEPHALY?

Presenter: Jacques van der Meulen, MD, PhD

Affiliation: Erasmus Medical Center

Authors: van der Muelen J, van der Vlugt JJ, Diamantopoulou S, Horstman EG, Coebergh RR, Hovius SE, Verhulst FC, Stolwijk A, Lequin M

4:05 - 4:15 pm

Discussion

4:15 pm

23

EFFECTS OF AGE AT THE TIME OF SURGERY ON LONG-TERM NEUROPSYCHOLOGICAL OUTCOMES IN SAGITTAL SYNOSTOSIS

Presenter: Anup Patel, MD, MBA

Affiliation: Yale University School of Medicine

Authors: Patel A, Hashim P, Yang J, Bridgett D, Losee J, Duncan C, Jane J, Persing JA

4:19 pm

24

ANAESTHESIA IN THE PRONE POSITION CAN SAFELY BE USED TO FACILITATE THE COMPLEX SURGICAL CORRECTION OF CRANIOSYNOSTOSIS. A SERIES OF 300 CONSECUTIVE PATIENTS IN 15 YEARS IN OXFORD, UK

Presenter: Russell G. Evans, MBBS, FRCA

Affiliation: The Oxford University Hospitals NHS Trust

Authors: Evans RG, Das S, Johnson D, Wall S



4:23 pm **25**
MATERNAL-FETAL COMPLICATIONS IN CRANIOSYNOSTOSIS
 Presenter: Jordan Swanson, MD
 Affiliation: Seattle Childrens Hospital
 Authors: Oppenheimer AJ, Swanson J, Al-Mufarrej F, Pet M, Saltzman B, Gruss J, Hopper R, Cunningham M, Birgfeld CB

4:27 pm **26**
RAISED INTRACRANIAL PRESSURE IN UNTREATED NON-SYNDROMIC SAGITTAL CRANIOSYNOSTOSIS: AN EIGHTEEN YEAR EXPERIENCE IN OXFORD
 Presenter: Steven A. Wall, MB BCh, FRCS, FRCPC, FCS(SA)plast
 Affiliation: Oxford Craniofacial Unit
 Authors: Wall SA, Thomas GP, Byren J, Jayamohan J, Johnson D, Magdum S, McAuley DJ, Richards PG

4:31 pm **27**
MINIMALLY INVASIVE SPRING-ASSISTED CORRECTION OF SAGITTAL SUTURE SYNOSTOSIS
 Presenter: MarieLise C. Van Veelen, MD
 Affiliation: Erasmus University Medical Center Rotterdam
 Authors: Van Veelen MC, Touw C, Mathijssen IM

4:35 pm **28**
THE ASSOCIATION OF CHIARI MALFORMATION TYPE I AND CLOSURE OF SAGITTAL SUTURE
 Presenter: Federico Di Rocco, MD, PhD
 Affiliation: Craniofacial Unit
 Authors: Di Rocco F, Chivoret N, Puget S, Zerah M, Sainte-Rose C, Arnaud E

4:39 pm **29**
COMPARISON OF SPRING CRANIOPLASTY AND PI PROCEDURE FOR SAGGITAL SYNOSTOSIS
 Presenter: Matthew Hansen, MBBS, FRACS
 Affiliation: Princess Margaret Hospital for Children
 Authors: Hansen M, Hewitt T

4:43 - 5:00 pm Discussion

5:30 - 9:00 pm **Welcome Reception** - Bridger Restaurant - Couloir
There will be a 15 minute gondola ride up to the restaurant

Thursday, 12 September

7:00 am - 5:00 pm Registration

7:00 - 8:00 am Continental Breakfast

8:00 - 8:45 am **Ortiz Monasterio Memorial Lecture**
 Moderator: Scott Bartlett, MD (USA)
Craniofacial Surgery: A Petit Histoire
 Speaker: Joseph McCarthy, MD

9:00 - 10:00 am **Concurrent Session 1A** - Walk Festival Hall
Basic Science Research
 Moderators: *Steven Buchman, MD, FACS (USA) & Cassio Raposo do Amaral, MD, PhD (Brazil)*

9:00 am **30**
ORGAN LEVEL TISSUE ENGINEERING USING NATIVE DECELLULARIZED MICROCIRCULATORY CONSTRUCTS AS VASCULARIZED BIOSCAFFOLDS
 Presenter: Lars H. Evers, MD
 Affiliation: Stanford University
 Authors: Evers LH, Findlay MW, Liu W, Simons D, Sorkin M, Rajadas J, Longaker MT, Gurtner GC

WITHDRAWN



- 9:04 am **31**
SUSTAINED, LOW-DOSE RHBMP-2 DELIVERY VIA PLGA MICROSPHERES PROVIDES FOR EQUIVALENT OSTEOGENESIS AND IMPROVED SIDE-EFFECT PROFILE
Presenter: Jason Wink, BA
Affiliation: Childrens Hospital of Philadelphia
Authors: Wink JD, Gerety PA, Sherif R, McGrath JL, Clarke NA, Rajapske C, Nah HD, Taylor JA
- 9:08 am **32**
BIOPRINTING TISSUE ENGINEERED INTRAMEMBRANOUS BONE CONSTRUCTS
Presenter: Darren M. Smith, MD
Affiliation: University of Pittsburgh Medical Center
Authors: Smith DM, Shakir S, Naran S, Campbell P, Losee JE, Cooper GM
- 9:12 am **33**
IN VIVO DIRECTED DIFFERENTIATION OF HUMAN EMBRYONIC-LIKE PLURIPOTENT CELLS INTO BONE
Presenter: Derrick Wan, MD
Affiliation: Stanford University
Authors: Chung MT, Paik KJ, Levi B, Montoro DT, Lo DD, Sun N, Wu JC, Longaker MT, Wan DC
- 9:16 am **34**
PREDICTIVE GENOMICS: VALIDATION OF A ROLE FOR MIRNA IN THE UNDERDEVELOPMENT OF THE MANDIBLE AND OTHER CRANIOFACIAL STRUCTURES
Presenter: Christopher Runyan, MD, PhD
Affiliation: Cincinnati Childrens Hospital Medical Center
Authors: Uribe-Rivera A, Aronow BJ, Billmire DA, Reyna-Rodriguez PX, Gordon CB
- 9:20 - 9:30 am Discussion
- 9:30 am **35**
SHOULD TRANS-GENDER FACIAL SKELETAL DONATION BE CONSIDERED FOR CRANIOMAXILLOFACIAL TRANSPLANTATION? A CADAVER STUDY
Presenter: Gabriel F. Santiago, MD
Affiliation: Johns Hopkins University and School of Medicine
Authors: Alrakan MA, Swanson EW, Susarla S, Santiago GF, Obrien D, Grant GT, Liacouras P, Armand M, Murphy R, Brandacher G, Andrew Lee WP, Gordon CR
- 9:34 am **36**
TRANSLATIONAL CADAVERIC STUDY OF COMPOSITE EYE AND PERIORBITAL ALLOTRANSPLANTATION FLAP FROM RAT MODEL
Presenter: Fatih Zor, MD
Affiliation: Gulhane Military Medical Academy
Authors: Bozkurt M, Zor F, Uygur S, Kulahci Y, Ozturk C, Djohan R, Siemionow M, Papay F
- 9:38 am **37**
FGFR2C(C342Y/+) MOUSE MODEL OF CROUZON SYNDROME TO STUDY CRANIOSYNOSTOSIS
Presenter: Xianxian Yang, MD, PhD
Affiliation: Shanghai Ninth Peoples Hospital/Shanghai Jiao Tong University School of Medicine
Authors: Yang X, Hatfield JT, Hinze SJ, Anderson PJ, Mu XZ, Powell BC
- 9:42 am **38**
FURTHER DEFINING THE ROLE OF RANK/RANKL/OPG AXIS IN CRANIAL SUTURE BIOLOGY
Presenter: Maureen R. Beederman, BA
Affiliation: University of Chicago
Authors: Beederman MR, Lee JC, Rajan M, Rogers MR, Kim S, He TC, Reid RR



- 9:46 am **39**
ANALYSIS OF THE AGING OF THE NASAL BONE AND NASAL PYRAMID STRUCTURES IN A CAUCASIAN POPULATION
Presenter: Can Ozturk, MD
Affiliation: Cleveland Clinic
Authors: Ozturk CN, Bozkurt M, Uygur HS, Sullivan TB, Djohan R, Papay F
- 9:50 - 10:00 am Discussion
- 9:00 - 10:00 am **Concurrent Session 1B** - Four Seasons Ballroom A
Craniofacial Orthodontics & Orthognathic Surgery
Moderators: *Patricia Glick, DMD (USA) & Pedro Santiago, DMD (USA)*
- 9:00 - 9:30 am **PANEL: 3D Computer Assisted Surgical Planning: Opportunities & Challenges. Part One**
Panelists: Patricia Glick, DMD (USA)
The Evolution in Craniofacial Surgical Planning: From the Stone Age to the Sophisticated 3D Digital Era
Barry Grayson, DDS (USA) & Pradip Sheyte, DDS (USA)
Use of Digital Design. A Paradigm Shift in Surgical Planning for Craniofacial Surgery. Preoperative Planning, Diagnosis of the Virtual Patient: Integrating Clinical Data with Static 3D Images.
Pedro Santiago, DMD (USA) & Jeff Marcus, MD (USA)
The Role of the Orthodontist and Surgeon in Computer Aided Surgical Planning
- 9:30 am **40**
EFFECTS OF NASAL ALVEOLAR MOLDING (NAM) ON INFANT WEIGHT GAIN AND TIMING OF PRIMARY BILATERAL CLEFT LIP REPAIR
Presenter: Michael R. Pharaon, MD
Affiliation: The University of North Carolina at Chapel Hill
Authors: Clayton J, Krochmal DJ, van Aalst JA
- 9:34 am **41**
EARLY CORRECTION OF THE MANDIBLE WITH UNI-VECTOR DISTRACTION OSTEOGENESIS IN HEMIFACIAL MICROSOMIA PATIENTS: ORTHODONTIC TECHNIQUE FOR PREVENTION OF MALOCCLUSION AND OPTIMIZING SURGICAL OUTCOME
Presenter: Patricia Glick, DMD
Affiliation: Barrow Childrens Cleft and Craniofacial Center Phoenix Childrens Hospital
Authors: Glick P, Singh DJ
- 9:38 am **42**
FACIAL BIPARTITION; THE ORTHODONTIC BURDEN OF CARE
Presenter: Daljit Gill, MSc, FDS, RCS(Orth)
Affiliation: Great Ormond Street Hospital for Children
Authors: Gill D, Dunaway DJ, Britto JA, Evans RD
- 9:42 am **43**
PREOPERATIVE SIMULATION AND PREDICTABILITY FOR THE FACIAL ASYMMETRY OF HEMIFACIAL MICROSOMIA PRUZANSKY GRADE I
Presenter: Suguru Kondo, PhD, DDS
Affiliation: Cleft Lip and Palate Center Fujita Health University School of Medicine
Authors: Kondo S, Okumoto T, Imamura M, Yoshimura Y
- 9:46 am **44**
VIRTUAL SURGICAL PLANNING FOR ORTHOGNATHIC SURGERY
Presenter: Alvaro A. Figueroa, DDS, MS
Affiliation: Rush University Medical Center
Authors: Figueroa AA, Polley JW
- 9:53 - 10:00 am Discussion



9:00 - 10:00 am

Concurrent Session 1C Four Seasons Ballroom B
Panel: Craniofacial Microsomia: Ethics, Embryos, Management and Outcomes
Moderator: *Michael Cunningham, MD, PhD (USA)*

Ethical Issues in the Management of Craniofacial Microsomia

Keynote Speaker: *Cassandra Aspinall, ACSW (USA)*

Panelists: *Paul Trainor, PhD (USA)*

Development of the Branchial Arches, Their Derivatives, and the Correlations to Craniofacial Microsomia

Carrie Heike, MD, MS (USA)

Efficient Interdisciplinary Management of Children with Craniofacial Microsomia

Nichola Rumsey, PhD (United Kingdom)

Appearance Related Outcomes in Children with Craniofacial Microsomia

10:00 - 10:30 am

Coffee Break

10:30 am - 12:00 pm

Concurrent Session 2A - Walk Festival Hall
Basic Science Research
Moderators: *James Bradley, MD (USA) & John van Aalst, MD, MA (USA)*

10:30 am

45

POSTOPERATIVE SERUM ALBUMIN LEVEL AND CLINICAL IMPLICATIONS IN PEDIATRIC CRANIOFACIAL SURGERY. A REVIEW OF 50 CASES IN OXFORD, UK

Presenter: *Sumit Das, MBBS, BSc, FRCA*

Affiliation: *Oxford Craniofacial Unit*

Authors: *Das S, Evans RG, Allison E, Johnson D, Wall S*

10:34 am

46

INTELLECTUAL OUTCOME IN 29 ADULT PATIENTS BORN WITH CRANIOSYNOSTOSIS

Presenter: *Robert Tovetjarn, MD*

Affiliation: *Institute of Clinical Sciences*

Authors: *Tovetjarn R, Maltese G, Andersson C, Kolby L, Tarnow P*

10:38 am

47

LOCAL FOREIGN-BODY REACTION TO COMMERCIAL BIODEGRADABLE IMPLANTS: AN IN VIVO ANIMAL STUDY

Presenter: *Amy Xue, MD*

Affiliation: *Baylor College of Medicine*

Authors: *Xue A, Koshy JC, Wolfswinkel EM, Weathers WM, Brown RH, Kaufman Y, Hollier LH*

10:42 am

48

HEALING OF CRITICAL CALVARIAL DEFECTS BY CIRCULATING MESENCHYMAL STEM CELLS

Presenter: *Michael Hu, MD, MPH, MS*

Affiliation: *Stanford University*

Authors: *Hyun JS, Wu JC, Lo DD, Li SW, Chung MT, Huang KJ, Hu M, Longaker MT, Lorenz HP*

10:46 am

49

3D CONSTRUCTS OF HUMAN SKELETAL MUSCLE-DERIVED CELLS AS A MODEL OF BONE FORMATION: OSTEOGENIC DIFFERENTIATION IN RESPONSE TO BMP₂

Presenter: *Tiffany Y. Lee, BA*

Affiliation: *University of Pittsburgh School of Medicine and Childrens Hospital of Pittsburgh*

Authors: *Lee TY, Meszaros LB, Tobita K, Kumar AR*

10:50 - 11:00 am

Discussion



11:00 am

50

APPLICATION OF LASER CAPTURE MICRODISSECTION TO CRANIOFACIAL BIOLOGY: CHARACTERIZATION OF ANATOMICALLY RELEVANT GENE EXPRESSION IN THE CRANIAL SUTURES

Presenter: Alex Rottgers, MD

Affiliation: University of Pittsburgh School of Medicine

Authors: Rottgers A, Gallo P, MacIsaac Z, Cray JJ, Smith DM, Kathju S, Mooney M, Losee JE, Cooper G

11:04 am

51

THE ABSOLUTE QUANTIFICATION OF OSTEOGENIC GENES IN RAT CALVARIAL CRITICAL SIZED DEFECTS

Presenter: Brendan J. Alleyne, BS

Affiliation: Case Western Reserve University School of Medicine

Authors: Alleyne BJ, Gliniak C, Varghai D, Askeroglu U, Tobin K, Zwiebel S, Sweeney WM, Cooper GM, Gosain AK

11:08 am

52

DELETION OF AXIN GENE IN MOUSE ADIPOSE-DERIVED STEM CELLS (MASCs) PROMOTES OSTEOGENESIS

Presenter: Kshemendra Senarath, MD

Affiliation: Stanford University

Authors: Senarath K, Chung M, Hasegawa M, Quarto N, Satoh K, Longaker MT

11:12 am

53

REPAIR OF A COMPLICATED CALVARIAL DEFECT: RECONSTRUCTION OF AN INFECTED WOUND WITH RHBMP-2

Presenter: Joseph E. Losee, MD

Affiliation: University of Pittsburgh

Authors: MacIsaac ZM, Shakir S, Naran S, Cray J, Nayar HS, Smith DM, Kinsella CR, Mooney MP, Cooper GM, Losee JE

11:16 - 11:30 am

Discussion

11:30 am

54

TRANSPLANTATION OF A TISSUE ENGINEERED, VASCULARIZED, MUCOSALIZED TRACHEAL HOMOGRAFT IN NEW ZEALAND WHITE RABBITS. COMPARISON OF SUBCUTANEOUS FLAP AND RECTUS MUSCLE FLAPS

Presenter: Christopher Gordon, MD

Affiliation: Cincinnati Childrens Hospital Medical Center

Authors: Gordon PX, Uribe-Rivera A, DeAlarcon A, Propst EJ, Lam DJ, Billmire DA, Aronow BJ, Reyna-Rodriguez XP, Rutter MJ, Gordon CB

11:34 am

55

LACUNOCANALICULAR FLOW AND DISTRACTION OSTEOGENESIS: THEORY AND THERAPEUTICS

Presenter: Edward H. Davidson, MA (Cantab), MBBS

Affiliation: University of Pittsburgh

Authors: Davidson EH, Butala P, Sultan SM, Knobel D, Tutela JP, Canizares O, Wagner IJ, Warren SM

11:38 am

56

PUTATIVE GENES DOWNSTREAM OF FGFR2 ASSOCIATED WITH CORONAL SUTURE SYNOSTOSIS IN A CROUZON SYNDROME (C342Y) MOUSE MODEL

Presenter: Samintharaj Kumar, MBBS, BDS, MRCS, MFDSRCS, MFDRCSI

Affiliation: University College London

Authors: Kumar S, Peskett E, Britto JA, Pauws E



- 11:42 am 57
ACCELERATION OF REGENERATE CONSOLIDATION UTILIZING PHARMACOTHERAPEUTICS DURING MANDIBULAR DISTRACTION OSTEOGENESIS
Presenter: Steven Buchman, MD
Affiliation: University of Michigan
Authors: Buchman S, Donneys A, Deshapande SS, Tchanque-Fossuo CN
- 11:46 am - 12:00 pm Discussion
- 10:30 am - 12:00 pm **Concurrent Session 2B** - Four Seasons Ballroom A
Craniofacial Orthodontics & Orthognathic Surgery
Moderators: *Patricia Glick, DMD (USA) & Pedro Santiago, DMD (USA)*
- 10:30 - 11:00 am **PANEL: 3D Computer Assisted Surgical Planning: Opportunities & Challenges. Part Two**
Panelists: Stephen Baker, MD, DDS (USA)
Virtual Surgical Planning Tools and Splint Construction for Craniofacial Surgery
Alvaro Figueroa, DDS (USA) & John Polley, MD (USA)
Clinical Case: Two Jaw Surgical Correction
Stephen Schendel, MD, DDS (USA)
Clinical Case: Vascularized Free Fibula Flap Design for Mandibular Reconstruction
- 11:00 am 58
STABILITY OF SINGLE STAGE MAJOR MAXILLARY ADVANCEMENTS IN UNILATERAL CLEFT PATIENTS
Presenter: Guy Watts, MBBS, FRACS
Affiliation: Hospital for Sick Children Toronto
Authors: Watts GD, Phillips J, Forrest C, Bigdoli-Moghaddam M, Antonarakis G
- 11:04 am 59
SHORT- AND LONG-TERM CHANGES OF CONDYLAR POSITION AFTER BILATERAL SAGITTAL SPLIT RAMUS OSTEOTOMY FOR MANDIBULAR ADVANCEMENT EVALUATED BY CONE-BEAM COMPUTED TOMOGRAPHY
Presenter: Shuo Chen, MD
Affiliation: School and Hospital of Stomatology Peking University
Authors: Chen S, Lei J, Li ZL, Liang C, Wang XX, Wang X, Fu KY, Farzad P, Yi B
- 11:08 am 60
3D FACIAL SIMULATION IN ORTHOGNATHIC SURGERY: EFFICACY AND ACCURACY
Presenter: Stephen A. Schendel, DDS, MD
Affiliation: Stanford University
Author: Schendel SA
- 11:12 am 61
MAXILLARY ADVANCEMENT AND MANDIBULAR RETRACTION USING NON-OSTEOTOMY TECHNIQUE WITH BOLLARD PLATES
Presenter: Eric Stelnicki, MD
Affiliation: Nova Southeastern University
Authors: Stelnicki E, Marchetto J, Almashat R
- 11:16 am 62
MAXILLOMANDIBULAR ADVANCEMENT SURGERY FOR OBSTRUCTIVE SLEEP APNEA SYNDROME AND LONG-TERM SURGICAL STABILITY IN CHINESE PATIENTS
Presenter: Yang Li, MD
Affiliation: Peking University School and Hospital of Stomatology
Author: Li Y
- 11:20 - 11:30 am Discussion



- 11:30 am **63**
ORTHOGNATHIC POSITIONING SYSTEM (OPS): LINKING VIRTUAL SURGICAL PLANNING (VSP) TO REALITY IN ORTHOGNATHIC SURGERY
Presenter: John W. Polley, MD
Affiliation: Rush University Medical Center
Author: Polley JW
- 11:34 am **64**
VIRTUAL PLANNING SOFTWARE FOR ORTHOGNATHIC SURGERY
Presenter: Earl A. Gage, MD
Affiliation: Mercy Childrens Hospital
Authors: Marsh JL, Gage EA
- 11:38 am **65**
MINIMAL INCISION LEFORT III OSTEOTOMY WITH RIGID EXTERNAL DISTRACTION: ANALYSIS AND CLINICAL OUTCOMES OF 12 SYNDROMIC PATIENTS
Presenter: Scott Rapp, MD
Affiliation: Cincinnati Childrens Hospital Medical Center
Authors: Rapp S, Uribe-Rivera A, Pan BS, Billmire DA, Gordon CB
- 11:42 am **66**
SURGERY-FIRST ORTHOGNATHIC SURGERY (WITHOUT PRESURGICAL ORTHODONTICS) -- EFFICIENCY AND STABILITY
Presenter: YuRay Chen, MD
Affiliation: Chang Gung Memorial Hospital
Authors: Chen Y, Liao YF, Ko EW, Wang YC, Huang CS
- 11:49 am - 12:00 pm Discussion
- 10:30 am - 12:00 pm **Concurrent Session 2C - Four Seasons Ballroom B**
Genetic and Non-Surgical Aspects of Craniofacial Conditions
Moderators: *Carrie Heike MD, MS (USA) & Michael Cunningham, MD, PhD (USA)*
- 10:30 am **67**
MUTATIONS IN TCF12 ARE A FREQUENT CAUSE OF CORONAL CRANIOSYNOSTOSIS
Presenter: Jacqueline A. Goos, MD
Affiliation: Erasmus Medical Center
Authors: Goos JA, Sharma VP, Fenwick A, Wilkie AO, Mathijssen IM
- 10:34 am **68**
CROUZON SYNDROME AND BENT BONE DYSPLASIA ASSOCIATED TO MUTATIONS AT THE SAME TYR-381 RESIDUE IN FGFR2 GENE
Presenter: Corinne Collet, MD
Affiliation: Craniofacial Unit
Authors: Collet C, Alessandri JL, Arnaud E, Cormier-Daire V, Di Rocco F
- 10:38 am **69**
NON-SCAPHOCEPHALIC SAGITTAL SYNOSTOSIS AND RAISED INTRACRANIAL PRESSURE ARE PART OF THE CLINICAL SPECTRUM OF FRANK-TER HAAR SYNDROME
Presenter: Charlotte L. Bendon, BA, BMBCh
Affiliation: Oxford University Hospitals
Authors: Bendon CL, Fenwick AL, Hurst JA, Nuernberg G, Nuernberg P, Wall SA, Wilkie AO, Johnson D
- 10:42 am **70**
HEMIFACIAL MICROSOMIA ASSOCIATED WITH A DUPLICATION OF 4P16
Presenter: Katrina M. Dipple, MD, PhD
Affiliation: University of California Los Angeles
Authors: Dipple KM, Lee Barber J, Bradley JP, Rivera-Quintero F



- 10:46 am **71**
CRANIOFACIAL ABNORMALITIES IN ASSOCIATION WITH THE 22Q11.2 DELETION SYNDROME: BEYOND CLEFTING
 Presenter: Donna McDonald-McGinn, MS, CGC
 Affiliation: Children’s Hospital of Philadelphia/University of Pennsylvania School of Medicine
 Authors: McDonald-McGinn DM, Jackson O, Passick C, Low D, Bailey A, Jethava N, Schultz P, Forbes BJ, Binnenbaum G, Whitaker L, Zackai EH, Bartlett SP
- 10:50 - 11:00 am Discussion
- 11:00 am **72**
PREVALENCE OF OBSTRUCTIVE SLEEP APNEA SYNDROME IN SYNDROMIC CHILDREN WITH CRANIOFACIAL ANOMALIES
 Presenter: Linlin Gao, MD
 Affiliation: Childrens Hospital of Pennsylvania
 Authors: Gao L, Paliga JT, Goldstein J, Cielo C, Marcus CL, Taylor JA
- 11:04 am **73**
TIMING OF AIRWAY OBSTRUCTION IN INFANTS WITH ROBIN SEQUENCE
 Presenter: Kelly N. Evans, MD
 Affiliation: Seattle Childrens Hospital and University of WA
 Authors: Evans KN, Saltzman BS, Sie KC
- 11:08 am **74**
PROGRESSIVE HEMIFACIAL ATROPHY: A NEW FINDING
 Presenter: Michael Friel, MD
 Affiliation: Indiana University
 Authors: Friel M, Havlik RJ
- 11:12 am **75**
FACIAL INFILTRATING LIPOMATOSIS IS CAUSED BY PIK3CA ACTIVATING MUTATIONS
 Presenter: Reid A. Maclellan, MD
 Affiliation: Boston Childrens Hospital/Harvard Medical School
 Authors: Maclellan RA, Kurek KC, Luks VL, Mulliken JB, Warman ML, Greene AK
- 11:16 am **76**
PLAGIOCEPHALY, TORTICOLLIS, LAMBDOID CRANIOSYNOSTOSIS: A SPECTRUM OF DISEASE. A REVIEW OF 9,683 PATIENTS WITH POSTERIOR PLAGIOCEPHALY
 Presenter: Robert Wood, MD
 Affiliation: Gillette Childrens Hospital
 Authors: Wood R, Chibbaro G, McGrory M
- 11:20 - 11:30 am Discussion
- 11:30 am **77**
CLEFT AND CRANIOFACIAL CLINIC FORMATS IN THE UNITED STATES: NATIONAL AND INSTITUTIONAL SURVEY
 Presenter: Haruko Okada, MD
 Affiliation: Case Western Reserve University
 Authors: Okada H, Alleyne B, Leuchtag RM, Lakin GE
- 11:34 am **78**
TELEMEDICINE IN CLEFT CARE: THE EXPERIENCE AT SHRINERS HOSPITAL FOR CHILDREN, LOS ANGELES
 Presenter: Melinda Costa, MD
 Affiliation: Shriners Hospital Los Angeles
 Authors: Costa M, Gillenwater JG, Taghva GT, Green TG, Magee WM



- 11:38 am **79**
POSITIONAL PLAGIOCEPHALY: EXPERIENCE WITH A PASSIVE ORTHOTIC MATTRESS
Presenter: Sasha Burn
Affiliation: Alder Hey Hospital for Children
Authors: Bruce S, Quirk D, Sinha A, Burn SC, Richardson D, Vaiude P, Duncan C
- 11:42 am **80**
DEVELOPMENT OF A PATIENT REPORTED OUTCOME MEASURE IN ADULT CLEFT LIP AND PALATE PATIENTS
Presenter: Sophie Ricketts, MD
Affiliation: Sunnybrook Health Sciences Center
Authors: Ricketts S, Fialkov J
- 11:46 am **81**
**“JUST BECAUSE SOMEONE’S A DOCTOR DOESN’T MEAN I JUST TRUST THEM”:
ENHANCING PATIENTS’ DECISION MAKING IN ELECTIVE RECONSTRUCTIVE SURGERY**
Presenter: Daniela Hearst, BSc, MPhil
Affiliation: Great Ormond Street Hospital for Children Foundation Trust
Authors: Hearst D, Mayers S, Liddle M, Thomas S, Shanly A, McKerracher G, Horton J, Knapp M, Kearney A
- 12:00 - 1:30 pm Lunch
- 12:00 - 1:30 pm ISCSF Biennial Business Meeting - Walk Festival Hall
- 1:30 - 3:30 pm **Concurrent Session 3A** - Walk Festival Hall
Aesthetic Surgery of the Craniofacial Skeleton & Craniofacial Trauma
Moderators: *Michael Yaremchuk, MD (USA) & Joseph Gruss, MD, FRCSC, FAAP (USA)*
- 1:30 pm **82**
MENTAL NERVE PATHWAY BEFORE MENTAL FORAMEN FOR GENIOPLASTY
Presenter: LunJou Lo, MD
Affiliation: Chang Gung Memorial Hospital
Authors: Kim HY, Kim SG, Huang CS, Chen YR, Lo LJ
- 1:34 pm **83**
AESTHETIC REFINEMENTS IN THE TREATMENT OF GRAVES’ OPHTHALMOPATHY
Presenter: Gaby Doumit, MD, MSc
Affiliation: Cleveland Clinic
Authors: Doumit G, Yaremchuk M
- 1:38 pm **84**
COSMETIC AND FUNCTIONAL TREATMENT OF GRAVES EXOPHTHALMOS AND RELATED EYELID MALPOSITION USING SELECTIVE OSTEOTOMIES
Presenter: Henry M. Spinelli, MD
Affiliation: Weill Medical College of Cornell University
Authors: Spinelli HM, Sackeyfio R
- 1:42 pm **85**
CORRECTION OF PROMINENT MALAR WITH FIBROUS DYSPLASIA BY L-SHAPE OSTEOTOMY
Presenter: Feng Niu, MD
Affiliation: Peking Union Medical College/Chinese Academy of Medical Sciences
Authors: Niu F, Gui L, Liu JF, Song T
- 1:46 pm **86**
IMPLEMENTING FAT GRAFTING IN MANAGING DIFFICULT CRANIOFACIAL PATIENTS
Presenter: Amir S. Elbarbary, MD
Affiliation: AinShams University
Author: Elbarbary AS



2:00 - 2:10 pm	Discussion
2:10 pm	<p>87 INFLUENCE OF THE MAXILLARY SINUS EXPOSURE AND PARTIAL MASSETER RESECTION IN AESTHETIC FACIAL CONTOUR SURGERY Presenter: Jie Yuan, MD, PhD Affiliation: Shanghai 9th Peoples Hospital, Shanghai Jiao Tong University School of Medicine Authors: Yuan J, Yu ZY, Xu L, Zhang Y, Wei M</p>
2:14 pm	<p>88 CUSTOMIZED 3D MODELING OF AUGMENTATION MENTOPLASTY Presenter: Robert Guryanov, MD Affiliation: Sechenov First Moscow State Medical University Authors: Guryanov R, Guryanov A</p>
2:18 pm	<p>89 ANALYSIS OF ADULT FACIAL SKELETAL CHANGE WITH AGE Presenter: Richard A. Levine, MD, DDS Affiliation: University of Texas Health Science Center at San Antonio Authors: Levine RA, Wang PT, Garcia A</p>
2:22 pm	<p>90 FAT GRAFTING IN THE CRANIOFACIAL PATIENT Presenter: Jorge E. Cabrera, MD Affiliation: Hospital General Dr Manuel Gea Gonzalez Authors: Cabrera JE, Aguilera A</p>
2:26 pm	<p>91 STRUCTURAL FAT GRAFTING OF THE CRANIO ORBITO FACIAL AREA VOLUMETRIC & MORPHO-AESTHETIC IMPLICATIONS Presenter: Riccardo Tieghi, MD Affiliation: S Anna Hospital and University of Ferrara Italy Authors: Tieghi R, Clauser LC</p>
2:30 - 2:40 pm	Discussion
2:40 pm	<p>92 A PRELIMINARY REPORT ON THE USE OF ANTIBIOTIC-IMPREGNATED METHYL METHACRYLATE IN SALVAGE CRANIOPLASTY Presenter: Anthony Wilson, MD Affiliation: University of Pennsylvania Authors: Hsu VM, Grady MS, Taylor JA</p>
2:44 pm	<p>93 TITANIUM CRANIOPLASTY IN PAEDIATRIC CRANIOFACIAL SURGERY - THE BIRMINGHAM EXPERIENCE Presenter: Anuradha Venugopal, MD Affiliation: Birmingham Childrens Hospital UK Authors: Dover S, Lloyd M, Ahmad F, Bhatia S, Evans M, White N, Solanki G, Rodrigues D, Nishikawa H</p>
2:48 pm	<p>94 ORBITAL FLOOR FRACTURES: AN OUTCOMES STUDY OF RECONSTRUCTION IN 364 CASES Presenter: John B. Turner, MD Affiliation: University of Kentucky Authors: Turner JB, Kirby EJ, Vasconez HC</p>



- 2:52 pm **95**
RECONSTRUCTION OF MAJOR CALVARIAL DEFECTS USING AUTOGENOUS MATERIAL
Presenter: Ramesh K. Sharma, MD
Affiliation: Postgraduate Institute of Medical Education and Research
Author: Sharma RK
- 2:56 pm **96**
POROUS HYDROXYAPATITE CUSTOM MADE CRANIOPLASTY IN 18 CHILDREN
Presenter: Gregoire Pech-Gourg, MD
Affiliation: Childrens Hospital La Timone
Authors: Pech-Gourg G, Di Rocco F, Arnaud E, Puget S, Zerah M, Scavarda D, Sainte-Rose C
- 3:00 - 3:10 pm Discussion
- 3:10 pm **97**
PATIENT-SPECIFIC ORBITAL IMPLANTS: DEVELOPMENT AND IMPLEMENTATION OF TECHNOLOGY FOR MORE ACCURATE ORBITAL RECONSTRUCTION
Presenter: Oleh Antonyshyn, MD, FRCSC
Affiliation: Sunnybrook Health Sciences Centre University of Toronto
Authors: Antonyshyn O, Mainprize J, Edwards G
- 3:14 pm **98**
A COST-BENEFIT ANALYSIS COMPARING CUSTOM COMPUTER-GENERATED CRANIOFACIAL IMPLANTS VS. TRADITIONAL BONY CRANIOPLASTY FOR CONGENITAL AND TRAUMATIC CRANIAL DEFORMITIES
Presenter: Omar A. Fouda Neel, MD, FRCSC
Affiliation: McGill University
Authors: Fouda Neel OA, Gilardino M, Karunanayake, Izadpanah A, Al-Ajmi S
- 3:18 pm **99**
TOTAL FACE, DOUBLE JAW, AND TONGUE TRANSPLANTATION: AN EVOLUTIONARY CONCEPT
Presenter: Amir H. Dorafshar, MBChB, FAAP
Affiliation: University of Maryland
Authors: Dorafshar AH, Bojovic B, Christy MR, Iliff NT, Borsuk DE, Rodriguez ED
- 3:22 - 3:30 pm Discussion
- 3:30 - 3:30 pm **Concurrent Session 3B - Four Seasons Ballroom A**
Craniofacial Microsomia/Related Conditions
Moderators: *Fernando Molina, MD (Mexico) & Sherard A. Tatum, MD (USA)*
- 1:30 pm **100**
FACIAL ANIMATION IN MÖBIUS SYNDROME
Presenter: Juan C. Rodriguez Sr., MD
Affiliation: Garrahan Pediatric Hospital
Author: Rodriguez JC
- 1:37 pm **101**
INVESTIGATING THE ETIOPATHOGENESIS OF OBSTRUCTIVE SLEEP APNEA IN PEDIATRIC PATIENTS WITH UNILATERAL CRANIOFACIAL MICROSOMIA
Presenter: Parit A. Patel, MD
Affiliation: New York University
Authors: Patel PA, Szpalski C, Fisher M, Wetterau M, Bernstein J, McCarthy JG, Warren SM
- 1:44 pm **102**
LONGITUDINAL GROWTH ANALYSIS OF MANDIBULAR ASYMMETRY IN UNOPERATED PATIENTS WITH UNILATERAL CRANIOFACIAL MICROSOMIA (UCFM)
Presenter: Pradip R. Shetye, DDS, MDS
Affiliation: New York University Langone Medical Center
Authors: Shetye PR, Grayson BG, Mccarthy JG



- 1:51 pm **103**
FINAL SKELETAL CORRECTION IN CRANIOFACIAL MICROSOMIA- STRATEGIES FOR MANAGING THE COSTOCHONDRAL-GRAFTED PATIENT
Presenter: Andrew A. Heggie, MD, DDS
Affiliation: Royal Childrens Hospital of Melbourne
Authors: Heggie AA, Shand JM
- 1:58 - 2:10 pm Discussion
- 2:10 pm **104**
248 YEARS OF EXPERIENCE CLASSIFYING THE MANDIBULAR DEFORMITY IN HEMIFACIAL MICROSOMIA: A CRITICAL ANALYSIS OF THE PRUZANSKY/KABAN CLASSIFICATION SYSTEM UTILIZING THREE-DIMENSIONAL COMPUTED TOMOGRAPHY
Presenter: Scott Bartlett, MD
Affiliation: Childrens Hospital of Philadelphia
Authors: Wink JD, Goldstein J, Paliga JT, Taylor JA, Bartlett SP
- 2:17 pm **105**
TOTAL AURICULAR CONSTRUCTION NEW APPROACH WITH SIX TYPES OF FRAMEWORK TEMPLATES BASED ON NORMAL CURVE VARIATIONS
Presenter: Akira Yamada, MD, PhD
Affiliation: Osaka Medical College
Authors: Yamada A, Ueda K, Otani K, Nuri T, Ohtsuki Y, Gosain A, Harada T, Salyer K
- 2:24 pm **106**
CUSTOMIZED POROUS POLYETHYLENE FRAMEWORK FOR EAR RECONSTRUCTION
Presenter: ZungChung Chen, MD
Affiliation: Chang Gng Memorial Hospital
Authors: Chen ZC, Chen YA
- 2:31 - 2:40 pm Discussion
- 2:40 pm **107**
PLANNING SURGICAL RECONSTRUCTION IN TREACHER - COLLINS SYNDROME USING GEOMETRIC MORPHOMETRICS
Presenter: Dariush Nikkhah, BM, MSc, MRCS
Affiliation: Great Ormond Street Hospital
Authors: Nikkhah D, Ponniah A, Ruff C, Dunaway D
- 2:44 pm **108**
A CRITICAL ANALYSIS OF THE RELATIONSHIP BETWEEN THE MAXILLA AND THE MANDIBLE IN HEMIFACIAL MICROSOMIA
Presenter: Jason Wink, BA
Affiliation: Childrens Hospital of Philadelphia
Authors: Wink JD, Goldstein J, Paliga JT, Taylor JA, Bartlett SP
- 2:48 pm **109**
A NOVEL APPROACH TO OSSEOINTEGRATED AURICULAR PROSTESIS RETENTION
Presenter: Drew Schnitt, MD
Affiliation: Joe DiMaggio Childrens Hospital
Authors: Schnitt D, Trainer D
- 2:52 pm **110**
"I KEEP IT IN THE DRAWER" - MINIMISING THE CONFLICT BETWEEN EAR RECONSTRUCTION AND BAHAS IN CHILDREN WITH MICROTIA - A SALUTORY EXPERIENCE FROM THE UK
Presenter: David Gault, MB ChB, FRCS
Affiliation: The London Centre for Ear Reconstruction
Authors: Gault DT, Cheang PP



2:56 pm

**III
MANAGEMENT OF SURVIVING PATIENTS WITH OTOCEPHALY: TREATMENT
ALGORITHM AND CORRECTION OF FACIAL DEFORMITIES**

Presenter: Kristin Yee

Affiliation: University of California Los Angeles

Authors: Yee K, Chawla R, Chan FC, Kawamoto HK, Bradley JP

3:00 - 3:10 pm

Discussion

3:10 pm

**II2
FREE PARASCAPULAR FLAPS FOR PRUZANSKI III HYPOPLASTIC MANDIBLES: OUR
EXPERIENCE WITH 7 PATIENTS**

Presenter: Christopher Gordon, MD

Affiliation: Cincinnati Childrens Hospital Medical Center

Authors: Rapp S, Uribe-Rivera A, Pan BS, Billmire DA, Gordon CB

3:14 pm

**II3
PSYCHOSOCIAL, FEEDING, SPEECH & DROOLING OUTCOMES IN CHILDREN WITH
BECKWITH WIEDEMANN SYNDROME FOLLOWING TONGUE REDUCTION SURGERY**

Presenter: Caroleen Shipster, MSc

Affiliation: Great Ormond Street Hospital for Children NHS Foundation Trust

Authors: Shipster C, Morgan A, Dunaway D

3:18 pm

**II4
EARLY FAT GRAFTING FOR AUGMENTATION OF ORBITOZYGOMATIC REGION IN
TREACHER COLLINS SYNDROME: A PRELIMINARY REPORT**

Presenter: Petros Konofaos, MD, PhD

Affiliation: University of Tennessee Health Science Center

Authors: Konofaos P, Diner PD, Arnaud EA

3:22 - 3:30 pm

Discussion

1:30 - 3:30 pm

**Concurrent Session 3C - Four Seasons Ballroom B
Craniofacial Imaging**

Moderators: *Alvaro Figueroa, DDS, MS & Mirko S. Gilardino, MD (Canada)*

1:30 pm

**II5
"BLACK BONE" MRI: CHANGING THE FACE OF CRANIOFACIAL IMAGING**

Presenter: Karen A. Eley, MBChB, MRCS, PGCTLCP, FHEA, MSc, DPhil

Affiliation: University of Oxford/Oxford Craniofacial Unit

Authors: Eley KA, Sheerin F, Watt-Smith SR, Golding SJ

1:37 pm

**II6
RADIATION REDUCTION PROTOCOL EFFECTIVELY REDUCES RADIATION EXPOSURE
DURING CT EVALUATION OF PEDIATRIC CRANIOFACIAL ANOMALIES**

Presenter: Christopher S. Zarella, MD

Affiliation: Oregon Health and Science University

Authors: Zarella CS, Didier R, Bardo D, Selden NR, Kuang AA

1:44 pm

**II7
AN OPEN-ACCESS, INTERNET-BASED, PLASTIC SURGERY SIMULATOR**

Presenter: Roberto Flores, MD

Affiliation: Indiana University

Authors: Flores R, Olikier A, Costa M, Cutting C



- 1:51 pm **118**
MEIN₃D. 12000 FACIAL SURFACE SCANS TO CREATE A MODEL OF NORMAL FACIAL VARIATION TO AID SURGICAL PLANNING
Presenter: Allan Ponniah, MBBS, MRCS, MSc
Affiliation: Great Ormond Street Hospital
Authors: Ponniah A, Ruff C, Angullia F, Dunaway D
- 1:58 - 2:10 pm Discussion
- 2:10 pm **119**
INCIDENCE OF CNS ABNORMALITIES ON BRAIN MRI IN INFANTS WITH PIERRE ROBIN SEQUENCE
Presenter: Arlene Rozzelle, MD
Affiliation: Childrens Hospital of Michigan Detroit Medical Center
Authors: Khan S, Pappas KP, Rozzelle AR
- 2:14 pm **120**
TRIGONOCEPHALY AND FRONTAL LOBE PERFUSION
Presenter: Giovanna Paternoster
Affiliation: Craniofacial Unit
Authors: Di Rocco F, Grevent D, Chivoret N, Paternoster G, Boddaert N, Brunelle F, Arnaud E
- 2:18 pm **121**
OUTCOME ANALYSIS AFTER HELMET THERAPY USING 3D PHOTOGRAMMETRY IN PATIENTS WITH DEFORMATIONAL PLAGIOCEPHALY: THE ROLE OF ROOT MEAN SQUARE
Presenter: Mahsa Bidgoli Moghaddam, MBBS
Affiliation: The Hospital for Sick Children University of Toronto
Authors: Bidgoli Moghaddam M, Brown TM, Clausen A, DaSilva T, Forrest CR
- 2:22 pm **122**
BEST FACE FORWARD: VIRTUAL MODELING AND CUSTOM DEVICE FABRICATION TO OPTIMIZE CRANIOFACIAL VASCULARIZED COMPOSITE ALLOTRANSPLANTATION (VCA)
Presenter: Jordan Jacobs, MD
Affiliation: New York Medical College
Authors: Jacobs J, Dec W, Levine JP, McCarthy JG, Weimer K, Moore K, Ceradini DJ
- 2:26 pm **123**
OPTICAL COHERENCE TOMOGRAPHY: A POTENTIAL OPTIC NERVE HEAD IMAGING MODALITY FOR CRANIOSYNOSTOSIS PATIENTS WITH SUSPECTED RAISED INTRACRANIAL PRESSURE
Presenter: William R. Katowitz, MD
Affiliation: The Childrens Hospital of Philadelphia
Authors: Katowitz WR, Cohen Y, Taylor J, Forbes BJ
- 2:30 - 2:40 pm Discussion
- 2:40 pm **124**
REAL TIME CINE-MRI IN FETUSES WITH HEAD AND NECK PATHOLOGY
Presenter: Oksana A. Jackson, MD
Affiliation: The Childrens Hospital of Philadelphia
Authors: Jackson OA, LaRossa D, Victoria T, Pollock A, Feygin T
- 2:44 pm **125**
COMPUTATIONAL FLUID DYNAMICS FOR AIR FLOW MODELING IN CRANIOFACIAL PATHOANATOMY
Presenter: Mark D. Fisher, MD
Affiliation: Duke University
Authors: Fisher MD, Patel PA, Szpalski C, Kassam S, McCarthy JG, Warren S, Kimbell J, Marcus J



2:48 pm

126

WITHDRAWN

**A CLASSIFICATION SYSTEM TO GUIDE ORBITO-ZYGOMATIC RECONSTRUCTION
INTREACHER - COLLINS SYNDROME**

Presenter: Ben Green, BSc

Affiliation: Great Ormond Street Hospital

Authors: Green B, Nikkhah D, Ponniah A, Dunaway D

2:52 pm

127

**CLINOCEPHALY AND CLOSURE OF LATERAL CALVARIAL SUTURES ON
VOLUME-RENDERED CT RECONSTRUCTIONS**

Presenter: Corbett Wilkinson, MD

Affiliation: University of Colorado School of Medicine Childrens Hospital Colorado

Authors: Wilkinson C, French BM, Serrano CA, Stence NV

2:56 pm

128

CRANIOFACIAL FIBROUS DYSPLASIA

Presenter: Thomas Satterwhite

Affiliation: Miami Childrens Hospital

Authors: Satterwhite T, Wolfe SA

3:00 - 3:10 pm

Discussion

3:10 pm

129

**INCIDENTAL FINDINGS AND THEIR CLINICAL SIGNIFICANCE IN PREOPERATIVE CT FOR
SINGLE SUTURE NON SYNDROMIC CRANIOSYNOSTOSIS**

Presenter: Keshav Magge, MD

Affiliation: Childrens National Medical Center

Authors: Magge K, Magge SN, Myseros JS, Keating RF, Rogers GF, Oh AK

3:14 pm

130

THE USE OF BRAINLAB NAVIGATION IN LEFORT III OSTEOTOMY

Presenter: Jeyhan Wood, MD

Affiliation: Wake Forest Baptist Health

Authors: Wood J, Thompson JT, David LR, Argenta LC

3:18 pm

131

**PREDICTING FACIAL SOFT TISSUE CHANGES FOLLOWING FACIAL OSTEOTOMIES
USING GEOMETRIC MORPHOMETRICS AND THIN PLATE SPLINE WARPING**

Presenter: Freida Angullia, MD

Affiliation: Great Ormond Street Hospital for Children

Authors: Angullia F, Ruff CF, Dunaway DJ

3:22 - 3:30 pm

Discussion

4:00 - 5:30 pm

Poster Session - Hotel Terra Ballroom/Exhibit Hall

1P

**SUCCESSFUL RECONSTRUCTION OF COMPLEX PEDIATRIC NASAL LESIONS:
IMPROVING OUTCOMES USING A DERMAL REGENERATIVE TEMPLATE IN PEDIATRIC
NASAL DEFECTS**

Presenter: Oluwaseun A. Adetayo, MD

Affiliation: University of Pittsburgh

Authors: Adetayo OA, Grunwaldt LJ, MacIsaac ZM, Losee JE, Kumar AR

2P

**FEMTOSECOND PLASMA MEDIATED LASER ABLATION HAS ADVANTAGES OVER
MECHANICAL OSTEOTOMY OF CRANIAL BONE**

Presenter: Kevin J. Paik, BS

Affiliation: Stanford University

Authors: Chung MT, Paik KJ, Mackanos MA, Hyun JS, Montoro DT, Grova M, Liu C, Wang J,

Chung AS, Palanker D, Connolly AJ, Longaker MT, Contag CH, Wan DC



3P

USE OF NOVEL BMP-2 MINICIRCLE PLASMID-RELEASING SCAFFOLDS FOR BONE ENGINEERING

Presenter: Michael T. Chung, BS

Affiliation: Stanford University

Authors: Chung MT, Paik KJ, Morrison SD, Zimmermann AS, Lo DD, Hyun JS, McArdle A, Walmsley G, Senarath-Yapa K, Montoro DT, Hu MS, Chen HH, Chung AS, Ransom RC, Longaker MT, Wan DC

4P

BONE MORPHOGENETIC PROTEIN RECEPTOR IB AS A MARKER FOR ENRICHMENT OF OSTEOGENIC PRECURSORS

Presenter: Kevin J. Paik, BS

Affiliation: Stanford University

Authors: Chung MT, Paik KJ, Lo DD, Hyun JS, Montoro DT, Walmsley G, Senarath-Yapa K, Zimmermann AS, Sorkin M, Rennert R, Hu MS, Chen HH, Chung AS, Longaker MT, Wan DC

5P

ISOLATION OF ADIPOSE-DERIVED STROMAL CELLS USING LASER-ASSISTED LIPOSUCTION SPECIMENS AND THEIR THERAPEUTIC POTENTIAL IN REGENERATIVE MEDICINE

Presenter: Michael T. Chung, BS

Affiliation: Stanford University

Authors: Chung MT, Paik KJ, Liu CJ, Morrison SD, Lo DD, Hyun JS, Montoro DT, McArdle A, Walmsley G, Sorkin M, Rennert R, Chen HH, Senarath-Yapa K, Chung AS, Vistnes D, Longaker MT, Wan DC

6P

ENHANCED ADIPOSE-DERIVED STROMAL CELL OSTEOGENESIS THROUGH SURFACE MARKER ENRICHMENT AND BMP MODULATION

Presenter: Kevin J. Paik, BS

Affiliation: Stanford University

Authors: Paik KJ, Morrison SD, Zimmermann AS, Lo DD, Hyun JS, McArdle A, Walmsley G, Senarath-Yapa K, Montoro DT, Sorkin M, Rennert R, Hu MS, Chen HH, Chung AS, Longaker MT, Wan DC

7P

ADVANCES IN 3D IMAGE FUSION FOR CRANIOFACIAL TRANSPLANTATION

Presenter: Darren M. Smith, MD

Affiliation: University of Pittsburgh Medical Center

Authors: Smith DM, Gorantla VS, Losee JE

8P

USABILITY OF 3D SIMULATION FOR CRANIO-MAXILLOFACIAL SURGERY

Presenter: Takayuki Okumoto, MD

Affiliation: Fujita Health University School of Medicine

Authors: Okumoto T, Yoshimura Y, Kondo S, Imamura M

9P

THREE-DIMENSIONAL MORPHOMETRIC ANALYSIS OF THE HYPOPLASTIC MANDIBLE

Presenter: Kevin J. Paik, BS

Affiliation: Stanford University

Authors: Paik KJ, Levi B, Hyun JS, Lo DD, Montoro DT, Lisiecki J, Chung AS, Bradley JP, Buchman SR, Longaker MT, Wan DC



10P

COMPARATIVE ANALYSIS OF THE MUSCULAR STRUCTURES OF THE NECK IN PATIENTS WITH PIERRE ROBIN SEQUENCE AND PATIENTS WITH NORMAL MANDIBULAR GROWTH

Presenter: Andres Bello, MD

Affiliation: Hospital General Dr Manuel Gea Gonzalez

Authors: Bello A, Lopez R, Molina F

11P

A THREE DIMENSIONAL ANALYSIS OF FACIAL-LABIAL FOLD AFTER MALAR AUGMENTATION OR REDUCTION

Presenter: Zheyuan Yu, MD

Affiliation: Shanghai Ninth People Hospital

Authors: Yu Z, Cao DJ, Chai G, Zhu YJ, Mu XZ

12P

BRAIN VOLUME AND SHAPE CHANGES IN MIDLINE NON-SYNDROMIC CRANIOSYNOSTOSIS PATIENTS

Presenter: John A. van Aalst, MD, MA

Affiliation: The University of North Carolina at Chapel Hill

Authors: Halevi A, Krochmal DJ, Paniagua B, Styner M, van Aalst JA

13P

DIFFERENCES IN MEASUREMENTS OF THE SKULL BASE AND FACIAL SKELETON IN PATIENTS WITH PLAGIOCEPHALY VERSUS PATIENTS WITH PLAGIOCEPHALY AND FACIAL SCOLIOSIS

Presenter: Cuauhtemoc Lorenzana, MD

Affiliation: Hospital General Dr Manuel Gea Gonzalez

Authors: Lorenzana C, Arrieta P, Francis FA, Molina F

14P

PATTERN OF DENTAL EXTRACTIONS IN PATIENTS WITH SYNDROMIC CRANIOSYNOSTOSIS UNDERGOING CRANIOFACIAL SURGERY

Presenter: Susana Dominguez-Gonzalez, PhD

Affiliation: Alder Hey Childrens NHS Foundation Trust

Authors: Dominguez-Gonzalez S, Richardson DR, Laraway DC

15P

SUTURAL DISTRACTION OSTEOGENESIS (SDO) OF INTERNAL DISTRACTOR TREATED UNILATERAL CORONAL SYNOSTOSIS AND IMPLANTED MEDPOR FOR BLOCK SKULL GROWTH AND REPAIR OF SKULL DEFECT: A PRIMARY CLINICAL REPORT

Presenter: Shen Weimin, MD

Affiliation: Nanjing Childrens Hospital affiliated with Nanjing Medical

Authors: Weimin S, Cui J, Jianbing CH, Jijun Z, Ji Y, Haini CH

16P

VIRTUAL DESIGN OF BILATERALLY DESTROYED TRAUMA CASE S BASED ON THREE DIMENSIONAL NORMAL SKULL DATABASE

Presenter: Xiaojing Liu, MD

Affiliation: Peking University School and Hospital of Stomatology

Authors: Liu X, Guo CB, Zhang Y, Chen L, Wang J

17P

COST ANALYSIS OF AUTOGENOUS VS. PREFABRICATED PATIENT-SPECIFIC ALLOPLASTIC CRANIOPLASTY

Presenter: Amir Mrad, MD, MBA, FRCSC

Affiliation: University of Toronto/Harvard University

Authors: Mrad A, Antonyshyn DR



18P

ZYGOMATIC FRACTURE: EXTENDED CLASSIFICATION FOR ADDITIONAL OPTION FOR THE IMMEDIATE MANAGEMENT

Presenter: Kazuhiro Otani, MD, PhD

Affiliation: Osaka Medical College Hospital

Authors: Otani K, Kyutoku S, Okada M, Yamada A, Ueda K

19P

USE OF CT DERIVED PROSTHETICS FOR INTRA-OPERATIVE GUIDANCE OF TUMOR RESECTION

Presenter: Christian J. Vercler, MD

Affiliation: University of Michigan

Authors: Vercler CJ, Kline S, Buchman SR

20P

BRANCHIAL ARCH ANOMALIES: RECURRENCE, MALIGNANT DEGENERATION AND OPERATIVE COMPLICATIONS

Presenter: Dave Stoddard, MD

Affiliation: Mayo Clinic

Authors: Al-Mufarrej F, Stoddard D, Bite U

21P

LATERAL TRANSZYGOMATIC APPROACH SPHENOID MENINGIOMA

Presenter: Henry M. Spinelli, MD

Affiliation: Weill Medical College of Cornell University

Authors: Spinelli HM, Hanasono MM, Langevin CJ

22P

MANAGEMENT OF MANDIBULAR HIGH-FLOW ARTERIOVENOUS MALFORMATIONS

Presenter: Jocelyn M. Shand, MBBS, MDSc, FRACDS(OMS), FDSRCS(Eng)

Affiliation: The Royal Childrens Hospital of Melbourne

Authors: Shand JM, Heggie AA

23P

EVALUATING THE EFFICACY OF AIRWAY EXPANSION USING TRANS-CRANIAL VERSUS SUB-CRANIAL FACIAL OSTEOTOMIES: A COHORT COMPARISON STUDY BETWEEN MONOBLOC FRONTOFACIAL ADVANCEMENT AND LE FORT III FACIAL ADVANCEMENT

Presenter: Oluwaseun A. Adetayo, MD

Affiliation: University of Pittsburgh

Authors: Adetayo OA, Rottgers SA, Miele LF, MacIsaac ZM, Davidson EH, Kumar AK

24P

A COMPARATIVE ANALYSIS OF COMPLICATIONS IN 55 MIDFACE DISTRACTION PROCEDURES IN PATIENTS WITH SYNDROMIC CRANIOSYNOSTOSIS

Presenter: Jesse A. Goldstein, MD

Affiliation: University of Pennsylvania School of Medicine & Childrens Hospital of Philadelphia

Authors: Goldstein JA, Taylor JA, Bartlett SP

25P

OUTCOMES ANALYSIS OF MANDIBULAR DISTRACTION OSTEOGENESIS FOR THE TREATMENT OF PIERRE ROBIN SEQUENCE ASSOCIATED WITH ADVANCED AIRWAY OBSTRUCTION

Presenter: Melinda Costa, MD

Affiliation: Indiana University School of Medicine

Authors: Murage KP, Tholpady SS, Friel M, Costa M, Havlik RJ, Flores RL



26P

EXPANSION OF THE POSTERIOR CRANIAL VAULT: CHARACTERIZATION OF THE ROLE OF DISTRACTION OSTEOGENESIS

Presenter: Raymond J. Harshbarger III, MD

Affiliation: Dell Childrens Medical Center

Authors: Harshbarger RJ, Combs PD, Kelley PK, George TM

27P

CAN WE PREDICT THE NEED FOR ICU ADMISSION AFTER CRANIOSYNOSTOSIS SURGERY?

Presenter: Susan Goobie, MD, FRCPC

Affiliation: Boston Childrens Hospital

Authors: Goobie S, Zurakowski D, Busa K, Proctor M, Meara J, Rogers G

28P

A 35-YEAR EXPERIENCE WITH SYNDROMIC CLEFT PALATE REPAIR: OPERATIVE OUTCOMES AND LONG-TERM SPEECH RESULTS

Presenter: Marten N. Basta, BS

Affiliation: University of Pennsylvania Perelman School of Medicine/Childrens Hospital of Philadelphia

Authors: Basta MN, Silvestre J, Solot C, Cohen M, Kirschner RE, Low DW, LaRossa D, Jackson OA

29P

THIRTY YEARS OF PRENATAL CLEFT DIAGNOSIS: WHAT HAVE WE LEARNED?

Presenter: Jordan P. Steinberg, MD, PhD

Affiliation: Lurie Childrens Hospital of Chicago

Authors: Steinberg JP, Gosain AK

30P

POST-OPERATIVE PYREXIA IN CHILDREN FOLLOWING TRANSCRANIAL SURGERY

Presenter: Fateh Ahmad, FRCS(Plast)

Affiliation: Birmingham Children's Hospital

Authors: Ahmad F, Qureshi H, Elserius A, Evans M, White N, Rodrigues D, Nishikawa H, Dover S, Solanki G

WITHDRAWN

31P

VISUAL FUNCTION AND OPERATIVE MORBIDITY IN THE CORRECTION OF HYPERTELORISM: REPORT OF 31 CASES

Presenter: Jonathan A. Britto, MB, MD, FRCS(Plast)

Affiliation: Great Ormond Street Hospital for Children

Authors: Britto JA, Dunaway DJ, Abela C, Schweibert K, Hon K

32P

USING PRINCIPAL COMPONENT ANALYSIS (PCA) TO QUANTIFY THE DEFORMITY IN CRANIOFRONTONASAL DYSPLASIA, SIMULATE CORRECTION AND AID SURGICAL PLANNING

Presenter: Nicola Bystrzonowski, MBBS, BSc, MRCS (Eng), MSc, DIC

Affiliation: Great Ormond Street Hospital

Authors: Bystrzonowski N, Ponniah A, Dunaway D

33P

LONG-TERM OUTCOMES FOLLOWING EXTENDED SAGITTAL SYNOSTECTOMIES

Presenter: Gary F. Rogers, MD

Affiliation: Royal Childrens Hospital Melbourne

Authors: Seruya M, DeFreitas T, Myseros JS, Yaun AL, Rogers GF, Keating RF



34P
MORPHOMETRIC ANALYSIS OF CORTICAL GRAY MATTER IN INFANTS WITH ISOLATED METOPIC CRANIOSYNOSTOSIS

Presenter: Arshad Muzaffar, MD
Affiliation: University of Missouri Columbia
Authors: Le MH, Aldridge KJ, Christ SE, Luo Y, Aviles DL, Cole KK, Muzaffar AR

35P
TRIGONOCEPHALY - TREATMENT USING DYNAMIC CRANIOTOMY AND EXPANDER SPRINGS

Presenter: Anderson Souza, MD
Affiliation: Hospital Beneficencia Portuguesa de Sao Paulo
Authors: Souza A, Silva AS, Figueiredo EG, Cardim VL

36P
A THREE DIMENSIONAL ANALYSIS OF NASAL AESTHETICS FOLLOWING LE FORT I ADVANCEMENT IN PATIENTS WITH CLEFT LIP AND PALATE

Presenter: Edward H. Davidson, MA (Cantab), MBBS
Affiliation: University of Pittsburgh
Authors: Davidson EH, Miele LF, Kumar AR

37P
SIMULTANEOUS ALVEOLAR FISTULA REPAIR AND BONE GRAFT DURING LEFORT I ADVANCEMENT IN CLEFT PATIENTS: IMPROVING OUTCOMES AND DECREASING MORBIDITY USING ALLOGRAFT ONLY BONE GRAFT

Presenter: Alex Rottgers, MD
Affiliation: University of Pittsburgh School of Medicine
Authors: Rottgers A, Miele L, MacIsaac Z, Kumar AR

38P
COMBINED CONTRACTION AND DISTRACTION OF THE FACIAL SKELETON IN STAGED TREATMENT OF AN ATYPICAL FACIAL CLEFT USING DISTRACTION DEVICES

Presenter: Robert D. Wallace, MD
Affiliation: University of Tennessee Memphis
Authors: Wallace RD, Alvarez S, Konofaos P

39P
LONG TERM FOLLOW UP OF MAXILAR RECONSTRUCTION IN CLEFT PATIENTS WITH RHBMP-2. COST BENEFITS ANALYSIS

Presenter: Nivaldo Alonso, MD, PhD
Affiliation: University of Sao Paulo
Authors: Alonso N, Lima Jr JE, Freitas RS

WITHDRAWN

40P
RARE FACIAL CLEFTS: 6 CASES EACH A DIFFERENT CHALLENGE FOR TREATMENT

Presenter: Diego J. Caycedo, MD, MSc
Affiliation: Universidad del Valle
Authors: Caycedo DJ, Cabal M

41P
8 TRANSVERSE CLEFT LIP REPAIR CASES - FOLLOW UP

Presenter: Penpak Krergmatukorn, MD
Affiliation: Queen Sirikit National Institute of Child Health
Author: Krergmatukorn P



42P

AGE-RELATED EFFECT OF MONOBLOC FRONTO-FACIAL DISTRACTION ON ORBITAL VOLUME, MORPHOLOGY, AND CLINICAL OUTCOME IN 29 CROUZON-PFEIFFER CASES: A CONTROLLED STUDY

Presenter: Jonathan A. Britto, MB, MD, FRCS(Plast)

Affiliation: Great Ormond Street Hospital for Children

Authors: Khonsari R, Karunakaran T, Ashraff S, Nysjo R, Nystrom I, Dunaway DJ, Evans RD, Britto JA

43P

CORRECTION OF CORONAL CRANIOSYNOSTOSIS FROM UCS TO KLEEBLATTSCHADEL

Presenter: Mckay Mckinnon, MD

Affiliation: Lurie Childrens Hospital

Author: Mckinnon M

44P

ORBITAL MORPHOLOGY IN APERT AND CROUZON-PFEIFFER SYNDROMES: AN AGE – MATCHED, CONTROLLED STUDY

Presenter: Jonathan A. Britto, MB, MD, FRCS(Plast)

Affiliation: Great Ormond Street Hospital for Children

Authors: Khonsari R, Way B, Matthews W, Nysjo R, Nystrom I, Evans RD, Britto JA

45P

CLOSURE OF THE SPHENO-OCCIPITAL SYNCHONDROSIS IN PATIENTS WITH CROUZON SYNDROME: A LINK TO MIDFACE HYPOPLASIA

Presenter: Jesse A. Goldstein, MD

Affiliation: University of Pennsylvania School of Medicine & Childrens Hospital of Philadelphia

Authors: Goldstein JA, Bartlett SP, Taylor JA

46P

HYDROCEPHALUS AND BASE SKULL ABNORMALITIES IN FGFR 2 CRANIOSYNOSTOSIS

Presenter: Coll Guillaume, MD

Affiliation: Necker Hospital

Authors: Guillaume C, Di Rocco F, Brunelle F, Collet C, Arnaud E

47P

MONOBLOC FRONTO FACIAL ADVANCEMENT WITH DISTRACTION OSTEOGENESIS FOR SINDROMIC CRANIOSYNOSTOSIS: COMPARISON BETWEEN 2 DISTRACTORS

Presenter: Cassio E. Raposo-Amaral Sr., MD, PhD

Affiliation: SOBRAPAR

Authors: Raposo-Amaral CE, Raposo-Amaral CA

48P

MAXILLARY DISTRACTION IN THE PATIENT WITH CLEFT LIP AND PALATE ANOMALIES: LESSONS LEARNED IN 74 CONSECUTIVE CASES

Presenter: Jordi Puente-Espel, MD

Affiliation: International Craniofacial Institute

Authors: Puente-Espel J, Genecov DG, Barcelo CR

49P

TREATMENT AND OUTCOME OF FIBROUS DYSPLASIA INVOLVING THE ORBIT: THE ROLE OF NEW TECHNOLOGIES

Presenter: Jordi Puente-Espel, MD

Affiliation: International Craniofacial Institute

Authors: Puente-Espel J, Barcelo CR, Genecov DG



- 6:30 - 7:30 pm **ASCFS Reception** - Four Seasons Ballroom Prefunction A
- 7:30 - 10:00 pm **ISCFS Council Dinner (by invitation)** - Four Seasons
- 8:00 - 10:00 pm **Seattle Children's Craniofacial Fellowship Alumni Dinner** - Alpenhof Lodge

Friday, 13 September

7:00 am - 12:00 pm Registration

7:00 - 8:00 am Continental Breakfast

7:30 - 9:30 am **Concurrent Session 4A** - Walk Festival Hall

Basic Science Research

Moderators: *Jesse Taylor, MD (USA) & Robert M. Menard, MD, FACS (USA)*

7:30 am

I32

LONG-TERM BIOMECHANICAL PROPERTIES OF BONE MORPHOGENETIC PROTEIN REGENERATED BONE IN FAVORABLE AND UNFAVORABLE CALVARIAL WOUNDS

Presenter: Zoe M. MacIsaac, MD

Affiliation: University of Pittsburgh

Authors: MacIsaac ZM, Henderson SE, Nayar HS, Shakir S, Naran S, Smith DM, Cray J, Mooney MP, Almarza A, Cooper GM, Losee JE

7:34 am

I33

EVALUATION OF CRANIAL TRANSPORT DISTRACTION WITH AND WITHOUT ADIPOSE GRAFTING

Presenter: James Clune, MD

Affiliation: Yale University School of Medicine

Authors: Yuhasz MM, Travieso R, Wong K, Clune J, Zuang ZW, Van Houten J, Steinbacher DM

7:38 am

I34

AN EXPERIMENTAL STUDY OF PARTICULATE BONE GRAFT FOR SECONDARY INLAY CRANIOPLASTY OVER SCARRED DURA

Presenter: Arin K. Greene, MD

Affiliation: Boston Childrens Hospital/Harvard Medical School

Authors: Maclellan RA, Hassanein AH, Mulliken JB, Rogers GF, Greene AK

7:42 am

I35

REPAIR OF A COMPLICATED CALVARIAL DEFECT: RECONSTRUCTION OF A WOUND COMPLICATED BY DURECTOMY AND INFECTION

Presenter: Sanjay Naran, MD

Affiliation: University of Pittsburgh

Authors: MacIsaac ZM, Shakir S, Naran S, Cray J, Smith DM, Kubala A, Kinsella CR, Mooney MP, Cooper GM, Losee JE

7:46 am

I36

IDENTIFICATION AND CHARACTERIZATION OF NEUROCRANIAL SKELETAL PROGENITOR CELLS IDENTIFICATION AND CHARACTERIZATION OF NEUROCRANIAL SKELETAL PROGENITOR CELLS

Presenter: Adrian McArdle, MD

Affiliation: Stanford University

Authors: McArdle A, Chan CK, Hyun JS, Chung MT, Montoro DT, Wan DC, Weissman IL, Longaker MT

7:50 - 8:00 am

Discussion



- 8:00 am **137**
EVIDENCE FROM THE FGFR₂C₃₄₂Y/C₃₄₂Y CROUZON MURINE MODEL ARGUES FOR A PRIMARY FAILURE IN CHONDROGENIC DEVELOPMENT
Presenter: Erwin Pauws, PhD
Affiliation: University College London
Authors: Kumar S, Peskett E, Britto JA, Pauws E
- 8:04 am **138**
OSTEOGENIC PERFORMANCE OF DONOR MATCHED HUMAN ADIPOSE AND BONE MARROW MSCS UNDER DYNAMIC CULTURE
Presenter: Miles Pfaff, MD
Affiliation: Yale University School of Medicine
Authors: Pfaff MJ, Zellner E, Niklason LE, Steinbacher DM
- 8:08 am **139**
NAGER SYNDROME DENTAL PULP STEM CELLS SHOW SUPERIOR OSTEOGENIC POTENTIAL COMPARED TO TREACHER COLLINS SYNDROME
Presenter: Joyce T. Yuan, MD
Affiliation: University of California Los Angeles
Authors: Yuan JT, Bueno D, Tabit CJ, Bradley JP
- 8:12 am **140**
HEDGEHOG GRADIENT MODULATES NASAL PROCESSES GROWTH AND FUSION BY ANTAGONIZING CANONICAL WNT SIGNALING
Presenter: Hiroshi Kurosaka, PhD, DDS
Affiliation: Stowers Institute for Medical Research
Authors: Kurosaka H, Trainor PA
- 8:16 am **141**
TRANSFORMING GROWTH FACTOR BETA τ IMPROVES BONE HEALING AND PROMOTES SUTURE REGENERATION
Presenter: Sameer Shakir, BS
Affiliation: University of Pittsburgh
Authors: Shakir S, MacIsaac ZM, Naran S, Gilbert JR, Shaw MA, Wang D, Kubala AA, Losee JE, Cooper GM
- 8:20 - 8:30 am Discussion
- 8:30 am **142**
CALVARIAL RECONSTRUCTION WITH BONE MARROW CELLS: CONCOMITANT TREATMENT WITH BMP-2
Presenter: Matthew Greives, MD
Affiliation: University of Pittsburgh
Authors: MacIsaac ZM, Shakir S, Naran S, Zammerilla L, Cooper GM, Losee JE
- 8:34 am **143**
TRANS-ORAL POSTERIOR MAXILLARY CRANIOFACIAL SURGERY TO PLACE A SPHENOPALATINE GANGLION (SPG) NEUROSTIMULATOR FOR TREATMENT OF CHRONIC CLUSTER HEADACHE (CCH): PATHWAY CH-1 STUDY SURGICAL EXPERIENCE
Presenter: Frank A. Papay, MD
Affiliation: Cleveland Clinic
Authors: Papay FA, Hillerup S, Wilmont A, Puche M, Pohlenz P, Muller O, Fontaine D, Blessman M, Caparso A



- 8:38 am **I44**
CELL-ASSISTED LIPOTRANSFER WITH BONE MORPHOGENETIC PROTEIN RECEPTOR IA+ ADIPOSE-DERIVED STROMAL CELL SUBPOPULATIONS
Presenter: Kevin J. Paik, BS
Affiliation: Stanford University
Authors: Paik KJ, Chung MT, Chen HH, McArdle A, Hu MS, Lo DD, Hyun JS, Montoro DT, Senarath-Yapa K, Zimmermann AS, Walmsley G, Sorkin M, Rennert R, Chung AS, Longaker MT, Wan DC
- 8:42 am **I45**
REFLECTANCE CONFOCAL MICROSCOPY MAY REDUCE THE NEED FOR SKIN BIOPSY IN COMPOSITE TISSUE ALLOTRANSPLANTATION
Presenter: Huseyin Karagoz, MD
Affiliation: Gulhane Military Medical Academy
Authors: Zor F, Karagoz H, Erdemir AV, Karslioglu Y, Acikel CH, Kapaj R, Guzey S, Gurel MS, Ozturk S
- 8:46 am **I46**
PREOPERATIVE LIP MEASUREMENT IN PATIENTS WITH UNILATERAL COMPLETE CLEFT LIP/PALATE AND ITS COMPARISON WITH NORMS
Presenter: PangYun Chou, MD
Affiliation: Chang Gung Memorial Hospital
Authors: Chou PY, Luo CC, Chen KT, Chen YR, Noordhoff MS, Lo LJ
- 8:50 am **I47**
BIOMECHANICAL ASSESSMENT OF CALVARIAL RECONSTRUCTION: REPAIR OF A CRANIAL DEFECT WITH CRYOPRESERVED AND FRESH AUTOLOGOUS GRAFT
Presenter: Seun Adetayo, MD
Affiliation: University of Pittsburgh
Authors: MacIsaac ZM, Shakir S, Henderson S, Naran S, Cray J, Smith DM, Mooney MP, Almarza A, Cooper GM, Losee JE
- 8:54 - 9:15 am Discussion
- 7:30 - 9:30 am **Concurrent Session 4B** - Four Seasons Ballroom A
Non-Syndromic Craniosynostosis II - A
Moderators: *Jie Yuan, MD, PhD (China) & Christopher Forrest, MD, MSc, FRCSC, FACS (Canada)*
- 7:30 am **I48**
EFFECTS OF HYPOTENSIVE ANESTHESIA ON BLOOD LOSS DURING CRANIOSYNOSTOSIS CORRECTIONS
Presenter: Kevin Cook, MD
Affiliation: The Craniofacial Center
Authors: Fearon JA, Cook TK, Herbert MA
- 7:34 am **I49**
EVALUATING THE SAFETY AND EFFICACY OF TRANEXAMIC ACID ADMINISTRATION IN PEDIATRIC CRANIAL VAULT RECONSTRUCTION
Presenter: John C. Crantford, MD
Affiliation: Wake Forest Baptist Health
Authors: Crantford JC, Claiborne JR, Wood BC, Ririe DG, Thompson JT, David LR
- 7:38 am **I50**
TECHNIQUES TO MINIMISE PATIENT EXPOSURE TO DONATED BLOOD; A 10 YEAR AUDIT OF 450 CONSECUTIVE, NON SYNDROMIC, PAEDIATRIC, CALVARIAL REMODELLING PROCEDURES
Presenter: Carol Millar, MD
Affiliation: Birmingham Childrens Hospital
Authors: Evans MJ, White N, Nishikawa H, Dover MS, Solanki G, Rodrigues D, Carver E, Millar C, Johnson D



- 7:42 am **151**
INTRAOPERATIVE BLOOD TRANSFUSION PATTERNS IN SURGERY FOR NON SYNDROMIC CRANIOSYNOSYOSIS
Presenter: Llewellyn C. Padayachy, MD
Affiliation: University of Cape Town
Authors: Padayachy LC, Fieggen AG, Figaji AA, Micheals J, Lechthape-Gruther R, Peter J
- 7:46 am **152**
EFFECTS OF TYPE OF SURGICAL INTERVENTION ON NEUROPSYCHOLOGICAL OUTCOMES IN SAGITTAL CRANIOSYNOSTOSIS
Presenter: Peter Hashim, MD
Affiliation: Yale University School of Medicine
Authors: Patel A, Yang J, Hashim P, Bridgett D, Losee J, Duncan C, Jane J, Persing JA
- 7:50 - 8:00 am Discussion
- ~~8:00 am **153**
EXAMINING THE UTILITY OF DYNAMIC CRANIOPLASTY FOR THE TREATMENT OF SAGITTAL SYNOSTOSIS: A RETROSPECTIVE COHORT COMPARISON STUDY BETWEEN REVERSE PI CRANIOPLASTY AND EXTENDED STRIP CRANIOPLASTY
Presenter: Christopher Bonfield, MD
Affiliation: University of Pittsburgh Medical Center
Authors: Bonfield C, Rottgers SA, MacIsaac ZM, Pollack IF, Tamber MS, Kumar AR~~
- WITHDRAWN**
- 8:04 am **154**
DIAGNOSIS SPECIFIC HEAD GROWTH CURVE IN CHILDREN UNDERGOING TOTAL CALVARIAL VAULT REMODELING (TCVR) FOR SAGITTAL SUTURE SYNOSTOSIS
Presenter: Peter D. Ray, MD
Affiliation: Childrens of Alabama Hospital
Authors: Ray PD, Slama RE, Huettner F, Dynda DI, Grant JH
- 8:08 am **155**
TOTAL CRANIAL VAULT REMODELING FOR ISOLATED SAGITTAL SYNOSTOSIS: PART I. POSTOPERATIVE CRANIAL SUTURE PATENCY
Presenter: David K. Chong, MD
Affiliation: Royal Childrens Hospital Melbourne
Authors: Seruya M, Tan SY, Wray AC, Penington AJ, Greensmith AL, Holmes AD, Chong DK
- 8:12 am **156**
EVALUATING THE EFFICACY OF DYNAMIC CRANIAL VAULT REMODELING FOR THE TREATMENT OF LATE PRESENTING SCAPHOCEPHALY: A COHORT COMPARISON STUDY BETWEEN PI CRANIOPLASTY AND SUBTOTAL CRANIOPLASTY
Presenter: Anand R. Kumar, MD
Affiliation: University of Pittsburgh School of Medicine
Authors: Rottgers A, MacIsaac ZM, Losee JE, Pollack IF, Tamber M, Kumar AR
- 8:16 am **157**
HOW DOES CEPHALIC INDEX AT THE SKULL VAULT RELATE TO CEPHALIC INDEX AT THE SKULL BASE IN CHILDREN WITH UNOPERATED, ISOLATED SAGITTAL SYNOSTOSIS?
Presenter: David Johnson, MD
Affiliation: Oxford University Hospitals
Authors: Bendon CL, Sheerin FB, Wall SA, Johnson D
- 8:20 - 8:30 am Discussion
- 8:30 am **158**
THE LIVERPOOL PROTOCOL FOR SCAPHOCEPHALY CORRECTION
Presenter: Christian Duncan, MD
Affiliation: Alder Hey Hospital for Children
Authors: Vaiude PN, Burn SC, Sinha A, Richardson D, Duncan C



- 8:34 am **159**
SPHENOID WING FLARE: FINDINGS AND IMPLICATIONS FOR TREATMENT OF SAGITTAL SUTURE SYNOSTOSIS
Presenter: Robert J. Havlik, MD
Affiliation: Indiana University
Authors: Havlik RJ, Friel M, Flores R, Smith J, Ackerman L
- 8:38 am **160**
INITIATION OF MULTI-MODEL THERAPY TO MINIMIZE ALLOGENEIC TRANSFUSION REQUIREMENTS DURING SURGICAL TREATMENT OF CRANIOSYNOSTOSIS AT A TEACHING HOSPITAL: LESSONS LEARNED
Presenter: Jennifer L. Rhodes, MD
Affiliation: VCU School of Medicine
Authors: Rhodes JL, Vega RA, Lyon C, Kierce JF
- 8:42 am **161**
INTERNATIONAL SURVEY OF PROTOCOLS FOR SAGITTAL SYNOSTOSIS IN INFANTS
Presenter: Uzoma Ben Gbulie, MD
Affiliation: Craniofacial Unit
Authors: Bengbulie U, Di Rocco F, Paternoster G, Baugnon T, Meyer P, Arnaud E
- 8:46 am **162**
SPLIT CRANIAL GRAFTING IN CHILDREN LESS THAN 3 YEARS OF AGE: >400 CONSECUTIVE CASES
Presenter: Christian J. Vercler, MD
Affiliation: University of Michigan
Authors: Vercler CJ, Sugg KB, Buchman SR
- 8:50 - 9:00 am Discussion
- 9:00 am **163**
ISOLATED FRONTOSPHEOIDAL SYNOSTOSIS: A RARE CAUSE OF SYNOSTOTIC FRONTAL PLAGIOCEPHALY
Presenter: Tina M. Sauerhammer, MD
Affiliation: Childrens National Medical Center
Authors: Sauerhammer TM, Oh AK, Boyajian M, Magge S, Myseros JS, Keating RF, Rogers GF
- 9:04 am **164**
UNILATERAL VERSUS BILATERAL CORRECTION OF UNICORONAL SYNOSTOSIS; AN ANALYSIS OF LONG-TERM RESULTS
Presenter: Martijn Cornelissen, MD
Affiliation: Erasmus Medical Center
Authors: Cornelissen M, van der Vlugt JJ, Willemsen J, Mathijssen I, van Adrichem L, van der Meulen JJ
- 9:08 am **165**
TREATMENT OF UNILATERAL CORONAL SYNOSTOSIS WITH ENDOSCOPIC STRIP CRANIECTOMY AND CRANIAL ORTHOSIS YIELDS SIGNIFICANTLY BETTER OPHTHALMIC OUTCOMES THAN TREATMENT WITH FRONTO-ORBITAL ADVANCEMENT
Presenter: Linda Dagi, MD
Affiliation: Boston Childrens Hospital
Authors: Dagi L, Rogers GR, Mackinnon S, Meara JG, Proctor MR
- 9:12 am **166**
A SINGLE CENTER'S EXPERIENCE WITH ISOLATED UNICORONAL CRANIOSYNOSTOSIS RECONSTRUCTION: LONG-TERM OUTCOMES OF 182 PATIENTS OVER 35 YEARS
Presenter: Ari M. Wes, BA
Affiliation: University of Pennsylvania School of Medicine & Childrens Hospital of Philadelphia
Authors: Wes AM, Goldstein JA, Whitaker LA, Bartlett SP, Taylor JA



- 9:16 am **167**
MINOR CRANIAL SUTURE CLOSURE: A MORPHOLOGICAL STUDY
Presenter: Wayne Ledinh, MD
Affiliation: Cleveland Clinic Foundation
Authors: Ledinh W, Papay FA, Doumit G
- 9:20 - 9:30 am Discussion
- 7:30 - 9:30 am **Concurrent Session 4C** - Four Seasons Ballroom B
Non-Syndromic Craniosynostosis II - B
Moderators: *Juan Martin Chavanne, MD (Argentina) & John Polley, MD (USA)*
- 7:30 am **168**
**ISOLATED METOPIC CRANIOSYNOSTOSIS IN THE FOURTH DIMENSION:
AN EVALUATION OF COMPLICATIONS, REVISIONS, AND LONG-TERM OUTCOMES IN
142 PATIENTS OVER 30 YEARS**
Presenter: J. Thomas Paliga, MD
Affiliation: University of Pennsylvania School of Medicine & Childrens Hospital of Philadelphia
Authors: Paliga JT, Wes AM, Goldstein JA, Whitaker LA, Bartlett SP, Taylor JA
- 7:34 am **169**
**CLASSIFICATION OF SEVERITY AND OPERATIVE DECISION-MAKING IN METOPIC
SYNOSTOSIS**
Presenter: Alexander C. Allori, MD, MPH
Affiliation: Duke University Hospital
Authors: Allori AC, Pourtaheri N, Meara JG, Rogers GF, Marcus JR
- 7:38 am **170**
**CLINICAL CHARACTERISTICS AND SURGICAL DECISION MAKING FOR INFANTS WITH
METOPIC CRANIOSYNOSTOSIS IN CONJUNCTION WITH OTHER CONGENITAL
ANOMALIES**
Presenter: Craig Birgfeld, MD
Affiliation: University of Washington Seattle Childrens Hospital
Authors: Birgfeld C, Heike CL, Saltzman BS, Hing AV
- 7:42 am **171**
**DIAGNOSIS OF METOPIC CRANIOSYNOSTOSIS FROM THE SYSTEMATIC ANALYSIS OF
CRANIAL MALFORMATION**
Presenter: Gary F. Rogers, MD
Affiliation: University of Sevilla Spain
Authors: Mendoza C, Safdar N, Rogers GF, Oh AK, Sauerhammer T, Keating R, Linguraru MG
- 7:46 am **172**
MANAGEMENT OF SAGITTAL SYNOSTOSIS: STANDARD OF CARE
Presenter: Gaby Doumit, MD, MSc, FRCSC, FACS
Affiliation: Cleveland Clinic
Authors: Doumit G, Moores N, Papay F
- 7:50 - 8:00 am Discussion
- 8:00 am **173**
**NAUTILUS: DYNAMIC CRANIOTOMY; NEW SURGICAL TECHNIQUE AND
PRELIMINARY RESULTS**
Presenter: Vera N. Cardim, MD, PhD
Affiliation: Hospital Beneficencia Portuguesa de Sao Paulo
Authors: Cardim VN, Silva AS, Dornelles RF, Salomons RL, Silva AL, Blom JO, Colange NZ



- 8:04 am **174**
BILATERAL LAMBDROID AND SAGITTAL SYNOSTOSIS (BLSS) – “MERCEDEZ BENZ” SYNDROME
Presenter: Hamilton Matushita, PhD
Affiliation: Sao Paulo University
Authors: Matushita H, Alonso N, Bueno MR, Cardeal DD, Andrade FG
- 8:08 am **175**
TRUE LAMBDROID SYNOSTOSIS: THE LIVERPOOL EXPERIENCE
Presenter: Jitske W. Nolte, MD, DDS
Affiliation: Alder Hey Childrens Hospital
Authors: Nolte JW, Sinha A, Burn S, Duncan C, Richardson D
- 8:12 am **176**
COMBINED METOPIC AND UNILATERAL CORONAL SYNOSTOSES: A PHENOTYPIC CONUNDRUM
Presenter: Kam Patel, MD
Affiliation: Childrens National Medical Center
Authors: Sauerhammer TM, Patel K, Oh AK, Proctor MR, Mulliken JB, Rogers GF
- 8:16 am **177**
PRIMARY CORRECTION OF NASAL ASYMMETRY IN PATIENTS WITH UNILATERAL CORONAL SYNOSTOSIS
Presenter: Arun K. Gosain, MD
Affiliation: Lurie Childrens Hospital of Northwestern University Feinberg School of Medicine
Authors: Gosain AK, Chepla KJ, Alleyne BJ
- 8:20 - 8:30 am Discussion
- 8:30 am **178**
A MODIFIED SURGICAL CORRECTION OF PLAGIOCEPHALY TO ACHIEVE SYMMETRY
Presenter: Hazem Ahmed Mostafa, MD
Affiliation: AinShams University
Authors: Elbarbary AS, Mostafa HA
- 8:34 am **179**
SPRING-ASSISTED CRANIOPLASTY: OUR FIRST EXPERIENCE
WITHDRAWN
Presenter: JanFalco Wilbrand, MD, DMD
Affiliation: University Hospital Giessen
Authors: Wilbrand J, Reinges M, Christophis P, Howaldt HP
- 8:38 am **180**
FAMILIAL NONSYNDROMIC CRANIOSYNOSTOSIS WITH SPECIFIC DEFORMITY OF THE CRANIUM
Presenter: Azusa Shimizu, MD
Affiliation: Juntendo University Shizuoka Hospital
Authors: Shimizu A, Komuro Y, Miyajima M, Arai H
- 8:42 am **181**
COMPARISON OF DIFFERENT SURGICAL STRATEGIES TO APPROACH UNILATERAL CORONAL SYNOSTOSIS
Presenter: Cesar A. Raposo-Amaral, MD
Affiliation: SOBRAPAR
Authors: Raposo-Amaral CA, Chammas D, Raposo-Amaral CE



- 8:46 am **r82**
EVALUATION OF SURGICAL OUTCOMES IN CRANIO-ORBITAL RESHAPING WITH NOVEL QUANTITATIVE CRANIOMETRIC ANALYSIS
Presenter: David Y. Khechoyan, MD
Affiliation: Texas Childrens Hospital
Authors: Khechoyan DY, Saber NR, Birgfeld CB, Gruss J, Saltzman B, Forrest CR, Phillips JH, Hopper RA
- 8:50 - 9:00 am Discussion
- 9:00 am **r83**
PREVENTING TEMPORAL HOLLOWING IN FRONTO-ORBITAL ADVANCEMENT USING POLYLACTIC ACID (PLA) MESH
Presenter: William Y. Hoffman, MD
Affiliation: UCSF
Authors: Hoffman WY, Bertrand AA, Nathan N
- 9:04 am **r84**
PREFABRICATED “PROFILE” CURVE TEMPLATES FOR FRONTORBITAL REMODELING IN CRANIOSYNOSTOSIS
Presenter: Kenneth Salyer, MD
Affiliation: Osaka Medical College
Authors: Yamada A, Ueda K, Kajimoto Y, Salyer K, Nuri T, Gosain A, Harada T
- 9:08 am **r85**
BILAMBDROID AND SAGITTAL SYNOSTOSIS : REPORT OF 31 CASES
Presenter: Rebecca Wyten, MD
Affiliation: APHP Hospital Necker Enfants Malades Paris France
Authors: Chivoret N, Di Rocco F, Wyten R, Selek L, Cuttaree H, Renier D, Duracher C, Arnaud E
- 9:12 am **r86**
COMPUTER-BASED QUANTITATIVE ASSESSMENT OF CRANIAL SHAPE FOR CRANIOSYNOSTOSIS
Presenter: Ben Wood, MD
Affiliation: University of Sevilla Spain
Authors: Mendoza C, Wood B, Safdar N, Myers E, Rogers GF, Oh AK, Sauerhammer T, Keating R, Linguraru MG
- 9:16 am **r87**
NEURODEVELOPMENTAL OUTCOME OF NON-SYNDROMIC CRANIOSYNOSTOSIS
Presenter: Dawid Larysz, MD, PhD
Affiliation: Medical University of Silesia
Authors: Larysz D, Larysz P, Dowgierd K
- 9:20 am **r88**
BENIGN RADIOGRAPHIC CORONAL SYNOSTOSIS AFTER SAGITTAL SYNOSTOSIS REPAIR
Presenter: Anna Kuang, MD
Affiliation: Oregon Health and Science University
Authors: Kuang A, Jenq T, Didier R, Moneta L, Bardo D, Selden NR
- 9:24 - 9:30 am Discussion



10:00 am - 12:00 pm
Walk Festival Hall

Innovation and Invention in Craniofacial Surgery - OPEN OPTIONAL SEMINAR

Speaker: *T. Forcht Dagi, MD, DMedSc, MPH, FAANS, FCCM, FRCSEd*
Harvard Medical School, MIT and Queen's University Belfast

This seminar outlines the process of developing and commercializing new technologies in craniofacial surgery. Based on the curriculum developed for the Harvard-MIT Program in Health Sciences and Technology, it introduces an integrative approach to biomedical innovation and invention, covering market assessment, intellectual property protection, prototyping, regulatory issues, reimbursement strategy, fund raising, the licensing vs. spinout decision, strategic relationships and company structure. The seminar is intended to familiarize the craniofacial surgeon with the issues to be confronted and the resources available in bringing new technologies to market.

7:00 - 9:00 pm

Penn Alumni Dinner - Mangy Moose

7:00 - 9:00 pm

WCF Partner Dinner (by invitation) - Alpenhof Lodge

Saturday, 14 September - Walk Festival Hall

7:00 am - 5:00 pm

Registration

7:00 - 8:00 am

Continental Breakfast

8:00 - 10:00 am

**General Session 3
Craniofacial Distraction**

Moderators: *Yu-Ray Chen, MD (Taiwan) & Craig Birgfeld, MD (USA)*

8:00 am

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FRONTOFACIAL DISTRACTION. A RISK - BENEFIT ANALYSIS OF 90 CONSECUTIVE CASES

Presenter: David Dunaway, MBChB, FDSRCS, FRCS(Plast)

Affiliation: Great Ormond Street Hospital for Children

Authors: Dunaway D, Britto J, Abela C, Evans R, Jeelani O

8:07 am

190

**THE BIRMINGHAM EXPERIENCE OF POSTERIOR CALVARIAL DISTRACTION:
A 6-YEAR REVIEW**

Presenter: Hiroshi Nishikawa, FRCS(Plast)

Affiliation: Birmingham Childrens Hospital

Authors: Nishikawa H, Solanki G, Rodrigues D, White N, Evans M, Ahmad F, Noons P, Dover MS

8:14 am

191

**NORMALIZING FACIAL RATIOS IN APERT SYNDROME PATIENTS WITH LEFORT II
MIDFACE DISTRACTION AND SIMULTANEOUS ZYGOMATIC REPOSITIONING**

Presenter: Richard A. Hopper, MD, MS

Affiliation: Seattle Childrens Hospital

Authors: Hopper RA, Kapadia H

8:21 am

192

**A CRANIOMETRIC ANALYSIS OF POSTERIOR CRANIAL VAULT DISTRACTION
OSTEOGENESIS**

Presenter: Jesse A. Goldstein, MD

Affiliation: University of Pennsylvania School of Medicine & Childrens Hospital of Philadelphia

Authors: Goldstein JA, Paliga JT, Wink J, Bartlett SP, Taylor JA

8:28 am

193

**THE ORTHOMORPHIC MONOBLOCK: A POWERFUL TECHNIQUE TO PRODUCE
CROUZON'S PATIENTS WITH NORMAL APPARIENCE**

Presenter: Fernando Molina, MD

Affiliation: Hospital General Dr Manuel Gea Gonzalez SS

Author: Molina F



8:35 - 8:45 am

Discussion

8:45 am

194

MANDIBULAR GROWTH AFTER DISTRACTION OSTEOGENESIS: CASES OF PIERRE ROBIN SYNDROME IN EARLY CHILDHOOD

Presenter: Masakazu Hasegawa, MD

Affiliation: Chiba University

Authors: Hasegawa M, Mitsukawa N, Saiga A, Kaneko T, Akita S, Satoh K

8:49 am

195

CORRECTING THE TYPICAL APERT FACE: COMBINING BIPARTITION WITH MONOBLOC DISTRACTION

Presenter: Aina V. Greig, MA, PhD, FRCS(Plast)

Affiliation: Great Ormond Street Hospital

Authors: Greig AV, Britto JA, Abela C, Witherow H, Richards R, Evans RD, Jeelani NU, Hayward RD, Dunaway DJ

8:53 am

196

DOUBLE JAW SURGERY AFTER ADOLESCENCE FOR HEMIFACIAL MICROSOMIA CASES IN WHICH DISTRACTION WAS APPLIED IN CHILDHOOD

Presenter: Yuhki Uchida, MD

Affiliation: Chiba University

Authors: Satoh K, Uchida Y, Mitsukawa N, Akita S, Hasegawa K

8:57 am

197

POSTERIOR POLE EXPANSION WITH SPRINGS WITHOUT OSTEOTOMIES: RESULTS AND PREDICTIVE FACTOR OF SUCCESS

Presenter: Monica Fawzy, MD

Affiliation: Craniofacial Unit

Authors: Di Rocco F, Bennet C, Jebblaoui Y, Fawzy M, Wyten R, Perie AC, Vecchione A, Renier D, Arnaud E

9:01 am

198

FRONTOFACIAL MONOBLOC ADVANCEMENT WITH DISTRACTION IMPROVES CONSIDERABLY THE APNEA AND APNEA-HYPOPNEA-INDEXES IN A 2-YEAR FOLLOW-UP PERIOD

Presenter: Junnu Leikola, MD, DDS, PhD

Affiliation: Craniofacial Unit

Authors: Leikola J, Morisseau-Durand MP, Guerin P, Viot-Blanc V, Fauroux B, Diner P, Tomat C, Pierrot S, Couloigner V, Manach Y, Di Rocco F, Arnaud E

9:05 am

199

IMPROVING NUTRITIONAL OUTCOMES IN SYMPTOMATIC PIERRE ROBIN SEQUENCE: A COHORT COMPARISON STUDY BETWEEN TONGUE LIP ADHESION AND MANDIBLE DISTRACTION

Presenter: Lorelei Grunwaldt, MD

Affiliation: University of Pittsburgh School of Medicine

Authors: Rottgers A, MacIsaac Z, Wine T, Grunwaldt L, Mehta D, Kumar AR

9:09 am

200

SUB-CRANIAL ROTATION DISTRACTION ADVANCEMENT OF THE ENTIRE FACE FOR THE TREATMENT OF SEVERE OBSTRUCTIVE APNEA ASSOCIATED WITH SYMMETRIC CRANIOFACIAL MICROSOMIA

Presenter: Hitesh Kapadia, DDS, PhD

Affiliation: Seattle Childrens Hospital

Authors: Kapadia H, Hopper RA



- 9:13 am **201**
OUR PROVISIONAL EXPERIENCE OF APPLYING DISTRACTION OSTEOGENESIS IN REMODELING CRANIOSYNOSTOSIS
Presenter: Masayuki Miyata, MD, PhD
Affiliation: Niigata University Graduate School of Medical and Dental Sciences
Authors: Miyata M, Sakamura R, Tobisawa Y, Shibata M, Nishiyama K, Yoshimura J
- 9:17 am **202**
EVALUATION OF THE AIRWAY AFTER MONOBLOC DO A TEN YEAR FOLLOW UP
Presenter: Carlos R. Barcelo, MD
Affiliation: International Craniofacial Institute
Authors: Barcelo CR, Genecov DG, Salyer KE
- 9:21 - 9:30 am Discussion
- 9:30 am **203**
FRONTO-ORBITAL ADVANCEMENT (FOA) VERSUS POSTERIOR ENLARGEMENT, WHICH IS FIRST?: OUR CURRENT TIMING FOR THE SURGERY IN SYNDROMIC CRANIOSYNOSTOSIS
Presenter: Kaneshige Satoh, MD
Affiliation: Chiba University
Authors: Satoh K, Mitsukawa N, Akita S, Kubota Y
- 9:37 am **204**
HOW DISTRACTION VECTORS SHOULD BE USED IN APERT SYNDROME FOR THE CORRECTION OF THE SKULL AND FACE
Presenter: Pablo Arrieta, MD
Affiliation: Hospital General Dr Manuel Gea Gonzalez SS
Authors: Molina F, Arrieta P
- 9:44 am **205**
THE ROLE OF POSTERIOR CRANIAL VAULT DISTRACTION FOR THE TREATMENT OF CRANIOSYNOSTOSIS
Presenter: Yuzo Komuro, MD
Affiliation: Juntendo University Urayasu Hospital
Authors: Komuro Y, Matsumoto S, Mochizuki M, Koizumi T
- 9:51 - 10:00 am Discussion
- 10:00 - 10:30 am Coffee Break
- 10:30 am - 12:00 pm **General Session 4**
Craniofacial Distraction
Moderators: *Eric Arnaud, MD (France) & David Dunaway, MBChB, FDSRCS, FRCS(Plast) (United Kingdom)*
- 10:30 am **206**
FRONTO-FACIAL MONOBLOC AND LE-FORT III DISTRACTION OSTEOGENESIS IN CRANIOSYNOSTOSIS-THREE-DIMENSIONAL EVALUATION OF TREATMENT OUTCOME AND THE NEED OF CENTRAL DISTRRACT
Presenter: ChingHsuan Hu, MD
Affiliation: Chan Guan Memorial Hospital
Authors: Hu CH, Chieh-Tsai WU, Wen-Ching KO, Kuo-Ting CH
- 10:37 am **207**
EARLY MIDFACIAL DISTRACTION FOR SYNDROMIC CRANIOSYNOSTOTIC PATIENTS WITH OBSTRUCTIVE SLEEP APNEA
Presenter: Nobuyuki Mitsukawa, MD, PhD
Affiliation: Chiba University
Authors: Mitsukawa N, Saiga A, Kaneko T, Akita S, Satoh K



- 10:44 am **208**
OUR NEW PROCEDURE FOR PROBLEMS AND COMPLICATIONS OF LEFORT MIDFACE DISTRACTION
Presenter: Keisuke Imai, MD
Affiliation: Osaka City General Hospital
Authors: Imai K, Takahashi M, Yamaguchi K, Ishise H, Takara A, Nochi A
- 10:51 am **209**
THE IMPACT OF POSTERIOR CALVARIAL DISTRACTION ON CHIARI MALFORMATIONS AND SYRINGOMYELIA
Presenter: Martin Evans, MD
Affiliation: Birmingham Children's Hospital
Authors: Ahmad F, Solanki G, White N, Evans M, Nishikawa H, Dover S, Rodrigues D
- 10:58 am **210**
LONG-TERM STABILITY OF ANTERIOR OPEN-BITE CLOSURE WITH DISTRACTION OSTEOGENESIS IN TREACHER COLLINS SYNDROME
Presenter: Jose Cortes-Arreguin, MD
Affiliation: Hospital General Dr Manuel Gea Gonzalez
Authors: Cortes-Arreguin J, Garcia-Garcia F, Molina F
- 11:05 - 11:15 am Discussion
- 11:15 am **211**
SURGICAL ANATOMY OF SPHENOMAXILLARY DISJUNCTION IN THE LE FORT III OSTEOTOMY
Presenter: William S. Tierney, MS
Affiliation: Cleveland Clinic Foundation
Authors: Tierney WS, Orra S, Doumit G
- 11:19 am **212**
ANATOMICAL STUDY USING CADAVERS FOR IMAGING OF LIFE-THREATENING COMPLICATIONS IN LE FORT III DISTRACTION
Presenter: Shinsuke Akita, MD, PhD
Affiliation: Chiba Cancer Center
Authors: Akita S, Mitsukawa N, Hasegawa M, Kubota Y, Satoh K
- 11:23 am **213**
BONE TRANSPORT OSTEOGENESIS FOR THE TREATMENT OF LARGE CALVARIAL DEFECTS: AN OVINE MODEL
Presenter: Patrick A. Gerety, MD
Affiliation: University of Pennsylvania
Authors: Gerety PA, Wink JD, Sherif R, Clarke NA, Nah HD, Taylor JA
- 11:27 am **214**
INITIAL EXPERIENCE WITH NEW INTRAORAL MIDFACE DISTRATOR, (IMD), INDICATIONS AND TECHNIQUE
Presenter: Fernando Burstein, MD
Affiliation: Emory University
Authors: Burstein F, Berhane C, Schoemann M
- 11:31 am **215**
IMAGE-GUIDED TRANSFACIAL PIN MIDFACE DISTRACTION IN INFANTS RESULTS IN OBSTRUCTIVE SLEEP APNEA RESOLUTION VERIFIED BY POLYSOMNOGRAPHY
Presenter: Robert M. Menard, MD, FACS
Affiliation: Surgical Director Northern California Kaiser Permanente Craniofacial Clinic
Authors: Menard RM



- 11:35 am **216**
A SYSTEMATIC REVIEW OF THE EFFECTIVENESS OF MANDIBULAR DISTRACTION IN IMPROVING AIRWAY OBSTRUCTION IN CHILDREN WITH MANDIBULAR HYPOPLASIA
Presenter: Youssef Tahiri, MD
Affiliation: McGill University
Authors: Tahiri Y, Viezel Mathieu A, Aldekhayel S, Lee J, Gilardino M
- 11:39 am **217**
ALSO FOR ADULTS WE PREFER DISTRACTION - TREATMENT OF SAOS
Presenter: Alberto Rocha Pereira, MD
Affiliation: Portuguese Armed Forces Hospital & Portuguese Oncological Institute
Authors: Rocha Pereira A, Matos I, Neves P, Montesuma N, Pires J, Ferreira R, Duarte JM
- 11:43 am **218**
NEONATAL MANDIBULAR DISTRACTION IN PIERRE ROBIN SEQUENCE: A VOLUMETRIC ANALYSIS OF THE MANDIBLE AND AIRWAY
Presenter: Patrick C. Hettinger, MD
Affiliation: Medical College of Wisconsin/Childrens Hospital of Wisconsin
Authors: Hettinger PC, Deschamps-Braly J, Denny AD
- 11:47 am - 12:00 pm Discussion
12:00 - 1:00 pm Lunch

1:00 - 3:00 pm **General Session 5**
Syndromic Craniosynostosis
Moderators: *Steven Wall, MD (United Kingdom) & Xiongzhen Mu, MD (China)*

- 1:00 pm **219**
DOES THE JUGULAR FORAMEN INFLUENCE INTRACRANIAL PRESSURE IN CRANIOSYNOSTOSIS PATIENTS?
Presenter: Joyce Florisson, MD
Affiliation: Erasmus Medidal Centre
Authors: Florisson J, Lequin M, Van Veelen ML, Bannink N, Mathijssen IM
- 1:07 pm **220**
THE ROLE OF EARLY ENDOSCOPIC RELEASE IN THE MANAGEMENT OF SYNDROMIC AND NON-SYNDROMIC BILATERAL CORONAL CRANIOSYNOSTOSIS
Presenter: Mark R. Proctor, MD
Affiliation: Boston Childrens Hospital
Authors: Proctor MR, Lohani S, Rogers GF, Meara JG
- ~~1:14 pm **221**
REGRESSION OF CHRONIC HINDBRAIN HERNIA AND SYRINGOMYELIA FOLLOWING POSTERIOR CALVARIAL AUGMENTATION IN CHILDREN: NEW INSIGHTS INTO PATHOLOGY OF HINDBRAIN HERNIA
Presenter: Guirish A. Solanki, MBBS, FRCSI, FRCS(SN)
Affiliation: Birmingham Childrens Hospital & University of Birmingham
Authors: Solanki GA, Farooq U, Lo W, Paternoster G, Davies P, Rodrigues D, White N, Evans M, Dover S, Nishikawa H~~
- WITHDRAWN**
- 1:21 pm **222**
SYNDROMIC CRANIOSYNOSTOSIS AND CHIARI TYPE I MALFORMATION: A VOLUMETRIC STUDY OF THE POSTERIOR FOSSA. DOES SIZE MATTER?
Presenter: Bianca Rijken, MD
Affiliation: Erasmus Medical Center University
Authors: Rijken B, Lequin MH, van der Lijn F, van Veelen-Vincent ML, Niessen W, Mathijssen IM



- 1:28 pm **223**
POSTERIOR CRANIAL VAULT DISTRACTION IN THE MANAGEMENT OF SYNDROMIC MULTI-SUTURE CRANIOSYNOSTOSIS: OUTCOMES AND 3D PHOTOGRAPHIC/CT-BASED MORPHOMETRIC ANALYSIS
Presenter: Brooke French, MD
Affiliation: The Hospital for Sick Children
Authors: French B, Clausen A, Forrest CR
- 1:35 pm **224**
CHARACTERISTICS OF POSTNATAL PROGRESSIVE PANSYNOSTOSIS
Presenter: Albert K. Oh, MD
Affiliation: Childrens National Medical Center
Authors: Oh AK, Sauerhammer TM, Magge KT, Magge SN, Myseros JS, Keating RK, Rogers GF
- 1:42 - 1:50 pm Discussion
- 1:50 pm **225**
ENHANCING THE PROCESS OF COMPLEX CRANIOFACIAL RECONSTRUCTION: LESSONS LEARNED IN 94 CASES USING PREOPERATIVE COMPUTER MODELING
Presenter: Patrick Kelley, MD
Affiliation: Dell Childrens Medical Center of Central Texas
Authors: Kelley P, Harshbarger RJ, Henry SL, Combs PD
- 1:54 pm **226**
ABNORMAL GROWTH OF THE FORAMEN MAGNUM IN CROUZON SYNDROME: SIZE AND CLOSING OF THE INTRA-OCCIPITAL SYNCHONDROSES
Presenter: Caroline Driessen, MD
Affiliation: Erasmus Medical Center University
Authors: Rijken B, Driessen C, Lequin MH, van Veelen-Vincent ML, Mathijssen IM
- 1:58 pm **227**
CLOSURE OF THE SPHENO-OCCIPITAL SYNCHONDROSIS IN PATIENTS WITH SYNDROMIC CRANIOSYNOSTOSIS: A LIKE TO MIDFACE HYPOPLASIA
Presenter: Jesse A. Taylor, MD
Affiliation: University of Pennsylvania School of Medicine & Childrens Hospital of Philadelphia
Authors: Goldstein JA, Paliga JT, Bartlett SP, Taylor JA
- 2:02 pm **228**
INTRACRANIAL PLATING WITH RESORBABLE PLATES FOR CRANIOFACIAL RECONSTRUCTION
Presenter: Peter T. Wang, MD, DMD
Affiliation: Pediatrix
Authors: Wang PT, Gennuso R, Megahead H, Garcia CA, Levine RA
- 2:06 pm **229**
PSEUDOMENINGOCELE WITH ORBITAL EXTENSION AS A COMPLICATION OF FRONTO-ORBITAL ADVANCEMENT AND REMODELLING IN CRANIOSYNOSTOSIS: A RETROSPECTIVE REVIEW
Presenter: Ajay Sinha, MD
Affiliation: Alder Hey Hospital for Children
Authors: Vaiude P, Sinha A, Burn SC, Richardson D, Sweeney E, Duncan C
- 2:10 - 2:20 pm Discussion
- 2:20 pm **230**
IDENTIFYING WHICH MORPHOLOGICAL ABNORMALITIES OF APERT SYNDROME ARE CORRECTED BY BIPARTITION DISTRACTION
Presenter: Nicki Bystrzonowski, MD
Affiliation: Great Ormond Street Hospital
Authors: Ponniah A, Bystrzonowski N, Verdoorn M, Nikkhah D, Ruff C, Dunaway D



2:24 pm	231 CHILDREN WITH CROUZON SYNDROME AS ADULTS. A FOLLOW-UP STUDY OF 31 SWEDISH PATIENTS Presenter: Lars Kolby, PhD Affiliation: Institute for Clinical Sciences Authors: Kolby L, Fischer S, Tovetjarn R, Maltese G, Sahlin PE, Tarnow P
2:28 pm	232 EFFECT OF MONOBLOC FRONTO-FACIAL BIPARTITION DISTRACTION ON ORBITAL VOLUME, MORPHOLOGY, AND CLINICAL OUTCOME IN 15 APERT SYNDROME CASES: A CONTROLLED STUDY Presenter: Roman Khonsari, MD Affiliation: Great Ormond Street Hospital Authors: Khonsari R, Karunakaran T, Way B, Matthews W, Nysjo J, Nystrom I, Dunaway DJ, Evans RD, Britto JA
2:32 pm	233 COMPLICATIONS AFTER FRONTOFACIAL MONOBLOC ADVANCEMENT WITH INTERNAL QUADRUPLE DISTRACTION IN CHILDREN Presenter: Eric Arnaud, MD Affiliation: Craniofacial Unit Hopital Necker Authors: Arnaud E, Saiepour D, Leikola J, Wyten R, Di Rocco F, Meyer PH, Sainte-Rose C
2:36 - 3:00 pm	Discussion
3:00 - 3:30 pm	Coffee Break
3:30 - 5:00 pm	General Session 6 Syndromic Craniosynostosis Moderators: <i>Stephen Dover, FDSRCS, FRCS (United Kingdom) & Kaneshige Satoh, MD (Japan)</i>
3:30 pm	234 WHERE ARE THEY NOW?: GEOGRAPHICAL AND SOCIAL OUTCOMES ON 102 ADULTS WITH CRANIOFACIAL SYNDROMES Presenter: Stephen Dover, FDSRCS, FRCS Affiliation: Birmingham Childrens Hospital UK Authors: Dover S, Venugopal A, Noons P, Evans M, White N, Solanki G, Rodrigues D, Nishikawa H
3:37 pm	235 PREOPERATIVE TREATMENT WITH MOLDING HELMET IMPROVES CEPHALIC INDEX IN BICORONAL SYNOSTOSIS Presenter: Arelene Rozzelle, MD Affiliation: Childrens Hospital of Michigan Authors: Rozzelle A, Sood S, Sofer JT
3:44 pm	236 FRONTO-ORBITAL EXPANSION AS KEY PROCEDURE IN CORRECTION OF CRANIOSYNOSTOSIS Presenter: Xiongzhen Mu, MD Affiliation: Huashan Hospital Fudan University Authors: Mu X, Yang JY, Bao N, Guo ZL
3:51 pm	237 ADVERSE EVENTS ASSOCIATED WITH THE USE OF SPRINGS FOR MOVING CRANIAL BONE Presenter: Roisin McNicholas, MD Affiliation: Great Ormond Street Hospital Authors: Jeelani O, Hayward R, McNicholas R
3:58 - 4:15 pm	Discussion



4:15 pm

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RADIOLOGICAL EVALUATION AFTER FRONTOFACIAL MONOBLOC ADVANCEMENT WITH INTERNAL QUADRUPLE DISTRACTION IN CHILDREN

Presenter: Daniel Saiepour, MD, PhD

Affiliation: Centre de Reference de Malformation Cranio Facialis

Authors: Saiepour D, Wyten R, Leikola J, Meyer PH, Oyama A, Di Rocco F, Arnaud E

4:22 pm

239

CENTRALIZED CARE FOR CRANIOSYNOSTOSIS

Presenter: Irene Mathijssen, MD, PhD

Affiliation: Erasmus MC

Author: Mathijssen I

4:29 pm

240

~~SIMULTANEOUS FRONTO-ORBITAL ADVANCEMENT AND DYNAMIC POSTERIOR CRANIAL VAULT EXPANSION IN APERT SYNDROME~~

WITHDRAWN

~~Presenter: Emmanuela Nadal Lopez, MD, PhD~~

~~Affiliation: Hospital de Pediatria Prof Dr Juan P Garrahan~~

~~Authors: Nadal Lopez E, Dogliotti P, Sabas M~~

4:36 pm

241

SPRING-ASSISTED POSTERIOR EXPANSION AND SIMULTANEOUS FRONTO-ORBITARY EXPANSION IN SYNDROMIC CRANIOSYNOSTOSIS

Presenter: Javier Gonzalez Ramos, MD

Affiliation: Garrahan Pediatric Hospital

Authors: Gonzalez Ramos J, Routaboule C, Zuccaro G

4:43 pm

242

'POSTERIOR CALVARIAL EXPANSION USING CUSTOM MADE SPRINGS'; THE EVOLUTION OF A SURGICAL TECHNIQUE

Presenter: Owase Jeelani, MBA, MPhil, FRCS

Affiliation: Great Ormond Street Hospital

Authors: Jeelani O, Hayward R

4:50 - 5:00 pm

Discussion

6:00 - 9:00 pm

Farewell Dinner - Commons Tent - "*Bandana Tickets*" Required



ABSTRACTS



1 FIBROUS DYSPLASIA OF THE ZYGOMATICOMAXILLARY REGION: OUTCOMES OF SURGICAL INTERVENTION

Presenter: Phuong D. Nguyen, MD
Authors: Andrews BT, Gabbay JS, Yuan JT,
Kawamoto HK, Bradley JP
University of California Los Angeles

Background: Fibrous dysplasia is the most common craniofacial tumor, presenting in both monostotic and polyostotic forms with varying degrees of severity. No consensus exists regarding the surgical management of craniofacial fibrous dysplasia, particularly in the zygomaticomaxillary region. The present study compared long-term outcomes of limited reduction burring vs. radical resection of zygomaticomaxillary fibrous dysplasia.

Methods: Patients with craniofacial fibrous dysplasia at the UCLA Craniofacial Center from 1982 to 2008 were studied based on demographics, treatment, and follow-up data including examinations, CT scans, photographs, physician Whitaker scoring, and patient surveys (n=97). Outcomes were compared for zygomaticomaxillary disease treated with radical resection with cranial bone graft reconstruction or limited reduction burring (n=58).

Results: 34% of patients had monostotic disease, 66% had polyostotic disease, 3% had McCune-Albright syndrome, and 2.1% had malignant degeneration into osteosarcoma. Most patient had surgical treatment (84.5%). Of the patients that required optic nerve decompression for vision changes (11.4%), most (75%) had vision stabilization postoperatively. Differences were recorded in zygomaticomaxillary disease treated with radical resection (63.8%) versus reduction burring (36.2%) according to: age (19.6 vs. 14.2 years), complications (13.5% vs. 4.8%), recurrence (66.7% vs. 24.3%), and number of subsequent procedures (2.8 vs. 4.0). There were similarities in Whitaker outcome score (1.3+0.3 vs. 1.5+0.6) and patient satisfaction (2.7+0.4 vs. 2.8+0.3).

Conclusions: While different approaches have been advocated to treat fibrous dysplasia, our data supports a more aggressive management for zygomaticomaxillary disease with radical resection and cranial bone graft reconstruction especially for more involved disease.

2 CRANIOFACIAL AND HEAD, AND NECK NEUROFIBROMATOSIS: CLINICAL CLASSIFICATION AS AN AID TO SURGICAL TREATMENT

Presenter: Joseph S. Gruss, MD, FRCSC, FAAP
Authors: Gruss JS, Latham KL, Buchanan EB,
Suver DS
Seattle Childrens Hospital

Craniofacial and Head and Neck Neurofibromatosis (NF) is common with variable presentation. Surgical treatment is challenging and the choice of surgical method controversial.

A 20 year review of patients treated surgically for complex head and neck neurofibromatosis was performed. 59 patients were identified and categorized into 5 distinct, but not exclusive, categories to assist with diagnosis and surgical management. 7 patients had plexiform NF. 3 had involvement of the supraorbital nerve with disease restricted to the course of the nerve. 24 patients had cranio-orbital NF, a very distinct clinico-pathological entity. The majority required multiple operations for resection and reconstruction of the involved soft tissue and bone. Patients with sphenoid wing dysplasia and pulsatile exophthalmos were corrected by a combined intra/extracranial approach. In patients requiring enucleation, superior esthetic results were obtained with complete orbital exenteration and reconstruction with a complete orbital prosthesis compared to enucleation and attempted preservation of abnormal eyelids. 4 patients were in the facial NF category. All these patients required multiple operations for debulking and soft tissue resuspension. Resection was often accompanied by major blood loss. 15 patients in the parotid auricular group had 16 facial nerve dissection (1 bilateral). Dissection with intraoperative monitoring was successful in all but 1 patient. All patients had occipital skull defects and required multiple surgeries. 9 patients had major neck involvement alone. 12 patients had neck involvement in addition to other involvement. 10 patients requiring brachial plexus dissection had relief of compressive neuropathy symptoms. Resection often required access osteotomies of the clavicle and sternum to provide access to the chest and mediastinum. None of the neck patients required reoperations for recurrences. A clinically based classification of Neurofibromatosis of the head and neck into 5 categories has facilitated surgical management to this challenging problem and improved functional and aesthetic outcome.



3 IMPROVEMENT OF TREATMENT OF FACIAL ARTERIO- VENOUS VASCULAR MALFORMATIONS BY MULTIPLE EMBOLIZATIONS: OUTCOMES IN A SERIES OF 31 PATIENTS

Presenter: Dov C. Goldenberg, MD, PhD
Authors: Goldenberg DC, Hiraki PY, Puglia P,
Caldas JG, Ferreira MC

Hospital das Clinicas University of Sao Paulo Medical School

Purpose: Combining superselective embolization and surgery is the most accepted method for treatment of arterio-venous malformations (AVM). Multiple sessions of embolizations may be required to control disease progression or hemorrhage, promoting transitory ischemia and fibrosis. The purpose of this study was to evaluate if surgical resection was facilitated by multiple embolization procedures.

Methods: 31 patients, operated between January 2000 and December 2012, after at least 2 embolization procedures were included. Mean age was 24.9 years. Lesions were analyzed according to anatomic compromise and functional impairment. Number of embolization per patient was evaluated. Type of resection and reconstruction, ease of resection, need of blood transfusion, Intensive Care Unit and hospital stay were evaluated. Morbidity/mortality rates, functional and esthetic improvement, need for additional procedures and re-growth were considered for outcome evaluation.

Results: Lesions were preferentially located at the orbits, cheeks and lips. 23 patients had functional impairment and 12 had a previous history of significant bleeding. All patients had esthetic problems. Number of embolization per patient increased with lesion complexity. In 24 cases (77.4%) resection was complete and in 7 cases subtotal resection was performed to favor function. It was possible to confirm a better identification of the vascular mass, surrounded by a fibrous tissue, making them more delimited from adjacent structures. In some cases, multiple embolizations simulated the formation of a capsule. Primary closure was performed in 20 cases, local flaps in 7 cases, regional flaps in 2 and free flaps in 2 cases. No mortality was observed in this series. Regrowth occurred in 7 cases. Esthetic improvement after surgical resection was observed in 26 from 31 patients. Functional improvement was observed in 21 patients.

Conclusion: Multiple therapeutic embolizations performed previously to surgical resection seems to increase safety in the treatment of high flow vascular malformations and suggest an additional positive effect besides bleeding control.

4 THE CHOICE OF SKULL BASE RECONSTRUCTION VARIANT IN THE TREATMENT OF TUMORS WITH CRANIOFACIAL LOCALIZATION

Presenter: Igor Reshetov, MD
Authors: Reshetov I, Chissov VI, Cherekaev VA,
Zaitzev AM, Belov AI, Polyakov AP,
Matorin OV, Ratushniy MV

Moscow P A Herten Cancer Research Institute

NOT PRESENTED



5 TREATMENT OF CRANIOMAXILLOFACIAL FIBROUS DYSPLASIA IN OUR DEPARTMENT

Presenter: Eiji Miyamoto, MD

Authors: Miyamoto E, Okumoto T, Yoshimura Y
Fujita Health University School of Medicine

Object: Fibrous Dysplasia is a benign osseous disease defined as a tumor-like osseous lesion in which normal bone is replaced by fibro-osseous tissue. In previous reports, radical resection is recommended because of the recurrence or the possibility of malignant change. But actually, radical resection is impossible particularly in the maxillofacial region. In our department, conservative shaving is selected aiming reduction of the tumor volume to achieve as normal facial aesthetics as possible. We will present our cases.

Methods: We have experienced eight cases of craniomaxillofacial fibrous dysplasia. Among them, six were treated by conservative shaving and two have been observed. Although optimal time of surgery is not defined, surgery was performed over approximately twenty years old when the osseous lesion seemed to stop progression.

Results: Six cases treated by conservative shaving showed no recurrence until now. In some cases postoperative observation is not long enough, so we should observe carefully from now on. Their facial contour had been improved remarkably.

Conclusion: Fibrous Dysplasia is a benign osseous disease, and commonly recommended to do radical resection because of the recurrence or possibility of malignant change. But in our department, conservative shaving is successfully applied to the maxillofacial lesions.

6 MODIFIED FACIAL TRANSLOCATION APPROACH IN ONE MONTH OLD WITH SKULLBASE TUMOR

Presenter: Sherard A. Tatum, MD

Author: Tatum SA
State University of New York

This is a case presentation of a neonate born with a germ cell tumor of the right middle cranial fossa, orbit, sphenoid, pharynx, and maxilla. The baby presented with dyspnea, poor feeding and right proptosis. Vision in that eye was thought to be minimal at best. Tracheostomy was required. CT imaging revealed a heterogenous mass in the area described above. Needle aspiration biopsy yielded a diagnosis of a teratoma/germ cell tumor hybrid. The Hematology/Oncology service instituted chemotherapy which had limited effect. Surgical intervention was requested. Further review of the radiographs informed that the lesion was contacting the right carotid and optic nerve. Endoscopic surgery was considered but not thought feasible. An extracranial approach was chosen with a goal of subtotal excision. A right modified Weber-Ferguson facial incision was performed traversing the lower lid just lateral to the punctum and extended as a transconjunctival incision then through the lateral canthus. The lip incision was extended just right of midline across the hard palate to the soft palate where it was turned transversely to the right to just behind the maxillary tuberosity. There was no soft tissue elevation. Osteotomies were then made following the incisions sparing the medial wall of the maxilla and the lacrimal system. Then pterygomaxillary disjunction was performed freeing the maxilla. The maxilla pedicled on the cheek was reflected laterally exposing the tumor. The tumor was resected with good exposure to the dura of the eroded middle cranial fossa. The carotid artery and orbital apex were visualized. The cheek flap and maxilla were returned to anatomic position. The bone was not fixated other than by the closure of the incisions. The patient tolerated the procedure well with estimated blood loss of 100 ml. Postoperative imaging revealed subtotal tumor removal. The facial incisions healed very well. A slit hard palate fistula developed that is planned to be repaired at one year of age. Chemotherapy was resumed, and the patient remains in remission. This approach proved direct and efficient for safely accessing this tumor.



7
REVIEW OF VARIOUS SURGICAL PROCEDURES IN RADIATED ORBIT RECONSTRUCTION

Presenter: Junyi Yang, MD
Authors: Mu X, Yang JY, Guo ZL, Yu ZY, Yang XX
Huashan Hospital Fudan University

Purpose: To review the different results with various surgical procedures in past 15 years in radiated orbit reconstruction.

Method: We classified three degree for indication of severity of radiated orbit based on the involved skeleton. Surgical procedures used in our series are one stage orbital implant with pedicel flap, orbital enlargement with free flaps coverage, orbital enlargement using distraction osteogenesis, injectable auto-fat transfer, and skin/mucosa graft in eye socket.

Result: Complicated orbital osteotomies with free flap coverage are worth to reconstruct the orbit dysplasia and eye socket atrophy but relapse still remain in severe cases of radiated orbit. One stage implants insertion with flaps is benefit for moderate dysplasia but 10% of complication of exposed material was happened in the long run.

8
MIDLINE FRONTAL NASAL DERMoids - A REVIEW OF 55 CASES AND A PROTOCOL FOR TREATMENT

Presenter: Mike A. Moses, BM BCh BSc FRCSEd(Plast)
Authors: Moses MA, Green BC, Cugno S, Jeelani NO, Bulsrode NW, Dunaway DJ
Great Ormond Street Hospital

Introduction: The incidence of midline frontonasal dermoids cysts has been estimated as 1 in 20000 to 1 in 40000. As a result of the embryology of this region, it is well-recognised that these lesions may have intracranial extension. In addition, skin involvement may also be extensive, with several punctums and hair-containing tracks located in the skin of the dorsum of the nose. Incomplete excision leads to recurrence in 50-100% of cases. Over the past 15 years we have developed a patient pathway to investigate and manage midline frontonasal dermoids.

Methods: Craniofacial, plastic surgery and neurosurgery databases were searched to identify patients who had undergone surgery for removal of a dermoid cyst. Those that were not in the midline, or not frontonasal (eg anterior fontanelle) were excluded. Preoperative imaging and indications for surgery were reviewed. Cases were grouped according to surgical approach, and outcomes and complications identified.

Results: Over 15 years, 55 patients with midline dermoids were treated at a nationally designated craniofacial unit. MRI or CT was used to delineate anatomy, and surgical excision was expedited if there was a clinical history of infection or if hairs were present at the sinus opening (especially if intracranial extension was apparent on imaging). 12 were treated endoscopically (one converted to open). 11 required transcranial approaches for intracranial extension (20%). Of these, one lesion breached the dura. The remaining 32 patients had dermoids excised with an open approach, which was either direct, bi-coronal or rhinoplasty. Overall there were no recurrences in the open group; one in the transcranial group. This was treated by re-excision. Another patient in the transcranial group was readmitted for treatment of an infection with intravenous antibiotics.

Conclusion: Midline dermoid cysts are relatively uncommon. However, with increasing experience it is possible to develop a safe and predictable approach to their management, resulting in low incidence of complications and recurrence.



9 LANGERHANS CELL HISTIOCYTOSIS IN THE PEDIATRIC POPULATION: HOW SHOULD WE TREAT ISOLATED CRANIOFACIAL LESIONS?

Presenter: Francesco Gargano, MD, PhD
Authors: Gargano F, Klinge PM, Welch J, Sullivan SR,
Taylor HO

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Background: Langerhans cell histiocytosis (LCH) incidence is 0.2 - 0.5 per 100,000 children per year in the US and commonly affects the craniofacial skeleton. Multifocal disease is typically responsive to chemotherapy, but treatment of unifocal craniofacial lesions is not as well delineated. Treatment algorithms for unifocal LCH vary dramatically. Little has been published on outcomes of isolated lesions, but one series suggests recurrence rates after curettage alone of 60%.

Materials and Methods: After IRB approval we reviewed records for patients with LCH treated at our center between 2000 and 2013. We recorded data on clinical presentation, location, extent, therapy, surgical time, transfusion rate, complications. Outcomes included response to treatment, recurrence, and bony healing.

Results: We identified twenty-five patients with LCH with mean age at diagnosis of 5.6 years (range 0.7-14). Fifteen patients (60%) had craniofacial involvement, while 10 (40%) had no craniofacial involvement with disease restricted to long bones (4), skin (3), lungs (2), and spine (1). Patients with multifocal bone disease, CNS-risk lesions or multi-system disease were treated with chemotherapy (vinblastine, prednisone, mercaptopurine). Six patients with isolated calvarial lesions were treated with craniectomy and cranioplasty. Three patients had mastoid lesions: one multifocal treated with chemotherapy alone and two unifocal treated with excision and chemotherapy. Mean follow-up was 3.9 years (range 0.4-12 years). No recurrence occurred in 17 patients treated with excision. Of six patients treated with curettage or excisional biopsy, 50% recurred. Two patients with multisystem disease had organ dysfunction.

Discussion: Many treatments are advocated for craniofacial LCH including observation, curettage, steroids, antibiotics, chemotherapy and resection. We had no recurrences in patients with unifocal calvarial lesions who underwent excisional biopsy for diagnosis and treatment by craniectomy with cranioplasty. Recurrences did occur in those undergoing curettage alone. Response to chemotherapy is excellent with minimal morbidity

10 COMPLICATIONS IN MEDIAL FACIOTOMY

Presenter: Sarah L. Versnel, MD, PhD
Authors: Versnel SL, Mathijssen IM
Erasmus Medical Center

Background: In 1979 medial faciotomy was described as a new technique for correction of hypertelorism. Since then multiple patients have been treated in our center with this technique. In this paper we discuss the minor and major complications on the short and long term of medial faciotomy/facial bipartition.

Methods: All patients who had a medial faciotomy for correction of hypertelorism in our center were analysed. Median, paramedian, and oblique facial clefts were included, as well as patients with a hypertelorism associated with craniosynostosis or acrocephalosyndactyly.

Results: In total 22 medial faciotomies were performed. All relevant data were included in the analysis, like bloodloss, infection rate, dural tears, rare complications, and long-term results.

Conclusion: Medial faciotomy has shown good and stable results over time. This technique benefited from distraction, which lowered the complication rates. Major complications are limited with this technique when performed by experienced surgeons.



II OPTIMIZING THE TIMING AND TECHNIQUE OF TREACHER COLLINS ORBITAL MALAR RECONSTRUCTION

Presenter: Justine C. Lee, MD, PhD
Authors: Fan KL, Federico C, Kawamoto HK,
Bradley JP
University of California Los Angeles

Background: The optimal timing and treatment of Treacher Collins syndrome with regard to zygomatico-orbital osseous reconstruction has not been fully established. Osseous reconstruction performed at an early age may result in bone graft resorption, but delays in surgical improvement may result in adverse psychosocial effects on the patient. In this study, we attempted to establish the optimal age at which a patient with Treacher Collins should undergo orbital/malar reconstruction based on bone resorption rate. We also surveyed the satisfaction of the patient/parents after the procedure.

Methods: To study the optimal age for reconstruction clinically, we examined three age groups based on timing of malar and eyelid reconstruction using a 3D CT scan and 3D photometric volume assessment. In addition, we collected outcome assessments from parents/patients using satisfaction surveys. Of 73 Treacher Collins patients at the UCLA Craniofacial Clinic, 45 had malar reconstruction with full-thickness parietal bone grafts, eyelid switch flaps and lateral canthopexies. The patients were separated into three groups: 1) Very Young = 0-5 years 2) Mid-childhood = 6-12 years, and 3) Adolescent/adult ≥13 years.

Results: As predicted, the Very Young group (with a mean age of 4.8 years) had the greatest percentage of complete bony resorption (77%) compared to Mid-childhood (mean age 9.9 years) with 4%, and Adolescent/adult (mean age of 16.4) with 0%. Partial bone resorption requiring fat grafting occurred in the Very Young (33%), Mid-Childhood (21%) and adolescents (14%). Complications (wound infection, vision problems, excessive scarring reoperations) were low and similar in all three groups. The Very Young group experienced the highest parent/patient satisfaction (3.8 vs. 3.5 and 3.0) possibly due to better psychosocial experience.

Conclusion: In treatment plans for Treacher Collins patients, the benefits of early surgical intervention such as improved psychological well being must be balanced with the disadvantages of undergoing a greater number of procedures like fat grafting after malar bone graft resorption.

I2 TESSIER 0-14 CLEFTS FROM BIRTH TO MATURITY: A REVIEW

Presenter: Walter J. Flapper, FRACS, MASurg
Authors: Flapper WJ, Pidgeon T, David DJ,
Anderson PJ
Australian Craniofacial Unit

Introduction: Rare craniofacial clefts are a rare set of malformations that represent an important segment of craniofacial pathology. Despite this, there remains a paucity of literature regarding management from birth to maturity. Large observational studies have shown that the midline (Tessier 0-14) clefts are the most commonly encountered of the rare clefts.

This study reviews the first cohort of Tessier 0-14 clefts that have completed management according to protocol from birth to skeletal maturity in a single centre.

Methods: A retrospective review of the clinical, photographic and radiographic records of all midline cleft patients was carried out. Strict inclusion and exclusion criteria were applied to ensure we only looked at patients with isolated Tessier 0-14 clefts.

Results: 4 patients (one male, three female) were identified as having completed protocol management at our unit for Tessier 0-14 midline clefting. Age range at their most recent follow-up was from 19.3 to 36.3 years. Three entered protocol management in infancy, the remaining patient presented to the unit at 13.8 years of age. Outcomes for development, hearing, speech and vision at maturity were all acceptable. Three patients attained a respectable educational and social status at maturity. When facial aesthetics were scored, the only significant difference after management was a significant increase in deformity in the region of the orbits (score increased from 3.25 to 5.50 after management, $P=0.02$, paired two-tailed student "t" test). The Whittaker grade for repeat surgery did however improve after management (3.25 pre- to 2.63 after management), but failed to attain significance ($P=0.14$).

Conclusion: Presented is the first cohort of midline Tessier 0-14 patients to have completed protocol management at a single craniofacial unit. Results are favourable, however much regarding an optimum protocol for midline cleft patients is still unknown.



13 RARE CRANIOFACIAL CLEFTS AND AGENESES

Presenter: S. Anthony Wolfe, MD
Authors: Wolfe SA
Miami Childrens Hospital

Rare Craniofacial Clefts and Ageneses: Dr. Tessier's unfinished and unpublished new classification.

All of us know the Tessier Classification of Facial Clefts (1976). It is the lingua franca worldwide for describing facial clefts.

In the last few years of his life Dr. Tessier came to believe that the 1976 classification was inadequate, and was working on a new form, with clefts in one category, and ageneses in a parallel, but different category.

Clinical examples with surgical results of all of the Tessier clefts, from 0 to 14, to 30, operated on by the author over a 37 year period, will be shown, with an incomplete number of the parallel ageneses.

14 SECONDARY CORRECTION OF HTO AVOIDING PROBLEMS DURING SKELETAL MOBILIZATION

Presenter: Juan M. Chavanne Sr., MD
Authors: Chavanne JM, Schauvinhold C, Steinberg D, Pringles D
WCF Bs As Craniofacial Center

Background: Secondary correction of HTO can represent a challenge for surgeons who have to deal not only with the malformed and mal positioned skeleton but also with the altered anatomy of the orbits very often destroyed or debilitated as result of a previous surgery. Under these circumstances re-addressing both orbits to the middle line to achieve a normal IOD, volume symmetry and aesthetic nose appearance can be very difficult. Fracture/disruption of the orbito-facial segment during the mobilization is one of the possible intraoperative complications. Although this is less frequent when using DOG process, it can still be present. This paper presents our experience using some guidelines to prevent these problems during operative procedure.

Methods: Eight patients received with residual HTO following primary surgery were operated. Patients were separated into 2 groups depending on the type of second procedure adopted: Group I: Six patients underwent a secondary facial bi-partition Group II: Two patients received orbital box transposition surgery. Prevention measures were taken before orbital mobilization including: 1-Transitory stabilization of inner and lateral facial buttress 2- Complete rebuilding of orbital walls. 3-Pterigo-maxillary disjunction prior the midline osteotomy.

Results: High-level lateral fractures often seen in secondary cases did not occur in any group. Mobilization of the orbits was achieved without fracture or disruption of the skeletal segments. Orbital volume and symmetry were satisfactory in most of the asymmetric cases. Restoration of the IOD and aesthetic nose improvement were obtained in all the patients.

Conclusions: Preserve the integrity of the osseous framework; re-building the entire orbital wall before the intraoperative mobilization can be helpful to avoid devastating complications during mobilization of the orbito-facial segments.



15
**TOWARDS PREDICTABLE AESTHETIC CHANGE IN
HYPERTELORISM SURGERY - A RADIOLOGIC STUDY
IN 18 PATIENTS AND 30 CONTROLS**

Presenter: Jonathan A. Britto, MB, MD, FRCS(Plast)
Authors: Syme Grant J, Karunakaran T, Abela C
Dunaway DJ, Evans RD, Britto JA
Great Ormond Street Hospital for Children

Surgical refinements in hypertelorism (HPT) correction aim to close the gap between patient expectation and operative outcome. We ask whether quantified orbital translocation can predict soft tissue change to improve prospective surgical design.

Methods: CT data in 18 hypertelorism patients ($m=11.2$ yrs, $r= 5-20$) were entered into Osirix software. Various parameters from pre/post-operative scans were analysed by two independent observers. Anatomical measurements were palpebral fissure width (PFW), inter-dacryon distance (IDD), inter-canthal distance (ICD), inter-centroid distance (distance between the midpoints of the globe, ICeD), and inter-lateral orbital wall distance (ILoD). Measurements were correlated, and assessed against osteotomy technique (frontofacial bipartition or 'box'), canthopexy details, and 30 controls from a neurosurgical population ($r= 2-17$ yrs).

Results: Palpebral fissure width is unchanged postoperatively, independent of surgeon/technique, providing a 'constant' reference value. Mean preoperative ratio IDD:PFW is 1.41 reducing to 0.94 postoperatively (mean control = 0.64). Mean preoperative ratio ICD:PFW of 1.89 reduces to 1.53 (mean control = 1.16). The ratios ICeD: PFW and ILoD:PFW reduce postoperatively as expected.

The correlation of ratio IDD:PFW to ICD:PFW is 0.89 preoperatively decreasing to 0.77 postoperatively, suggesting that the medial canthus and dacryon move imperfectly together. However, the correlation of IDD:PFW with ICeD:PFW of 0.88 preoperatively is 0.85 postoperatively, suggesting that translocation of the 'effective orbit' predictably translocates the globe. Preoperative IDD:PFW correlation to ILoD:PFW is 0.91 decreasing to 0.78; reflecting movement of the lateral wall through an arc in translocation.

Discussion: Orbital translocation to reduce IDD predictably translocates the globe. Change in ICD:PFW does not reach 'control', however it more highly correlates the amount of midline bone resected than lateral orbital wall movement. These data facilitate surgical planning in HPT phenotypes in which orbital translocation is multiplanar.

16
**OUR THERAPEUTIC EXPERIENCE OF ORBITAL
HYPERTELORISM: A STUDY OF 23 CHINESE CASES**

Presenter: Min Wei, MD
Authors: Wei M, Yuan J, Yu ZY, Xu L
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Orbital Hypertelorism is defined by the increasing of the inter orbital distance (IOD), which always occurs with the midline or paramedian cleft in many craniofacial anomalies. Recently, the main surgical methods to correct the hypertelorism include the U-shaped osteotomy, box osteotomy and facial bipartition. These methods could effectively decrease the IOD. But it is still a challenge that how to acquire a satisfied and stable effect for the nasal deformity correction during the procedure. Since 2005, 23 patients (8 female and 15 male) with hypertelorism have been treated in the craniofacial Unit in Shanghai 9th hospital. 8 cases were moderate and 15 cases were severe among them. The age ranged from 16 to 28 years, with an average age of 18.9 years. 3 cases received U-shaped osteotomy, 18 cases received box osteotomy, 2 cases received facial bipartition. All the patients were undergone the surgical correction. As a key point, the correction of the nasal deformity were well done in the surgical procedure including nasal augmentation with rib bone grafting and reconstruction of the subunit of the nose. We did the tip extension, the columella elongated, the alar cartilage reposition and suspension, etc. All the patients were satisfied with the improvement of the post-operative appearance. Furthermore, followed with 3 to 7 years, most patients still showed favorable and stable results except 1 case. In conclusion, it is important to correct the nasal deformity in the hypertelorism correction surgery. We thought reconstruction of the nasal subunit in early stage could get a stable and favorable results in long-term follow-up.



17
**PROTOCOL MANAGEMENT AND LONG TERM
OUTCOME OF TESSIER CLEFTS 3, 4, 5, 6 AND 7**

Presenter: David J. David, MD, FRCS, FRACS

Authors: David DJ, Flapper WJ
Womens and Childrens Hospital

Of the 458 rare craniofacial clefts on the ACFU database since 1975, the author has reported on 62 cases of Treacher Collins and 258 cases of hemifacial microsomia. Of the residuum, we present 57 clefts affecting the orbits and middle third of the face (Tessier clefts 3, 4, 5, 6 and 7.)

We present the birth to maturity protocol with emphasis on the specific strategies necessary to preserve vital functions in the early post natal period. The need to manage growth and dental development in an appropriate way so that definitive surgery can be best performed at the time of skeletal maturity.

Multiple surgical modalities are often necessary to achieve the intended result at maturity and their integration and timing in the protocol is discussed.

Of the 57 patients, there is a cohort that have undergone management by the same team from birth to maturity. These are presented and their care map demonstrates the health care burden that must be endured by patient surgeon and society to achieve the results.

18
**SHUNT-INDUCED CRANIOSYNOSTOSIS: DIAGNOSIS
AND SURGICAL MANAGEMENT**

Presenter: Matthieu Vinchon, MD, PhD

Authors: Vinchon M, Guerreschi P, Wolber A,
Baroncini M, Pellerin P

Lille University Hospital

Background: Chronic overdrainage caused by cerebrospinal fluid (CSF) shunts in children is a rare cause of microcrania with craniocerebral disproportion. Shunt-induced craniosynostosis (SIC) is difficult to diagnose and its management is debated. We report on our protocol for the surgical management of SIC.

Material and Methods: We reviewed our database which includes 1712 patients shunted before the age of 18, and followed for an average duration of 14.8 years. We selected patients having required cranioplasty for SIC. The diagnosis of SIC was made because of symptoms of raised intracranial pressure without ventriculomegaly, fingerprinting of the calvaria and/or stagnation of the head circumference. Whenever possible, intracranial pressure was monitored for 24-48 hours before deciding to perform a craniotomy. We then performed bifrontal craniotomy and advancement (BFCA) followed by shunt revision with insertion of a flow-regulated valve.

Results: 26 patients developed a SIC requiring BFCA, after a mean 8.4 years of shunting. The cause of hydrocephalus was neonatal hemorrhage and meningitis in 12/26 and 4/26 cases respectively. In all but two cases, the initial valve had been pressure-related. Fifteen patients were explored with intracranial pressure monitoring: the median maximal pressure in this group was 50 mm Hg. The symptoms resolved in all cases after surgery and the cosmetic results were good.

Conclusions: SIC is a rare complication of shunts, which appears almost specific of pressure-related valves. Calvarial augmentation and insertion of a flow-regulated valve appear to be both necessary in order to restore physiological intracranial hydrodynamics.



19

LONG TERM OUTCOMES OF EARLY CORRECTION OF SAGITTAL SYNOSTOSIS VIA ENDOSCOPICALLY ASSISTED CALVARIAL SYNOSTECTOMY AND POST SURGICAL HELMETING

Presenter: Davinder J. Singh, MD
Authors: Singh DJ, Beals SP, Joganic EF, Manwaring K, Bristol R

Barrow Childrens Cleft and Craniofacial Center Phoenix Childrens Hospital

Purpose: Endoscopically assisted synostectomy for treatment of sagittal synostosis still remains controversial, with many authors claiming that the results are suboptimal in comparison to open approaches. The purpose of this paper is to review the surgical protocol, and to present long-term outcomes for this patient group.

Materials and Methods: A retrospective review over a nine year period included 82 patients with sagittal synostosis who were treated via endoscopically assisted synostectomy and post surgical helmeting. Treatment protocol involves endoscopic-assisted strip craniectomy of 4 cm width from anterior to posterior fontanelle followed by 1.5 cm width wedge excisions posterior to coronals and anterior to lambdoid sutures. The patients are cast 7-10 days post-op after reduction of swelling. The orthotic is worn 23 hours a day with weekly adjustments for an average duration of 3.5 months.

Results: To date, 82 patients have undergone this surgery: 56 males and 26 females. The mean surgical age is 3.3 months with range of 2.5 to 7 months. Treatment time with orthotic was 3.5 months with range of 1.25 to 7 months. The mean pre-surgical cephalic index (CI) was 64.8, post-surgical was 69.1, and post banding was 80.2. This represents an increase in CI of 15.4. Follow up mean was 5.7 years, with range of 4 months to 9 years. There was a mean regression in CI of 6.5 at nine years from the post banding CI. Based on photographic assessment, all patients had improvement in the reverse inclination of the forehead plane, frontal bossing, and bitemporal narrowing.

Conclusions: Endoscopic calvarial synostectomy in conjunction with postsurgical helmeting is a valid technique if limited to patients with sagittal synostosis who are under four months of age. The regression in cephalic index noted in long-term follow up is comparable to that documented after open techniques. Patients who undergo this surgical procedure and adhere to the orthotic protocol experience an improved CI and forehead appearance, thus potentially alleviating any need for a second stage procedure as is needed with open approaches.

20

RAISED INTRACRANIAL PRESSURE IN NON SYNDROMIC SAGITTAL CRANIOSYNOSTOSIS FOLLOWING CORRECTIVE SURGERY

Presenter: Greg Thomas, PhD, FRCS (Plast)
Authors: Thomas G, Wall SA, Jayamohan J, Johnson D, Magdum S, Richards PG
Oxford Craniofacial Unit

Estimates of the incidence of raised intracranial pressure (ICP) in patients presenting with non syndromic sagittal craniosynostosis vary widely in the literature. In general, higher rates are reported in later presenting patients, suggesting an aetiological association between untreated sagittal craniosynostosis and intracranial hypertension. Thus one of the goals of corrective surgery must be to reduce the risk of a subsequently raised ICP, although the incidence of this following surgery is rarely reported.

We performed a retrospective review of all patients with a diagnosis of non-syndromic craniosynostosis who underwent primary surgery at the Oxford Craniofacial Unit from 1994 to 2012. In total 292 patients had either a modified strip craniectomy (n=102, mean age 5.8 months) or a more extensive total or subtotal calvarial remodelling procedure (n=190, mean age 27 months). Patients are closely followed up until skeletal maturity. Forty eight patients (16%) presented with symptoms and signs sufficient to warrant intraparenchymal ICP monitoring, at a mean of 52 months (19-111 months) following surgery. ICP was raised in 17 patients (6%), necessitating a calvarial expansion (CE) procedure. Three patients required ICP monitoring post CE of which one was elevated, undergoing a further CE.

Patients who had a modified strip craniectomy were more likely to have post operative pressure monitoring (n=32, 31.3%) and to have raised ICP (n=16, 15.6%) than those treated by a calvarial remodelling procedure (monitoring = 16 (8.4%) [P<0.0001], raised ICP = 1 (0.5%) [P<0.0001]). This relationship remained statistically significant even when only those patients with 4 or more years follow up were considered.

In conclusion, the overall incidence of proven raised ICP post primary surgery was 6%. The incidence of raised ICP was significantly higher following modified strip craniectomy than total or subtotal calvarial remodelling.



21

AGE-BASED APPROACH TO SAGITTAL SYNOSTOSIS: APPLICATION OF EXTENDED STRIP CRANIECTOMY (ESC) AND TOTAL CRANIAL VAULT RESHAPING (TCVR) TECHNIQUES

Presenter: Christopher R. Forrest, MD, MSc, FRCSC, FACS
Authors: Forrest CR
The Hospital for Sick Children

Purpose: Little consensus exists as to the procedure of choice in the management of sagittal synostosis. The purpose of this study was to compare outcomes and complications following a TCVR procedure (> 6 months of age) and ESC with post-operative molding helmet therapy (< 6 months of age) using morphometric CT-based/3D camera analysis.

Method: 129 patients with sagittal synostosis seen at the Hospital for Sick Children, Toronto between 1999-2011 underwent surgical treatment. Due to limitations in application of the cranial index, cranial shape was also assessed by CT-based morphometric analysis or 3D (3dMD, Atlanta, GA) camera analysis. Complications were tabulated.

Results: 84 patients (68M;16F, 5.3 months old) underwent ESC and 45 patients (38M;7F, 26.1 months old) underwent TCVR (mean follow-up of 35.2 ± 11 months). No mortalities were encountered. Surgical cost, length of stay, surgical time, transfusion rates and major complications were greater in the TCVR group ($p < 0.05$). 1 year post-op cranial index was within normal range in both groups. CT-based outcomes analysis demonstrated excellent improvement in sagittal profile at 1-year post-op in both groups. 3D photometry demonstrated pre and post-op changes in cranial form with no differences between surgical technique.

Conclusions: Surgical management of sagittal synostosis should be based upon patient age. ESC is effective for infants 6 months of age and under taking advantage of rapid brain growth and thin bone to allow for physiologic remodeling of the infant calvarium. Vault reshaping techniques are applicable in those over 6 months of age.

22

CAN COGNITIVE FUNCTIONING BE PREDICTED BY PRE-OPERATIVE (CT-SCAN) ANOMALIES IN PATIENTS WITH TRIGONOCEPHALY?

Presenter: Jacques van der Meulen, MD, PhD
Authors: van der Meulen J, van der Vlugt JJ, Diamantopoulou S, Horstman EG, Coebergh RR, Hovius SE, Verhulst FC, Stolwijk A, Lequin M

Erasmus Medical Center

Objectives: The aim of this prospective study was to examine the associations between pre-operative (CT-scan) anomalies (i.e. stenosis of the frontal bone, intracranial volume, brain anomalies) and cognitive functioning in childhood in a sample of patients with trigonocephaly. We expected that severe pre-operative frontal stenosis, small intracranial volume and/or brain anomalies might be associated with poor cognitive functioning in childhood.

Methods: We performed a prospective study in 72 patients with trigonocephaly from the Dutch Craniofacial Center of the Erasmus Medical Center in Rotterdam, the Netherlands. The severity of the frontal stenosis, intracranial volume and the presence of brain anomalies were determined on preoperative CT-scans (mean age at time of CT-scan = 7 months; SD 3.2 months). Intelligence levels were assessed using validated instruments. Mean age at the time of cognitive assessment was 6.0 years (SD 3.0 years).

Results: A significant association was found between the width of the preoperative ventricles and IQ at childhood. Wider ventricles were associated with lower IQ at later age. None of the other CT-scan measurements were associated with IQ.

Conclusion: The findings of this study show that preoperative wider ventricles are associated with lower IQ levels in patients with trigonocephaly. Severe pre-operative frontal stenosis and other brain anomalies are not associated with cognitive problems in childhood as compared to patients with less severe preoperative frontal stenosis or without anomalies.



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EFFECTS OF AGE AT THE TIME OF SURGERY ON LONG-TERM NEUROPSYCHOLOGICAL OUTCOMES IN SAGITTAL SYNOSTOSIS

Presenter: Anup Patel, MD, MBA

Authors: Patel A, Hashim P, Yang J, Bridgett D, Losee J, Duncan C, Jane J, Persing JA

Yale University School of Medicine

Background: The relationship between age at the time of surgery and long-term neuropsychological (NP) outcomes in sagittal craniosynostosis (SGC) remains equivocal. (1-3) In current clinical practice, whole-vault cranioplasty (WVC) is performed for a wide age range of individuals with SGC as no consensus exists on the optimal timing of surgery. This study examines the relationship between age at the time of WVC and long-term NP function using comprehensive, longitudinal neurological testing.

Methods: This multi-institutional study consisted of 72 surgically treated SGC patients. The subset of patients who underwent WVC (n=42) was subdivided by age at the time of surgery: younger than 6 months (n=20) vs. older than 6 months (n=22). All patients underwent a battery of neurodevelopmental tests evaluating various domains of NP function (Beery VMI, WASI, and Wechsler Fundamentals).

Results: Patients treated after 6 months of age demonstrated poorer visual-motor integration than those treated before 6 months ($p<0.05$). Individuals treated after 6 months also scored lower on Verbal IQ and Performance IQ ($p<0.05$). In addition, those who were treated at later ages demonstrated poorer accomplishment in Word Reading, Reading Comprehension, Spelling, and Numerical Operations ($p<0.05$). All analyses statistically controlled for full-scale IQ, with the exception of analyses comparing subgroups on IQ variables.

Conclusion: These results demonstrate that patients who underwent WVC before 6 months have significantly better cognitive outcomes compared to those who underwent surgery at later ages. This data strongly suggests that surgical intervention prior to six months of age imparts greater long-term NP benefits than delayed treatment.

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ANAESTHESIA IN THE PRONE POSITION CAN SAFELY BE USED TO FACILITATE THE COMPLEX SURGICAL CORRECTION OF CRANIOSYNOSTOSIS. A SERIES OF 300 CONSECUTIVE PATIENTS IN 15 YEARS IN OXFORD, UK

Presenter: Russell G. Evans, MBBS FRCA

Authors: Evans RG, Das S, Johnson D, Wall S
The Oxford University Hospitals NHS Trust

Prone positioning allows improved surgical access for calvarial remodelling in patients with craniosynostosis. Complications and hazards related to prone positioning have been reported. We have reviewed the intraoperative records of 300 paediatric patients undergoing trans cranial surgery in the prone position over the past 15 years in our institution. Anaesthesia charts and operative notes were reviewed by two Consultant Paediatric Craniofacial Anaesthetists in 300 patients having calvarial remodelling in the prone position. Patient demographics, use of extended sphinx position and intraoperative complications were recorded. Overall intraoperative complications related to patient position were recorded and subdivided into airway/ventilation (1%), vascular access/monitoring (1%) and tissue pressure damage (1%) with no long term sequelae reported. 30% of patients were placed in the extended sphinx position compared to standard prone position with no significant difference in complications between the two positions. The aim of this review is to demonstrate that whilst there are complications related to placing these patients in the prone position it is a safe technique to use in order to allow enhanced surgical access for calvarial remodelling in the paediatric craniofacial patient.



25
**MATERNAL-FETAL COMPLICATIONS IN
CRANIOSYNOSTOSIS**

Presenter: Jordan Swanson, MD
Authors: Oppenheimer AJ, Swanson J, Al-Mufarrej F,
Pet M, Saltzman B, Gruss J, Hopper R,
Cunningham M, Birgfeld CB

Seattle Childrens Hospital

Background: Teleologically, the cranial sutures facilitate deformation of the neonatal skull, allowing passage through the birth canal. Premature fusion of these sutures may preclude the normal birthing process. The purpose of this study is to determine the incidence of puerperal maternal-fetal complications in cases of craniosynostosis. Our global hypothesis is that craniosynostosis incurs an increased risk of unplanned cesarean section and neonatal birth trauma due to cephalo-pelvic disproportion.

Materials & Methods: Following institutional IRB approval, a retrospective chart review was performed at Seattle Childrens Hospital for 300 children with a diagnosis of craniosynostosis. Intake records were reviewed, with particular attention directed to birth history and birth-related complications. The incidence of unplanned cesarean section in primiparous women with standard-risk pregnancy was obtained from the literature (5-10%) for comparison.

Results: Two-hundred and eighty (280) complete records were obtained for non-syndromic craniosynostosis cases. Unplanned cesarean sections were carried out on 66 women (24%) for failure to progress in labor; no difference in cesarean section rates were observed based on suture phenotype. One cephalohematoma and 5 subgaleal hematomas were recorded. Among 102 primiparous women, 39 (38%) required unplanned cesarean section for failure to progress in labor.

Conclusion: Women carrying children with craniosynostosis have an increased risk of unplanned cesarean section; primiparous women, in particular, may have 4- to 8-fold higher risk of unplanned cesarean section when compared to rates from the literature. Cephalo-pelvic disproportion may mediate this association. Based on this information, a prenatal diagnosis of craniosynostosis could influence decision-making in the management of labor.

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**RAISED INTRACRANIAL PRESSURE IN UNTREATED
NON SYNDROMIC SAGITTAL CRANIOSYNOSTOSIS:
AN EIGHTEEN YEAR EXPERIENCE IN OXFORD**

Presenter: Steve A. Wall, MB BCh, FRCS, FRCPCH,
FCS(SA)
Authors: Wall SA, Thomas GP, Byren J, Jayamohan J,
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Oxford Craniofacial Unit

Patients presenting with non syndromic sagittal craniosynostosis (SC) beyond an age suitable for strip craniectomy under the Oxford Craniofacial Unit protocol (developed over 18 years) are routinely offered formal intracranial pressure (ICP) monitoring when there is insufficient justification for surgery on morphological grounds alone or in any case where, despite morphological indications, there is a parental reluctance for surgical intervention.

From 1994 to 2012 318 children with SC presented for assessment. Two hundred and fourteen cases presented too late for strip craniectomy (n=104). On the basis of our protocol 49 patients underwent initial ICP monitoring. Of these 19 had confirmed raised ICP (39%). By extrapolation this represents a 9% minimum risk of raised ICP in later presenting patients alone and 6% of the total SC population treated over the period. Whilst it cannot be claimed that 39% of all untreated cases will develop raised ICP, this finding indicates that the risk is substantially greater than figures currently quoted in the craniofacial literature.

There was no significant correlation between the severity of scaphocephaly and the incidence of raised ICP.

We recommend that ICP monitoring should be strongly considered for all patients with SC in whom a conservative management path is considered, or where additional validation of the indications for surgery is required.



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MINIMALLY INVASIVE SPRING-ASSISTED CORRECTION OF SAGITTAL SUTURE SYNOSTOSIS

Presenter: MarieLise C. Van Veelen, MD
Authors: Van Veelen MC, Touw C, Mathijssen IM
Erasmus University Medical Center Rotterdam

The technique, results and advantages and limitations of minimally invasive spring-assisted cranioplasty for sagittal suture synostosis are presented.

Technique: Access is gained by two small incisions perpendicular to the sagittal suture. These incisions provide sufficient access to perform two parasagittal craniotomies and place two distractive springs. Springs are manufactured from 1.22 mm medical steel wire, are 8.9 cm wide and generate 9 Newton of distractive force.

Results: We analysed the results of the first 45 patients treated with this technique at our institution, between January 2010 and June 2012. The cephalic index (CI) at presentation was 67 (sd 3.9), after surgery CI increased to 75 (sd 4.3), at one year after surgery CI dropped to 74 one year, after that CI remained stable at 74 (sd 3.3). Mean blood loss was 78 ml (sd 83). One patient developed a dural tear, one patient had insufficient distraction.

Conclusion: Spring-assisted cranioplasty requires only two small incisions and is at least as effective as other techniques with respect to the cephalic index. Blood loss, operative time and complication rate are reduced. The most important disadvantage is the need to remove the springs in a second intervention. A second drawback is the fact that expansion of the spring is not controllable after placement. This can be partially intercepted by adjusting the spring (or the craniotomy) to the patients specific features.

28

THE ASSOCIATION OF CHIARI MALFORMATION TYPE I AND CLOSURE OF SAGITTAL SUTURE.

Presenter: Federico Di Rocco, MD PhD
Authors: Di Rocco F, Chivoret N, Puget S, Zerah M, Sainte-Rose C, Arnaud E

Craniofacial Unit

Background: The association between Chiari Malformation type I (CM1) and non-syndromic craniosynostosis is reported in the literature (4.4% - 9.1%) especially concerning the sagittal suture. However, the real incidence of the association of CM1 and early closure of the sagittal suture are still debated.

Methods: To study the association of CM1 and sagittal suture closure, we realised:

- 1 - a 15-years retrospective review of 950 consecutive surgically corrected scaphocephalic patients;
- 2 - a 5-years retrospective review of 92 consecutive operated on CM1 patients;
- 3 - a review of 100 TDMs controls.

The TDMs reconstructions were analysed to define: the presence/absence of sagittal suture, the cephalic index (CI), the measurement of Foramen Magnum, in particular its width (FMw), its antero-postero length (FMA-p) and its cross-sectional area (FMsurf).

Results: The measurement could be obtained in all scaphocephalic patients and in 31/92 CM1 patients (mainly studied by craniovertebral junction MRIs). We found that:

- 1- 3/950 (0.3%) of scaphocephalic patients present with a CM1 at diagnosis (mean age 3 months);
- 2- 8/31 (25.8%) of symptomatic CM1 patients present a closure of sagittal suture
- 3- 3/100 (3%) of controls showed the absence of sagittal suture, none had a CM1.

In particular, we compare the isolated CM1 group and the CM1 patients presenting a closure of sagittal suture group and we report: tonsillar herniation degree (12.1 mm vs 15.3mm), mean age (85.7 vs 83.3 months), mean CI (78.4 vs 75.8), presence of syrinx (39.1% vs 50%), bone anomalies/syndrome associated (17.4% vs 37.5%), measurement of FM (FMw 27.3 mm vs 28.18mm-FMap 28.98mm vs 30.32 mm-FMsurf 515.16mm² vs 600.79mm²) and presence of Hydrocephalus (17.4% vs 12.5%)

Discussion: CM1 associated to the fusion of the sagittal suture seems to be a different nosological entity that a "classical" scaphocephaly (with an early fusion diagnosed in infancy) and isolated symptomatic CM1. This distinction highlights the importance of complete workout in patient presenting with CM1.



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**COMPARISON OF SPRING CRANIOPLASTY AND PI
PROCEDURE FOR SAGGITAL SYNOSTOSIS**

Presenter: Matthew Hansen, MBBS, FRACS

Authors: Hansen M, Hewitt T

Princess Margaret Hospital for Children

Spring assisted cranioplasty has been used for the treatment of selected cases of saggital craniosynostosis in our unit since 2007. It has replaced our previous technique of Pi -plasty in the management of early presenting scaphocephaly. The aim of this study was to compare the results of the two approaches. To do this we did a retrospective review of the first 30 patients who underwent spring assisted cranioplasty for saggital craniosynostosis. We compared this to the first 30 patients who underwent the Pi procedure for the same condition. Data was collected for demographics, operative time, length of hospital stay, length of ICU stay, transfusion requirements, post operative morbidity, complications and outcome. We found the spring group was superior to the Pi group with respect to length of stay both hospital and ICU, operative time and need for transfusion. Subjective assesment of outcome has necessitated one revision of Pi for undercorrection but no need for revision of any of the spring group. We conclude that the spring assisted cranioplasty delivers at least equivalent correction of scaphocephaly when compared to the Pi group but with significantly less surgical, ICU and hospital time and with much less transfusion requirement.

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**ORGAN LEVEL TISSUE ENGINEERING USING
NATIVE DECELLULARIZED MICROCIRCULATORY
CONSTRUCTS AS VASCULARIZED BIOSCAFFOLDS**

Presenter: Lars H. Evers, MD

Authors: Evers LH, Findlay MW, Liu W, Simons D,
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Stanford University

WITHDRAWN



31
SUSTAINED, LOW-DOSE RHBMP-2 DELIVERY VIA PLGA MICROSPHERES PROVIDES FOR EQUIVALENT OSTEOGENESIS AND IMPROVED SIDE-EFFECT PROFILE

Presenter: Jason Wink, BA
Authors: Wink JD, Gerety PA, Sherif R, McGrath JL, Clarke NA, Rajapske C, Nah HD, Taylor JA

Childrens Hospital of Philadelphia

Background: The use of recombinant human bone morphogenetic protein 2(rhBMP2) is limited in the craniofacial region due to dose-dependent side effects-namely heterotopic ossification(HO), craniosynostosis, and seroma- when delivered in high doses on a collagen sponge at a single time point. Our aim is to investigate a novel, low-dose, sustained-delivery system for rhBMP2 utilizing poly-lactic-co-glycolic acid (PLGA) microspheres in a rabbit cranial model.

Methods: 17 adult New Zealand white rabbits underwent bilateral 10mm craniectomy. Defects were loaded with a collagen scaffold containing one of the following: free rhBMP2 (1ug, 0.1ug, 30ug), PLGA-rhBMP2 (0.1ug), with and without allogeneic bone marrow derived mesenchymal stem cells(BM-MSC). Appropriate positive and negative controls were performed. Bone volume and surface area (SA) were assessed using microCT. Finite element analysis (FEA) was performed on a region of interest within each defect site and native calvarium to assess the mechanical competence of bone. Specimens were evaluated qualitatively with H&E and trichrome staining.

Results: In acellular groups, PLGA-rhBMP2 resulted in higher volumes of new bone than an equal dose of free rhBMP2 ($p=0.049$). Significant variability was found in our experiments containing BM-MSCs, and PLGA-rhBMP2 groups without BM-MSCs yielded more bone than their acellular counterpart ($p=0.034$). HO and craniosynostosis were observed only in the 30ug free rhBMP2/implant group. New bone in all groups formed in a woven fashion with disorganized trabeculae and the presence of neovascularization. FEA indicated that the mechanical competence measured using the regional elastic modulus did not differ between groups with or without exposure to rhBMP2 ($p=0.89$).

Conclusions: Sustained, low-dose rhBMP2 delivery via PLGA microspheres provides for equivalent osteogenesis as high-dose, free rhBMP2, but with an improved side-effect profile. Future work will focus on optimal dosing and scaffold delivery of PLGA microsphere delivery of rhBMP2.

32
BIOPRINTING TISSUE ENGINEERED INTRAMEMBRANOUS BONE CONSTRUCTS

Presenter: Darren M. Smith, MD
Authors: Smith DM, Shakir S, Naran S, Campbell P, Losee JE, Cooper GM
University of Pittsburgh Medical Center

Purpose: Pediatric craniofacial reconstruction is complicated by a limited supply of bone. Tissue engineering is a potentially valuable strategy in this regard. Many promising results have involved BMP-2-generated endochondral bone. Most calvarial bone, however, is intramembranous in origin. We hypothesize bioprinting precise doses and patterns of proteins known to play a key role in intramembranous ossification can more efficiently generate calvarial bone by favoring intramembranous rather than endochondral osteogenesis with lower (safer) doses of potent morphogens, including BMP-2 and other members of the TGF-B superfamily.

Methods: Cranial defects in a mature murine model were filled with a bioprinted acellular dermal matrix (ADM) disc. Each half of each ADM disc was bioprinted with 1 of 4 combinations of BMP-2 and TGF-B1 (Treatment 1: 0 overprints (OP) BMP-2/30 OP TGF-B1; Treatment 2: 30 OP BMP-2/50 OP TGF-B1; Treatment 3: 50 OP BMP-2/0 OP TGF-B1; Treatment 4: 50 OP BMP-2/30 OP TGF-B1). Histologic and radiographic analyses were performed after 4 weeks.

Results: The two treatment groups with less than 50 OP BMP-2 (Treatment 1 and 2) demonstrated similarly weak osteogenesis (average 17.3% and 14.3%). In contrast, both treatment groups with 50 OP BMP-2 (Treatment 3 and 4) demonstrated meaningful and comparable osteogenesis (82.1% and 77.3%). H&E staining showed that bone generated in response to Treatment 4 (50 OP BMP-2/30 OP TGF-B1) demonstrated a more compact and robust lamellar pattern than that generated in response to Treatment 3 (50 OP BMP-2/0 OP TGF-B1).

Conclusions: BMP-2 confers a clear osteogenic advantage at 50 OP. While TGF-B1 does not lead to more osteogenesis at the doses and concentrations employed in this series of experiments, it does seem to affect the quality of the bone generated. Bone generated in response to Treatment 4 appeared more compact, lamellar, and indeed more mature at 4 weeks than that generated in response to Treatment 3. Taken together, these findings may imply that TGF-B1 accelerates or enhances osteogenesis with a certain threshold dose of BMP-2.



33 IN VIVO DIRECTED DIFFERENTIATION OF HUMAN EMBRYONIC-LIKE PLURIPOTENT CELLS INTO BONE

Presenter: Derrick Wan, MD
Authors: Chung MT, Paik KJ, Levi B, Montoro DT,
Lo DD, Sun N, Wu JC, Longaker MT,
Wan DC

Stanford University

Introduction: Pluripotent embryonic stem (ES) cells have shown promise for bone tissue engineering, however, these cells raise ethical concerns and form teratomas when placed in vivo. To mitigate these concerns, we set out to derive induced pluripotent stem cells (iPSCs) and directly differentiate them down an osteogenic lineage in vivo without pre-differentiation.

Methods: iPSCs were derived from hASCs using the Yamanaka factors and characterized for pluripotency. In vitro bone differentiation was confirmed through q-PCR, western blotting, and immunocytochemistry. Next, iPSCs were seeded on an osteogenic microniche consisting of a hydroxyapatite coated BMP-2 releasing scaffold and implanted into a 4 mm critical size calvarial defect. Implanted cell viability and calvarial healing was followed through bioluminescence, microCT, and histology.

Results: We demonstrate that iPSCs undergo osteogenic differentiation in vitro, as shown by numerous gene markers and alizarin red staining. We also demonstrate that iPSCs seeded on an osteoinductive scaffold survive and participate in de novo bone formation. Finally, the rate of teratoma formation was reduced using our osteoinductive scaffold.

Conclusion: These data support that an osteogenic microniche scaffold placed into a macro-environmental niche of a skeletal defect promotes in vivo-directed iPSC osteogenic differentiation without teratoma formation. We believe this is the first example of directed lineage differentiation of pluripotent cells in vivo.

34 PREDICTIVE GENOMICS: VALIDATION OF A ROLE FOR MIRNA IN THE UNDERDEVELOPMENT OF THE MANDIBLE AND OTHER CRANIOFACIAL STRUCTURES

Presenter: Christopher Runyan, MD, PhD
Authors: Uribe-Rivera A, Aronow BJ, Billmire DA,
Reyna-Rodriguez PX, Gordon CB

Cincinnati Childrens Hospital Medical Center

Background and Purpose: MicroRNAs (miRs) are short, non-protein-coding RNAs that control gene regulation by targeting messenger RNAs. They are found in highly conserved clusters within groups of known cranial morphogens such as HOX genes, and have been implicated in maintenance of neural crest and skeletal morphogenesis. They affect expression of groups of downstream targets, rather than regulate single gene transcripts. We hypothesized that miRs may be regulators of families of genes implicated in sporadic human craniofacial disorders such as hemifacial microsomia and Pierre Robin sequence which anatomical structure such as mandible is underdeveloped.

Methods: Using a systems biology approach we identified miRs that could result in craniofacial syndromes. We identified families of known craniofacial genes with potential miR binding sites in their 3'UTRs. These miRs were then validated in zebrafish embryos for craniofacial expression using LNA in situ hybridization and miR microarrays. Of these, the member of the miR23 and miR27 family, such as miR23ab and miR27ab family's gene targets overlapped with gene networks known to produce phenotypes found in hemifacial microsomia and Pierre Robin sequence, respectively. Antagomirs to miR 23 and miR27 were injected into zebrafish embryos.

Results: Approximately 1/3 (85%) of miR23 treated fish embryos had asymmetric micrognathia, cleft palate, fusion of otoliths and midfacial asymmetry/hypoplasia. Similarly, 1/3 (87%) of the miR27 group demonstrated profound micrognathia and variably severe midfacial/cranial hypoplasia, and facial clefting. In several, there was mandibular agenesis. We could rescue the phenotype of the injected embryos for both miR23ab and miR27ab with separately microinjection of the antagomirs and mimic MO. In situ hybridization and immunohistochemistry analysis implicate a role for miR in dHAND and endothelin signaling.

Conclusions: In summary, we have predicted a role for miRNA as master regulators of craniofacial development and blocked their expression to duplicate the human phenotype.



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SHOULD TRANS-GENDER FACIAL SKELETAL DONATION BE CONSIDERED FOR CRANIOMAXILLOFACIAL TRANSPLANTATION? A CADAVER STUDY

Presenter: Gabriel F. Santiago, MD
Authors: Alrakan MA, Swanson EW, Susarla S, Santiago GF, Obrien D, Grant GT, Liacouras P, Armand M, Murphy R, Brandacher G, Andrew Lee WP, Gordon CR

Johns Hopkins University and School of Medicine

Background: Donor-recipient gender mismatch is considered a contraindication to facial transplantation. Gender-specific anthropometrics, skin texture/adnexae mismatch, and social apprehension have prevented transgender facial transplantation from evolving. However, the scarce donor pool and extreme waitlist times are currently suboptimal. Our objective was to: 1) perform and assess cadaveric facial transplantation for each gender-mismatch scenario, 2) review the advantages/disadvantages of transgender facial transplantation, and 3) to raise awareness of its potential benefits.

Methods: Transgender facial transplantation feasibility was evaluated through two mock, double-jaw, Le Fort-based cadaveric allotransplants, including female donor-to-male recipient (T1-FM) and male donor-to-female recipient (T2-MF). Hybrid facial-skeletal relationships were investigated using cephalometric measurements, including sellion-nasion-A point (SNA) and sellion-nasion-B point (SNB) angles, and lower-anterior-facial-height to total-anterior-facial-height ratio (LAFH/TAFH).

Results: Skeletal proportions and facial-aesthetic harmony of the cadaveric transplants [n=2] were found to be equivalent to all reported experimental/clinical gender-matched cases by using custom cutting guides. Cephalometric measurements are shown in Table 1 relative to Eastman Normal Values.

Conclusions: Based on our results, we believe that transgender facial transplantation can offer equivalent outcomes to those of gender-matched pairs. Lack of literature discussion of transgender facial transplantation highlights the general stigmata encompassing the subject. We hypothesize that concerns over gender-specific anthropometrics, skin texture/adnexae disparity, and increased immunological resistance have prevented full acceptance thus far. Advantages of transgender facial transplantation include an increased donor pool with expedited reconstruction, as well as size-matched donors. Appropriately size-matching donor-recipient pairs for skeletal dimensions should take precedence over gender-matching.

36
TRANSLATIONAL CADAVERIC STUDY OF COMPOSITE EYE AND PERIORBITAL ALLOTRANSPLANTATION FLAP FROM RAT MODEL

Presenter: Fatih Zor, MD
Authors: Bozkurt M, Zor F, Uygur S, Kulahci Y, Ozturk C, Djohan R, Siemionow M, Papay F
Gulhane Military Medical Academy

Composite tissue allotransplantation (CTA) is a novel alternative for reconstruction of severe periorbital defects with functional recovery which can offer a new hope for restoring vision to these patients. The aim of this study is to describe a composite tissue allotransplantation model of eyeball and periorbital tissues on cadavers. Study is performed on 5 fresh cadavers. Skin island of the composite eyeball and periorbital allotransplantation flap was planned. Borders of the flap were infraorbital rim at inferior, nasal dorsum at medial, eyebrow at superior and lateral orbital rim at lateral. External carotid artery was found with anterior neck incision. The pedicle of the skin island of the flap was included facial artery and superficial temporal artery and external jugular vein. The skin and subcutaneous tissues of the periorbital region were incised according to the planned skin island and bony tissue was reached. Later, a coronal incision was performed and frontal bone was exposed. Following a frontal osteotomy, frontal lobe of the brain was reached. Frontal lobe was retracted exposing internal jugular vein and ophthalmic artery which is a branch of internal carotid artery. Ophthalmic nerve and oculomotor nerve were also dissected and included in the flap. Finally, a “box osteotomy” was performed to the orbit and the dissection was ended. Indocyanin was injected from the pedicle of the flap. The perfusion of the flap was confirmed with SPY Elite System to identify perfusion zones and to visualize micro-surgical arterial inflow to identify flap design and perfusion. Composite tissue allotransplantation model including eyeball and periorbital tissues was described for the first time. Perfusion of the flap was confirmed showing this transplantation is possible in the future. Although harvesting of this flap is difficult to perform, it constitutes a new alternative for reconstruction of periorbital region.



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FGFR2C(C342Y/+) MOUSE MODEL OF CROUZON SYNDROME TO STUDY CRANIOSYNOSTOSIS

Presenter: Xianxian Yang, MD PhD
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Background: Crouzon syndrome patients exhibit premature fusion of coronal sutures resulting in craniosynostosis during early childhood. A previous microarray study identified a number of genes that were downregulated during this process, and the *Fgfr2cC342Y/+* Crouzon mouse model was chosen to further explore the roles of these genes during suture fusion.

Methods: The onset of cranial suture fusion in *Fgfr2cC342Y/+* mice was assessed from E16.5 to P10 using microCT and histology. Expression levels of target genes and osteogenic markers during coronal suture fusion were determined using RT-qPCR. The proliferation rates of coronal suture-derived and parietal bone-derived cell cultures from wildtype and *Fgfr2cC342Y/+* mice were assessed by DNA assay.

Results: *Fgfr2cC342Y/+* mice exhibited coronal suture fusion from E18.5 with complete fusion by P10, while wildtype remained patent. During coronal suture fusion the *Gpc3* and *C1qtnf3* genes were downregulated in *Fgfr2cC342Y/+* mice compared to wildtype, mimicking our human data. A significant increase in coronal suture cell proliferation in mutant mice was observed but no change was observed for parietal bone cells.

Conclusions: Coronal suture fusion occurs embryonically and may relate to an increase in suture cell proliferation.

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FURTHER DEFINING THE ROLE OF RANK/RANKL/OPG AXIS IN CRANIAL SUTURE BIOLOGY

Presenter: Maureen R. Beederman, BA
Authors: Beederman MR, Lee JC, Rajan M, Rogers MR, Kim S, He TC, Reid RR

The University of Chicago

Introduction: Craniosynostosis (CS), or the premature fusion of cranial sutures, is a condition that affects approximately 1 in 2,500 live births worldwide. Much research on this topic has shown that osteoblasts are implicated in the complex mechanisms that lead to early suture fusion. However, relatively little is known about the role of osteoclasts in this process. Osteoclasts are regulated by TNF- α superfamily members, receptor activator of NF- κ B (RANK) and RANK ligand (RANKL), as well as osteoprotegerin (OPG), a soluble inhibitor of RANK. Past work from our lab suggests that the RANK-RANKL-OPG pathway plays a key role in patients with CS. Our work further examines the role of OPG in this process using knockout technology.

Methods: Animal and human studies were approved by the University of Chicago Animal Care and Use Committee and the University of Chicago Institutional Review Board, respectively. Fused and patent human suture samples were collected during cranial reconstruction surgery and were analyzed for RANK and OPG protein expression using immunohistochemistry and Western blotting (WB) techniques. To further define the role of OPG in suture homeostasis, wild-type, OPG $^{+/-}$, and OPG $^{-/-}$ (KO) mice were bred and imaged by serial microCT scans at 3, 5, 7, 9, and 12 weeks. Posterofrontal suture density measurements were performed at these time points.

Results: WB results show that OPG protein expression is increased in the human fused sutures compared to patent control samples. Microwestern array data yields quantitative assessment of protein expression of these samples. Preliminary data from OPG KO mice suggest that perturbations in osteoclastogenesis (in this case, upregulation of osteoclast activity), lead to alterations in cranial and suture morphology. MicroCT analysis of the posterofrontal suture indicates a decrease in bone density in these KO mice.

Conclusions: OPG, the soluble inhibitor of the RANK-RANKL signaling pathway, appears to aid in regulation of suture patency and cranial morphology in the mouse. Further studies focusing on osteoclast biology in diseased and patent sutures are warranted.



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ANALYSIS OF THE AGING OF THE NASAL BONE AND NASAL PYRAMID STRUCTURES IN A CAUCASIAN POPULATION

Presenter: Can Ozturk, MD
Authors: Ozturk CN, Bozkurt M, Uygur HS, Sullivan TB, Djohan R, Papay F

Cleveland Clinic

Purpose: The aim of this study was to compare various nasal bone and nasal pyramid morphometric measurements between different age groups of caucasians and to analyze the effect of aging over these parameters.

Methods: Ninety caucasian skulls were used for morphometric analysis. The specimens were grouped according to three age groups: Group I (20-40), Group II (41-64) and Group III (> 65). Each group consisted of 15 male and 15 female skulls. The length and width of each nasal bones, nasion (N) to anterior nasal spine (ANS), ANS to supradentale (SD), height and width of pyriform aperture, pyriform aperture angle and circumference, relation between septum and inferior nasal concha (turbinate) were measured by using a high precision measurement tool, the MicroScribe®, a ruler and goniometer. Three age groups were compared, male and female separately, as well as symmetry were compared between left and right sides.

Results: The width of pyriform aperture increased significantly ($p=0.024$) with increasing age (Group III versus Group I) in males. Besides this, the angle of pyriform aperture significantly ($p=0.032$) increased (Group II versus Group I) in males. Additionally, the vertical distance from nasion to ANS and angle of pyriform aperture increased significantly with increasing age ($p=0.045$ - Group III versus Group I and $p=0.014$ - Group II, III versus Group I, respectively) in both genders. When the relationship between pyriform aperture width and height was analyzed, there was no statistically significant result. However, the correlation of increase distance between nasion to ANS and ANS distance to SD was found statistically significant ($p=0.004$). When symmetry was analyzed, the left nasal bone distal length was found to be significantly larger than on the right side in both genders ($p=0.044$).

Conclusions: The morphologic measurements and relationships of nasal bones, nasal pyramide and pyriform apertures were found to demonstrate changes in different age groups. These skeletal changes in different age groups should be considered when performing nasal airway and aesthetic surgeries.

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EFFECTS OF NASAL ALVEOLAR MOLDING (NAM) ON INFANT WEIGHT GAIN AND TIMING OF PRIMARY BILATERAL CLEFT LIP REPAIR

Presenter: Michael R. Pharaon, MD
Authors: Clayton J, Krochmal DJ, van Aalst JA
The University of North Carolina at Chapel Hill

Background: NAM proponents report the following benefits in patients with bilateral cleft lip and palate (BCLP): retroclining of the pre-maxillary segment, lateral segment alignment, nasal contouring, and columellar lengthening. Proponents also suggest an improvement in oral intake similar to that obtained using a feeding obturator. However, little objective data exists regarding the effects of NAM on infant feeding. Because patients with BCLP are generally more likely to have feeding difficulties, we elected to study the impact of NAM on infant weight gain and timing for primary lip repair in a cohort of patients with BCLP.

Methods: We performed a retrospective chart review ($n=77$) based on hospital administrative data (CPT code 40701) during a 7-year period from 2005-2012. Inclusion criteria included patients with BCLP with lip repair performed at a single intuition. Exclusion criteria included, lip repair after 1 year-of-age, and use of TPN or feeding tubes. Data included gestational age, age at time of primary lip repair, and weight. Corrections for age were made based on gestational age. All data are reported as a mean \pm standard deviation. Statistical comparisons were generated using a Mann-Whitney two sided t-test. Statistical significance was set at a p value < 0.05

Results: Fifty-six patients met the inclusion criteria, of which 6 patients underwent NAM. The age in days for primary cleft lip repair in NAM patients was 124.2 ± 31.5 ; the non-NAM group was 120 ± 50.5 ($p = 0.4996$). The weight (kg) of the patient at time of surgery in the NAM group was 6.19 ± 0.86 and in the non-NAM group was 5.58 ± 1.24 ($p = 0.0941$).

Conclusion: This study attempts to explore the relationship between the selective use of NAM, pre-operative weight gain, and timing of primary lip repair in patients with BCLP. We did not detect a statistically significant difference in the groups compared. This data suggests that infants with NAM and non-NAM were similar in weight gain and timing to lip repair.



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EARLY CORRECTION OF THE MANDIBLE WITH UNI-VECTOR DISTRACTION OSTEOGENESIS IN HEMIFACIAL MICRO SOMIA PATIENTS: ORTHODONTIC TECHNIQUE FOR PREVENTION OF MALOCCLUSION AND OPTIMIZING SURGICAL OUTCOME

Presenter: Patricia Glick, DMD
Authors: Glick P, Singh DJ
Barrow Childrens Cleft and Craniofacial Center Phoenix Childrens Hospital

Purpose: Early mandibular surgery with distraction osteogenesis in patients with hemifacial microsomia has received criticism for creating a malocclusion and resulting in long term relapse. Proponents of early surgery believe that distraction osteogenesis can be carried out successfully without generating a malocclusion, and that by leveling the maxillary occlusal plane, the mandibular lengthening is more stable. The purpose of this paper is to present an orthodontic technique that needs to be implemented in conjunction with mandibular distraction with the aim of preventing a malocclusion and leveling the maxillary occlusal plane.

Materials and Methods: Patients have orthodontic hardware placed several days prior to surgery. Mandibular corticotomy and placement of a uni-vector semi buried device is performed. Activation of the device is initiated on post operative day 5. Elastic traction is applied to previously placed orthodontic hardware as soon as active distraction ends. The rigidity of the titanium rod of the distractor protects the newly formed osteoid from resorption. Elastic traction is placed from the lingual of the maxillary arch to the buccal of the mandibular arch. Primary or permanent teeth can be utilized. This configuration is used to avoid the narrowing of the maxillary arch that often accompanies inter-arch elastic use.

Results: To date, this technique has been employed in six patients. The average age of the patient is 6.3 years, the average duration of activation is 30 days, and the average duration of consolidation is 60 days. With good cooperation, closure of the open bite often occurs in three months.

Conclusions: The technique of closing the lateral and posterior open bite in the recently distracted hemifacial microsomia patient is efficient and easily managed by the family. This not only accomplishes and maintains good occlusion for the child, but also aids in maintaining stability for the mandibular lengthening.

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FACIAL BIPARTITION; THE ORTHODONTIC BURDEN OF CARE

Presenter: Daljit Gill, MSc, FDS, RCS(Orth)
Authors: Gill D, Dunaway DJ, Britto JA, Evans RD
Great Ormond Street Hospital for Children

The surgical correction of hypertelorism may be carried out using either the facial bipartition (FB) or box osteotomies. The FB, in this unit, is regarded as being more suitable for the deformity associated with Apert syndrome and has shown to be an effective procedure (Greig et al 2013). Our current practice is to create space between the central incisors using either full or sectional arch fixed appliances to facilitate the midline osteotomy and reduce the dental morbidity to a minimum. Whilst the FB may be considered for patients with hypertelorism associated with other syndromes e.g. craniofrontonasal dysplasia, the orthodontic management of the dentition needs to be considered very carefully in terms of the burden of care and iatrogenic damage. Pre- and post- surgical orthodontic treatment, on what are frequently well aligned dental arches with no significant occlusal discrepancies, can take up to 48 months. The post surgical phase is most challenging as treatment is essentially aimed at returning the occlusion to the pre surgical position whilst affording the opportunity to fully correct any alignment or occlusal issues. Closing the midline space requires careful skeletal and dental control and the possibility of cancellous bone grafts cannot be discounted. Whilst all this is possible the frequency of iatrogenic damage i.e. decalcification, caries, gingival inflammation and root resorption increase the longer the patients are in treatment. Our preferred strategy is to factor in the orthodontic considerations during the surgical planning process, so as not to compromise the health and long term prognosis of the dentition. Cases will be presented to illustrate our approach. Greig et al Correcting the typical Apert face: combining bipartition with monobloc distraction. *Plast Reconstr Surg* 2013; 131:219e



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**PREOPERATIVE SIMULATION AND PREDICTABILITY
FOR THE FACIAL ASYMMETRY OF HEMIFACIAL
MICROSOMIA PRUZANSKY GRADE I**

Presenter: Suguru Kondo, PhD, DDS
Authors: Kondo S, Okumoto T, Imamura M,
Yoshimura Y

*Cleft Lip and Palate Center Fujita Health University School of
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Objective: Hemifacial microsomia is a congenital disorder in which hypoplasia of bones and soft tissues derived from the first and second brachial arches result in facial asymmetry. In the past, we have proposed and recommended the treatments with full consideration to the degree of mandibular deformities and postoperative control of the occlusion. We hereby present our method of preoperative simulation and outcomes of the treatments in Pruzansky grade I Hemifacial microsomia.

Methods: We retrospectively selected two cases with one-stage elongation procedure for mandibular hypoplasia. We constructed a real-scale model using preoperative computed tomography three-dimensional reconstruction data and underwent preoperative simulation. Care was taken not to cause lateral cross bite of the normal side while lowering the underdeveloped side of the mandibular molars. Precisely fit bite block was then created using an occlusion device, and applied to the maxilla after vertical osteotomy of the affected side of the mandible, just before its bone plate fixation, and the mandibular position was then guided by the bite block. Lengthening of the mandible and bone fixation were carried out to the most three-dimensionally fit position. One month after the operation, the bite block fixed to the maxilla was changed to a removable type and subsequently, underdeveloped side of the maxillary molars' downward traction was started.

Results: Six months after the procedure, the gap between the upper and lower molar teeth of the affected side had been considerably reduced and stable occlusion of the upper and lower teeth was obtained.

Conclusions: In order to prevent early postoperative relapse, meticulously planned preoperative simulation should be carried out so that malocclusion that might occur could be kept to the minimal. Preoperative simulation performed as such has high predictability of the operative outcomes and therefore, allows early acquisition of the satisfactory occlusion during post-operative orthodontics treatment.

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**VIRTUAL SURGICAL PLANNING FOR
ORTHOGNATHIC SURGERY**

Presenter: Alvaro A. Figueroa, DDS, MS
Authors: Figueroa AA, Polley JW
Rush University Medical Center

Treatment planning for orthognathic surgery (OS) has been greatly enhanced through the use of computer-aided design/computer-aided manufacturing (CAD/CAM) technology. The new CAD/CAM approach to plan OS has proven efficient and highly accurate. It is rapidly replacing the use of traditional two-dimensional cephalometric analysis and mock surgery on dental models. CAD/CAM virtual surgical planning (VSP) has improved not only the preoperative planning but also the understanding of the surgical anatomy, and has facilitated the digital preparation of traditionally used surgical splints/as well as newly developed intraoperative surgical positioning guides. In this presentation the step-by-step approach utilized by the authors to plan OS will be discussed. This presentation on VSP will familiarize those clinicians in surgery and orthodontics interested in incorporating current three-dimensional diagnostic and planning approaches into their orthognathic surgical practices.



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POSTOPERATIVE SERUM ALBUMIN LEVEL AND CLINICAL IMPLICATIONS IN PEDIATRIC CRANIOFACIAL SURGERY. A REVIEW OF 50 CASES IN OXFORD, UK

Presenter: Sumit Das, MBBS BSc FRCA
Authors: Das S, Evans RG, Allison E, Johnson D, Wall S

Oxford Craniofacial Unit

Following an incident of phenytoin toxicity in a postoperative craniofacial patient, it was noted that patients undergoing craniofacial reconstruction surgery in our institution, show a statistically significant reduction ($p=0.0018$) in serum albumin levels in the immediate postoperative period.

In order to identify a potential cause for this, we conducted a literature search and found very little reported regarding perioperative albumin levels in any population. We hypothesise that loss of albumin through blood volume loss and the dilutional effect of large blood volume transfusion is the likely mechanism for the drop in albumin. The use of albumin-poor fluids and packed red blood cells is likely to be responsible for the effect.

We reviewed our units statistics on blood loss and replacement fluid for 50 paediatric patients undergoing major craniofacial reconstruction surgery. This information was then statistically analysed to determine if there was any correlation between degree of percentage blood volume loss, fluid types and volumes used and level of albumin drop.

Post-operative hypoalbuminemia has not been previously reported in children undergoing craniofacial surgery. Its relevance may extend beyond our population, and may be a feature in other patient groups who experience major peri-operative blood volume loss. The change in practice from transfusing whole blood to packed red cells is a likely contributing factor.

Since hypoalbuminemia has important clinical implications, such as toxicity of highly protein-bound drugs (e.g. Phenytoin, Carbamazepine, Valproic acid, Phenobarbital). We would like to highlight this as a feature of the post-operative period in this patient population. We would be interested to see if there is variation between institutions that may use alternative transfusion strategies.

Keywords: serum albumin, craniosynostosis, syndromic, non-syndromic, craniofacial surgery, pediatric anesthesia, high protein-binding

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INTELLECTUAL OUTCOME IN 29 ADULT PATIENTS BORN WITH CRANIOSYNOSTOSIS

Presenter: Robert Tovetjarn, MD
Authors: Tovetjarn R, Maltese G, Andersson C, Kolby L, Tarnow P

Institute of Clinical Sciences

Background: The intellectual outcome in adult patients born with craniofacial anomalies needs further investigations both to evaluate the consequences of their anomalies and the craniofacial surgical procedures performed in early life.

Objectives: The aim of this study was to present the intellectual outcome, in terms of psychological scores, extracted from the Swedish Defence Recruitment Agency, in adult patients born with craniosynostosis.

Patients and Methods: A series of patients from the Gothenburg craniofacial registry with metopic synostosis, sagittal synostosis, and Saethre-Chotzen syndrome, and having psychological score from the Swedish Defence Recruitment Agency were included. The psychological abilities were tested by four parts; logistic-inductive, verbal, spatial, and theoretical-technical. As control material served all Swedish males that performed the recruitment in year 2000. Student's t-test was used for statistical analyses.

Results: Twenty-nine adult patients, all males and having psychological scores from the recruitment test could be identified. The diagnoses were: 15 metopic synostosis, 6 sagittal synostosis, and 8 Saethre-Chotzen syndrome. The mean psychological score in the patient group was 4.9 ± 2.4 (mean \pm SD). In the metopic group the score was 5.1 ± 2.1 , in the sagittal group 6.5 ± 2.6 , and in the Saethre-Chotzen group 3.4 ± 1.9 . The control group ($n=37\ 717$) had a mean score of 4.9 ± 2.0 . Only patients with Saethre-Chotzen syndrome differed significantly from controls ($p < 0.05$).

Conclusion: In this small study sample, adult patients with metopic and sagittal synostosis seem to have an intellectual function comparable to the normal population, while Saethre-Chotzen syndrome patients don't. Further long-term follow-up studies are encouraged.



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LOCAL FOREIGN-BODY REACTION TO COMMERCIAL BIODEGRADABLE IMPLANTS: AN IN VIVO ANIMAL STUDY

Presenter: Amy Xue, MD
Authors: Xue A, Koshy JC, Wolfswinkel EM, Weathers WM, Brown RH, Kaufman Y, Hollier LH

Baylor College of Medicine

Background: Resorbable plates have been used extensively in fracture fixation since the 1960s. They rarely cause stress-protection atrophy or problems requiring secondary plate removal, common complications seen with metallic plates. However, aseptic foreign-body reactions have been reported, sometimes years after the original implantation. Both inadequate polymer degradation and debris accumulation have been implicated. The current generation of commercial biodegradable plates is formulated to minimize this complication by altering the ratio of polylactic and polyglycolic acids. This in vivo study compares the degree of local foreign-body reaction of two commercially available resorbable plates in rabbits.

Method: Two types of resorbable plates were examined: poly-D/L-lactide (PDLLA) and polylactide-coglycolide (PLGA). Each plate was placed into a periosteal pericalvarial pocket created beneath the anterior or posterior scalp of a rabbit. Sacrifice occurred at 3, 6 and 12 months postoperatively. Foreign-body reaction was evaluated histologically.

Result: The PLLA plates demonstrated marked local foreign-body reactions within the implant capsule as early as 3 months after implantation, with presence of inflammatory cells and granulomatous giant cells in close association with the implant material. All local foreign body reactions were subclinical with no corresponding tissue swelling requiring drainage. PLGA plates did not demonstrate any signs of inflammatory reactions. In addition, the PLGA plates did not appear to resorb or integrate at 12 months. Neither PLLA nor PLGA plate demonstrated inflammation of the soft tissue or adjacent bone outside the implant capsule.

Conclusion: In our study, the PLLA plates demonstrated histological evidence of foreign body reaction that is confined within the implant capsule, which was not seen with the PLGA plates. This finding may be attributable to the lack of significant resorption seen in the PLGA plates. Both PLLA and PLGA plates were biocompatible with the rabbit tissue environment and should be considered for continued use in craniomaxillofacial reconstruction.

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HEALING OF CRITICAL CALVARIAL DEFECTS BY CIRCULATING MESENCHYMAL STEM CELLS

Presenter: Michael S. Hu, MD, MPH, MS
Authors: Hyun JS, Wu JC, Lo DD, Li SW, Chung MT, Huang KJ, Hu M, Longaker MT, Lorenz HP
Stanford University

Introduction: An urgent need exists for new strategies to accelerate bone regeneration in skeletal defects. Mesenchymal stem cells (MSCs) are promising in bone tissue engineering for their biological ability to differentiate into osteogenic lineages. Herein, we demonstrate the potential of peripheral blood-derived MSCs (PB-MSCs) for bone tissue engineering.

Methods: Circulating PB-MSCs were obtained from peripheral blood of 2-month-old CD1 mice by a novel method utilizing OptiPrep density gradient isolation and hepatocyte stimulation. Cells were characterized by fluorescence-activated cell sorting (FACS). Potential for osteogenic differentiation in vitro was demonstrated by alizarin red staining. For evaluation of in vivo osteogenesis, critical-sized (4 mm) calvarial defects in 2-month-old female CD1 athymic nude mice were treated with hydroxyapatite-poly (lactic-coglycolic acid) (HA-PLGA) scaffold with or without PB-MSCs. Healing was followed using micro-CT scans for 8 weeks.

Results: Alizarin red staining demonstrated in vitro osteogenesis of PB-MSCs with an optical density (OD) at 450 nm of 0.415 versus 0.009 ($p=0.005$) in controls. Quantification of in vivo healing demonstrated increased bone formation in critical-sized calvarial defects treated with HA-PLGA scaffold seeded with PB-MSCs versus HA-PLGA scaffold alone at 8 weeks postoperatively ($p=0.01$).

Conclusions: We demonstrate osteogenic potential in vitro. Utilizing an in vivo calvarial defect model, we show enhanced healing of defects with our PB-MSCs. This novel approach may provide a promising alternative to available techniques for bone tissue repair.



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3D CONSTRUCTS OF HUMAN SKELETAL MUSCLE-DERIVED CELLS AS A MODEL OF BONE FORMATION: OSTEOGENIC DIFFERENTIATION IN RESPONSE TO BMP₂

Presenter: Tiffany Y. Lee, BA
Authors: Lee TY, Meszaros LB, Tobita K, Kumar AR
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In craniofacial surgery, surgeons are challenged with a shortage of bone tissue at the site of defect. Human skeletal muscle-derived cells (skMDCs) undergo osteogenic differentiation in response to bone morphogenetic proteins (BMP) and have been explored for use in bone reconstructions. Before cell therapy can be considered for clinical use, delivery of cells and control of bone growth must be optimized. The aim of this study was to determine the effect of BMP₂ on differentiation of skMDCs fabricated into 3D constructs, thereby validating this novel model of bone formation. 3D constructs provide a more accurate representation of in vivo setting than monolayer cultures and may be used in bone grafting without additional structural support. skMDCs from 20, 25, and 32 year-old male donors were purchased from Cook MyoSite and cultured in growth media. 1x10⁵ skMDCs were fabricated into 3D constructs with ECM components using Flexcell Tissue Train System then cultured in osteogenic media with or without BMP₂ (50ng/mL) for 3 days. RNA isolation and reverse transcription were then performed. Marker genes of osteogenic and myogenic differentiation, *Osx* and *MyoD*, respectively, were evaluated via qPCR. Their expression was normalized to GAPDH, and fold difference of BMP₂ groups over their controls were computed. The data was analyzed with ANOVA and Tukeys test with $P < 0.05$ considered statistically significant. BMP₂ treatments resulted in statistically significant difference in both *Osx* and *MyoD* expression. *Osx* expression was increased with BMP₂ treatment (mean fold difference/control \pm SD): 5.12 \pm 1.01 for 20yo, 2.85 \pm 0.10 for 25yo, and 3.67 \pm 1.09 for 32yo. *MyoD* expression was decreased with BMP₂ treatment: 0.61 \pm 0.06 for 20yo, 0.51 \pm 0.10 for 25yo, and 0.25 \pm 0.11 for 32yo. The result indicates that BMP₂ promotes osteogenic differentiation of skMDC 3D constructs as evidenced by the increased *Osx* expression and decreased *MyoD* expression with BMP₂ treatment. The 3D constructs showed an osteogenic response to BMP₂ comparable to other models and can therefore serve as an excellent in vitro model of bone formation.

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APPLICATION OF LASER CAPTURE MICRODISSECTION TO CRANIOFACIAL BIOLOGY: CHARACTERIZATION OF ANATOMICALLY RELEVANT GENE EXPRESSION IN THE CRANIAL SUTURES

Presenter: Alex Rottgers, MD
Authors: Rottgers A, Gallo P, MacIsaac Z, Cray JJ, Smith DM, Kathju S, Mooney M, Losee JE, Cooper G
University of Pittsburgh School of Medicine

Background: Determining how and when the component tissues of cranial sutures express different signaling factors will help determine how they interact to maintain suture patency and trigger fusion. Previous technologies applied to studying these behaviors are difficult to reproduce and subject to bias. Laser capture microdissection (LCM) allows precise isolation of small homogenous cell populations from frozen histology sections. RNA extracted from these samples can be analyzed using qPCR to measure factor expression within different tissues for comparison. Previous studies have implicated a role of TGF β signaling in suture fusion in our colony of craniosynostotic rabbits and shown Noggin to be ineffective in preventing their suture fusion.

Methods: Coronal sutures from 10 day old wild-type (wt) and craniosynostotic (cs) New Zealand White rabbits were prepared and LCM was used to isolate RNA samples from suture, osteogenic fronts, dura, and periosteum. Taqman probes for FGF₂, FGFR₂, TGF β -2, TGF β -3, BMP-2, BMP-4, and Noggin were tested on each sample using probes for 18s RNA and GAPDH as endogenous controls. Relative expression was determined using $2^{-\Delta\Delta Ct}$ calculations.

Results: TGF β -3 expression was 15x less in cs osteogenic fronts than wild-type. Craniosynostotic suture contents expressed 5x more TGF β -2 than wild-type. Noggin expression was elevated in 10x cs sutures and diminished 13x in wt dura. Finally FGF₂ expression was diminished in both cs suture and dura while its receptor FGFR₂ was increased in cs suture contents. BMP-2 and BMP-4 expression was variable between tissues.

Conclusion: The tissue-specific differences in growth factor expression demonstrated above are unlikely to be appreciated without the specificity achieved with LCM. The data corroborates previous evidence that altered TGF β signaling plays a role in causing suture fusion in this colony of craniosynostotic rabbits, and that the FGF/Noggin signaling pathway described in other models is either secondary or unaffected in this model of suture fusion.



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THE ABSOLUTE QUANTIFICATION OF OSTEOGENIC GENES IN RAT CALVARIAL CRITICAL SIZED DEFECTS

Presenter: Brendan J. Alleyne, BS
Authors: Alleyne BJ, Gliniak C, Varghai D, Askeroglu U, Tobin K, Zwiebel S, Sweeney WM, Cooper GM, Gosain AK

Case Western Reserve University School of Medicine

Purpose: A rat critical sized defect model was used to test the levels of osteogenic genes in the dura mater in rats of varying age. The purpose of this study was to identify factors that are upregulated in dura underneath critical sized defects and to identify whether age or time after wounding impacts these factors.

Methods: Unilateral parietal defects sized 5, 6 & 8 mm were created in P6, P20 & 12 week old rats respectively. Defect size was determined as a ratio of defect size to Lambda-Bregma distance. Dura was harvested at 3, 7 or 14 days after surgery. Dura was collected from beneath defects and from the intact parietal bone on the contralateral side. Absolute qPCR was performed for BMP2, FGF2, IGF-1 & TGF- β 1 and the mRNA copy number was expressed per 100 copies of GAPDH control. Fisher's t-test was performed for statistical significance.

Results: In P6 rats 3 days after surgery there was a significant increase of BMP2, FGF2 & TGF- β 1 in dura under defects compared with control dura. The percent increase of factors in defect dura was 176% ($p < 0.019$), 235% ($p < 0.005$) and 545% ($p < 0.017$) for BMP2, FGF2 & TGF- β 1 respectively. In control dura we observed a ratio of BMP2, FGF2, IGF-1 & TGF- β 1 at 21.87-6.59-1.91-1 respectively, which was different than the ratio for defect dura, 9.35-3.40-1-1. Gene expression in dura under defects was highest for post-operative days 3 & 7 in P6 rats.

Conclusion: Our results indicate an increase in BMP2, FGF2 & TGF- β 1 mRNA in dura underlying defects, but not IGF-1. This indicates that while increased production of growth factors is important to healing, the decrease of BMP2 & FGF2 in the ratio in healing defects may also encourage regeneration. These data provide valuable insight into the mechanism of healing of critical sized defects.

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DELETION OF AXIN GENE IN MOUSE ADIPOSE-DERIVED STEM CELLS (MASCs) PROMOTES OSTEOGENESIS

Presenter: Kshemendra Senarath, MD
Authors: Senarath K, Chung MT, Hasegawa M, Quarto N, Satoh K, Longaker MT

Stanford University

Introduction: Wnt pathway has been shown to be important for the osteogenic differentiation of mesenchymal stem cells, bone development, and bone homeostasis. However, the role of Wnt signaling in osteogenesis remains a controversial matter. We investigated how loss of Axin2, a negative regulator of Wnt signaling pathway, influences proliferation and osteogenic differentiation of mASCs.

Methods: Mouse ASCs were harvested from Axin2 null (Axin2 $^{-/-}$) transgenic mice and their corresponding background CD-1 mice. For the growth curve assay, cell counting was performed in triplicate. Cells were cultured in osteogenic differentiation media (ODM) with or without retinoic acid (RA) and with or without Wnt3a treatment. Mineralization of extracellular matrix was assessed by Alkaline phosphatase and Alizarin red staining. Analysis of genes associated with osteogenic differentiation (Runx2, Alpl) and BMP signaling (Bmp2, Bmp4, Smad1, Smad5) was performed by quantitative PCR (qPCR).

Results: A higher proliferative activity in Axin2 $^{-/-}$ ASCs was revealed as compared to CD-1 ASCs. Axin2 $^{-/-}$ ASCs showed more robust mineralization of the extracellular matrix than CD-1 ASCs in ODM with retinoic acid (RA). Interestingly, Axin2 $^{-/-}$ ASCs in the absence of RA also underwent osteogenic differentiation, whereas, control WT ASCs failed. Furthermore, qPCR analysis showed that genes associated with BMP signaling, such as Bmp2, Bmp4, Smad1 and Smad5, were upregulated, specifically in Axin2 $^{-/-}$ ASCs treated with Wnt3a.

Conclusion: Our results indicate that loss of a negative Wnt regulator, such as Axin2, promotes cell proliferation and higher osteogenic potential on Axin2 $^{-/-}$ ASCs. Moreover, these results also suggest the occurrence of a cross talk between Wnt and BMP signaling which is responsible for the greater osteogenic potential of these cells.



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**REPAIR OF A COMPLICATED CALVARIAL DEFECT:
RECONSTRUCTION OF AN INFECTED WOUND WITH
RHBMP-2**

Presenter: Joseph E. Losee, MD
Authors: MacIsaac ZM, Shakir S, Naran S, Cray J,
Nayar HS, Smith DM, Kinsella CR,
Mooney MP, Cooper GM, Losee JE

University of Pittsburgh

Background: Management of craniofacial defects remains a challenge, especially following infection. Optimal therapy would provide replacement identical to that of native calvarial bone, without need for donor site morbidity. The purpose of this study was to compare the effectiveness of recombinant human bone morphogenetic protein-2 (rhBMP-2) mediated bone regeneration with the gold standard of autologous bone graft for repair of previously infected calvarial defects.

Methods: Nineteen adult New Zealand White rabbits underwent subtotal calvariectomy. Bone flaps were inoculated with *S. aureus* and immediately replaced. Following 6 days of infection, bone flaps were removed and wounds debrided, followed by 10 days of antibiotic treatment. After six weeks of recovery, animals underwent reoperation for scar debridement followed by definitive reconstruction in one of four groups: empty control (n=3), vehicle control (buffer solution on absorbable collagen sponge (ACS), n=3), autologous bone graft (n=3), or rhBMP-2 repair (rhBMP-2/ACS, n=10). Animals underwent CT imaging at 0, 2, 4 and 6 weeks postoperatively, followed by euthanization and histological analysis. Percent healing was determined by 3-dimensional analysis. A (time x group) 2-way ANOVA was performed on healing versus treatment group and postoperative time.

Results: At six weeks postoperatively, rhBMP-2/ACS and autologous bone graft resulted in 93% and 68% healing, respectively, while the empty and vehicle control groups resulted in 27% and 26% healing ($p < 0.001$). Histologically, compared to autologous bone graft reconstruction, bone in the rhBMP-2/ACS group was more cellular and more consistently continuous with wound margins. Compared to immediate favorable reconstruction (96.8% healing), rhBMP-2 in this setting was similarly effective ($p > 0.05$).

Conclusions: rhBMP-2 therapy is effective in achieving radiographic coverage of calvarial defects complicated by previous infection. Future studies may employ manipulations such as additional growth factors/cell therapy to further improve results in this difficult scenario.

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**TRANSPLANTATION OF A TISSUE ENGINEERED,
VASCULARIZED, MUCOSALIZED TRACHEAL
HOMOGRAFT IN NEW ZEALAND WHITE RABBITS.
COMPARISON OF SUBCUTANEOUS FLAP AND
RECTUS MUSCLE FLAPS**

Presenter: Christopher Gordon, MD
Authors: Gordon PX, Uribe-Rivera A, DeAlarcon A,
Propst EJ, Lam DJ, Billmire DA, Aronow BJ,
Reyna-Rodriguez XP, Rutter MJ, Gordon CB

Cincinnati Childrens Hospital Medical Center

Long segment tracheal stenosis or tracheal atresia is a life-threatening condition with few options for treatment. The purpose of this study is to compare two flaps and develop a durable, biocompatible, vascularized, mucosalized and epithelialized trachea for transplantation into long tracheal defects.

Methods: Tracheas of 26 New Zealand white rabbits were removed and processed using a gentle decellularization process that maintains glycosaminoglycans and structure using a proprietary method. Processed tracheas were implanted into the subcutaneous fascia n=14 and n=12 into the rectus muscle of live rabbits. All processed tracheas were injected with a mixture of mucosal cells (biopsied during bronchoscopy), abdominal fat (source of stem cells) and phosphate buffered saline (PBS). Six tracheas underwent radiation during the process of decellularization. Two tracheas served as controls and were not injected. Animals were survived for 8 weeks while tracheal constructs were incubated. Tracheal constructs embedded in rectus muscle and subcutaneous fascia were rotated into a created tracheal defect (mean 5.75 +/- 1.5 tracheal rings) and animals were extubated immediately postoperatively and survived for 8 weeks, at which point they were euthanized and the trachea was examined histopathologically.

Results: Rabbits transplanted with tracheal lumen injected with mucosal cells, fat and PBS had less respiratory compromise, survived longer, and histopathologically maintained greater airway patency with less scar formation compared to those receiving additional radiation, not significant differences were observed in those tracheas implanted into rectus muscle or subcutaneous fascia. These rabbits also demonstrated remucosalization of their newly transplanted trachea with evidence of numerous beating cilia confirmed on video movement caption and electron microscopy. RNA seq demonstrated a pattern of ciliated respiratory epithelium.

Conclusions: It is possible to create a durable, biocompatible, vascularized, mucosalized and epithelialized trachea using tissue tissue engineering



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**LACUNOCANALICULAR FLOW AND DISTRACTION
OSTEOGENESIS: THEORY AND THERAPEUTICS**

Presenter: Edward H. Davidson, MA (Cantab), MBBS
Authors: Davidson EH, Butala P, Sultan SM,
Knobel D, Tutela JP, Canizares O, Wagner IJ,
Warren SM

University of Pittsburgh

Introduction: Our hypothesis is that the tension stress of activation increases lacunocanalicular flow and upregulates the mechanotransductive osteogenic pathway. Furthermore, we hypothesize improving vascularization by endothelial progenitor cell (EPC) mobilization enhances lacunocanalicular flow in the consolidation period, maintaining upregulation of the mechanotransductive pathway to accelerate osteogenesis.

Methods: Sprague-Dawley rats were subjected to mandibular osteotomy alone or distraction with 3-day latency and 7-day activation periods. At day 10, subjects were injected with fluorescent reactive red lacunocanalicular tracer prior to sacrifice. FAK (integrin cascade regulator), SUN, lamin, nesprins (cytoskeletal mediators), sox 9, osterix, runx2, BMP 2, 4, Smads 1,5,8 (osteogenic markers) were measured with western blotting. Additional animals, having undergone latency and activation, underwent a 28-day consolidation period, receiving daily injections of either AMD3100 (bone marrow EPC mobilizing agent) or saline. Animals were sacrificed on postoperative day 38. Reactive red tracing and western blotting for mechanotransductive proteins were performed. Bony regeneration was assessed (μ -CT, DEXA, immunohistochemistry, mechanical testing).

Results: Fluorescent intensity of reactive red was 4.5% following mechanical activation vs 1.6% following osteotomy alone. Mechanotransductive mediators in the activated group were upregulated eg FAK (13.9×10^5 vs 8.9×10^5), SUN (12.1×10^5 vs 6.6×10^5) etc.

Fluorescent intensity was 1.9% with AMD3100 vs 0.1% with saline. Upregulation of mechanotransductive mediators was seen with progenitor cell mobilization eg FAK (3.9×10^6 vs 1.4×10^6), SUN (3.7×10^6 vs 1.2×10^6), Nesprin (11.4×10^5 vs 6.5×10^5) etc. μ -CT and immunohistochemistry demonstrated AMD3100 treatment increased bone formation (8.93 vs 3.94%). Greater force was required to break AMD3100-supplemented bone (98 vs. 60N).

Conclusion: This is the first description of lacunocanalicular flow as the transducer of mechanical tension stress to osteogenesis in distraction and introduces a novel mechanism to accelerate osteogenesis.

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**PUTATIVE GENES DOWNSTREAM OF FGFR2
ASSOCIATED WITH CORONAL SUTURE SYNOSTOSIS
IN A CROUZON SYNDROME (C342Y) MOUSE MODEL**

Presenter: Samintharaj Kumar, MBBS, BDS, MRCS,
MFDSRCS, MFDRCSI

Authors: Kumar S, Peskett E, Britto JA, Pauws E
University College London

Crouzon syndrome is most commonly caused by the FGFR2C342Y/+ mutation. Much is known about FGFR signalling in membranous osteogenesis, and its downstream biochemical effects, however, the associated molecular genetic mechanisms are poorly understood.

Morphological differences in the Crouzon mouse model between wild-type (Wt) and Fgfr2C342Y/+ (Mut) calvaria were found after embryonic day (E)17.5, suggesting enrichment for genes responsible for coronal synostosis.

Coronal sutures were micro-dissected from E17.5 Wt/Mut mouse calvaria and gene expression profiles were examined using an Affymetrix Mouse Exon microarray. 51 genes were found to be differentially expressed by at least 1.5-fold ($p < 0.05$); of which five; Dpt, Osr1, Nov, Dlk1 and Kera, are downregulated up to 3.5-fold in the fusing mutant suture. Validation of these findings by qPCR, implies functional significance for these genes in craniosynostosis.

In situ hybridisation of these genes at different embryonic stages of development (E15.5-E18.5) showed complementary spatio-temporal expression in dura and periosteum in calvarial parasagittal paraffin sections. In addition, RNAi knockdown of Fgfr2c in the MC3T3 osteoblast cell line identified transcriptional changes in a subset of these genes ($p < 0.05$).

We have identified novel genes, downstream of FGFR2 signalling, that are implicated in the pathogenesis of murine coronal synostosis. These genes and their related pathways may identify key targets for genetic diagnosis and pharmacotherapy in syndromic craniosynostosis.



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ACCELERATION OF REGENERATE CONSOLIDATION UTILIZING PHARMACOTHERAPEUTICS DURING MANDIBULAR DISTRACTION OSTEOGENESIS

Presenter: Steven Buchman, MD
Authors: Buchman S, Donneys A, Deshapande SS, Tchanque-Fossuo CN

University of Michigan

Objective: An inherent limitation of mandibular Distraction Osteogenesis (DO) is the length of time required for successful consolidation. This drawback subjects patients to vulnerability by prolonged return to activities of normal daily living. Finding techniques to reduce consolidation periods could be quite beneficial. Deferoxamine (DFO) is a pharmacotherapeutic, already on formulary, that triggers the HIF-1R± pathway through a mechanism of localized iron depletion. We previously established the effectiveness of DFO in enhancing regenerate vascularity at a full consolidation period (28 days). To investigate whether this augmentation in vascularity would function to accelerate consolidation without compromising regenerate quality or strength, we progressively shortened consolidation periods prior to microCT imaging and biomechanical testing.

Methods: 3 time points (28d, 14d and 7d) were selected and 6 groups of Sprague-Dawley rats (n=60) were divided into control (C) and experimental (E) groups for each time period. All groups underwent external fixator placement, mandibular osteotomy, and a 5.1 mm distraction. During distraction, the experimental groups were treated with DFO injections into the distraction gap. After consolidation, mandibles were imaged and tension tested to failure. Statistical comparisons were conducted with an independent samples t-test, and $p < 0.05$ was considered statistically significant.

Results: At 14 days of consolidation the experimental group demonstrated a striking 100% increase in the number of bony unions (C:4/10 vs. E:8/10). Significant increases in Bone Volume Fraction (BVF), Bone Mineral Density (BMD) and Ultimate Load (UL) were observed when comparing 14d C to 14d E. While significant decreases in Total Volume, Bone Volume, BMD, Yield, UL and Stiffness were observed when comparing 28d C to 14d C, no significant differences were observed when comparing 28d C to 14d E.

Conclusions: We contend that augmentation of vascular density through localized DFO injection delivers an efficient means for accelerating bone regeneration without significantly impacting bone quality or strength.

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STABILITY OF SINGLE STAGE MAJOR MAXILLARY ADVANCEMENTS IN UNILATERAL CLEFT PATIENTS

Presenter: Guy D. Watts, MBBS, FRACS
Authors: Watts GD, Phillips J, Forrest C, Bigdoli-Moghaddam M, Antonarakis G

Hospital for Sick Children Toronto

Purpose: There is controversy about the need for distraction in Le Fort I maxillary advancements >6mm to achieve both the surgical movement and to maintain long-term stability. This paper looks at the long-term stability of single stage Le Fort I advancements >10mm in the unilateral cleft population.

Methods: A retrospective review of the stability of Le Fort I advancement (>10mm) for maxillary hypoplasia in unilateral cleft patients between 2002 and 2010 at The Hospital for Sick Children. 20 patients (6 females 14 males) were identified with complete data sets including pre-operative T₁, early postoperative T₂ and long-term T₃ (>12 months) lateral cephalometric studies. The population was subdivided into two groups; two-piece segmental advancement with immediate alveolar bone graft (n=16) and a single piece advancement group that had been previously bone grafted (n=4). The institutions cephalometric analysis was performed with the dentofacial planner 7.2. Skeletal and dental measures were used to assess long-term stability.

Results: Change between T₁ and T₂ indicated surgical movement and T₂ to T₃ indicated relapse. Mean horizontal surgical movement was 11.8 mm (range 10.0-15.4) and mean vertical movement 1.9mm (range 1.1-2.7). Mean horizontal relapse 2 mm (range 1-3.1) and mean vertical relapse 0.6 mm (0-1.39). Statistically significant relapse was noted for all maxillary outcome measures except palatal plane. Significant clinical relapse was defined as >2 mm or >2 degrees, a value deemed orthodontically salvageable for occlusal outcomes. When this was incorporated into statistical calculations there were no significant relapses for maxillary outcomes measures. This was confirmed with dental measurements demonstrating no statistically significant relapse during follow-up. These results were compared to a previous study at the same institution with <10 mm advancements. The results did not demonstrate any significant difference in relapse rates between the two groups.

Conclusion: Greater than 10 mm advancements can be achieved and are stable at long-term followup in unilateral cleft patients.



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SHORT- AND LONG-TERM CHANGES OF CONDYLAR POSITION AFTER BILATERAL SAGITTAL SPLIT RAMUS OSTEOTOMY FOR MANDIBULAR ADVANCEMENT EVALUATED BY CONE-BEAM COMPUTED TOMOGRAPHY

Presenter: Shuo Chen, MD
Authors: Chen S, Lei J, Li ZL, Liang C, Wang XX, Wang X, Fu KY, Farzad P, Yi B
School and Hospital of Stomatology Peking University

Purpose: Bilateral Sagittal split ramus osteotomy (BSSRO) may change condylar position, which can be one of the factors contributing to skeletal relapse. This study aimed to evaluate short- and long-term changes in condylar position using cone-beam computed tomography (CBCT) and to investigate changes in temporomandibular joint (TMJ) signs, after BSSRO for mandibular advancement.

Materials and Methods: Thirty-five patients were included, and CBCT data of 70 TMJs were collected before surgery (T₀), immediately after surgery (T₁), 3 months after surgery (T₂) and at the last follow-up 12.1±2.9 months after surgery (T₃). Condyle-fossa relationship was evaluated by Pullinger and Hollender's method. Clinical examination with special focus on signs of temporomandibular disorder (TMD) was documented at T₀, T₂, and T₃. A repeated-measures analysis of variance (ANOVA) (P= 0.05) and chi square test was performed (P= 0.05).

Results: Data of twenty-nine patients were used for statistical analysis. Values from Pullinger formula changed significantly with time but there was no significant difference between right and left condyles. Condyles moved infero-posteriorly immediately after the surgery (T₀-T₁) followed by antero-superior movement 3 months after surgery (T₁-T₂). The superimposed effect demonstrated postero-superior movement compared with the initial position before surgery (T₀-T₂) and this position remained stable at 1-year follow-up (T₂-T₃). A decrease of TMD signs over time from 21.4% (T₀) to 12.9% (T₂) and 8.6% (T₃) was observed, which showed no statistical significance.

Conclusions: There were obvious changes in condylar position after BSSRO. Condyles tended to be located in a concentric position in relation to the glenoid fossa 3 months after surgery and maintained stable during 1-year follow-up. These changes did not cause an increase of TMD signs.

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3D FACIAL SIMULATION IN ORTHOGNATHIC SURGERY: EFFICACY AND ACCURACY

Presenter: Stephen A. Schendel, DDS, MD
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Stanford University

Introduction: The development of Cone Beam Computed Tomography (CBCT) and 3D facial scanning have permitted the creation of fused images thus creating a virtual 3D patient which is called the patient specific anatomical reconstruction. (PSAR) 3D Simulation of the desired surgical result both from a skeletal and soft tissue standpoint is thus now possible. The importance of 3D virtual surgical planning increases with the complexity of the deformity and reconstruction needed for the correction. In this study we evaluated the accuracy and predictability of 3D computer soft tissue simulation in orthognathic surgery.

Materials & Method: 23 consecutive patients who underwent orthognathic surgery were scanned by the 3dMD (3dMD Inc., Atlanta, Georgia) photogrammetric facial scanner and a CBCT scan was also obtained prior to and after surgery. The facial scan was then fused to the CBCT scan creating a patient specific image, according to the Vultus protocol. The surgery was then simulated in 3D from the pre-operative fused image and the resultant simulated soft tissue face compared to the actual facial scan obtained post-operatively. The simulated soft tissue image is created by the mass springs model where multiple springs (>500,000) connect the hard tissue polygons to the soft tissue polygons. This generates 3D deformable tissue models that include spring-based force computations to model the physical characteristics of real tissue reactions. This mechanism simulates deformable tissues constructed from 3D nodes, faces, tetrahedral and edges, which include stiffness parameters to determine the ease of tissue deformation since it is well known that soft tissues in different areas respond differently.

Results: The greatest difference found between anatomical points on the face was 0.6 mm and most points were considerably below this, below 0.4 mm. The clinical rule of thumb for soft tissue simulation systems to be accurate enough for clinical use has routinely been stated as 0.5 mm or less for the variables.



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MAXILLARY ADVANCEMENT AND MANDIBULAR RETRACTION USING NON-OSTEOTOMY TECHNIQUE WITH BOLLARD PLATES

Presenter: Eric Stelnicki, MD
Authors: Stelnicki E, Marchetto J, Almashat R
Nova Southeastern University

Introduction: In this study, we analyze the effect of maxillary advancement in CL III skeletal patients using a novel approach. Basically, a non-osteotomy two jaw surgery that produces combined maxillary advancement and mandibular retraction via growth orthopedics using Bollard plates is applied. The purpose of this study was to evaluate the change in the WITS appraisal and ANB values for each patient before and after treatment.

Methods: A total of 14 patients were enrolled in the study as early as possible, cervical vertebral maturation stage CVM2. Bilateral maxillary FDA approved miniplates (Bollard plates) were surgically inserted in the infrazygomatic crest of the maxilla and bilateral mandibular plates in the anterior mandible between the canine and the lateral incisor area. The patients were treated by intermaxillary elastics. Cephalometric analysis was done before, during and at the end of the treatment. Intraoral and extraoral photographs were taken as well.

Results: The WITS appraisal value showed a remarkable increase from -9 to -1 in some patients. Most of the patients showed an increase in the ANB values by 2 or 3 degrees. There was a 100% success in changing the mandible and the maxilla relation. There were minimum complications with 4 patients including loosening of the plates which were easily treated. On the other hand, none of the patients suffered infection, dental complications or nerve injury.

Conclusion: Because of the minimum complications and the maximum benefits related to the Bollard plates, this novel treatment approach shows promising future results for the treatment of CL III skeletal patients.

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MAXILLOMANDIBULAR ADVANCEMENT SURGERY FOR OBSTRUCTIVE SLEEP APNEA SYNDROME AND LONG-TERM SURGICAL STABILITY IN CHINESE PATIENTS

Presenter: Yang Li, MD
Author: Li Y
Peking University School and Hospital of Stomatology

Objective: A prospective study to evaluate the maxillomandibular advancement (MMA) for the treatment of obstructive sleep apnea hypopnea syndrome (OSAHS) in Chinese patients. This study also examined the long-term surgical stability and assessed the acceptance of perceived facial changes.

Methods: 15 patients underwent MMA for the treatment of OSAHS. All the patients underwent polysomnography, cephalometric analysis and 3-dimensional CT at T₁ (3 months before surgery), T₂ (3-6 months postsurgery) and T₃ (3 years postsurgery).

Results: Mean apnoea/hypopnoea index (AHI) decreased from 51.5 events h⁻¹ before therapy to 6.8 events h⁻¹ 3 months postoperatively, and 11.5 events h⁻¹ after 3 years. Oxygen saturation significantly increased following surgery. No significant changes were found comparing the 3 months versus the 3 years follow-up data. The position of the maxilla and mandible measurements in upper airway and objective results assessment show relatively stable over long term. Although extreme maxillomandibular protrusion was found from cephalometric analysis in some patients, favorable esthetic results were achieved in most patients.

Conclusions: The MMA technique for the treatment of OSAHS is highly effective in severe OSAHS in china. The results from this study indicate that large surgical advancements in OSAHS patients result in relatively stable repositioning of the maxilla and mandible over the long term.



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**ORTHOGNATHIC POSITIONING SYSTEM (OPS):
LINKING VIRTUAL SURGICAL PLANNING (VSP) TO
REALITY IN ORTHOGNATHIC SURGERY**

Presenter: John W. Polley, MD
Author: Polley JW
Rush University Medical Center

Virtual Surgical Planning (VSP) has revolutionized treatment planning in orthognathic surgery. VSP allows an unprecedented understanding of planned surgical procedures enabling the surgeon to visualize complicated skeletal movements and the resultant effects on related skeletal structures and soft tissues. Despite this advancement for preoperative planning, intraoperatively surgeons still rely on traditional methodologies of multiple splints, intermaxillary fixation, and manual guessing of proximal segment condylar positioning. The purpose of this presentation is to introduce a new concept and methodology entitled Orthognathic Positioning Systems (OPS) that bridge the gap between VSP and actual surgery. OPS consists of virtually designed, intraoperative splint supported guides, that translate the virtual plan to the operating room. OPS are designed by the surgeon during VSP and manufactured through stereolithographic technology. OPS allows a precise translation of the planned skeletal movements to the operating room. OPS also eliminates multiple traditional intraoperative steps including the need for intermaxillary fixation, eliminates the need for intermediate splints even during 2-jaw surgery, eliminates manual guessing of centric relation and eliminates the dependency on external reference landmarks.

OPS has been used in 30 cases including single and multi-piece Le Fort I, mandibular sagittal split (SSO), and inverted L osteotomies, and genioplasty. Diagnoses include cleft lip and palate, hemifacial microsomia, facial asymmetries, and nonsyndromic dental facial deformities. All patients in this series had a successful treatment outcome. The concept of OPS will be presented and its application illustrated in clinical cases.

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**VIRTUAL PLANNING SOFTWARE FOR
ORTHOGNATHIC SURGERY**

Presenter: Earl A. Gage, MD
Authors: Marsh JL, Gage EA
Mercy Childrens Hospital

Orthognathic surgery has traditionally been planned 2-dimensionally with a 3-dimensional assist from dental study models. The advent of computer-assisted medically imaging and 3D reformation software now makes it possible for patient-specific anatomically-accurate computer simulation of complex maxillofacial dentoskeletal movements including fabrication of intermediate and final intra-operative occlusal splints.

We present our experience with the VSP Orthognathics surgical planning and splint fabrication system over the past 2 years in 17 patients. The process will be described: 1. head CT data, dental study models and the surgical plan are sent to the vendor; 2. the dental models are laser scanned and the data embedded into the CT data; 3. the vendor's surgical simulator segments the facial skeleton per the surgeon's plan and simulates the requested movements; 4. the simulator conducts a web conference with the surgeon reviewing the simulation and making adjustments, as directed by the surgeon, in real-time; 5. the surgeon approves the surgical plan; 6. the vendor CAD/CAM produces the surgical splint(s) and digital and hardcopy surgical plan output that are forwarded to the surgeon for intra-operative use. Outcome (photographic and radiographic) will be presented. This process eliminates the need for traditional stone model surgery and face-bow transfers, and accurately predicts bony gaps and overlaps improving patient outcome and saving valuable OR time. The process optimizes the surgeon's time and is cost-effective.

While helpful for simple (one-jaw, 2D movement) orthognathic cases, 3D computer simulation and surgical splint fabrication is invaluable for complex, asymmetric, two-jaw, 3D dentoskeletal movements.



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MINIMAL INCISION LEFORT III OSTEOTOMY WITH RIGID EXTERNAL DISTRACTION: ANALYSIS AND CLINICAL OUTCOMES OF 12 SYNDROMIC PATIENTS

Presenter: Scott Rapp, MD
Authors: Rapp S, Uribe-Rivera A, Pan BS, Billmire DA, Gordon CB
Cincinnati Childrens Hospital Medical Center

Purpose: Lefort III distraction is an effective tool to address midface hypoplasia and obstructive sleep apnea in syndromic patients. Conventional osteotomies require extensive dissection and devascularization of the segments. This may affect stability, growth and lead to increased morbidity. The authors report a minimally invasive approach with transconjunctival osteotomies and external rigid distractors to minimize morbidity, improve stability and airway patency in a syndromic population.

Materials and Methods: From 2004 to 2013, 12 syndromic patients with proven obstructive sleep apnea and failed conservative management underwent Lefort III osteotomies through extended transconjunctival and 1 cm vestibular incisions. Ages ranged from 3-15 yrs. Osteotomies were performed with the Sonopet Ultrasonic Aspirator (Stryker) and subsequent rigid external device (KLS-Martin) fixation. Distraction was performed at 2 mm a day and discontinued when airway was deemed improved by direct microlaryngoscopy. Linear bone movements were measured by weekly cephalographs.

Results: 2-D cephalometrics were used to assess pre-operative and post-operative midface movement up to 2 years following surgery. The average pre and post-operative posterior airway space (PAS) increase in distance is 8.99+ 5.4 mm (43.5%). The average pre and post-operative menton to hyoid (Me-Hy) increase in distance was 19.72 + 9.8 mm (47.9%). The average SNA angle increase is 13.4 + 7.5 degrees post-distraction. Average blood loss was 295 mL. Comparative outcome data will be presented regarding operative cost, ICU and overall hospital stay, blood loss, transfusion rates, decannulation rates and peri-operative complications.

Discussion: Drawbacks of LeFort III distraction relate to extensive degloving during hardware placement and osteotomy. We developed a transconjunctival approach to address these issues utilizing ultrasonic osteotomy and limited dissection. Early outcomes appear to be superior to open procedures with improved stability and decreased morbidity.

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SURGERY-FIRST ORTHOGNATHIC SURGERY (WITHOUT PRESURGICAL ORTHODONTICS) -- EFFICIENCY AND STABILITY

Presenter: YuRay Chen, MD
Authors: Chen Y, Liao YF, Ko EW, Wang YC, Huang CS
Chang Gung Memorial Hospital

Purpose: Studies were designed to compare the postsurgical facial aesthetics, occlusion, stability (transverse, anterior-posterior and vertical dimensions) and efficiency with or without presurgical orthodontics.

Material and Method: A retrospective cohort study was carried out to compare the final outcomes of orthognathic correction of skeletal Class III between one group with presurgical orthodontic treatment and the other group without. The final outcomes were assessed by photography, dental casts and cephalometric analysis at one year after the surgery.

Results: The improvements in facial esthetics and dental occlusion were very similar in both groups. However the treatment time was about 6 months shorter in the group without presurgical orthodontics. The skeletal stability was assessed in transverse, vertical and anteroposterior planes. In transverse plane (Wang et al, 2010), molar width was decreased and canine width was increased in both groups after the surgery. Eventually, maxillary and mandibular molar inclinations and canine inclinations will be the same for both groups at one year after the surgery. In vertical plane (Liao et al, 2010), vertical control of B point and Pogonion was better in the group without presurgical orthodontics. Better vertical control of B point and pogonion could increase the overbite to prevent the relapse of anterior open bite after the surgery. In anteroposterior plane (Ko et al, 2011), the presurgical orthodontic movements were mainly in dental tipping movements which could be easily accomplished by postsurgical orthodontic treatment. There was no difference for skeletal and dental relapse in the three planes between these two groups after the surgery.

Conclusion: In surgical-orthodontic correction of skeletal class III, presurgical orthodontics has no clinically significant effects on facial aesthetics, occlusion or stability, and had longer treatment time compared to those receiving no presurgical orthodontics.



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MUTATIONS IN TCF12 ARE A FREQUENT CAUSE OF CORONAL CRANIOSYNOSTOSIS

Presenter: Jacqueline A. Goos, MD

Authors: Goos JA, Sharma VP, Fenwick A, Wilkie AO, Mathijssen IM

Erasmus MC

Background: Although several genes are known to cause craniosynostosis (e.g. FGFR2, FGFR3 and TWIST1), many craniosynostosis patients suffer from an unidentified genetic cause. Possibly, more genetic causes can be identified by next generation sequencing. The identification of TCF12 as a new craniosynostosis gene was also made possible by this technique.

Methods: Whole exome sequencing was performed in seven patients with bicoronal synostosis. Three out of seven carried a mutation in Transcription Factor 12 (TCF12). A search for more patients with mutations in this gene was carried out (e.g. in other exome and whole genome projects, in 297 patients with different types of craniosynostosis, in the Oxford birth cohort, and in an independent cohort of fifty patients with coronal synostosis). The family members of the TCF12-positive patients were also tested. And the phenotypes of mice heterozygous for Tcf12 or Twist1 were compared to the phenotype of mice doubly heterozygous for Tcf12 and Twist1.

Results: Mutations in TCF12 were found in 38 patients. All patients had coronal synostosis and a benign course of the surgical trajectory. None of the patients had an interval of raised intracranial pressure. 35 TCF12-positive family members were also identified, but only 16 of them had craniosynostosis or other suspicious clinical features. In addition, mice doubly heterozygous for Tcf12 and Twist1 suffered from severe coronal synostosis.

Discussion: Patients with mutations in TCF12 have coronal synostosis and a benign surgical course. They don't suffer from raised intracranial pressure. Mutation-positive individuals can also have a normal phenotype, indicating substantial non-penetrance. The mouse model shows that the total quantity of TCF12/TWIST1 heterodimers plays an important role in coronal suture development. In conclusion, a new craniosynostosis gene is identified: TCF12.

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CROUZON SYNDROME AND BENT BONE DYSPLASIA ASSOCIATED TO MUTATIONS AT THE SAME TYR-381 RESIDUE IN FGFR2 GENE

Presenter: Corinne Collet, MD

Authors: Collet C, Alessandri JL, Arnaud E, Cormier-Daire V, Di Rocco F

Craniofacial Unit

Crouzon syndrome has an high penetrance and an extreme clinical variability. The majority of the mutations responsible for his syndrome are located in the exon 8 (IIIa) and 10 (IIIc) of the fibroblast growth factor receptor 2 gene (FGFR2) corresponding to an immunoglobulin domain. We report here a familial Crouzon syndrome associated to a new p.Tyr381Asn mutation in exon 11 of FGFR2 gene. The proband was suspected of this syndrome at the age of 3 years. At that time he presented with adolichocephaly with bilateral moderate exophthalmia, parrot-beaked nose and frontal bossing. A skull CT examination was performed at age 5 confirming the closure of the sagittal suture associated a facial retrusion and inverted bite. His father and his sister displayed a milder phenotype. The Crouzon syndrome was confirmed after detection of novel heterozygous p.Tyr381Asn (c.1141T>C) mutation in FGFR2 gene located in the transmembrane domain for the 3 affected members of the family, segregating with the craniosynostosis phenotype. This mutation was located at the same position (p.Tyr381Asp(c.1141T>G)) that the recently described perinatal lethal bent bone dysplasia which is characterized by coronal craniosynostosis associated with hypoplastic clavicle, bent long bones in the lower extremities, hypoplastic pubis and abnormal phalanges. Ours cases did not show any appendicular bony abnormality on X-rays. This observation highlights the need of further researches to explain such major clinical difference between those two syndromes associated with a FGFR2 mutation affecting the same Tyr-381 residue.



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NON-SCAPHOCEPHALIC SAGITTAL SYNOSTOSIS AND RAISED INTRACRANIAL PRESSURE ARE PART OF THE CLINICAL SPECTRUM OF FRANK-TER HAAR SYNDROME

Presenter: Charlotte L. Bendon, BA, BMBCh
Authors: Bendon CL, Fenwick AL, Hurst JA, Nuernberg G, Nuernberg P, Wall SA, Wilkie AO, Johnson D

Oxford University Hospitals

Background: Frank-ter Haar syndrome is a rare disorder associated with craniofacial features including hypertelorism, brachycephaly and a wide anterior fontanelle. The underlying genetic defect in Frank-ter Haar syndrome is thought to be a mutation in the SH3PXD2B gene on chromosome 5q35.1. Premature fusion of the sagittal suture classically produces a long, narrow, scaphocephalic skull deformity, and is associated with raised intracranial pressure in 14% of cases. Non-scaphocephalic sagittal synostosis is rare, and understanding of the condition is limited. Craniosynostosis, or premature fusion of the calvarial sutures, has not been reported previously in patients with Frank-ter Haar syndrome.

Case Presentation: We present three affected siblings born to consanguineous parents with cardiac, skeletal and craniofacial features in keeping with a diagnosis of Frank-ter Haar syndrome, including hypertelorism, brachycephaly, a prominent forehead and broad mouth. These three siblings are all homozygous for a de novo deletion of exon 13 of the SH3PXD2B gene demonstrated by PCR amplification of SH3PXD2B. In addition to craniofacial features previously reported in Frank-ter Haar syndrome, two of the three siblings also have non-scaphocephalic sagittal synostosis, both complicated by raised intracranial pressure.

Conclusion: Craniosynostosis and raised intracranial pressure in a family with a genetic diagnosis of Frank-ter Haar syndrome expands the phenotype of the disease. The involvement of the sagittal suture in a mouse model of the disease suggests that the association is more than coincidental. In addition this study supports the evidence that raised intracranial pressure still occurs in non-scaphocephalic sagittal synostosis, and suggests that these patients could be at increased risk.

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HEMIFACIAL MICROSOMIA ASSOCIATED WITH A DUPLICATION OF 4P16

Presenter: Katrina M. Dipple, MD, PhD
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Hemifacial microsomia, HFM, is characterized by asymmetry of the face including unilateral hypoplasia of the maxilla or mandible and microtia. It can be part of oculo-aurico-vertebral syndrome (OAVS) or isolated HFM/craniofacial microsomia. The majority of cases are sporadic, and most familial cases are inherited in an autosomal dominant manner. We report a 14 month old male with congenital right hemifacial microsomia including hypoplasia of the right mandible, right lateral zygomatic arch, and lateral temporal bone; and grade three right microtia with complete absence of the right external auditory canal. He also has congenital heart disease (ASD, VSD) and grade 3 hypospadias. Chromosomal microarray revealed a 1.5 Mb heterozygous copy number gain at 4p16.1 - a region previously reported as important in craniofacial anomalies (Balikova et al., Am J Hum Genet 82, 2008). Updated mapping places the HMX1 gene in this region. Schorder et al. attributes a familial homozygous deletion of HMX1 to Oculo-Auricular Syndrome (Am J Hum Genet 82: 2008). Furthermore, mutations in rat (dmbo) and mouse (dmbo, mpe) affecting Hmx1 gene expression result in eye and ear malformations (Quina et al., Dis Model Mech, 2012). Our patient has a duplication in this region confirming that copy number variants of HMX1 are important in craniofacial development and OAVS. The patient's mother has a similar copy number gain in this region, but with normal ears and hearing. Further careful phenotyping of the mother is underway. This suggests that other genetic and/or environmental factors in the presence of the 4p16.1 duplication are contributing to the HFM. While the HMX1 gene lends explanation for the microtia seen in all three families, our patient is the only one with HFM. Unique to our patient's region is the CPZ1 gene, which modulates Wnt-4 signaling (Wang, et al J Bone Miner Res 24, 2009). Despite etiologic heterogeneity, these familial cases and genetic linkages can expand our understanding of the development of the first and second branchial arches in humans.



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CRANIOFACIAL ABNORMALITIES IN ASSOCIATION WITH THE 22Q11.2 DELETION SYNDROME: BEYOND CLEFTING

Presenter: Donna McDonald-McGinn, MS, CGC
Authors: McDonald-McGinn DM, Jackson O, Passick C, Low D, Bailey A, Jethava N, Schultz P, Forbes BJ, Binnenbaum G, Whitaker L, Zackai EH, Bartlett SP

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Introduction: The 22q11.2 contiguous gene deletion syndrome (22q11.2DS) is quintessentially a multisystem disorder involving haploinsufficiency of 30-40 genes and is most frequently associated with the triad of congenital heart disease, palatal abnormalities and immunodeficiency. Additional common findings include hypocalcemia, feeding/swallowing issues, renal problems, learning differences and psychiatric illness. Craniofacial teams are generally familiar with the presenting palatal phenotype, such as velopharyngeal dysfunction, overt cleft palate, and occasionally cleft lip and palate, but are unlikely to consider the diagnosis in the presence of less frequently occurring craniofacial anomalies. Here we report additional associated features, beyond clefting, which often result in referral to Plastic Surgery as the initial point of contact. Familiarity with such associations will likely advance detection of the underlying etiology; improve coordinated care and genetic counseling; and ultimately contribute to important genotype-phenotype correlations.

Methods: 1077 individuals with 22q11.2DS have been evaluated by the 22q and You Center at the Children's Hospital of Philadelphia since 1992. Uncommon craniofacial findings, such as craniosynostosis, were noted and have been reported previously (McDonald-McGinn 2005; Forbes 2007; Binnenbaum 2008; Digilio 2009). In addition, 836 records had suitable data to assess the presence or absence of asymmetric crying facies.

Results: Important infrequent (<10%) craniofacial findings included: bilateral and unicoronal craniosynostosis (4); Goldenhar syndrome/Hemifacial microsomia; ptosis (4) and sclerocornea (3). More prevalent, asymmetric crying facies was noted in 14% (117/836).

Conclusion: This study indicates that there are a number of significant craniofacial abnormalities, beyond palatal differences, found in association with 22q11.2DS which may well bring patients to initial attention in the Plastic Surgery/Craniofacial Clinic setting. Additional reports support this observation with 22q11.2DS and: craniosynostosis (Al-Hertani 2013); Goldenhar syndrome/Hemifacial microsomia (Ryan 1997; Derbent 2003; Xu 2008); sclerocornea (Casteels 2005);

and asymmetric crying facies (Bawle 1998). Thus, based on our data and in combination with the literature, we suggest that providers remain alert for findings classically associated with 22q11.2DS such as congenital heart disease, palatal anomalies, feeding difficulties, hypocalcemia, and chronic infection when patients present with craniosynostosis, Goldenhar syndrome/Hemifacial microsomia, ptosis, or sclerocornea. Moreover, caregivers should remain mindful of 22q11.2DS as a potential etiology when standard genetic studies are negative; for example when FGFR1, 2, 3 and TWIST mutational analyses are normal in a patient with craniosynostosis. Additionally, based on this evidence, we strongly recommend evaluation for associated features, such as congenital heart disease, as well as 22q11.2 deletion studies, as a first line of investigation in all patients with asymmetric crying facies. Lastly, rare findings in association with 22q11.2DS may well inform our understanding of the etiology of apparently isolated anomalies, via genotype-phenotype correlations, much like the recent report of four children with 22q11.2DS and uncommon features, including one with bilateral cleft lip and palate, and both a 22q11.2 deletion and a mutation in SNAP29 on the remaining 22q11.2 allele (McDonald-McGinn, 2012) hence providing a convincing argument for tracking and reporting atypical patients.



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PREVALENCE OF OBSTRUCTIVE SLEEP APNEA SYNDROME IN SYNDROMIC CHILDREN WITH CRANIOFACIAL ANOMALIES

Presenter: Linlin Gao, MD

Authors: Gao L, Paliga JT, Goldstein J, Cielo C, Marcus CL, Taylor JA

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Objective: The prevalence of obstructive sleep apnea (OSA) in syndromic children with craniofacial anomalies is unknown.

Methods: The Pediatric Sleep Questionnaire (PSQ) was developed to help identify patients with OSA. The survey includes questions related to snoring, daytime sleepiness and associated behavioral problems. A fractional score of 0.33 on the PSQ is used as a cutoff for detecting an $AHI > 5$. Parents of patients seen at the Children's Hospital of Philadelphia (CHOP) craniofacial clinic completed the PSQ as part of the clinical assessment. We reviewed the charts to determine which patients have also had polysomnograms.

Results: Over a period of 18 months, 156 consecutive pediatric sleep questionnaires were completed. The average fractional score was 0.23 ± 0.12 . We found a statistically significant difference in PSQ scores between patients with DiGeorge syndrome without VPI (0.21 ± 0.14) compared to with VPI (0.33 ± 0.14), $p < 0.01$. More DiGeorge patients with VPI screened positive for OSA (61%) using PSQ compared to those without VPI (13%). There were no statistically significant difference found between patients with Apert's syndrome, who had fractional scores of 0.41 ± 0.16 (mean \pm SD), compared with patients with Crouzon's syndrome (0.24 ± 0.22), $p < 0.10$, nor between patients with Goldenhar's syndrome (0.30 ± 0.20) vs. hemifacial microsomia (0.19 ± 0.14), $p < 0.26$, nor between patients with Pierre Robin Sequence (0.22 ± 0.15) vs. Treacher-Collins (0.24 ± 0.21), $p < 0.75$. A total of forty-four patients (28%) had a PSQ score consistent with OSA. Of these patients, ten patients had polysomnograms and three patients had apnea-hypoxia index (AHI) greater than 5/hour (30%). Of the patients who had negative PSQ screening, fifteen patients had polysomnograms and six patients had AHI greater than 5/hour.

Conclusions: In syndromic patients with craniofacial anomalies, a large portion surveyed positive for OSA using PSQ. OSA may be under diagnosed in this patient population. However, additional research is needed to validate PSQ as a screening tool for OSA.

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TIMING OF AIRWAY OBSTRUCTION IN INFANTS WITH ROBIN SEQUENCE

Presenter: Kelly N. Evans, MD

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Background: The clinical presentation of upper airway obstruction (UAO) in infants with Robin sequence (RS) varies. Anecdotal experience at our institution has led to heightened surveillance between 4-8 weeks of age. However, little evidence supports monitoring strategies and gaps regarding factors that predict severe or progressive UAO remain.

Methods: We conducted a retrospective review of 167 children (born 1990-2011) with RS and > 2 of the following: micrognathia; glossoptosis; secondary cleft palate; airway obstruction. We describe infants' characteristics, associated anomalies and diagnoses, UAO severity and treatments. Characterization of UAO (mild, moderate, severe) was determined by work of breathing, hypoxia and CO_2 retention. We explored associations between UAO severity and phenotypic characteristics (jaw severity, cleft shape), clinical characteristics (growth failure, concomitant feeding problems, gastroesophageal reflux), and perinatal characteristics (prematurity, pregnancy complications). We describe the trajectory of airway obstruction over the 1st 6 months of life, and the relationship to treatments received.

Results: Peak airway obstruction in the 1st 6 months was categorized as none (8%), mild (31%), moderate (26%), severe (31%), and peaked earliest (< 2 weeks old) in infants with severe UAO. Infants with severe UAO were more likely to have associated anomalies, severe micrognathia, dysphagia and tube feeding. While UAO improved in most infants, 13% had progressive UAO between 2-4 weeks. Among infants with no or mild UAO in the 1st month of life ($n=57$), 6 developed more severe UAO treated with nasopharyngeal airway (4), mandibular distraction (1) and tracheotomy (1). Compared with infants whose UAO peaked > 2 weeks, infants who peaked < 2 weeks were twice as likely to have surgery for airway management.

Conclusion: These data suggest the highest risk period for airway compromise in infants with RS is within the 1st 2 weeks of life. Prospective studies confirming predictors of severe or progressive UAO will inform monitoring strategies and enhance opportunities to reduce the sequelae of chronic airway obstruction.



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PROGRESSIVE HEMIFACIAL ATROPHY: A NEW FINDING

Presenter: Michael Friel, MD
Authors: Friel M, Havlik RJ
Indiana University

Introduction: Progressive hemifacial atrophy (PHA), or Parry-Romberg Syndrome, is a rare but well-known “acquired” facial deformity. The coup de sabre line of involvement is pathognomonic for this disorder. Despite genetic and molecular analysis, this disorders pathogenesis has escaped elucidation. Infectious, lymphovascular, and neural mechanisms have been postulated, but no single mechanism has been clearly established. The disorder’s most striking features may be attributable to soft tissue changes, including changes in skin, subcutaneous fat, and muscle, but boney and cartilaginous changes can contribute to the disfigurement. We report the first reported finding of a case of intracranial involvement of the dura mater in Parry-Romberg Syndrome, which has potential implications for pathogenesis.

Case Description: A 10 year old African-American male presented with a three year history of facial disfigurement. His findings included a classic coup de sabre involvement of the right forehead, brow, orbit and maxilla. The underlying brain was normal on CT scan. He underwent anterior cranial vault reconstruction as a component of his rehabilitation. His frontal bone flap was removed in standard fashion, and his dura mater was found to have an area of increased opacity directly beneath the line of the coup de sabre, consistent with intra-cranial involvement.

Discussion: To our knowledge, dura mater atrophy in Parry-Romberg Syndrome is a finding that has not been reported previously in the literature. In our patient, as with others with the syndrome, significant soft tissue and bony hemiatrophy of the face was present at time of evaluation. Atrophy of the zygoma, temporal, and frontal bone has previously been reported, but never the intracranial manifestations of the disease. While causality is neither implied nor demonstrated, the finding does have potential implications for the pathogenesis of the disorder. It is difficult to advance theories of infectious, lymphovascular or neural mechanisms of pathogenesis being transmitted through intact cranial bone. Implications of this finding will be discussed.

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FACIAL INFILTRATING LIPOMATOSIS IS CAUSED BY PIK3CA ACTIVATING MUTATIONS

Presenter: Reid A. Maclellan, MD
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Background: Facial infiltrating lipomatosis (FIL) is a rare, congenital, non-heritable, disorder characterized by hemifacial soft-tissue and skeletal overgrowth, precocious dental development, macrodontia, hemimacroglossia, and mucosal neuromas. It has been hypothesized that FIL is caused by a somatic mutation, with regional expression, that arose during embryonic development. The purpose of this study was to search for causative somatic mutations in patients with FIL by using massively parallel sequencing.

Methods: Human FIL tissue was obtained prospectively from 4 patients during a clinically-indicated procedure and stored frozen. DNA was extracted from these specimens to produce massively parallel sequencing libraries that were enriched for coding sequences from genes involved in pathways that control cell growth using targeted capture. We massively parallel sequenced the enriched libraries and analyzed the sequence data for mutations that appeared to be mosaic and unique to the affected tissue.

Results: We identified a different missense mutation in PIK3CA in each patient’s affected tissue. PIK3CA encodes the catalytic subunit of the enzyme phosphoinositide-3-kinase (PI3K), which promotes cell growth. One patient had a nucleotide transition that changed a histidine to an arginine codon at the amino acid residue 1047 (p.H1047R), the other patients had different amino acid mutations: p.H1047L, p.E453K, or p.E542K. Each mutation is predicted to significantly increase enzymatic activity. The frequency of mutant cells in the affected tissue ranged from 12% to 68%, compatible with their representing somatic mosaic rather than germline mutations.

Conclusions: Somatic mosaic mutations in PIK3CA cause FIL. Interestingly, similar mosaic mutations have recently been identified in patients with other overgrowth disorders including CLOVES syndrome, hemimegalencephaly, and segmental fibroadipose hyperplasia. PIK3CA inhibitors are currently in clinical trials for cancers containing PIK3CA mutations; they may be efficacious in patients with FIL and other PIK3CA-associated overgrowth syndromes.



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**PLAGIOCEPHALY, TORTICOLLIS, LAMBDROID
CRANIOSYNOSTOSIS: A SPECTRUM OF DISEASE.
A REVIEW OF 9,683 PATIENTS WITH POSTERIOR
PLAGIOCEPHALY**

Presenter: Robert Wood, MD
Authors: Wood R, Chibbaro G, McGrory M
Gillette Childrens Hospital

Patients with posterior plagiocephaly or torticollis seen by the senior author between July, 1999 and November 2012 were retrospectively reviewed. Data collected included diagnosis, treatment modality, treatment duration, age and sex.

9,683 patients with plagiocephaly were seen with a mean age of 7.4 months. A total of 5,792 patients with torticollis were seen during this time frame. Of the 9,683 patients with plagiocephaly, 5150 (53.2%) had torticollis. Physical therapy for torticollis averaged 146.6 days and 94.7% were treated with physical therapy alone. 327 (5.6%) were deemed refractory to therapy and underwent release of the sternocleidomastoid.

6,220 patients (64%) with plagiocephaly elected to receive an orthotic molding helmet. Helmet therapy averaged 96.4 days.

36 patients (0.4%) were noted to have lambdoid craniosynostosis. Of these, 30 (83%) had torticollis.

Conclusion: Plagiocephaly and torticollis are common diagnoses and so frequently related that with either diagnosis the other should be specifically sought. Most torticollis can be treated with physical therapy alone. Surgery for torticollis should be relatively rare and any child with refractory torticollis should first have their therapy reviewed. Orthotic molding helmets are an attractive option to the majority of families with deformational plagiocephaly when available.

Lambdoid craniosynostosis is very rare in the plagiocephaly population. This may have implications for how and by whom orthotic molding helmets are supplied in the future. Torticollis is normally present and should be specifically ruled out in lambdoid craniosynostosis patients. We feel persistent or undiagnosed torticollis is a frequent issue in the residual asymmetry and relapse seen in lambdoid craniosynostosis patients.

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**CLEFT AND CRANIOFACIAL CLINIC FORMATS IN THE
UNITED STATES: NATIONAL AND INSTITUTIONAL
SURVEY**

Presenter: Haruko Okada, MD
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Case Western Reserve University

Background: Craniofacial/cleft teams employ multidisciplinary clinics to optimize care for their patients. Different clinic formats exist and surveys were used to study family preferences and compare them to current U.S. national practices.

Method: An institutional survey was distributed to families of our team clinic patients from November 2012 to February 2013. The survey was given after a clinic format change from patients moving between rooms to see providers, to providers moving between rooms to see patients. This survey focused on patient satisfaction, preference of clinic format, and their perception of efficiency. A second, national survey was distributed to 161 American cleft/craniofacial teams approved by the American Cleft Palate–Craniofacial Association. This survey queried clinic formats, provider satisfaction, and experience with other formats.

Results: 39 out of 54 (72.2%) families responded to the institutional survey. Providers moving between rooms while patients remained stationary was positively associated with satisfaction (mean 4.97 out of 5, 5 being most satisfied), shorter perceived clinic time (76.9%), and patient comfort (84.6%). The national survey had 91 responses out of 161 (56.5%). 54.9% of respondents have providers rotating between exam rooms, and 32.9% have patients moving between rooms. Other formats include entire team moving as a group between rooms (10.99%), and specialties sitting together in one room while patients rotate (8.97%). Respondents were satisfied with current formats (mean 4.24 out of 5, 5 being most satisfied). 21.5% had tried a different format previously.

Conclusion: The most common American cleft/craniofacial clinic format is providers moving between rooms, however all formats have high provider satisfaction. At our institution, there is high family satisfaction rate where providers move between rooms and this is preferred to patients moving between rooms. Patients cite increased privacy and efficiency as reasons for this preferred format.



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TELEMEDICINE IN CLEFT CARE: THE EXPERIENCE AT SHRINERS HOSPITAL FOR CHILDREN, LOS ANGELES

Presenter: Melinda Costa, MD

Authors: Costa M, Gillenwater JG, Taghva GT, Green TG, Magee WM

Shriners Hospital Los Angeles

Background: This hospital cares for patients from remote areas. Conventionally, patients travel to the primary clinic or are evaluated in outreach clinics. Video conferencing allows for evaluation of patients pre- and postoperatively, decreasing the need for travel by families and the cleft team.

Methods: A retrospective review of cleft patients evaluated via video conferencing from 2007 – 2012 was conducted. The number of telemedicine visits, clinic sites, and patient demographic information were collected. A postoperative protocol was developed for primary cleft lip and palate repairs and revision surgery.

Results: Over the five-year period, 519 cleft patients were evaluated. 94 (18.1%) had at least one teleconferencing visit. Of these patients, 36 (38.3%) had a diagnosis of unilateral cleft lip and or palate, 21 (22.3%) bilateral cleft lip and or palate, 18 (19.1%) isolated cleft palate and 19 (20.2%) isolated cleft lip. They attended 7 primary clinic sites for the teleconferences. The postoperative regimen for primary cleft lip and nasal repair included an overnight stay to avoid long distance travel the day of surgery. Nasal stents were evaluated on postoperative day 1 to confirm positioning. A phone call was made at one week. Patients were seen at 4 weeks postoperatively via telemedicine for nasal stent removal and again via telemedicine to plan for palatoplasty. Patients undergoing primary palatoplasty stayed overnight and received a phone call at 1 week, then seen via telemedicine 3 weeks postoperatively prior to advancing the diet. At 6 months, patients were seen in person at an outreach clinic to evaluate for complications and were subsequently seen annually for speech evaluation via telemedicine. All patients were successfully managed postoperatively using teleconferencing.

Conclusions: Videoconferencing can be used in the perioperative care of patients who live long distances from a cleft center. Telemedicine is a reliable resource for resource for health care delivery that can decrease the burden on families, allowing the surgeon to effectively practice regionally without compromising care.

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POSITIONAL PLAGIOCEPHALY: EXPERIENCE WITH A PASSIVE ORTHOTIC MATTRESS

Presenter: Sasha Burn

Authors: Bruce S, Quirk D, Sinha A, Burn SC, Richardson D, Vaiude P, Duncan C

Alder Hey Hospital for Children

Introduction: Positional plagiocephaly (PP) is the commonest head shape deformity which presents to craniofacial units. Management options include advice and repositioning, orthotic mattresses and helmet therapy, the latter being the most expensive.

Objective: To document changes in head-shape associated with use of a passive orthotic mattress for PP.

Methods: A consecutive case series of 40 patients were managed with the orthotic mattress. Anthropometric head measurements were recorded using Bertillon callipers and Cranial Vault Assymetry (CVA) was calculated and recorded before treatment and after 6 months of follow up.

Results: 26 males and 14 females with a mean age of 4.5 months were entered into the study. Mean pre-treatment CVA was 16 mm (range 4-43 mm). Mean post-treatment CVA was 5 mm (range 0-18 mm) ($P < 0.05$). There were 10 patients who had torticollis as part of their initial presentation. A similar improvement in CVA to the non-torticollis group occurred in these patients. There were no complications and compliance was 100%.

Conclusion: This observational study demonstrated a significant improvement in CVA of PP in patients aged less than 6 months treated with an orthotic mattress. This occurred irrespective of whether the patients had torticollis or not and the improvement was equivalent to what has been reported with helmet therapy. Other advantages of the orthotic mattress included high compliance and parent satisfaction combined with low cost.



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DEVELOPMENT OF A PATIENT REPORTED OUTCOME MEASURE IN ADULT CLEFT LIP AND PALATE PATIENTS

Presenter: Sophie Ricketts, MD
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Background: Conventional methods of evaluating treatment outcomes in the cleft lip and palate (CLP) patient group have been objective measures determined by practitioners. In an effort to evaluate quality of life (QOL), patient reported outcome (PRO) measures are gaining increasing attention as valid measures of surgical efficacy. It is now accepted that systematic development should be employed when generating PRO measures to ensure reliability and validity. This process of development involves three phases; 1. Item generation, 2. Item reduction and 3. Psychometric evaluation. Adult CLP (as distinct from children or adolescents) encounter varied psychosocial and vocational QOL issues associated with their cleft deformity. This may prompt them to seek secondary revisionary surgery. Outcomes of this surgery in relation to QOL can be assessed with a PRO. We outline Phase 1 of PRO development; the generation of a set of scales that measures outcomes after surgery in this patient group.

Methods: Phase 1 in the development of a PRO measure includes literature review, patient interviews and expert panels. The literature review identifies existing questionnaires or PROs that help to formulate an interview guide. Patient interviews allow issues important to the group to be explored in detail. These are recorded and transcribed before being coded for generation of a pool of items or questions for inclusion in the PRO. The expert panel then reviews the pool of items and ensures all concepts considered important are included.

Results: Results of Phase 1, or Item Generation will be presented.

Conclusion: There is currently no specific validated PRO measure that addresses the issues important to the adult CLP patient population who often seek secondary revisionary surgery for reasons that may differ considerably from the paediatric and adolescent populations. We place considerable worth in the development of such a measure according to the rigorous guidelines now accepted. This will therefore ensure scientific soundness and confidence in the evaluation of interventions and outcomes in the adult CLP patient group.

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“JUST BECAUSE SOMEONES A DOCTOR DOESN’T MEAN I JUST TRUST THEM”: ENHANCING PATIENTS’ DECISION MAKING IN ELECTIVE RECONSTRUCTIVE SURGERY

Presenter: Daniela Hearst, BSc, MPhil
Authors: Hearst D, Mayers S, Liddle M, Thomas S, Shanly A, McKerracher G, Horton J, Knapp M, Kearney A
Great Ormond Street Hospital for Children Foundation Trust

Background: Clinical observation indicates that adolescents, particularly those with syndrome related cognitive impairment frequently experience difficulties formulating and communicating their wishes and fears surrounding facial change. Clinic structure and quality of communication are key elements in the decision making process for adolescents being offered surgery to improve facial appearance. Clinicians need a better understanding of the decision making process, how they can assist with this and improve management of patient expectation. A preliminary survey of parents and children aged 7-18, using 4 point Likert scales, conducted over 4 months in 2012 yielded 170 responses with 2 consistent themes: patients wanted more information about their treatment plan and more detail about who was in the consulting room. Subsequently a study was conducted across all 4 UK supraregional craniofacial centres to investigate the decision making process and what constitutes a “good” outpatient experience.

Methodology: 42 patients, aged 10-20 years, with both syndromic and nonsyndromic diagnoses, participated in 4 focus groups and individual interviews. Additionally 46 parents and guardians took part in 4 adult focus groups and individual interviews. Conversations were recorded and Interpretive Phenomenological Analysis was used to analyse the data.

Results: Adolescents wanted their surgeons to use simpler verbal descriptions and more visual aids. They want to be involved from a younger age and take more responsibility for decisions. Internet blogs and forums become increasingly important with age as does the possibility of meeting others who have undergone surgery. The clinic environment is key to discussions; the presence of many professionals is intimidating. Adolescents are very concerned that if they show any sign of hesitation or doubts, the choice of surgery will be removed from them. Parents’ responses were similar; they wanted more preparation to adapt to their child looking like a different person after surgery.

Conclusions: More visual aids and facilitator led support groups will be introduced into clinic practice.



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MENTAL NERVE PATHWAY BEFORE MENTAL FORAMEN FOR GENIOPLASTY

Presenter: LunJou Lo, MD

Authors: Kim HY, Kim SG, Huang CS, Chen YR, Lo LJ
Chang Gung Memorial Hospital

Background: The mental nerve is purely related to the sensation of the lower face. During genioplasty, incidence of mental nerve injury is reported to be about 10%. Previous studies have revealed that the area 5mm below the mental foramen is safe, but sometimes it may not be enough. This study was planned to evaluate the course of the mental nerve around the mental foramen using 3 dimensional analysis with iCAT Vision software, and to compare the difference between Class I, Class II, Class III and cleft patients.

Methods: From 2007 to 2011, a total of 101 cone-beam computed tomography (CBCT) data (202 hemimandibles) was evaluated. Using iCAT Vision, we performed analysis of the mental nerve pathway around the mental foramen.

Results: There were no differences between the left and right side, or between different patient groups. In about 25% of all cases, no loop was observed, while the majority of CBCT data reveals an anterior loop. The range of distance between the nerve and the mental foramen was from 0.8mm to 10.8mm. With the 5mm safety margin, only 63.86% of patients are risk-free from mental nerve injury.

Conclusion: Preoperative CBCT evaluation can be a good method of screening before genioplasty. If the CBCT is not available, a safety margin of at least 8mm below the mental foramen is rendered safe in about 98% of patients.

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AESTHETIC REFINEMENTS IN THE TREATMENT OF GRAVES' OPHTHALMOPATHY

Presenter: Gaby Doumit, MD, MSc

Authors: Doumit G, Yaremchuk M
Cleveland Clinic

Graves' ophthalmopathy is a chronic, multisystem, autoimmune disorder characterized by increased volume of intraorbital fat and hypertrophic extra-ocular muscles. Clinical findings of proptosis, impaired ocular motility, diplopia, lid retraction, and impaired visual acuity are treated with orbit decompression and fat reduction. We present the addition of skeletal augmentation to further improve periorbital aesthetics.

Through a transconjunctival with lateral canthotomy incision, a balanced orbital decompression was executed removing the medial and lateral walls, and medial floor. Intraorbital fat was excised. All patients underwent placement of porous polyethylene infraorbital rim implants and midface soft tissue elevation increasing floor projection and improving globe-cheek relationship. From 2009-2012, 13 patients (11 females, 2 males, 26 eyes) with Graves' ophthalmopathy underwent surgery at two institutions. Outcomes were evaluated, specifically for improvements of proptosis, diplopia, dry eye symptoms, and cosmetic satisfaction.

Postoperative follow up ranged from 0.5-10 yrs (median 1.5 yrs). The mean improvement in Hertel exophthalmometer was 54.1 mm. Diplopia resolved in 3/13 (23%) cases. No patients had worsening diplopia, and 12/13 (92%) discontinued use of eye lubricants. All patients had cosmetic satisfaction. One patient suffered temporary paresthesia to the inferior orbital nerve. There were no infections, hematomas or ocular complications.

Skeletal augmentation is a useful adjunct to orbital decompression and fat excision for treating Graves' ophthalmopathy. Balanced orbital decompression with infraorbital rim implants are reliable, effective, and safe, with good, lasting results. Improvements are made towards the resolution of ocular symptoms and in the patient's personal well-being and social life, with a high benefit-to-low-risk ratio.



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COSMETIC AND FUNCTIONAL TREATMENT OF GRAVES EXOPHTHALMOS AND RELATED EYELID MALPOSITION USING SELECTIVE OSTEOTOMIES

Presenter: Henry M. Spinelli, MD
Authors: Spinelli HM, Sackeyfio R
Weill Medical College of Cornell University

Introduction: The most common underlying condition associated with exophthalmos is Graves disease. Graves presents with a spectrum of infiltrative disorders. Ranging from eyelid retraction to severe exophthalmos with corneal exposure and even optic nerve compression. The later should not be treated with simple gross fractures into sinuses and/or soft tissue “masking procedures”. Ideal treatment of graves should address bone and soft tissue concomitantly with a single procedure.

Methods: We present a series of 26 patients (14 Bilateral) treated over 10 years (2001-2011), using extraanatomic zygomatic osteotomies selective medial and orbital floor resection and lateral retinacular tightening. The relevant bone and soft tissue anatomy, technical nuances and theoretical considerations of the procedure will be described based on cadaveric skull and clinical dissections.

Results: All patients had significant correction of their exophthalmos averaging 6.8mm by Hertel. No visual loss, diplopia or other major complications were noted. All had markedly improved eyelid position and corneal coverage. All 26 patients noted significant cosmetic and functional improvement; confirmed by independent examination.

Conclusion: Selective well executed supra anatomic osteotomies are the procedure of choice for moderate to severe orbital Graves disease and satisfactory correct exophthalmos and eyelid malpositions in a cosmetic fashion.

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CORRECTION OF PROMINENT MALAR WITH FIBROUS DYSPLASIA BY L-SHAPE OSTEOTOMY

Presenter: Feng Niu, MD
Authors: Niu F, Gui L, Liu JF, Song T
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Objective: To present a new method for correction of prominent zygomaticomaxillary complex with fibrous dysplasia by L-shape osteotomy through an intraoral incision.

Methods: There were 23 patients with zygomaticomaxillary complex fibrous dysplasia, which resulted in asymmetrical midface deformity. Based on the anatomical characteristics of the malar complex, we designed a ‘L’-shape osteotomy for malar eminence reduction. The procedure includes oblique osteotomy of the upper part of the malar, vertical osteotomy of the anterior part of the mala and “greenstick” fracture of the zygomatic arche. According to the severity of malar prominence, we resect part of the anterior-inferior part of the malar and lower the malar complex by “L”-shape osteotomy. Internal fixation with small splint and excision of partial hyperplastic bone were performed.

Results: 23 cases with zygomaticomaxillary complex fibrous dysplasia resulting in asymmetric deformity were surgically treated. All patients obtained satisfactory aesthetic and functional results after treatment.

Conclusion: “L”-shape osteotomy for correction of prominent malar complex with fibrous dysplasia is a relatively ideal surgical method with the advantages of simpler manipulation, fewer complications, better result, and ensuring the intactness of the structural characteristics of the malar complex.

Keywords: Prominent malar; fibrous dysplasia; L-shape osteotomy; Intraoral approach



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IMPLEMENTING FAT GRAFTING IN MANAGING DIFFICULT CRANIOFACIAL PATIENTS

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Fat grafting has been serving as the primary procedure or an adjunct to other procedures in facial aesthetic surgery. In reconstructive surgery, several techniques were used to correct soft tissue deficiency with variable degrees of successes and problems. Attempts of using fat grafting have been limited to hemifacial atrophy cases. Its application to the more complex deformities with skeletal deficiencies is limited in the literature. This work emphasizes the implementation of fat grafting in the management of difficult craniofacial cases. Coleman's principles for structural fat grafting were used to replenish volume in different facial aesthetic units of 43 patients with facial soft tissue deficiencies &/or skeletal deficiencies. 5 patients had only soft tissue deficiencies and they all belonged to the hemifacial atrophy category and received only fat grafting. The remaining 38 patients had both skeletal & soft tissue deficiencies and included repaired cleft lip & palate, Treacher Collins syndrome, frontal plagiocephaly, anophthalmic sockets, hemifacial microsomia, and jaw deformity cases. With the exception of 5 patients whom declined bony work & underwent only fat grafting as a camouflage tool, the remaining received fat grafting after establishing their bony framework. Individualized planning was formulated for each patient to achieve realistic aesthetic goals. The results of this work demonstrated 89% high satisfaction rate of patients up to three years follow up. Fat grafting offered simple, natural, and highly predictable outcomes. It restored volume and enhanced the results in patients with different craniofacial deformities with overall regain of facial balance. Fat grafting can be implemented efficiently into the difficult and complex craniofacial patients. Its use becomes a logical approach to treat cases with only soft tissue deficiencies with the ideal goal being "replacement of like with like", cases with combined skeletal & soft tissue deficiencies after establishing the bony foundation to enhance on aesthetic outcome upon the bony work, or as a camouflage tool for cases declining major surgeries.

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INFLUENCE OF THE MAXILLARY SINUS EXPOSURE AND PARTIAL MASSETER RESECTION IN AESTHETIC FACIAL CONTOUR SURGERY

Presenter: Jie Yuan, MD, PhD

Authors: Yuan J, Yu ZY, Xu L, Zhang Y, Wei M
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The oriental face is usually with a prominent zygomatic body and arch. As an oval face is considered ideal, reduction malarplasty is always requested by oriental people. Currently, L-shaped zygomatic osteotomy is often reported in literature yet no previous study has documented the exact outcome of this approach after exposure of the maxillary sinus. The mandibular angle also plays an important role in determining the asian facial attractiveness. Mandibular angle osteotomy is often reported as the correction of square face. However, no previous study has documented the long-term outcome of partial masseter muscle resection along with reduction mandibular angle.

Objective: The study was to investigate the outcome and complications of intraoral L-shaped zygomatic osteotomy with the opening of maxillary sinus. Further, To demonstrate the long-term effect of partial masseter muscle resection along with reduction mandibular angle.

Method: 101 patients undergone reduction malarplasty were subjected to computed tomography before and 12 months after surgery. And 151 patients undergone partial masseter muscle resection along with reduction mandibular angle were also subjected to computed tomography before and 3 years after surgery. All parameters were obtained with morphometric analysis for clinical evaluation, so as to reveal the change in maxillary sinus, the change of masseter muscle, postsurgical results, and complications.

Results: Both group patients were satisfied with the optimal outcome post-operation. Morphometric analysis indicated that the volume and surface area of maxillary sinus statistically significantly changed before and after surgery as well as the change of masseter muscle volume. Complications included deeper nasolabial fold, infraorbital nerve injury for reduction malarplasty, and swelling, low lip numbness for reduction mandibular angle and partial masseter muscle.

Conclusions: Intraoral L-shaped osteotomy of the zygoma, partial masseter muscle resection along with reduction mandibular angle can improve the appearance of the patients greatly. Both methods caused only a few complications.



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**CUSTOMIZED 3D MODELING OF AUGMENTATION
MENTOPLASTY**

Presenter: Robert Guryanov, MD

Author: Guryanov R, Guryanov A

Sechenov First Moscow State Medical University

Abnormalities in the development of the facial bones may be reason for seeking plastic surgery. Violation of the classical proportions of the face in the form of underdevelopment of the chin gives a characteristic appearance. Young people with such deformities feel unhappy with their appearance and are often not socially adapted. At this age, orthodontic treatment does not give the desired results and orthognathic surgery appears to be too traumatic. The use of chin implants in these patients allows us to solve the problem of harmonizing the face. Because of the high aesthetic requirements to the face, patients want to get an idea of his/her new appearance before surgery. Since the human face is inherently three-dimensional, finding a common platform for a visual dialogue between doctor and patient is not a trivial task. Further, use of standard chin implants is very practical, but still often requires fitting them in place because of non-compliance of the lower jaw, frequent asymmetries, bony protrusions, sharp transitions between the implant and the surface of the jaw. Reliable fixation of prostheses is problematic, since mismatch between bone and prosthesis profiles may lead to undesirable shifts. A lot of empirical experience is required to select the appropriate size and shape of the implant and correctly predict the future shape of the chin. In contrast, customized implants can be optimized in shape and size, they do not require adjustment during the operation, can simply be aligned with bones and rarely move. Here we present an overall scheme for 3D modeling and fabrication of chin implants with the predefined impact on skin contours. Virtual models of the patients anatomy are generated on the basis of CT and optical surface scans. Changes of skin contours are simulated using the Finite Element modeling of soft tissue mechanics. The simulation results are evaluated by direct comparison with 3-6 months postsurgery data. From our experience, application of 3D modeling dramatically increases quality assurance by augmentation mentoplasty as well as the patients satisfaction with the aesthetic surgery outcome.

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**ANALYSIS OF ADULT FACIAL SKELETAL CHANGE
WITH AGE**

Presenter: Richard A. Levine, MD,DDS

Authors: Levine RA, Wang PT, Garcia A

University of Texas Health Science Center at San Antonio

NOT PRESENTED



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FAT GRAFTING IN THE CRANIOFACIAL PATIENT

Presenter: Jorge E. Cabrera, MD

Authors: Cabrera JE, Aguilera A

Hospital General Dr Manuel Gea Gonzalez

Grafted fat tissue has been used for years as an excellent filler to treat volume and contour deficiencies in aesthetic and reconstructive surgery. Patients with congenital craniofacial malformations and trauma sequelae present with complex challenges for reconstruction. Individualized treatment often involves rebuilding of the facial skeleton, as well as correcting the overlying soft tissue deficiencies in final stages. Serial autologous fat grafting performed during staged reconstruction is the preferred method in the craniofacial clinic at General Hospital Dr. Manuel Gea Gonzalez. 97 patient retrospective study with craniofacial malformation diagnosis including Parry Romberg Sd. (n=36), craniofacial Microsomia (n=13), Facial clefts (n=17), Non syndromic (n=10), Syndromic craniosynostosis (n=8), and trauma sequelae (n=13) treated by fat grafting between March 2010 and March 2013. The most common donor sites were the upper abdominal area and groin. The fat was harvested through the same incisions made for infiltration of anesthetic solutions of 1:200,000 of epinephrine in Ringer's Lactate. A blunt tip harvesting cannula was 2mm in diameter and 15cm in length, connected to a 10-mL syringe. Preparation was performed media washing using Ringer's solution. Application of the graft was made with a 17-gauge blunt cannula connected to a 1-cc syringe and using a "fanning out" technique in the supraperiosteal, intramuscular and superficial planes. The mean overall age was 24.5 years. Analysis was conducted on standardized pre and Postoperative photographs to determine facial symmetry and correction of soft tissue defects. The mean procedures overall was 2.3 and the time period between each one was 6 months. The average quantities of injected fat per facial area was 5.3 Cm³. Malar, Temporal and forehead regions were the most grafted. Based on the HGDMGG experience there is an important improvement in facial morphology, shape and volume in patient's appearance. facial asymmetry and contour correction with fat grafting is attainable and must be individualized according to patient needs and characteristics.

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STRUCTURAL FAT GRAFTING OF THE CRANIO ORBITO FACIAL AREA VOLUMETRIC & MORPHO-AESTHETIC IMPLICATIONS

Presenter: Riccardo Tieghi, MD

Authors: Tieghi R, Clauser LC

S Anna Hospital and University of Ferrara Italy

Keywords: Lipostructure, fat grafting, regenerative medicine, stem cells, facial reconstruction

Summary: The authors overview the application of structural fat grafting in the management of volumetric deficit of the cranio-orbito-facial area in congenital craniofacial patients (Crouzon, Apert, Plagiocephaly, Hypertelorbitism, Treacher Collins, Parry-Romberg syndrome).

Introduction: Autologous transplantation of fat tissue is not a new technique. Structural fat grafting was introduced as a way to improve facial aesthetics, and in recent years has evolved into applications in cranio-orbito-facial reconstructions.

Methods: The fat is aspirated using very thin liposuction cannulas attached to a 10 cc syringe. The fat must be gently woven in several layers to pick up a blood supply, allowing the fat cells to survive and maintain the filamentous architecture. The fat is slowly absorbed by the body, although the amount of reabsorption is sometimes unpredictable; however, this percentage varies from patient to patient. If a significant amount of fat is reabsorbed, a second or third procedure may be considered to improve the final result. In fact, using more fat tissue in a single-step corrective procedure can cause poor vascularization and more resorption, particularly in areas covered by a thin layer of soft tissue, such as the maxillofacial area.

Results: The primary indication for structural fat grafting in the cranio-orbito & maxillofacial area are for restoration and reshaping of different facial and craniofacial defects. Recent applications include the correction of localized tissue atrophy, loss of substance due to trauma, post tumor and congenital complex orbito craniofacial deformities, burns and hemifacial atrophy such as Romberg syndrome and scleroderma.

Conclusion: Fat grafting can be an excellent means for facial and craniofacial volumetric restoration. Recent studies have proved that human adipose tissue represents a rich source of mesenchymal stem cells. The Authors analyze indication, surgical technique and details and patients with a long term follow-up.



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A PRELIMINARY REPORT ON THE USE OF ANTIBIOTIC-IMPREGNATED METHYL METHACRYLATE IN SALVAGE CRANIOPLASTY

Presenter: Anthony Wilson, MD

Authors: Hsu VM, Grady MS, Taylor JA
University of Pennsylvania

Background: Many traumatic brain injury patients require the removal of calvarial bone to alleviate elevated intracranial pressure. Current practice is to replace the bone flaps when no further intracranial interventions are planned. These devascularized bone flaps can become colonized and develop osteomyelitis, necessitating their removal. These patients then have variable rates of success with subsequent alloplastic cranioplasty.

Objective: The purpose of this study is to investigate the use of antibiotic-impregnated methyl methacrylate for salvage cranioplasty in patients who have failed prior reconstruction.

Methods: The authors performed a retrospective review of a single surgeon's experience in vancomycin and tobramycin-impregnated methyl methacrylate for salvage cranioplasty. Vancomycin (1 gm) and tobramycin (1 gm) were mixed in methyl methacrylate, which was then applied to a rigidly-fixed titanium mesh for reconstruction (MMT). The indications and outcomes of this technique were evaluated.

Results: Eight patients (5 males, 3 females, mean age: 51 years) underwent vancomycin and tobramycin-impregnated MMT reconstruction for salvage cranioplasty. The indications for initial craniotomy were bleeding due to trauma, ruptured aneurysm, or anticoagulation, and tumor resection. On average, these patients underwent 4.1 procedures (range: 1-15), including repeat craniotomy, debridements, and failed reconstruction over the course of 3.9 years (range: 7 months - 14 years) prior to salvage cranioplasty. All patients required salvage cranioplasty due to infection. The most common bacteria isolated in culture were MRSA, P.acnes, and Enterobacter. The average size of the craniectomy defect was 130 cm². In early follow-up after MMT (mean duration: 3.5 months), there were no incidences of postoperative infection or need for revisions.

Conclusions: Vancomycin and tobramycin-impregnated MMT reconstruction is a technique that can be used successfully in salvage cranioplasty. Our early report represents a "proof of concept"-the true test is whether these short-term successes translate to long-term stable results.

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TITANIUM CRANIOPLASTY IN PAEDIATRIC CRANIOFACIAL SURGERY - THE BIRMINGHAM EXPERIENCE

Presenter: Anuradha Venugopal, MD

Authors: Dover S, Lloyd M, Ahmad F, Bhatia S, Evans M, White N, Solanki G, Rodrigues D, Nishikawa H

Birmingham Childrens Hospital UK

Introduction: Calvarial defects can be congenital or acquired. Various materials have been used over the years to cover or fill these defects including autologous bone, acrylic, metals (including titanium) and synthetic materials. We present our experience of the use of titanium cranioplasty plates in children at our craniofacial unit.

Methods: A retrospective review was undertaken to identify patients in whom titanium cranioplasties were performed over a 13-year period. Demographics, indications, outcomes and complications were recorded.

Results: Within our paediatric practice, 20 patients underwent titanium cranioplasties over this period. Complete data were available for 15 of these cases. The average age for titanium cranioplasty was 9 years (4-15 years). Nine cranioplasties (60%) were performed for defects following calvarial remodelling procedures. In 3 patients, the procedure was required following traumatic skull injury and 3 cases were performed for congenital parietal foramina. In 2 cases, autologous bone had initially been used as a cranioplasty but had resorbed. There were no intra-operative complications. There was one post-operative infection that required washout only. There was no clinical or radiological evidence of skull growth problems associated with the plates on mean follow-up of 4.2 years (1-12 years) and none of the cranioplasties have had to be removed to date.

Conclusion: This technique avoids the need for a bone graft, is relatively cheap, quick, reliable and effective. The procedure has evolved over the years and can include 3DCT scanning and CAD/CAM derived models. The custom designed plates now have multiple flanges and screw holes, allowing for a flexible approach to plate placement and improved results. The plate fabrication, technique, results and complications will be presented.



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ORBITAL FLOOR FRACTURES: AN OUTCOMES STUDY OF RECONSTRUCTION IN 364 CASES

Presenter: John B. Turner, MD
Authors: Turner JB, Kirby EJ, Vasconez HC
University of Kentucky

Objective: There has been considerable debate regarding the optimal management of orbital floor fractures. A retrospective chart review of patients undergoing surgery for orbital floor fractures was performed including 317 patients and 364 orbital floor fractures.

Methods: We reviewed the mechanism of injury, classification of fracture, and material used for reconstruction. We focus on outcomes of treatment comparing autologous bone reconstruction with alloplastic materials for reconstruction and examine the repercussions of negative exploration of the orbital floor.

Results: Indication for surgery were floor defect of >1 cm², entrapment of orbital contents, early enophthalmos or repair of associated injuries. Surgery was performed on average 8.6 days after injury. Mean follow-up was 38.8 weeks.

Alloplastic material was used in 169 patients and 71 repaired with autologous bone graft. Alloplastic materials demonstrated no statistically significant differences to bone graft in reoperation (20% vs. 24% $p=0.44$), enophthalmos (14% vs. 24% $p=0.11$), restriction of motility (3% vs. 7% $p=0.32$) and diplopia (14% vs. 17% $p=0.31$). Alloplastic materials demonstrated superiority in the category of orbital dystopia (6% vs. 14% $p=0.04$). Both materials were equivalent in removal for infection (6% vs. 6% $p=0.93$). No fracture was identified in 5.2% of explorations and 22.5% ($n=82$) warranted no repair. At >4 weeks from operation 36.8% of this group noted clinically significant complications including diplopia (21.1%), enophthalmos (10.6%), orbital dystopia (10.5%), persistent paresthesia (15.8%), chronic pain (21.8%), restriction of motility (10.5%), and entropion (5.3%). Reoperation rate for this non-interventional subgroup was 19.2%.

Conclusion: This study is the largest recent comparison of modern materials used to repair orbital floor defects. This study has shown newer alloplastic materials are comparable and in some ways superior to autologous bone which has long been regarded as the gold standard in reconstruction. In addition negative explorations are not without significant complications.

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RECONSTRUCTION OF MAJOR CALVARIAL DEFECTS USING AUTOGENOUS MATERIAL

Presenter: Ramesh K. Sharma, MD
Author: Sharma RK
Postgraduate Institute of Medical Education and Research

Background: Major calvarial defects can result following treatment of head and neck trauma or excision of tumors. The resultant defects need to be reconstructed both for aesthetic and functional needs. The conventional teaching is that we can repair only relatively small defects with autogenous bone and larger defects necessitate use of some alloplastic material. The authors have challenged this tenet and have reconstructed large defects with autogenous sources using in-situ split cranial bone graft and occasionally supplemented with rib grafts.

Materials and Methods: A total of 60 large defects have been treated in the last 12 years. The grafts were immobilized with minimum hardware. The age of patients varied from 6 months to 55 years. A long term follow up of these patients is discussed. The site of split cranial bone graft was decided depending upon the contour of the recipient side and on many occasions crossed midline. Large size grafts were harvested using contouring burr and osteotome.

Results: The bone grafts survived in all but one patient who had postoperative infection following exposure of graft because of tight envelope. The follow up in children shows that the reconstructed area has grown proportionately maintaining the contours and symmetry. The long term follow up of the donor site confirmed no morbidity associated with the donor site.

Conclusion: Autogenous bone is the best source for reconstruction of cranial bone defects. Almost all defects of any size can be managed with autogenous bone sources.



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POROUS HYDROXYAPATITE CUSTOM MADE CRANIOPLASTY IN 18 CHILDREN

Presenter: Gregoire Pech-Gourg, MD

Authors: Pech-Gourg, Di Rocco F, Arnaud E, Puget S, Zerah M, Scavarda D, Sainte-Rose C

Childrens Hospital La Timone

Custom made implants in porous hydroxyapatite seem to be a good solution for cranioplasty. Their supposed property to biocompatibility by bone integration is of major interest in children cases. A retrospective study of children who had undergone custom-made porous hydroxyapatite bioceramic cranioplasty in two french departments of pediatric neurosurgery between 2005 and 2012 was carried out. 19 patients were reviewed. The children's ages ranged from 4 to 16 with a mean age of 10.8. There were 3 girls and 16 boys. The causes of the bone defects were decompressive craniotomy, head trauma with comminute fracture, failure of previous cranioplasty, reabsorption of the autologous bone flap, cranial vault tumors. Preoperatively, all the patients underwent a three-dimensional (3D) CT scan. Custom made devices were obtained using 3D stereolithography. A model of the patients skull with the implant was manufactured in order to be accepted by the surgeon. The definitive porous hydroxyapatite bioceramic was then produced. The following criteria were recorded: sex, age, etiology for skull defect, time since craniectomy, dimensions of the implant, difficulties encountered during surgery, wound healing, cosmetic result, complications, clinical and CT scan follow-up. One asymptomatic fracture of a bioceramic appeared during the follow-up, secondary to a minor trauma. We highlight the difficulties to demonstrate skull bone integration in a hydroxyapatite cranioplasty.

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PATIENT-SPECIFIC ORBITAL IMPLANTS: DEVELOPMENT AND IMPLEMENTATION OF TECHNOLOGY FOR MORE ACCURATE ORBITAL RECONSTRUCTION

Presenter: Oleh Antonyshyn, MD, FRCSC

Authors: Antonyshyn O, Mainprize J, Edwards G
Sunnybrook Health Sciences Centre University of Toronto

Precise reconstruction of both the volume and geometry of the orbital cavity is of critical importance in the correction of post-traumatic enophthalmos. However, restoration of pre-injury anatomy is virtually impossible. Limited visualization, as well as the complex morphology of the deep orbit, defy the surgeon's ability to shape or place an implant with any degree of accuracy. This paper describes a novel technique which is specifically designed to allow intraoperative shaping of a patient-specific implant, and to guide implant placement within the orbit, to restore pre-injury orbital anatomy with unprecedented accuracy. The method comprises the following steps: 1. Computer-assisted modeling of the missing orbital geometry based on the anatomy of the uninjured contralateral orbit. Algorithms which facilitate segmentation of the thin bony walls of the orbit are employed. 2. Rapid prototyping of a patient-specific orbital mold and forming tool. 3. Intraoperative molding of a patient-specific orbital implant with built-in registration features to ensure anatomical positioning of the implant within the orbital cavity. The clinical application of the technology is demonstrated in a pilot series of patients with unilateral primary orbital fractures, and established post-traumatic enophthalmos. Results are assessed in terms of restoration of symmetry in orbital morphology. All patients underwent CT imaging preoperatively and postoperatively. Symmetry is assessed in terms of globe position and projection, orbital volume, and orbital shape as determined by registration of the operated and control orbits and distance mapping of corresponding surface topography.



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A COST-BENEFIT ANALYSIS COMPARING CUSTOM COMPUTER-GENERATED CRANIOFACIAL IMPLANTS VS. TRADITIONAL BONY CRANIOPLASTY FOR CONGENITAL AND TRAUMATIC CRANIAL DEFORMITIES

Presenter: Omar A. Fouda Neel, MD, FRCSC
Authors: Fouda Neel OA, Gilardino M, Karunanayake, Izadpanah A, Al-Ajmi S
McGill University

Purpose: Cranioplasty for cranial defects can be performed either with gold-standard autologous bone grafts and osteotomies or alloplastic materials in skeletally mature patients. Recently, custom computer-generated implants (CCGI) have gained popularity due to their patient-specific, pre-operatively designed anatomic forms and obviated donor site morbidity. Besides their apparent advantages in properly selected patients, however, a cost-benefit analysis has not been performed. The purpose of the present study was to compare the perioperative cost-benefit of cranioplasties performed with autologous techniques versus CCGI for both traumatic and congenital reconstructive indications.

Methods: A retrospective chart review of patients who underwent cranioplasty from 1990 to January 2013 at our centre was performed. Costs were calculated to include operative lengths, length of hospital/ICU stay, hardware utilized and need for transfusion.

Results: The average total cost per case for autologous reconstruction (n=16) was 3.7% higher than CCGI (n=5) cranioplasty (\$25,528.34 compared to \$24,592.80). Operative and anesthesia time were 51% and 37% less, respectively, in the CCGI cases. The average length of hospitalization was 49% less in CCGI cases. Blood transfusion was required in two autologous cases but was not required in any CCGI cases. None of the CCGI cases required revision for contour deformity or resorption whereas 38% of autologous reconstructions required a revision cranioplasty and 31% demonstrated bony resorption.

Conclusion: The analysis of our experience demonstrated an overall minor cost advantage and a significant patient benefit (decreased OR time, hospitalization and revision rate) of CCGI cranioplasty over traditional autologous bone techniques. These promising preliminary results warrant future studies to determine a definitive cost-benefit and complication comparison between the techniques.

Learning Objectives: 1. To compare the potential cost-benefit of CCGI vs. autologous cranioplasty for both traumatic and congenital reconstructive indications.

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TOTAL FACE, DOUBLE JAW, AND TONGUE TRANSPLANTATION: AN EVOLUTIONARY CONCEPT

Presenter: Amir H. Dorafshar, MBChB, FAAP
Authors: Dorafshar AH, Bojovic B, Christy MR, Iliff NT, Borsuk DE, Rodriguez ED
University of Maryland

Introduction: The central face high-energy avulsive injury has been repetitively encountered and predictably managed. Despite our significant surgical advances and multiple surgical procedures, the ultimate final result continues to reveal an inanimate, insensate and sub-optimal aesthetic result.

Methods: To effectively address this perplexing deformity a comprehensive multidisciplinary approach was devised. The strategy involved the foundation of a basic science laboratory; the cultivation of a supportive institutional clinical environment; the innovative application of technologies; cadaveric simulations; a real-time clinical rehearsal; and an informed and willing patient who had the characteristic deformity.

Results: Following Institutional Review Board and organ procurement organization approval, a total face, double jaw and tongue transplantation was performed on a 37-year-old male with a central face high-energy avulsive ballistic injury.

Conclusion: This facial VCA represents the most comprehensive transplant performed to date. Through a systematic approach and clinical adherence to fundamental principles of aesthetic, craniofacial, and microsurgery as well as the innovative application of technologies, restoration of human appearance and function for individuals with a devastating composite disfigurement is now a reality for our patient population.



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FACIAL ANIMATION IN MÖBIUS SYNDROME

Presenter: Juan C. Rodriguez Sr., MD

Author: Rodriguez JC

Garrahan Pediatric Hospital

Möbius Syndrome is a congenital anomaly that compromises the abducens and facial nerves, affecting speech, oral competence and most importantly facial animation.

Different procedures have been tried to improve the lack of facial musculature. Microsurgery, specially the gracilis muscle transfer with the use of nerve grafts, improved the possibilities to reconstruct facial palsy.

Based on the study by Zuker et al. we present our experience in segmental gracilis muscle transplant using facial or temporal vessels for revascularization and the motor nerve to the masseter for reinnervation.

Others possibilities are innervate the muscle with a segment of the hypoglossal nerve, but the most of patients have some degree of paralysis or incoordination of XII nerve. The accessory nerve is not affected in Möbius Syndrome, but is far from the face and thus may require a nerve graft to activate the muscle.

The other cranial nerve that could activate the muscle and is not involved in Möbius Syndrome is the trigeminal nerve. Twenty six patients with Möbius Syndrome, 20 unilateral and 6 bilateral, underwent gracilis muscle transfer. To reduce time of surgery, two teams worked simultaneously, one lifting the gracilis muscle and the other dissecting receptor vessels and the masseteric nerve. The muscle is anchored into the corner of the mouth and upper lip providing sufficient tension. In all children facial animation improved with good excursion of the oral commissure. In all cases difficulties eating and drinking and drooling improved significantly. Complications were minor in spite of the complexity and duration of the surgery.

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INVESTIGATING THE ETIOPATHOGENESIS OF OBSTRUCTIVE SLEEP APNEA IN PEDIATRIC PATIENTS WITH UNILATERAL CRANIOFACIAL MICROSOMIA

Presenter: Parit A. Patel, MD

Authors: Patel PA, Szpalski C, Fisher M, Wetterau M, Bernstein J, McCarthy JG, Warren SM

New York University

Background: Bilateral craniofacial microsomia has been determined to be a major risk factor for obstructive sleep apnea (OSA). We hypothesize that unilateral craniofacial microsomia (UCFM) can also be an unrecognized cause of OSA.

Purpose: To determine and characterize the etiopathogenesis of OSA in patients with UCFM through the use of three dimensional airflow analysis.

Materials and Methods: All pediatric patients diagnosed with UCFM at a tertiary level institution from 1990 to 2010 were reviewed. Patients with confirmed diagnoses of OSA (Apnea Hypopnea Index > 1) were compared to control patients with UCFM without OSA otherwise randomly selected. Three dimensional analysis of airflow with a computational fluid analysis model was performed (MIMIC software) on a sample of both affected as well as unaffected individuals. CT data was converted into a 3-dimensional computational model where the Navier-Stokes equations governing fluid flow can yield a variety of objective measures of airflow in the given anatomy.

Results: 62 patients were identified as having UCFM, 7 patients were further diagnosed with OSA. 11.5% of the patients diagnosed with UCFM were diagnosed with OSA, compared to 2.5% in the healthy pediatric population ($p=0.01$). 100% of patients with OSA were Pruzanski grades IIB or higher. OSA presenting symptoms included snoring (71.4%), failure to thrive (FTT) (57.1%), chronic respiratory infections (42.8%), adenotonsillar hypertrophy (28.6%) or loud breathing (28.5%). Snoring ($p<0.001$), presence of Goldenhaar features ($p=0.001$) and FTT ($p=0.004$) were identified as significant predictors for OSA in patients with UCFM. Race, obesity, cleft lip/palate, upper respiratory complications, presence of adenotonsillar hypertrophy and side of UCFM were not predictors of OSA in our cohort. Patients with UCFM and OSA had significantly lower total flow rate compare to controls.

Conclusions: The prevalence of OSA in UCFM patients is 3.9 times greater than in the otherwise healthy population. Snoring, presence of Goldenhaar features and FTT were shown to be predictive factors for OSA in the presence of UCFM.



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LONGITUDINAL GROWTH ANALYSIS OF MANDIBULAR ASYMMETRY IN UNOPERATED PATIENTS WITH UNILATERAL CRANIOFACIAL MICROSOMIA (UCFM)

Presenter: Pradip R. Shetye, DDS, MDS

Authors: Shetye PR, Grayson BG, McCarthy JG
New York University Langone Medical Center

Purpose: To examine the longitudinal growth changes in facial asymmetry in unoperated children with UCFM and to determine the rate of growth of the ramus height on the affected and on the unaffected side of the mandible.

Methods: This is a serial retrospective longitudinal growth study of 30 untreated patients (21 males and 9 females) with unilateral craniofacial microsomia (age range from 5 years to 14 years). The inclusion criteria were that patients had to have a lateral and posteroanterior (PA) cephalograms at 2 time points separated by at least a 2 year interval without any surgical intervention. The mean age of patients was 8.5 years and the mean follow-up records were 3.7 years. There were 13 patients in group I with a Pruzansky Type I mandible and 17 patients in group II with a Pruzansky Type II mandible. Fourteen patients had the right side of the mandible affected and sixteen patients had the left side of the mandible affected. The unaffected side of the mandible served as a control. Eighteen parameters were examined at each of the two time intervals.

Results: In patients with Pruzansky Type I mandible the affected ramus grew on average 1.41 mm per year; during the same period the unaffected ramus grew 1.66 mm per year. In patients with Pruzansky Type II mandible the affected ramus grew on average 0.84 mm per year; during the same period unaffected ramus grew 1.79 per year. When the growth rate of the ramus height on the affected side was compared to unaffected side, there was no statistical significant difference in Pruzansky Type I mandible ($p > 0.05$), and there was statistical significant difference in Pruzansky Type II mandible ($p < 0.05$).

Conclusion: The growth rate discrepancy of the affected ramus height was more severe in Pruzansky Type II mandible when compared to Type I mandible. Unoperated patients with unilateral craniofacial microsomia and Pruzansky Type II mandible had progressive facial asymmetry. These findings must be considered in treatment decisions in growing patients with UCFM.

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FINAL SKELETAL CORRECTION IN CRANIOFACIAL MICROSOMIA- STRATEGIES FOR MANAGING THE COSTOCHONDRAL-GRAFTED PATIENT

Presenter: Andrew A. Heggie, MD, DDS

Authors: Heggie AA, Shand JM
Royal Childrens Hospital of Melbourne

The spectrum of soft tissue and skeletal deficiency in craniofacial microsomia dictates the need for a range of surgical strategies to manage individual cases. While the advent of mandibular distraction was initially recommended for this condition, many units are re-evaluating this approach as a substitute for traditional techniques that remain indicated. Costo-chondral grafting to establish a more functional ramus/condyle unit remains our preferred reconstruction for the medially-placed (Kaban) Type IIB and Type III cases. However, the combination of inherited skeletal patterns and unpredictable graft growth can make the final skeletal deformity a major surgical challenge for correction.

The records of patients who underwent costo-chondral grafting and then reached skeletal maturity were retrieved for the past 15 years. Of the 11 cases identified, all required orthognathic surgery (bimaxillary-7 mandible-4, genioplasty-10). In several patients, difficulties were encountered in the sagittal section of the previous ramus/body graft site due to the altered anatomy. The neurovascular bundle is at considerable risk as its path varies considerably and is usually encased within cortical bone. Levelling of the occlusal plane in extreme cases may be unstable due to difficulty in overcoming muscular forces during mandibular repositioning. The magnitude of chin repositioning is much greater than may be anticipated as the soft tissue drape follows only in a most reduced hard to soft tissue ratio. Fat grafting on the new skeletal framework as a "fine-tuning" procedure has made a significant contribution to improving facial balance by replacing the missing subcutaneous tissue on the affected side.

Several cases illustrating the final skeletal correction will be presented together with strategies for managing the traditional osteotomies. Patients and families must be warned that a number of procedures at maturity may be required to achieve the best symmetry possible with a functional outcome.



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248 YEARS OF EXPERIENCE CLASSIFYING THE MANDIBULAR DEFORMITY IN HEMIFACIAL MICROSOMIA: A CRITICAL ANALYSIS OF THE PRUZANSKY/KABAN CLASSIFICATION SYSTEM UTILIZING THREE-DIMENSIONAL COMPUTED TOMOGRAPHY

Presenter: Scott Bartlett, MD
Authors: Wink JD, Goldstein J, Paliga JT, Taylor JA, Bartlett SP

Childrens Hospital of Philadelphia

The Kaban modification of the Pruzansky classification (KPC) is based on the severity of the mandibular deformity in Hemifacial Microsomia (HFM) as assessed by clinical exam and plain x-ray. Despite advances in medical imaging, the KPC continues to be the benchmark for the classification of HFM. Our goal is to examine HFM using current 3DCT technology to determine its relationship to traditional KPC as well as its reproducibility between individual raters.

An IRB approved retrospective review of all patients with a diagnosis of HFM and a preop 3DCT was performed. A gold-standard score based on consensus between surgeons at our institution stratified the population into Mild (0-1), Moderate (2a), and Severe (2b-3). Clinical KPC score was used as a second comparison. 3DCT scans were evaluated by surgeons and rated according to the KPC. Percent agreement was compared between these standards and the scores of our raters. ANOVA was used for statistical significance.

16 craniofacial surgeons with 248 yrs (avg 15.5 yrs) of experience from 11 institutions were surveyed. 41 patients met criteria including 38 patients with documented clinical scores. When comparing the raters' 3DCT-based classification to the clinical KPC scores, the average agreement was 38.1% (range: 27.3% for Type2 to 57.9% for Type3). There was improved rater identification of Type 3 mandibles ($p < .001$), however, as a group all raters were equally unable to accurately identify mandibular severity as compared to clinical assessment ($p = .90$). When comparing the raters' 3DCT-based classification to our gold standard, the average total agreement was 67.8% (range: 40.0% Moderate to 84.6% Severe) with improved identification of Mild and Severe mandibles ($p < .001$). As a group all raters were equally unable to accurately identify mandibular severity as compared to the gold standard ($p = .97$).

The introduction of 3DCT into the diagnostic paradigm highlights the inaccuracy and variability of traditional classification systems. Our results question the accuracy and reproducibility of the current clinical paradigm suggesting the need to reexamine the classification of HFM.

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TOTAL AURICULAR CONSTRUCTION NEW APPROACH WITH SIX TYPES OF FRAMEWORK TEMPLATES BASED ON NORMAL CURVE VARIATIONS

Presenter: Akira Yamada, MD, PhD
Authors: Yamada A, Ueda K, Otani K, Nuri T, Ohtsuki Y, Gosain A, Harada T, Salyer K

Osaka Medical College

Background: Total auricular construction is still one of the most challenging procedures in craniofacial deformities. We found that normal auricle has three major types of helix-lobule curve, and two major types of concha outline. Based on these findings, we developed new auricular frame templates with normal curve variations, and applied to total auricular construction.

Patient and Methods: 24 patients (25 auricles) were employed in this study. The patients consist of 15 lobule type, four concha type4, one anotia type, and three secondary cases, and one partial reconstruction. The curve analysis of normal side revealed that Helix-lobule curve consist of TypeA; 19, TypeB; 4, and TypeC; 2., and concha outline consists of Type1; 15, Type2; 10. We evaluated postoperative morphology of the auricle with the six types of auricular framework templates.

Results: This new six types of auricular frame templates were successfully applied to patients with various ethnic background, in various country (e.g.: Japan, China, USA, Argentina, Mexico, Oman, and India), and satisfactory outcome was achieved.

Conclusion: Based on pre and postoperative curve analysis of auricular shape, the new six types of auricular frame templates can be applied to patients with various ethnicities.



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CUSTOMIZED POROUS POLYETHYLENE FRAMEWORK FOR EAR RECONSTRUCTION

Presenter: ZungChung Chen, MD

Authors: Chen ZC, Chen YA

Chang Gng Memorial Hospital

Since porous polyethylene (Medpor) was introduced to ear reconstruction, it has become one of the most popular techniques in microtia reconstruction. Compare with the traditional method with autologous costal cartilage as the framework, using this alloplastic substitute avoids the harvesting of ribs. Moreover, the surgery can be performed at earlier age with relative consistent results. However, the uniform appearance and the higher extrusion rate still cannot be widely accepted.

Patients and Methods: Since Dec 2010 to Jun 2012, 13 microtia patients received primary ear reconstruction in one-stage using customized Medpor as the framework. The mean age is 19 years old (range 8-51). All patients were follow-up regularly (7-20 months) for observation of wounds. The Medpor block (4cm x 6cm x 1cm) was used then carved by scalpel and sculpture knife according to the appearance of contralateral ear by single expert surgeon. A piece of Medpor was shaped to fit the posterior surface of the framework to provide adequate projection. Wide temporoparietal fascia (TPF) including superficial temporal artery was raised to cover the entire Medpor. Skin graft was harvested from scalp to TPF surface. Tie over was used without any drainage.

Results: Wounds healed uneventfully in 4 patients. Hematoma was noted in one case and drainage was done at post-operative day 3. Medpor extrusion was noted in 3 cases. One was over upper helical rim 1 year after surgery and another 2 cases happened over tragus 6 months post-operatively. Once the medpor was exposure, surgery was indicated using random fascial flap for coverage. Small skin loss was noted in 5 patients and managed with skin graft to ensure wounds healing. All patients were satisfied with the reconstructed ear and their appearances were quite similar to the normal side.

Discussion: Customized Medpor framework is available by carefully sculpturing. The entire framework must be completely covered by TPF. Once Medpor was exposure or the wound not healed as expectation, surgical intervention should be performed aggressively without hesitance. Therefore, a better result can be achieved.

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PLANNING SURGICAL RECONSTRUCTION IN TREACHER - COLLINS SYNDROME USING GEOMETRIC MORPHOMETRICS

Presenter: Dariush Nikkhah, BM, MSc, MRCS

Authors: Nikkhah D, Ponniah A, Ruff C, Dunaway D

Great Ormond Street Hospital

Background: Treacher-Collins syndrome (TCS) is a rare autosomal dominant condition of varying phenotypic expression. The surgical correction in this syndrome is difficult; approach varies between craniofacial departments worldwide. We aimed to design standardized tools for planning orbito-zygomatic and mandibular reconstruction in TCS using geometric morphometrics.

Methods: The Great Ormond Street Hospital database was retrospectively identified for patients with TCS. 13 children (2 - 15 yrs) who had suitable un-operated three-dimensional computed tomography head scans (3DCT) were included. The TCS 3DCT scans were compared using a template of 96 anatomically defined landmarks to 26 age-matched normal dry skulls.

Results: Thin-plate spline movies illustrated the characteristic deformities of retromicrognathia, maxillary and orbito-zygomatic hypoplasia in the TCS population. Geometric morphometrics was used to quantify the changes required in the reconstruction of the orbito-zygomatic and mandibular region in TCS patients. Intra and inter-rater reliability of the landmarks was acceptable and within a sd < 1mm on 97% and 100% of 10 repeated scans respectively.

Conclusions: Virtual normalisation of the TCS skull effectively describes characteristic skeletal deformities and provides a useful guide to surgical reconstruction. Size matched stereolithographic templates derived from thin plate spline warps can provide effective intraoperative templates for zygomatic and mandibular reconstruction in the TCS patient.



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A CRITICAL ANALYSIS OF THE RELATIONSHIP BETWEEN THE MAXILLA AND THE MANDIBLE IN HEMIFACIAL MICROSOMIA

Presenter: Jason D. Wink, BA

Authors: Wink JD, Goldstein J, Paliga JT, Taylor JA, Bartlett SP

Childrens Hospital of Philadelphia

Hemifacial Microsomia (HFM) commonly manifests as a heterogeneous mandibular deformity. The most cited theory involves a vascular insult to the first and second branchial arches. Based on this theory, the severity of maxillary deformity should correlate with the degree of mandibular deformity. Our goal was to use 3D computed tomography (3DCT) and image segmentation to better understand the maxillary deformity in relation to mandibular disease.

An IRB approved retrospective review of all patients with a diagnosis of HFM and a preoperative 3DCT was performed. Volumetric analysis was performed by segmentation of the mandible, maxilla and maxillary sinus (Mimics). Patients were stratified into groups based on the Kaban modification of the Pruzansky classification: Mild (0-1), Moderate (2a), Severe (2b-3) as well as rank ordering based on overall severity. Analyses involved paired T-Tests within severity groups, one-way ANOVA when assessing across groups (Ipsilateral/Contralateral ratio) and regression to assess for trends.

30 patients (16M, 14F) were identified including 4 with mild (11.6 yrs), 12 with moderate (17.5 yrs), and 14 with severe (2.3 yrs) disease. The mandibular volume ratio appeared to differ across all patient groups ($p < .001$) and trend with our rank ordering ($p < .001$). Maxillary bony volume ratio across all patient groups appeared equal ($p = 0.16$). In patients with severe disease, the maxillary bone volume was found to be significantly decreased on the ipsilateral as compared to the contralateral side ($p = .0123$). There was no difference in maxillary sinus volume between ipsilateral and contralateral sides within any patient groups or when comparing across groups ($p = 0.10$). There appeared to be no trend in the volume ratio of mandible and maxilla ($p = .41$).

Our analysis found a bony deficiency of the mandible and maxilla in the most severe HFM patients. No differences in maxillary sinus volume were found. There appeared to be no trend in severity between mandible and maxilla thus providing evidence contrary to the existing theory of a vascular insult to the 1st and 2nd branchial arch as the most likely etiology for HFM.

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A NOVEL APPROACH TO OSSEOINTEGRATED AURICULAR PROSTESIS RETENTION

Presenter: Drew Schnitt, MD

Authors: Schnitt D, Trainer D
Joe DiMaggio Childrens Hospital

Background: The application of extraoral implants for the prosthetic rehabilitation of auricular defects is widely accepted. The traditional technique of implant retention using the bar and clip provides good retention but the bar prevents easy patient access for cleaning and is more costly and cumbersome. An alternative method of using magnetic retention is limited by the corrosive nature of the magnet and its low breakaway strength. We present a new approach for auricular prosthesis retention using the Baha® snap attachment method which promotes ease of hygiene, improved retention, and increased breakaway strength for active patients.

Methods: A group of patients are presented who received prosthetic rehabilitation of the auricle. After all reconstructive options were discussed, both patients selected and underwent a single stage osseointegrated auricular prosthetic implant placement with the Baha® snap attachment method.

Results: The results of the Baha® snap attachment method for auricular prosthesis retention are presented as a photographic analysis and description of technique. This attachment method provides a high retentive force of 30 N per implant and patients report a high degree of satisfaction with ease of prosthesis placement and hygiene of the implant at one year follow-up.

Conclusions: We present the first known report of extraoral retention of an auricular prosthesis using the Baha® snap attachment method. This technique is lower in cost with relative ease of placement and improves upon the current bar and clip and magnet methods with easier access for hygiene and greater retention breakaway strength and a arguably a better aesthetic outcome than most autologous reconstructions.



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“I KEEP IT IN THE DRAWER” - MINIMISING THE CONFLICT BETWEEN EAR RECONSTRUCTION AND BAHAS IN CHILDREN WITH MICROTIA - A SALUTORY EXPERIENCE FROM THE UK

Presenter: David T. Gault, MB ChB FRCS

Authors: Gault DT, Cheang PP

The London Centre for Ear Reconstruction

Children with microtia present to many different clinics. Those with bilateral microtia require early help with hearing and the use of a bone-anchored hearing aid (introduced in 1977) is the most popular approach in the United Kingdom. Pleas for a cogent approach to hearing aid placement mindful of later autogenous ear reconstruction (Bajaj 2005, Sabbagh 2004) still go unheeded. An additional population of patients with unilateral microtia who are fitted with a hearing aid have increased the number of those whose ear reconstruction is unnecessarily complicated by injudicious placement. Presenting with a scarred and tethered operative field, many of these patients will respond, when questioned about the whereabouts of their BAHA, “I keep it in the drawer”. Nineteen consecutive microtia ear reconstructions in sixteen patients (3 had bilateral microtia) are reviewed. BAHA's had been fitted in ten of these 19 ears, and all were located in the zone of reconstruction. All required removal and four were later relocated. Temporo-parietal flaps were required at the first stage in two cases, but in all 10 cases fitted with BAHA's, both the first and second stage procedures were more difficult and longer than usual. Surgical coping techniques are important but no substitute for an early multidisciplinary team approach, especially for those with concomitant craniosynostosis when bicoronal incisions can compromise the TP flap.

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MANAGEMENT OF SURVIVING PATIENTS WITH OTOCEPHALY: TREATMENT ALGORITHM AND CORRECTION OF FACIAL DEFORMITIES

Presenter: Kristin Yee

Authors: Yee K, Chawla R, Chan FC, Kawamoto HK, Bradley JP

University of California Los Angeles

Background: Otocephaly is a rare developmental defect with severe hypoplastic and retrognathic mandible that poses an immediate airway threat to the newborn infant. This syndrome is associated with hypoplasia of the entire lower face and neck structures. The aim of this study is to examine the long-term outcomes of surviving patients and to provide an understanding of its management and compulsory mandibular reconstruction. We present the management and reconstruction of previously unreported cases of surviving otocephaly.

Methods: We conducted a retrospective study of surviving otocephalic patients between 2002 and 2012 with long-term follow up at University of California, Los Angeles Craniofacial Center (n=3). Patient information was recorded including: diagnosis, deformities, surgical procedures, indications for surgery, and surgical outcomes. We also review and update the management of such a rare group of patients.

Results: The patients range between 8 to 14 years of age with a minimum five year follow-up. All patients had various degrees of facial deformities (severe micrognathia, mastoid/cranial base anomalies, hypoplastic floor of mouth, tongue and laryngopharyngeal regions) consistent with their common diagnosis of otocephaly. Initial management of immediate tracheostomy and gastrostomy facilitated survival. Tessier craniofacial clefts and cleft palates were repaired at a mean of 16 months of age. Serial mandibular distraction procedures were necessary (mean of 2.8; range 1-4) in all cases (2 external; 7 internal devices). Temporomandibular joint reconstruction with bilateral rib grafts and Matthew's distraction device was performed in two patients. One patient required supraclavicular island flaps for neck reconstruction.

Conclusion: For patients surviving with otocephaly, multiple reconstructive procedures and long-term therapy is necessary for improved function. A treatment algorithm with craniofacial distraction and correction of craniofacial deformities is outlined based on our experience.



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FREE PARASCAPULAR FLAPS FOR PRUZANSKI III
HYPOPLASTIC MANDIBLES: OUR EXPERIENCE WITH
7 PATIENTS

Presenter: Christopher Gordon, MD

Authors: Rapp S, Uribe-Rivera A, Pan BS,
Billmire DA, Gordon CB

Cincinnati Childrens Hospital Medical Center

Purpose: Distraction osteogenesis is a well established approach in correction of the hypoplastic mandible. It has been less successful in the treatment of Pruzanski Class III deformities. Despite early overcorrection, there is poor subsequent growth owing to absence of the condylar growth center. Various free tissue transfers have been utilized for pediatric ramus reconstruction, including rib, fibula, iliac crest, and scapula. Only scapula appears to include a viable growth center. We describe our experience utilizing the parascapular osteocutaneous free flap for mandibular reconstruction.

Materials and Methods: From 1994 to 2013, 7 patients with grade III hypoplastic mandibles were candidates for mandibular reconstruction with a parascapular osteocutaneous flap. Flaps were performed at two different institutions and by two different senior surgeons. The average age at time of initial surgery is 5.3 years old. Distraction was performed on 2 patients in an effort to improve obstructive sleep apnea symptoms. Bone surveillance was conducted through cephalograms and computed tomography at average of 3.5 years follow-up (range 0.2 to 9.3 years). All patients received 1 week of perioperative cephalosporin antibiotic coverage.

Results: All seven patients underwent successful transfer and no bone flap loss with stable clinical fixation noted up to 9.3 years of follow-up. One patient with bilateral Goldenhar Syndrome underwent bilateral flaps 6 months apart. Complications include, hematomas (n=1), and TMJ limited aperture (n=1).

N=1 syndromic patient was diagnosed within this cohort with Goldean Har syndrome (Pruzanski III) and Pierre Robin Sequence.

Discussion: There are few reports of long term success in treating the severely hypoplastic mandible with free osteocutaneous scapula transfer. Our results suggest that the parascapular osteocutaneous free flap represents an effective surgical option with minimal donor site morbidity, good long term growth and appropriate qualities to make it our preferred method of mandibular reconstruction.

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PSYCHOSOCIAL, FEEDING, SPEECH & DROOLING
OUTCOMES IN CHILDREN WITH BECKWITH
WIEDEMANN SYNDROME FOLLOWING TONGUE
REDUCTION SURGERY

Presenter: Caroleen Shipster, MSc

Authors: Shipster C, Morgan A, Dunaway D

Great Ormond Street Hospital for Children NHS Foundation Trust

Tongue reduction surgery (TRS) is recommended for children who have macroglossia associated with Beckwith Wiedemann Syndrome (BWS) to overcome, or reduce, the effects of macroglossia. These include altered facial appearance, feeding difficulties, speech difficulties, dental and occlusal anomalies and drooling. Only two studies report the psychosocial, feeding, speech, and drooling outcomes both pre and post TRS longitudinally within the same cohort N=10 (Shipster et al., 2006; Shipster et al., 2012). The aim of this study was to report findings in a larger cohort.

30 children were assessed using a parental questionnaire and a variety of clinical measures pre-operatively -T1; 3 months post-operatively -T2 and at long term follow up -T3 (2 years + following surgery). All were operated on by the same surgeon. The procedure involved excision of an anterior modified keyhole pattern. Age at surgery ranged from 1;4 to 5;1 years.

Parents reported that macroglossia had a negative impact cosmetically that was ameliorated following surgery. At T1 excessive drooling was present in 75%; feeding difficulties were present in 100%; the placement of anterior consonants during speech production was disrupted in 100%. At T2, drooling, and feeding difficulties were completely resolved. At T2, speech errors related to occlusion and delayed developmental were present in 50%. At T3, speech errors related to occlusion and delayed development were present in 20%.

Pre-operatively, a common profile of feeding, speech and drooling difficulties was found with negative effects on cosmetic appearance. TRS has a positive impact on these features with good outcomes for children with BWS.



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EARLY FAT GRAFTING FOR AUGMENTATION OF ORBITOZYGOMATIC REGION IN TREACHER COLLINS SYNDROME: A PRELIMINARY REPORT

Presenter: Petros Konofaos, MD, PhD
Authors: Konofaos P, Diner PD, Arnaud EA
University of Tennessee Health Science Center

Treacher Collins (TC) syndrome requires various steps of reconstructions including soft tissues and bone. Bone grafts have been proven to resorb frequently. Recently fat grafting has been proposed as an adjunct in the therapy sequence. Our preliminary report was to evaluate a very early fat grafting in infants in order to: 1) Prepare a better soft tissue environment for the bone graft and 2) Allow spontaneous repair of the skull after fat grafting. Three infants with TC were primarily treated with sequential fat grafting according to Coleman technique as early as 6 months of age. Two to three sessions were performed prior to bone grafts, which could be done at age 12, 14 and 20 months. After the calvarial bone grafts, another fat grafting was indicated in two out of the three patients. Maximum follow-up was two years. In all patients, the early fat grafting improved the quality of the atrophic skin and the morphological appearance of both the eyelids and the zygomatic region. In two patients, there was not any need for further reconstruction of the lower eyelid with an upper eyelid transposition flap. Following fat grafting, bone grafting with calvarial graft was performed in all patients. After one year of follow-up, the bone graft was partially resorbed in the two youngest patients. Simultaneously, the two youngest patients show almost spontaneous complete repair of the skull defects. Very early fat grafting in infants was proven effective and safe after six months of age. Marked improvement in soft tissues allowed simple bony repair with calvarial bone grafts, as well as spontaneous bony healing of the skull in two patients. In the youngest patients, no further step of reconstruction of the eyelids was necessary. It is too early to observe a reduction in the overgrowth of the medial third of those patients, which is needed to be confirmed.

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“BLACK BONE” MRI: CHANGING THE FACE OF CRANIOFACIAL IMAGING

Presenter: Karen A. Eley, MBChB, MRCS, PGCTLCP, FHEA, MSc, DPhil
Authors: Eley KA, Sheerin F, Watt-Smith SR, Golding SJ
University of Oxford/Oxford Craniofacial Unit

Introduction: CT with 3D reconstruction is currently the imaging modality of choice in craniosynostosis, despite increasing concerns regarding the potential deleterious effects of ionising radiation. The objectives of this study were to explore the potential of MRI in diagnosis and 3D reconstruction in craniosynostosis, thus establishing a non-ionising alternative to CT.

Methods: We developed the “Black Bone” MRI sequence to minimise soft tissue contrast and enhance the bone-soft tissue boundary. Following optimisation, this novel technique was utilised in 12 children with a clinical diagnosis of craniosynostosis. The anonymised “Black Bone” datasets were independently assessed for accuracy of diagnosis and subsequently correlated with CT findings. To provide comparative analysis the appearance of normal cranial sutures in infants (< 1 year) on 100 anonymised datasets across 5 routine MRI sequences were independently assessed. Techniques were subsequently developed to segment the craniofacial skeleton on “Black Bone” datasets to create 3D rendered images, using a range of commercially available software.

Results: On the routine MRI sequences the cranial sutures were visualised as an area of signal void, often difficult to clearly delineate and were not consistently identified in all children. The “Black Bone” MRI sequence provided improved identification of the cranial sutures. The cranial sutures appeared as areas of increased signal, clearly visible against the dense black cortical bone. In craniosynostosis, the fused suture could be distinguished from remaining patent sutures with high levels of accuracy. Three dimensional rendering of the “Black Bone” images improved identification of the cranial sutures, permitting visualisation throughout their course. On comparison with the 3D CT images from the same infant, the 3D “Black Bone” images approached the quality of those from CT.

Conclusions: “Black Bone” MRI offers a reliable non-ionising alternative to CT in children with craniosynostosis and demonstrates considerable potential in changing the face of craniofacial imaging.



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RADIATION REDUCTION PROTOCOL EFFECTIVELY REDUCES RADIATION EXPOSURE DURING CT EVALUATION OF PEDIATRIC CRANIOFACIAL ANOMALIES

Presenter: Christopher S. Zarella, MD
Authors: Zarella CS, Didier R, Bardo D, Selden NR, Kuang AA

Oregon Health and Science University

Background: There has been growing concern with radiation exposure during computed tomography (CT) evaluation in children. Using a radiation reduction protocol we developed and described in 2010 in *Pediatric Radiology*, we have previously demonstrated that optimal head position decreased the overall effective radiation dose received while maintaining diagnostic quality. We now prospectively evaluate the effectiveness of this protocol in a large group of patients.

Methods: This single-institution prospective cohort study examined effective radiation dose and image quality for craniofacial CT evaluation of children between January 2010 and April 2012 with comparison to patients in a pilot radiation reduction group from October 2008 to January 2010 as well as historical age matched controls.

Results: We had previously shown an 18% reduction in effective radiation dose in pilot patients after initial implementation of our protocol. Currently, 305 patients in the study group experienced an average CT effective radiation dose of 2.32 mSv, compared to 1.86 mSv in 91 pilot group patients, a 20% additional reduction, and a 34% overall reduction compared to historical controls. Dose reduction was equivalent for males and females. The clinical utility of radiation reduction protocol examinations was maintained despite a slight decrease in the quality of the brain windows.

Conclusion: Altering the position of the head using neck extension as part of a radiation reduction protocol diminishes the effective radiation dose by more than one third, while maintaining the diagnostic quality and clinical utility of pediatric craniofacial CT imaging.

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AN OPEN-ACCESS, INTERNET-BASED, PLASTIC SURGERY SIMULATOR

Presenter: Roberto Flores, MD
Authors: Flores R, Olikier A, Costa M, Cutting C
Indiana University

Background: Plastic surgery education was revolutionized by Smile Train's Virtual Surgery Videos. Since its original release, over 10 years ago, improvements in internet technology have allowed browsers to display interactive digital 3D environments, providing opportunity to enhance the educational quality and outreach of this surgical educational tool. We report our experience in capitalizing on these recent internet advances in creating the first web-based interactive cleft surgery simulator.

Methods: Previously constructed digital animations of cleft surgery, were upgraded in Maya 10 (Autodesk, San Raphael, CA) in preparation for web-based simulation. New animations were also created demonstrating the surgical markings as well as the normal and pathologic anatomy relevant to the procedures. These animations were exported into a web-based, interactive surgical simulator produced by BioDigital Systems Inc. (New York, NY). High-definition intra-operative video recordings of all procedures were edited in Adobe Premiere CS5.5 (Adobe, San Jose, CA) and exported into the simulator with the addition of voiceover. Test questions were produced for each surgical module.

Results: Five cleft surgical repairs are demonstrated in this web-based interactive surgical simulator: Extended Mohlar unilateral cleft lip, Cutting bilateral cleft lip, Furlow cleft palate, Interverlar veloplasty cleft palate, superiorly-based pharyngeal flap and secondary cleft rhinoplasty. Through a customized digital interface the user can manipulate the 3D simulations in real-time including the ability to alter perspective, pace and order of the operation as well as zoom in on specific anatomy during the simulation. High-definition intra-operative video footage compliments the critical steps of each procedure demonstrated in the simulation. A voiceover and text guides the user through each tutorial. A test is included at the end of each simulation. There is no need for specialized software or downloads and access is zero cost.

Conclusions: We present the first open access, internet-based plastic surgery simulator.



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MEIN₃D. 12000 FACIAL SURFACE SCANS TO CREATE A MODEL OF NORMAL FACIAL VARIATION TO AID SURGICAL PLANNING

Presenter: Allan Ponniah, MBBS, MRCS, MSc

Authors: Ponniah A, Ruff C, Angullia F, Dunaway D
Great Ormond Street Hospital

Introduction: The problem with correcting facial deformity is that there is such great variation in what is considered a balanced face and therefore acceptable outcome. As such it is difficult to quantify outcomes of craniofacial surgery. Although normal values of facial proportions are useful, facial balance is a phenomenon that involves the whole face and not just the individual components. In addition to this, the surgery required to correct a deformity towards the traditional normal values may be more than that required to restore facial balance. This study demonstrates how Principal Component Analysis (PCA) could be used to create a virtual ideal 3D outcome that in shape terms is near to the patient's face. PCA has been previously used by this group in published work to demonstrate how the deformity of the Apert skull can be corrected.

Material and Methods: 12000 3D face scans were collected. These were male and female volunteers from all age groups and ethnicities. It is hoped that this is the start of a growing database which will collect enough data to capture facial variation in the whole human population. Of these a subset of 100 volunteers was used to create a model of normal variation using PCA.

Results: When introducing a patient with a facial deformity (midface hypoplasia due to Crouzon Syndrome) the model selects a balanced face shape that is closest to the patient's face. The mean landmark movement required to achieve the mean face was 15mm whereas to achieve the selected face by the model was 9mm. This selected outcome would require less surgery to achieve in comparison with working towards the the mean face shape.

Conclusion: This technology allows creation of virtual ideal outcomes in craniofacial surgery. It can also be used to assess the degree to which the deformity has been corrected. It would be useful in auditing craniofacial outcomes in order to help refine techniques. Ultimately it could be used to plan craniofacial surgery.

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INCIDENCE OF CNS ABNORMALITIES ON BRAIN MRI IN INFANTS WITH PIERRE ROBIN SEQUENCE

Presenter: Arlene Rozzelle, MD

Authors: Khan S, Pappas KP, Rozzelle AR
Childrens Hospital of Michigan Detroit Medical Center

Objective: Infants with Pierre Robin Sequence (micrognathia, glossoptosis, upper airway obstruction) may have co-morbidities (CNS, cardio, GERD, lower airway) that contribute to their apnea. At Children's Hospital of Michigan, we do a comprehensive work-up to determine the etiology and severity of apnea, including MRI of the brain to evaluate CNS abnormalities. Though there are isolated case reports in the literature of CNS anomalies in infants with Pierre Robin Sequence, no large patient series has been described. The objective of this study is to identify and quantify CNS co-morbidities found on MRI in patients with Pierre Robin Sequence treated at our institution.

Method: A retrospective chart review (with IRB approval) of all patients presenting to our institution with Pierre Robin Sequence since 1995, was carried out identifying patients who had brain MRI's performed. MRI results were reviewed, quantifying the incidence of brain abnormalities, types of abnormalities, and possible influence on apnea.

Results: Records were reviewed for 187 patients with the diagnosis of Pierre Robin Sequence. Of these, 84 patients (45%) had an MRI of the brain. Abnormalities were noted on 43 of the 84 (51%). Abnormalities included Chiari, Dandy-Walker, hypoxic injury and, most commonly, microcephaly and ventricular dilatation.

Conclusion: In this large series of infants with Pierre Robin Sequence who underwent brain MRI, a large proportion (51%) had brain abnormalities, some of which may cause apnea. Brain imaging is an important element of the comprehensive work-up of infants with Pierre Robin Sequence and necessary to delineate the etiology of their apnea and to plan appropriate treatment.



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TRIGONOCEPHALY AND FRONTAL LOBE PERFUSION

Presenter: Giovanna Paternoster

Authors: Di Rocco F, Grevent D, Chivoret N,
Paternoster G, Wyten R, Boddaert N,
Brunelle F, Arnaud E

Craniofacial Unit

Introduction: Trigenocephaly is associated to a reduction of frontal volume that can lead to an impaired cerebral development with cognitive impairment. Our previous study showed a reduction of the cerebral perfusion of frontal region in trigonocephaly compared to controls where the cerebral perfusion was homogene. In this study, we assessed the cerebral perfusion in trigonocephaly before and after surgical treatment comparing the perfusion in the frontal region and the occipital region in the same patient.

Methods: 49 trigonocephalic children undergoing surgery were prospectively studied by MRI with perfusion analysis before and after surgery (early post-operative period and at 3 months). There were 33 boys and 16 girls, mean age at surgery was 10 months. Neuroimaging protocol included a 3DT1, 3DT2 and ASL (Arterial Spin Labelling). Cortical blood flow was studied using regions of interest of 100 mm². A t-test was used for statistic analysis.

Results: In trigonocephalic children, the cortical perfusion was reduced in the frontal region compared to others regions (-21% to -49%, mean : -34%, $p < 0.01$). After surgery, this frontal hypoperfusion is corrected (0 to -31% mean : -8%, $p < 0.05$) at 3 months, that is confirmed at 12 months.

Conclusions: Though intracranial pressure is rarely increased in trigonocephaly, this study demonstrates that bony frontal stenosis may induce frontal lobes hypoperfusion which can explain the cognitive impairment. Furthermore, this study established that an early surgery can normalize the frontal perfusion.

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OUTCOME ANALYSIS AFTER HELMET THERAPY USING 3D PHOTOGRAMMETRY IN PATIENTS WITH DEFORMATIONAL PLAGIOCEPHALY: THE ROLE OF ROOT MEAN SQUARE

Presenter: Mahsa Bidgoli Moghaddam, MBBS

Authors: Bidgoli Moghaddam M, Brown TM,
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Deformational plagiocephaly (DP) is a multifactorial non-synostotic cranial deformity with a reported incidence as high as 1 in 7 infants in North America. Treatment options have focused on non-operative interventions including head repositioning and the use of an orthotic helmet device. Previous studies have used linear and two dimensional outcome measures to assess changes in cranial symmetry after helmet therapy. Our objective was to demonstrate improvement in head shape after treatment with a cranial molding helmet by using Root Mean Square (RMS), a measure unique to 3D photogrammetry, which takes into account both changes in volume and shape over time. Three dimensional photographs were obtained before and after molding helmet treatment in 40 infants (4-10 months old) with deformational plagiocephaly. Anatomical reference planes and measurements were recorded using the 3dMD Vultus® analysis software. RMS was used to quantify symmetry by superimposing left and right quadrants and calculating the mean value of aggregate distances between surfaces. Over 95% of the patients demonstrated an improvement in symmetry with helmet therapy. Furthermore, when the sample of infants was divided into two treatment subgroups, a statistically significant correlation was found between the age at the beginning of treatment and the change in the RMS value. When helmet therapy was started before 7 months of age a greater improvement in symmetry was seen. This work represents the first application of the technique of RMS analysis to demonstrate the efficacy of treatment of deformational plagiocephaly with a cranial molding helmet.



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BEST FACE FORWARD: VIRTUAL MODELING AND CUSTOM DEVICE FABRICATION TO OPTIMIZE CRANIOFACIAL VASCULARIZED COMPOSITE ALLOTRANSPLANTATION (VCA)

Presenter: Jordan Jacobs, MD
Authors: Jacobs J, Dec W, Levine JP, McCarthy JG, Weimer K, Moore K, Ceradini DJ

New York Medical College

Craniofacial vascularized composite allotransplantation (VCA) is especially challenging when bony components are required. Matching the complex 3D anatomy of the donor and recipient craniofacial skeletons to optimize bony contact and dental occlusion is a time consuming step in the operating room. Currently few tools exist to facilitate this process. We describe the development of a virtual planning protocol and patient specific device design to efficiently match the donor and recipient skeletal elements in craniofacial VCA. The protocol was validated in a cadaveric transplant. This innovative planning method allows a “snap-fit” reconstruction using custom surgical guides while maintaining facial height and width, as well as a functional occlusion. Postoperative overlay technology in the virtual environment provides an objective outcome analysis.

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OPTICAL COHERENCE TOMOGRAPHY: A POTENTIAL OPTIC NERVE HEAD IMAGING MODALITY FOR CRANIOSYNOSTOSIS PATIENTS WITH SUSPECTED RAISED INTRACRANIAL PRESSURE

Presenter: William R. Katowitz, MD
Authors: Katowitz WR, Cohen Y, Taylor J, Forbes BJ
The Childrens Hospital of Philadelphia

Craniosynostosis can cause an increased intracranial pressure (ICP). An elevated ICP (>15mm Hg) occurs in craniosynostosis patients at a rate of 8-16% in single suture fusion, up to 42% in multiple suture fusion and as high as 68% in patients with Crouzon syndrome. Ophthalmologists are often consulted to diagnose optic nerve edema, a sign in children with suspected raised ICP. However, up to 68% of children with craniosynostosis and a raised ICP do not have visually appreciable optic nerve edema on fundoscopic visualization. [1] Other testing modalities exist to detect optic nerve dysfunction associated with raised ICP, which include visual evoked potentials (VEP), orbital ultrasound, optic nerve evaluation on MRI, and optical coherence tomography (OCT). These ancillary tests complement direct optic nerve head visualization, but at this time are not a replacement for an evaluation by an ophthalmologist. While VEP can show great sensitivity to optic nerve dysfunction, it requires a dedicated electrophysiology lab. In addition, multiple readings are necessary as each patient serves as his or her own baseline. [2] OCT is a newer imaging technology that uses an optical signal acquisition and processing method. OCT can detect subtle optic nerve changes to the retinal nerve fiber layer, which can be thickened in optic nerve edema due to raised ICP. [3] We present our initial experience in using OCT in patients with craniosynostosis and suspected raised ICP.1. Tuite GF, Chong WK, Evanson J, et al. The effectiveness of papilledema as an indicator of raised intracranial pressure in children with craniosynostosis. *Neurosurgery*. 1996 Feb;38(2):272-8.2. Liasis A, Thompson DA, Hayward R, Nischal KK. Sustained raised intracranial pressure implicated only by pattern reversal visual evoked potentials after cranial vault expansion surgery. *Pediatr Neurosurg*. 2003 Jul;39(2):75-80.3. Skau M, Yri H, Sander B, et al. Diagnostic value of optical coherence tomography for intracranial pressure in idiopathic intracranial hypertension. *Graefes Arch Clin Exp Ophthalmol*. 2013 Feb;251(2):567-74.



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REAL TIME CINE-MRI IN FETUSES WITH HEAD AND NECK PATHOLOGY

Presenter: Oksana A. Jackson, MD

Authors: Jackson OA, LaRossa D, Victoria T, Pollock A, Feygin T

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Introduction: Real-time cine-MRI is being used with increasing frequency for the prenatal diagnosis and characterization of functional anomalies of the heart and other organ systems. The purpose of this study was to determine the feasibility and applications of this technique in fetuses with head and neck pathology.

Methods: Real-time cine-MRI evaluations were performed in 370 pregnant patients in addition to routine fetal MR evaluation of different organ systems, and the studies were reviewed by two pediatric neuroradiologists. Different patterns of fetal swallowing were identified in fetuses with head and neck pathologies. Comparison was performed to the normal motion of the fetal tongue, soft palate, hypopharynx, and epiglottis during swallowing. Post-natal follow-up was performed to confirm the diagnosis and correlate findings with outcomes post-delivery.

Results: The spectrum of pathologies noted with abnormal patterns of swallowing included giant fetal neck masses, cleft lip and palate, brainstem anomaly, syndromic craniosynostoses, and micrognathia with visualized glossoptosis. 70 patients with cleft lip and palate were studied and an abnormal pattern of swallowing characterized by velopharyngeal incompetence and hypopharyngeal pooling was noted in most.

Conclusion: Real-time cine-MRI methods are technically feasible and valuable tools in prenatal assessment, with increasing applications in the functional evaluation of head and neck pathology. Further characterization of fetal swallowing patterns may yield prognostic information that can be used for parental counseling and the planning of post-natal care.

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COMPUTATIONAL FLUID DYNAMICS FOR AIR FLOW MODELING IN CRANIOFACIAL PATHOANATOMY

Presenter: Mark D. Fisher, MD

Authors: Fisher MD, Patel PA, Szpalski C, Kassam S, McCarthy JG, Warren S, Kimbell J, Marcus J

Duke University

Background: A growing literature in the computational fluid dynamics (CFD) of nasal airflow has demonstrated a powerful new means of elucidating the relationship between anatomy and airway function. Along the same lines, computational fluid dynamics offers a new avenue for objectively studying the impact of our operations on airway function. Over the last two years we have developed a multi-institutional collaboration to bring the same technology to bear on problems pertinent to the craniofacial surgeon including the cleft lip nasal deformity (CLND) and hemifacial microsomia (HM).

Methods: CFD analysis was performed in a total of 10 individuals with the cleft nasal deformity or hemifacial microsomia (n=5 each). CT data were used to produce a 3D model using the MIMICS software. These were then converted into a computational mesh for solution of the Navier-Stokes equations governing fluid flow (ANSYS ICEM CFD Software). A simulation corresponding to resting breathing was then performed and analyzed to identify the impact of the anatomy on airway function (Fluent Software).

Results: CFD analysis allowed the objective demonstration of airflow patterns in both the CLND and in HM. In the case of the CLND, the hallmark areas impacting airflow included the cleft-side external valve, the internal valve, the septum and the inferior turbinate. The location of greatest impingement on flow was the internal valve due to bowing of the septum towards relatively oversized inferior turbinates. In the most severely affected of cases total nasal air flow was 49.5% of expected. Only 17.3% of total airflow passed through the cleft side.

Conclusions: Our early experience demonstrates the promise of computational fluid dynamics in elucidating the impact of craniofacial pathoanatomy on airway function. In both the CLND and in HM we have demonstrated patterns through which characteristic anatomy influences airway function. Future efforts examining pre- vs post-operative CT data will expand the present work through the analysis of the impact of surgery on airway function in craniofacial problems.



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A CLASSIFICATION SYSTEM TO GUIDE ORBITO-ZYGOMATIC RECONSTRUCTION IN TREACHER-COLLINS SYNDROME

Presenter: Ben Green, BSc

Authors: Green B, Nikkhah D, Ponniah A, Dunaway D
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WITHDRAWN

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CLINOCEPHALY AND CLOSURE OF LATERAL CALVARIAL SUTURES ON VOLUME-RENDERED CT RECONSTRUCTIONS

Presenter: Corbett Wilkinson, MD

Authors: Wilkinson C, French BM, Serrano CA, Stence NV

University of Colorado School of Medicine Childrens Hospital Colorado

Recently, we treated an infant with familial sagittal craniosynostosis, clinoccephaly, and bilateral parietomastoid suture synostosis, a pattern previously unreported. This suggests two questions: 1) Does craniosynostosis, particularly with clinoccephaly, often involve the lateral calvarial sutures? 2) When do these sutures normally close? Neither question is well-answered.

Methods: We reviewed all head CT volume-rendered reconstructions performed January 2010 through September 2012 at Childrens Hospital Colorado in 2 patient groups: 1) infants who subsequently underwent initial surgery for craniosynostosis 2) trauma patients, aged 0-18 years. Each parietomastoid, squamosal, and sphenoparietal suture was characterized as open, partially-closed, or closed. In the latter group, preexisting cerebrospinal fluid shunts, craniosynostosis, and possible involvement of a suture with fracture were exclusion criteria.

Results: Of 59 craniosynostosis subjects, there were 5 parietomastoid sutures, and no squamosal or sphenoparietal sutures closed. There were 3 cases of clinoccephaly; only the index case had parietomastoid suture fusion. Of 338 trauma patients, 89 sutures were excluded, including in a patient with undiagnosed sagittal synostosis, clinoccephaly, and bilateral parietomastoid suture fusion. Fusion typically began before 18 months at the anterior squamosal suture. Over 80% of sphenoparietal sutures were completely closed after 14 years; only 10% of parietomastoid sutures were completely closed by 17-18 years.

Conclusions: Lateral calvarial sutures are uncommonly involved in craniosynostosis. The current study is insufficiently powered to determine whether premature lateral suture fusion is associated with clinoccephaly. Normally, these sutures fuse in a characteristic pattern, often beginning at the anterior squamosal suture.



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CRANIOFACIAL FIBROUS DYSPLASIA

Presenter: S. Anthony Wolfe, MD
Authors: Satterwhite T, Wolfe SA
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Craniofacial Fibrous Dysplasia: Pick Your Neurosurgeon if you have a Contrarian View

From 1975 to 2012 we have treated 41 patients with craniofacial fibrous dysplasia, in conjunction with 5 different neurosurgeons. 39 patients were monostotic or polyostotic, and 2 were McCune Albright. 11 (26%) of the patients underwent optic nerve unroofing (ONU). Since this was a procedure performed by the neurosurgeon, we adhered to the indications of the particular neurosurgeon for ONU.

4 of the neurosurgeons were willing to perform ONU if the nerve was surrounded by fibrous dysplasia and surgery was being performed in adjacent areas to correct morphological deformation, without signs of compressive optic neuropathy (CON) such as visual acuity change, change in visual fields, red color desaturation change, and change in optical coherence tomography. The other neurosurgeon required objective signs of CON before he would perform ONU.

Results: 3 patients had total visual loss. One patient, with McCune Albright syndrome, underwent emergent bilateral ONU because of rapid deterioration of vision. He had some fluctuating vision in the first week postoperatively, but ended up with bilateral NLP. A second patient underwent 2 ONU's, with preservation of vision. She was being followed in an Eye Institute, and when seen 2 years later, had NLP in that eye. A third patient, also McCune Albright, underwent bilateral ONU after decompensation of visual acuity was noted. These procedures were performed a year apart. Because of further deterioration of vision on one side, and second ONU was performed. There is vague light perception in this eye now, but no useful vision.

We conclude: 1. The decision whether to perform ONU rests with the neurosurgeon

2. In no case did we have a deterioration or loss of vision when ONU was performed prophylactically.
3. ONU however, may not prevent eventual deterioration of vision when performed when visual changes have already been noted.
4. Fibrous dysplasia does not need aggressive treatment when involving tooth roots. The worst that may happen is tooth loss.

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INCIDENTAL FINDINGS AND THEIR CLINICAL SIGNIFICANCE IN PREOPERATIVE CT FOR SINGLE SUTURE NON SYNDROMIC CRANIOSYNOSTOSIS

Presenter: Keshav Magge, MD
Authors: Magge K, Magge SN, Myseros JS, Keating RF, Rogers GF, Oh AK
Childrens National Medical Center

Background and Purpose: While the diagnosis of single suture nonsyndromic craniosynostosis (SSNSC) can usually be made by clinical examination, computed tomography (CT) is often used in preoperative evaluation. The prudence of this practice has been questioned in light of recent studies that document a small increased risk of malignancy from CT-associated radiation exposure. The purpose of this study was to examine whether preoperative CT images for patients with SSNSC provided any clinically important information beyond the presence of the craniosynostosis.

Methods: We performed a retrospective analysis of all patients with SSNSC undergoing cranial vault remodeling at our center from 3/99-3/11. Only patients with complete preoperative CT scans available for review were included. Blinded to the patients' diagnosis and radiologists' official report, staff pediatric neurosurgeons analyzed the CT images and documented site of synostosis and any incidental findings.

Results: Of 231 patients, 80 met inclusion criteria. Sites of synostosis included: sagittal (51 patients), coronal (17 patients), metopic (11 patients), and frontosphenoidal (1 patient). Clinical diagnosis correlated with radiographic site of fusion in all patients except the patient with frontosphenoidal synostosis. Incidental findings were documented in over 50% of patients including: prominent extra-axial CSF (n=36; 45%); ventriculomegaly (n=5; 6.25%); choroid fissure cyst (n=2) cavum septum pellucidum (n=2), Chiari malformation (n=1), and prominent perivascular space (n=1). Incidental findings led to additional follow up or management in 5 patients (6.25%).

Conclusions: Our findings support preoperative imaging in this population to identify intracranial anomalies that cannot be discerned by clinical exam. Some of these findings are not clinically important, but may increase the likelihood of operative complications (e.g., prominent extra-axial CSF and possible increased subdural bleeding risk). While preoperative MRI could negate the cancer risk of CT, it is more costly and require general anesthesia (which may lead to neural apoptosis).



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THE USE OF BRAINLAB NAVIGATION IN LEFORT III OSTEOTOMY

Presenter: Jeyhan Wood, MD

Authors: Wood J, Thompson JT, David LR,
Argenta LC

Wake Forest Baptist Health

Background: LeFort III osteotomy is commonly used in the surgical management of patients with midface hypoplasia associated with syndromic craniosynostosis. LeFort osteotomies can be associated with significant complications, including hemorrhage. One main precipitating factor is related to nasofrontal and pterygomaxillary dysjunction and the force required for down-fracturing. Brainlab, a technology first developed to improve neurosurgical navigation, has been applied to numerous surgical subspecialties. The aim of this study is to report our experience using the Brainlab VectorVision2 system as an intraoperative guidance system for osteotomy placement during LeFort III advancement.

Methods: Three pediatric patients with Pfeiffer syndrome and midface hypoplasia scheduled to undergo LeFort III advancement were scanned using a CT scanner with 0.6mm cuts and these images uploaded to the Brainlab system preoperatively. All surgeries commenced with rigid fixation of the Brainlab registration device to the patients skull. The navigation system was used intraoperatively to accurately determine osteotomy sites.

Results: The first patient underwent a LeFort III osteotomy and placement of an external distractor. The second and third patients underwent LeFort III osteotomy and placement of both internal and external distractors. All surgeries were completed without complication. While there is additional time required for setup of the system, Brainlab image-guidance ultimately shortens overall operative time by eliminating the guesswork of osteotomy placement. We also noted a decreased transfusion requirement from non Brainlab assisted similar procedures.

Conclusions: The application of BrainLab technology to LeFort III advancement allows for accurately placed and completely performed osteotomies, especially in difficult to visualize areas. This can expedite the procedure, decrease the force required for down-fracturing of the midface and limit collateral tissue damage, which minimizes intraoperative blood loss. We recommend the use of a navigation system for image-guided osteotomy placement in LeFort III advancement.

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PREDICTING FACIAL SOFT TISSUE CHANGES FOLLOWING FACIAL OSTEOTOMIES USING GEOMETRIC MORPHOMETRICS AND THIN PLATE SPLINE WARPING

Presenter: Freida Angullia, MD

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Introduction: Predicting the overlying soft tissue changes resulting from facial osteotomies has proved to be a difficult task. To date, attempts to inform these predictions have relied on describing the degree to which overlying soft tissues move in response to advancement of a limited number of specific bony points defined on plain radiographs or CT scans. These techniques describe movements of specific points on the face but do not describe global changes that occur.

We describe a technique using geometric morphometrics and thin plate spline warping (TPSW) to predict the soft tissue changes that occur with facial osteotomies.

Method: 3D CT scans of 20 patients undergoing frontofacial distraction were analysed. Each pre and post operative scan was landmarked with predetermined validated homologous bony landmarks. The movements of these landmarks from pre to post surgery were analysed. TPSW is a technique predicting changes that occur to a surface by assuming materials in the model have uniform elasticity. In our model, the bony landmarks are warped and the extent to which overlying soft tissues move will depend on their thickness. Thicker soft tissues (eg. on the cheek) will move less than thinner soft tissues (eg. over the nose) mimicking the effects observed clinically. The bony movements undertaken were used to predict overlying soft tissue changes using TPSW then compared to actual soft tissue changes achieved.

Results: The predicted results generally matched the actual results within a few millimetres. The model was most effective over areas with the most reliable landmarks and where the tissues have a fairly uniform consistency (eg. central face and cheeks). The model was least effective in the orbital region where soft tissue anatomy is complex and does not move uniformly with the surrounding bone.

Discussion: Geometric morphometrics and thin plate spline warping provide a novel and promising approach to predicting soft tissue changes following facial osteotomies. Further adjustment to the mathematical algorithms used is needed to refine the model.



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LONG-TERM BIOMECHANICAL PROPERTIES OF BONE MORPHOGENETIC PROTEIN REGENERATED BONE IN FAVORABLE AND UNFAVORABLE CALVARIAL WOUNDS

Presenter: Zoe M. MacIsaac, MD
Authors: MacIsaac ZM, Henderson SE, Nayar HS, Shakir S, Naran S, Smith DM, Cray J, Mooney MP, Almarza A, Cooper GM, Losee JE
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Purpose: The limitations of autologous and alloplastic reconstruction for craniofacial defects have created a clinical need for viable tissue engineering strategies. While bone morphogenetic protein-2 (rhBMP-2) has shown promise in this setting, we predict that the biomechanical properties of the regenerate differ depending on wound environment.

Methods: 12-week old New Zealand white rabbits underwent subtotal calvariectomy. In Group 1 (Favorable), animals underwent immediate reconstruction with rhBMP-2 on an absorbable collagen sponge (rhBMP-2/ACS). In Group 2 (Unfavorable), bone flaps were inoculated with *S. aureus* and replaced. After 2 weeks of infection, wounds were debrided, bone flaps discarded, and antibiotic treatment instituted. Following a 6-week recovery, final debridement/reconstruction with rhBMP-2/ACS was performed. Defect healing was assessed with CT. An unconfined compression test was performed on three groups of bone. Samples were compressed at 0.1mm/min to 80% of initial thickness, followed by a 1-hour equilibration, then compression by 1800N. Results were normalized into stress and strain and compared by 1-way ANOVA.

Results: At 1 year, 100% and 99% healing was achieved in favorable and unfavorable defects. At 6 weeks, favorable reconstruction had less internal strength and was more compressible than native bone. At 6 months, favorable reconstruction gained equivalent internal strength, but remained significantly more compressible through 1 year. Bone in unfavorable reconstruction had less internal strength compared to native bone only at 6 months, but was significantly more compressible at 6 months and at 1 year. There were no significant differences between favorable and unfavorable groups.

Conclusion: Despite providing radiographic coverage, the biophysical properties of rhBMP-2 bone differ significantly from native. Bone generated in both favorable and unfavorable wounds remained more compressible through 1 year postoperatively. Further studies are warranted to determine how these properties affect overall strength and structural integrity.

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EVALUATION OF CRANIAL TRANSPORT DISTRACTION WITH AND WITHOUT ADIPOSE GRAFTING

Presenter: James Clune, MD
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Background: Transport distraction osteogenesis (DO) can be used to autologously reconstitute calvarial defects. However, distraction gap biology in transport DO has not been adequately described. The purpose of this study is to histomorphologically interrogate osteogenic formation during cranial transport distraction using a novel device. We also evaluate the effect of fat grafting on the regenerate during distraction.

Methods: This study was approved by Yale IACUC (# 2011-11393). Ten male New Zealand white rabbits (3 months; 3.5 kg) were used (8 treatment, 2 control). A 16x16 mm defect was created abutted by a 10x16 mm transport disc. The device was fixated anteroposteriorly. Four animals were fat-grafted using 2cc of subdermal intrascapular fat deposited along the distraction site. Latency (1d), active distraction (12-14d) (1.5 mm/day), and consolidation (4wks) followed. Calcein and xylene orange fluorochromes were injected subcutaneously during and post-distraction to mark sites of bone formation. Following sacrifice, osteogenesis was assessed using microCT, histology, and fluorescence.

Results: No perioperative complications were experienced. Treatment animals demonstrated regenerate bone between distracted segments on microCT. MicroCT analysis of fat-grafted and non-fat grafted animals revealed a mean density of 2271.95 mgHA/ccm and 2254.27 mgHA/ccm ($p=0.967$), respectively, and defect bone versus total volume (BV/TV) of 0.0999 and 0.0766 ($p=0.5979$), respectively. Controls had minimal reossification. Histologically, mean densities measured 43.63% and 8.19% for non-fat and fat grafted animals, respectively. Density ratios (regenerate:native bone) were 53.96% and 23.71%, respectively. Fluorescent microscopy revealed ossification from the callus as well as bone fronts emanating from dura and periosteum.

Conclusions: Transport distraction is effective to reconstruct critically-sized rabbit calvarial defects. Regenerate bone arises predominantly from the callus with contribution from surrounding dura and periosteum. Adipose grafting is well tolerated but does not enhance osseous regeneration



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AN EXPERIMENTAL STUDY OF PARTICULATE BONE GRAFT FOR SECONDARY INLAY CRANIOPLASTY OVER SCARRED DURA

Presenter: Arin K. Greene, MD
Authors: Maclellan RA, Hassanein AH, Mulliken JB, Rogers GF, Greene AK
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Background: Inlay cranioplasty in children is difficult because autologous bone is limited. Cranial particulate bone graft effectively closes defects when placed over normal dura. The purpose of this study was to determine if particulate bone graft will heal when used for secondary cranioplasty over scarred dura.

Methods: A 17mm x 17mm critical-sized defect was made in the parietal bone of 12 rabbits and allowed to heal. Sixteen weeks post-operatively the 17mm x 17mm critical-sized defect was recreated and managed in two ways: Group I (no implant) (n=6) and Group II (particulate bone graft) (n=6). Particulate graft was obtained using a brace and bit from the frontal bone and placed over the scarred dura. Gross analysis and micro-computed tomography were performed 16 weeks following the cranioplasty to determine the: (1) area of critical-sized defect ossification and (2) thickness of the healed bone graft.

Results: Critical-sized defects treated with particulate bone graft grossly exhibited superior ossification (96.0%; range, 86.5%-100%) compared to those managed without an implant (49.9%; range, 42.6%-54.6%) ($p < 0.0001$). MicroCT examination showed critical-sized defects treated with particulate bone graft healed 91.1% (range, 79.0-97.2%) of the area, while control defects demonstrated inferior ossification 56.9% (range, 40.0-68.3%) ($p < 0.0001$). Critical-sized defects treated with particulate bone graft exhibited thinner bone (2.42mm; range, 1.69-3.30mm) compared to the normal adjacent parietal cranium (4.33mm; range, 3.28-6.20mm) ($p < 0.0001$).

Conclusions: Particulate bone graft ossifies inlay calvarial defect area over scarred dura, although the bone is thinner than the normal cranium. Clinically, particulate bone graft may be efficacious for secondary inlay cranioplasty.

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REPAIR OF A COMPLICATED CALVARIAL DEFECT: RECONSTRUCTION OF A WOUND COMPLICATED BY DURECTOMY AND INFECTION

Presenter: Sanjay Naran, MD
Authors: MacIsaac ZM, Shakir S, Naran S, Cray J, Smith DM, Kubala A, Kinsella CR, Mooney MP, Cooper GM; Losee JE
University of Pittsburgh

Background: Recombinant human bone morphogenetic protein-2 (rhBMP-2) has been shown to be an effective therapy in the acute, uncompromised calvarial defect. This study compared the efficacy of rhBMP-2 mediated bone regeneration with the gold standard of autologous bone graft for repair of calvarial defects complicated by 1) previous infection and 2) durectomy with dural repair.

Methods: 18 adult New Zealand White rabbits underwent subtotal calvariectomy and dural removal, followed by dural repair. Bone flaps were cryopreserved. After 6 weeks, animals received bone flaps inoculated with *S. aureus*. After 6 days of infection, bone flaps were removed and wounds debrided, followed by 10 days of antibiotics. After 6 additional weeks of recovery, defects were debrided and definitive reconstruction performed in one of 4 groups: empty (no treatment, n=3), vehicle (buffer on an absorbable collagen sponge (ACS), n=2), autologous bone graft (cryopreserved calvarial bone graft, n=3), or rhBMP-2 (rhBMP-2 on ACS, n=10). Animals underwent CTs at 0, 2, 4 and 6 weeks. At 6 weeks, defects were examined histologically. Percent healing was determined by image analysis. A (time x group) 2-way ANOVA was performed on healing vs. group and postoperative time.

Results: At 6 weeks, rhBMP-2/ACS and autologous bone groups were statistically equivalent with 80% and 96% healing respectively. Empty and vehicle groups, (14% and 18% healing respectively) at 6 weeks, were inferior to the rhBMP-2/ACS and autologous bone graft groups at each timepoint ($p < 0.00$). Histologically, bone in the autologous bone graft group was less trabecular and less cellular than the bone formed in the experimental treatment group (rhBMP-2/ACS). rhBMP-2 performed equivalently in this complicated defect compared to immediate calvarial repair ($p=0.97$).

Conclusions: Compared to cryopreserved autologous bone graft, rhBMP-2 regenerated bone resulted in equal defect coverage and similar thickness with varying bony architecture, with greater cellularity. Further studies are necessary to demonstrate the long-term viability and remodeling of this treatment modality.



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IDENTIFICATION AND CHARACTERIZATION OF NEUROCRANIAL SKELETAL PROGENITOR CELLS

Presenter: Adrian McArdle, MD

Authors: McArdle A, Chan CK, Hyun JS, Chung MT, Montoro DT, Wan DC, Weissman IL, Longaker MT

Stanford University

Introduction: Many recent studies have focused on osteogenic cellular sources for cell-based bone tissue engineering with variable success. However, a distinct source of skeletogenic-specific progenitor cells has not been identified. Since the developing neurocranium is a particularly dynamic structure that undergoes rapid bone formation, we hypothesized that it should be replete with progenitor cells that possess accelerated bone regenerative capabilities. In this study, we prospectively isolated multiple novel progenitor subtypes from developing mouse neurocranium to characterize their specific skeletogenic potential.

Methods: Skeletal elements from the calvaria of GFP transgenic mice were dissected from early postnatal mice (P1~P3) and digested in collagenase. Collagenase-dissociated cells were then stained with fluorochrome-conjugated antibodies against cell surface proteins that are differentially expressed on skeletogenic populations and fractionated by FACS sorting. Each prospectively isolated population was compared on the basis of their ability to differentiate into bone and cartilage both *in vitro* and *in vivo*, by transplantation into a 4-mm critical-sized calvarial defect in nude mice.

Results: We identified four functionally distinct skeletal progenitor populations based on differential expression of CD45, CD31, CD105, Thy1.1, and integrin α v. Noticeably, the CD105+Thy1.1- progenitor population consistently gave rise to donor-derived endochondral bone with a marrow cavity. In contrast, CD105+Thy1.1+ progenitors resulted in *de novo* donor-derived ectopic bone formation absent of marrow and a cartilaginous intermediate reminiscent of intramembranous ossification. All other fractions failed to consistently produce any donor-derived bone.

Conclusions: Our preliminary data support the presence of distinct skeletal progenitor subsets that reside in early post-natal neurocranium with the capability of *de novo* bone formation. Importantly, we have identified specific progenitors for both endochondral ossification and intramembranous ossification – the two major types of osteogenesis in the neurocranium.

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EVIDENCE FROM THE FGFR2C342Y/C342Y CROUZON MURINE MODEL ARGUES FOR A PRIMARY FAILURE IN CHONDROGENIC DEVELOPMENT

Presenter: Erwin Pauws, PhD

Authors: Kumar S, Peskett E, Britto JA, Pauws E
University College London

Crouzon syndrome, most commonly caused by the FGFR2C342Y/+ mutation, is characterised by developmental pathology in both endochondral and membranous skeletogenesis. Spatio-temporal correlation of the pathogenesis of features characterising the membranous cranial vertex and the chondrogenic cranial base and other structures in the axial and appendicular skeleton, has not been defined at embryonic stages. We contrast membranous/endochondral phenotypes in the mouse heterozygote Fgfr2C342Y/+ and homozygote Fgfr2C342Y/C342Y against wild-type controls between embryonic day (E)15.5 to E18.5.

Whole mount staining of CD1 wild-type, Fgfr2C342Y/+ and Fgfr2C342Y/C342Y mouse embryos (E13.5-E18.5) with Alcian Blue and Alizarin Red was performed and studied together with parasagittal sections of the cranial base. *In situ* hybridisation for Sox9 was undertaken in the developing trachea and cranial base.

The differences observed in Fgfr2C342Y/C342Y embryos include: (1) anterior-posterior shortening of the cranial base with cartilaginous disorganisation (2) delayed ossification of the posterior cranial base (3) expansion of the basicranial cartilaginous precursor resulting in fusion with the tympanic bullae (4) abnormal rib patterning with a midline sternal defect (5) delayed intramembranous ossification of the bony calvaria (6) cervical and thoracic vertebrae fusions (7) tracheal cartilaginous sheet in homozygote embryos, and an intermediate phenotype in heterozygotes showing fused rings in the proximal part of the trachea. The Sox9 expression domain in tracheal specimens at E13.5 in both Fgfr2C342Y/+ and Fgfr2C342Y/C342Y is expanded, correlating with the phenotypic observations.

A spectrum of phenotypic variations observed in the Fgfr2C342Y/C342Y murine embryo paves the way towards better understanding the extracranial features seen in the human Crouzon syndrome. FGFR2c gain of function results in impaired skeletogenesis, however our findings suggest that the phenotypic aberrations stem from a primary failure of chondrogenic growth and links SOX9, principal regulator of chondrogenic differentiation, to FGFR2c.



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OSTEOGENIC PERFORMANCE OF DONOR MATCHED HUMAN ADIPOSE AND BONE MARROW MSCS UNDER DYNAMIC CULTURE

Presenter: Miles Pfaff, MD

Authors: Pfaff MJ, Zellner E, Niklason LE, Steinbacher DM

Yale University School of Medicine

Introduction: Tissue engineered bone holds translational promise for myriad applications in reconstructive surgery. Both adipose-derived and bone marrow-derived stem cells (ADSCs and BMSCs) have been used for bone regeneration, and can be seeded on a variety of rigid scaffolds. This study aims to compare ADSCs and BMSCs from the same donor in three distinct bioreactor settings to create the most viable osseous engineered construct. We hypothesize that physiologic flow dynamics will optimize osteogenic cell viability and function, which are prerequisites to successful human tissue implantation.

Methods: Human ADSCs and BMSCs were isolated from the same donor, then cultured and seeded on decellularized porcine bone constructs. The constructs were subjected to static or dynamic (stirring or perfusion bioreactor) culture conditions for 7 to 21 days. The constructs were then analyzed for cell adhesion and distribution using histology and electron scanning microscopy. Proliferation and osteogenic differentiation were gauged using DNA quantification, alkaline phosphatase (ALP) assay, immunostaining for osteocalcin and real-time-PCR, and calcium deposition assay.

Results: hADSCs demonstrated higher seeding efficiency and proliferation in static culture than hBMSCs. However, dynamic culture, driven by stirring or perfusion flow, significantly increased BMSCs proliferation more than ADSCs proliferation. The highest cellularity was seen in the stirring bioreactor. In all conditions, BMSCs demonstrated stronger osteogenic activity compared to ADSCs, in ALP activity assay and gene expression for various bony markers. Conversely, ADSCs expressed more collagen I. In all constructs (ADSC and BMSC), dynamic conditions enhance overall osteogenic gene expression. BMSCs in the stirring bioreactor exhibited the greatest calcium production, likely secondary to the greater cell proliferation and osteogenic function.

Conclusions: Scaffolds seeded with BMSCs in dynamic conditions exhibit the greatest osteogenic proliferation and function. In particular, the stirring bioreactor optimizes the bone engineered construct.

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NAGER SYNDROME DENTAL PULP STEM CELLS SHOW SUPERIOR OSTEOGENIC POTENTIAL COMPARED TO TREACHER COLLINS SYNDROME

Presenter: Joyce T. Yuan, MD

Authors: Yuan JT, Bueno D, Tabit CJ, Bradley JP
University of California Los Angeles

Background: Nager syndrome patients with severe micrognathia, malar hypoplasia, cleft palate (at times), and radial club hand deformities require bone reconstruction for severe deformities. Treacher Collins patients have similar symptoms without hand abnormalities. Novel stem cell therapies may be used to decrease donor site morbidity in these patients. We isolated stem cells from dental pulp tissue of Nager and Treacher Collins patients and looked at osteogenesis.

Methods: Deciduous teeth in Nager, Treacher Collins, and normal patients were used to isolate dental pulp stem cells (DPSCs) with a pre-plating technique. Harvested DPSCs were confirmed with stem cell antigens (CD29, CD90, CD105, CD166) and tested for hematopoietic cell markers (CD34, CD45) and endothelial cell markers (CD31) to confirm mesenchymal origin. After 7 days in osteogenic media, osteogenesis was tested with von Kossa staining and RT-PCR (runx2, c-fos, osteocalcin, osteonectin and osteopontin). Nager and Treacher Collins cells were also suspended in a 3D collagen matrix and stressed in an in vivo microdistractor to study osteogenic potential.

Results: Isolated Nager and Treacher Collins DPSCs were positive for mesenchymal stem cell antigens while negative for hematopoietic cell markers and endothelial cell marker. Cells were capable of osteogenic differentiation as evidenced by positive von Kossa staining and real time PCR for osteogenic genes: runx2 (3.6/2.4 fold), c-fos (2.2/1.8 fold), osteocalcin (12/7 fold), osteopontin (3.1/2.0 fold). Comparison with preosteoblasts and normal DPSCs confirmed differential osteogenic expression of Nager and Treacher Collins cells. Linear stress of Nager but not Treacher Collins cells within the microdistractor showed earlier osteogenic expression compared with non-stressed cells and controls.

Conclusions: Mesenchymal cells from the dental pulp of Nager patients have osteogenic potential that can be used to correct hypoplastic mandibles. Isolated Treacher Collins cells did not have the same robust osteogenic potential. Exfoliated deciduous teeth are a promising source of stem cells in syndromic patients.



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**HEDGEHOG GRADIENT MODULATES NASAL
PROCESSES GROWTH AND FUSION BY
ANTAGONIZING CANONICAL WNT SIGNALING**

Presenter: Hiroshi Kurosaka, PhD, DDS
Authors: Kurosaka H, Trainor PA
Stowers Institute for Medical Research

Craniofacial development depends on the proper fusion of distinct embryonic processes. It is well known that during fusion, epithelial cells at the tip of each process are eliminated by several mechanisms including cell death and EMT. Removal of this periderm results in the formation of direct mesenchymal bridges or connections between each process. Failure to complete fusion leads to various types of orofacial clefting such as cleft lip and palate (CLP). One of the most common sites of fusion failure is at the lambdoidal junction between the maxillary, and medial and lateral nasal processes. In the present study, we examined the spatiotemporal requirement for Hh signaling in the etiology and pathogenesis of CLP through synergistic mutations in Hhat and Patched1. Compound mutant mice exhibit primary cleft palate due to failure of the nasal processes to fuse at the lambdoidal junction. Altered domains of cell death together with persistence of the periderm domain in the lambdoidal region mechanistically underlies the pathogenesis of this defect. To investigate the gene regulatory network governing this phenotype, we performed RNA-seq analyses and discovered that canonical Wnt signaling is reduced in the lamdoidal junction of compound Hh signaling mutants. Furthermore, we identified several Wnt inhibitory genes such as the sFRP family whose expression domains are expanded in Hh signaling mutants. Thus a precise level of Hh signaling to control canonical Wnt signaling at lambdoidal junction for proper nasal processes development and fusion.

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**TRANSFORMING GROWTH FACTOR BETA 1
IMPROVES BONE HEALING AND PROMOTES SUTURE
REGENERATION**

Presenter: Sameer Shakir, BS
Authors: Shakir S, MacIsaac ZM, Naran S, Gilbert JR,
Shaw MA, Wang D, Kubala AA, Losee JE,
Cooper GM
University of Pittsburgh

Background: Craniosynostosis (CS) is a congenital disorder defined by premature fusion of the cranial sutures. Conventional therapy for CS requires surgical intervention to decrease intracranial pressure and improve craniofacial development, but outcomes are frequently complicated by the recurrence of fusion. We hypothesized that tailored growth factor therapies would facilitate calvarial repair and restore suture function.

Methods: Suturectomies (3x15 mm) were performed in 10-day-old New Zealand White rabbits (n=19) and treated with acellular dermal matrix (3x15 mm) biopatterned with 350ng BMP2, 200ng TGFβ1, 350ng BMP2 + 200ng TGFβ1, or nothing (control). Rabbits were euthanized 4 weeks post-surgery. Bone healing was quantified by micro-CT and tissue morphology was assessed histologically.

Results: Statistical analysis of micro-CT data revealed significant differences in the degree of bone healing among the experimental therapies compared to the control; however, no significant differences were observed when comparing the different therapeutic interventions. Histologically, control animals exhibited extensive fibrous tissue within regenerating defects. Bone repair within the BMP2 group occurred through endochondral ossification. The TGFβ1 treatment group produced bone similar to native bone in terms of cellularity and matrix density. Animals within the TGFβ1 treatment group also showed evidence of suture-like tissue within the defect margin comprising collagen and blood vessels. Suture-like tissue was not observed in the other groups.

Conclusions: TGFβ1 growth factor delivery augmented bone healing and produced bone through intramembranous ossification, unlike BMP2-generated bone. Furthermore, TGFβ1 treatment allowed for suture-like tissue regeneration in this model of craniofacial reconstruction. Interestingly, the presence of BMP2 did not allow for suture regeneration and did not augment defect healing when compared to other experimental treatments. Therefore, TGF-beta treatment may be a viable therapeutic option to enhance healing after craniofacial surgery to manage CS.



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CALVARIAL RECONSTRUCTION WITH BONE MARROW CELLS: CONCOMITANT TREATMENT WITH BMP-2

Presenter: Matthew Greives, MD
Authors: MacIsaac ZM, Shakir S, Naran S, Zammerilla L, Cooper GM, Losee JE
University of Pittsburgh

Background: While bone morphogenetic protein-2 (BMP-2) demonstrates promise as a therapy for calvarial bone regeneration, its application remains controversial due to concern for side effects such as heterotopic ossification or malignant transformation. Meanwhile, bone marrow cells (BMCs) have demonstrated therapeutic promise, but therapies remain to be optimized. The aim of this study was to augment the efficacy of bone marrow cell mediated bone regeneration through application of a low dose of rhBMP-2.

Methods: Subtotal calvariectomy defects measuring 7.5 x 7.5 mm² were created in 12-week old New Zealand White rabbits. Defects were reconstructed in one of three groups: Group 1, acellular dermal matrix (ADM) soaked in PBS overnight [ADM/PBS, n=3]; Group 2, ADM soaked overnight in DMEM with 150,000 bone marrow cells from the femur of a New Zealand White rabbit [ADM/BMCs, n=3]; and Group 3, ADM and BMCs with 1.78 μ g of BMP-2 applied by ink-jet based biopatterning [ADM/BMCs/BMP-2 n=3]. Animals underwent serial CT imaging at 0, 2, 4 and 6 weeks postoperatively, followed by euthanization and histological analysis. CTs were reconstructed utilizing Amira software (Visage Imaging, San Francisco, CA, USA), and areas of translucency were calculated using ImageJ (NIH) to determine change in defect area relative to time zero (baseline value).

Results: At 6 weeks compared to time zero, ADM/PBS resulted in 45.2% healing (standard deviation 30.1%), and ADM/BMCs resulted in 54.1% healing (standard deviation 12.7%). Treatment with ADM/BMCs/BMP-2 resulted in 78.6% healing (standard deviation 11.6%); there was no significant difference between groups. Histologically, bone in each group was similar, consisting of islands of compact, cellular bone.

Conclusions: Compared to treatment with ADM/PBS only, addition of BMCs for reconstruction of an acute calvarial defect trended toward better results, and addition of BMP-2 to this regimen further optimized outcomes. Combination cell/growth factor therapy may enable successful reconstruction of calvarial defects without side effects related to higher dose.

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TRANS-ORAL POSTERIOR MAXILLARY CRANIOFACIAL SURGERY TO PLACE A SPHENOPALATINE GANGLION (SPG) NEUROSTIMULATOR FOR TREATMENT OF CHRONIC CLUSTER HEADACHE (CCH): PATHWAY CH-1 STUDY SURGICAL EXPERIENCE

Presenter: Frank A. Papay, MD
Authors: Papay FA, Hillerup S, Wilmont A, Puche M, Pohlenz P, Muller O, Fontaine D, Blessman M, Caparso A

Cleveland Clinic

Aim: The pain and autonomic symptoms of cluster headache may result from activation of the trigeminal parasympathetic reflex, mediated through the sphenopalatine ganglion (SPG). We aimed to investigate the surgical safety of SPG stimulation for the acute treatment of cluster headaches.

Material and Methods: In a multi-center study subjects are implanted with a miniaturized neurostimulator using a trans-oral technique. The neurostimulator is implanted such that the electrodes are placed in close proximity to the SPG. The body of the neurostimulator is placed on the lateral posterior maxilla and the fixation plate is used to anchor the neurostimulator on the superior lateral zygomaticomaxillary buttress. The procedure is performed under general anesthesia and utilizes fluoroscopic or CT imaging.

Results: 43 subjects have undergone the implantation procedure for the ATITM Neurostimulator. In open label use, 81% of subjects have achieved pain relief in >50% of acute headaches and/or have experienced a >50% reduction in headache frequency with SPG stimulation. Three subjects (7%) were explanted due to early lead migrations or misplacement of the neurostimulator and 4 subjects (9%) underwent a revision procedure due to lack of efficacy and incorrect electrode locations. In 1 subject (2%) the procedure was not completed due to anatomical limitations. Within the first 30 days post surgery, 20 subjects (47%) reported mild to moderate numbness in the second division of the trigeminal nerve. 62% of these events, in 12 subjects, resolved within 90 days of onset on average. 38% are currently un-resolved, in 8 subjects, with an average duration of 163 days. Additionally, no infections resulting in explant have occurred.

Summary: The initial experience using the minimal invasive implantation procedure for the ATITM Neurostimulator has shown an acceptable safety profile that is comparable to standard trans-oral surgery. Advancements in surgical instruments and further surgical experience has had a positive impact in overall subject experiences.



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CELL-ASSISTED LIPOTRANSFER WITH BONE MORPHOGENETIC PROTEIN RECEPTOR IA+ ADIPOSE-DERIVED STROMAL CELL SUBPOPULATIONS

Presenter: Kevin J. Paik, AB
Authors: Paik KJ, Chung MT, Chen HH, McArdle A, Hu MS, Lo DD, Hyun JS, Montoro DT, Senarath-Yapa K, Zimmermann AS, Walmsley G, Sorkin M, Rennert R, Chung AS, Longaker MT, Wan DC

Stanford University

Background: Due to significant resorption rates (20-80%) with autologous fat grafting, there is a need to develop techniques that increase the retention of graft volume over time. Yoshimura previously investigated the efficacy of supplementing fat grafts with ASCs, a strategy known as cell-assisted lipotransfer (CAL). As one of the believed roles ASCs play in fat grafts is to differentiate into fat cells themselves, the present study took a more targeted look at the CAL strategy by enriching supplemental ASCs for BMPR-IA, a marker associated with adipocyte differentiation.

Methods: BMPR-IA+ and BMPR-IA- ASCs were sorted using magnetic activated cell sorting (MACS) and verified by fluorescence activated cell sorting (FACS). After treatment with adipogenic differentiation media in vitro for seven days, Oil Red-O staining and quantification were conducted on both subpopulations and unsorted ASCs, as was qRT-PCR for adipogenic gene expression (AP2, LPL, and PPAR-c). In vivo, human fat grafts prepared from lipoaspirate samples using the Coleman technique were injected subcutaneously into the scalps of nude mice with BMPR-IA+ ASCs, BMPR-IA- ASCs, or without ASCs (n=4 per group). Micro-CT was performed after three days for baseline, and then every two weeks for eight weeks. Fat volume was rendered by reconstructing a 3D surface through cubic-spline interpolation. At Week 8, fat volumes were explanted for gross inspection, weighing, and histology.

Results: BMPR-IA+ cells showed higher adipogenic gene expression in qRT-PCR. Oil Red-O staining and quantification showed more lipid droplet formation in BMPR-IA+ cells. Over the course of eight weeks, the BMPR-IA+ enriched fat grafts were found to consistently show greater volume retention than the BMPR-IA- enriched fat grafts, the fat grafts with unsorted ASCs, or the fat grafts alone.

Conclusions: Our findings demonstrate that subpopulations of ASCs may be identified with enhanced adipogenic capacity and suggest potential clinical benefit in performing cell-assisted lipotransfer with BMPR-IA+ cells for soft tissue augmentation.

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REFLECTANCE CONFOCAL MICROSCOPY MAY REDUCE THE NEED FOR SKIN BIOPSY IN COMPOSITE TISSUE ALLOTRANSPLANTATION

Presenter: Huseyin Karagoz, MD
Authors: Zor F, Karagoz H, Erdemir AV, Karslioglu Y, Acikel CH, Kapaj R, Guzey S, Gurel MS, Ozturk S

Gulhane Military Medical Academy

Composite tissue allograft can undergo immune-mediated rejection therefore routine biopsies are needed during the follow-up period. These biopsies enable the determination of the severity of the rejection and provide adjustment of the drugs. However, skin biopsy is an invasive method, needs a processing time to obtain the results and taking biopsy from a flap in a lifelong period is not feasible. Thus, noninvasive methods of determination of the severity of the rejection are needed. In this study, a noninvasive method of determination of rejection severity is developed by using reflectance confocal microscopy (RCM) and the results are compared with Banff classification. Five composite tissue allograft transfers were performed between HLA mismatched male Sprague Dawley rats. The rats were given 16 mg/kg CsA for 10 days postoperatively and resolution of postoperative edema was waited. At day 10th postoperative day, CsA was stopped and the rejection process is enabled. First evaluation was performed at 10th postoperative day and repeated each other day until the rejection is completed. The rejection severity is determined with two different techniques: In vivo RCM observation and skin biopsy. Reflectance confocal microscopy is an in vivo imaging system which enables imaging of skin at histologic resolution. During the evaluation, in vivo observation with RCM is performed and skin biopsy was taken from the observation site. During the RCM observations epidermal appearance, circulation, inflammatory cells, ground glass appearance and collagen structure was evaluated. A scoring system is developed which has lowest score "0" as normal skin and highest score "10" as acute rejection. A total of 17 specimens were evaluated and both RCM scores and Banff grades of the specimens are determined. Spearman and Kendal's tau correlation tests revealed correlation between RCM scores and Banff grades (Correlation Coefficient=0.653) and the correlation was significant at the 0.01 level. In this study, a noninvasive method of composite tissue allograft rejection grading system based on reflectance confocal microscopy is developed.



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PREOPERATIVE LIP MEASUREMENT IN PATIENTS WITH UNILATERAL COMPLETE CLEFT LIP/PALATE AND ITS COMPARISON WITH NORMS

Presenter: PangYun Chou, MD

Authors: Chou PY, Luo CC, Chen KT, Chen YR, Noordhoff MS, Lo LJ

Chang Gung Memorial Hospital

Purpose: There is prominent lip asymmetry in patients with unilateral complete cleft lip and palate. Measurement of lip on cleft and non-cleft sides provides appraisal of the lip deformity and information for planning of surgical correction. The purpose of this retrospective study is to evaluate the lip deformity and to compare with normative data.

Materials and Methods: From 1983 to 1997, data from a total of 168 patients with unilateral complete cleft lip and palate were collected. There was no other associated craniofacial anomaly in this patient group. The measurement was performed under general anesthesia by a senior surgeon using a caliper prior to the first lip repair. Corresponding normative data were collected from 2002 to 2003 on 50 patients who had normal facial appearance prior to hernia repair. The measurements included lip height, lip width, philtrum length, and vermilion thickness. Comparisons were made between cleft side and non-cleft side, as well as between cleft patients and norms.

Results: Comparisons between cleft and non-cleft sides revealed significantly longer lip on the non-cleft side, including lip height from alar base to Cupid's bow, lip width from Cupid's bow to commissure, and the vermilion thickness. The lip measurements on the norms were longer than those on the cleft side of the lip, but were similar to the non-cleft side.

Conclusion: A wide variety of tissue growth asymmetry is observed between non-cleft and cleft sides, indicating a deficiency of tissue development associated with cleft deformity. These data can provide a fundamental basis for presurgical orthopedic treatment, surgical planning, execution of surgery, postoperative assessment, and a predictor for the treatment outcome.

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BIOMECHANICAL ASSESSMENT OF CALVARIAL RECONSTRUCTION: REPAIR OF A CRANIAL DEFECT WITH CRYOPRESERVED AND FRESH AUTOLOGOUS GRAFT

Presenter: Seun Adetayo, MD

Authors: MacIsaac ZM, Shakir S, Henderson S, Naran S, Cray J, Smith DM, Mooney MP, Almarza A, Cooper GM, Losee JE

University of Pittsburgh

Background: Of the materials available for cranioplasty, autograft has consistently been the gold standard due to its superior incorporation and lack of immune response. The purpose of this study was to compare biomechanical properties of fresh, autologous reconstruction, versus reconstruction with cryopreserved autologous graft.

Methods: 16 adult New Zealand White rabbits underwent subtotal calvariectomy. In Group 1 and 2, bone flaps were immediately replaced to repair the defect. In Group 3, defects were repaired with cryopreserved calvarial bone graft (in-colony), which had been stored at -80°C. Animals underwent imaging at 0 and 6 weeks, and in Group 2, at 6 months, followed by histological analysis. Biomechanical analysis with an unconfined compression test was performed on the three groups of bone. Samples were compressed at 0.1 mm/min to 80% initial thickness, followed by a 1-hour equilibration at 80% initial thickness, then compression by 1800N. Results were normalized into stress and strain and compared by 1-way ANOVA. Percent healing was determined by 3-dimensional analysis.

Results: Groups 1 and 2 demonstrated 87.3% and 82.4% healing at 6 weeks postoperatively; Group 3 demonstrated 100% healing at 6 months. Histologically, lacunae appeared more cellular at both time points for fresh autologous reconstruction, compared to cryopreserved. Pentachrome staining revealed more remodeling in cryopreserved reconstruction. Results of biomechanical testing are reported in Table 1. There were no significant differences between time points for fresh autologous graft, nor were there significant differences between fresh and cryopreserved bone graft at 6 weeks.

Conclusions: Both fresh and cryopreserved calvarial graft provided radiographic coverage; both methods resulted in equivalent biomechanical properties. At 6 weeks, cryopreserved graft exhibited more remodeling than did fresh autologous graft. Further studies are warranted to determine how biomechanical properties evolve over the long term to effect on overall strength and structural integrity.



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EFFECTS OF HYPOTENSIVE ANESTHESIA ON BLOOD LOSS DURING CRANIOSYNOSTOSIS CORRECTIONS

Presenter: Kevin Cook, MD

Authors: Fearon JA, Cook TK, Herbert MA
The Craniofacial Center

Purpose: Hypotensive anesthesia (MAP = 50mmHg) is routinely used to reduce blood loss during craniostomosis corrections. Utilizing Near-Infrared Spectroscopy (NIR) we noted cerebral oxygenation levels often fell below recommended levels during craniostomosis corrections. Based on these observations, we sought to measure the effects of standard vs. minimal hypotension on blood loss during craniostomosis corrections.

Methods: 75 children undergoing craniostomosis corrections were prospectively randomized into two groups: MAP = 50 mmHg or 60 mmHg. Aside from the anesthesiologists, all caregivers were blinded and strict transfusion criteria were followed. Multiple variables were analyzed including blood loss and transfusions, and appropriate statistical testing was performed.

Results: Our analyses revealed that both standard and minimal hypotension groups were similar; no statistically significant differences noted with respect to: mean age (43 vs. 50 months), weight (20.1 vs. 18.5 kgs.), procedure (anterior remodeling (25 vs. 25) vs. posterior (12 vs. 15)), preoperative hemoglobin levels (12.9 vs. 13gm/dl.). There were also no differences noted in calculated blood loss as ascertained by captured cell saver products (245 vs. 274cc.), nor in postoperative hemoglobin levels (8.8 vs. 9gm/dl.). Eleven of 75 patients (14.7%) received allogenic transfusions, but there were no statistically significant differences found between transfusion rates in the standard (7/39, 18.0%) vs. minimal hypotension (4/36, 11.1%) groups.

Conclusions: This prospective, blinded, randomized study found no statistically significant differences in intraoperative blood loss and transfusion requirements between two target levels for hypotensive anesthesia (standard vs. minimal hypotension). Based on our observations of occasional cerebral hypoxia by NIR monitoring, and the absence of statistically significant differences in blood loss at higher MAP's, we recommend that surgeons consider performing craniostomosis corrections with minimum hypotension to improve cerebral blood flow and total body perfusion.

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EVALUATING THE SAFETY AND EFFICACY OF TRANEXAMIC ACID ADMINISTRATION IN PEDIATRIC CRANIAL VAULT RECONSTRUCTION

Presenter: John C. Crantford, MD

Authors: Crantford JC, Claiborne JR, Wood BC, Ririe DG, Thompson JT, David LR
Wake Forest Baptist Health

Background: Blood loss is the leading cause of mortality following major craniofacial surgery. Autologous blood donation, short-term normovolemic hemodilution, and intraoperative blood salvage have shown low efficacy in decreasing transfusions. Tranexamic acid (TXA) is a synthetic antifibrinolytic drug that competitively decreases the conversion of plasminogen to plasmin, thereby suppressing fibrinolysis. The purpose of this study was to investigate the impact TXA administration has on intraoperative blood loss and blood product transfusion in pediatric patients undergoing cranial vault reconstruction.

Methods: An IRB-approved retrospective study was conducted on pediatric patients undergoing cranial vault reconstruction from January 2009 to June 2012. Twenty consecutive patients that received TXA at the time of cranial vault reconstruction were compared to twenty patients that did not receive TXA. Criteria for blood product transfusion were identical for both groups. Outcomes including perioperative blood loss, volume of blood transfused, and any adverse effects were analyzed.

Results: The TXA group had a significantly lower preoperative hematocrit (33.8 g/dL vs 35.7 g/dL, $p < 0.042$), longer duration of surgery (199 mins vs 170 mins, $p < 0.042$), lower perioperative blood loss (9.6 mL/kg vs 20.5 mL/kg, $p < 0.0003$), and lower volume perioperative mean blood product transfusion (13.4 mL/kg vs 26.5 mL/kg, $p < 0.0018$) compared to the non-TXA group. There was no significant difference in demographic data, infection rate, postoperative hematocrit, change in preoperative to postoperative hematocrit, or complication rates between the TXA and non-TXA groups. No drug-related adverse effects were identified in patients that received TXA.

Conclusions: The use of TXA in pediatric cranial vault reconstruction significantly reduces perioperative blood loss and blood product transfusion requirements, despite the TXA group undergoing significantly longer operative times. TXA administration is safe and may improve patient outcomes by decreasing the likelihood of adverse effects related to blood product transfusion.



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TECHNIQUES TO MINIMISE PATIENT EXPOSURE TO DONATED BLOOD; A 10 YEAR AUDIT OF 450 CONSECUTIVE, NON SYNDROMIC, PAEDIATRIC, CALVARIAL REMODELLING PROCEDURES

Presenting: Carol Millar, MD
Authors: Evans MJ, White N, Nishikawa H, Dover MS, Solanki G, Rodrigues D, Carver E, Millar C, Johnson D

Birmingham Childrens Hospital

Introduction: Surgery for craniosynostosis is associated with significant blood loss that can vary, ranging from 20-500% of estimated blood volume (EBV). Factors such as age of patient, length of procedure and whether the craniosynostosis is syndromic or non-syndromic are important.

Methods: We have prospectively collected data over 10 years relating to blood loss, blood products used and whether autotransfusion has been used for more than 450 consecutive calvarial remodelling procedures.

Results: During the study period, blood loss per patient was in the range 43-172% of EBV. Most patients have required a transfusion of packed red cells but the number of units transfused per patient has reduced during the 10 years, particularly in cases where cell salvage has been utilised for autotransfusion. Use of fresh frozen plasma has increased, but platelet transfusion and cryoprecipitate use has been low and remains unchanged. Recently we have used peroperative Tranexamic Acid as an adjunct and comparison of its efficacy in this cohort of patients will be illustrated.

Conclusion: This paper highlights the techniques used to minimize exposure to donated blood products in our 10 year series of FOAR and calvarial remodelling procedures for non syndromic craniosynostosis. The patient safety issues will be highlighted.

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INTRAOPERATIVE BLOOD TRANSFUSION PATTERNS IN SURGERY FOR NON SYNDROMIC CRANIOSYNOSYOSIS

Presenting: Llewellyn C. Padayachy, MD
Authors: Padayachy LC, Fieggen AG, Figaji AA, Micheals J, Lechthape-Gruther R, Peter J
University of Cape Town

Introduction: Blood transfusion during craniosynostosis almost routine. The blood product used, transfusion threshold and volume of blood transfused, however, differ significantly. We have analysed data from 45 patients undergoing surgery for primary craniosynostosis repair at our institution, from January 2008 to December 2012. Packed red cells were used in 97%, with whole blood used in 3%. The mean pre-operative hemoglobin (Hb) was 11.46 g/dL, mean post-operative Hb was 11.35 g/dL. The mean blood volume transfused was 233.34 ml (25.56ml/kg). The mean operating time was 171.20 minutes, demonstrating a positive predictive value of blood volume transfused of 83.8% (sensitivity: 97.8%, specificity: 78.5%), irrespective of the type of surgery being performed. Intra-operative complications, as well as any episodes of hemodynamic instability, requiring additional blood were noted and controlled for in the analyses. Blood transfusion was considered appropriate if the post-operative Hb was within 15% of the pre-operative value. Post-operatively 45% of patients receiving blood transfusions were transfused appropriately, with 75% of patients transfused to within 20% of their pre-operative value. No complications related purely to volume of blood transfusion were noted. Blood transfusion practices at our institution are appropriate, with length of operating being the strongest predictor of blood volume transfused.



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EFFECTS OF TYPE OF SURGICAL INTERVENTION ON NEUROPSYCHOLOGICAL OUTCOMES IN SAGITTAL CRANIOSYNOSTOSIS

Presenter: Peter Hashim, MD
Authors: Patel A, Yang J, Hashim P, Bridgett D, Losee J, Duncan C, Jane J, Persing JA
Yale University School of Medicine

Background: The relationship between type of surgical intervention and long-term neuropsychological (NP) outcomes in sagittal craniosynostosis (SGC) remains a source of debate.⁽¹⁻²⁾ The optimal surgical approach in treating younger patients (i.e., under 6 months of age) proves controversial with a paucity of data related to long-term neuropsychological (NP) outcomes. This study compares the effect of performing whole-vault cranioplasty (WVC) versus strip craniectomy (SC) on long-term NP function using comprehensive, longitudinal neurological testing.

Methods: This multi-institutional study consisted of 72 surgically treated SGC patients. The subset of patients who underwent surgery prior to 6 months of age (n=45) was subdivided by type of surgery: WVC (n=20) vs. SC (n=25). All patients underwent a battery of neurodevelopmental tests evaluating various domains of NP function (Beery VMI, WASI, and Wechsler Fundamentals).

Results: Patients who received WVC obtained better outcomes relative to SC patients on Verbal IQ scores ($p < 0.05$) and Beery VMI ($p < 0.05$). A trend was also observed such that WVC patients performed better than SC patients on the Wechsler Fundamentals Word Reading task ($p < .10$). Differences in other domains of NP function (Full Scale IQ, Performance IQ, Spelling, Reading Comprehension, and Numerical Operations) were not significant. All analyses statistically controlled for full scale IQ, with the exception of analyses comparing subgroups on IQ variables.

Conclusion: These results demonstrate that the type of surgical intervention in treating SGC impacts long-term NP outcomes. Patients undergoing WVC attained higher intellectual and achievement scores in relative to those undergoing SC. This study highlights that performing WVC compared to SC prior to 6 months of age may be advantageous in terms of improved long-term NP outcomes.

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EXAMINING THE UTILITY OF DYNAMIC CRANIOPLASTY FOR THE TREATMENT OF SAGITTAL SYNOSTOSIS: A RETROSPECTIVE COHORT COMPARISON STUDY BETWEEN REVERSE PI CRANIOPLASTY AND EXTENDED STRIP CRANIOPLASTY

Presenter: Christopher Bonfield, MD
Authors: Bonfield C, Rottgers SA, MacIsaac Z, Pollack IF, Tamber MS, Kumar AR
University of Pittsburgh Medical Center

WITHDRAWN



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DIAGNOSIS SPECIFIC HEAD GROWTH CURVE IN CHILDREN UNDERGOING TOTAL CALVARIAL VAULT REMODELING (TCVR) FOR SAGITTAL SUTURE SYNOSTOSIS

Presenter: Peter D. Ray, MD
Authors: Ray PD, Slama RE, Huettner F, Dynda DI, Grant JH

Childrens of Alabama Hospital

Background: The head circumference of children with surgically corrected craniosynostosis is currently monitored using standard growth charts based on age and sex. These charts are derived from a selection of children living under conditions most favorable to obtain full genetic growth potential, according to the World Health Organization. Monitoring these children over time, the authors noted that these individuals seemed to follow a different overall head growth curve relative to unaffected individuals.

Objective: The objective of this presentation is to explain the current model monitoring the growth curve and why it may be sub-optimal for children undergoing surgical correction of sagittal craniosynostosis.

Design/Methods: A single surgeon, IRB approved, retrospective chart review identified 429 open craniofacial cases at Children's of Alabama between 1997 and 2012. Of the 429 charts reviewed, 275 underwent correction of synostosis. Of these, 115 sagittal synostosis patients were identified for the study. Age in months (X-axis) vs. head circumference in centimeters (Y-axis) were plotted separately for pre-operative and post-operative visits. A logarithmic trend line was added and compared with The Center for Disease Control Growth Charts.

Results: Both male and female sagittal synostosis patients who underwent TCVR had preoperative head circumference measurements approaching the 90th percentile. Their trend line over time remained relatively unchanged post-operatively as it tended to track between the 75th and 90th percentiles.

Conclusions: Children with craniosynostosis have head circumference growth that is unique to their condition and reflects a distinct subset of the general population. The development of diagnosis specific head circumference growth curves for each type of craniosynostosis may therefore help maximize the care of these patients and reduce the risk of "falling off the curve" when being monitored by physicians unfamiliar with the condition.

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TOTAL CRANIAL VAULT REMODELING FOR ISOLATED SAGITTAL SYNOSTOSIS: PART I. POSTOPERATIVE CRANIAL SUTURE PATENCY

Presenter: David K. Chong, MD
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Background: Total cranial vault reconstruction addresses all phenotypic aspects of scaphocephaly. The clinical implications of remodeling across open cranial sutures, however, remain unclear. The purpose of this study is to assess patency of unaffected sutures following total vault remodeling for isolated sagittal synostosis.

Methods: The authors reviewed all patients who underwent total vault remodeling for isolated sagittal synostosis between 2004-2008, a period when craniofacial computed tomographic (CT) scans were routinely performed postoperatively. Degree of patency of coronal and lambdoidal sutures was scored by a single reviewer as: 0 = closed; 1 = partial; and 2 = open. Individual suture scores were tallied for a total sutural patency score. CT scans were also categorized by postoperative time and by craniofacial surgeon.

Results: 42 patients met the inclusion criteria. Individual sutural closure rates were 42.6%, 38.3%, 74.5%, and 74.5% for right coronal, left coronal, right lambdoidal, and left lambdoidal sutures, respectively. Lambdoidal sutures had a significantly higher rate of closure than coronals (ORClosure 4.3, 95% C.I. 2.3 – 8.0, $p < 0.001$); lambdoidal patency significantly changed over time (Chi square = 9.9, $p = 0.04$). Across craniofacial surgeons, coronal and lambdoidal patency were equivalent. The total sutural patency score did not significantly correlate with postoperative time, surgical age, preoperative cephalic index, or craniofacial surgeon.

Conclusions: Total vault remodeling for isolated sagittal synostosis results in a high degree of secondary craniosynostosis. Lambdoidal sutures are especially prone to closure, with their patency diminishing over time. The long-term implications of these findings are being evaluated by growth, morphometric, and neurocognitive studies.



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EVALUATING THE EFFICACY OF DYNAMIC CRANIAL VAULT REMODELING FOR THE TREATMENT OF LATE PRESENTING SCAPHOCEPHALY: A COHORT COMPARISON STUDY BETWEEN PI CRANIOPLASTY AND SUBTOTAL CRANIOPLASTY

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Purpose: Optimal treatment for late presenting sagittal synostosis remains controversial. The efficacy of dynamically shortening the AP dimension has been demonstrated in younger patients, but not in this older cohort. This study aims to compare the ability of either a dynamic Pi/Hungspenting sagittal synostosis patients (>12 months old) was performed comparing cranial index (CI) and naso-frontal angle (NFA) changes between patients treated with PHC and STC with CT data. Non-syndromic sagittal synostosis patients < 12 months old treated with reverse Pi craniopalsty (CONTROL) were used as a control population.

Results: Six PHC patients from August 2011-May 2012 (5 male and 1 female, age 59 months), 5 STC patients from April 2006- May 2012 (4 male and 1 female, age 31 months), and 5 CONTROL patients from August 2010- May 2012 (5 male and 0 female, age 7 months) were identified. The median PHC CI increased from 0.71 to 0.82 ($p=0.028$), STC CI increased from 0.67 to 0.71 ($p=0.225$), and CONTROL CI increased from 0.74 to 0.80 ($p=0.008$). The median delta-CI was significantly greater for PHC compared to STC ($p=0.004$) and CONTROL ($p=0.004$). The NFA significantly increased with both PHC and CONTROL (PHC; pre-op: 140.5, post-op: 143; $p=0.046$) (CONTROL; pre-op: 124; post-op: 139; $p=0.042$) but not with STC (pre-op: 140; post-op 140; $p=0.713$). NFA increased significantly more in CONTROL patients compared to STC ($p=0.016$) but not PHC ($p=0.082$).

Conclusions: PHC more effectively corrected cranial disproportion compared to STC for late presenting scaphocephaly. PHC also better corrected CI when compared to a younger CONTROL population. Later age of treatment and rigid fixation of the outfractures may explain this finding. Dynamic cranioplasty in both younger and older cohorts significantly improved NFA. The improvement in CONTROL patients was significantly greater than older STC patient.

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HOW DOES CEPHALIC INDEX AT THE SKULL VAULT RELATE TO CEPHALIC INDEX AT THE SKULL BASE IN CHILDREN WITH UNOPERATED, ISOLATED SAGITTAL SYNOSTOSIS?

Presenter: David Johnson, MD

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Introduction: The skull vault is scaphocephalic in sagittal synostosis but the severity of the associated skull base deformity is unknown, and cannot be measured clinically. There is much debate over the timing of intervention, and the optimal procedure for the correction of isolated sagittal synostosis, and information on the extent and progression of the skull base deformity might influence decision making. We used 3-dimensional CT (3DCT) scans to correlate deformity at the vault, base and posterior fossa in isolated sagittal synostosis.

Methods: Cephalic index (CI) was measured from 3DCT craniofacial scans in 34 consecutive cases of unoperated isolated sagittal synostosis, and 16 controls without craniosynostosis. CI was measured from the 3DCT volume data set using pre-determined landmarks in planes generated by a Vitrea™ workstation at three levels corresponding to the skull vault, base and posterior fossa. Measurements at the skull base and posterior fossa were below the level affected by calvarial remodelling. Data were analysed by student's t-test and Pearson coefficient.

Results: Mean CI in isolated sagittal synostosis was 0.67, 0.72 and 0.70 at the skull vault, base and posterior fossa, respectively. CI was significantly reduced compared to controls who had a mean CI of 0.82 at all three levels ($p<0.0001$). Ratios of CI between the vault and base, and the vault and posterior fossa were significantly reduced in sagittal synostosis ($p<0.0001$ and $p=0.0031$), implying a milder deformity at the base and posterior fossa compared to the skull vault. However there was strong positive correlation between CI at the vault and base in unoperated sagittal synostosis ($r=0.77$, $p<0.0001$).

Conclusions: This is the first study to show that the extent of deformity at the base is less severe but still correlates with the vault in unoperated sagittal synostosis. The clinical significance of the skull base deformity, its progression over time, and response to surgical correction at the skull vault, remain uncertain, however these results provide a framework for future studies.



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THE LIVERPOOL PROTOCOL FOR SCAPHOCEPHALY CORRECTION

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Introduction: Scaphocephaly is a common craniosynostosis with variable presentation including a long thin head, frontal bossing, bitemporal pinching, posterior bullet formation, increased calvarial height and anterior displacement of the vertex. Many techniques including passive cranioplasty, traditional active remodelling and spring assisted cranioplasty have been described but no procedure alone reliably addresses all aesthetic aspects of the condition while optimising risk versus benefit for each patient. In the Merseyside Supra Regional Craniofacial Unit, the presented protocol for management of scaphocephaly offers a comprehensive range of operations which optimises aesthetic outcomes while balancing risk versus return.

Methods: The protocol offers extended pi craniectomy and micro-barrel staving to children younger than 6 months and active vault remodelling in the older than 6 month group, usually carried out at a year of age. A modified Melbourne technique which avoids removal of the posterior bullet and therefore avoids excess bleeding risk, is used in the majority of these patients with total vault remodelling used in a selected cohort only.

Results: 109 patients underwent scaphocephaly correction between April 2009 and December 2012. 54 underwent extended Pi craniectomy, 41 underwent subtotal vault remodelling and 14 underwent total vault remodelling. Aesthetic outcomes in the pi craniectomy group have been previously reported with mean cephalic index improving from 66.8 pre op to 73.7 post op. Mean CI pre-operatively in patients having subtotal vault remodelling was 66.2 (SD: 3.1), increasing to 75.2 (SD: 4.6) at six months post-operatively. In those patients undergoing total vault remodelling the cephalic index improved from 66 (SD: 3.6) to 75 (SD: 2.3). Transfusion requirements have been kept low with median donor exposure = 1 and 25% of patients avoiding allogenic transfusion.

Conclusions: This protocol effectively addresses all the stigmata of scaphocephaly while achieving excellent aesthetic outcomes and minimising morbidity. Transfusion requirements are low, and there has been no mortality.

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SPHENOID WING FLARE: FINDINGS AND IMPLICATIONS FOR TREATMENT OF SAGITTAL SUTURE SYNOSTOSIS

Presenter: Robert J. Havlik, MD

Authors: Havlik RJ, Friel M, Flores R, Smith J, Ackerman L

Indiana University

Introduction: Non-syndromic sagittal suture synostosis (SSS) is the most frequent form of craniosynostosis, with findings of scaphocephaly, frontal bossing, and occipital bone deformity. Many techniques have been described for treatment of this skull deformity. Early presentation of SSS is frequently treated with an extended strip craniectomy. Late presentation of SSS is routinely treated with more extensive cranial vault remodeling. We have routinely found that SSS includes posterior extension of the orbital roof and the sphenoid, or a condition which we term “sphenoid flare”, which has implications for treatment of this disorder.

Methods/Results: Fifty-one patients between 2007 and 2012 presented at over six months of age with scaphocephaly and posterior sphenoid flare and were treated with a combination of anterior cranial vault reconstruction and an interdigitating CRAB technique for subtotal cranial vault reconstruction. All had abnormal fusion of the sphenoid to the temporal bone with posterior extension of the sphenoid flare into the sulcus between the frontal and temporal lobes of up to 2 cm. All underwent resection of the posterior flare of the sphenoid, as well as the posterior orbital wall adjacent to the temporal lobe. More severe cases also clearly had intradural bone within the sulcus. Results to date have been favorable, without the late development of bi-temporal “pinch”.

Discussion: We have identified a posterior flare of the sphenoid wing in all 51 patients with late presentation of SSS. These findings are consistent with Moss’ postulated mechanism of pathogenesis of SSS. In these cases, we believe that resection of the flare and the expansion of the temporal space is important in correction of bitemporal narrowing, and prevention of the “pinched” look in the temporal region that can be seen in those with SSS that have undergone reconstruction following growth. Moss states “It is as true for any scientific or professional discipline as it is for political societies that those who forget their past are condemned to repeat it”.



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INITIATION OF MULTI-MODEL THERAPY TO MINIMIZE ALLOGENEIC TRANSFUSION REQUIREMENTS DURING SURGICAL TREATMENT OF CRANIOSYNOSTOSIS AT A TEACHING HOSPITAL: LESSONS LEARNED

Presenter: Jennifer L. Rhodes, MD
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Surgery for craniosynostosis may be associated with significant blood loss and significant rates of allogeneic blood transfusions. The risks of blood transfusions are well known, and include transmission of viruses, bacterial contamination, acute hemolytic reactions, transfusion-related lung injury, and inflammation. To minimize blood transfusions, we instituted a protocol including preoperative erythropoietin therapy, intraoperative blood recycling; and acceptance of a lower hemoglobin level.

This study examined patients who underwent craniosynostosis repair and completed the protocol after initial implementation. A control group consisted of similar patients prior to the protocol's initiation. The protocol included erythropoietin preoperatively, intraoperative autologous blood recycling, and postoperative transfusion threshold of 7 gm/dl hemoglobin. Transfusion rates were examined with a subgroup analysis performed based on age at surgery and date of surgery.

32 patients completed the protocol and 25 patients were included in the control group. Discrete variables were compared using Fisher's Exact test. Continuous variables were compared using Student's t test. There was a significant decrease in the rate of transfusion between groups (96% vs 56%, $p=0.0006$) and in hospital length of stay (3.9 vs 2.6 days, $p=0.0013$). There was no difference in age (mean age 7 vs 8 mo, $p=0.1$). Children ≥ 6 mo at surgery were transfused 70% and >6 mo 50%; however, this did not achieve significance likely due sample size. With age, weight, and estimate blood loss similar, the transfusion rate was of 70% in first half of study and 44% in last half, although this did not achieve significance likely due to small sample size ($p=0.16$). Protocols that include preoperative erythropoietin, autologous blood recycling, and acceptance of lower hemoglobin levels may be effective at decreasing transfusions in craniosynostosis surgery. The concomitant decrease in transfusion-related complications may lead to better outcomes. The introduction of a novel protocol may be associated with an institutional "learning curve".

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INTERNATIONAL SURVEY OF PROTOCOLS FOR SAGITTAL SYNOSTOSIS IN INFANTS

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Craniofacial Unit

This study reviews current techniques and protocols used in young infants (aged 6 months or less), as well as the outcomes in terms of re-operation rates. A short questionnaire was designed including questions about the preferred surgical techniques, transfusion protocols and re-operation rates. Surgeons from the International Society of Craniofacial Surgery (ISCFS) and the International Society for Pediatric Neurosurgery (ISPN) were requested to respond to this questionnaire online or by email. Responses over a two weeks period were collated and analyzed using a Fisher's exact test. Ninety-one surgeons responded from craniofacial centers around the world, of which 93.4% completed the questionnaire. Most respondents were from North America and Europe (35 and 20%, respectively). The operative volume was less than 15 cases per year in 57%. The Bicoronal skin incision was most commonly used (82%). Post-operative drainage was not performed by 59.5%, but was statistically more common with use of the Bicoronal incision ($p = 0.029$). Two third of the respondents used calvarial remodeling, and 34% strip craniectomy. Blood was most commonly transfused at a hemoglobin level under 8b/dl (31%) with a mean transfusion rate of 65%. Almost half of the respondents transfused in more than 90% of cases, while only 18% transfused in 20% or less of cases. The mean reoperation rate for secondary fusion was 1.7%, and 41% respondents claimed a 0% re-operation rate. No statistical correlation was found with the type of surgical technique but a statistically higher frequency of reoperation was reported by centers with a caseload over 15 cases per year ($p = 0.035$), suggesting a lack of strict monitoring in most centers.

In conclusion, our survey shows that for young infants treated for Scaphocephaly the Bicoronal incision remains the most commonly used. A greater number of surgeons do not employ drains. A secondary surgery may be needed in 1.7% of the cases. This study presents a snapshot of the current surgical treatment and should serve as a basis for quality improvement and outcomes monitoring in their surgical management.



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SPLIT CRANIAL GRAFTING IN CHILDREN LESS THAN 3 YEARS OF AGE: >400 CONSECUTIVE CASES

Presenter: Christian J. Vercler, MD

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University of Michigan

Introduction: Traditional teaching has maintained that split cranial bone grafting cannot reliably be performed before the age of three. This notion has become absorbed into the mainstream of plastic surgery teaching as evidenced by a question in the most recent in-service examination that ruled out the possibility of split cranial grafting in a patient less than 3 years of age. This commonly held belief is incorrect. To set the record straight and facilitate an evidence-based practice in craniofacial surgery, we here present 432 consecutive cases in which cranial bone was split in patients under the age of 3 years.

Methods: We reviewed a single surgeon's experience with performing split cranial bone grafting in cases of craniosynostosis from 1995 to 2013. After the neurosurgeon performs the craniotomy, the sections of calvaria removed are split along a plane between the inner and outer cortex using a series of small (2mm – 4mm) sharp osteotomes. The split bone is then used as bone graft in the reconstruction of the cranial vault.

Results: Between 1995 and 2013, 432 cranial vault remodeling procedures utilizing split cranial bone graft were performed on patients with both syndromic and non-syndromic craniosynostosis. In each case the bone splitting technique was performed by a trainee under the direct supervision of the senior surgeon. Only the presence of Lückenschadel, which obliterates the plane between the inner and outer cortex, prevented these areas from separating.

Conclusion: Contrary to popular belief, the calvaria of children under the age of three can reliably be split into an inner and outer cortex. This expands the amount of rigid bone available in cranial vault remodeling and is a feasible technique easily taught to residents and fellows.

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ISOLATED FRONTOSPHEOIDAL SYNOSTOSIS: A RARE CAUSE OF SYNOSTOTIC FRONTAL PLAGIOCEPHALY

Presenter: Tina M. Sauerhammer, MD

Authors: Sauerhammer TM, Oh AK, Boyajian M, Magge S, Myseros JS, Keating RF, Rogers GF

Childrens National Medical Center

Background: Unilateral fusion of the frontoparietal suture is the most common cause of synostotic frontal plagiocephaly. Localized fusion of the frontosphenoïdal suture is rare but can lead to a similar, but subtly distinct, phenotype.

Methods: A retrospective chart review of our craniofacial database was performed. Patients with isolated frontosphenoïdal synostosis on computed tomography (CT) imaging were included. Demographic data, as well as the clinical and radiographic findings, were recorded.

Results: Three patients were identified. All patients were female and none had an identifiable syndrome. Head circumference was normal in each patient. Mean age at presentation was 4.8 months (range, 2.0-9.8); two fusions were on the right side. Frontal flattening and recession of the supraorbital rim on the fused side were consistent physical findings. No patient had appreciable facial angulation or orbital dystopia, and two patients had anterior displacement of the ipsilateral ear. All three patients were initially misdiagnosed with unilateral coronal synostosis and CT imaging at mean age of 5.4 months (range, 2.1-10.8) was required to secure the correct diagnosis. CT findings included: patency of the frontoparietal suture; minor to no anterior cranial base angulation; vertical flattening of the orbit without sphenoid wing elevation on the fused side. One patient had a CT at 2.1 months of age that demonstrated a narrow, but patent, frontosphenoïdal suture. Assumed to be a deformational process, the patient underwent six months of unsuccessful helmet therapy. Repeat CT at 10.7 months of age demonstrated the synostosis. All three patients underwent fronto-orbital correction at mean age of 12.1 months (range, 7.8-16.1).

Conclusions: Isolated frontosphenoïdal synostosis should be considered in the differential diagnosis of atypical frontal plagiocephaly.



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UNILATERAL VERSUS BILATERAL CORRECTION OF UNICORONAL SYNOSTOSIS; AN ANALYSIS OF LONG-TERM RESULTS

Presenter: Martijn Cornelissen, MD
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Introduction: Hollowing of the temporal region is a common problem after cranioplasty for unicoronal synostosis. In this study the development of temporal hollowing pre- and postoperatively was evaluated, in relation to the two operative techniques used and their timings.

Methods: From 1979 to 2010 194 patients with unicoronal synostosis were operated at our center. Patients were treated with either a unilateral or a bilateral correction of the supraorbital rim. Forty-eight patients qualified for the present study with a mean age at follow-up of 7.5 years. Cephalic landmarks were identified on radiographs prior to and after surgery, to determine the growth of the forehead. For visual analysis, two independent observers evaluated normal photographs for the presence and severity of temporal hollowing.

Results: Preoperative osseous asymmetry improved significantly after surgery. Twenty-one patients show an increase of temporal hollowing on photographs after surgery (46%). In 35 out of 48 patients postoperative temporal hollowing was noted (73%). Bilaterally treated patients showed more severe temporal hollowing compared to unilaterally treated patients, however not significantly (23% vs 6%, $p=0.229$). Timing of surgery (before or after the age of one year) did not influence the occurrence of severe temporal hollowing.

Conclusions: Fronto-supraorbital advancement was unable to achieve normal growth in the temporal region in a large proportion of patients, although more symmetry was achieved. The operative technique itself did not seem to influence the occurrence of temporal hollowing, nor did the timing of surgery.

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TREATMENT OF UNILATERAL CORONAL SYNOSTOSIS WITH ENDOSCOPIC STRIP CRANIECTOMY AND CRANIAL ORTHOSIS YIELDS SIGNIFICANTLY BETTER OPHTHALMIC OUTCOMES THAN TREATMENT WITH FRONTO-ORBITAL ADVANCEMENT

Presenter: Linda Dagi, MD
Authors: Dagi L, Rogers GR, Mackinnon S, Meara JG, Proctor MR

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Purpose: Unilateral coronal synostosis (UCS) is highly associated with V-pattern strabismus, astigmatism and amblyopia. We compared long-term ophthalmic outcomes in infants with UCS treated with endoscopic strip craniectomy and post-operative helmet therapy (ESC+HT) to those managed by fronto-orbital advancement (FOA).

Methods: Retrospective chart review identified all patients with isolated UCS treated at our institution. We reviewed ophthalmic records seeking evidence of amblyopia, astigmatism and anisometropia, strabismus and strabismus surgical intervention for all included subjects.

Results: Between 2004 and 2010, 22 patients were treated by FOA (mean follow-up of 21.5 months), and 21 patients with ESC+HT (mean follow-up of 23.5 months). The mean aniso-astigmatism was equal; however, the standard deviation was greater for those treated by FOA ($P < 0.05$). Patients treated with FOA had a more severe pattern of strabismus ($P < 0.0001$), higher rates of amblyopia ($P = 0.0015$), and were more likely to require surgical correction of their strabismus (odds ratio 6.3:1).

Conclusions: Children with UCS treated with ESC+HT had less severe V-pattern strabismus, amblyopia, extremes of astigmatism, and less need for strabismus surgery than in those treated by FOA. Though the reason for these more favorable outcomes remains uncertain, we speculate that the earlier timing of endoscopic strip craniectomy, or differences in the anatomical changes resulting from the two procedures may play a role. These findings may support early intervention to reduce ophthalmic morbidity.



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A SINGLE CENTERS EXPERIENCE WITH ISOLATED UNICORONAL CRANIOSYNOSTOSIS RECONSTRUCTION: LONG-TERM OUTCOMES OF 182 PATIENTS OVER 35 YEARS

Presenter: Ari M. Wes, BA

Authors: Wes AM, Goldstein JA, Whitaker LA, Bartlett SP, Taylor JA

University of Pennsylvania School of Medicine and Childrens Hospital of Philadelphia

Purpose: Assess long-term outcomes of patients with isolated unicoronal synostosis treated at our institution over a thirty-five year period.

Methods: A retrospective review was performed of patients with unicoronal synostosis from 1977 to 2012. Patients were excluded with prior outside intervention, or diagnosis of syndromic or multisutural synostosis. Demographic, operative/post-operative data and outcomes were analyzed with chi-squared and Fisher's exact test for categorical data and Wilcoxon rank-sum and Kruskal-Wallis rank for continuous data.

Results: Over 35 years, 182 patients were treated for unicoronal synostosis and 156 met inclusion criteria. The patient population was predominantly female (65%, n=101) with synostosis predominantly on the right (60%, n=93). Patients presented with supraorbital retrusion (95%, n=148), orbital dysmorphology (71%, n=110), compensatory bossing (51%, n=79), nasal root deviation (38%, n=59), occipital irregularity (12%, n=18), and midface asymmetry (8%, n=12). Primary intervention included 55 (35%) unilateral fronto-orbital advancements (FOA) with unilateral frontal craniotomy, 54 (35%) incomplete bilateral FOA with bilateral frontal craniotomy, 42 (27%) unilateral FOA with bilateral frontal craniotomy, and 5 (3%) bilateral FOA with bilateral frontal craniotomy at a mean age of 0.98 ± 1.0 years. There were 3 acute surgical complications (3%). Mean follow up was 5.9 ± 5.0 years, and in patients with ≥ 1 year follow-up (n=129), 57 (44%) required second intervention. At definitive intervention, 70 (54%) patients were Whitaker class I, 6 (5%) class II, 50 (39%) class III, and 3 (3%) class IV. Recurrence of the preoperative abnormality was noted in 71 (55%) patients at latest follow-up. Patients with ≥ 5 years follow-up were more often class III ($p < .001$) and less often class I ($p < .001$) and developed supraorbital retrusion following primary intervention more often than their counterparts with < 5 year follow-up ($p < .001$).

Conclusions: In the largest evaluation of isolated unicoronal synostosis, we critically evaluate outcomes to help shape expectations that may be useful when counseling patients and families.

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MINOR CRANIAL SUTURE CLOSURE: A MORPHOLOGICAL STUDY

Presenter: Wayne Ledin, MD

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Context: Craniosynostosis is a not uncommon premature fusion of one or more cranial sutures affecting 1:2000 live births. The more common synostoses involve the sagittal and coronal sutures, which are both major cranial sutures. There have been few reports of craniosynostosis involving the squamosal, sphenofrontal, and other minor cranial sutures. Controversy exists in the anthropological literature regarding the use of cranial suture closure for age determination. It is not entirely clear when, or if, any of the minor cranial sutures normally close.

Objectives: To determine whether and at what age the minor cranial sutures close.

Materials & Methods: 114 skulls in the Hamann-Todd collection at the Cleveland Museum of Natural History (ages 1-50) were examined. Closure of the minor sutures (intraoccipital, parietomastoid, occipitomastoid, sphenoparietal, squamosal, sphenofrontal, and sphenosquamosal) was graded. Cranial dimensions were measured and cephalic indices were calculated.

Results: Data analysis indicates that the intraoccipital suture is the only minor suture to close in the pediatric age group. Median closure is at age 4, with 95% confidence interval between ages 1 and 5. The median closure for the sphenofrontal suture is age 26 (95% confidence interval between ages 26 and 44). Closure of the occipitomastoid suture is at the median age of 20 (95% confidence interval between ages 17 and 32). The remaining minor cranial sutures show no evidence of closure in our dry skull model.

Conclusions: Closure of the minor cranial sutures, excepting the intraoccipital suture, in the pediatric population may be abnormal. Consideration of minor suture craniosynostosis should be investigated in the following two patient populations: (1) those diagnosed with positional plagiocephaly for whom conservative management has been ineffective and (2) patients with abnormal skull shapes that do not fit any of the classically described morphologies for the varying craniosynostoses.



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ISOLATED METOPIC CRANIOSYNOSTOSIS IN THE FOURTH DIMENSION: AN EVALUATION OF COMPLICATIONS, REVISIONS, AND LONG-TERM OUTCOMES IN 142 PATIENTS OVER 30 YEARS

Presenter: J. Thomas Paliga, MD

Authors: Paliga JT, Wes AM, Goldstein JA, Whitaker LA, Bartlett SP, Taylor JA

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Purpose: Assess long-term outcomes of patients with isolated metopic synostosis through the evolution of treatment at a single institution.

Methods: IRB approved retrospective chart review performed on consecutive patients with metopic synostosis from 1978 to 2012 at the Children's Hospital of Philadelphia. Inclusion criteria required treatment of metopic synostosis, complete medical record and ≥ 1 year follow-up. Exclusion criteria involved prior outside intervention, multisutural synostosis, or syndromic diagnosis. Demographic, operative and post-operative data were collected. Outcomes were reported as Whitaker class and clinical features at latest follow-up. Chi-squared and Fisher's exact test were run on categorical data. Wilcoxon rank-sum and Kruskal-Wallis rank tests were run on continuous data.

Results: Over 34 years, 142 patients were treated for metopic synostosis and 113 met inclusion criteria. Mean age at surgery was 0.83 years (range: 0.3-4.7), and mean follow-up was 6.0 years (range: 1.0-17.8). There were 10 (8.8%) surgical complications; 3 (2.6%) major and 7 (6.1%) minor. 13 (21.0%) patients required secondary intervention including 2 (3.2%) complete revisions, 6 (9.7%) hardware removals, 5 (8.1%) bony contouring and 1 (1.6%) soft tissue augmentation. At follow-up, 63 (55.8%) patients were Whitaker class I, 6 (5.3%) class II, 41 (36.3%) class III and 3 (2.7%) class IV. Patients with temporal hollowing ($p=0.001$) or lateral orbital retrusion ($p<0.001$) had higher Whitaker class and more interventions over time ($p=0.0012$). Patients with ≥ 5 years follow-up ($n=54$) were more likely to have temporal hollowing (OR 3.6, 95%CI 1.7-7.9, $p=0.001$), lateral orbital retrusion (OR 7.4, 95%CI 3.2-17.2, $p<0.0001$) and a Whitaker class III or IV (OR 6.2, 95%CI 2.6-14.5, $p<0.0001$) compared to patients with <5 year follow-up ($n=59$).

Conclusions: In the largest outcome evaluation of isolated metopic synostosis to date, we characterize the complication and reoperation rates and demonstrate a clear trend toward worsening outcomes over time. This may impact patient counseling and underscores the need of follow-up to physical maturity.

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CLASSIFICATION OF SEVERITY AND OPERATIVE DECISION-MAKING IN METOPIC SYNOSTOSIS

Presenter: Alexander C. Allori, MD, MPH

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Background: Most clinicians intuitively agree when classifying mild and severe cases of metopic synostosis, but there exists a difficult "gray zone" of moderate-severity cases for which appropriate management is not so obvious. In this study, a previously validated method for quantifying the interfrontal angle is assessed as an aid to operative decision-making.

Methods: Craniofacial CTs of mild to severe metopic synostosis were obtained, and the interfrontal angle (IFA) was computed as previously described. An expert panel of pediatric craniofacial surgeons and neurosurgeons was convened. Panelists were asked to grade severity on a scale of 1-4 (mild to severe) and to decide on operative or non-operative management. Concordance for each severity group was assessed by ANOVA and pairwise t-testing. Operative vs. non-operative groups were compared using Student's t-test. Logistic regression and receiver-operator characteristic analysis were used to assess the utility of the IFA in predicting management.

Results: Thirty-five cases were selected with IFA ranging from 92.3° (severe) to 140.6° (mild). Expert severity scores were higher for more acute IFAs: A severity score of 1 correlated with an IFA of 132.1° \pm 9.2°; 2 with 122.1° \pm 9.9°; 3 with 116.0° \pm 9.6°; and 4 with 109.7° \pm 9.7°. High concordance was observed for IFA 136.1°-140.6° with a median severity score of 1 and also for IFA 92.3°-114.3° with a median severity score of 4 ($p<0.0001$). A gray zone with severe discordance in rankings existed between 114.3°-136.1°. An operative threshold of 118.2°, with the IFA able to predict the expert panels decision to proceed with surgery 87.6% of the time.

Conclusion: The IFA was previously validated as an accurate, reproducible, and practical means for diagnosing trigonocephaly. In this work, we demonstrate that IFA is also capable of predicting expert rankings of mild, moderate, and severe trigonocephaly; moreover, it was able to reliably predict the expert panel's decision for operative or non-operative management. This simple measure may serve to a useful adjunct to help guide clinical decision making.



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CLINICAL CHARACTERISTICS AND SURGICAL DECISION MAKING FOR INFANTS WITH METOPIC CRANIOSYNOSTOSIS IN CONJUNCTION WITH OTHER CONGENITAL ANOMALIES

Presenter: Craig Birgfeld, MD

Authors: Birgfeld C, Heike CL, Saltzman BS, Hing AV
University of Washington Seattle Childrens Hospital

Background: Metopic craniosynostosis can occur in isolation or in conjunction with other congenital anomalies. The surgical decision making and outcomes between these two groups are analyzed.

Methods: A retrospective review of all children evaluated in the craniofacial clinic at Seattle Children's Hospital for metopic craniosynostosis between 2004-2009 was performed. Physical exam and CT scan characteristics were analyzed as were the treatment decisions and surgical outcomes.

Results: From 2004-2009, 282 patients were evaluated and 100 were determined to have metopic craniosynostosis. Of these, 19 patients were found to have additional congenital anomalies. Review of these patients' CT scans revealed 13 with classic trigonencephaly, 3 with microcephaly and 3 with narrow frontal bones, abnormal orbits, and small anterior fossa. 90% of patients with isolated metopic craniosynostosis underwent cranial vault expansion, whereas only 63% of the complex group did so. The complex metopic group had a longer hospital stay (5 days vs. 3.4 days), more intraoperative complications and required more repeat surgery.

Conclusion: Patients with metopic craniosynostosis and additional anomalies require special consideration when deciding upon surgical intervention and should be cared for by a multidisciplinary team to address their additional needs.

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DIAGNOSIS OF METOPIC CRANIOSYNOSTOSIS FROM THE SYSTEMATIC ANALYSIS OF CRANIAL MALFORMATION

Presenter: Gary F. Rogers, PhD

Authors: Mendoza C, Safdar N, Rogers GF, Oh AK, Sauerhammer T, Keating R, Linguraru MG
University of Sevilla Spain

Background and Purpose: The metopic suture closes normally in early infancy. Thus, the diagnosis of metopic craniosynostosis (MS) hinges on cranial shape and not the presence of a fused suture, and is largely subjective. The purpose of this study was to create a simple, reproducible radiographic method to quantify forehead shape and distinguish normal variation from abnormal inter-frontal narrowing.

Methods: 111 head CT scans from subjects aged 1-12 months (mean 6 mo.) were acquired from our institutional PACS: a normative data base of 93 CT (average age, 6 months) was compared to the scans of 18 patients with a clinical diagnosis of MS (average age, 4 mo.). All CTs were aligned to a cranial template to correct for scale and pose, using the skull base. From the aligned normal scans, a statistical shape model was constructed, and for each MS subject the deformation fields (distance to the closest normal variant) were obtained. By computing the average deformation field over all MS subjects, optimal and simplified (co-planar) angles were found from the three points of maximum average deformation. Both angles were compared to each other, and area-under-the-curve (AUC) of receiver-operator characteristic curves for diagnosis of MS using both angles were computed.

Results: The analysis yielded an angle centered at a landmark on the metopic suture that optimally describes recedence of the frontal bones and the protrusion of the sutural area. The two lateral landmarks on the left and right frontal bones were at the point of maximal divergence from normal. The AUC for the optimal and simplified angles were 0.998 and 0.996, respectively; there was no significant difference between optimal and simplified angles in identifying MS ($p=0.87$). The average simplified angles for normal and MS patients were 145 ± 7 and 121 ± 6 degrees, respectively.

Conclusions: A systematic method for quantifying the severity of frontal narrowing based on cranial shape analysis may help reduce over diagnosis of MS. The proposed method uses a simple planar angle measurement on CT that is reproducible and accurate.



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MANAGEMENT OF SAGITTAL SYNOSTOSIS: STANDARD OF CARE

Presenter: Gaby Doumit, MD, MSc, FRCSC, FACS

Authors: Doumit G, Moores N, Papay F
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In the craniofacial surgery literature there is a wide disparity of opinions regarding the appropriate treatment of non-syndromic sagittal synostosis. With the lack of level one evidence to support a particular treatment, our study aims to elucidate the current state of practice among craniofacial surgeons. A cross-sectional questionnaire based survey.

An internet based survey was sent to 102 craniofacial surgeons in fourteen countries on four continents. Surgeons were queried regarding preoperative, intraoperative and post operative protocols for patients presenting with non-syndromic isolated sagittal synostosis with no sign of increased intracranial pressure.

After two mailings the response rate was 58% (59/103). For 63 % of respondents, skull deformity was the primary indication for treatment of craniosynostosis. 71% of surgeons will obtain a pre operative computed tomography irrespective of presentation. For non syndromic cases of sagittal synostosis presenting prior to 4 months of age, 75.8% of surgeons elect immediate operative management. 35% of craniofacial surgeons chose an endoscopic surgical approach. Total or sub total cranial vault remodeling was the procedure of choice for open surgical intervention. Forty three percent of respondents report utilization of transfusion in 91-100% of open synostosis repairs. Surgeons that choose total calvarial remodeling as their operation of choice and to perform these open operation in children at or younger than 6 months of age were more likely to require transfusions. 78% of craniofacial surgeons are satisfied with their surgical technique. However, craniofacial surgeons that utilize endoscopic methods at less than 4 months of age self reported with a statistical significance a higher satisfaction rate when compared to the open methods at the same age. (95% vs. 71%, p value 0.04).

Our survey supports the hypothesis that there exists a wide disparity of opinions regarding both the timing and method of therapy for non-syndromic sagittal synostosis.

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NAUTILUS: DYNAMIC CRANIOTOMY; NEW SURGICAL TECHNIQUE AND PRELIMINARY RESULTS

Presenter: Vera N. Cardim, MD, PhD

Authors: Cardim VN, Silva AS, Dornelles RF,
Salomons RL, Silva AL, Blom JO,
Colange NZ

Hospital Beneficencia Portuguesa de Sao Paulo

Introduction: Craniosynostosis is basically a suture defect where the main challenge of treatment is to release an entrapped brain from a closed compartment that does not have the required compliance to accompany its growth. The objective is to restore the complacency of the stenotic suture region and to correct the compensatory cranial deformity. This paper proposes a helicoid osteotomy associated to spring mediated osteogenic distraction to reshape craniofacial defects caused by craniosynostosis.

Method: During the period of July-2010 to April 2012, 10 patients with craniosynostosis were submitted to surgery: turricephaly: 01; trigonocephaly: 01; oxicephaly: 05 and scaphocephaly: 03. The treatment consisted of dynamic spring mediated osteotomies described by Lauritzen to correct the primary craniosynostosis deformity and a helicoid craniotomy "Nautilus-shaped" at the secondary compensatory deformation of the skull, without dural detachment.

Results: Resolution of the cranial deformity as remission of clinical signs of intracranial hypertension were observed. None of the patients presented complications (CSF fistula, local infection, seroma or hematoma).

Conclusion: The association of a helicoid osteotomy with spring mediated bone distraction or contraction promoted active skull remodeling, facilitating the accommodation of the brain in the cranial continent.



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BILATERAL LAMBDROID AND SAGITTAL SYNOSTOSIS (BLSS) – “MERCEDEZ BENZ” SYNDROME

Presenter: Hamilton Matushita, PhD

Authors: Marushita H, Alonso N, Bueno MR, Cardeal DD, Andrade FG

Sao Paulo University

Introduction: Multi-suture craniosynostosis are uncommon, constituting 0.7% of all craniosynostosis. A peculiar type of craniosynostosis consists of a combination of bilateral lambdoid and sagittal synostosis (BLSS). The tri-radiate ridging of the synostotic sutures resembles the emblem of “Mercedes Benz”. To date, fewer than 30 patients with craniosynostosis named “Mercedes Benz” or Craniofacial Dysynostosis have been reported. Few of them have complete clinical and radiological documentation, as well as the best surgical technique is still controversial.

Objectives: The authors present a series of 9 children with multi-suture craniosynostosis with involvement of bilateral lambdoid and sagittal sutures, in order to help defining its clinical features and treatment.

Material and Methods: All patients were examined for general clinical and neurologic aspects, morphometric measurement, and computed tomographic and magnetic resonance imaging of the head.

Results: The age ranged from 5 to 72 months (mean = 21.6m), gender distribution was M/F = 5/4, and there was no specific ethnic preference. The major cranial deformities observed were brachycephaly (cephalic index ranged from 0.79-1.06; mean=0.91), increased frontal bossing 6/9 (66.6%), increased occipital concavity in 6/9 (66.6%). Neuropsychological delay was observed in 5/9 (55.5%). Other malformations observed: Chiari I malformation in 4/9 (44.4%), agenesis of the corpus callosum in 1/9 (11.1%), ear malformation in 1/9 (11.1%), and ventriculomegaly was observed in 2/9 (22.2%). Genetic evaluation identified 3/9 (33.3%) cases associated with syndrome. Surgical treatment aimed at the expansion and remodeling of the parietal, occipital, and suboccipital regions. No major surgical complications occurred.

Conclusion: Our results showed etiologic heterogeneity of this rare type of craniosynostosis and demonstrated a varied phenotype. Neurological impairment affected both syndromic and non-syndromic BLSS. Surgical treatment with remodeling of the posterior cranium showed effective morphological correction.

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TRUE LAMBDROID SYNOSTOSIS: THE LIVERPOOL EXPERIENCE

Presenter: Jitske W. Nolte, MD, DDS

Authors: Nolte JW, Sinha A, Burn S, Duncan C, Richardson D

Alder Hey Childrens Hospital

Introduction: True lambdoid synostosis is a rare condition, with sparse detailed reports in literature. The incidence is estimated to be around 1% of all synostosis. We share our experience of 12 true lambdoid synostosis patients treated at a supraregional craniofacial unit in UK.

Methods: Clinical records of all patients who were referred to the Supraregional Craniofacial Unit Liverpool for possible lambdoid craniosynostosis in the period of 2004-2012 were analysed retrospectively. The diagnosis of true lambdoid synostosis was confirmed by clinical observation, CT-scan and at surgery. Gender, affected side and age at presentation were noted, as well as medical conditions, pregnancy and delivery details. Clinical characteristics regarding ear position, ipsilateral occipito-mastoid bulge, compensatory contralateral parietal bossing, frontal bossing, facial twist and hemifacial asymmetry were noted. The surgical details were evaluated and pre- and postoperative cranial vault asymmetry was recorded. Patients were screened for any pre- and/or postoperative developmental delay.

Results/Conclusions: A total of 8 patients were operated, while 3 are listed for surgery and 1 with mild vault asymmetry was managed conservatively. 9 cases were isolated lambdoid synostosis while 3 had multiple suture synostosis. Marked clinical features in isolated lambdoid synostosis were ipsilateral occipital-mastoid bulge, contralateral parietal bossing and contralateral hemifacial deficiency. A preference for male gender was noted. There were no significant perioperative complications. Our surgical technique and the post-operative aesthetic outcome will be discussed along with a review of literature.



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COMBINED METOPIC AND UNILATERAL CORONAL SYNOSTOSES: A PHENOTYPIC CONUNDRUM

Presenter: Kam Patel, MD

Authors: Sauerhammer TM, Patel K, Oh AK, Proctor MR, Mulliken JB, Rogers GF

Childrens National Medical Center

Background: Most types of craniosynostosis cause predictable changes in cranial shape. However, the phenotype of combined metopic and unilateral coronal synostoses is anomalous. The purpose of this observational study is to better clarify the clinical and radiographic features of this rare entity.

Methods: A retrospective review of a craniofacial database was performed. Patients with combined metopic and unilateral coronal synostoses were included in this study. Data collected included demographic information, physical and radiographic findings, genetic evaluation, treatment, and operative outcomes.

Results: Of 687 patients treated between 1989 and 2010, only three patients had combined metopic and unilateral coronal synostoses. All patients were diagnosed by computed tomography on the first day of life. Phenotypic features included: 1) narrowed forehead with a prominent midline ridge; 2) severe bilateral brow retrusion with an acute indentation on the side of the patent coronal suture; 3) facial and nasal angulation similar to isolated unilateral coronal synostosis; and 4) anterior displacement of the ear on the fused side. In addition, the cranial vertex was deviated toward the side of the open coronal suture. Two patients had a head circumference below the 25th percentile; two of the three had a TWIST gene mutation consistent with Saethre-Chotzen syndrome. One patient was managed by fronto-orbital advancement and required a revision. The other two patients had early endoscopic release followed by post-operative helmet therapy; one improved, but still required open cranial remodeling. The other has near normal phenotype and no further surgery is planned.

Conclusions: Combined metopic and unilateral coronal synostoses present a rare and unusual phenotype. While early intervention improves the deformity, revisional procedures are usually required.

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PRIMARY CORRECTION OF NASAL ASYMMETRY IN PATIENTS WITH UNILATERAL CORONAL SYNOSTOSIS

Presenter: Arun K. Gosain, MD

Authors: Gosain AK, Chepla KJ, Alleyne BJ

Lurie Childrens Hospital of Northwestern University Feinberg School of Medicine

Background: Unilateral coronal synostosis (UCS) results in cranial vault asymmetry and ipsilateral deviation of the nasal radix. Traditional surgical correction fails to address nasal root deviation. Without primary correction this nasal deformity can persist and may require secondary surgical correction in up to 50% of patients.

Methods: We report on a consecutive series of patients with UCS operated on by a single surgeon in which nasal osteotomies for correction of nasal root deviation was performed in conjunction with fronto-orbital advancement before age one year. The technique evolved from complete removal of the nasal bones with ex vivo repositioning (EVR) to an in vivo repositioning with osteotomy of the nasal bones without removing them from the surrounding soft tissue envelope (IVR). In both groups the nasion is repositioned in the midline and secured to the supraorbital bandeau. Clinical photographs and 3D CT scans were used to calculate nasal angulation preoperatively and at one year follow-up. Clinical follow-up has ranged from one to 5 years.

Results: Six consecutive patients (3 in each group) are reported. No patient demonstrated progressive nasal asymmetry on clinical follow-up. There was no statistically significant difference in nasal angulation between the groups preoperatively, postoperatively, or degree change in nasal angulations by either soft tissue or CT analysis. All patients demonstrated a significant reduction in postoperative nasal angulation by both soft tissue analysis (7.78 v. 2.00 degrees, $p=0.002$) and CT analysis (9.95 v. 2.78 degrees, $p=0.0002$).

Conclusion: Nasal root asymmetry in patients with UCS can be corrected before one year of age in conjunction with fronto-orbital advancement. Limited dissection of the nasal bones with in vivo repositioning is as effective as EVR for correction of nasal angulation at one-year follow-up. Since IVR better preserves nasal architecture and is associated with minimal perinasal tissue disruption; this is now our preferred technique for primary correction of nasal asymmetry in UCS.



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**A MODIFIED SURGICAL CORRECTION OF
PLAGIOCEPHALY TO ACHIEVE SYMMETRY**

Presenter: Hazem Ahmed Mostafa, MD

Authors: Elbarbary AS, Mostafa HA

AinShams University

Plagiocephaly is a descriptive term that denotes an oblique or twisted “plagio” head “cephale” shape. This abnormality may result from craniosynostosis or secondary deformational forces without synostosis. Techniques of managing plagiocephaly have improved overtime and comprised unilateral or bilateral fronta-orbital bar advancement and frontal bone advancement/rotation using bone-molding forceps or distraction devices. However, reestablishment of normal aesthetic units & complete symmetry of the forehead & orbits are quite challenging. It is still considered unachievable in one stage and patients tend to undergo secondary & touch-up procedures. This study sought to present a modification in the surgical technique of these cases to achieve symmetry. In principle, a suitably curved frontoparietal bone, based temporally, was harvested from the non-synostotic side to reconstruct the new forehead. Its frontal component was placed to substitute the old forehead on the synostotic side and benefit from its curvature. Orbital reconstruction was carried out by inserting a bone graft within the supraorbital bar to increase its width, which was then advanced & caudally fixed. In non-synostotic older cases, orbital reconstruction was carried out to mimic the normal side. Twenty three cases with age ranging between 6 months & 18 years underwent this modification by the same craniofacial team. All patients underwent in-depth clinical evaluation including anthropometric craniofacial measurements and received CT scans prior to surgery. Preoperative ophthalmologic evaluation & genetic counseling were conducted routinely. Up to four years of follow-up, this technique achieved symmetry, normalization of the cranial shape & cephalic index in all cases with negligible complications reported. There were no complications related to using the resorbables for fixation. In conclusion, reestablishment of normal aesthetic units and achieving symmetry of the forehead & orbits in plagiocephalic patients is attainable through temporally basing a suitably curved frontoparietal bone from the non-synostotic side to reconstruct the new forehead.

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**SPRING-ASSISTED CRANIOPLASTY: OUR FIRST
EXPERIENCE**

Presenter: JanFalco Wilbrand, MD, DMD

Authors: Wilbrand J, Reinges M, Christophis P,
Howaldt HP

University Hospital Giessen

WITHDRAWN



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FAMILIAL NONSYNDROMIC CRANIOSYNOSTOSIS WITH SPECIFIC DEFORMITY OF THE CRANIUM

Presenter: Azusa Shimizu, MD

Authors: Shimizu A, Komuro Y, Miyajima M, Arai H
Juntendo University Shizuoka Hospital

Introduction: Craniosynostosis syndrome is a congenital anomaly of the cranium with an incidence of 4–5 per 10000 live births. We treated a young boy who presented with the specific cranial deformity. His family had similar deformities of the cranium, so we examined the genetic characteristics of this familial case.

Case Report: An otherwise healthy, developmentally normal 3-week-old male was referred by his physician for evaluation of cranial deformation and protrusion of the forehead which was noticed immediately after birth. Physical examination and computed tomography revealed complex multi-suture craniosynostosis involving the metopic suture and bilateral coronal sutures with frontal prominence and hypotelorism. Frontal craniectomy and bilateral fronto-orbital advancement remodeling were performed at the age of 5 months. Postoperative course was uneventful. His father and grandfather had similar specific deformities of the cranium, but no anomaly of the extremities was found, and conversation suggested intelligence was normal, probably excluding the possibility of syndromic craniosynostosis. DNA analysis of the patient and his family revealed a common copy number loss area on chromosome 4 p14 which includes NEDD4 binding protein 2 (N4BP2), such a copy number loss was absent in normal Japanese population. To examine further genetic backgrounds, we screened for mutations in FGFR1-3 genes by using direct DNA sequencing. As a result, no FGFR mutation was identified in this family. Our investigation of this family revealed other female relative with similar cranial deformity, confirming the presence of autosomal dominant inheritance.

Conclusion: Non-syndromic craniosynostosis involving only dysmorphology of the cranium usually occurs as solitary cases, whereas familial syndromic craniosynostosis is associated with other anomalies like facial hypoplasia or anomaly of the extremities. Therefore, the present familial case of non-syndromic craniosynostosis is rare. We plan further genomic analysis of this family and long-term observation of the craniofacial deformity of this patient.

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COMPARISON OF DIFFERENT SURGICAL STRATEGIES TO APPROACH UNILATERAL CORONAL SYNOSTOSIS

Presenter: Cesar A. Raposo-Amaral, MD

Authors: Raposo-Amaral CA, Chammas D, Raposo-Amaral CE

SOBRAPAR

Introduction: The premature fusion of unilateral coronal suture can cause a significant asymmetry of the craniofacial skeleton, with an oblique deviation of the cranial base that impacts soft tissue symmetry of the face. The aim of this study was to assess the craniofacial symmetry obtained with 2 different techniques to treat patients with unilateral coronal synostosis. Additionally, neurological status, length of stay in the hospital, blood units transfused during surgery and complication rate of each group were assessed.

Methods: This prospective study was conducted during a two years period and received approval by the Ethic Committee of the Institution. The patients with unilateral coronal synostosis were consecutively, randomly divided into two groups. Group 1: Patients who underwent total frontal reconstruction and transferring of onlay bone grafts to the recessive superior orbital rim, without any osteotomies in the orbital cone (n=7). Group 2: Patients who underwent total frontal reconstruction and unilateral fronto-orbital advancement (n=5). To assess craniofacial symmetry, a cephalic index (relation between anteroposterior and bi-temporal distances) and an oblique index (relation of major and minor oblique cranial distance) were obtained through anthropometric measurements using a spreading caliper and compared. Measurements were performed by two craniofacial surgeons based on previously determined anatomical points. The results have undergone statistical analysis using the Mann-Whitney test.

Results: Patients of groups 1 and 2 presented average cephalic index of 85% and 80%, respectively and an average oblique index of 1.03 and 1.01, respectively. Both indices did not statistically differ between groups. Neurological status was normal for all patients of both groups. Average blood units transfusion were 1.42 units and 1 unit for groups 1 and 2, respectively. Average length of hospital stay was 3.85 days and 5.33 days for groups 1 and 2, respectively. No complications were observed in this series of patients.

Conclusion: Similar craniofacial symmetry was obtained using both techniques.



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EVALUATION OF SURGICAL OUTCOMES IN CRANIO-ORBITAL RESHAPING WITH NOVEL QUANTITATIVE CRANIOMETRIC ANALYSIS

Presenter: David Y. Khechoyan, MD
Authors: Khechoyan DY, Saber NR, Birgfeld CB, Gruss J, Saltzman B, Forrest CR, Phillips JH, Hopper RA

Texas Childrens Hospital

Background: Surgical outcomes in cranio-orbital reshaping operations for metopic and unilateral coronal synostoses have been poorly quantified in the past. We present our two-center experience with devising and applying novel objective quantification strategies to more accurately evaluate the success of surgical correction.

Methods: Patients treated at The Hospital for Sick Children (HSC), Toronto, Canada, and Seattle Children's Hospital (SCH) were included in this study. For patients treated at HSC, a normative bandeau template was utilized to guide surgical re-shaping for a subset of patients (template, $n = 14$; no template, $n = 23$). In all patients, a virtual, computational version of the template was utilized as an outcome assessment tool to calculate an intervening area under the curve (AUC) between the normative template and patient's reconstructed supra-orbital bar on a CT scan. At SCH, patients (unilateral coronal, $n = 24$; metopic, $n = 32$) underwent craniometric axial radial vector analysis on CT scans at pre-operative, post-operative, and two-year follow-up time points. The radial vectors were converted into a Cartesian coordinate system, and quadrangular AUCs were calculated under areas of most conspicuous deformity (lateral brow; central forehead).

Results: For patients treated at HSC, the use of an intra-operative bandeau template led to a greater reduction in AUC (74% vs. 56%, $p = 0.016$), indicating better conformity between the reconstructed supra-orbital bar and the ideal, normal bandeau shape. For SCH patients, the stability of correction in UCS was more strongly predicted by pre-operative degree of deformity (Coef. = 0.88, $p < 0.01$) versus post-operative shape (Coef. = 0.60, $p < 0.01$), with a definite threshold to benefit of overcorrection. Conversely, for metopic suture synostosis, post-operative metrics were more closely associated with eventual outcome (Coef. = 0.76, $p < 0.001$) with a linear benefit for overcorrection.

Conclusion: Evaluation of surgical outcomes in craniosynostosis surgery is no longer relegated to subjective, unreliable ranking scales or anecdotal reports.

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PREVENTING TEMPORAL HOLLOWING IN FRONTO-ORBITAL ADVANCEMENT USING POLYLACTIC ACID (PLA) MESH

Presenter: William Y. Hoffman, MD
Authors: Hoffman WY, Bertrand AA, Nathan N
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Introduction: Temporal hollowing is often apparent in patients following fronto-orbital advancement. We have used a technique of placing polylactic acid (PLA) mesh over the osseous gap posterior to the terminus of the frontal bandeau when there is inadequate bone for reconstruction of the temporal region. This study reviews the outcome of this technique compared to autologous reconstruction.

Materials/Methods: We performed a retrospective chart review of all patients who underwent fronto-orbital advancement for coronal or metopic craniosynostosis from 1997 to 2010. We excluded syndromic patients, those who had other craniofacial abnormalities or had undergone prior surgeries at other centers. Operative technique, complications, and photographic outcome were recorded. The average length of photographic follow-up was 38.3 months. Outcome classification was determined by 2 independent observers using Bartlett's criteria. Statistical analysis was performed using t tests and linear regression.

Results: Fifty-seven patients were included in the study. 63% had a diagnosis of coronal synostosis ($N=36$) and 37% metopic synostosis ($N=21$). 56% were male ($N=32$) and 44% ($N=25$) were female. The mean age at the operative date was 7.65 months (range 3-18). 77% ($N=44$) of outcomes fell under I or IIA classification, and 98% when considering I, IIA, and IIB. Only one patient required a second surgery to obtain an improved cosmetic result. Surgical site infections occurred in 2 (3%) patients. One was a simple cellulitis while the other required surgical re-exploration and drainage. The use of PLA mesh resulted in outcomes equivalent to that seen with autologous bone ($p=0.74$). There was no statistically significant difference in outcome when considering coronal vs. metopic synostosis, age or gender.

Conclusions: In this study, we demonstrate that the use of PLA mesh gives equivalent outcomes to those of autologous bone reconstruction, with 77% of patients having excellent results. This is a simple technique that provides "guided bone regeneration" to improve results of fronto-orbital advancement.



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PREFABRICATED “PROFILE” CURVE TEMPLATES FOR FRONTORBITAL REMODELING IN CRANIOSYNOSTOSIS

Presenter: Kenneth Salyer, MD

Authors: Yamada A, Ueda K, Kajimoto Y, Salyer K, Nuri T, Gosain A, Harada T

Osaka Medical College

Background: Despite the well-known fact that forehead shape is one of the key of attractive face, only a few attempted to develop guide for remodeling frontoorbital shape. We, therefore, developed “profile” curve template, based on age-matched normal, and applied it to cranial vault remodeling in craniosynostosis.

Patient and Methods: We applied “profile” template for two cases: case 1; a four-year old boy with sagittal synostosis was presented with secondary deformity of the skull, especially unusually high forehead, after remodeling at other hospital. We performed anterior two-thirds remodeling, utilizing profile template to correct forehead profile. Case 2; we performed anterior cranial vault remodeling for 9-months old boy with trigonocephaly. We utilized age-matched bandeau template, as well as forehead profile template for remodeling.

Results: We were able to create smooth, nice profile of the forehead with the guidance of “profile template”.

Conclusion: Prefabricated “profile” template is a powerful tool to guide the optimal shape in cranial vault remodeling.

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BILAMBDROID AND SAGITTAL SYNOSTOSIS: REPORT OF 31 CASES

Presenter: Rebecca Wyten, MD

Authors: Chivoret N, Di Rocco F, Wyten R, Selek L, Cuttaree H, Renier D, Duracher C, Arnaud E

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Introduction: Bilambdoid and sagittal synostosis or «Mercedes pattern» is a multisutural craniosynostosis that has been described as a specific entity. However this synostotic pattern can also be found in faciocraniosynostosis. In order to better define this entity we reviewed our cases of bilambdoid and sagittal synostosis.

Patients and Methods: We searched our prospective database for cases of bilambdoid and sagittal synostosis among all types of craniosynostosis. We reviewed clinical findings, associated diseases and management.

Results: 31 patients were diagnosed with bilambdoid and sagittal synostosis among 4250 cases of craniosynostosis treated in our department over a period of 37 years. The mean age at presentation was 26 months. 74 % was male. Raised ICP was present in 13% of cases. A tonsillar prolapse was found in 18 cases. Among them, 7 were finally diagnosed as Crouzon or Pfeiffer syndromes. Several surgical techniques were used: isolated biparietal vault remodeling, posterior vault remodelling, posterior vault expansion with internal distraction. 3 of them required several surgeries. In 3 of them a craniovertebral junction decompression was performed. The mean follow-up was 71 months.

Conclusion: This study is the largest published series. Bilambdoid and sagittal synostosis constitute an isolated entity in almost 80% of the cases whereas in the remaining 20% it is part of a faciocraniosynostosis syndrome. Several techniques can be used in such condition. Tonsillar prolapse is found in some cases and seldom requires a specific treatment.



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COMPUTER-BASED QUANTITATIVE ASSESSMENT OF CRANIAL SHAPE FOR CRANIOSYNOSTOSIS

Presenter: Ben Wood, MD

Authors: Mendoza C, Wood B, Safdar N, Myers E, Rogers GF, Oh AK, Sauerhammer T, Keating R, Linguraru MG

University of Sevilla Spain

Purpose: Cranial shape is central to the diagnosis, treatment, and follow-up of children with craniosynostosis (CS). Nevertheless, shape is largely gauged by subjective measures that lack precision and prevent meaningful comparison. The purpose of this study was to determine the effectiveness of CT cranial shape analysis for quantitating the severity, location, and direction of cranial shape abnormalities consequent to CS.

Methods: An automated system was developed to quantify the cranial phenotypes of 141 subjects aged 0-12 months from thin-slice CT data (90 normal and 51 CS: 27 sagittal, 8 unicoronal, 16 metopic). All subjects were aligned using a registration procedure to correct for differences in scale and pose. Anatomic landmarks were automatically obtained on all CTs: posterior clinoid processes of the dorsum sellae (S1, S2), opisthion (OP), frontal, parietal and occipital bones. A statistical shape model was constructed and curvature/deformation fields for each of the cranial bones and sutures were obtained. A fusion index was derived from the number of adjacent bone voxels at sutures. Statistics of the shape fields computed over the different anatomical regions, and fusion indices on all sutures were compared between normal and CS subjects using Mann-Whitney tests. Finally, machine learning was used to diagnose and classify CS according to all the obtained features.

Results: Significant differences ($p < 0.05$) were found between fusion indices from normal and CS subjects. Deformation and curvature were significantly different in normal and CS subjects over most bone and suture regions. Combining all features it was possible to detect CS with average 92.7% sensitivity and 98.9% specificity. The probability of correctly classifying a subject into normal, sagittal, unicoronal and metopic CS, using the fusion indices of the sutures and the deformation and curvature fields of the different regions, was 95.7%.

Conclusion: Quantitative analysis of cranial shape is a viable approach for assessing the location, degree, and direction of abnormal anatomy and may assist in planning and assessing operative reconstruction

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NEURODEVELOPMENTAL OUTCOME OF NON-SYNDROMIC CRANIOSYNOSTOSIS

Presenter: Dawid Larysz, MD, PhD

Authors: Larysz D, Larysz P, Dowgierd K
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Non-syndromic craniosynostosis could be associated with developmental problems including improper speech acquisition, dyspraxia, learning disabilities and behavioral problems. Many hypotheses were proposed to explain aforementioned phenomena, concerning associated intracerebral anomalies, abnormal shape of brain in malformed skull and local intracranial hypertension with local hypoperfusion. Aim of the study was prospective analysis of frequency and type of neurodevelopmental problems in children with non-syndromic craniosynostosis and attempt to find factors that could influence their development. Studied group consisted of 124 children with non-syndromic craniosynostosis. Inclusion criteria consisted of: age before one year, confirmation of CSO in CT examination, absence of associated brain malformations and absence of other skull and other congenital anomalies. Developmental examinations were performed by team consisted of pediatric neuropsychologist, speech therapist, neurosurgeon and occupational therapist. We used Polish version of MFED and DSR tests, neuropsychological and speech evaluation, before surgery, than each 3 months until 2 years, and then each 6 months. Mean follow-up was 3 years (ranged from 1 – 6 years). Proper development was found in unoperated children in 68,97%. In nearly 27% of children multimodal developmental delay was noted. The most frequent were speech acquisition problems and dyspraxia. The highest percentage of developmental problems was noted in children with bilateral coronal and multisuture CSO. All children improve after multimodal therapy, but the highest rate of improvement was achieved in sagittal CSO, and children operated before 9 months of age. Developmental problems could affect children with non-syndromic craniosynostosis preoperatively. The most frequent abnormalities are speech acquisition delay and motor development abnormalities. Children operated before 9 months of age seem to have faster recovery period.



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BENIGN RADIOGRAPHIC CORONAL SYNOSTOSIS AFTER SAGITTAL SYNOSTOSIS REPAIR

Presenter: Anna Kuang, MD

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Whether cranial vault remodeling surgery for non-syndromic, isolated sagittal suture synostosis affects the patency of initially normal, unaffected sutures is unknown. The influence of coronal and lambdoidal suture patency after cranial vault remodeling on the trajectory of subsequent cranial growth is also unknown. Disruption of normal sutural anatomy during cranial vault reconstruction could influence the incidence of secondary craniosynostosis and need for reoperation. We performed a retrospective review of patients less than 1 year of age with non-syndromic sagittal synostosis treated at a single tertiary referral pediatric hospital from 2005 to 2010. Computed tomographic images obtained pre-operatively, immediately and 2 years post-operatively were evaluated for the occurrence of secondary synostosis of initially non-synostotic sutures. Surgical and ophthalmologic follow-up records were also analyzed for the occurrence of radiographic cranial restenosis, clinical or ophthalmological signs of intracranial hypertension, and reoperation. Fifty-one patients under 1 year of age underwent primary surgical repair of isolated, non-syndromic sagittal suture synostosis. Thirty-seven of these patients (71%) completed 2-year clinical and radiographic follow-up by the time of analysis. The average age at surgery was 5.4 months (range: 3.1-11.5 mo). Thirty-three of the 37 study patients (89%) showed radiographic evidence of bilateral secondary coronal synostosis (SCS). Five patients (15%) additionally showed partial lambdoid synostosis. One patient with radiographic SCS (3%) required reoperation for radiographic cranial restenosis, clinical signs and symptoms of intracranial hypertension, and papilledema. There is a high incidence of secondary coronal suture synostosis following cranial vault remodeling for isolated, non-syndromic sagittal synostosis. Post-operative secondary coronal synostosis was only rarely associated with secondary radiographic cranial stenosis, clinical or ophthalmological signs of intra-cranial hypertension and the need for re-operation.

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FRONTOFACIAL DISTRACTION. A RISK - BENEFIT ANALYSIS OF 90 CONSECUTIVE CASES

Presenter: David Dunaway, MBChB, FDSRCS, FRCS

Authors: Dunaway D, Britto J, Abela C, Evans R, Jeelani O

Great Ormond Street Hospital for Children

Purpose: To document the benefits and complications of frontofacial distraction and determine patient groups with the best risk-benefit ratios.

Method: A review of 90 consecutive cases of frontofacial distraction at Great Ormond Street Hospital. Information was collected prospectively from a standardised assessment protocol. Patients were assessed preoperatively and 1, 6 and 12 months postoperatively.

Results: There were 29 Apert and 61 Crouzon/Pfeiffer patients aged between 4 months to 22 years. 66 monobloc and 24 bipartition distractions were performed. Mean advancement was 16.6 mm (12-22 mm). Globe protection was achieved in all cases and papilloedema resolved when present. Airway obstruction improved in 85%. Younger patients achieved greater airway improvement than adolescents and bipartition was more effective than monobloc. All patients achieved an aesthetic improvement.

70 patients suffered complications. There were 13 major complications and 1 death. Major haemorrhage occurred in 9%. 26% suffered CSF leaks. The infection rate was 6.3%. Patients under 9 years were twice as likely to suffer CSF leaks, half as likely to suffer infections and had greater functional improvements than older patients.

Discussion: The benefits of frontofacial distraction are demonstrated along with the complication risk. The functional benefits of monobloc distraction in infants are significant and in the severely affected there may be no practical alternative. The benefits of bipartition distraction in Apert syndrome are also clear. For these two groups frontofacial distraction offers advantages that exceed the risks.

In older Crouzon's patients where the indications are aesthetic, the situation is unclear. Aesthetic outcomes of frontofacial distraction are excellent, but many authors report good results using a two stage frontal advancement and Le Fort III osteotomy with lower complication rates.

Conclusion: Frontofacial distraction is an effective procedure with a moderate complication risk. This risk -benefit ratio must be assessed against lower risk procedures, before recommending frontofacial distraction.



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THE BIRMINGHAM EXPERIENCE OF POSTERIOR CALVARIAL DISTRACTION: A 6-YEAR REVIEW

Presenter: Hiroshi Nishikawa, FRCS(PLAST)

Authors: Nishikawa H, Solanki G, Rodrigues D, White N, Evans M, Ahmad F, Noons P, Dover MS

Birmingham Childrens Hospital

Introduction: Posterior calvarial distraction osteogenesis was first carried out at the West Midlands Craniofacial Centre in 2006. It is a method of increasing cranial volume for the treatment of raised intracranial pressure (ICP), Chiari malformation and syrinx associated with multi-sutural craniosynostosis. The evolution of this technique, complications and outcomes are presented.

Method: Thirty four patients underwent posterior distraction between October 2006 and February 2013. Prospective Data was collected regarding age, diagnosis, indication for surgery, length of hospital stay, number and type of cranial distractors used, distraction distance achieved, consolidation period, complications, and outcomes.

Key Results: Median age at surgery was 19 months (4-230 months). 20 patients had a confirmed syndrome. Indications for surgery included raised ICP (n=22), unilateral lambdoid with Chiari malformation (n=2), Chiari malformation with syrinx (n=2), Chiari malformation alone (n=4), brachycephaly (n=4), pansynostosis (n=2). Three different distractor types were utilized. Posterior osteotomy cuts were supra-torcula (n=19) or infra-torcula (n=15). The mean distraction distance achieved was 21 mm (16-35 mm). Definitive improvement of Chiari malformation was observed in 3 patients and significant improvement of syrinx in 1 patient. Two patients subsequently underwent fronto-orbital advancement and one on to a monobloc. There was one repeat posterior distraction. Some complications encountered were incomplete osteotomy (n=1); early distraction was stopped in two cases because of significant CSF leak (n=1) and infection (n=1). There were 4 cases of CSF leak which required treatment but did not prevent completion of distraction. Some mechanical problems with distractors used early in the series (n=5) did not lead to distraction failure.

Conclusions: Posterior calvarial distraction osteogenesis permits controlled stable major increases in cranial volume that would be difficult to achieve using other techniques. The procedure can be repeated and allows further frontal transcranial surgery to be carried out if required.

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NORMALIZING FACIAL RATIOS IN APERT SYNDROME PATIENTS WITH LEFORT II MIDFACE DISTRACTION AND SIMULTANEOUS ZYGOMATIC REPOSITIONING

Presenter: Richard A. Hopper, MD, MS

Authors: Hopper RA, Kapadia H
Seattle Childrens Hospital

Background: Lefort 3 distraction (LF3D) advances the Apert midface, but leaves the concavity seen on “worms eye” view and the vertical compression of the central midface seen on frontal view untreated. We propose that Lefort 2 distraction and simultaneous zygomatic repositioning (LF2ZR) can move the central midface independent of the lateral orbits to treat the facial deformity in all three dimensions. The purpose of this paper was to determine if LF2ZR results in more normal facial proportions in Apert syndrome than LF3D.

Method: CT scan analyses were performed before and after distraction in patients undergoing LF3D (n=5) and LF2ZR (n=5). Unoperated Crouzon (n=5) and normal (n=6) controls were analyzed for comparison. Movement of the central compared to lateral face was measured relative to skull base using Dolphin software. Axial facial ratios (AFR) representing “worms eye” curvature, and vertical facial ratios (VFR) representing relative vertical facial height on frontal view were also calculated.

Results: With LF3D, facial ratios did not change with surgery and remained lower ($p < 0.01$; ANOVA with 2 sided t test) than normal and Crouzon controls. Although the face was advanced, its shape remained abnormal. With LF2ZR however the central face advanced 12.8 mm and vertically lengthened 12.4 mm more than the lateral orbits. In addition the medial canthi inferior movement leveled the palpebral dystopia, and the maxillary rotation closed the open bite deformity and increased nasal length. This differential movement changed the abnormal Apert facial ratios that were present before surgery ($p < 0.01$) into ratios that were not significantly different from normal controls.

Conclusion: The Apert face has distinct central to lateral facial proportions. Compared to LF3D, LF2ZR normalizes the position and the shape of the Apert face, resulting in more desirable periorbital and facial balance. LF2ZR has replaced LF3D in the treatment of the Apert midface in our center and is an alternate treatment to bipartition monobloc distraction in this patient group.



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**A CRANIOMETRIC ANALYSIS OF POSTERIOR
CRANIAL VAULT DISTRACTION OSTEOGENESIS**

Presenter: Jesse A. Goldstein, MD
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Background: Posterior cranial vault distraction osteogenesis (PVDO) has replaced fronto-orbital advancement in some centers as the first-line treatment in patients with syndromic craniosynostosis. Despite this fact, little has been written about the craniometric effects of PVDO on children with syndromic craniosynostosis. We present craniometric outcomes on the largest series of PVDO cases to date.

Methods: An IRB approved retrospective review of all patients who have undergone PVDO at our center was performed. Patient demographic, peri-operative data, and pre- and post-op CT scans performed as part of normal patient care were reviewed. Volumetric and craniometric indices were calculated and measured using commercial 3D imaging software.

Results: From 2008 to 2012, 22 patients underwent PVDO for suspected intracranial hypertension (ICH) (11 patients) or severe turribrachicephaly (11 patients). Diagnoses included Apert syndrome (N=3), Muenke syndrome (N=2), Saethre-Chotzen syndrome (N=5), Crouzon syndrome (N=4), Pfeiffer syndrome (N=2), VACTERL syndrome (N=1). 5 patients had unique genetic syndromes. In 13 patients, this was the first cranial vault procedure performed, while 8 had previous fronto-orbital advancement, and 1 had parieto-occipital reshaping. 50% of patients underwent PVDO prior to age 1; average age at surgery was 2.3 (0.3 to 14.1) years and distraction length averaged 27.3 mm (19 to 35) [“mm). Average length of surgery was 2.9 (1.6 to 3.8) hours, and average blood loss was 400 ml (200 to 600 ml). Total treatment length was 91 (to 147) days. Distraction length ranged from 19-35 mm (average 27.3 mm). Intracranial volume increase averaged 21.5% (7.5 to 70.0%; $p < 0.0001$) and 28.4% (10.8 to 66.0%; $p = 0.01$) in the subset of patients less than 1 year old. Posterior cranial height increased 12.2% (range: 0 to 35%, $p = 0.002$) and basofrontal angle decrease averaged 3.9% (range: 0 to 12%; $p = 0.003$) indicating a decrease in cranial height trajectory and improvement in frontal bossing.

Conclusions: PVDO is a safe/effective operation which may lower the risk of ICH and control turribrachicephaly.

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**THE ORTHOMORPHIC MONOBLOCK: A POWERFUL
TECHNIQUE TO PRODUCE CROUZON'S PATIENTS
WITH NORMAL APPEARANCE**

Presenter: Fernando Molina, MD
Author: Molina F

Hospital General Dr Manuel Gea Gonzalez SS

Traditionally the treatment of Crouzon disease include skull deformity correction, exorbitism and midface retrusion. Most techniques obtaine a satisfactory correction. However most of the time still patients look syndromic. We present a series of 36 Crouzon's patients, 20 male, 16 female. A Orthomorphic Monoblock was performed ages from 2-8 years. The technique includes the maxillae, orbits and the lower portion of the frontal bone “on block”, followed by osteotomies of the remaining frontal bone. A pair of internal distractors malar-zygoma are inserted and when moving the “on Block” osteotomy, the rest of the frontal bone moves in a passive way. The advancement overcorrecte the maxillae molar class II, the advanced orbits correct exorbitism and the forehead and skull normalize its forms. Initial results produce aesthetic and functional correction of the patients, providing an appearance of “normal people”, obtaining from a deformity the concept of orthomorphia. The overcorrection in molar class II, plus functional orthodontic mantains the maxillae in very satisfactory a forward position, widening the air track and correcting different degrees of hypoxemia during night. Maximum follow-up is 10 years and the analysis included cephalometric measurements correlated with cranial base. The study assess for a CF growth very close to normal for the orbital, malar and maxillary areas.



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MANDIBULAR GROWTH AFTER DISTRACTION
OSTEOGENESIS: CASES OF PIERRE ROBIN
SYNDROME IN EARLY CHILDHOOD

Presenter: Masakazu Hasegawa, MD
Authors: Hasegawa M, Mitsukawa N, Saiga A,
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Chiba University

Objective: We performed mandibular distraction osteogenesis in infants and toddlers with obstructive sleep apnea (OSA) caused by micrognathia. In the present study, we examined pre- and postoperative mandibular growth in 9 such patients with Pierre Robin syndrome.

Subjects and Methods: The subjects were 9 patients with Pierre Robin syndrome who were 1 month to 3 years old and who had OSA since birth due to micrognathia. All patients underwent bilateral mandibular distraction osteogenesis using internal fixation devices. Cephalograms were used to measure the lengths of mandibles over time to examine their postoperative growth. Then the mandibular lengths were compared with data of standard mandible growth. The postoperative follow-up period ranged from 4 years to 9 years 6 months.

Results: The mandibles were clearly small and underdeveloped preoperatively. In most patients, the mandibular lengths increased after distraction osteogenesis and thereafter the mandibles continued to grow without falling greatly below the standard growth data of mandibles. However, mild OSA recurred in some patients accompanying mandibular underdevelopment, and two of these patients underwent mandibular distraction osteogenesis again.

Discussion: There are very few reports examining the bone growth of mandibles after distraction osteogenesis. In the present study, we examined over time the mandibular lengths of 9 infant and toddler patients with Pierre Robin syndrome after they underwent mandibular distraction osteogenesis. Our results showed that the mandibular growth was not inhibited after the surgery, and the mandibles grew at rates close to normal. That is, it was speculated that some type of catch up growth was in effect due to mandibular distraction osteogenesis. However, there have been clearly more cases with underdevelopment of the mandibles compared to maxillary growth, and mild OSA has recurred postoperatively for patients who had underdeveloped mandibles. Thus, it is important to overcorrect in distraction osteogenesis.

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CORRECTING THE TYPICAL APERT FACE:
COMBINING BIPARTITION WITH MONOBLOC
DISTRACTION

Presenter: Aina V. Greig, MA PhD FRCS(Plast)
Authors: Greig AV, Britto JA, Abela C, Witherow H,
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Great Ormond Street Hospital

Background: Bipartition distraction is a novel procedure combining frontofacial bipartition and monobloc distraction. Apert syndrome and other syndromic craniofacial dysostoses are often characterised by hypertelorism, with a negative canthal axis and counter-rotated orbits. Central midface hypoplasia can result in a biconcave face in both midsagittal and axial planes. Bipartition distraction can correct these facial abnormalities.

Objectives: To examine the functional and aesthetic changes in patients undergoing bipartition distraction and to assess outcomes and complications.

Methods: Twenty patients (19 Apert, 1 Pfeiffer; 1.6 - 21 years) underwent bipartition distraction using the RED II frame. The severity of appearance was graded pre- and postoperatively as mild, moderate or severe. Functional problems were documented pre- and postoperatively by a multidisciplinary team during a full craniofacial assessment. 3D CT and OsiriX software were used to measure central and lateral midface skeletal advancement. Follow up ranged from 15 months to 7 years.

Results: Bipartition distraction consistently produced more central than lateral facial advancement. Mean central advancement at sella-nasion was 13.2mm (SD 5.9mm) and at sella-A point was 11.7mm (SD 5.4mm). Lateral advancement was 4.7mm (SD 2.8mm). Unbending the face improved aesthetic appearance. Functional issues in terms of airway, eye exposure and elevated ICP were improved. Complications included: 6 temporary CSF leaks (4 needing a lumbar drain), 5 patients with post-op seizures, 5 patients requiring RED frame repositioning, 1 palatal fistula, 1 velopharyngeal incompetence, 5 pin site infections, 1 abscess under frontal bone, 3 sepsis, 9 worsened strabismus, 2 enophthalmos, 1 partial visual field loss, 3 re-intubations due to aspiration.

Conclusions: Bipartition distraction is an effective procedure to differentially advance the central face in Apert syndrome and improves both function and aesthetics.



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**DOUBLE JAW SURGERY AFTER ADOLESCENCE
FOR HEMIFACIAL MICROSOMIA CASES IN WHICH
DISTRACTION WAS APPLIED IN CHILDHOOD**

Presenter: Yuhki Uchida, MD
Authors: Satoh K, Uchida Y, Mitsukawa N, Akita S,
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Chiba University

Background: Since the report of McCarthy for the application of distraction for hemifacial microsomia, distraction has been widely spread in the world. Although distraction technique tremendously augments the bone volume actually in the distracted area, the facial skeletal harmony has not always been maintained as the time goes by. Herein the authors report the cases of double jaw osteotomy after adolescence for the deformed facial skeleton in which distraction was applied in childhood.

Materials: Distraction was applied to 38 cases of hemifacial microsomia since 1988. 6 Type I, 17 Type IIA, 10 Type IIB, and 5 Type III are included. The post-distraction growth has not always been caught up with in some cases, and some exhibited facial deformity with occlusal inclination and or hypo-plastic cheek. Among them, double jaw osteotomy was applied to 5 cases (1 Type IIA, 3 Type IIB, and 1 Type III).

Results: Occlusal inclination and facial skeletal harmony has been improved much for all the osteotomized cases. Alloplastic materials has been implanted as bony augmentation to obtain the facial symmetry in some cases.

Conclusion: Although distraction technique has been extremely effective procedure for deformed hemifacial microsimia, facial skeletal growth has not been always maintained, and occlusal inclination has recurred and hypo-plastic cheek was getting figured out as the growth has progressed in some cases. Double jaw osteotomy and or touch up surgery to obtain the facial symmetry as much as possible are required in some of the cases in which distraction was applied in childhood.

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**POSTERIOR POLE EXPANSION WITH SPRINGS
WITHOUT OSTEOTOMIES: RESULTS AND PREDICTIVE
FACTOR OF SUCCESS**

Presenter: Monica Fawzy, MD
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Craniofacial Unit

Introduction: Infants affected with either brachycephaly or Apert syndrome present frequently a posterior flattening of the cranial vault, with open lamboid sutures. This posterior deformation may lead to the development of a turriccephalic morphology. An early posterior remodeling of the posterior pole has been proposed for diminish this risk. To achieve this, trans lamboid springs can be utilized across open lamboid sutures.

Surgical Technique: The patient is positioned in decubitus ventral and both lamboids sutures and the lambda are exposed. The springs are molded with 1.5 mm steel cables, U-shaped and positioned across the suture. Its not necessary to perform osteotomies. During the post operative period, the infant is placed in decubitus ventral or lateral (decubitus dorsal is avoided), and after 3-6 months the springs are removed by small incisions (frequently at the time of the surgery of fronto-facial advancement).

Results: We have used this technique in 15 infants (7 Apert, 7 brachicephaly, 1 Saethre Chotzen). We had 2 complications, namely one case of spring displacement and one case of cutaneous perforation. A volumetric analysis was available for 11 patients operated between 2006 and 2010. At 30 days the mean gain in intracranial volume was of 7,48%, a 70 days the mean gain in intracranial volume was of 14,72% (max 23%, min 6%).

The treatment with springs was effective in preventing a turriccephalic morphology: the ratio between the vertical height of the cranium (basion to vertex) and the anterior-posterior diameter (nasion to inion) diminished after the treatment (from 1,05 to 0,97 at the time of the last tomographic control, mean: 111 days after surgery).

Children with >10% volume increase at 70 days had a significative smaller age at intervention (mean 199 and 176 days) than the group with <10% increase (mean age at intervention: 313 days, p=0,033).

Conclusion: The spring assisted technique has allowed a trans suture distraction with a small morbidity, and it is an effective option for a volumetric expansion allowing to delay the anterior cranial surgery for a most favorable condition.



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FRONTOFACIAL MONOBLOC ADVANCEMENT WITH DISTRACTION IMPROVES CONSIDERABLY THE APNEA AND APNEA-HYPOPNEA-INDEXES IN A 2-YEAR FOLLOW-UP PERIOD

Presenter: Junnu Leikola, MD, DDS, PhD
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Craniofacial Unit

Background: Patients with faciocraniosynostosis are likely to suffer from obstructive sleep apnea (OSA) due to midface hypoplasia. Frontofacial monobloc osteotomy with osteodistraction (FFMO) is an effective procedure to correct such severe midface retrusion. Polysomnography (PSG) has widely replaced oxymetry in the assessment of respiratory outcome after FFMO.

Methods: 106 patients with Crouzon, Apert or Pfeiffer syndrome underwent FFMO between September 2000 and July 2010 at the craniofacial unit of Necker Enfants Malades Hospital, Paris. They were prospectively evaluated for the respiratory outcome. To minimize the effect of surgical technique, operations were performed by the same craniofacial surgeon with a standardized protocol. Respiratory function was evaluated by PSG preoperatively and at 6 months, 1 and 2 year recordings. PSGs were quantitatively evaluated to assess the severity of OSA.

Results: 39 preoperative PSGs were recorded. 18 patients had evaluation at 6 months, 18 at 1 year and 6 at 2 years. The Apnea Index was improved in 84% at 6 months, 78% at 1 year and 83% of the cases at the 2 years PSG compared with preoperative figures. Even superior improvement to preoperative Apnea-hypopnea Index was found in 88%, 89%, of the recorded cases, respectively. The mean saturation, percentage of time recorded with saturation under 90% and the overall desaturation recorded showed minimal differences at the postoperative recordings compared with the preoperative ones.

Conclusions: Apnea-hypopnea Index appears to improve considerably after FFMO. Along with equally improved Apnea Index it seems that FFMO provides a consistent and stable improvement to OSA for at least a 2 year time period.

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IMPROVING NUTRITIONAL OUTCOMES IN SYMPTOMATIC PIERRE ROBIN SEQUENCE: A COHORT COMPARISON STUDY BETWEEN TONGUE LIP ADHESION AND MANDIBULAR DISTRACTION

Presenter: Lorelei Grunwaldt, MD
Authors: Rottgers A, MacIsaac Z, Wine T, Grunwaldt L, Mehta D, Kumar AR

University of Pittsburgh School of Medicine

Background and Purpose: The optimal treatment of Pierre Robin Sequence (PRS) associated dysphagia remains poorly characterized. This study aims to compare the feeding outcomes of symptomatic PRS patients treated with tongue lip adhesion (TLA) or mandibular distraction (DOG).

Methods: A retrospective cohort study of symptomatic PRS patients treated from August 2008 till June 2012 was performed to compare feeding outcomes between TLA (Group 1) and DOG (Group 2)

Results: Eleven TLA and ten DOG patients were identified. The average age at TLA was 0.9 months (0.07-2.2) and 0.8 months (0.03-1.7) at DOG. The average follow-up was 33 months (12.9-43.5) for TLA and 12.5 months (2.1-22.5) for DOG. Three (27%) TLA and four (40%) DOG patients were syndromic. The post-TLA gastrostomy rate was 54.5% (6 of 11) compared to the post-DOG gastrostomy rate of 20% (2 of 10); yielding an odds ratio of 4.8 ($p=0.183$). In Group 1 (TLA) 60% (3 of 5) of patients grew faster than their projected growth curve after surgery, 20% (1 of 5) grew parallel to their growth curve, and 20% (1 of 5) grew slower than their growth curve. In Group 2 (DOG) 50% (4 of 8) of patients grew faster than their projected growth curve after surgery, 25% (2 of 8) grew parallel to their growth curve, and 25% (2 of 8) grew slower than their growth curve. No statistical difference in growth rates was observed ($p=1.0$). The major complication rate was 8% in Group 1 (TLA), including one mortality and 0% in Group 2 (DOG). Minor complication rates were 9% in Group 1 (TLA) with labial mucosal redundancy and 20% in Group 2 (DOG), including 1 pin site infections treated successfully with drainage, antibiotics and hardware retention, and one transient marginal mandibular nerve palsy.

Conclusions: Mandibular distraction may be superior compared to tongue lip adhesion in avoiding gastrostomy for supplemental alimentation in symptomatic Pierre Robin Sequence. This would be a significant decrease in health care burden for the patient and family in the neonatal period. Growth rates were similar for both treatment modalities in patients where gastrostomy was avoided.



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**SUB-CRANIAL ROTATION DISTRACTION
ADVANCEMENT OF THE ENTIRE FACE FOR THE
TREATMENT OF SEVERE OBSTRUCTIVE APNEA
ASSOCIATED WITH SYMMETRIC CRANIOFACIAL
MICROSOMIA**

Presenter: Hitesh Kapadia, DDS, PhD
Authors: Kapadia H, Hopper RA
Seattle Childrens Hospital

Background: Craniofacial Microsomia (CFM) has variable presentation and can affect any part of the craniofacial skeleton. Frequently, there is a differential effect on the maxilla-mandible relationship, resulting in malocclusion, asymmetry and a distinct clinical phenotype. We present two cases of CFM with symmetric involvement, acceptable maxillo-mandibular relationship, and severe obstructive sleep apnea (OSA). Airway evaluation demonstrated multiple levels of obstruction requiring tracheostomy. Cephalometric evaluation revealed a sub-cranial clockwise rotation hypoplasia deformity that was not clinically evident.

Purpose: To describe the rotation deformity and evaluate the success of simultaneous subcranial Lefort III rotation advancement with bilateral mandible ramal distraction lengthening treatment for their OSA.

Methods: Superimposition skullbase analysis was performed on lateral cephalograms pre- and post-treatment. OSA was measured using polysomnography. Both patients were treated with subcranial Lefort III rotation advancement hinged at nasion along with simultaneous bilateral ramal lengthening. They were maintained in MMF during activation and the movement was driven by an external halo based device with two buried mandible distracters.

Results: Superimposition of Bolton standards revealed a pre-operative subcranial normo-occlusive rotation deformity. Post-operative analysis demonstrated successful counterclockwise rotation of the entire sub-cranial facial skeleton while maintaining pre-operative occlusion. Polysomnography showed a drop in AHI from 38 and 84 pre-operation to 4.7 and 1.3 post-treatment. Both patients remained decannulated and without positive pressure support on last follow-up 4 and 1.5 years post-treatment.

Conclusions: Simultaneous clockwise rotation of the entire subcranial facial skeleton is possible using distraction. This differential advancement and the posterior lengthening of the maxilla and mandible had a dramatic effect on multi-level OSA, and has implications for other craniofacial conditions with rotation deformities like Treacher Collins.

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**OUR PROVISIONAL EXPERIENCE OF APPLYING
DISTRACTION OSTEOGENESIS IN REMODELING
CRANIOSYNOSTOSIS**

Presenter: Masayuki Miyata, MD, PhD
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Background: Although distraction osteogenesis has been used for patients with craniosynostosis more than 10 years in Japan, its application does not spread worldwide. The technique itself has the potential advantages such as the decrease of postoperative relapse, the easy overcorrection, the avoidance of epidural dead space and the potential for infection, tension free wound closure and the elimination of scalp wide scar. But we recognized some problems in clinical use in our limited cases.

Aim: The purpose of our study was to evaluate the effectiveness of distraction osteogenesis for remodeling the craniosynostosis in our provisional cases. We also present an important aspect of the operative procedure refined by ourselves.

Method: Last 10-year period, we treated 24 children with craniosynostosis. Among them, 8 patients underwent cranial remodeling by gradual distraction. Four cases were syndromic (2 Crouzon, 2 Apert) and 4 were nonsyndromic (2 sagittal, 1 bicoronal, 1 unicoronal and unilambdoid). The patients consisted of 5 boys and 3 girls and ranged in age from 7 to 47 months at the first surgery with a median follow-up of 85 months (range, 30 to 120 months). Simulated surgery was performed on a 3D model in last 4 cases. The effect of surgery was assessed by comparing the preoperative and postoperative CT scans.

Results: Remodeling of deformities and gaps occur after removal of the distraction devices, but the postoperative cranial configuration was favorable in just 4 cases. The irregularities were conspicuous in 2 nonsyndromic cases and the re-advancement was needed for 2 syndromic cases. We resigned distraction and changed to the conventional method in 2 cases because of the dura damage during the undermining at the anterior cranial base.

Conclusion: In unfavorable syndromic cases, we noticed undercorrection not keeping up with the growth. We also identified our osteotomy lines were too simple to correct complex configuration in nonsyndromic cases. We must stress the importance of preoperative planning when applying distraction osteogenesis in both syndromic and nonsyndromic craniosynostosis.



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EVALUATION OF THE AIRWAY AFTER MONOBLOC DO A TEN YEAR FOLLOW UP

Presenter: Carlos R. Barcelo, MD

Authors: Barcelo CR, Genecov DG, Salyer KE
International Craniofacial Institute

Summary: Upper airway stenosis in patients with syndromic craniosynostosis is very common and often severe. Midface advancement with distraction can improve respiration dramatically and prevent a tracheotomy or result in its removal. In this study we reviewed the respiratory outcome with mono-bloc advancement with distraction in 6 faciocraniosynostotic patients over a 10 year period. Early respiratory results of were very good and remain stable at long-term follow up.

Materials and Methods: A retrospective study was performed on 6 patients with a craniosynostosis and associated midface retrusion (Apert, Crouzon or Pfeiffer syndrome) they all had a Monobloc advancement with distraction between 2001 and 2003 at the International Craniofacial Institute Dallas Texas. The preoperative and postoperative respiratory status was studied using Polysomnography, Nasoendoscopy and 3D CT evaluation of the airway. The first postoperative respiratory assessment was undertaken 3 months after the procedure. Long-term evaluation was performed at least 12 months after the intervention and at least 6 months after the removal of the distractors and then once a year for the last ten years. Relapse of respiratory symptoms was defined as worsening of symptoms at the late evaluation compared to the early postoperatively. Mean age at surgery was 4 years (range 7 months–12 years) 100% had respiratory problems preoperatively.

Results: Within the early postoperative period 4 out of 6 patients were decanulated 1 patient never had a tracheotomy, 1 patient failed decanulation and required a Le Fort I advancement which then resulted in decanulation. The decanulated patients kept a normal breathing pattern.

Conclusions: The respiratory improvement takes places in the early postoperative phase and the results are stable over a long period of time The patient that required secondarily a Le Fort I advancement was directly related to lack of movement on the center of the midface more likely due to incomplete disjunction of the midface. Consolidation period time of more than 3 months is mandatory for monobloc distraction.

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FRONTO-ORBITAL ADVANCEMENT (FOA) VERSUS POSTERIOR ENLARGEMENT, WHICH IS FIRST?: OUR CURRENT TIMING FOR THE SURGERY IN SYNDROMIC CRANIOSYNOSTOSIS

Presenter: Kaneshige Satoh, MD

Authors: Satoh K, Mitsukawa N, Akita S, Kubota Y
Chiba University

Background: In syndromic craniosynostosis, multiple suture-synostosis are often encountered. Posterior decompression by removing substantial amount of occipital bone is followed by FOA for long. In our series, distraction (DO) devices are very often applied for the craniosynostosis, and it is advantageous that the enlargement amount can be controlled by the surgeon. Here the current timing for the surgery (FOA or posterior enlargement. Which is first?) will be discussed in our recent 19 series.

Method: The study consists of 19 cases with a range of 8 years follow-up. 10 Crouzon, 3 Apert, 5 Pfeiffer, and 1 Sathre-Chozen syndrome cases are included. First surgery was conducted in 5-12 months of age in all the cases.

Result: In 16 cases, FOA was firstly conducted before occipital enlargement, and among them, in 4 cases, occipital surgery is under consideration. In 2 cases, occipital enlargement is followed by FOA. One is Pfeiffer with hydrocephalus and severe intracranial hypertension with multiple honey comb appearance. The other one exhibited severe posterior honey comb. In 19 FOA conducted cases, DO was applied for 10 cases, and conventional procedure was applied for 9 cases. Occipital enlargement by DO was applied for 13 cases except one in 14. As for FOA, although DO was basically applied for FOA to obtain and maintain much advanced volume, the conventional procedure was applied for infants with fragile calvarial bone or with asymmetric plagiocephalic forehead.

Conclusion: While in a typical syndromic craniosynostosis, simple FOA is firstly done in early infancy, in severe cases with multiple calvarial honey combs defects and/or prolapsing exophthalmos it is controversial which should be first. FOA or posterior enlargement, which should firstly done will be different individually. Although FOA by conventional osteotomy and reshaping is workhorse, FOA or posterior enlargement by distraction technique is actually advantageous to some extent. Large amount of enlargement can be guaranteed by distraction for severe cases with multiple calvarial honey combs defects.



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HOW DISTRACTION VECTORS SHOULD BE USED IN APERT SYNDROME FOR THE CORRECTION OF THE SKULL AND FACE

Presenter: Pablo Arrieta, MD

Authors: Molina F, Arrieta P

Hospital General Dr Manuel Gea Gonzalez SS

Apert syndrome is one of the syndromic craniosynostosis with more difficulties during treatment. Contemporary treatment include occipital expansion in the first months of life and then a monoblock advancement for functional and aesthetic correction of the forehead and facial deformity. We present a series of 9 Apert patients, treated between 6 months to 4 years of age with a combine occipital expansion and monoblock. 5 female and 4 male were include. First surgical procedure is to remove the synostotic occipital cranial sutures and the insertion of "springs". Then 8-10 months later, a second surgical procedure includes: to remove the "springs" and to perform a monoblock distraction advancement. Together with the oblique lateral distraction vectors, a central one should be include in order to promote a more effective interorbital and frontal advancement to correct the supraorbital flattening characteristic of this syndrome. An extra wire traction has been used in 5 kids and a simple posterior frontal bone weakening in 4. The combined treatment produce a highly satisfactory result with a nice reshaping of the posterior skull aspect, and achieving an efective fronto-orbital-maxillae advancement ranged from 20 to 32 mm at the orbital level. The extra interorbital advancement corrects the supraorbital hollowing and producing a nice curvature that follows the new orbital position and giving the impression that we correct interorbital distance with a very pleasant aesthetic result, all of this together with the correction of midface and exorbitism. Surgical technique details, pre and post CT-Scan, intracranial views will be present. Maximum follow up is 34 months and clinical pictures will be presented also.

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THE ROLE OF POSTERIOR CRANIAL VAULT DISTRACTION FOR THE TREATMENT OF CRANIOSYNOSTOSIS

Presenter: Yuzo Komuro, MD

Authors: Komuro Y, Matsumoto S, Mochizuki M, Koizumi T

Juntendo University Urayasu Hospital

Background: In cases of posterior skull stenosis, particularly those with increased intracranial pressure (ICP) and those with chronic tonsillar herniation, it is necessary to attempt posterior skull expansion. Aim of this paper is to present our experience based on 13 cases.

Methods: From 1998 to 2013, 13 patients with craniosynostosis (four Crouzon syndrome, two Pfeiffer syndrome, two clover-leaf skull, two multiple suture synostosis, one Saethre-Chotzen syndrome, one unilambdoid synostosis, and one cranial cleft) presenting with severe occipital flattening underwent posterior skull expansion. Mean age at surgery was 17 months (min: 2, max: 91). After 2001, we applied the distraction technique for occipital expansion in 8 of 12 cases (Komuro, et al. J Craniofac Surg 2009).

Results: The posterior expansion obtained with conventional methods were insufficient except one case which showed elevated ICP. By using distraction technique, we were able to perform bone extension of 30 mm or more and achieve sufficient skull expansion for the occipital regions without bone defects.

Conclusions: In cases of surgery for craniosynostosis with posterior flattening, it is not possible to achieve sufficient expansion of the skull through fronto-orbital advancement alone. Although it is necessary to expand the occipital region, the surgery is risky and highly invasive. We applied the distraction osteogenesis technique for occipital expansion. The present method is extremely useful for skull formation in cases of craniosynostosis with posterior flattening.



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FRONTO-FACIAL MONOBLOC AND LE-FORT III DISTRACTION OSTEOGENESIS IN CRANIOSYNOSTOSIS-THREE-DIMENSIONAL EVALUATION OF TREATMENT OUTCOME AND THE NEED OF CENTRAL DISTRACT

Presenter: ChingHsuan Hu, MD

Authors: Hu CH, Chieh-Tsai WU, Wen-Ching KO, Kuo-Ting CH

Chan Guan Memorial Hospital

Purpose: The objectives of this study were to investigate the treatment effect, stability and outcome of frontofacial monobloc or Le-Fort III distraction osteogenesis in syndromic or non-syndromic Craniosynostosis by Three-dimensional Evaluation.

Materials and Methods: Thirteen consecutive patients who underwent FF monobloc or Le-Fort III distraction during 2003 to 2010 were included with age ranged from 4.8 to 18.4 year old. The diagnosis consists of eight Crouzon, two Apert, one Pfeiffer syndromes, one bicoronal synostosis and one lymphoma. The evaluation included clinical records, and computerized tomography for at least one year follow up (average 40.6 months). The pre-treatment and post-treatment changes were measured. The intracranial volume, upper airway volume and globe protrusion were calculated from computerized tomography by AVIZO software before and after treatment. Complication will also evaluate and discuss.

Results: The overall distraction period was 113 +/-24 days. After distraction, the intracranial volume, upper airway volume and globe protrusion all had significant increase compared to pre-operation status. Bilateral eminence advancement was significant more than central advancement. The overall complication including zygomatico-maxillary suture or other skull bone fracture (3/13), depressed nose (2/13), trismus due to temporalis calcification, severe temporal hollowing with hypernasality (1/13), one molar impaction into sinus (1/13), and one device failure (1/13).

Conclusion: Monobloc and Le-Fort III distraction is effective for relieving related symptoms and signs through differential external distraction at different vertical levels of the face which also improve patients' general appearance and result in better psychological and mental development. Central advancement is still not as the same as bilateral eminence. Central distraction is considerable in future.

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EARLY MIDFACIAL DISTRACTION FOR SYNDROMIC CRANIOSYNOSTOTIC PATIENTS WITH OBSTRUCTIVE SLEEP APNEA

Presenter: Nobuyuki Mitsukawa, MD, PhD

Authors: Mitsukawa N, Saiga A, Kaneko T, Akita S, Satoh K

Chiba University

Syndromic craniosynostosis is known to be associated with various obstructive respiratory disorders, including sleep apnea. We performed early midfacial distraction in 11 syndromic craniosynostotic patients with obstructive respiratory apnea and obtained good results. There were 4 patients with Crouzon syndrome, 3 patients with Pfeiffer syndrome, and 4 patients with Apert syndrome. Their ages ranged from 7 months to 3 years, 9 months (mean: 2 years, 5 months). Midfacial distraction was performed using an internal or external device to improve obstructive respiratory disorders, such as sleep apnea, and to avoid tracheostomy. Evaluation was performed by comparison of the pre- and postoperative polysomnograms (PSGs) and cephalograms. All patients had markedly improved respiratory disorders during sleep and avoided tracheostomy. Preoperative PSGs and cephalograms were compared with those from 1 year to 1 year, 6 months postoperatively. The postoperative PSGs and cephalograms showed marked improvements compared with preoperative PSGs and cephalograms. Syndromic craniosynostotic patients have facial bone hypoplasia, particularly of the maxilla. The tongue and parapharyngeal soft tissue become enlarged, and the enlarged tissues can cause airway obstruction. In this study, marked improvements were observed in physical evaluation using PSGs and morphological evaluation using cephalograms. Early midfacial distraction is thought to be a very useful method to reconstruct a large pharyngeal cavity, to enlarge the airway, to improve obstructive respiratory disorders, and to avoid tracheostomy. It is important to select a device suitable for each case when surgery and distraction are performed.



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OUR NEW PROCEDURE FOR PROBLEMS AND COMPLICATIONS OF LEFORT MIDFACE DISTRACTION

Presenter: Keisuke Imai, MD
Authors: Imai K, Takahashi M, Yamaguchi K, Ishise H, Takara A, Nochi A

Osaka City General Hospital

Recently, craniofacial distraction osteogenesis has become a valuable technique. Midface distraction osteogenesis serves as the primary surgical treatment for children with severe midface hypoplasia. External distractors have several points of fixation, providing multiple levels of traction. However, some problems and complications have been reported. Since 1997, 212 craniofacial gradual distractions have been performed; 35 of these were LeFort midface distractions. In our LeFort experience, there were some problems and complications; inappropriate change of force application level and vector control, eventually leading to improper rotation of the osteotomized bony segment and an unwanted dentoalveolar effect, fixation failure due to fracture of the zygomatic maxillary suture and intracranial fixation pin migration and/or slipping. We devised new procedures for this technique to avoid these problems and to continue using the distraction procedure after overcoming the complications.

For vector control, dentoalveolar problems and fixation failure, it was useful to pull the titanium plate fixed on the piriform margin through the nasal cavity, and we developed an external device with or without transmalar pinning for this purpose. After intracranial fixation pin migration, it was possible to continue LeFort III midface distraction by using a device with a plaster cast as anchorage.

In these results it was easier to achieve vector control and it was possible to avoid some problems (improper rotation of the osteotomized bony segment and an unwanted dentoalveolar effect, fixation failure due to fracture of the zygomatic maxillary suture, etc.). However, it was difficult to achieve elongation exceeding 25mm. In a cranial bone fracture case, distraction was completed to achieve the ideal advancement. In conclusion, our new procedure can serve as a highly effective fixation approach to obtain more accurate vector control. It is useful for continued distraction after intracranial fixation pin migration to perform distraction with a plaster cast as anchorage.

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THE IMPACT OF POSTERIOR CALVARIAL DISTRACTION ON CHIARI MALFORMATIONS AND SYRINGOMYELIA

Presenter: Martin Evans, MD
Authors: Ahmad F, Solanki G, White N, Evans M, Nishikawa H, Dover S, Rodrigues D

Birmingham Children's Hospital

Introduction: Treatment options for Chiari malformation and syringomyelia include conservative management, foramen magnum decompression with or without a diversion procedure and syringo-subarachnoid shunting (SSS). Posterior fossa craniectomy and duroplasty has also been performed to create more room within the posterior fossa.

Methods: Two patients with syndromic pan-craniosynostosis, Chiari I malformation and syringomyelia underwent posterior calvarial expansion using internal distractors. Following maximum distraction of 30mm and the consolidation phase, the patients were followed up in a multidisciplinary craniofacial clinic and MRI imaging was performed at about a year post-surgery.

Results: The ages of the patients were 6 years and 8 years. There were no intra-operative or post-operative complications. The procedure and distraction were well tolerated. Developmental assessment, fundoscopy and general behaviour were normal at follow-up. Imaging demonstrated regression of the cerebellar tonsillar descent as well as near total resolution of the syrinx and an improvement in intracranial venous hypertension.

Conclusion: In a selected group of patients, with or without craniosynostosis, distraction osteogenesis is a safe and effective method for treating Chiari malformation and syringomyelia with less morbidity than other traditionally used methods.



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LONG-TERM STABILITY OF ANTERIOR OPEN-BITE CLOSURE WITH DISTRACTION OSTEOGENESIS IN TREACHER COLLINS SYNDROME

Presenter: Jose Cortes-Arreguin, MD

Authors: Cortes-Arreguin J, Garcia-Garcia F, Molina F
Hospital General Dr Manuel Gea Gonzalez

Background: The deficient mandible in Treacher Collins Syndrome conveys a convex profile, lack of chin projection, presence of redundant submental soft tissue, and a class II malocclusion. The sagittal retrusion at pogonion is made worse by the clockwise rotation of the mandibular body. An anterior open bite is typically present. After conventional osteotomies to correct the anterior open bite deformities, the contracted muscles and overlying soft-tissue envelope act as a counterforce to the bony advancement, often causing bony relapse and necessitating multiple procedures to achieve optimal aesthetic results. The aim of this study was to evaluate the stability after distraction osteogenesis procedures for correction of anterior open bite deformities in Treacher Collins Syndrome.

Methods: From January 1994 to July 2011, 36 patients with Treacher Collins Syndrome underwent distraction osteogenesis procedures in our center to correct the anterior open bite. The sample was divided into three groups: Group A (3-8 yrs.): bilateral bidirectional distraction osteogenesis (18 cases), Group B (>13 yrs.): bilateral unidirectional distraction osteogenesis plus bilateral mandibular body osteotomy plus osseous genioplasty (10 cases) and Group C (>13 yrs.): Bilateral Sagittal Split Mandibular Osteotomy (BSSO) plus distraction osteogenesis with elastics traction. We report surgical technique, complications, and long-term outcomes through cephalometric and 3D CT analysis.

Results: From January 1994 to July 2011, 36 patients underwent distraction osteogenesis procedures with a mean follow-up of 18 months. Treatment resulted in good occlusion, enhancement of the facial profile, and good dental aesthetic appearance. The treatment outcome was satisfactory without bony relapses at long term follow up.

Conclusions: Distraction osteogenesis for correction of anterior open bite deformities in Treacher Collins syndrome is a technique capable of solving definitively the malocclusion with the benefits of bone distraction of tissue elongation impacting in a better cosmetic result and tissue stability.

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SURGICAL ANATOMY OF SPHENOMAXILLARY DISJUNCTION IN THE LE FORT III OSTEOTOMY

Presenter: William S. Tierney, MS

Authors: Tierney WS, Orra S, Doumit G
Cleveland Clinic Foundation

Background: Le Fort III osteotomy (LFIII) represents the foundation of surgical correction for midface hypoplasia. One serious complication of LFIII is severing the maxillary artery or its branches (MA) during osteotome advancement for sphenomaxillary osteotomy. This study seeks to characterize relevant anatomy of the infratemporal fossa (IFF) and MA as it enters the sphenopalatine fissure (SPF) to better inform operative planning.

Methods: Bilateral midface dissections were performed on 10 fresh, adult cadavers without dysmorphic pathology. Zygomatic arch and mandible were removed for complete visualization and measurement of 14 relationships. JMP statistical package used to calculate descriptive statistics and generate predictive linear statistical models.

Results: All measurements are given as mean and standard deviation. Measurement from anterior-superior aspect of the zygomatic arch to the sphenopalatine artery (SPA) entering the SPF: 38.9 ± 3.2 mm. Alveolar process of the maxillary bone to the SPA entry into SPF: 30.3 ± 6.4 mm. The zygomaticofrontal suture was 43.4 ± 8.5 mm from SPA entry into the SPF, 58.8 ± 8.0 mm from the sphenomaxillary junction, and 74.9 ± 6.5 mm from the maxillary alveolar process. The SPA to posterior superior alveolar artery (PSAA): 14.4 ± 4.1 mm. Elevation of MA from lateral pterygoid plate: 5.8 ± 2.5 mm.

Discussion: This study characterizes the surgical anatomy of the IFF to better inform LFIII surgical approach. Elevation of MA off the pterygoid plate makes it vulnerable during osteotome advancement. Also, PSAA is at risk for avulsion if the osteotome is advanced along the maxilla. If pursuing a bitemporal approach, the osteotome should be elevated from bony surfaces and advanced 40mm inferiorly beyond the superior border of the zygomatic arch to avoid arterial structures. The gingivobuccal approach offers decreased risk of arterial rupture during osteotome placement, and our data supports limiting advancement to no more than 30 mm beyond the inferior surface of the alveolar process of the maxilla. This research assessed adult cadavers, and further study is needed to address pediatric LFIII cases.



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ANATOMICAL STUDY USING CADAVERS FOR IMAGING OF LIFE-THREATENING COMPLICATIONS IN LE FORT III DISTRACTION

Presenter: Shinsuke Akita, MD, PhD
Authors: Akita S, Mitsukawa N, Hasegawa M, Kubota Y, Satoh K

Chiba Cancer Center

Background: Few reports describe devastating complications with conventional Le Fort III osteotomy; however, life-threatening complications have been reported occasionally with Le Fort III distraction. An anatomical study using cadaveric Le Fort III osteotomy models was performed to investigate the causes of untoward fractures that might induce devastating complications.

Methods: The study sample comprised 30 cadavers (60 sides). Specimens were separated into six groups depending whether osteotomy of the lateral maxillary wall from the inferior orbital fissure to the pterygomaxillary junction (A) and separation of pterygomaxillary junction (B) were performed completely, incompletely, or not performed at all. All osteotomy and fracture lines including the skull base and orbit were examined by computed tomography and direct observation. The separation or fracture type of the pterygoid plate of the sphenoid bone was categorized into four groups, i.e., ideal separation, low-level fracture, high-level fracture, and others. Frequency of each type of pterygoid plate fracture between controls and each group was compared.

Results: High-level fractures occurred more frequently in groups with intact pterygomaxillary junctions. All specimens with untoward fractures of the sphenoid bone leading to the skull base or carotid canal accompanied high-level pterygoid fractures, occurring in groups without sufficient pterygomaxillary separation. An extraordinary orbital fracture was observed when neither A nor B were performed.

Conclusion: Precise separation of the pterygomaxillary junction is primarily of importance preventing devastating complications of Le Fort III osteotomy and Le Fort III distraction. Osteotomy of the lateral maxillary wall is also necessary to minimize this risk.

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BONE TRANSPORT OSTEOGENESIS FOR THE TREATMENT OF LARGE CALVARIAL DEFECTS: AN OVINE MODEL

Presenter: Patrick A. Gerety, MD
Authors: Gerety PA, Wink JD, Sherif R, Clarke NA, Nah HD, Taylor JA

University of Pennsylvania

Purpose: To determine the limits of osteogenesis and distraction kinetics using bone transport osteogenesis (BTO) to treat large cranial defects in a sheep model.

Methods: Defects (3.5 x 3.5 cm) were created in the cranium of sheep and a transport segment (3.5 x 2 cm) traversed the defect at varying distraction rates (0.5, 1.0, 1.5 mm/day) after a 5 day latency. The sheep underwent consolidation for 6 weeks. Analysis included CT, mechanical testing, and histology. CT data was analyzed with Mimics/3Matic (Materialise, Leuven, Belgium). A three-point bending test was performed to assess flexural modulus. The samples were decalcified and histologic sections were stained.

Results: Eleven sheep were used. There were 3 controls and 2 animals each in the different distraction rate groups. After sacrifice, gross examination revealed that control animals had fibrous nonunion while distraction animals had ossified defects. Cross sectional and 3D reconstruction revealed good bone formation in distraction animals and fibrous non-union in controls. Analysis revealed that midline bone thickness varied between native calvarium and bone regenerate ($p < 0.001$). There were not statistically significant differences between the groups. Volume of regenerated bone was also calculated and found to be 2041, 1359, 2029 mm³ respectively ($p = 0.88$). The volume formed was not significantly different between groups. The flexural modulus (MPa) of non-decalcified samples from the control cranium, transport segment, and bone regenerate was found to be 4.50, 6.17, and 4.14 respectively. These values were not significantly different ($p = 0.24$). Histologic sections revealed bone formation in distracted specimens and fibrous non-union in control animals.

Conclusion: This experiment provides proof of concept for BTO in a large animal model. While differences in distraction rate have significant effects on experimental duration they do not appear to affect regenerate quantity/quality. Future plans include use of novel distractors, three-dimensional contouring, complex defects, and enhanced transport segment docking.



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INITIAL EXPERIENCE WITH NEW INTRAORAL MIDFACE DISTRACTOR, (IMD), INDICATIONS AND TECHNIQUE

Presenter: Fernando Burstein, MD

Authors: Burstein F, Berhane C, Schoemann M
Emory University

Distraction of the facial skeleton has gained increasing popularity as a therapeutic technique over the last two decades. Both syndromic and non syndromic individuals may benefit from distraction of the facial bones for functional as well as aesthetic reasons. Mandibular distraction devices have been technically easier to develop and apply to clinical practice compared to midface and orbital distraction devices. Indications for midface distraction include obstructive sleep apnea, severe class III malocclusion, and selected patients with symptomatic proptosis. Although effective external midface distractors have not gained wide patient acceptance due to their cumbersome and visible characteristics. Our study reports on a new push type internal midface distraction device, (IMD), capable of providing up to 30 mm of linear distraction of the maxilla, and if necessary the inferior/medial orbits.

Methods: Twenty consecutive patients, 12 males, 8 females ages six to sixteen, (mean 10.24 yrs), underwent midface distraction with the IMD. Three patients had Cruzons, 12 had unilateral clefts and 5 had bilateral clefts. All had severe class III malocclusion, over 15 mm, symptoms of obstructive sleep apnea and visible deformities. In addition the three Cruzons patients had proptosis with corneal exposure symptoms. All underwent intraoral placement of the IMD device with distraction of 15-30 mm over a period of 10-15 days with a five week consolidation period. Bone morphogenic protein was placed in the osteotomy sites with 5 mm of intraoperative distraction. The lower orbital rim was included in the osteotomy design in the three Cruzons patients.

Results: Mass midface movement was achieved with the IMD in all cases, ranging from 15-30 mm. Obstructive apnea symptoms were improved in all, proptosis was improved in all three Cruzons patients. There were no deaths or major complications. In three patients we failed to achieve the desired 30mm of distraction. In two converging vectors limited distraction to 15 and 18 mm respectively while one would not tolerate turning after 15 mm. There has been no significant relapse to date.

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IMAGE-GUIDED TRANSFACIAL PIN MIDFACE DISTRACTION IN INFANTS RESULTS IN OBSTRUCTIVE SLEEP APNEA RESOLUTION VERIFIED BY POLYSOMNOGRAPHY

Presenter: Robert M. Menard, MD, FACS

Author: Menard RM

Surgical Director Northern California Kaiser Permanente Craniofacial Clinic

In 2001, Pellerin et al. published their series of 4 children treated without pterygomaxillary osteotomies utilizing an external distractor with a transfacial pin. They presented their expanded series of 11 patients at the 2003 ISCFS meeting in Monterey, and published their series of 17 patients in 2012. Given the positive results seen in these publications, we set out to develop a means of performing infant midface distraction using readily available equipment to allow for a more precise means of placing the transfacial pin to optimize results and safety. Our first patient, now with 5 years follow up, was presented at the 2009 ISCFS meeting in Oxford. Precise placement of the 2mm transfacial pin was achieved using the Stryker image-guided surgical navigation system, and the patients midface was advanced a total of 21 mm with a stable result. A total of 4 infants (3 Aperts, one Jackson-Weiss) have been treated with image-guided transfacial pin midface distraction. The two most recent patients underwent formal in-house pre- and post-operative polysomnography, and the results of these studies are presented herewith. Both patients were distracted as previously described, at an average age of 8 months. They were distracted an average of 23 mm. A mean objective improvement, measured as a percentage decrease in pre-intervention to post-intervention values, was seen with a 56% decrease in Apnea-Hypopnea Index and 55% decrease in Respiratory Disturbance Index, down to normal levels in one and near normal levels in the other. Infant midface distraction with a transfacial pin can be performed accurately and safely using readily available distraction devices and navigation systems, avoiding injury to the orbit and teeth, and addressing issues such as orbital exposure and obstructive apnea; five year follow up of our first patient has shown stability of this procedure. Pre- and post-intervention in-house polysomnography demonstrates the efficacy of this procedure towards correcting obstructive sleep apnea in these syndromic infants, and the technique should be considered in these infants with midfacial hypoplasia.



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A SYSTEMATIC REVIEW OF THE EFFECTIVENESS OF MANDIBULAR DISTRACTION IN IMPROVING AIRWAY OBSTRUCTION IN CHILDREN WITH MANDIBULAR HYPOPLASIA

Presenter: Youssef Tahiri, MD

Authors: Tahiri Y, Viesel Mathieu A, Aldekhayel S, Lee J, Gilardino M

McGill University

Background: Distraction Osteogenesis (DO) is an effective technique to elongate the deficient mandible. The purpose of the present study was to specifically evaluate the effectiveness of DO in the treatment of airway compromise in patients with mandibular hypoplasia.

Method: A comprehensive literature review was performed. Inclusion criteria were studies involving isolated distraction of the mandible, English language, and results that included data about airway changes. Selected manuscripts were analyzed with regards to patient demographics, principle diagnosis, distractor type, distraction protocol, pre and post-distraction airway status and complications.

Results: 49 manuscripts met inclusion criteria, resulting in 611 patients with craniofacial malformations who underwent mandibular DO. The mean age of the patients at the time of the procedure was 17.4 months. The most common diagnoses were Pierre Robin sequence (52%), mandibular hypoplasia (11%), Goldenhar Syndrome (6%), and Treacher-Collins Syndrome (6%). In 91.4% of cases, mandibular DO was found to be successful in the treatment of airway obstruction. 148 (88.6%) of the 167 patients who were tracheostomy dependent at the time of the procedure were successfully decannulated. DO was reported to avoid the need for tracheostomy in 270 of 444 (60.8%) patients. Among the patients who had obstructive sleep apnea, only two cases had persistent symptoms post-mandibular DO (96.4% resolved). A 26% overall complication rate was noted. The mean time of follow up was 26.7 months.

Conclusions: In addition to its positive effect on facial appearance, mandibular DO is an effective procedure for the treatment of airway obstruction associated with congenital craniofacial defects involving mandibular hypoplasia.

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ALSO FOR ADULTS WE PREFER DISTRACTION - TREATMENT OF SAOS

Presenter: Alberto Rocha Pereira, MD

Authors: Rocha Pereira A, Matos I, Neves P, Montesuma N, Pires J, Ferreira R, Duarte JM

Portuguese Armed Forces Hospital and Portuguese Oncological Institute

Purpose: Presenting authors clinical experience with 15 cases of adult OSAS successfully treated by multidisciplinary approach using DO, showing important functional and aesthetic results.

Introduction: After two decades, DO has proven that important skeletal advances with good stability can be achieved in facial reconstruction. After appearance of new totally intraoral distraction devices, better tolerated by adult active patients, authors started applying DO in adults with poor adaptation to CPAP. Advantages of this surgical option are: gradual advancement allowing intermediate polissonography during distraction phase, minimally invasive technique and reduced risk of postoperative airway compromise.

Material and Methods: Retrospective clinical study of 15 non-syndromic adults, presenting moderate to severe OSAS, all treated with DO. Ages ranging from 17 to 72, 11 males and 4 female, presented with apparent normal (5), retrognathic (8) or asymmetric (2) faces. Retrolingual hypopharyngeal space ranged from 2 to 7mm and hypopnea-apnea index from 28 to 68. Standard approach was: 1st surgical procedure for mandible angle osteotomies and transbuccal fixation of two internal (intraoral activation) distractors. After classical distraction protocol and consolidation verified, devices were removed and additional case demand procedures were performed (7 patients Le Fort I osteotomy, 4 patients advancement genioplasties, 1 segmental maxillary osteotomy, 1 tonsillectomy). Mean length of distraction was 18mm (12-60mm). Follow-up ranged from 8 month to 4 years.

Results and Conclusions: All patients were successfully treated with this approach, one tracheotomised patient was decannulated, all others discontinued CPAP conservative treatment, having postoperative polissonography with HAI inferior to 10 events, no roncopathy or desaturations. Cephalometric xR showed retrolingual space superior to 11mm in all patients. All patients improved facial aesthetics and functional dental occlusions were obtained with stable skeletal and upper airway space. DO provides an excellent treatment for OSAS also for adult patients.



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NEONATAL MANDIBULAR DISTRACTION IN PIERRE ROBIN SEQUENCE: A VOLUMETRIC ANALYSIS OF THE MANDIBLE AND AIRWAY

Presenter: Patrick C. Hettinger, MD
Authors: Hettinger PC, Deschamps-Braly J, Denny AD

Medical College of Wisconsin/Childrens Hospital of Wisconsin

Background: Mandibular distraction is an accepted method of treatment for airway obstruction associated with Pierre Robin Sequence. The purpose of this study is to review our experience of neonatal mandibular distraction using volumetric analysis along with previously validated methods.

Methods: Following IRB approval, a retrospective chart review was conducted to include all neonates with Pierre Robin Sequence treated with mandibular distraction prior to 6 weeks of age. Pre- and post-operative sleep studies and high resolution CT scans were required for inclusion in this study. CT DICOM data was analyzed with AMIRA imaging software to calculate pre- and post-operative mandible and airway volumes. Results were then compared with age matched controls.

Results: A total of 12 patients met inclusion criteria. Pre-operative CT scans were obtained at an average age of 2.2 weeks, while distraction was performed at an average age of 3 weeks. Distraction distance measured 13.6 mm (range 11-16 mm). Post-operative CT scans were performed at an average age of 35.4 weeks. Pre-operative mandible volume averaged 6.36 cc (range 5.40-7.49 cc) while post-operative volume averaged 18.28 cc (range 12.25-25.15 cc) ($p < 0.005$). Preoperative airway volume measured 1.65 cc (range 1.01 - 2.65 cc), while post-operative airway volume measured 3.22 cc (range 1.70-6.63 cc) ($p=0.005$). When compared with age matched controls, preoperative airway and mandible volumes were significantly smaller in the treated group ($p=0.002$ and $p=0.04$). Post-operatively, there was no statistically significant difference in mandible or airway volume when comparing the two groups ($p=0.21$ and $p=0.07$). All twelve patients were successfully extubated during the activation period. Furthermore, all patients showed improvements in apnea-hypopnea index on sleep study post-operatively.

Conclusions: Mandibular distraction is a proven method of treatment for neonatal airway obstruction in patients with Pierre Robin sequence. In this study, we offer volumetric analysis to quantify both airway and mandible volume changes following mandibular distraction.

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DOES THE JUGULAR FORAMEN INFLUENCE INTRACRANIAL PRESSURE IN CRANIOSYNOSTOSIS PATIENTS?

Presenter: Joyce Florisson, MD
Authors: Florisson J, Lequin M, Van Veelen ML, Bannink N, Mathijssen IM

Erasmus Medical Centre

Introduction: Why craniosynostosis patients develop elevated ICP is still an unsolved mystery. Multiple factors are investigated and a reduced jugular foramen diameter is one of the factors cited in literature. This prospective study aims to test the hypothesis that bony narrowing of the jugular foramen causes elevated intracranial pressure in children with syndromic or complex craniosynostosis. Additionally we investigated the jugular vein and the anatomical variations of the venous drainage system in syndromic craniosynostosis.

Methods: 41 patients with a syndromic or complex craniosynostosis were included and underwent a 3D- CT-Angiography. We describe the anatomic course of the jugular vein, we investigated the diameter and the surface of the jugular foramen and how age influences the diameter of the jugular foramen. Moreover we tested the different collateral patterns and the attendance in having a Chiari I Malformation our patients.

Results: 14.6% of our patients has an aberrant course of the jugular vein. We found that the diameter and the surface did not correlate with papilledema. Neither having occipital collaterals is a predictive factor of developing papilledema. Hence, age seems to be the most influencing parameter. Older children have a larger jugular foramen diameter and do more often have occipital collaterals.



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THE ROLE OF EARLY ENDOSCOPIC RELEASE IN THE MANAGEMENT OF SYNDROMIC AND NON-SYNDROMIC BILATERAL CORONAL CRANIOSYNOSTOSIS

Presenter: Mark R. Proctor, MD

Authors: Proctor MR, Lohani S, Rogers GF, Meara JG
Boston Childrens Hospital

Background: The premature closure of the coronal sutures results in a characteristic malformation of the skull called turricephaly. Conventionally, fronto-orbital advancement has been the keystone of treatment, but repair of the turricephaly remains problematic. More recently, surgeons have realized that early surgery can minimize the progression of turricephaly, and various early techniques using distractors or springs have been advocated, even preferentially advancing the occiput. In this paper we report our results of early endoscopic coronal suturectomy with post-operative helmet therapy as a treatment option in these children.

Methods: This is a retrospective study of all patients with bicoronal synostosis that underwent endoscopic release and helmet therapy between January 2005 and December 2012 at our institution. A preoperative CT scan was performed in all patients. Endoscopic suturectomy was performed between the age of one to four months. Cranial remodeling helmets were fitted within a week of surgery and continued for 6-9 months.

Results: There were 18 patients; eight males and 10 females. Eight patients were syndromic: three Saethre-Chotzen, two Apert, two Muenke, and one Pfeiffer syndromes. Mean age at surgery was 2.5 months. Duration of surgery was 73.3 minutes. Mean blood loss was 40 cc (range 20 to 100 cc). Two patients needed blood transfusion. Mean duration of hospital stay was 1.2 days (range 1-2 days). One child needed reoperation for CSF leak. Mean follow up duration was 18.3 months. Mean cranial index prior to surgery was 0.94 which improved to 0.84 post-operatively, and mean head circumference percentile went from 31% preoperatively to 46% postoperatively. Turricephaly improved in all patients. Three patients have needed secondary fronto-orbital advancement due to sutural re-fusion.

Discussion: Early endoscopic suturectomy with post-operative helmet therapy is an excellent treatment alternative to avoid secondary turricephaly, and should be considered as an alternative early release procedure.

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SYNDROMIC CRANIOSYNOSTOSIS AND CHIARI TYPE I MALFORMATION: A VOLUMETRIC STUDY OF THE POSTERIOR FOSSA. DOES SIZE MATTER?

Presenter: Bianca Rijken, MD

Authors: Rijken B, Lequin MH, van der Lijn F, van Veelen-Vincent ML, Niessen W, Mathijssen IM

Erasmus Medical Center University

Introduction: Patients with craniosynostosis syndromes are at risk for increased intra cranial pressure, ventriculomegaly and Chiari malformation type I (CMI). It remains unclear whether or not the presence of CMI is related to a reduced volume of the posterior fossa (PF) and/or to an abnormal volume of the cerebellum. Therefore, we investigated cerebellar and PF volumes as well as their volume ratio and related that to the position of the cerebellar tonsils.

Methods: In 113 patients (mean= 7.2 years, range: 0-18 years) cerebellar and PF volumes were measured in 3D SPGR T1-weighted MR scans. Control data were collected from MR scans of 34 patients without any congenital disorder or brain anomaly (mean= 4.8 years, range: 0-15 years). Differences in volumes between patients and controls were tested after correction for age and gender.

Results: Patients with Apert and Muenke syndrome develop a larger PF within the first few years of life compared to controls (181ml (SD:47) and 183ml (SD:46) vs. 153ml (SD:44), $p < 0.01$), while Muenke patients also develop a larger cerebellum (135ml (SD:31) vs. 115ml (SD:34), $p < 0.01$). The volumes of PF and cerebellum were not significantly different between patients with other craniosynostosis syndromes and controls. However, the ratio cerebellum/ PF was significantly higher in patients with a CMI than in those without a herniation (0.78 versus 0.73 $p < 0.001$), even though their cerebellar volume and PF volume did not differ from controls. A close relation was noticed between the presence of papilledema and CMI (40% vs. 11%, $p = 0.015$).

Conclusions: Craniosynostosis patients with CMI have a significantly higher ratio between cerebellar and PF volumes. Additional factors to this intrinsic anomaly appear to be essential to the development of a CMI or not. Such a factor might be elevated ICP since a close relation is found between the presence of papilledema and CMI. Longitudinal data are necessary to determine whether elevated ICP induces CMI or whether CMI causes elevated ICP by obstructing outflow of blood and CSF.



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POSTERIOR CRANIAL VAULT DISTRACTION IN THE MANAGEMENT OF SYNDROMIC MULTI-SUTURE CRANIOSYNOSTOSIS: OUTCOMES AND 3D PHOTOGRAPHIC/CT-BASED MORPHOMETRIC ANALYSIS

Presenter: Brooke French, MD

Authors: French B, Clausen A, Forrest CR

The Hospital for Sick Children

Purpose: Posterior cranial vault distraction has been popularized as a first intervention technique in the management of syndromic multi-suture craniosynostosis. The purpose of this study was to present our experience with posterior cranial vault distraction in the management of these complex patients.

Methods: 17 patients (7M, 10F; age 4 months to 149 months) with syndromic craniosynostosis (3 Apert, 3 Pfeiffer, 2 chromosomal anomaly, 2 Muenke, 2 Crouzon, 2 combined sagittal/lambdoid, 3 unknown) underwent posterior craniotomy with application of KLS-Martin distractors for correction of elevated intracranial pressure (n=9) and/or correction of turribrachycephaly (n=15). Distraction (0.5 mm bid) commenced on the first post-operative day and consolidation period was 8 to 10 weeks (follow-up 8 to 68 months). CT-based intracranial volumes and sagittal ray analysis pre- and post-distraction were analyzed with GE Healthcare AW VolumeShare2 software.

Results: Posterior cranial vault distraction was successful in achieving normocephaly in 8 patients. Two patients underwent mono-bloc fronto-facial advancement and 6 patients underwent subsequent planned fronto-orbital advancement. Elevated ICP was resolved in 5/6 patients. Two patients required VP shunt for persistent CSF leak. Complications included 1 technical failure requiring redo surgery, 2 skin breakdowns overlying the distractor, 1 infection and 3 CSF leaks. Mean intracranial volume increase was 34% (range 24-42%). Sagittal ray analysis demonstrated mean reduction on vertical height of the cranium of 7.7% based on a standard cranial index of vertical height over transverse width.

Conclusions: Posterior cranial vault distraction provides a controlled expansion of the brachycephalic cranium with correction of turribrachycephaly and resolution of intracranial hypertension in select cases. Disadvantages include removal of the distractors and cost of hardware outweighed by the advantages of shorter procedure time, less blood loss, no need to remove bone over the confluence of the venous sinuses and spontaneous remodeling of the forehead.

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CHARACTERISTICS OF POSTNATAL PROGRESSIVE PANSYNOSTOSIS

Presenter: Albert K. Oh, MD

Authors: Oh AK, Sauerhammer TM, Magge KT, Magge SN, Myseros JS, Keating RK, Rogers GF

Childrens National Medical Center

Background: Postnatal progressive pansynostosis (PPP) is uncommon. The insidious onset and relatively normal cranial shape often leads to delay in or failure of diagnosis. The risk of increased intracranial pressure (ICP) is significant. The purpose of this study was to review our series of patients with PPP to provide a more detailed description of this form of synostosis.

Methods: The authors reviewed their ongoing prospective craniofacial database (2/97-7/12) to document patients with PPP. Variables included age at diagnosis, syndromic association, head shape, serial head circumference (HC), radiographic findings, and operative interventions. Inclusion criteria included: 1) complete or partial fusion of all major cranial sutures on high resolution CT scan; 2) no prior craniofacial procedures at the time of CT diagnosis.

Results: A total of 10 out of 292 patients (3.5%) were identified. Each patient had confirmed fusion of all major cranial sutures. Mean age at diagnosis was 59.4 ± 42.3 months (range: 19-151 months). A syndrome was documented in 60% of patients, with Crouzon syndrome being most common (3 patients). Head shape was reported as normal or near normal in all patients. Serial HC, available in 90% of patients, documented sustained growth deceleration at the time of diagnosis. All patients had signs of increased ICP, such as severe endocortical erosion and loss of cisternal spaces on CT; 6/7 patients with MRI had Chiari malformation. ICP monitoring in 4 patients revealed high pressures ranging from 5 to >35 cm H₂O. Calvarial expansion was performed in 70% of patients, two are awaiting definitive surgical repair, and one patient had only Chiari decompression and clinical monitoring.

Conclusion: The gradual progression of fusion in PPP results in a slow decrease in HC percentile and a relatively normal head shape, leading to delayed diagnosis and increased ICP. There should be a high clinical index of suspicion in children with a known craniosynostosis syndrome (especially Crouzon syndrome) and normal head shape. Any sustained deceleration in serial HC should prompt CT evaluation.



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ENHANCING THE PROCESS OF COMPLEX CRANIOFACIAL RECONSTRUCTION: LESSONS LEARNED IN 94 CASES USING PREOPERATIVE COMPUTER MODELING

Presenter: Patrick Kelley, MD

Authors: Kelley P, Harshbarger RJ, Henry SL, Combs PD

Dell Childrens Medical Center of Central Texas

Computed tomography (CT) is essential in providing detailed anatomical data to enable complex diagnosis and treatment planning. Yet, heretofore, the utility of this information in the actual execution of reconstruction was limited to the artistic imagination of the surgeon. Recently, advances in computer-aided design software (CAD) have enabled the surgeon to translate the data obtained from CT into workable images.

The integration of scanned dental models with CT images and registration with the natural head position generates the perfected craniofacial image (PCI). Virtual osteotomies create units that can be manipulated to simulate surgical maneuvers whereby the surgeon can view possible interferences, relationships to other structures, soft tissue response, and changes relative to orthognathic standards. "Mirror imaging" and "cohort substitution" is used to create structures de novo, thereby providing a template image that guides movements or de novo creation of structures. Surgical cutting guides, reconstruction plate templates, oclusal splints, and positioning guides fabricated from stereolithography facilitate translation and execution of the digital plan to the patient.

Over the past six years, we have completed 94 craniofacial cases that utilized preoperative computer modeling, employing traditional osteotomies, distraction, grafting, and free tissue transfer. We have perceived the following advantages: (1) enhanced accuracy of the reconstruction (as documented by congruence with postoperative imaging); (2) improved time to union and completeness of union (due to improved precision of osteotomies and hence bone-to-bone apposition); (3) reduced operative time (secondary to the elimination of freehand whittling to shape the bone segments); (4) reduced external scar burden (as the precise precutting of the bone segments better permits their alignment and fixation through intraoral approaches); and (5) qualitative improvements in aesthetic outcome (documented by post-operative photography). In this presentation we will demonstrate these advantages and discuss the lessons learned through our series.

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ABNORMAL GROWTH OF THE FORAMEN MAGNUM IN CROUZON SYNDROME: SIZE AND CLOSING OF THE INTRA-OCCIPITAL SYNCHONDROSES

Presenter: Caroline Driessen, MD

Authors: Rijken B, Driessen C, Lequin MH, van Veelen-Vincent ML, Mathijssen IM

Erasmus Medical Center University

Introduction: Patients with Crouzon have prematurely closed skull vault and skull base sutures. Therefore these patients have a high risk for developing hydrocephalus, disturbances in cerebrospinal fluid or venous outflow and Chiari malformation type I (CMI). In this study we investigated the size of the foramen magnum (FM) as well as the closure of posterior and anterior intra-occipital synchondroses (PIOS and AIOS), which are possibly involved in common problems of Crouzon patients.

Methods: The area and anterior-posterior (AP) diameter were measured in 3D-CT scans of 29 Crouzon patients (mean age: 4.6 years) and 29 age-matched trauma-patients (mean age: 4.8 years). Moreover, synchondroses were graded by Madeline and Elster Grading system (Madeline and Elster 1995). Size differences were tested with the independent sample t-test, while ordinal logistic regression analysis was performed for studying the closure of synchondroses.

Results: Crouzon patients have a smaller FM than controls (area of 598 mm vs. 768 mm and AP-diameter of 31 mm vs. 35 mm, $p < 0.001$).

PIOS close earlier than AIOS in both groups, while they did not close earlier in Crouzon patients than in controls ($p = 0.062$ and $p = 0.787$, respectively).

Conclusions: Patients with Crouzon syndrome have a smaller FM than controls; this difference is already present at birth. Surprisingly, we could not confirm that intra-occipital synchondroses in Crouzon patients close earlier than those in controls.



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**CLOSURE OF THE SPHENO-OCCIPITAL
SYNCHONDROSIS IN PATIENTS WITH SYNDROMIC
CRANIOSYNOSTOSIS: A LIKE TO MIDFACE
HYPOPLASIA**

Presenter: Jesse A. Taylor, MD

Authors: Goldstein JA, Paliga JT, Bartlett SP, Taylor JA
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Background: The spheeno-occipital synchondrosis (SOS) is important for facial and cranial base growth, and its premature fusion is implicated in midface hypoplasia in animals. The current study characterizes SOS in patients with Crouzon, Apert and Pfeiffer syndrome and correlates early fusion with the presence of midface hypoplasia.

Methods: A case-control study was performed of Crouzon, Apert, and Pfeiffer patients. CT scans performed as part of normal care were analyzed for status of SOS patency, and matched control CTs identified from trauma patients were also assessed. CTs were categorized with either open, partially fused, or completely fused SOS. Midface hypoplasia determined by SNA measurement prior to midface surgery was correlated with SOS status. McNemars test for matched pairs and linear regression were used to assess significance.

Results: During the study period, 54 patients with 206 CT scans were compared with 206 age/gender matched control scans. Average age at CT scan was 6.1 years. The earliest age of partial and complete fusion was 1.1 and 7.0 years, among cases and 6.2 and 12.7 among controls. In 67 case-control pairs, there was discordance between patency and fusion with the odds of SOS fusion in cases of 66.0 (95%CI 9.2-475.5, $p < 0.000001$) times that of controls. 45 patients underwent cephalometric analysis with an average SNA angle of 68 (range: 58-80, $p < 0.0001$) degrees. In the 22 cases who initially demonstrated open SOS, average age of initial SOS closure was 3.5 (range: 0.5-6.0) years. In this group, average SNA at time of midface surgery was 67.5 (range: 58-76) degrees. Linear regression analysis demonstrated positive correlation with earlier age of first SOS closure and midface hypoplasia ($r = 0.48$, $p = 0.028$).

Conclusion: The SOS closes significantly earlier in patients with syndromic craniosynostosis compared with age-matched controls. There is positive correlation between earlier evidence of SOS closure and degree of midface hypoplasia, though definitive causality cannot be concluded by this methodology.

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**INTRACRANIAL PLATING WITH RESORBABLE PLATES
FOR CRANIOFACIAL RECONSTRUCTION**

Presenter: Peter T. Wang, MD, DMD

Authors: Wang PT, Gennuso R, Megahead H,
Garcia CA, Levine RA

Pediatrics

Resorbable plates have become the standard of care over the last ten years for cranial vault reconstruction in the pediatric patient. Currently, the majority of plates are placed on the outside of bone relying on resorption of the plate in one to two years. During this period, the plate maybe palpable and occasionally cause bony overgrowth at the nasofrontal region. Further, plate resorption may leave an indentation on the bony surface. We propose that placing the resorbable plates on the inner surface of the bone flaps can safely overcome these current limitations while maintaining the desired results.

We describe our experience with intracranial plating for cranial vault reconstruction performed over the last 5 years at Methodist Children's Hospital in San Antonio, Texas.

All patients undergoing intracranial plating for cranial vault reconstruction since 2008 were identified and records reviewed. Data evaluated included diagnosis, age at time of surgery, surgical technique, pre-operative & post-operative cranial proportions, patient/family satisfaction, and complications. A minimum of 6 months follow up was required for inclusion. Post-op CT's were obtained annually from the time of surgery until plate resorption and boney closure noted. Resorbable plates were placed internally for the supraorbital ridge, forehead, and vault reconstruction in 42 patients. There were 13 sagittal, 10 metopic, 7 unicoronal, 2 bi-coronal, 3 lambdoid, 4 Crouzon's, and 3 multi-suture synostoses. Patient's age ranged from 4 to 16 months, mean (8). Average follow up is 2 years, with a minimum of 6 months. There was one scalp infection due to bony prominence on the incision and tight closure. The forehead and vault contours were smooth in all patients and family satisfaction is high. Patients whose follow up was longer than 2.5 years all had resorption of their plates demonstrated by CT's and no granulomatous or intracranial problems noted. Intracranial plating is a safe and successful technique for craniofacial reconstruction, which can overcome some of the limitations of resorbable plating systems.



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**PSEUDOMENINGOCELE WITH ORBITAL
EXTENSION AS A COMPLICATION OF FRONTO-
ORBITAL ADVANCEMENT AND REMODELLING IN
CRANIOSYNOSTOSIS: A RETROSPECTIVE REVIEW**

Presenter: Ajay Sinha, MD
Authors: Vaiude P, Sinha A, Burn SC, Richardson D,
Sweeney E, Duncan C

Alder Hey Hospital for Children

Aim: We present a series of patients who developed a pseudomeningocele following fronto-orbital advancement and remodelling (FOAR), describing clinical presentation, investigation and management. Risk factors are identified and preventative strategies suggested.

Materials & Methods: All patients who developed a pseudomeningocele post-FOAR from 2002-2012 at our centre were identified and studied.

Results: 236 FOAR operations were carried out over 12 consecutive years. 61 of these patients were syndromic. Pseudomeningocele occurred in 6 patients all of whom were syndromic. Of affected patients - 4 had raised intracranial pressure (ICP) pre-operatively, 4 had a recognised dural tear at FOAR and 4 had an infection post-surgery. Clinically they presented with orbital swelling, ptosis and proptosis. Details of management will be presented. Decompression of the pseudomeningocele with excision and duraplasty was carried out in all 6 patients. 4 patients had a calvarial graft cranioplasty and a further 2 had a titanium mesh. 1 patient developed epilepsy. No other ocular, aesthetic or recurrence related complication was noted.

Conclusion: Pseudomeningocele has not previously been described in FOAR as a large series of consecutive patients. We have identified a 2.5% overall incidence which increases to 10% in the syndromic population. The risk factors include syndromic craniosynostosis, dural tear, hydrocephalus or raised ICP, infection, persistent CSF leak or presence of dead space. Preventative strategies include CSF management prior to or post-FOAR. The ultimate treatment of the pseudomeningocele and growing fracture involves surgical decompression of the collection, duraplasty, reconstruction of the orbital roof and temporary CSF diversion. Diagnosis and treatment of a pseudomeningocele should be prompt to prevent long term complications.

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**IDENTIFYING WHICH MORPHOLOGICAL
ABNORMALITIES OF APERT SYNDROME ARE
CORRECTED BY BIPARTITION DISTRACTION**

Presenter: Nicki Bustrzonowski, MD
Authors: Ponniah A, Bustrzonowski N, Verdoorn M,
Nikkhah D, Ruff C, Dunaway D

Great Ormond Street Hospital

Background: Apert syndrome is a congenital disorder characterized by craniosynostosis and midface hypoplasia. This study looks to identify to what extent bipartition distraction corrects the morphological abnormalities of this condition.

Methods: Pre and post - operative three dimensional computed tomography (3DCT) scans of 10 patients with Apert Syndrome, (12-21 years) were identified from the Great Ormond Street Hospital database. 98 landmarks were used to analyse pre and post-operative scans and 13 normal skulls.

Principal component analysis (PCA) was used to analyse patterns in the datasets. Within each group, eigenvectors were identified that demonstrated the aspects of the skull where the most variation was found. The analysis allowed both global shape measurement and local proportions.

Results: Inter and intra reliability of landmark positioning was within 1mm for the majority of the landmarks. Post-operative and normal scans both showed close agreement in the first three principal components of variation, demonstrating a good global normalisation of the face by bipartition distraction. Warping from pre- to post-operative illustrates midface advancement and inward rotation of the orbits. Post-operative to normal warps demonstrate that bipartition distraction effectively advances the midface and corrects orbital position but fails to adequately correct interzygomatic width and midfacial height.

Conclusion: This study allows us to understand the way bipartition distraction corrects the abnormalities of the Apert skull. Bipartition distraction significantly improves Apert craniofacial morphology, but is unable to fully harmonise facial width or height.



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CHILDREN WITH CROUZON SYNDROME AS ADULTS. A FOLLOW-UP STUDY OF 31 SWEDISH PATIENTS

Presenter: Lars Kolby, PhD

Authors: Kolby L, Fischer S, Tovetjarn R, Maltese G, Sahlin PE, Tarnow P

Institute for Clinical Sciences

Background: Crouzon syndrome presents with craniosynostosis, maxillary hypoplasia and exophthalmus. In addition the neuropsychological development is often hampered resulting in mild to moderate reduction of intelligence and social short-comings. The aim of the present study was to evaluate the life situation for adult patients with Crouzon syndrome.

Patients & Methods: Forty patients with Crouzon syndrome born before 1990 could be identified in the Goteborg Craniofacial Registry. A questionnaire dealing with education, employment, social relations, and quality of life was used. A matched control group was created for comparison. Logistic regression, correcting for the influence of age and sex, was used to compare patients and controls.

Results: Thirty-one (11 female, 20 male, mean age 35 years) of the patients answered the questionnaire. The control group consisted of 285 persons. The level of education was lower in patients than in controls ($p < 0.015$) but there was no significant difference in the extent of employment between the two groups. Patients were less often married or had a partner ($p = 0.059$), had fewer children of their own ($p = 0.004$), and had less experience of a sexual relationship ($p < 0.001$). The difference in housing was not significant and only one patient lived in a care centre and three patients required a personal assistant to manage activities of daily living. The patients' estimation of their somatic health was equal to that of controls but patients more often used anti-epileptic medication ($p = 0.003$). Periods of depressive mood were more common in patients ($p = 0.001$), but there was no difference between the groups regarding a general positive attitude to life.

Conclusion: Patients with Crouzon syndrome often have intellectual and social short-comings that negatively effect their lives. However, the range of abilities is wide in this group with some patients in need of substantial supportive efforts whereas others have high education, work full time and have a flourishing social life.

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EFFECT OF MONOBLOC FRONTO-FACIAL BIPARTITION DISTRACTION ON ORBITAL VOLUME, MORPHOLOGY, AND CLINICAL OUTCOME IN 15 APERT SYNDROME CASES: A CONTROLLED STUDY

Presenter: Roman Khonsari, MD

Authors: Khonsari R, Karunakaran T, Way B, Matthews W, Nysjo J, Nystrom I, Dunaway DJ, Evans RD, Britto JA

Great Ormond Street Hospital

Frontofacial bipartition and advancement by distraction (BpD) is a valuable strategy in the management of Apert syndrome (AS) for functional and aesthetic gain. We ask whether BpD achieves a stable resolution of oculo-orbital disproportion in AS.

Methods: CT radiologic data of 15 Apert (AS) patients (30 orbits, R=2-21yr) were assessed against similar data from 40 age-matched control scans from a neurosurgical population, and 18 patients with different forms of unoperated hypertelorism. Globe volume, orbital volume, orbital morphology and globe position were obtained by manual segmentation with inter- and intra- observer control, verified by a mesh-based semi-automatic technique (Nystrom et al). Palpebral fissure width (PFW), intercanthal distance (ICD), and inter-dacryon distance (IDD) were measured. Patient data included pre-operative, six-week post-BpD, and delayed postoperative (8-18 mnth) scans, correlated with contemporaneous ophthalmic examination findings.

Results: Mean AS orbital volume is 22.9cc (11-21 yrs), excluding two 23 and 28 month old patients at 11cc and 20cc; and therefore matches controls, in which orbital volume rises from 14-21cc aged 2-10 yrs and plateaus at 22cc aged >10yrs. Control globe and orbit volumes are symmetrical, whereas AS orbits show greater symmetry of volume than shape. BpD normalises globe protrusion in AS, which preoperatively significantly exceeds control value (which is symmetrical, constant, and independent of age). The ratios ICD:PFW and IDD:PFW are consistent in control (mean 1.16 and 0.64), whereas AS demonstrates higher ratios (1.37 and 0.76) approaching those of hypertelorid patients (1.89 and 1.41). Longitudinal surface rendering studies indicate that BpD provides a stable augmentation of the medial orbital wall in AS and volume expansion compared to control.

Discussion: BpD is a valuable strategy to resolve oculo-orbital disproportion in AS, addressing medial volume and shape insufficiency. BpD allows "unbending" of the midface and addresses the tendency in AS towards a hypertelorid phenotype.



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**COMPLICATIONS AFTER FRONTOFACIAL
MONOBLOC ADVANCEMENT WITH INTERNAL
QUADRUPLE DISTRACTION IN CHILDREN**

Presenter: Eric Arnaud, MD
Authors: Arnaud E, Saiepour D, Leikola J, Wyten R,
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Craniofacial Unit Hospital Necker

Even reduced by distraction, some various risks still exist for monobloc advancement.

Patients: This study prospectively evaluated the complications in a cohort of 105 faciocraniosynostotic children treated with a frontofacial monobloc advancement with internal distraction. Mean age at surgery was 3.8 years. There were 65 Crouzon, 21 Apert, 18 Pfeiffer and 1 Sprintzen-Goldberg syndrome. Maximum follow-up was 12 years.

Results: At the beginning of the experience, one patient died a few hours after an uneventful surgery from a misdiagnosed tonsillar herniation. Since that, all patients were operated on of FM decompression and/or posterior expansion in case of Chiari malformation. No further lethal complications was encountered.

Suspicion of CSF leakage through the nose was evoked in 37/104 patients but was confirmed in only half of the cases after lumbar puncture during 3 days was proved unsuccessful. Transient lumbar drains were implanted to stop the leakage. In two patients (in the group without periosteal flaps) a persistent leakage in the posterior rhinopharynx was treated endoscopically. The infection of at least one distractor was found frequently (15 cases) but only 8/104 necessitated some kind of reoperation for removal of one distractor. The osteonecrosis of the frontal bone was found in 4 patients, three of them being secondary cases, one of them being a patient in which traction had been simultaneously applied with a rapid distraction and traction. One partial frontal brain atrophy in one of the patient who lost the forehead and in whom the cranioplasty repair was delayed after one year. Two transient reduction of vision on one eye during the course of distraction, recovered after contraction. A tendency to trismus occurred during distraction and necessitated active reeducation.

Discussion: Chiari malformation should be screened and treated with posterior surgery prior to monobloc. The reduction of morbidity was confirmed in terms of reduction of infection but secondary patients and rapid distraction were were risk factors of osteonecrosis. Vision problems were transient if immediately detected.

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**WHERE ARE THEY NOW?: GEOGRAPHICAL
AND SOCIAL OUTCOMES ON 102 ADULTS WITH
CRANIOFACIAL SYNDROMES**

Presenter: Stephen Dover, FDSRCS, FRCS
Authors: Dover S, Venugopal A, Noons P, Evans M,
White N, Solanki G, Rodrigues D,
Nishikawa H

Birmingham Childrens Hospital UK

The surgical management of patients with syndromic craniosynostosis has changed little over the past 40 years. More recently, there has been greater emphasis on the importance of psychological assessment, speech and language input and educational support. Ultimately, all healthcare efforts are with a view to maximising a child's potential and equipping them as best as possible for adult life. So how well are our adult patients doing?

103 adult patients with syndromic craniosynostosis have been identified from the department database. Many had been discharged from clinical care and some had been lost to follow up. The purpose of this paper was to find out what had happened to this adult cohort and how they had progressed in life.

The cohort included 50 male and 53 female patients with the following diagnoses, Crouzon's n=55, Apert's n = 19, Saethre-Chotzen n=21, Muenke n=4, Pfeiffers n=2, Carpenter's n=1. Only patients with confirmed diagnoses were included in the study.

Details of our more recent patients were most accurate and these patients were easiest to track. Overall, 41 adults were in full time or part time higher education, 5 were at home with children and 4 worked in manual trades. Only 2 were unemployed. 31 patients could not be contacted, but efforts to do so will continue. The remainder were in a wide variety of roles and occupations.

If successful craniofacial management could be measured only by employment or educational outcomes in adult life, then this initial and on-going work would appear to suggest that we are succeeding. However, this does not address how these patients feel about themselves and their place in the community. This is an area of our research work that is on-going.



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PREOPERATIVE TREATMENT WITH MOLDING HELMET IMPROVES CEPHALIC INDEX IN BICORONAL SYNOSTOSIS

Presenter: Arlene Rozzelle, MD
Authors: Rozzelle A, Sood S, Sofer JT
Childrens Hospital of Michigan

Infants with bicoronal synostosis have brachycephaly with a cephalic index (CI) (ratio of width to length of the calvarium) above the normal range of .75 to .85. Traditionally, surgical correction includes the advancement of the frontal-orbital area during infancy without addressing the posterior fossa constriction.

Based on our success using preoperative helmet to improve the CI in sagittal synostosis, we have used preoperative helmet to improve the CI in bicoronal synostosis.

Methods: A chart review was carried out (2010-2012) of infants with bicoronal synostosis. CI was measured on presentation in infancy and at every visit, including the one immediately prior to surgery. Molding helmet was initiated at 2-3 months of age and surgery typically carried out between 6 and 12 months of age. CI at presentation was compared in the helmet treated and non-helmet treated groups and immediately prior to surgery.

Results: Six infants were identified with bicoronal synostosis. Three infants had preoperative treatment with the molding helmet and three did not have preoperative treatment with the molding helmet. CI at presentation was .95 in the helmet group and .96 in the non-helmet group. CI immediately pre-op was .90 in the helmet group and .97 in the non-helmet group. Thus, the helmet group CI was improved both compared to the CI at presentation and to the non-helmet group immediate pre-op. There was no change in the non-helmet group CI between presentation and immediate pre-op. Long term, CI in the helmet group (.86) continued to be better than the non-helmet group (.92).

Conclusion: Preoperative treatment with a molding helmet in infants with brachycephaly due to bicoronal synostosis is effective in improving the head shape as demonstrated by improved cephalic index compared to infants who did not have a preoperative helmet. The use of the molding helmet preoperatively during the rapid growth phase in infancy should give a better long-term result with less posterior fossa constriction.

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FRONTO-ORBITAL EXPANSION AS KEY PROCEDURE IN CORRECTION OF CRANIOSYNOSTOSIS

Presenter: Xiongzhen Mu, MD
Authors: Mu X, Yang JY, Bao N, Guo ZL
Huashan Hospital Fudan University

Purpose: To point out the key procedure in correction of craniosynostosis.

Method: Review of 45 cases in craniosynostosis correction in this list. There are 8 cases of secondary craniosynostosis which only be performed with molding craniotomies in other centers. Each case in our experience is employed fronto-orbital advancement with various kinds of remodeling craniotomies. Pre and post operative 3D CT scans were collected as the data of analysis. At least 12 months Followup is in this list.

Result: In 8 cases of secondary craniosynostosis, the reunion of cranial sutures is still found after craniotomies without fronto-orbital advancement. The remodeling craniotomies with fronto-orbital advancement were employed in our 45 cases including 8 cases of secondary craniosynostosis. There are no sign of reunion of cranial suture have been found in followup list.



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ADVERSE EVENTS ASSOCIATED WITH THE USE OF SPRINGS FOR MOVING CRANIAL BONE

Presenter: Roisin McNicholas, MD

Authors: Jeelani O, Hayward R, McNicholas R
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Introduction: The use of springs in craniofacial surgery was pioneered by Lauritzen in Gottenberg. We have found that they provide an efficient way of moving cranial bone for distances that were unachievable using previous methods that relied on holding mobilised bone stable by per-operative fixation – something of particular use in vault expansion procedures in which the amount of bone movement is severely restricted by the lack of immediate elasticity of the scalp. The purpose of this communication is to audit in more detail the incidence and clinical importance of those adverse events that can be attributed to the use of springs and point to ways in which their technology can where possible be modified to improve their effectiveness.

Methods: All spring cases performed over the first 5 years of use were analysed using a comprehensive contemporaneously collated surgical database.

Results: 1. Between 2008 and 2013 a total of 501 springs were inserted in 173 patients.

2. Their diagnoses/indications for operation were a. Sagittal synostosis - 77 OR 45% of patient group b. Posterior vault expansion – 75 OR 43% of patient group c. Other – 23 – OR 13% of patient group.

3. The average number of springs per patient has gone down steadily from 6 per patient in 2008 to 2 per patient since 2011, reflecting the learning curve.

4. Mean hospital stay of 2 nights.

5. 38% of patients require a blood transfusion (the average red cell replacement was 40%).

6. Mean surgical time 75mins.

7. 1 patient had a Grade 3 adverse event, 13% had a Grade 2 and 11% a Grade 1 adverse event. (see appendix)

8. 14% required an early removal of one/more springs

9. “Morbidity” due to aberrant/undesired bone movement by the springs was seen in 9% (n=17) cases. 53% of the cases were Sagittals, 35% PVE & 12%.

Other Discussion: This study confirms that the use of springs as a less invasive method for moving cranial bone with comparable morbidity compared with traditional techniques.

Conclusion: Springs are a definite spring forward but “misdirection” of bone movement can be an issue. Use as few as possible in each case.

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RADIOLOGICAL EVALUATION AFTER FRONTOFACIAL MONOBLOC ADVANCEMENT WITH INTERNAL QUADRUPLE DISTRACTION IN CHILDREN

Presenter: Daniel Saiepour, MD, PhD

Authors: Saiepour D, Wyten R, Leikola J, Meyer PH, Oyama A, Di Rocco F, Arnaud E

Centre de Reference de Malformation Cranio Facialis

In this study, the long-term radiological results of frontofacial monobloc advancement in children with faciocraniosynostosis were evaluated.

Patients and Methods: We prospectively evaluated the ossification of the anterior fossa, the frontal bone and the expansion of the frontal lobe in 105 patients with syndromic craniosynostosis after frontofacial monobloc advancement. All patients were operated at Hospital Necker-EnfantsMalades, Paris, France between 2000 and 2011. The group consisted of 65 patients with Crouzon, 21 patients with Apert, 18 patients with Pfeiffer and one patient with Shprintzen-Goldberg syndrome. In all patients, a frontal bone flap was separated anteriorly. A bilateral pedicled periosteal flap was raised and sutured to seal the anterior fossa. Four internal distractors were placed and the distraction rate was 0,5 mm/day. The distractors were removed after a mean of 6 months consolidation period. The long-term radiological evaluation was done at least one year after the operation (mean 5 years, range 1-11 years) using 1 mm CT scans.

The frontal lobe expansion was evaluated using postoperative CT-scans and measuring the distance between the frontal bone and the frontal lobe.

Results: Complete frontal lobe expansion was observed in all patients but 5 (95%). Four patients (3.8%) had an infection of the frontal bone and underwent surgical removal of the infected bone followed by cranioplasty. Further the ossification of the osteotomy gaps in the anterior fossa and the lateral aspect of the frontal bone were evaluated. Eighty-four percent of patients had “good” or “fair” ossification of the anterior fossa whereas 58 percent had good or fair ossification of the frontal bone.

Conclusions: In this long-term radiological evaluation of frontofacial monobloc advancement we could observe full expansion of the frontal lobe in the majority of the cases, despite the frontal bone flap. The anterior fossa bone seems to consolidate better than the frontal gaps in the coronal region. The periosteal flap could partly explain the enhanced ossification of the anterior fossa that was observed in this study.



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CENTRALIZED CARE FOR CRANIOSYNOSTOSIS

Presenter: Irene Mathijssen, MD, PhD

Author: Mathijssen I

Erasmus MC

The Netherlands has a population of 16 million people and yearly approximately 120 new patients with craniosynostosis are born. The care for children with craniosynostosis was given in 5 university centers, with a large spread in treated numbers ranging from less than 5 to 100 a year. As we felt the need for more centralized care, the initiative was undertaken to develop an evidence based guideline on craniosynostosis. Members of all 5 teams were invited to participate in this project. In the guideline we defined which type of care should be provided at what ages, and how team management should be organized. In the end there was consensus to recommend that only 2 centers would provide this care; 1 center for both syndromic and non-syndromic cases and 1 center for non-syndromic only. This guideline was then sent to all the boards of the involved scientific societies for approval, which was obtained.

Next, an independent audit team was established with the participation of a neurosurgeon, a plastic surgeon, a maxillofacial surgeon and the president of the patients society. They asked all teams to fill out a questionnaire on several aspects regarding care. Based on these results, they gave the recommendation on which 2 teams should continue their activities. Although the 3 centers which were not assigned were disappointed, the decision was made in an open way and based on agreed upon criteria.

The Netherlands is now one of a few countries in which care for craniofacial malformations is restricted to designated centers, next to Great Britain and Sweden.

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SPRING-ASSISTED POSTERIOR EXPANSION AND SIMULTANEOUS FRONTO-ORBITARY EXPANSION IN SYNDROMIC CRANIOSYNOSTOSIS

Presenter: Javier Gonzalez Ramos, MD

Authors: Rodriguez JC, Gonzalez Ramos J,

Routaboule C, Zuccaro G

Garrahan Pediatric Hospital

Syndromic craniosynostosis is a difficult-to-resolve congenital anomaly generally requiring several surgical procedures. Expansion of the posterior cranial vault diminishes intracranial hypertension and its deleterious effect on the brain. The use of spring-assisted distraction for posterior vault expansion showed to be an effective and stable method with lower morbidity than that seen in traditional procedures. The combination of spring-assisted posterior vault expansion and fronto-orbital advancement in a single stage before one year of life decreases the number of surgical interventions necessary and allows for adequate remodeling of the cranial vault.

The traditional technique of occipital remodeling cause significant blood loss and relapse. We retrospectively present an evaluation of three children with syndromic craniosynostosis who underwent spring assisted distraction for posterior vault expansion leading to good esthetic and functional results.

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'POSTERIOR CALVARIAL EXPANSION USING CUSTOM MADE SPRINGS'; THE EVOLUTION OF A SURGICAL TECHNIQUE

Presenter: Owase Jeelani, MBA, MPhil, FRCS

Authors: Jeelani O, Hayward R

Great Ormond Street Hospital

The process of Calvarial Expansion has evolved over the past few decades, at our institution. Craniectomies and Biparietal expansions were the procedure of choice, followed by a Frontal Expansion and most recently a Posterior Calvarial Expansion. Posterior expansions were initially performed as large craniotomies with rigid fixation in an expanded position. This technique has gradually evolved with the use of springs acting as internal distractors and limited access approaches utilising endoscopes. We have done 56 cases in the past 3 years utilising the most recent technique and a total of 87 cases of PVE utilising springs. This paper aims to details the evolution behind this technique, the rational for the changes and expand on the surgical details of the current technique. The complications associated with this technique are presented as a separate paper.

Method: The patient is positioned prone and a bicoronal incision is fashioned. The skin and muscle dissection ranges from turning the flap down to the Nuchal line and below for Foramen Magnum decompressions to limiting the flaps to 3-4 cm around the bicoronal incision and completing the lower bone cuts below the retromastoid region using limited access endoscopic techniques. The bone cuts are made vertex down to the retromastoid with a craniotome under direct vision Cuts are then made horizontally or vertically towards the foramen magnum using an endoscope or minimal access techniques. The bone is removed completely in one or two pieces and reattached and springs are then placed. In the recent technique the bone is not lifted off the dura but instead green stick fractured out before spring placement.

Results: Ophthalmic surveillance was used to monitor ICP and an improvement was seen in 89% of the cases. The complication rate was comparable to our traditional cohort. The surgical time and length of stay was shorter with a transfusion rate of 68% compared with 89% for the earlier cases.

Conclusion: Limited access Posterior Vault Expansion using Springs is a safe, efficient and effective way of increasing the Calvarial Volume.

POSTERS



1P
SUCCESSFUL RECONSTRUCTION OF COMPLEX PEDIATRIC NASAL LESIONS: IMPROVING OUTCOMES USING A DERMAL REGENERATIVE TEMPLATE IN PEDIATRIC NASAL DEFECTS

Presenter: Oluwaseun A. Adetayo, MD
Authors: Adetayo OA, Grunwaldt LJ, MacIsaac ZM, Losee JE, Kumar AR

University of Pittsburgh

Background: The safety and efficacy of regenerate templates combined with full thickness skin grafts (FTSG) for pediatric facial defects is not widely published. The aim of this study is to report the safety and efficacy of pediatric nasal defect reconstruction using regenerative templates/FTSG.

Methods: A retrospective review of one institutions pediatric nasal defects secondary to nevi or vascular lesions treated with regenerative templates was performed. All patients (n=4) were treated with a multistage protocol. Two reviewers independently assigned Visual Analogue Cosmetic Scale (VACS) scores. Standardized photographs (anteroposterior, oblique, lateral and worm's eye view) were assigned VACS scores according to a 100 point scale: "Abhorrent", 0-24; "Poor", 25-49; "Moderate", 50-74; and "Excellent", 75-100. Statistical analysis was performed using Mann-Whitney U and Wilcoxon Paired Signed Rank tests, with significance $p < 0.05$.

Results: Four patients (two male and two female, average age 6.8 years) met the inclusion criteria. A total of five nasal lesions (two Spitz nevi, one vascular lesion, two congenital nevus) were removed. All patients underwent a two-stage procedure. Stage I included lesion excision and regenerative template placement. Stage II included silicone sheeting removal and FTSG placement. The average operative time per lesion was 65.5 minutes for Stage I and 93.3 minutes for Stage II. The average time between both operations was 13.8 days (range 10-19 days). The average graft area was 1.53 cm² (range 0.80-2.00 cm²). The average duration of follow up after Stage II was 175.8 days (range: 51-328 days). Complete skin graft take occurred in all patients (100%). The preoperative VACS score was 45.2 (range 5-70), compared to 84.5 (range 45-100) postoperatively ($p < 0.000$), indicating excellent results in all patients. There were no wound infections, bleeding requiring reoperation, or mortalities.

Conclusions: This technique is associated with low morbidity, short operative times, and high rates of skin graft survival resulting in significant improvement in Visual Analog Cosmetic scores.

2P
FEMTOSECOND PLASMA MEDIATED LASER ABLATION HAS ADVANTAGES OVER MECHANICAL OSTEOTOMY OF CRANIAL BONE

Presenter: Kevin J. Paik, AB
Authors: Chung MT, Paik KJ, Mackanos MA, Hyun JS, Montoro DT, Grova M, Liu C, Wang J, Chung AS, Palanker D, Connolly AJ, Longaker MT, Contag CH, Wan DC

Stanford University

Background: Although mechanical osteotomies are frequently made on the craniofacial skeleton, collateral thermal and mechanical trauma to adjacent bone tissue causes cell death and may delay healing. The present study evaluated the use of plasma-mediated laser ablation using a femtosecond laser to circumvent thermal damage and improve bone regeneration.

Methods: Critical-size circular calvarial defects were created with a trephine drill bit or with a Ti:Sapphire femtosecond pulsed laser. Healing was followed using micro-CT scans for 8 weeks. Calvaria were also harvested at various time points for histological analysis. Finally, scanning electron microscopy was used to analyze the microstructure of bone tissue treated with the Ti:Sapphire laser and compared to that treated with the trephine bur.

Results: Laser-created defects healed significantly faster than those created mechanically at 2, 4, and 6 weeks post-surgery. However, at 8 weeks post-surgery, there was no significant difference. In the drill osteotomy treatment group, empty osteocyte lacunae were seen to extend $699 \pm 27 \mu\text{m}$ away from the edge of the defect. In marked contrast, empty osteocyte lacunae were seen to extend only $182 \pm 22 \mu\text{m}$ away from the edge of the laser-created craters. Significantly less ossification and formation of irregular woven bone was noted on histological analysis for drill defects.

Conclusions: We demonstrate accelerated bone healing after femtosecond laser ablation in a calvarial defect model compared to traditional mechanical drilling techniques. Therefore, improved rates of early regeneration make plasma-mediated osteotomies of the craniofacial skeleton advantageous.



3P

USE OF NOVEL BMP-2 MINICIRCLE PLASMID-RELEASING SCAFFOLDS FOR BONE ENGINEERING

Presenter: Michael T. Chung, BS

Authors: Chung MT, Paik KJ, Morrison SD, Zimmermann AS, Lo DD, Hyun JS, McArdle A, Walmsley G, Senarath-Yapa K, Montoro DT, Hu MS, Chen HH, Chung AS, Ransom RC, Longaker MT, Wan DC

Stanford University

Background: BMP-2 has very strong osteoinductive activity and is known to facilitate bone regeneration. However, a single exposure to BMP-2 may not be sufficient to stimulate and sustain adequate bone growth. A gene-therapy approach to BMP manipulation may offer a more cost-effective alternative, considering the safety issues of viral vectors, a non-integrating technology would have the safest risk profile to enhance BMP signaling. The present study evaluated the potential of a gene-therapy approach employing a novel BMP-2 minicircle (mc)-releasing scaffold to promote bone regeneration.

Methods: A novel mc-releasing scaffold was prepared using supercritical CO₂ (scCO₂). Luciferase mc was lyophilized with 10% hydroxyapatite (HA) powder and pulverized PLGA. A PicoGreen® assay was conducted to determine release kinetics of Luc-mc. To evaluate the time course of mc DNA expression, Luc-mc-releasing scaffolds were implanted into calvarial defects in nude mice. Transgene expression was analyzed by in vivo bioluminescence imaging. For evaluation of in vivo osteogenesis, critical-sized (4-mm) calvarial defects in nude mice were treated with scaffolds containing BMP-2 mc DNA and rhBMP-2 protein for comparison.

Results: Scaffolds formed with scCO₂ gas foaming had a sustained release of plasmid for at least six weeks with retention of DNA integrity. Over the course of eight weeks, defects treated with the BMP-2 mc releasing scaffolds were found to consistently outperform defects treated with scaffolds alone. The mechanical strength of bone regenerated with delivery of BMP-2 mc DNA was comparable to that of rhBMP-2 regenerated bone.

Conclusions: Scaffold-based delivery of BMP-2 mc enhanced osteogenic capacity and facilitated more rapid regeneration of critical-sized calvarial defects. The ultimate goal would be to bring this non-integrating technology to the bedside to overcome the risk of insertional mutagenesis associated with viral-mediated gene delivery.

4P

BONE MORPHOGENETIC PROTEIN RECEPTOR IB AS A MARKER FOR ENRICHMENT OF OSTEOGENIC PRECURSORS

Presenter: Kevin J. Paik, AB

Authors: Chung MT, Paik KJ, Lo DD, Hyun JS, Montoro DT, Walmsley G, Senarath-Yapa K, Zimmermann AS, Sorkin M, Rennert R, Hu MS, Chen HH, Chung AS, Longaker MT, Wan DC

Stanford University

Background: Cell sorting has identified subpopulations of cells with enhanced osteogenic potential. However, some of these markers undergo a phenotypic drift in vitro. Type I BMP receptors play critical roles in the specification of osteoblasts and adipocytes, and may offer a more reliable cell surface marker. The present study evaluated the potential of a combinatorial approach, employing both FACS and manipulation of the BMP pathway, to enhance bone formation.

Methods: Flow cytometry was used to separate BMPR-IB+, BMPR-IB-, and unsorted ASCs. Phenotype was analyzed at Days 0, 7, and 14 using fluorochrome-conjugated antibodies. Each group was then treated with or without Noggin suppression, and cultured in ODM. Alkaline phosphatase and quantification were performed on Day 7, alizarin red staining and quantification on Day 14. Osteogenic gene expression was examined by qRT-PCR. In vivo, critical-sized calvarial defects were created in nude mice, and repair was performed using the above-mentioned subpopulations delivered on a HA-PLGA scaffold with or without BMP-2. Healing was monitored using micro-CT scans for eight weeks. Calvaria were harvested at Week 8, and sections were stained with Movats Pentachrome.

Results: Staining assays and qRT-PCR revealed that BMPR-IB+ ASCs were more osteogenic than BMPR-IB- or unsorted ASCs. Suppression of Noggin in ASCs resulted in increased osteogenic gene expression and in vitro osteogenic differentiation. In vivo, similar results were found, with Noggin-suppressed BMPR-IB+ ASCs seeded on BMP-2 scaffolds experiencing the greatest skeletal healing compared to the other groups over eight weeks.

Conclusions: Our findings demonstrate that subpopulations of ASCs based on BMPR-IB expression may be identified with enhanced osteogenic capacity and that ASC-mediated bone formation can be further promoted through manipulation of the BMP pathway. Use of BMPR-IB+ as a cell surface marker effectively identified an enriched group of cells that could facilitate more rapid regeneration of skeletal defects.



5P
ISOLATION OF ADIPOSE-DERIVED STROMAL CELLS USING LASER-ASSISTED LIPOSUCTION SPECIMENS AND THEIR THERAPEUTIC POTENTIAL IN REGENERATIVE MEDICINE

Presenter: Michael T. Chung, BS
Authors: Chung MT, Paik KJ, Liu CJ, Morrison SD, Lo DD, Hyun JS, Montoro DT, McArdle A, Walmsley G, Sorkin M, Rennert R, Chen HH, Senarath-Yapa K, Chung AS, Vistnes D, Longaker MT, Wan DC

Stanford University

Background: Liposuction method may impact the cellular characteristics of subsequently harvested ASCs. A previous study in our lab found that ultrasound-assisted liposuction (VASER), does not negatively affect the osteogenic potential of ASCs when compared to suction-assisted liposuction (SAL). However, a comparison between laser-assisted liposuction (SMARTLipo) and SAL has yet to be made. In this study, we investigated whether differences in yield, proliferative capacity, and differentiation potential exist between ASCs obtained by means of laser-assisted liposuction and SAL.

Methods: ASCs were harvested from patients undergoing elective SAL and laser-assisted lipoaspiration, then were stained with trypan blue and counted. The ASC phenotypical marker profile was determined using FACS, and cell proliferation was measured using an XTT assay. ASCs were differentiated under adipogenic and osteogenic conditions, and respective gene expressions were each examined by qRT-PCR at days 7 and 14. For evaluation of in vivo osteogenesis, critical-sized (4-mm) calvarial defects in nude mice were treated with HA-PLGA scaffolds seeded with uncultured, freshly isolated ASCs. Healing was monitored using micro-CT scans for eight weeks. Calvaria were harvested at week 8, and sections were stained with Movats Pentachrome.

Results: Laser-assisted liposuction resulted in lower total cell yield and reduced ASC viability and proliferation when compared with SAL. In addition, laser-assisted liposuction resulted in a lower frequency of cells with the ASC phenotype (CD34+CD31-CD45-). ASCs harvested from both techniques demonstrated adipogenic and osteogenic differentiation potential. Quantification of osseous healing by micro-CT revealed more healing with ASCs harvested via SAL relative to laser-assisted liposuction at the Weeks 4, 6, and 8 time points.

Conclusions: We found that adipose tissue obtained via SAL provides higher frequencies of rapidly growing ASCs than does adipose tissue via laser-assisted liposuction. Therefore, SAL seems to be preferable to laser-assisted liposuction for tissue-engineering purposes.

6P
ENHANCED ADIPOSE-DERIVED STROMAL CELL OSTEOGENESIS THROUGH SURFACE MARKER ENRICHMENT AND BMP MODULATION

Presenter: Kevin J. Paik, AB
Authors: Paik KJ, Morrison SD, Zimmermann AS, Lo DD, Hyun JS, McArdle A, Walmsley G, Senarath-Yapa K, Montoro DT, Sorkin M, Rennert R, Hu MS, Chen HH, Chung AS, Longaker MT, Wan DC

Stanford University

Background: Stem cell-based bone engineering has shown great promise as an approach for treating patients with skeletal defects. FACS has identified CD90+ ASCs with enhanced osteogenesis. More recently, inhibition of Noggin (NOG) has been shown to leave endogenously produced BMPs relatively unopposed, also resulting in enhanced osteogenesis. The present study investigated the potential of a combinatorial approach, employing both an isolation of CD90+ ASCs via FACS and modulation of BMP signaling, to optimize bone regeneration.

Methods: Magnet assisted transfection was used to deliver minicircle (MC) NOG shRNA into CD90+ and unsorted cells. These cells were then treated with ODM in vitro. Alkaline phosphatase and quantification were performed on Day 7, alizarin red staining and quantification on Day 14. Osteogenic gene expression was examined by qRT-PCR. For evaluation of in vivo osteogenesis, critical-sized calvarial defects in nude mice were treated with a novel MC-releasing HA-PLGA scaffold prepared using supercritical CO₂. MC plasmids were labeled with magnetic nanoparticles, and an external magnet was used to transfect ASCs in vivo. Healing was followed using micro-CT scans for eight weeks. Calvaria were harvested at Week 8, and sections were stained with Movat's Pentachrome.

Results: Transcriptional analysis revealed that the CD90+ ASCs were enriched for a more osteogenic subtype. Suppression of NOG resulted in increased osteogenic gene expression and in vitro osteogenic differentiation, as demonstrated by alkaline phosphatase and alizarin red staining. In vivo, over the course of eight weeks, the NOG-suppressed CD90+ subpopulation was found to consistently outperform other defects treated with the NOG-suppressed unsorted cells.

Conclusions: Our findings demonstrate that the use of CD90-selected ASCs may facilitate more rapid regeneration of skeletal defects. Furthermore, NOG knockdown may serve to augment bone differentiation through an increase in BMP signaling. The integration of these two strategies may lead to developing promising treatments for clinical translation.



7P

ADVANCES IN 3D IMAGE FUSION FOR CRANIOFACIAL TRANSPLANTATION

Presenter: Darren M. Smith, MD

Authors: Smith DM, Gorantla VS, Losee JE
University of Pittsburgh Medical Center

Purpose: Face transplantation has become a clinical reality. Sophisticated imaging modalities can be employed to maximize outcomes in surgical planning for three-dimensionally complex composite craniofacial defects. Skeletal, soft tissue, and neurovascular structures are imaged toward this end via techniques including surface scans, MRI, 3DCT and tractography. These data, however, are relegated to separate platforms and are often not compatible with real-time user interaction and modification. Here, we integrate data from multiple imaging sources into a single 3D representation of donor or recipient anatomy that supports real-time user interaction and modification.

Methods: Three-dimensional models of the craniofacial skeleton were generated as polygonal frameworks by thresholding and “stacking” dicom images from CT scans. Muscles were extracted from either the same dicom dataset as the bone data or from MRI, depending on data quality. If data quality permitted, blood vessels relevant to the model were extracted automatically from a CT angiogram. If the dataset was not of sufficient quality for this approach, key slices were imported into a 3D package and models manually segmented as above. Nerves were modeled as non-uniform rational basis splines based on tractography data.

Results: We successfully integrated disparate and unwieldy CT, surface scan, CTA, MRI and tractography data into detailed 3D anatomical polygonal meshes compatible with real-time end-user manipulation and modification. Important relationships between osseous, soft tissue, and neurovascular structures were visualized from a novel perspective.

Conclusions: Craniofacial transplantation is a complex procedure. Critical insight into 3D anatomy is afforded by powerful imaging techniques. We integrate classically disparate data into a single interactive 3D representation of donor or recipient anatomy compatible with real-time user interaction and modification. Procedural planning may be enhanced by allowing preoperative virtual interaction with simultaneously visualized skeletal, soft tissue, and neurovascular anatomy.

8P

USABILITY OF 3D SIMULATION FOR CRANIO-MAXILLOFACIAL SURGERY

Presenter: Takayuki Okumoto, MD

Authors: Okumoto T, Yoshimura Y, Kondo S, Imamura M

Fujita Health University School of Medicine

Object: Surgical planning is the most important part as well as the accuracy of the operation in cranio-maxillofacial surgery. The conventional cephalogram is useful for a two-dimensional surgical planning. Recently, a three-dimensional CT image is available easily, making it possible to comprehend the deformity intuitively. But on planning the treatment for the asymmetric cases mainly composed of three-dimensional distortions, the three-dimensional CT images output on films or a monitor are useless for the simulation as with the cephalogram, because they are essentially two-dimensional. In such cases, a three-dimensional simulation is extremely useful, so it should be promoted more in clinical use.

Methods: We use an image processing software for 3D design and modeling, Mimics® (Materialise NV, Belgium) for simulation. CT image data output by DICOM format are imported into the software, and 3D skeletal images are calculated excluding the unnecessary data such as artifacts after extracting only bone images. Furthermore, these 3D images can be cut like a real surgery using optional planes on three-dimensional space or cutting templates for osteotomy set in advance such as Le Fort I osteotomy, SSRO etc. Each cut image is recognized as a different 3D object and we are able to move it freely on a three-dimensional space. Both the direction and the distance in the movement of the objects should be assessed in reference to the measurement results of a cephalogram or the dental cast model.

Result: We have used this method on over 100 cases. This method was particularly useful in treatment planning of the asymmetric cases, and could give us more information such as the contact or interference condition between each bone fragment after movement.

Conclusion: Three-dimensional simulations enable us to make the reasonable plan, and it is thought that safety and certainty in the real surgery rise drastically.



9P
**THREE-DIMENSIONAL MORPHOMETRIC ANALYSIS
OF THE HYPOPLASTIC MANDIBLE**

Presenter: Kevin J. Paik, AB
Authors: Paik KJ, Levi B, Hyun JS, Lo DD,
Montoro DT, Lisiecki J, Chung AS,
Bradley JP, Buchman SR, Longaker MT,
Wan DC

Stanford University

Background: Pierre Robin sequence and Treacher Collins syndrome are both associated with mandibular hypoplasia. However, it has been hypothesized that the mandible may be differentially affected. The purpose of this study was therefore to compare mandibular morphology in children with Pierre Robin sequence to children with Treacher Collins syndrome using three-dimensional analysis of CT scans.

Methods: A retrospective analysis was performed identifying children with Pierre Robin sequence and Treacher Collins syndrome receiving CT scans. Three-dimensional reconstruction was performed and the following measurements were taken: ramus height, mandibular body length, and gonial angle. These were then compared with the clinical norm corrected for age and sex based on previously published measurements. Three children with Pierre Robin sequence (six hemi-mandibles), four children with Treacher Collins syndrome (eight hemi-mandibles), and two control children (four hemi-mandibles) were identified. The mean age for patients with Pierre Robin sequence was 10.61 years. The mean age in the Treacher Collins syndrome group was 12.07 years. The mean age in the control group was 10.74 years.

Results: In our study, mandibular body length was significantly shorter for children with Pierre Robin sequence while ramus height was significantly shorter for children with Treacher Collins syndrome. This resulted in distinctly different ramus height/mandibular body length ratios. In addition, the gonial angle was more obtuse in both the Pierre Robin sequence and Treacher Collins syndrome groups compared with the controls.

Conclusions: Three-dimensional morphometric analysis of mandibles in patients with Pierre Robin sequence and Treacher Collins syndrome revealed distinctly different patterns of mandibular hypoplasia relative to normal controls. These findings underscore distinct considerations that must be made in surgical planning for reconstruction.

10P
**COMPARATIVE ANALYSIS OF THE MUSCULAR
STRUCTURES OF THE NECK IN PATIENTS WITH
PIERRE ROBIN SEQUENCE AND PATIENTS WITH
NORMAL MANDIBULAR GROWTH**

Presenter: Andres Bello, MD
Authors: Bello A, Lopez R, Molina F
Hospital General Dr Manuel Gea Gonzalez

Patients with Pierre Robin sequence have microretrognathia and studies prove that they never achieve normal mandibular development. These observations lean towards the idea that the suprahyoid muscles are shorter and play a crucial role in mandibular growth. Even though patients are treated with different surgical modalities, there is a tendency towards failed mandibular growth. Because of this, it's necessary to study the dimensions of the suprahyoid muscles that directly interact with the mandible. Besides, there are no quantitative reports of the degree of affectation of this muscle group. Using tomography images, the suprahyoid muscles in Pierre Robin patients were measured and compared to similar age patients without the anomaly. Results show that the most affected muscles are the geniohyoid and the anterior belly of the digastric muscle. This explains the natural tendency towards micrognathia and microgenia observed in these patients during development. The affectation of the mylohyoid explains the light concavity observed in the mandible body of Pierre Robin patients. This study proves the important role of the suprahyoid muscle group that, through their short length and contraction, secondarily affects mandibular growth.



11P

A THREE DIMENSIONAL ANALYSIS OF FACIAL-LABIAL FOLD AFTER MALAR AUGMENTATION OR REDUCTION

Presenter: Zheyuan Yu, MD

Authors: Yu Z, Cao DJ, Chai G, Zhu YJ, Mu XZ
Shanghai Ninth People Hospital

Object: to investigate the three dimensional changes of facial-labial fold for patients after malar augmentation or reduction.

Methods: 47 patients who accepted malar augmentation or reduction surgeries were divided into four groups. Group 1: malar augmentation for zygoma complex fracture (n=16). Group 2: aesthetic malar augmentation (n=6). Group 3: malar arch reduction (n=9). Group 4: total malar reduction (n=16). All patients accepted a spiral CT scan before and 3-6 months after surgery. The facial surface was reconstructed and separated from raw data by Rapidform2006. A three dimensional analysis was then applied to find out the changes of facial-labial fold.

Results: An increasing depth of facial-labial groove was observed in all patients after malar augmentation or reduction. In Group 1 and Group 2, the depth increased $2.3\text{mm}\pm 1.4\text{mm}$ averagely and the facial-labial fold maintained in U shape. In Group 3, the increase was insignificant. In Group 4, it increased $1.6\text{mm}\pm 1.2\text{mm}$ and the topography of facial-labial groove presented trends to V shape.

Conclusion: Deepen of facial-labial fold in malar augmentation seemed to be formed by the upwards and anterior deformation of midface muscles while in total malar reduction it was formed by loose of midface muscle attach. This might help us to improve the details in malar surgeries.

12P

BRAIN VOLUME AND SHAPE CHANGES IN MIDLINE NON-SYNDROMIC CRANIOSYNOSTOSIS PATIENTS

Presenter: John A. van Aalst, MD, MA

Authors: Halevi A, Krochmal DJ, Paniagua B, Styner M, van Aalst JA

The University of North Carolina at Chapel Hill

Background: Sagittal suture synostosis has historically constituted approximately 60% of patients in large series of non-syndromic single suture synostosis; however, a recent increase in metopic synostosis has been documented. Patients with sagittal synostosis often have no measurable developmental delays; however, delays are often noted in patients with metopic synostosis. These delays may not be ameliorated by surgical volume expansion. Surgical changes in intracranial volume (ICV) are poorly characterized in these patients, with contradictory findings in the literature. This pilot study evaluates pre- and post-surgical ICVs derived from 3D computerized tomography (CT) reconstructions of patients with non-syndromic sagittal and metopic synostosis for comparison with age and sex-matched normal patients.

Methods: All study patients (n = 15; 8 metopic and 7 sagittal) underwent CT scans with 3D reconstruction pre- and post-surgically. An age and sex-matched ICV regression curve was generated using a unique 350-patient normal MRI database (Infant Brain Imaging Study.) A similar process was used to compute total ventricular volume (n=11). A validation study with CT and MRI imaging modalities demonstrated limited variability (1.54%), confirming that these modalities are comparable. CT-derived head circumference was compared to computed ICV and clinically-derived HC.

Results: All 15 cases demonstrated increased ICV compared to healthy controls (independent of gender, age, and diagnosis) both pre- and post-surgically; brain parenchymal volume findings were similar. Ventricular volume increased post-operatively more significantly in patients with metopic synostosis. CT-derived HC had a high correlation (0.85) with pre-surgical and moderately (0.59) with post-surgical ICV. CT-derived and clinical head circumference had a strong correlation both pre- and post-surgically (0.77 and 0.91).

Conclusions: The use of volumetric measurements suggests that brain volume is not normal in patients with single suture craniosynostosis, even post-surgically. Clinical HCs are reasonable surrogates for ICV.



13P
DIFFERENCES IN MEASUREMENTS OF THE SKULL BASE AND FACIAL SKELETON IN PATIENTS WITH PLAGIOCEPHALY VERSUS PATIENTS WITH PLAGIOCEPHALY AND FACIAL SCOLIOSIS

Presenter: Cuauhtemoc Lorenzana, MD
Authors: Lorenzana C, Arrieta P, Francis FA, Molina F
Hospital General Dr Manuel Gea Gonzalez

Investigations suggest that in patients with plagiocephaly the affection is not only in the cranial vault, but also in the skull base, and that the higher number of sinostotic sutures of coronal ring, the greater the degree of affectation and more likely to develop facial asymmetries. In the literature we can find numerous descriptions of malformations found in patients with anterior plagiocephaly, but most of these reports focus on the orbitofrontal deformity and displacement of the ears and few describe quantitatively the skull base, facial skeleton and its alterations, and none offers comparison between measures of skull morphology, skull base and facial skeleton that include measurements of the cranial fossas, maxilar, external acoustic meatus, mandibular dimensions to detect where the problem sets with plagiocephaly patients who develop facial scoliosis. We analyzed by preoperative tomographic measurements, the Skull characteristics and the coronal ring sutures affected in patients with plagiocephaly, for so in that way, obtain the morphologic differences that allow us consider the posterior development of facial scoliosis. 29 patients were found with plagiocephaly with or without facial scoliosis. In our observation we found that the greater difference is in: 1) the situation of the external acoustic meatus, 2) the position and dimension of the mandible and glenoid fossa, and 3) the affection of 3 or more coronal ring sutures are factors that differentiate patients with plagiocephaly and facial scoliosis and are associated with the development of facial scoliosis. Therefore, concluded that through intracranial and extracranial quantitative studies, we can differentiate the patients with plagiocephaly that will develop facial scoliosis.

14P
PATTERN OF DENTAL EXTRACTIONS IN PATIENTS WITH SYNDROMIC CRANIOSYNOSTOSIS UNDERGOING CRANIOFACIAL SURGERY

Presenter: Susana Dominguez-Gonzalez, PhD
Authors: Dominguez-Gonzalez S, Richardson DR, Laraway DC
Alder Hey Childrens NHS Foundation Trust

Objectives: Children with syndromic craniosynostosis often have narrow, crowded dental arches, and require extensive postoperative orthodontic work. The treatment usually involves several dental extractions to create space in the arch for the alignment of teeth, to achieve a satisfactory occlusion. The aim of this paper was to look at the maxillary and mandibular extraction patterns in our cohort of patients, and to compare it to that of the general orthodontic population.

Materials and Methods: Alder Hey Hospital is a supra-regional craniofacial centre in the UK, which has been carrying out mid-face advancement surgeries since 2000. The unit has complete orthodontic records (study models, photographs and panoramic radiograph) of 15 patients who had undergone craniofacial surgery (Le Fort III osteotomy, Monoblock frontofacial advancement, Facial bipartition), followed by either distraction or rigid fixation. 9 patients had Apert Syndrome, 4 Crouzon, 1 Saethre-Chotzen and 1 Fronto-Nasal dysplasia. One Crouzon and the Fronto-Nasal dysplasia patients had standard Le Fort III advancement and direct fixation, and all others underwent rigid external fixation. The orthodontic case notes were studied to obtain the information on dental extractions.

Results: All patients required four or more dental extractions. 6 patients had 4 maxillary and 2 mandibular teeth extracted, 6 had 2 upper and two lower teeth extracted, 2 patients had 3 maxillary and 2 mandibular teeth extracted and 1 had 3 upper and 1 lower teeth extracted. The teeth more frequently extracted were the maxillary premolar and lateral incisors and lower premolars and canines.

Conclusions: All patients required at least four extractions, in contrast to the general population, in which only 28% of patients needed four (1). This study shows that patients with syndromic craniosynostosis have a much higher incidence of multiple maxillary extractions during orthodontic treatment than patients undergoing orthodontic care in the general population.

Reference: Proffit WR. Forty-year review of extraction frequencies at a university orthodontic clinic. *Angle Orthod*, 1994



15P
**SUTURAL DISTRACTION OSTEOGENESIS (SDO)
OF INTERNAL DISTRACTOR TREATED UNILATERAL
CORONAL SYNOSTOSIS AND IMPLANTED MEDPOR
FOR BLOCK SKULL GROWTH AND REPAIR OF SKULL
DEFECT: A PRIMARY CLINICAL REPORT**

Presenter: Shen Weimin, MD
Authors: Weimin S, Cui J, Jianbing CH, Jijun Z, Ji Y,
Haini CH

Nanjing Childrens Hospital affiliated with Nanjing Medical

Objective: This study was conducted to review the changes in forehead after unilateral forehead anterior advancement by Sutural Distraction Osteogenesis in patients with unilateral coronal synostosis. Implanted medpor for block skull growth and repair of skull defect.

Materials and Methods: Five cases were unilateral coronal synostosis, the distraction system used of internal distraction and a suturectomy is needed, we did coronal suture strip craniectomy with surgybone (Silfradent S.r.l.), the width of strip was 2cm. During surgery, we assessed the synostosis of the coronal suture on the left side and placed two internal distracters beside left coronal suture. On the second day after the surgery, the distraction of 1.2 mm per day started and continued for 15 days, and 18 mm forward distraction was obtained. The children with plagiocephaly were treated by suturectomy and sutural distraction. After remove distraction, we had filled a medpor in the skull defect site and repaired the skull defect caused by distraction. After being distracted, the patients established harmonious facial profiles and normal foreheads.

Results: Five unilateral coronal synostosis had cured after one year, shape of fronthead were good.

Conclusions: The technique of suture osteotomy and distraction osteogenesis is suitable for infants. The technique of sutural distraction osteogenesis is suitable for young infant, the ages were below 6 month. implanted medpor can block skull growth and repair of skull defect. Medpor can repaired skull defect due to distraction and improved temporal profile Radiographic examination showed balanced advancement of the skeleton. It is suggested that the treatment of plagiocephaly in infant by the technique of suture osteotomy sutural distraction osteogenesis and implanted medpor is to be preferred because of its simplicity and relative minimal invasive. Thus, the authors suggest that plagiocephaly should be treated at a young infant by this technique.

Keywords: sutural distraction osteogenesis, unilateral coronal synostosis, medipor.

16P
**VIRTUAL DESIGN OF BILATERALLY DESTROYED
TRAUMA CASE S BASED ON THREE DIMENSIONAL
NORMAL SKULL DATABASE**

Presenter: Xiaojing Liu, MD
Authors: Liu X, Guo CB, Zhang Y, Chen L, Wang J
Peking University School and Hosptial of Stomatology

Background: Mirror imaging is often applied in virtual surgical planning of maxillofacial trauma patients. However, the technique doesn't work in bilaterally destroyed cases.

Objects: The object of this study was to investigate the feasibility of designing bilateral destroyed trauma cases based on three dimensional skull database.

Methods: A relational database based on MySQL was developed for the storage and retrieval of three dimensional skulls. During 2009 to 2012, 120 spiral CT data were gathered and imported into database. 72 anatomic bony land marks were manually identified on each skull after 3D reconstruction. Relevant distances, angles and distance ratios were calculated. All the skulls were labeled and could be retrieved by general information and quantized descriptions.

Up to now, five cases of trauma cases, including 2 cases of sever midface fracture, 2 cases of mandible defects and 1 case of maxilla defect, were virtually designed based on the database. At the first stage of virtual design, points were indentified at the remained area of patients skull. Similar normal skulls were retrieved and listed according to similar ratio and the final reference skull was decided by doctors.

The second stage of virtual design involved the surgical simulation assisted by reference normal skull. DICOM data of both trauma and reference skull were imported into I plan System 3.01 (BrainLab, Germany). The two sets of data were automatically overlapped using "auto image fusion" module. Virtual surgical plan including osteotomy, reposition and design for custom implant were performed according to the contour of reference skull.

Results: All the five cases were successfully operated following preoperative virtual design. None complication occurred. All the patients were satisfied with the outcome.

Conclusion: Normal skull database could serve as reference during the virtual surgical design for bilaterally destroyed trauma cases.



17P
**COST ANALYSIS OF AUTOGENOUS VS.
PREFABRICATED PATIENT-SPECIFIC ALLOPLASTIC
CRANIOPLASTY**

Presenter: Amir Mrad, MD, MBA, FRCSC
Authors: Mrad A, Antonyshyn DR
University of Toronto/Harvard University

Purpose: To compare costs associated with autogenous cranioplasty vs. Prefabricated Patient-specific Alloplastic cranioplasty

Methods: Retrospective chart review of a series of patients that underwent autogenous cranioplasty and PEEK cranioplasty. Data abstracted from the charts will include demographic data, particulars of surgery, details of post-op recovery and duration of surgery. Also included are the expenses related to duration of surgery, ICU stay, duration of hospital admission(?), severity and impact of complications and cost of hardware. Outcomes will be assessed in terms of skull contour preservation and complications. Eleven patients underwent PEEK cranioplasty at our institute between July 2009 and June 2011. We identified 11 additional patients who underwent split skull autogenous bone graft cranioplasty, that were matched for age and skull defect size. Similar data was abstracted for both groups. The data was analyzed to compare costs of Prefabricated Patient-specific Alloplastic implants to costs of autogenous reconstruction.

Results: The results of this study will provide valuable information that may guide distribution of hospital resources or funding of technology at ministry of health level.

Teaching Objectives: The participant will gain knowledge of long term outcomes of both autogenous and prefabricated alloplastic cranial vault reconstruction. The participant will be able to guide his future practice in terms of which cranioplasty procedure to choose from based on hospital resources or funding available.

18P
**ZYGOMATIC FRACTURE: EXTENDED
CLASSIFICATION FOR ADDITIONAL OPTION FOR THE
IMMEDIATE MANAGEMENT**

Presenter: Kazuhiro Otani, MD, PhD
Authors: Otani K, Kyutoku S, Okada M, Yamada A,
Ueda K
Osaka Medical College Hospital

Night & North reported a classification of the zygomatic fracture, as an anatomical disconnection to surrounding structure, in 1961, and it has widely been applied for simple fixation. A new concept for the comprehensive classification of comminuted zygomatic fractures combined with skull base, medial orbit and maxilla into 4 types, is presented; type Z is so-called zygoma fracture of Night & North type III, IV, V (with or without arch fracture), type A is upper and posterior extension (orbital Apex, spheno-zygomatic and skull base area), type B is inferior extension (maxillary Buttress and Le Fort I, II combination) and type C is medial extension (naso-ethmoidal and Canthal complex). The concept of the clinical alternative to Night & Norths classification is to arrange its approach according to its extension and to repair the additional anatomical displacement and malfunction; surgeon should prepare the comminuted problems from simple trauma, clearing an acute embellishment at ER and immediate required fixation lessen the secondary annoying operation. In the last decade, we encountered 104 zygomatic fractures (12 in NK type I, 13 in II, 44 in III, IV, V, 16 in VI and 15 comminuted and 17 included blow-out fractures) out of 333 facial trauma in our group. Representative case of each type are illustrated.



19P

USE OF CT DERIVED PROSTHETICS FOR INTRA-OPERATIVE GUIDANCE OF TUMOR RESECTION

Presenter: Christian J. Vercler, MD

Authors: Vercler CJ, Kline S, Buchman SR
University of Michigan

Introduction: Patients with neurofibromatosis of the face present a challenging deformity as there is a combination of both bony and soft tissue abnormalities that must be resected and reconstructed. Cutting edge technology has now made it possible for surgeons and prosthetists to combine their expertise by utilizing 3D CT-derived models to create patient-specific prostheses and implants for craniofacial rehabilitation. Using this technique to create intra-op osteotomy guides is now commonplace, but we are using this modality to aid in both bony and soft tissue reconstruction.

Methods: This approach is well-suited for patients with neurofibromatosis who require further debulking of the orbitozygomaticomaxillary complex with the specific goal of having an orbital prosthesis fabricated. To ensure that we remove adequate bony overgrowth and neurofibroma so the area will accommodate an orbital prosthesis, we use CT-derived intra-operative guides. These guides are created by our anaplastologist who makes models of the patient's ideal post-surgical orbital region, including unilateral and bilateral periorbital soft tissue, and a mask from the forehead down to stomion—all based on a mirror image projection from the unaffected side of the face.

Results: These custom anatomic models allow for a precise resection to an endpoint identified by the anaplastologist as ideal for subsequent orbital prosthesis creation and placement. Without these intra-operative guides we would have to rely on our aesthetic judgment, which may or may not achieved the optimum result.

Conclusion: Three-dimensional reconstruction of CT scan and cone-beam CT (CBCT) scan images have contributed significantly to pre-operative craniomaxillofacial surgery planning. At our institution we work closely with our anaplastology team to create intra-operative aides that allow for precise soft tissue and bone resection and reconstruction. This interdisciplinary approach takes advantage of the latest advances in imaging and biomedical engineering with the goal of achieving better outcomes for the patient with fewer subsequent operations.

20P

BRANCHIAL ARCH ANOMALIES: RECURRENCE, MALIGNANT DEGENERATION AND OPERATIVE COMPLICATIONS

Presenter: Dave Stoddard, MD

Authors: Al-Mufarrej F, Stoddard D, Bite U
Mayo Clinic

Introduction: Branchial arch anomalies (BAA) represent one of the commonest pediatric neck masses, but large case series are lacking.

Methods: From 1/1/1976-7/29/2011, 421 subjects underwent BAA excision at our institution. Records were retrospectively reviewed. Features studied include age, gender, location, BAA type, symptoms, recurrence, preoperative management, extent of surgery, pathology as well as presence of tracts. Associations with tracts, operative complications, and recurrence were evaluated.

Results: Frequencies of first, second, third and fourth BAA were 20%, 75%, 5% and 1%, respectively. 41% of study participants had BAA associated with tracts. Mean age for the subjects with tracts was 15.3 years compared with 33.2 years for others ($p < 0.001$). Of the 420 subjects with available pathology, 60.2% had inflammatory changes or cartilage with no identifiable epithelium. 4 cases (mean age 60.3 years) demonstrated malignant degeneration. Among the 370 subjects with no previous surgery, 13 recurred at a mean of 47.1 months postoperatively. Recurrence was more likely with preoperative incision and drainage (hazard ratio 3.55; $p = 0.035$). 2% experienced complications. Age, BAA type, preoperative imaging and extent of surgery did not affect recurrence or complication rates.

Conclusion: Patients with history of preoperative incision and drainage should be followed closely for recurrence the first 3 years postoperatively. Early BAA excision is not associated with higher complication rate. Extent of resection should be determined by gross margins of BAA. Older patients presenting with should raise suspicion for malignancy, and a thorough work-up is important for correct diagnosis.



21P

LATERAL TRANSZYGOMATIC APPROACH SPHENOID MENINGIOMA

Presenter: Henry M. Spinelli, MD

Authors: Spinelli HM, Hanasono MM, Langevin CJ
Weill Medical College of Cornell University

Objectives: Sphenoid wing meningiomas are slow growing, well circumscribed, and histologically benign lesions. The recurrence rate is low if removed completely at the time of surgery. Adequate surgical exposure with minimal morbidity is a challenge for those treating these lateral skull base lesions.

Methods: A retrospective review of the records of 34 patients who underwent sphenoid wing meningioma resection via a lateral transzygomatic approach between 1997 and 2011 was performed. A confirmatory cadaver dissection was performed to illustrate the anatomic nature and reproducibility of the technique.

Technique: To achieve maximal exposure and minimal brain retraction, a lateral transzygomatic approach with osteotomies of the entire zygoma, which remains pedicled on the masseter muscle, was used. A skull can be selectively resected when necessary.

Results: Thirty-four patients with sphenoid wing meningioma underwent resection via a lateral transzygomatic approach. Complete resection of the meningioma was achieved in 31 cases. Morbidity consisted of temporary frontal nerve weakness (57.9%), mild to moderate temporalis atrophy (36.8%) and diplopia (15.8%). There were no cases of wound infection, bone malunion or resorption. A mean follow up period of 43.1 months (range of 12 to 85 months) revealed no recurrences following surgery as demonstrated by computed tomography or magnetic resonance imaging.

Conclusions: The lateral transzygomatic approach to the sphenoid wing can be performed safely with minimal morbidity and facilitates complete resection of the tumor. Complete removal at an early stage is the best prognostic factor in treating sphenoid wing meningioma. This approach is based on fundamental anatomic principals and can routinely be performed by the average craniofacial surgeon employing a steep learning curve. The lateral transzygomatic approach belongs in the armamentarium of all surgeons who are involved in the resection of skull base neoplasms.

22P

MANAGEMENT OF MANDIBULAR HIGH-FLOW ARTERIOVENOUS MALFORMATIONS

Presenter: Jocelyn M. Shand, MBBS, MDSc,
FRACDS(OMS), FDSRCS(Eng)

Authors: Shand JM, Heggie AA
The Royal Childrens Hospital of Melbourne

Large arteriovenous malformations (AVMs) of the mandible are rare and potentially life-threatening lesions. Pulsatile expansion of the buccal cortex, interdental gingival bleeding and socket haemorrhage post-extraction are typical features of high flow lesions. Due to the risk of catastrophic haemorrhage, these vascular malformations are a major challenge that require an intergrated team approach with close cooperation between surgeons, interventional radiology and anaesthesia. Embolization and complete surgical resection of AVMs has been considered as the treatment of choice yet there remains a risk of recurrence. However, in the case of an extensive lesion, surgical removal of the nidus may be contraindicated due to the potential for a major, disfiguring deformity. Our experience in managing two extensive, life threatening AVMs of the mandible, in an 11- and a 15-year old male, will be presented and the multiple stages in their management discussed.



23P

EVALUATING THE EFFICACY OF AIRWAY EXPANSION USING TRANS-CRANIAL VERSUS SUB-CRANIAL FACIAL OSTEOTOMIES: A COHORT COMPARISON STUDY BETWEEN MONOBLOC FRONTOFACIAL ADVANCEMENT AND LE FORT III FACIAL ADVANCEMENT

Presenter: Oluwaseun A. Adetayo, MD

Authors: Adetayo OA, Rottgers SA, Miele LF, MacIsaac ZM, Davidson EH, Kumar AR

University of Pittsburgh

Background: Differential airway volume expansion comparing transcranial versus subcranial facial advancement procedures remains understudied. The aim of this study is to compare differences in airway changes between monobloc/bipartition advancements versus Lefort III advancements.

Methods: A 16-month retrospective cohort study comparing airway changes using radiographic data (total airway volume (TAV), nasopharyngeal airway volume (NAV), oropharyngeal airway volume (OAV), and minimal airway areas in the nasopharynx and oropharynx in patients treated with monobloc/bipartition advancements (Group 1) and Le Fort III advancements (Group 2) was performed. Kawamoto internal distraction devices (KLS Martin, Jacksonville, FL) were used.

Results: In Group 1, four patients (1 female, 3 males, all syndromic) and in Group 2, three patients (3 males, all syndromic) were identified who were treated for airway obstruction and signs of increased cranial pressure (Group 1). Average age was 8.9 years in Group 1 and 15 years in Group 2 ($p=0.13$). The average radiographic advancement (body of C2-A) was 12.4 mm (10-17) in Group 1 and 12.27 mm (9.9-14.3) in Group 2 ($p=0.14$). The average increase in NAV, OAV, and TAV was 5,374 mm³, 3,463 mm³, and 8,837 mm³ in Group 1 and 5,786 mm³, 2,758 mm³, and 8,433 mm³ in Group 2 ($p=0.186$, 0.734, 0.586). The minimal nasopharyngeal airway area increased in Group 1 (63.7 mm² to 194.6 mm², $p=0.068$) and in Group 2 (59.6 mm² to 104 mm², $p=0.109$). All patients in both groups demonstrated resolution of clinical airway obstruction. One major complication occurred in Group 1 (cerebral salt wasting syndrome/seizure) and no major complications occurred in Group 2.

Conclusions: Airway changes within the nasopharynx, oropharynx, and total upper airway were similarly expanded using either a transcranial or subcranial facial advancement in two patient populations with similar preoperative airway volumes. Transcranial expansion should be considered for the treatment of concurrent cranial constriction, as transcranial and subcranial facial expansion are equally effective treatment for nasopharyngeal airway expansion.

24P

A COMPARATIVE ANALYSIS OF COMPLICATIONS IN 55 MIDFACE DISTRACTION PROCEDURES IN PATIENTS WITH SYNDROMIC CRANIOSYNOSTOSIS

Presenter: Jesse A. Goldstein, MD

Authors: Goldstein JA, Taylor JA, Bartlett SP

University of Pennsylvania School of Medicine & Childrens Hospital of Philadelphia

Background: This study was designed to compare the peri-operative complications between buried and halo-type distraction osteogenesis of the midface.

Methods: A retrospective review was performed on all patients with syndromic craniosynostosis who underwent midface distraction with buried or halo-type external distractors. Demographic information and operative/post-operative course were reviewed. Complications were categorized either as major (requiring additional intervention) or minor (requiring medication only). Chi-squared and Fisher's exact test were used to compare variables.

Results: From 1999 to 2012, 54 patients underwent midface distraction osteogenesis including 23 patients with Aperts, 18 Crouzon, 10 Pfeiffer and 3 with other craniofacial syndromes. 33 patients underwent a total of 34 subcranial Le Fort III distraction procedures and 21 underwent 21 monoclock distraction procedures. Average age of surgery was 8.0 (range: 4.0-17.7) years, while average time between distractor placement and removal was 102.9 days. 30 procedures were performed with external halo-type distractors (18 Le Fort III and 12 monoclocks), while 25 were performed with buried distractors (16 Le Fort III and 9 monoblocks). There were no significant differences in diagnoses or operation type between distraction techniques. Of the 19 distractor related complications, there were a total 10 (33%) in the halo group including 5 (16%) requiring separate operative intervention and 9 (39%) in the buried distractor group including 6 (26%) requiring separate operative intervention. Serious infections were the more common in the buried distraction group ($n=8$) compared to the halo distractor group ($n=3$) ($p=0.048$). There were four (13.3%) patients in the halo groups who had malposition or transcranial pin migration related to post-operative positioning or falls and required operative repositioning.

Conclusions: Midface distraction is safe but poses some risks. Higher rates of halo displacement requiring surgery are off set with lower rates of infections compared to buried distractors.



25P

OUTCOMES ANALYSIS OF MANDIBULAR DISTRACTION OSTEOGENESIS FOR THE TREATMENT OF PIERRE ROBIN SEQUENCE ASSOCIATED WITH ADVANCED AIRWAY OBSTRUCTION

Presenter: Melinda Costa, MD

Authors: Murage KP, Tholpady SS, Friel M, Costa M, Havlik RJ, Flores RL

Indiana University School of Medicine

Background: Mandibular distraction osteogenesis (MDO) is an established technique used to treat infants with Pierre Robin Sequence (PRS) associated with severe airway obstruction. Despite its widespread use, there is little information on the limitations and morbidity associated with MDO in this patient population. We report an outcomes analysis focusing specifically on our failures and complications.

Methods: A 7-year retrospective review of all patients with PRS treated with MDO. Recorded variables included: need for tracheostomy, complications, prematurity, low birth weight, cleft palate, genetic syndromes, heart and airway abnormalities, gastroesophageal reflux disease (GERD), need for feeding tube, Nissen fundoplication, late operation (2 weeks or greater at the time of MDO) and pre-operatively intubated. Failure in MDO was defined as need for tracheostomy post MDO. The association of failure of MDO with the listed preoperative variables was determined using a Fisher exact test.

Results: 50 patients were identified for this study. Patient characteristics included: mean gestational age 37 wks; Prematurity (22%); mean birth weight 2.98kg; low birth weight (20%); cleft palate (84%); genetic syndrome (22%); cardiac anomalies (12%); pulmonary abnormalities (26%); GERD (26%); gastrostomy tube (58%); Nissen (12%), late presentation (76%); and pre-operatively intubated (4%). The most common complication was infection (22%) and all cases except one were successfully treated using antibiotics alone. Other common complications included self extubation (4%) and device fracture (2%).

Four patients (8%) required tracheostomy post MDO. The only variables demonstrating statistical association with MDO and tracheostomy were in patients with no cleft palate ($p=0.0003$), GERD ($p=0.003$) and Nissen fundoplication ($p=0.00006$).

Conclusion: MDO can be safely applied to infants with PRS. The most common complication is infection and most cases can be treated non-surgically. Although MDO is an effective technique, absence of a cleft palate, GERD, and need for Nissen fundoplication are associated with failure.

26P

EXPANSION OF THE POSTERIOR CRANIAL VAULT: CHARACTERIZATION OF THE ROLE OF DISTRACTION OSTEOGENESIS

Presenter: Raymond J. Harshbarger III, MD

Authors: Harshbarger RJ, Combs PD, Kelley PK, George TM

Dell Childrens Medical Center

Introduction: Expansion of the posterior cranial vault can be used to treat cephalocranial disproportion (CcD) and cranial vault dysmorphology. Techniques to achieve expansion include: traditional osteotomies with remodeling, springs, and distraction osteogenesis (DO). While initial studies have shown achievability of posterior cranial vault (PCV) DO in syndromic and late presenting sagittal synostosis, better defined uses for this technique have not been described. The goal of this study was to further elucidate clinical situations in which PCV DO is useful.

Patients/Methods: A retrospective review was conducted at Dell Children's Medical Center identifying 16 patients who underwent 18 PCV DO. Diagnosis, operative notes, perioperative course, distraction protocol, complications, and devices were reviewed.

Results: Categorization by diagnosis: Syndromic -4, Nonsyndromic multisuture synostosis-6, Single suture synostosis- 3, Shunt related/slit ventricle syndrome- 2, Post-traumatic deformity-1. 14/16 patients had increased ICP by observation/measurement. All patients were distracted successfully with adequate consolidation and improvement of symptoms +/- ICP. 38% (6/16) had prior CVR. 38% underwent PCV DO, staged before anterior vault (ACV) surgery. Overall complication rate: 22%, last 10 cases 10%.

Conclusions: Based on our cohort, PCV DO is achievable in 4 main diagnostic groups: 1) CcD, 2) Repeat expansion, 3) Staging PCV expansion prior to anterior procedures in severe deformities, 4) Late presentation of craniosynostosis with mild deformity. Patients with CcD, not requiring reshaping, improve with PCV DO. 14 patients (87%) with evidence of high ICP, all had improvement after DO. PCV DO works in areas of scarring requiring repeat expansions. 6 (38%) patients with prior CVR had successful PCV DO without complication. In severe deformities, staging surgery, PCV DO performed first, with subsequent ACVR, was achieved in 6 patients. This study further characterizes the uses of PCV DO in the arsenal of techniques for expansion of the posterior vault.



27P

CAN WE PREDICT THE NEED FOR ICU ADMISSION AFTER CRANIOSYNOSTOSIS SURGERY?

Presenter: Susan Goobie, MD, FRCPC

Authors: Goobie S, Zurakowski D, Busa K, Proctor M, Meara J, Rogers G

Boston Childrens Hospital

Infants undergoing craniostyostosis surgery are at risk for significant postoperative events. We conducted a retrospective chart review to investigate patient factors which may increase/decrease the risk for these events & thus influence the decision re the need for postoperative ICU.

Methods: With IRB approval, the records of infants undergoing open craniostyostosis repair between 2002-12 at Boston Childrens Hospital were reviewed.

Results: 225 successive patient charts were analyzed. The median age of surgery was 12 mos (IQR 10-26). The avg wt was 11.6 +/-7.2kg. The median LOS in ICU was 1 day (IQR 1-2). 91% received PRBC intraop. 65% received Tranexamic acid (TXA). 4 major postoperative events which might require ICU stay were defined: major respiratory event (13%), major cardiac event (1.3%), postop PRBC transfusion (16%) for significant hypotension/severe anemia (Hct <24), and postop hemostatic agent (i.e. platelets, FFP, and/or cryoppt) transfusion to treat significant coagulopathy (13%). Demographic data & intraoperative factors were compared between those having a major postop event & those not. Univariate analysis identified 7 variables which were associated with major postop events; age, weight, intraop complication, intraop PRBC, albumin and/or hemostatic agent transfusion, and TXA administration (the latter having a protective effect). All 7 variables significant by univariate analysis were tested using multivariable logistic regression analysis to determine independent risk factors of a major postoperative event. Five variables are significant: lower body weight, intraoperative complication, intra-operative blood loss > 60ml/kg, postoperative hemostatic agent given, & TXA as a protective factor.

Conclusion: Infants undergoing craniostyostosis repair likely need an ICU bed postop if they are <10kg, have an intraoperative complication, have blood loss > half their total blood volume, or have FFP, platelets, cryoppt administered intraoperatively. TXA administration is associated with less postop major events & may reduce the requirement for ICU admission (1). Ref:1.Goobie et al. Anesth. 2011;114(4)

28P

A 35-YEAR EXPERIENCE WITH SYNDROMIC CLEFT PALATE REPAIR: OPERATIVE OUTCOMES AND LONG-TERM SPEECH RESULTS

Presenter: Marten N. Basta, BS

Authors: Basta MN, Silvestre J, Solot C, Cohen M, Kirschner RE, Low DW, LaRossa D, Jackson OA

University of Pennsylvania Perelman School of Medicine/ Childrens Hospital of Philadelphia

Background: Associated medical comorbidities and developmental delays place syndromic patients with cleft palate at risk for poor speech outcomes. Elevated rates of velopharyngeal insufficiency (VPI) have been reported, however few studies provide long-term follow-up. The purpose of this study was to describe one institution's long-term experience with syndromic patients undergoing cleft palate repair.

Methods: A retrospective review was performed of all patients with syndromic diagnoses who underwent primary palatoplasty at a large pediatric center from 1975-2011. Collected outcome measures included post-operative oronasal fistula and need for secondary surgery for VPI. Speech scores for verbal patients 5 years or older were collected via the Pittsburgh Weighted Values for Speech Symptoms Associated with VPI, and based on total score, outcomes were categorized as competent, borderline, or incompetent. Outcomes were analyzed by syndrome, association with Pierre Robin Sequence (PRS), Veau cleft type, and age at repair.

Results: 141 patients were included with average age at palatoplasty of 20.6 months (6-154). Composition of cleft type was 9% submucosal, 66% Veau classes I and II, 12% class III and 13% class IV. The overall oronasal fistula rate was 4.5%. PRS was diagnosed in 46 patients, of which 32 had Stickler syndrome, and 22q deletions in 18 patients. 88 patients (62.4%) had speech records available at a minimum of age 5, with average age at last assessment of 10.8 years (5 - 21). Overall, 49.1% of patients had a competent velopharyngeal mechanism and 11.3% required secondary VPI surgery. Patients with 22q deletions had the worst speech outcomes with 27.8% requiring secondary VPI surgery (p=.04). Patients with syndromic PRS and Stickler syndrome only had significantly better outcomes with 69.4% and 74.1% demonstrating a competent mechanism (p=.08, p=.05) and 13% and 15.6% rate of secondary VPI surgery, respectively.

Conclusion: This study highlights long-term outcomes in syndromic patients after palatoplasty and describes which factors are associated with worse post-operative outcomes in this population.



29P
**THIRTY YEARS OF PRENATAL CLEFT DIAGNOSIS:
WHAT HAVE WE LEARNED?**

Presenter: Jordan P. Steinberg, MD, PhD
Authors: Steinberg JP, Gosain AK
Lurie Childrens Hospital of Chicago

Purpose: Since the first description of prenatal ultrasound diagnosis (PUSD) of cleft lip \pm palate (CL \pm P) in 1981, many reports have detailed detection rates and accuracy. We review current knowledge of PUSD so as to help clinicians interpret the biases in prior reports and to utilize the information toward improved counseling.

Methods: A PubMed search was performed to determine estimates of cleft incidence as well as to review reports of PUSD of CL \pm P.

Results: We infer the following: (1) the best estimates of cleft incidence today from Scandinavian studies are consistent with historical estimates of 1:1000 live births with multifactorial inheritance; (2) reports may grossly underestimate incidence due to exclusion of fetal demise or pregnancy terminations; (3) original reports of PUSD in high-risk populations indicate frequent associated anomalies (> 60%), but rates in low-risk screened populations are more consistent with classical estimates (33% of CL \pm P, 50% of CP alone); (4) in the absence of concurrent structural anomalies, karyotypic errors are rare; (5) rates of detection (10-90%) are highly variable across studies; (6) reporting errors range from 10-60%, with most errors related to inaccurate characterization of the secondary palate; (7) pregnancy termination for isolated cleft remains rare, but data from underdeveloped nations are lacking.

Conclusions: PUSD of CL \pm P has received little attention. Our review underscores the variability in PUSD and the potential for misinterpretation. Heterogeneity in screening methodology, sonographer experience, and technology contributes to large differences in detection rates. As evidenced in low-risk populations, cases of CL \pm P detected prenatally are not inherently associated with more chromosomal anomalies, obviating karyotyping in all cases. A lack of common databases linking pre- and postnatal records has impeded careful analysis of PUSD in many countries. Informed counseling on the basis of prospective data can have a major impact on cleft incidence, particularly in less developed nations where the incidence is subject to decline with misuse of PUSD.

30P
**POST-OPERATIVE PYREXIA IN CHILDREN
FOLLOWING TRANSCRANIAL SURGERY**

Presenter: Fateh Ahmad, FRCS(Plast)
Authors: Ahmad F, Qureshi H, Elserius A, Evans M,
White N, Rodrigues D, Nishikawa H,
Dover S, Solanki G
Birmingham Children's Hospital

WITHDRAWN



31P
VISUAL FUNCTION AND OPERATIVE MORBIDITY IN THE CORRECTION OF HYPERTELORISM: REPORT OF 31 CASES

Presenter: Jonathan A. Britto, MB, MD, FRCS(Plast)
Authors: Britto JA, Dunaway DJ, Abela C, Schweibert K, Hon K

Great Ormond Street Hospital for Children

The correction of hypertelorism (HPT) requires translocation of the effective orbit in pursuit of improved facial balance. We quantify peri-operative morbidity and visual risk in a series of 31 orbital translocations.

Methods: Complete clinical records were reviewed 31 HPT patients (range 2-20 years), from a range of diagnostic groups (CFND 22, FC 6, others 3). Complete specialist orthoptic evaluations were available in 15 patients, within the wider serial 'craniofacial assessment'. Visual outcome and perioperative morbidity were documented.

Results: Orbital translocation was undertaken by frontofacial bipartition (13) or box osteotomy (18). Adverse events were assessed as major (n=2; major haemorrhage), moderate (n=3; CSF leak, bone loss), intermediate (n=3; re-operation), and minor (n=6; dressings/infections).

Four of fifteen patients with complete orthoptic record had preoperative convergent squint, maintained postoperatively, of which one required re-exploration for entrapment. 5/15 patients maintained a small angle exotropia (divergence) postoperatively. 2/15 patients reduced a large angle exotropia by HPT correction, facilitating strabismus surgery for aesthetic purposes.

4/15 patients had preoperative visual fusion for effective binocularity and depth perception (stereopsis). Three maintain postoperative stereopsis of which one, following a temporary postoperative entrapment and horizontal diplopia has required temporary adjuvant prism-lens rehabilitation and is now lens free. The fourth patient with entrapment and vertical diplopia has required functional strabismus surgery.

Discussion: HPT correction carries major to moderate risk in 16% of cases. Binocularity cannot be 'rescued' in patients with no fusion history. Visual fusion may be present in a third of HPT patients, posing high risk for visual disruption. In these, postoperative entrapment with vertical diplopia requires early operative release, horizontal diplopia has wider tolerance for ophthalmic rehabilitation. Intra-operative strategies to reduce osteotomy 'step-off' and entrapment may reduce visual risk.

32P
USING PRINCIPAL COMPONENT ANALYSIS (PCA) TO QUANTIFY THE DEFORMITY IN CRANIOFRONTONASAL DYSPLASIA, SIMULATE CORRECTION AND AID SURGICAL PLANNING

Presenter: Nicola Bystrzonowski, MBBS, BSc, MRCS (Eng), MSc, DIC

Authors: Bystrzonowski N, Ponniah A, Dunaway D
Great Ormond Street Hospital

Introduction: Craniofrontonasal dysplasia (CFND) is an X-linked genetic condition, resulting from mutations on the *EFNB1* gene. The main manifestations include hypertelorism, bifid nasal tip, asymmetry of the face, frontal bossing, craniosynostosis (in females).

Aims: Using a mathematical model to describe the changes between a CFND skull and a normal skull and to use the differences to visually guide clinicians in planning corrective surgery.

Methods: The Great Ormond Street Hospital database found 7 patients who had complete pre-operative three-dimensional CT (3DCT) scans, four of whom were age matched (12-17 years). These scans were compared to 40 dry skulls and soft tissue face scans using 87 anatomical landmarks. Principle component analysis (PCA) based on the differences between normal and CFND skulls allows a mathematical description of abnormalities seen in CFND. Thin plate spline movies demonstrated the corrective effects of facial bipartition box osteotomies.

Results: PCA demonstrated that patients with CFND demonstrated true hypertelorbitism. There was a consistent outward rotation of the orbit and in general orbital height was reduced. CFND patients demonstrate greater asymmetries than the normal population.

Choice of corrective procedure depends on the degree of deformity. Generally, bipartition was the procedure of choice. Box osteotomies were more effective at treating asymmetries, particularly where there was a vertical component.

Conclusion: PCA based on geometric morphometrics usefully describes the anatomical anomalies of CFND, aids surgical planning and the assessment of surgical outcome. Facial bipartition is the procedure of choice to correct these deformities except where there are significant asymmetries.



33P
**LONG-TERM OUTCOMES FOLLOWING EXTENDED
SAGITTAL SYNOSTECTOMIES**

Presenter: Gary F. Rogers, MD
Authors: Seruya M, DeFreitas T, Myseros JS, Yaun AL,
Rogers GF, Keating RF
Royal Childrens Hospital Melbourne

Background: While approaches to surgical correction for sagittal synostosis are variable, ranging from endoscopic to total calvarial reconstruction and include extended synostectomies (Pi procedure), there is a paucity of objective long-term outcome data. We reviewed our experience for isolated sagittal synostosis over 14 years at a single institution to evaluate objective criteria for improvement and longevity of correction.

Methods: Infants with isolated sagittal synostosis who underwent Pi procedures from 1997-2011 at a single institution were evaluated by pre and postoperative cephalic indices, head circumference percentiles, and morbidity/mortality.

Results: Thirty of 79 infants (38%) who underwent Pi procedures had both pre and postoperative cephalometric data over an average follow-up of 45.0 months. Average surgical age was 5.3 (2.6- 11.5) months. Operative duration was 1.4 ± 0.4 hours and hospital length of stay was 2.3 ± 0.5 days. Average cephalic index preoperatively was 68.7 ± 4.1 , which significantly improved to 72.8 ± 4.4 postoperatively ($p < 0.001$) and remained constant over follow-up. Median head circumference preoperatively was at the 90.5 percentile, stabilizing to the 89th percentile postoperatively ($p = 1.00$). 5/79 patients (6.3%) underwent a major reoperation between 12-60 months after the primary surgery; indications included scapho-trigonocephaly ($n=2$), biparietal narrowing, frontal/occipital bossing, and intracranial hypertension. There were no minor reoperations or mortalities.

Conclusions: Extended sagittal synostectomies (Pi procedures) secure appreciable correction of scaphocephaly that persists over time. This is concomitant with a stable head circumference. In the setting of a low reoperation rate and no appreciable morbidity or mortality, the Pi procedure should be considered a credible surgical therapeutic option for scaphocephaly.

34P
**MORPHOMETRIC ANALYSIS OF CORTICAL GRAY
MATTER IN INFANTS WITH ISOLATED METOPIC
CRANIOSYNOSTOSIS**

Presenter: MyChi H. Le, MD
Authors: Le MH, Aldridge KJ, Christ SE, Luo Y,
Aviles DL, Cole KK, Muzaffar AR
University of Missouri Columbia

Background: Craniosynostosis (CSO) has been hypothesized to constrain the growth of the underlying brain. This is thought to result in the region of the brain adjacent to the prematurely fused suture being predominantly affected. Previous work has shown altered morphology of the brain in infants with single suture CSO. Following this hypothesis, isolated metopic synostosis (IMS) should be associated with reduction of the underlying frontal lobe. In this study, we test two hypotheses: 1) overall brain size is reduced in IMS, and 2) frontal lobe size is reduced in IMS.

Methods: Our study sample consists of magnetic resonance images (MRIs) from infants between 30 and 64 weeks old, including infants with IMS ($N=10$) and age-matched unaffected infants (UN, $N=10$). Three-dimensional reconstructions of the surface of the brain were produced from the MRIs. Volumes of the whole brain (WBV) and total cerebral cortical gray matter (CXV) were measured from these reconstructions. Frontal lobe gray matter (FCV) was then manually segmented from the CXV volume. These volumes were then statistically compared between IMS and UN using Mann-Whitney U tests.

Results: WBV and CXV are slightly reduced in IMS relative to UN ($<5\%$), though these differences are not statistically significantly different. FCV is slightly increased in IMS (approximately 8%). However, when FCV is considered as a percentage of CXV, it does not differ from that of the UN group (34.7% and 35.2% of CXV, respectively).

Conclusions: Our results suggest that overall brain size and total cortical gray matter is slightly reduced in infants with IMS. However, the proportion of gray matter that comprises the frontal lobe appear similar in infants with IMS and UN. These results indicate that premature fusion of the metopic suture affects the overall shape of the brain, but it does not affect the relative size of the frontal lobe gray matter. These findings challenge the conventional wisdom that CSO results in restriction of growth of the underlying brain in the region of the fused suture. Further investigation into the functional implications of this finding is warranted.



35P
TRIGONOCEPHALY - TREATMENT USING DYNAMIC CRANIOTOMY AND EXPANDER SPRINGS

Presenter: Anderson Souza, MD
Authors: Souza A, Silva AS, Figueiredo EG, Cardim VL

Hospital Beneficencia Portuguesa de Sao Paulo

Summary: Introduction-The premature closure of the metopic suture causes a lateral-lateral fronto temporal depression causing a “delta” deformity which is called trigonocephaly. The incidence of trigonocephaly has increased and is currently the second most common type of craniosynostosis. The treatment, usually performed within one year of age with the advancement and frontorbital remodeling has received modifications in an attempt to reduce its morbidity. The use of bone distraction with springs expander inserted in the sides of the sphenoid wing offers a proposition to reduce the morbidity of this procedure. This work demonstrates the use for craniotomy and dynamic expander springs in the treatment of a case of trigonocephaly.

Method: Case report of a seven months old male patient diagnosed with moderate trigonocephaly without deficits of development and normal neurological examination, no indirect signs of intracranial hypertension appeared in tomography. Underwent Nautilus shaped spiral osteotomy in fronto-temporo-parietal bilateral and placement of expander springs in the region of Pterio at both sides.

Results: There was a improvement in the patient cranial shape, increased frontal angle (angle between two lines through Pterion and Nasio) and normalization of the ratio between the distance and interparietal intercoronal. There were no postoperative complications.

Conclusion: the use of osteotomy dynamics associated with the use of springs expander allowed indirect cranial remodeling is showing promise in the treatment of trigonocephaly.

36P
A THREE DIMENSIONAL ANALYSIS OF NASAL AESTHETICS FOLLOWING LE FORT I ADVANCEMENT IN PATIENTS WITH CLEFT LIP AND PALATE

Presenter: Edward H. Davidson, MA (Cantab) MBBS
Authors: Davidson EH, Miele LF, Kumar AR
University of Pittsburgh

Background: This study evaluates changes in nasal aesthetics using 3D photography after Le Fort I advancement osteotomies (single- or two-piece) in patients with non-syndromic cleft-related maxillary hypoplasia.

Methods: Cephalometric parameters were recorded pre- and postoperatively (overjet, overbite, soft tissue advancement). 3D photogrammetric imaging (Vectra XT Imaging System, Canfield Scientific, Inc., Fairfield, NJ) analyzed changes in nasal interalar width (IAW), internostril width (INW), nasal tip projection (NTP), columellar length (CL), nasal labial angle (NLA), and nasal length (NL). Statistical significance between pre- and postoperative data was determined using T-tests for each parameter.

Results: Eleven patients underwent either single piece Lefort I osteotomy and advancement, (3 bilateral, 4 unilateral cleft lip and palate), or two-piece advancement (2 bilateral, 2 unilateral). Average age at orthognathic surgery was 18.1 years. Average follow-up was 5.1 months. Average cephalometric advancement at maxilla central incisor edge was 6.4mm. Average nasal soft tissue changes were IAW 1.9mm (0.4-4.2), INW -0.2mm (-2.8-1.6), NTP -1.0mm (-4.0-2.0), CL -0.7mm (-2.9-1.5), NLA -0.2 degrees (-13.9-15.1) and NL -0.7mm (-4.3-1.5) (p=0.001, 0.6, 0.08, 0.01, 0.9, 0.2). For single-piece osteotomy alone changes were IAW 2.1mm (0.6-4.1), INW -0.6mm (-2.8-1.7), NTP -1.9mm (-4.0--0.3), CL -1.2mm (-2.9--0.03), NLA -1.3 degrees (-13.9-15.0) and NL -1.1mm (-4.3-0.7) (p=0.007, 0.3, 0.009, 0.0002, 0.7, 0.2). For two-piece osteotomy alone changes were IAW 1.6mm (-0.4-3.3), INW 0.5mm (0.4-1.6), NTP 0.5mm (-1.1-2.0), CL 0.2mm (-1.4-1.5), NLA 2.8 degrees (-7.6-10.1) and NL -0.1mm (-1.4-1.5) (p=0.2, 0.4, 0.5, 0.6, 0.5, 0.9).

Conclusions: Cleft-related scarring and malposition affect changes in nasal aesthetics following maxillary advancement. Patients with cleft lip and palate demonstrate a predictable increase in interalar width and decrease in columellar length as well as a trend to decrease nasal tip projection. Two-piece Lefort I increases variability of changes compared with single-piece advancement.



36P

AN INTERNATIONAL SURVEY OF CRANIOFACIAL SURGEONS: CURRENT TRENDS IN PRACTICE

Presenter: Patrick A. Gerety, MD

Authors: Gerety PA, Serletti JM, Taylor JA
University of Pennsylvania

Background: Craniofacial surgery is a diverse sub-specialty of plastic surgery that focuses on a wide range of head and neck pathology in children and adults. The purpose of this study was to define the characteristics of this group of surgeons and to compare sub-groups within the specialty.

Methods: A 36 question, anonymous, electronic survey was sent to 403 craniofacial surgeons; the response rate was 30% (121). Distribution was to members of the International Society of Craniofacial Surgeons (ISCFS) and to graduates of fellowships recognized by the American Society of Craniofacial Surgeons (ASCFS). Data was collected and analyzed for surgeon demographics, geography, practice setting (academic vs. private), case mix and volume, and career satisfaction. Comparisons were made between U.S. and international surgeons, males and females, and surgeons of different ages.

Results: Craniofacial surgeons in this study ranged in age from 29 to 75 years (mean = 53 years). 92% were male; 8% were female. They are largely academic (69%), high in academic rank (54% full professors), predominantly male (92%), and actively practicing craniofacial surgery. There are significant differences between international and domestic surgeons in terms of training background (64 vs 36% plastic surgery residency, $p=0.003$), and volume of craniofacial surgery (56 vs 26% performing >5 complex craniofacial procedures per year, $p=0.002$).

Conclusions: Craniofacial surgeons are a highly successful academic group with high career satisfaction. There are significant differences between U.S. and international craniofacial surgeons in terms of demographics and practice, with more U.S. surgeons performing fewer major craniofacial osteotomies. There is a significant gender disparity which warrants further study.

37P

SIMULTANEOUS ALVEOLAR FISTULA REPAIR AND BONE GRAFT DURING LEFORT I ADVANCEMENT IN CLEFT PATIENTS: IMPROVING OUTCOMES AND DECREASING MORBIDITY USING ALLOGRAFT ONLY BONE GRAFT

Persenter: Alex Rottgers, MD

Authors: Rottgers A, Miele L, MacIsaac Z, Kumar AR
University of Pittsburgh School of Medicine

Purpose: Cleft patients requiring maxillary advancement with persistent alveolar defects after failed prior bone grafting (ABG) present a unique reconstructive challenge. This study aims to demonstrate use of allograft bone alone for fistula repair at the time of Lefort I surgery is safe, efficacious, and avoids the morbidity of iliac crest harvest.

Methods: A 30-month (Jul 2010-Jan 2013) retrospective review of patients undergoing Lefort I advancement with simultaneous ABG using allograft bone was performed. Demineralized bone matrix with cancellous chips (DBX Mix, Synthes, Inc., West Chester, PA, USA) was used. Statistical significance of Enemark score improvement was assessed with the Wilcoxon Signed Rank Text.

Results: Ten patients (7 male, 3 female) underwent 11 Lefort I advancements with concurrent ABG of 15 alveolar gaps. All patients had nonsyndromic cleft-related class III malocclusion, previous failed ABG, and persistent alveolar fistulas. Seven two-piece and three one-piece Lefort I advancements were performed. An average 4.25 cc (2.5-7) DBX Mix was used per alveolar gap. Alloderm was implanted in two patients (20%). Average age was 19.35 (17-23) years, and radiologic follow-up was 6.2 (2-10) months. Average Enemark bone graft scores improved from 3.4 pre-op to 1.2 post-op ($p<0.0001$). Bone union was achieved in 9 (90%) patients and 14 alveolar bone gaps (93%). Bone graft and hardware extrusion occurred in 2 patients (20%); one patient was regrafted. A persistent alveolar fistula occurred in 1 patient (10% of patients; 7% of alveolar bone gaps). Revision Lefort I osteotomy was required in 1 patient (10%) due to relapse. No wound infections, bleeding requiring transfusion/reoperation, or mortalities occurred.

Conclusions: The use of allograft alone to reconstruct a persistent alveolar cleft at the time of Lefort I advancement is effective and provides a safe alternative to traditional bone grafting. The use of allograft alone is associated with low morbidity, significantly improved Enemark scores, low relapse rates, and high bone union rates in patients that have previously failed ABG.



38P

COMBINED CONTRACTION AND DISTRACTION OF THE FACIAL SKELETON IN STAGED TREATMENT OF AN ATYPICAL FACIAL CLEFT USING DISTRACTION DEVICES

Presenter: Robert D. Wallace, MD

Authors: Wallace RD, Alvarez S, Konofaos P
University of Tennessee Memphis

Atypical craniofacial clefts represent a complex and unique set of treatment challenges. The majority of these patients require multiple staged procedures over the course of their infant and childhood years to achieve some degree of facial harmony. There is no consensus on the optimal treatment algorithm for some of the more complex facial cleft. Included in the techniques and goals of this complex treatment algorithm are the needs of restoring the craniofacial skeleton and symmetric repositioning of key facial soft tissue landmarks while adhering to the aesthetic subunit principle. We present the case of a female born with a Tessier 2/12 craniofacial cleft, left anophthalmos and microtia. The first step towards the creation of facial harmony is the midline closure of the soft tissues. In order to accomplish this in this extremely wide facial cleft it was necessary to first more closely approximate the bony cleft. We describe a novel use of mandibular distraction devices in a reverse manner for the re-approximation of the craniofacial skeleton allowing for closure of the soft tissue component of this cleft. The changes in the bony portions of the cleft were accomplished without osteotomies taking advantage of the mobility of the facial bones involving both normal bony sutures and the cleft bone. The bone movement allowed for easier adhesion of the midline lower facial soft tissue during the first year of life. During the second stage traditional osteotomies and distraction were done to position the lip, nose, and palate for definitive closure. This new use of distraction devices in a reverse manner to contract the craniofacial skeleton without osteotomies to the authors knowledge has not been previously reported and adds a valuable tool to closing large facial clefts.

40P

RARE FACIAL CLEFTS: 6 CASES EACH A DIFFERENT CHALLENGE FOR TREATMENT

Presenter: Diego J. Caycedo, MD, MSc

Authors: Caycedo DJ, Cabal M
Universidad del Valle

The challenge presented by a rare facial fissure that compromises one or more tissues demands from the craniofacial surgeon precise determinations with a surgical plan for each particular case developing the capacity to face the comprehensive management of these patients. There are surgical patterns described in literary sources that can help determine a specific procedure, but in the majority of situations a technique that is based on experience and is able to solve the problem, is required. Nevertheless, it is imperative to also have the capacity to determine when nothing is to be done. We present a series with 6 patients with rare facial clefts, of different severity and clinical presentation, that where intervened and its evidenced the form how their medical and surgical treatment developed, the short and long term results, and specially when due to the complexity of the situation, no procedure was undertaken.



41P

8 TRANSVERSE CLEFT LIP REPAIR CASES - FOLLOW UP

Presenter: Penpak Krergmatukorn, MD

Author: Krergmatukorn P

Queen Sirikit National Institute of Child Health

During the last 4 years we repaired eight transverse cleft lip cases by a craniofacial plastic surgeon. The age of our patients when repaired was vary between 4 months old to 4 years and 3 months old. There are 2 cases that have bilateral transverse cleft. The technic was 3 layers repair: buccal mucosa, orbicularis muscle and outer vermillion/skin respectively. Wound closure appeared like lazy linear wound. When follow the results there was one case that we feel unsatisfied so we repeat the surgery 9 months later. This case was the one we repaired at more than 4 years old age. For the others,we and the parents accepted with the result.

We learn from our cases that careful measurement/design and muscle layer repair is an important factor to get a satisfied result. Repairing time at the age between 3-8 months may effect a good recovery of the orbicularis function...result in more favour outcome than late surgery.

42P

AGE-RELATED EFFECT OF MONOBLOC FRONTO-FACIAL DISTRACTION ON ORBITAL VOLUME, MORPHOLOGY, AND CLINICAL OUTCOME IN 29 CROUZON-PFEIFFER CASES: A CONTROLLED STUDY

Presenter: Jonathan A. Britto, MB, MD, FRCS(Plast)

Authors: Khonsari R, Karunakaran T, Ashraff S, Nysjo R, Nystrom I, Dunaway DJ, Evans RD, Britto JA

Great Ormond Street Hospital for Children

Monobloc frontofacial advancement by distraction (MBD) is a valuable strategy in the management of Crouzon-Pfeiffer syndrome (CPS) for functional and aesthetic gain. We ask whether MBD undertaken at infancy to adolescence achieves a stable volume expansion of the orbit in the treatment of symptomatic oculo-orbital disproportion.

Methods: CT radiologic data of 29 CPS patients (58 orbits, R=3m-17 yr), were assessed in Osirix software against that from 30 age – matched controls from a neurosurgical population. Parameters of globe volume, orbital volume, orbital morphology and globe position in the orbit were obtained by manual segmentation, verified by a mesh-based semi-automatic technique (Nystrom et al). Patient data included pre-operative, six-week post-MBD, and delayed postoperative (8-18 mnth) scans, correlated with contemporaneous ophthalmic examination findings.

Results: Control globe and orbit volumes are symmetrical. Volume increase of the orbit rises from 14-21cc aged 2-10 yrs and plateaus at 22cc aged >10 yrs. Control globe protrusion is symmetrical, constant, and independent of age. CPS orbits are more symmetrical of volume than shape. Mean unoperated CPS orbital volume is 10.7 mls (<10 years) rising to 17.9 mls (>10 yrs). Crouzon globe volume approximates control, but globe protrusion significantly exceeds control ($p<0.05$). Monobloc advancement expands CPS orbital volume by a mean of 84.5% at 6 weeks. ($p<0.05$) to approximate age-matched control. Globe protrusion reduces to control value. Orbital expansion and shape change is mostly maintained at 8-18 months and correlates with symptomatic relief. Delayed volume relapse, where it occurs, is a mean of 0.49 cm³ in 31% cases <10 yrs, and 0.26 cm³ in 19% cases >10 yrs, and is neither clinically nor statistically significant.

Discussion: Monobloc frontofacial advancement relieves syndromic oculo-orbital disproportion and globe protrusion to approximate normal. Surgical relapse characterises younger patients, nonetheless for whom functional necessity is paramount.



43P

CORRECTION OF CORONAL CRANIOSYNOSTOSIS FROM UCS TO KLEEBLATTSCHADEL

Presenter: Mckay Mckinnon, MD

Author: Mckinnon M

Lurie Childrens Hospital

Purpose: Coronal craniosynostosis presents a primary disturbance of the cranial base. Current treatment of coronal synostosis is controversial due to time of intervention, fixation of fragments, and use of distraction devices, internal and external. Recent studies report significant rates of resynostosis, reoperation and other complications. This study attempts to review one treatment, standard osteotomies and surgical reshaping of the involved skeleton, followed by helmet guided wire distraction, during a 15 year period.

Method: An IRB approved review was conducted of patients treated for coronal craniosynostosis between 1997-2012 at two institutions by one craniofacial surgeon. 56 patients underwent standard osteotomies and skeletal reshaping, plus use of distraction wire(s) attached to a custom helmet and bar. Distraction of about 12 days was followed by maintenance wire tension up until post-op day 30, at which point all wires were removed as an office procedure. Follow-up was between one and 12 years.

Results: Mean age at operation was 7 months. 14 of the 56 patients were documented as syndromic, eight were undetermined or had multiple suture synostosis (including coronal), and 34 patients had either unilateral or bilateral coronal synostosis. There were no deaths and no cases of resynostosis. Five patients underwent reoperation for only residual cranial defects by age four. Orthotists' helmet manufacture averaged less than \$3000.

Conclusions: Coronal craniosynostosis can affect the entire anterior cranial base and fronto-orbital skeleton, thus producing an alteration of normal craniofacial tension. Correction should: release the closed suture(s), normalize the cranial base shape, and restore normal bony position and craniofacial tension. The combination of standard osteotomies with bone remodeling and non-plate internal fixation, accompanied by short-term external helmet anchored wire distraction, has proven to be a simple, safe and effective treatment for both syndromic and non-syndromic cases of coronal synostosis. This treatment permits results which exceed previous reports.

44P

ORBITAL MORPHOLOGY IN APERT AND CROUZON-PFEIFFER SYNDROMES: AN AGE – MATCHED, CONTROLLED STUDY

Presenter: Jonathan A. Britto, MB, MD, FRCS(Plast)

Authors: Khonsari R, Way B, Matthews W, Nysjo R, Nystrom I, Evans RD, Britto JA

Great Ormond Street Hospital for Children

Oculo-orbital disproportion in FGFR2- related syndromic craniosynostosis tends to exorbitism and symptomatic palpebral incompetence. We report an age-matched, controlled study of orbital morphology and globe position in unoperated Apert (AS) and Crouzon-Pfeiffer (CPS) syndromes, resulting from distinct sub-types of activating FGFR2 mutation, and ask whether syndromic differences in orbito-facial morphology justify different approaches in orbital remodelling.

Methods: CT radiologic data from 29 unoperated Crouzon-Pfeiffer (CPS) patients (58 orbits, R=3m-17 yr), and 15 unoperated Apert (AS) patients (30 orbits, R=2-21 yr) were assessed against similar data from 40 age-matched control scans from a neurosurgical population. Globe volume, orbital volume, orbital morphology and globe position were obtained by manual segmentation with inter- and intra-observer control, verified by a mesh-based semi-automatic technique (Nystrom et al). Palpebral fissure width (PFW), intercanthal distance (ICD), and inter-dacryon distance (IDD) were measured, and all data correlated to patient genotype.

Results: Control globe and orbit volumes are symmetrical. Orbital volume rises from 14.21cc aged 2-10 yrs and plateaus at 22cc aged >10 yrs. Control globe protrusion is symmetrical, constant, and independent of age. Orbital volume in CPS is 10.7cc (<8 years) rising to 17.9cc (>8 yrs). Mean AS orbital volume is 22.9cc (11-21 yrs), excluding two 23 and 28 month old patients at 11cc and 20cc. Both AS and CPS orbits show greater symmetry of volume than shape, and globe protrusion significantly exceeds control. We have found PFW to be a reliable constant in orbital translocation surgery. ICD:PFW and IDD:PFW are equal in CPS and control, whereas AS demonstrates higher ratios characterising hypertelorism patients.

Discussion: Symptomatic exorbitism in AS and CPS does not equally result from orbital hypoplasia. Oculo-orbital disproportion in CPS reflects a hypoplastic orbit. Globe position in AS reflects genotype/phenotype specificity, with normal volume, and a tendency to hypertelorism. Surgical strategies are optimally directed in a bespoke manner.



45P
**CLOSURE OF THE SPHENO-OCCIPITAL
SYNCHONDROSIS IN PATIENTS WITH CROUZON
SYNDROME: A LINK TO MIDFACE HYPOPLASIA**

Presenter: Jesse A. Goldstein, MD
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Purpose: Premature fusion of the spheno-occipital synchondrosis (SOS) has recently been described in the syndromic craniosynostosis subpopulations with Apert syndrome and Muenke syndrome. The current study aims to characterize SOS fusion in patients with Crouzon syndrome, a syndrome with high rates of midface hypoplasia.

Methods: A retrospective case-control study was performed in patients with Crouzon syndrome treated at a large craniofacial center between 1984 and 2012. Inclusion criteria were a diagnosis of Crouzon syndrome and at least one high-quality CT scan in which SOS patency could be assessed. Age/gender matched control CT scans were identified and assessed for status of SOS patency. Three independent reviewers with high inter-rater reliability ($\kappa=0.88$) graded SOS patency on axial images as open, partially fused, or completely fused SOS. Wilcoxon Rank-Sum test was used to compare the Pfeiffer group to controls.

Results: Over the study period, 30 patients were identified with Crouzon syndrome. A total of 24 patients with 112 head CT scans met inclusion criteria. All patients with Crouzon syndrome had some degree of midface hypoplasia. Accordingly, 112 age/gender-matched control CT scans were assessed, and no patient in the control group had midface hypoplasia. Within the Crouzon group, the average age of complete closure (14.33 ± 3.43 years; $n=31$) evident on CT scan was significantly younger than the control group (16.55 ± 2.15 years; $n=18$) ($p=0.0155$). The average age of partial closure evident on CT scan was significantly younger (5.57 ± 2.04 years; $n=43$) within the Crouzon group compared to the control group (10.65 ± 2.44 years; $n=18$) ($p=0.0001$). The average age of scans showing complete patency of the SOS in the Crouzon group (1.32 ± 1.07 years; $n=38$) was significantly younger than the control group (3.22 ± 2.30 ; $n=76$) ($p=0.0001$).

Conclusions: The SOS closes significantly earlier in patients with Crouzon syndrome compared to age/gender-matched controls. Although causality cannot be concluded, there exists a strong correlation between midface hypoplasia and premature SOS closure in Crouzon syndrome.

46P
**HYDROCEPHALUS AND BASE SKULL
ABNORMALITIES IN FGFR 2 CRANIOSYNOSTOSIS**

Presenter: Coll Guillaume, MD
Authors: Guillaume C, Di Rocco F, Brunelle F,
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Subject: Hydrocephalus and tonsillar prolapse (TP) are commonly found in syndromic faciocraniosynostosis. We analyzed the correlations between the abnormalities of the skull base and the associated cerebrospinal fluid disorders found in children with FGFR 2 mutations (Crouzon, Apert and Pfeiffer syndromes).

Patients and Methods: We compared a group of 31 children under 2 years of age with craniofacial synostosis due to genetically confirmed mutation of FGFR 2 (14 children with Crouzon syndrome, 11 with Apert syndrome and 6 with Pfeiffer syndrome) to a control group of 17 children without craniofacial abnormalities. The area of Foramen Magnum (FM), the posterior fossa volumes (PFV), cerebellum (CV) and jugular foramen areas (JF) were studied on millimeter CT scan slices performed prior to any surgery.

Results: The patients with Crouzon syndrome and Pfeiffer had a small FM (area and diameter) ($p < 0.05$). All syndromic patients had small JF. There were no difference for the PFV and CV between control children and syndromic children. There were no hydrocephalus in the Apert group. Hydrocephalus was associated with a small FM ($p < 0.05$). Hydrocephalus and PT were correlated ($p = 0.02$).

Discussion and Conclusion: Our study shows that, in FGFR2 related faciocraniosynostosis, the area of the FM seems to be a determining factor in the formation of hydrocephalus and not the PFV or JF diameters.



47P
MONOBLOC FRONTO FACIAL ADVANCEMENT WITH DISTRACTION OSTEOGENESIS FOR SYNDROMIC CRANIOSYNOSTOSIS: COMPARISON BETWEEN 2 DISTRACTORS

Presenter: Cassio E. Raposo-Amaral Sr., MD, PhD
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SOBRAPAR

Introduction: Distraction Osteogenesis revolutionized the treatment of syndromic craniosynostosis. Less complication rate and decreased morbidity were observed with monobloc fronto facial advancement with distraction osteogenesis technique. The objective of this study was to compare the role of two types of distractors as an adjuvant in the monobloc fronto-facial advancement.

Methods: Eight patients with syndromic craniosynostosis underwent to monobloc fronto facial advancement with distraction osteogenesis and were divided into 2 groups. In group I (n=4), the advancement was performed using an external univector device. The mean age of group I was 10.5 years. The mean follow up of group I was 11 years. In group II (n=4), the advancement was performed using the internal univector device. The mean age of group II was 8.7 years. The mean follow-up of group II was 3 years. The cephalometric tracings were analyzed according to Ricketts cephalometric standards.

Results: The average midface advancement in group I was 18.3 mm. The cephalometric variation in group I was: SNA: 70, SNB: -30. The average midface advancement in group II was 10.7 mm. The cephalometric variation in group II was SNA: 8.10, SNB: -3.40. In both groups, all patients end up with temporary enophthalmia and class II occlusion. In groups I and II, patients changed from class II to class I during the follow-up period.

Conclusion: All patients had satisfactory results. Exorbitism, maxillary retrusion and breathing impairment were adequately corrected by monobloc fronto facial advancement with distraction osteogenesis technique, regardless of the device used. Higher midface advancement was needed in group I, probably because the lower bone stability generated by the first device.

48P
TREATMENT AND OUTCOME OF FIBROUS DYSPLASIA INVOLVING THE ORBIT: THE ROLE OF NEW TECHNOLOGIES

Presenter: Jordi Puente-Espel, MD
Authors: Puente-Espel J, Barcelo CR, Genecov DG
International Craniofacial Institute

Classically, fibrous dysplasia (FD) involving the orbit has been treated with either resection and primary reconstruction or conservative debulking. The objective of the study was to evaluate how current technologies impact the planning and prediction of treatment and reconstruction in patients (pts) with FD involving the orbit.

Patients and Methods: A retrospective analysis of 16 pts (F: 5, M: 11; age 3-41 yr, avg 18 yr), 2004 - 2012 period, at the International Craniofacial Institute, is provided. Monostotic (MFD) 10 pts, polyostotic (PFD) 6 pts. The areas involved were documented according to the Chen-Noordhoff classification. We divided pts in 2 groups (gp): gp1, 12 pts who underwent radical resection and primary 3-D reconstruction; gp2, 4 pts submitted to conservative debulking.

Preoperative planning used: stereolithographic models [Medical Modeling Inc, CO], computer planning [Med Cad Inc, TX] and reverse templates [Med Cad Inc]. Intraop support used: CRS reinforced bone cement [Synthes, PA], demineralized bone graft [Skye Orthobiologics, CA] and surgical navigation [Stryker navigation system, MI].

Results: In gp1, 58% of pts achieved complete excision; 6 were submitted to optic nerve decompression (OND). 3/12 showed residual deformity, 2 required secondary surgery. Gp2, 4 pts with PFD; 1 underwent OND.

The orbital reconstruction outcome was analyzed according to 3 functional (permanent diplopia, permanent visual acuity deficits and enophthalmos) and 3 aesthetic variables (bossing, indentations and dystopia). In gp1, 1 pt (8.3%) appeared mildly enophthalmic postoperatively. 3 pts (25%) showed temporary decreased visual acuity, 2 (16%) with temporary diplopia, 1 pt (8.3%) a 2 mm proptosis. No long-term deficits in gp2. Aesthetic outcome: in gp1, 1 pt with indentations (8.3%), 1 pt with FD regrowth. 3 pts in gp2 showed orbital/forehead bossing.

Conclusions: The proper use of current preop and transop technologies provides the craniofacial surgeon with the adequate tools to extend the limit of resection, thus, pts with MFD and selected PFD can be approached with total resection and primary reconstruction.



49P
**MAXILLARY DISTRACTION IN THE PATIENT WITH
CLEFT LIP AND PALATE ANOMALIES: LESSONS
LEARNED IN 74 CONSECUTIVE CASES**

Presenter: Jordi Puente-Espel, MD

Authors: Puente-Espel J, Genecov DG, Barcelo CR
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The objective of the following study was to study long-term stability and relapse rates [RR] in unilateral CLP [UCLP] and bilateral CLP [BCLP] patients [pts] with class III malocclusion submitted to internal and external distraction osteogenesis [DO].

Material and Methods: A retrospective study of 74 pts with UCLP or BCLP, treated at the International Craniofacial Institute, Dallas, TX, from 2004 to 2012. All the pts had any of the following indications for Lefort 1 with DO: severe overjet (>12 mm negative overjet), obstructive sleep apnea [OSA] in pt with maxillary deficiency, maxillary advancement >12 mm, re-operative maxillary surgery in presence of severe scarring of the maxilla. Two groups (gp) were created: gp 1, pts with internal distraction devices and, gp 2, pts with external distraction devices.

The results were evaluated based on cephalometric analysis (before, immediately after DO and at 1 year follow-up); mandibular growth [MG] was measured as well. Gp 1, 42 pts: ages 6.2-25 yr, mean 14.3 yr; M:31, F:10; UCLP: 32 and BCLP: 10. Pts in this group had a negative overjet of 8-21 mm (mean 12.7 mm) and an advancement of 11-27 mm (mean 15.7 mm). Gp 2, 32 pts: ages 8-45 yr, mean 15.1 yr, F: 5 M: 10; UCLP: 20 and BCLP: 12. A negative overjet of 5-14 mm (mean 8.7 mm) and an advancement 8-22 mm (mean 14.2 mm) was shown.

In gp 1, an overcorrection of 2-10 mm (mean 5.5 mm), consolidation period [CP] 6-14 wks (mean 9.5 wks), RR: 0-14 mm (mean 1.3 mm [9.1%]), MG: 0-11 mm (mean 3 mm), occlusion class I-III; and non union in 4/42 pts. Gp 2 demonstrated an overcorrection of 2-5 mm (mean 3.2 mm), CP: 4-12 wks (mean 6.2 wks), RR: 1-10 mm (mean 2.1 mm [10.3%]), MG: 0-4 mm (mean 1.4 mm), occlusion class I-III; and non union in 2/32 pts.

No significant difference in sex, cleft type or initial negative overjet. Significant difference ($p<0.05$) in distraction distance, overcorrection, consolidation period, presence of alveolar ridge bone grafting and age. The fact that internal or external distraction devices are used, if an appropriate consolidation period is followed, does not have long term effects in the outcome.

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