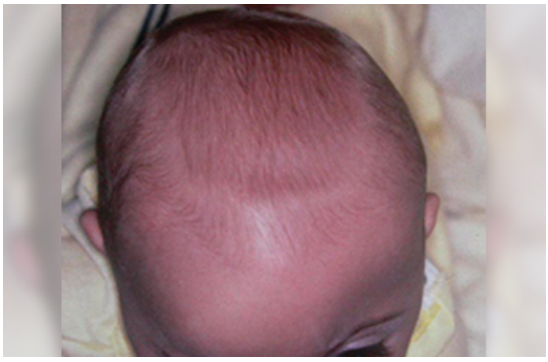
AAP News™

Is this head shape abnormal? Clinical report guides referral decisions

August 31, 2020
Mark S. Dias, M.D., FAANS, FAAP
Article type: AAP Clinical Report
Topics: Neurological Surgery



One of the priorities of the AAP surgical specialty sections is to educate pediatric care providers on identifying and referring children with common surgical conditions to appropriate specialists for evaluation and care. A new clinical report addresses head shape abnormalities that pediatricians are likely to encounter and offers guidance on when to refer patients to a craniofacial surgical team.

The report, *Identifying the Misshapen Head: Craniosynostosis and Related Disorders* from the Section on Neurological Surgery and Section on Plastic and Reconstructive Surgery, is available at https://doi.org/10.1542/peds.2020-015511 and will be published in the September issue of *Pediatrics*.

The clinical report reviews the identification, pathophysiology and clinical consequences of both isolated and syndromic craniosynostosis, occipital deformational plagiocephaly/brachycephaly, as well as microcephaly and fontanelle closure. Photographs and radiographs illustrate the common types of craniosynostotic and deformational head shapes. The report reviews how to differentiate single suture synostosis from other conditions (including deformational conditions), how and when to further evaluate the misshapen head, and when to refer to a surgical specialist for further evaluation and treatment.

Early identification vital

Craniosynostosis and craniofacial disorders are relatively uncommon conditions, and a delayed diagnosis can result in more complicated, extensive and riskier surgical repairs. Treatment delays also can be associated with increased intracranial pressure with central nervous system and/or visual complications. Early recognition is vital so that appropriate referrals can be made to surgical specialists for timely repair — usually within the first several months of life.

Occipital deformational plagiocephaly and brachycephaly, due to supine positioning, are conditions much more commonly encountered by pediatric care providers. Although the overwhelming majority of children with these deformities are readily diagnosed by visual examination alone, a small proportion continue to undergo computed tomographic evaluations (some with 3-D reconstructions) with concomitant radiation exposure. It also is imperative to differentiate children who should not be treated surgically from those with true synostotic deformities who should.

Treatment (positional changes and molding helmets) for deformational conditions continues to be hotly debated among craniofacial specialists. AAP-endorsed guidelines are available from the Congress of Neurological Surgeons at https://bit.ly/3enKW9K.

Finally, misinformation surrounding the role of craniosynostosis in both microcephaly and the timing of fontanelle closure continues to influence the clinical management of these children. Although it is important to identify the rare child for whom craniosynostosis is a potential cause of microcephaly or accelerated fontanelle closure, it also is vital to identify when they are not, so as not to cause undue family stress and unnecessary referrals.

The clinical report provides information that will increase the expertise and comfort of pediatric care providers tasked with caring for children with misshapen heads.

Key points

- Children with craniosynostosis most commonly present with stereotypically shaped heads, each associated with particular sutural fusions:
  - Long (scaphocephaly, sagittal)
  - Short (brachycephaly, bicoronal or bilambdoid)
  - Anteriorly pointed (trigonocephaly, metopic)
  - Asymmetrical (plagiocephaly, unilateral coronal or lambdoid)
- Deformational plagiocephaly/brachycephaly is the most common head shape abnormality, recognized by its parallelogram shape and lack of retroauricular bulge. In 80% of cases, the deformation was not present at birth.
- Syndromic craniosynostosis most commonly manifests with bicoronal synostosis, midface hypoplasia and shallow orbits with exorbitism and strabismus.
- Surgery often is performed within the first 8 to 10 weeks of life for sagittal synostosis repairs, endoscopic procedures and raised intracranial pressure. Orbitofrontal advancements for coronal and metopic synostosis are performed most often between ages 6-10 months. Early referrals to craniofacial teams are encouraged to allow early identification and repair.

Dr. Dias, a lead author of the report, is former chair of the AAP Section on Neurological Surgery Executive Committee.

Resources

- The clinical report offers resources, including photos and radiographs that illustrate common types of craniosynostotic and deformational head shapes.