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New Pediatric Subspecialty

Opening of the Craniofacial, Plastic and Reconstructive Surgery Center at Children's Hospital of Austin

By Patrick Kelley, MD, Director

Craniofacial surgery is a relatively new subspecialty of plastic surgery that focuses on reconstruction of the head and face. The specialty was born within the past 25 years, after Paul Tessier, a French surgeon, developed a number of surgical techniques that transcended traditional boundaries between neurosurgeons, ophthalmologists, otolaryngologists

and plastic surgeons. Plastic and reconstructive surgeons around the world immediately embraced his revolutionary advances, and the field has blossomed since its induction.

At the core of craniofacial surgery is osteotomies of the cranium and face that allow movement and rearrangement of bony structures that are malformed, malpositioned or deformed by cancer or trauma.

In its infancy craniofacial surgery dealt with congenital malformations of the head and face like Crouzon, Pfeiffer and Apert syndromes. These syndromes are marked by early fusion of skull sutures (craniosynostosis) and severe midface hypoplasias. (Figures 1A-1C) These complex patients develop early intracranial hypertension; if left untreated, it leads to significant neuropsychiatric consequences including mental retardation. Additionally,

the severe midface hypoplasia contributes to difficulties with exorbitism with the consequence of corneal exposure, upper airway obstruction (tracheostomy dependence or severe sleep apnea) and difficulties with oral intake and speech.

As craniofacial surgeons have refined the techniques of cranial and facial osteotomies, these techniques have found further use in the treatment of other congenital malformations. Because of the close association of craniofacial surgery with congenital malformation and pediatrics, many craniofacial surgeons in the United States have become defacto pediatric plastic surgeons involved in the treatment of malformation and maladies throughout the body, as well.

With the tremendous advances in CT scanning technology, we are now able to assess the cranial sutures as well as cranial and facial form in much greater detail with less radiation exposure to the patient. New spiral CT scanners available at Children's Hospital of Austin also have the additional advantage of being able to acquire a complete high-resolution scan within seconds, which greatly reduces movement artifact as well as limits the amount of sedation required for many patients. (Figure 2)

The principles of craniofacial surgery also have led to tremendous advances in the treatment of facial trauma with the advent of rigid fixation techniques and the concepts of early, aggressive open reduction and internal fixation. Additionally, we have learned that some of the cranial and facial osteotomies used in treatment of malformations provided incredible access to regions previously considered

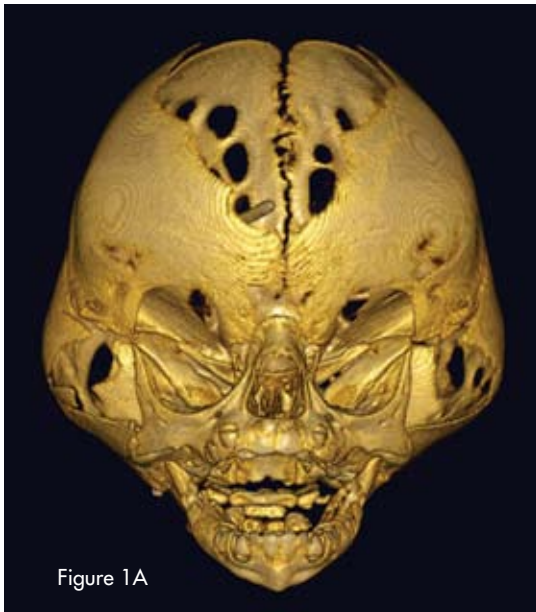


Figure 1A

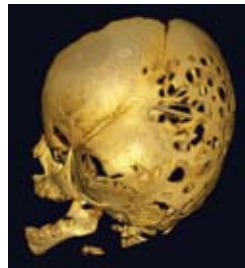


Figure 1B

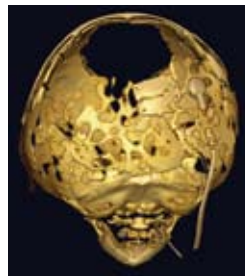


Figure 1C

Figures 1A, 1B and 1C

1A-1B. Patient with Crouzon Syndrome at three months of age developing bilateral coronal and bilateral lambdoid craniosynostosis with the classic "clover leaf" skull deformity. The "swiss cheese" appearance of the skull is caused by elevated intracranial pressure as brain growth exceeds that of the skull with the multiple fused sutures. The pulsating brain literally wears holes into the overlying bone. Note the associated midface hypoplasia that leads to severe exorbitism (proptosis) and corneal exposure.

1C. Posterior view of skull after first stage cranial vault expansion of the posterior cranium. The second stage completed several months later focuses on expansion of the orbits and forehead.



Figure 2

Patient has a rare form of plagiocephaly ("crooked head") related to fusion of the intraoccipital synchondrosis seen on the posterior view of the skull. Right arrow indicates fused synchondrosis and left arrow indicates normal side. Advanced CT scanning allows for evaluation of cranial base sutures in detail that has not been possible until now. Most patients with minor suture fusions will not need surgery, but should be followed closely.

unapproachable for surgical intervention. We are now capable of reliably resecting tumors at the base of the anterior and middle cranial fossae, which were previously considered



Figure 3A



Figure 3B

uncureable.

The newest advancement in craniofacial surgeons' armamentarium is the technique of distraction osteogenesis. With bony distraction techniques, we are now able to achieve advancements of the facial bones and jaws well beyond that of traditional techniques. This has allowed us to prevent tracheostomy in the neonatal period for patients born with hypoplastic mandibles like Pierre-Robin Sequence. This is achieved by advancing the lower jaw forward within a matter of days from the time of birth. We are also able to satisfactorily treat patients with severe midface retrusion, especially patients with cleft lip and palates with a single staged jaw advancement as opposed to

multiple jaw procedures as previously required. (Figures 3A-3D)

Because craniofacial surgery involves so many regions of the head and neck, we regularly interact with the other specialists in this region. We rely heavily on the expertise of neurosurgeons, ophthalmologists, head and neck

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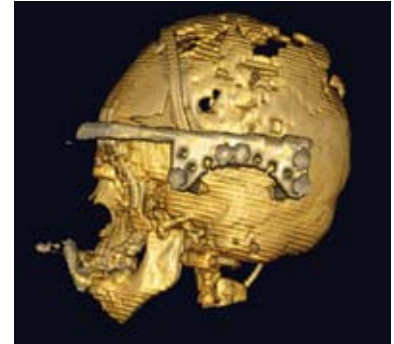


Figure 3D

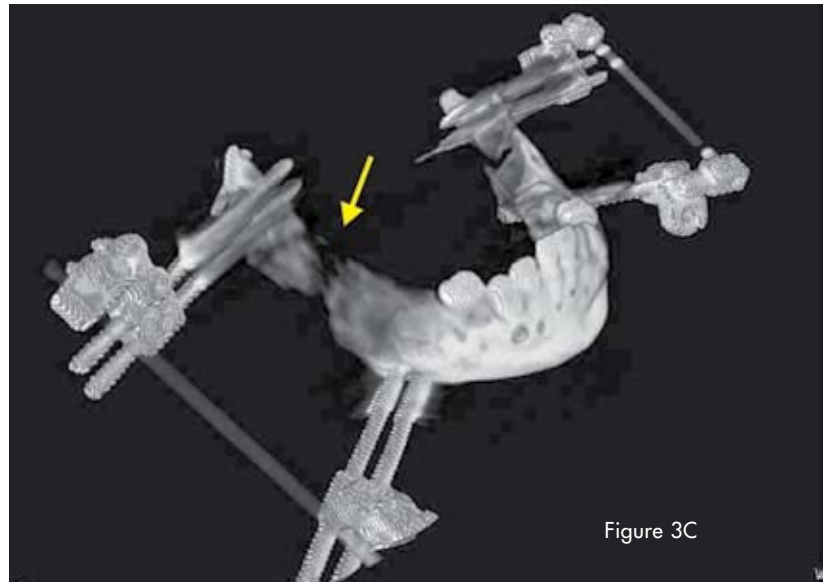


Figure 3C

Figures 3A, 3B, 3C and 3D

3A. Patient born with Goldenhar Syndrome with congenital micrognathia and airway obstruction secondary to retropositioning of the base of tongue (glossoptosis). Although the patient did not present sufficiently early in life to avoid tracheostomy, bilateral mandibular distraction is performed to advance the lower jaw and allow removal of the tracheostomy.

3B. AP CT showing mandibular distractors in place.

3C. Close-up view of mandible showing generated bone in distraction gap (arrow). Distraction osteogenesis involves performing a bony cut followed by slowly moving the ends apart from one another. New bone is generated in the gap creating a larger mandible made from the patient's own tissues.

3D. Patients with severe midface hypoplasia often require movement of the entire face (Lefort III) or entire face and forehead (Monobloc). This patient is seen immediately post-op after a Monobloc operation with the external distraction device used slowly to bring the face and forehead forward over a couple of weeks. This device is amazingly well-tolerated. This procedure resolves symptoms related to ocular exposure, airway obstruction, elevated intracranial pressure, speech and oral intake all in one operation.

The center offers multidisciplinary expertise in the treatment of complex congenital malformations of the head and neck including:

- Comprehensive cleft lip and palate treatment
- Presurgical molding (“NAM”)
- Speech disorders (velopharyngeal insufficiency)
- Plagiocephaly
- In-utero or postnatal positional molding
- Craniosynostosis (premature fusion of skull sutures) (Figures 4A-4B)
- Torticollis
- Complex neonatal airway treatment (Pierre-Robin)
- Sleep apnea related to jaw deformities and upper airway obstruction
- Cranial and facial clefting syndromes (Amniotic Band Syndrome)
- Dentofacial malocclusions including severe “under- and over-bites”
- Occlusal disorders and chin deformities
- Eyelid and orbital deformities (blepharophimosis)
- Common and complex external ear deformities
- Treacher-Collins
- Marshall-Stickler, Stickler, Binder
- Velocardiofacial syndrome
- Van der Woude
- Crouzon, Apert, Pfeiffer, Jackson-Weiss, Saethre-Chotzen
- Hemifacial Microsomia (Goldenar Syndrome)

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surgeons (otorhinolaryngologists) and oral surgeons. These physicians have expertise in their respective organ systems that is well beyond that of the craniofacial surgeon and as such the craniofacial surgeon often finds himself or herself relying heavily on their expertise. Likewise, the genes for many of the craniofacial malformations have been identified and craniofacial surgeons have contributed to these advancements, but by no means do craniofacial surgeons consider themselves geneticists.

Children’s Hospital, which will become the Dell Children’s Medical Center of Central Texas in spring 2007, has made a tremendous commitment to the development of a craniofacial

center to service and support the Central Texas region. The center will coordinate the comprehensive needs of these complex patients including comprehensive orthodontic care through a full-time craniofacial orthodontist as well as care from medical specialists including neurosurgery, otorhinolaryngology, ophthalmology, speech therapy and audiology. The Craniofacial Center is a truly multidisciplinary center coordinating care of patients from birth through



Figure 4A

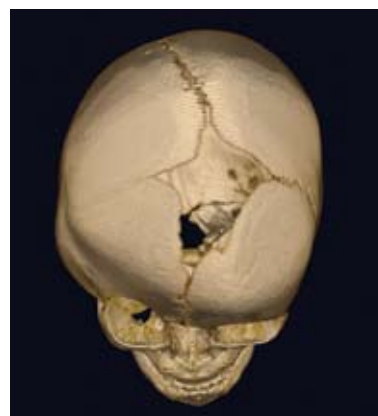


Figure 4B

Figures 4A-4B

Patient with right unicoronal craniosynostosis with the associated fusion of the right coronal suture, retrusion of the ipsilateral brow and forehead, bulging of the ipsilateral temporal region and deviation of the facial structures, particularly the root of the nose toward the synostosis. Patients with craniosynostosis are capable of developing elevated intracranial pressure and should be monitored closely and ideally treated within the first year of life.

adulthood. We offer neonatal feeding and complex airway assessment consultations at the time of birth and counseling to patients as they begin to transition to independent living.

To refer patients to the Craniofacial Center, please contact Mary Breen, Craniofacial Clinical Coordinator, or Patrick Kelley, MD, Director, Department of Craniofacial, Plastic and Reconstructive Surgery, at 512-324-8000.