



Pediatric Clips

The Flat Head Controversy

Positional Plagiocephaly vs Lambdoidal Craniosynostosis

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Pediatric Clips from The Children's Medical Center are quick reviews of common pediatric conditions.

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CASE: A FIVE-MONTH-OLD PRESENTED WITH FLATTENING OF THE BACK OF THE HEAD

A five-month-old, developmentally normal infant presents with flattening of the back of the head. The infant's family felt there was

some flattening a month or two ago, but didn't feel that it was serious. Now it has progressed.

They want to know what is going on and what you are going to do about it.

EVALUATION/RECOMMENDATIONS

Deformational plagiocephaly has been recognized for thousands of years. The diagnosis and treatment of the majority of craniosynostotic deformities involving the saggital, metopic and coronal sutures are straightforward. It is the lambdoidal suture that is problematic, yielding a posterior flattening which has created controversy in recent years. The majority of cases of posterior plagiocephaly do not represent a true synostosis, but rather positional molding.

EPIDEMIOLOGY

Nonsyndromic craniosynostosis affects approximately one in 2,000 live births. The incidence of true lambdoidal synostosis among all craniosynostosis is less than three percent. Interestingly, however, one in 300 births manifest some degree of posterior plagiocephaly. The incidence of positional molding has skyrocketed since the early 1990s when the American Academy of Pediatrics instituted recommendations for supine positioning of infants to prevent sudden infant death syndrome. In some craniofacial centers, referral patterns for plagiocephaly without synostosis has risen six-fold.

DIFFERENTIAL DIAGNOSIS

It is critical to differentiate between true synostosis and positional molding. This can often be done on a clinical basis without radiographic testing. This differential is crucial because true synostosis remains a surgical disease that will worsen over time, while positional molding requires no intervention or subspecialty referral.

From an etiologic point of view, positional molding occurs from constraint in utero or posturing in the postnatal period. Torticollis may be a contributing factor in 10% of cases. It is a result of either sternocleidomastoid imbalance or congenital muscular torticollis which occurs in about three in 1,000 live births and is not related to a traumatic delivery. Self-limiting external hydrocephalus may also be a factor leading to plagiocephaly by contributing to greater compliance and malleability of the skull.

The difference between the two is comparable to the difference in a parallelogram and trapezoid. Characteristically, true synostosis will have palpable ridging or piling up of bone in the region of the abnormal suture, although on skull

x-ray at times, there may be sclerosis on either side of the suture in positional molding. In true synostosis the head appears trapezoidal in configuration and viewed from behind has a skull base tilt. In positional molding, the skull appears normal from behind. Both will have a contralateral posterior bossing. In the true synostosis, the bossing is displaced into the parietal region. There is an ipsilateral occipital-mastoid bulge and the ear is displaced posteriorly and inferiorly. In positional molding, the ear is displaced anteriorly. Due to the effect in the ipsilateral coronal suture, there is contralateral frontal bossing in true synostosis whereas in positional molding there is an ipsilateral frontal bossing. In severe cases of true synostosis, there may be scoliosis of the face and mandibular malalignment, which reflects changes involving not only the suture but also the skull base. When viewed from above and behind, the true synostosis appears like a trapezoid and from above positional molding looks like a parallelogram.

Continued on the reverse side.

Continued from the front.

TREATMENT

The treatment for the true synostosis is surgical. The technique employed will vary according to the patient's age. The treatment for positional molding is as controversial as the diagnosis. Most agree that it is a nonsurgical deformity. With rare exceptions conservative management with positional change will suffice. A significant differential result between active counter-positioning and dynamic orthotic cranioplasty (DOC band) and/or helmets has been found to be wanting. Others believe that quicker and better results occur with an external orthotic device. As the list of externally applied devices begins to expand (assistive device, DOC band, helmets, thermoplastic orthotic device), one must take into account the collateral effects of their use. Costs can range from \$500 to \$2,000. Upgrading adds a substantial cost. Third-party payers frequently refuse to cover these, even with a prescription, and the burden of cost falls to the family. Patient compliance and comfort should also play a role in decision-making. The window of opportunity is somewhat narrow for using externally applied devices as the actively growing brain and, therefore, skull are prerequisites for their employ. By the end of the first year (some feel up to eight months) they would have little utility. The general use of these devices seems to be falling from favor

in the mainstream of pediatric neurosurgery.

Unlike some case of true synostosis, which may be associated with increased intracranial pressure and possible developmental problems, there have been no reported cases of developmental delay or brain injury associated with positional plagiocephaly. Active repositioning of the infant as well as the crib location, modification of feeding patterns, sleep positioning in the crib with head turning, and the prone position for those after three months may be more than adequate intervention for the majority of cases. A short course of physical therapy intervention may also be helpful particularly if there is a question as to neck musculature as a contributing factor.

It is important to counsel the family so they understand that they are not facing a serious neurologic/cosmetic threat. They need to understand that their role is an active one in the amelioration of the problem. Finally, it is important to convey that the majority of children with this problem go on to have normal-appearing heads given time and active intervention on the family's part.

Featured specialist



Laurence Kleiner, MD, is the director of the department of neurosurgery at The Children's Medical

Center. Dr. Kleiner received his medical degree from Temple University. He completed his fellowship in pediatric neurosurgery/CFS physiology at Brown University School of Medicine. His specialty interests include brain tumors, endoscopy with hydrocephalus.

REFERENCES/ RESOURCES

References are available on request by calling The Children's Medical Center, marketing communications department at 937-641-3666.



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