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Maximizing Outcomes of Pediatric Craniofacial Surgery

· Craniosynostosis

· Craniofacial Microsomia

Assistant Professor
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FUNDING

NIH

Craniofacial surgery involves the treatment of congenital deformities of the head and face, such as craniosynostosis, cleft lip and palate, and craniofacial microsomia as well as acquired deformities of the face through trauma or tumor extirpation. Our research focuses on the outcomes of our treatments for patients with craniosynostosis, cleft lip and palate, craniofacial microsomia, and facial trauma.

Craniosynostosis

Craniosynostosis is the premature fusion of one or more cranial sutures. These sutures are vital for normal development of the rapidly growing infant brain. Premature fusion can occur sporadically, or as part of an inherited syndrome and carries with it the risk of elevated intracranial pressure, developmental delay and blindness. Treatment options vary, but generally involve expansion of the cranium. Our research focuses on the outcomes of these treatments.

SAFETY

We are currently analyzing the safety of the surgical approaches offered at Seattle Children's Hospital. Factors such as blood loss, intraoperative events and post-operative complications are analyzed along with anesthesia protocols to establish the safest practice. We have found that early transfusion of blood and plasma have improved the safety of these procedures and shortened the length of hospital stay.

NEURODEVELOPMENT

An important outcome measure in craniosynostosis surgery is the child's neurocognitive development. Coupled with our research into the safety of our procedures is development of the patients after surgery. This project seeks to determine the effects of anesthesia, surgery, and post-operative complications on the development of children during the first decade after birth. Neurocognitive tests are administered at various time points during childhood and compared to controls to determine the effects of craniosynostosis and its surgical treatment.

MORPHOLOGY

One component of the surgical treatment of craniosynostosis is alteration of cranial morphology. Abnormal skull shape is a hallmark of craniosynostosis, and altering the shape and establishing a normal morphology is one goal of our surgical correction. Our outcomes research includes analysis of 3-dimensional CT scan data before and after surgery as well as long term follow up (Figure 1). These measurements are used to predict severity of phenotype, success of surgery, and stability of our reconstruction.

Craniofacial microsomia

Craniofacial microsomia (CFM) is one of the most common congenital deformities of the face treated in the Craniofacial Center at Seattle Children's Hospital. This condition affects 1 in 3,000 live births. It presents a wide spectrum of phenotypic severity, as it is known to affect development of the orbits, the jaws, the facial musculature and soft tissue, the ears, and the facial nerve in varying degrees. Because of this wide spectrum, craniofacial microsomia is difficult to treat and requires a team approach to care, similar to that in cleft lip and palate. Surgical treatments for CFM vary and are constantly evolving. Newer techniques, such as distraction osteogenesis and facial reanimation, improve options for these patients and likely improve their outcomes.

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PHENOTYPE

Classification of patients with CFM is challenging due to the wide spectrum of phenotype. This, historically, has made research and treatment planning difficult. A multi-specialty group at Seattle Children's Hospital, including pediatricians, geneticists, psychiatrists, surgeons, speech pathologists, and epidemiologists, are currently working on improving the care provided for these patients. Our research has created a phenotyping tool, the PAT-CFM (Phenotypic Assessment Tool for Craniofacial Microsomia) that utilizes a pictorial to classify patients of varying severity in an effort to streamline research and treatment (Figure 2). This tool is currently undergoing inter- and intra-rater reliability studies and is being used in an NIH-funded multi-center study (FACIAL network). Additionally, we are using this tool to assess patients with CFM using a 2-dimensional photographic protocol as well as 3-dimensional photos using the 3dMD camera system (Figure 3). With this tool, we hope to quantify severity of phenotype, group similar phenotypes to facilitate multi-center research, and analyze outcomes of our surgical interventions.

QUALITY OF LIFE

As surgeons, we assume that improvements we make on patients' medical conditions will improve the quality of their lives. Yet, for conditions such as CFM, multiple surgeries through sensitive developmental periods of life are necessary to approach normal form and function. It is possible that the surgeries themselves, along with the child's fear and pain during recovery, have negative effects on their quality of life which may supersede any improvements our surgeries make. Our research is, therefore, including a component to measure quality of life. We are administering a quality of life tool (the Y-QOL), initially developed at the University of Washington and modified for online use, to assess the changes in life quality seen in our patients before and after surgery.

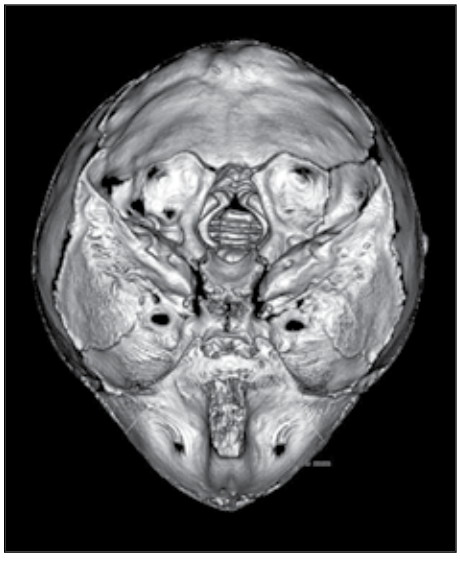

Figure 1. Top down view of the skull base in a patient with metopic craniosynostosis. Analytic software is used to measure angles and distances from the skull base to the frontal bones and orbital bandeau. These measurements are used to document phenotypic severity, measure degree of surgical improvement, and monitor amount of relapse when applied to pre-op, post-op and 2-year follow-up CT scans.

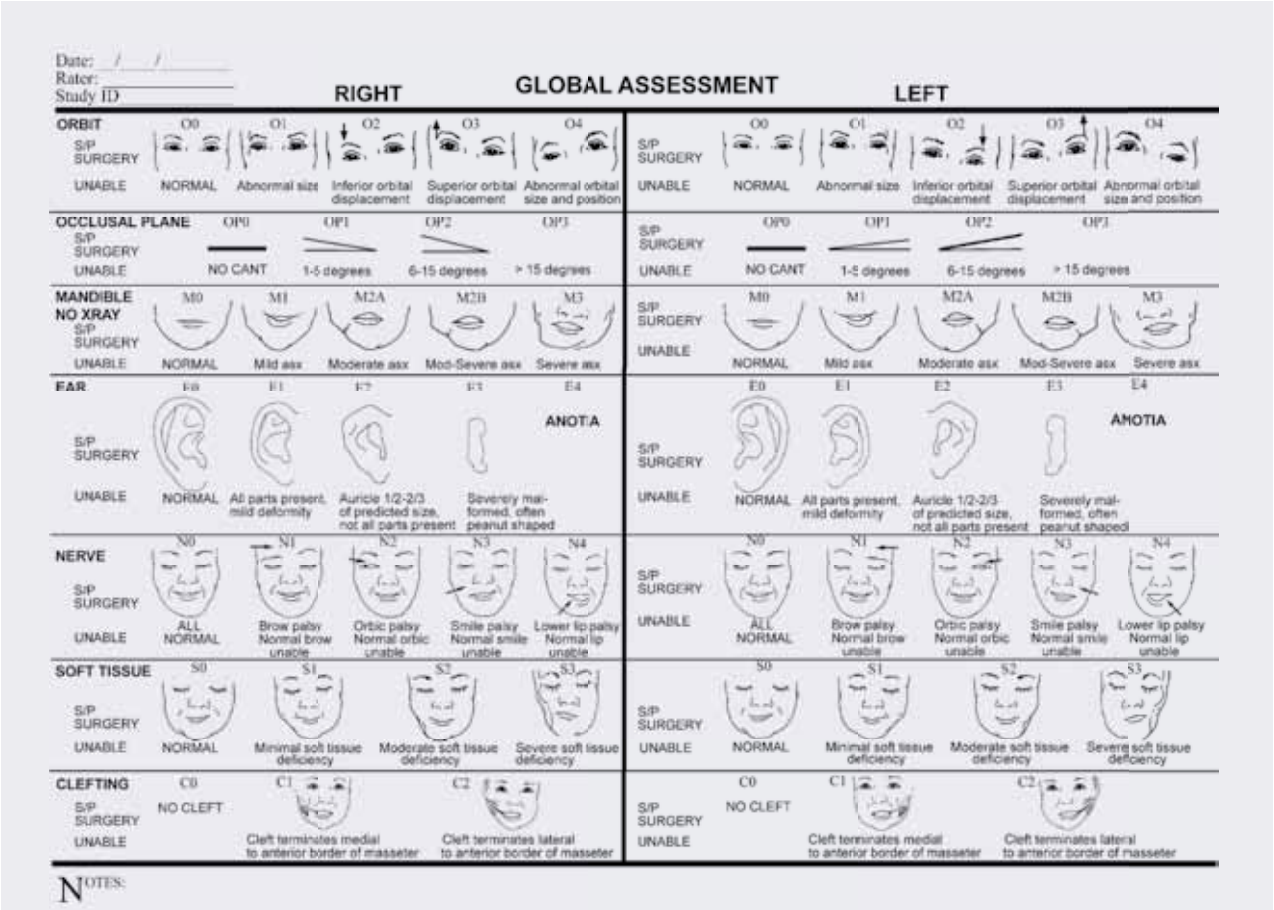

Figure 2. above **Page 1 of the Phenotypic Assessment Tool for Craniofacial Microsomia (PAT-CFM); Global Assessment. This tool was created to document phenotypic severity in patients with craniofacial microsomia and was modified to improve ease of use in the clinic and with both 2D and 3D photographs. Severity of Phenotype can then be compared to post-surgical results to monitor quality of care and can be compared to quality of life scores to help predict outcomes.**

Figure 3. left **Example of a 3D photo taken using the 3dMD camera system. This camera system is being used to document patients with various craniofacial anomalies, especially craniofacial microsomia both to document phenotypic severity and to track and measure surgical outcomes.**

RELATED PUBLICATIONS

1. Scott JR, Isom CN, Gruss JS, Salemy S, Ellenbogen RG, Avellino A, Birgfeld C, Hopper RA. Symptom outcomes following cranial vault expansion for craniosynostosis in children older than 2 years. *Plast Reconstr Surg* 123:289-299, 2009.
2. Birgfeld CB, Luquetti DV, Gougoutas AJ, Bartlett SP, Low DW, Sie KC, Evans KN, Heike CL. A phenotypic assessment tool for craniofacial microsomia. *Plast Reconstr Surg* 127:313-320, 2011.
3. Birgfeld CB, Neligan PC. Treatment of facial nerve disorders. *Skull Base: an Interdisciplinary Approach*. Accepted for publication, October 2010
4. Birgfeld CB, Gruss JS. The importance of early, accurate bony reconstruction in orbital injuries with globe loss. *Craniomaxillofacial Trauma and Reconstruction*. Submitted for publication October 2010.