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The Misshapen Head
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The Misshapen Head

The evaluation and management of infants with potentially misshapen heads presents many challenges. Diagnostic dilemmas for the pediatric primary care provider can be significant. Is a particular infant's head shape normal or abnormal? How does one define geometric and anthropomorphic features? Does the head circumference measurement bear any significance? Should skull radiographs be obtained? What about computed tomography scans? To whom should this infant be referred: a neurosurgeon, plastic surgeon, physical therapist, or geneticist? And, perhaps most difficult, what does one tell the parents?

Pediatric subspecialists caring for these patients must be dedicated to a comprehensive and compassionate multidisciplinary approach. Primary objectives include the early and thorough evaluation of the suspected cranial abnormality and its optimal management, resulting in the greatest patient benefit at the lowest risk. Establishing an effective line of communication among family and health care team members is imperative. A rewarding part of our jobs as care providers is the alleviation of fear and anxiety that haunts parents who believe their infant may be "deformed." Thus, clarification and education become paramount.

After the introduction of the "Back to Sleep" program for the prevention of sudden infant death syndrome, evaluation of the misshapen head has become a very common referral problem for pediatric neurosurgeons. The vast majority of these represent positional plagiocephaly—flattening of the occipital region produced by chronic pressure effects on the calvarium in infants who lie supine. Because the cranial sutures remain open in this entity, nonsurgical management is generally highly effective. This includes positional alteration; physical therapy if any underlying torticollis is present; and the application of a cranial orthosis (remolding helmet) in more severely affected cases. Reassurance can be offered that the cosmetic deformity will correct (albeit incompletely) over the course of time, and not interfere with the normal growth and development of the brain. This realization, in and of itself, is powerful medicine to many families.

Although less common than positional plagiocephaly, the true craniosynostoses certainly are not rare. The most prevalent is sagittal synostosis that occurs with a reported incidence of 1 case per 1000 live births. This entity, which has a narrow, keel-shaped vertex, can be readily diagnosed by clinical impression alone, obviating the need for confirmatory radiographs in the first half of infancy. Unilateral, bilateral, or multisutural craniosynostosis frequently occurs in the context of craniofacial syn-

dromes; again, clinical impression is paramount and radiographs are used primarily to define the extent of calvarial and skull base deformities. True lamdoidal (posterior) and metopic (forehead) synostoses are quite rare; positional plagiocephaly is most often confused with the former. Most cases of true craniosynostosis require operative intervention. Optimal cosmetic results can be achieved by operating on infants affected by sagittal synostosis at ~3 to 4 months of age. Surgery for the other varieties is generally deferred until later in infancy or childhood to provide the best-sustained correction.

A plethora of surgical techniques have been advanced for the correction of the craniosynostoses. The nature of these interventions has mirrored surgical philosophy and technology available at the time of their introduction. For example, simple synostectomy (surgical excision of the affected suture) was described centuries ago by the Greeks and remained the standard until the 1970s when the field of craniofacial surgery emerged. A sophisticated approach to anthropomorphics, calvarial and facial anatomy, as well as the emergence of microsurgical techniques and instrumentation produced an abundance of complex remodeling procedures. Although each progenitor has espoused cosmetic superiority for his or her particular technique, strong scientific evidence has been sorely lacking. Complicating the situation is the absence of a uniform system for describing cranial morphologic alterations and the results of their surgical correction. Almost by definition, cranial surgery is a discipline oriented toward aesthetics and subjective impressions. What is beautiful to one set of parents may be unacceptable to another. Additionally, families who have invested emotionally in the decision-making process and who subject their infant to surgery tend to be highly accepting of the outcome.

In this issue of *Pediatrics*, Jimenez et al¹ present the results of surgical management of the craniosynostoses using an endoscopic-assisted technique followed by cranial orthotic therapy. Data were collected prospectively for 100 patients with sagittal, lamdoidal, coronal, or metopic synostosis undergoing these procedures. The results demonstrated that this technique could be applied safely, with minimal operative time, small blood loss volumes, and infrequent need for perioperative transfusions. Although not well-defined, the authors report good cosmetic outcomes in the vast majority of cases. They advocate their technique over more extensive surgical remodeling procedures that have reported longer operative times, larger blood loss volumes, and increased need for perioperative transfusions.

Despite its strong surgical orientation, this report is important to the pediatric health care provider for several reasons. First, the use of the cranial orthosis for positional plagiocephaly has been popularized and seemingly well-accepted by clinicians and families. The use of helmets for remodeling or maintaining results after surgery, as outlined by Jimenez et al,¹ is novel and appears to be an effective adjunct to surgery. Thus, primary pediatric health care providers will certainly be exposed to an increasing number

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of helmeted infants. It is incumbent on both surgeons and primary care providers to establish a dialogue and comfort with the fitting and use issues of such devices. For example, when should one be concerned about scalp erythema that develops at “pressure points?” When do helmets need to be resized or remanufactured?

The pediatric health care provider must also understand that the early diagnosis and management of the craniosynostoses is imperative to produce optimal cosmetic benefit of therapy. Although it may be disturbing to discuss a seemingly misshapen head with parents of a potentially affected child, failure to do so is an unacceptable omission. Many pediatric neurosurgeons and plastic surgeons have developed a comfort with discussing these issues; therefore, please make use of their consultative skills.

Is also important to understand that “lesser” is not always better for patients with regard to a surgical procedure. The safety and well-being of patients is always paramount, however parameters such as lower length of operating time, operative blood loss, and transfusion rates do not, de facto, connote overall patient benefit. Any amount of blood loss—especially that which may be unrecognized because of its occult location beneath a surgical wound—may place a young infant at risk for rapid and catastrophic cardiovascular collapse. Although all surgeons try to minimize blood loss and avoid transfusions, sensible infusions of blood products on a routine basis have made many varieties of surgery (for example, open-heart procedures) highly safe.

Along similar lines, a minimally invasive procedure is of tremendous benefit if it affords the same short- and long-term outcome as a more invasive one. In the case of craniosynostosis repair, correction of the cranial/craniofacial deformity is the obvious primary outcome. Jimenez et al¹ document an outcome commensurate with other cranial remodeling procedures. In the case of Dr Jimenez’s technique, however, there is an absolute requirement for weeks to months of cranial orthosis use. For the more extensive cranial remodeling procedures, the optimal cosmetic result is realized immediately after postoperative swelling subsides. As alluded to above, helmeting also involves issues of proper fitting and maintenance that can be time-consuming, intensive for the families and the patients, and costly.

As a final note, it must be realized that each medical community may approach the management of craniosynostosis differently. The skill level and comfort for an individual procedure or approach will vary among surgeons. Jimenez et al¹ have presented a novel means for safely and effectively managing infants affected by various forms of craniosynostosis. They are to be congratulated on their work and its presentation to the pediatric community.

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Coding for Mental Health and Behavioral Problems: The Arcane Elevated to the Ranks of the Scientific

ABBREVIATIONS. CPT, *Current Procedural Terminology*; ICD-9-CM, *International Classification of Diseases, Ninth Revision, Clinical Modification*; DSM-IV, *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition*; ADHD, attention-deficit hyperactivity disorder; MBD, minimal brain dysfunction.

In this issue of *Pediatrics electronic pages*, Rushton and colleagues¹ report their experience regarding the coding practices of pediatric generalists, developmental and behavioral pediatricians, and child and adolescent psychiatrists as related to behavioral and mental health issues. In their review, the phrase “alternate coding” is used to characterize the reality that physicians in different specialties will use different coding strategies to report the same patient encounter and diagnostic reality. There is some moralizing about the right way to do things and whether or not alternate coding can be characterized “simply as miscoding, gaming, or fraud.”

*The Merriam Webster Dictionary*² defines alternate as “substitute” or “that which is chosen in place of something else.” There is nothing in this characterization that suggests either appropriateness or accuracy. To determine whether or not “alternate coding” is an acceptable practice, it would be germane to review the existing administrative data and coding systems. Reporting data of a medical office encounter consists of 2 numeric codes—a 5-digit procedural code (*Current Procedural Terminology, Fourth Edition [CPT-IV]*)³ and a 3-, 4-, or 5-digit code that reports a diagnosis (*International Classification of Diseases, Ninth Edition, Clinical Modification [ICD-9-CM]*). CPT reports **what** was done and ICD-9-CM reports **why** it was done. In a simple and perfect world, this would be enough. Ironically, however, the CPT system is not used in its entirety by insurers and codes, modifiers, and entire procedural sections are either not included by health plans or not honored when reported by the medical community. A subcategory of ICD-9-CM is the *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition (DSM-IV)* mental health coding system and its complementary volume, the *Diagnostic and Statistical Manual for Primary*

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