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Posterior plagiocephaly: craniosynostosis or skull molding?

Abstract The surgical indication in posterior plagiocephaly has been and still is a subject of discussion. Unlike other types of craniosynostosis, this particular type of cranial deformity does not show the typical radiological findings that are encountered in cases of prematurely fused cranial sutures. Furthermore, in most cases even the inspective evaluation during the surgical operation fails to demonstrate the actual fusion of the lambdoid suture. The mild clinical manifestations of the condition and the observation that physical maneuvers, such as, for example, wearing constrictive helmets, may suffice to correct the malformation add further uncertainties to surgical correction. In recent years, there has been an apparent increase in the incidence of posterior plagiocephaly following changes in the sleeping habits of infants as a result of the recommendation of the American Task Force on Infant Position and Sudden Infant Death Syndrome, which has favored a supine position. The phenomenon has prompted a series of reports aimed at differentiating posterior plagiocephaly resulting from a true early fusion of the lambdoid suture, which requires early surgical correction, from a deformational modification of the calvarium caused by prolonged and excessive pressure exerted on the posterior aspect of the skull in early infancy, which may not need surgical treatment. Unfortunately, in spite of great interest in the problem, the differential diagnosis between the two conditions remains obscure. In clinical practice, the surgical indication appears still to be based mainly on the evidence of a rapidly worsening cranial deformation rather than on sound objective criteria.

Key words Craniosynostosis · Posterior plagiocephaly · Lambdoid suture · Helmet · Subarachnoid space · Skull morphometric analysis

Introduction

In the early 1990s, formal actions were undertaken in several countries to encourage parents to avoid placing their infants in a prone position following a variety of publications supporting a possible relationship between the prone position and sudden infant death syndrome (SIDS). Such a policy was favored by the report of the AAP (American Academy of Pediatrics) Task Force on Infant Positioning and SIDS, published in 1992, which recommended a nonprone sleeping position for healthy infants, while indicating the
prone position as the position of choice only for premature infants with respiratory distress, gastroesophageal reflex, and some upper-airway anomalies.

Several studies have demonstrated that babies placed in either supine or prone positions will generally remain in that position throughout sleep, at least during the first months of life. A possible role of the supine position in inducing molding of the posterior aspect of the head was suggested by some investigations as early as the end of the 1970s. In recent years, an apparent increase in the relative incidence of posterior plagiocephaly has been recorded in some series of children operated on under the diagnosis of craniosynostosis. On the grounds of these observations, pediatric neurosurgeons have begun to express concern about the possible relationship of this apparent increase with the new policy of placing babies in a supine position for sleep. In some cases, the diagnosis which led to the operation has been questioned by authors who have taken into account the possibility that some of these children with “posterior plagiocephaly” had actually been unnecessarily operated on because of a molded skull, a condition easily treatable by nonsurgical methods. Further support for this hypothesis has been provided by reports demonstrating that the great majority of infants with flattened skull may be simply corrected using physical measures such as changing the sleep position or having them wear corrective helmets for a certain period of time. In February 1997 in Phoenix, a symposium sponsored by the Plastic Surgery Education Foundation, the American Society of Maxillo-facial Surgeons, and the Joint Section on Pediatric Neurological Surgeons discussed the problem with the aim of identifying reliable specific criteria for differentiating true craniosynostosis and simple skull molding.


Information. During the period 1991–1994, 102 infants with posterior plagiocephaly were referred to the craniofacial program at the Children’s Hospital and Medical Center, Seattle (Wash., USA) for the management of abnormal head shape, characterized primarily by abnormal head shape, characterized primarily by posterior plagiocephaly. Patients believed to be affected by positional molding were treated either conservatively or with helmet therapies. However, subjects with progressive positional molding and patients with true lambdoid synostosis were considered surgical candidates. Of the 102 infants, 98 were diagnosed as affected by posterior plagiocephaly secondary to positional molding and only 4 – all with a unilateral malformation – as affected by true lambdoid synostosis. These 4 subjects represented an incidence of 3.1% of a series of 130 children with craniosynostosis surgically treated in the same period of time. Three of the 98 infants with positional molding also underwent surgical treatment owing to more severe and progressive skull deformities. Operative findings demonstrated the patency of the lambdoid suture in the 3 children with positional molding and the unequivocal fusion of the same suture in the cases with true synostosis. In the latter patients, the complete bony fusion of the suture was confirmed by the subsequent histological examination. The clinical features of the patients with positional molding differed from those of children with true unilateral lambdoid synostosis, although both groups were characterized by unilateral occipitoparietal flattening and contralateral posterior bossing. In cases of true synostosis, the fused suture was palpable as a thick ridge; the contralateral posterior bossing mainly involved the parietal region; there was an obvious occipital-mastoid bulge; and the ipsilateral ear when seen from the vertex appeared displaced posteriorly and when seen from a posterior view, inferiorly. A tilt of the skull base and a twist of the cervical spine, visible on the radiograph, were also typical associated anomalies. In deformational cases, the lambdoid suture was not palpable, the contralateral posterior bossing remained confined to the occipital region, the ipsilateral ear was displaced anteriorly, and the skull base was horizontal and symmetrical. The diagnostic imaging also differed in the two groups: the head shape from the posterior view remained normal in the positional molding cases, whereas it assumed a form like a parallelogram in the cases of true unilambdoid synostosis. The pathological lambdoid suture appeared clearly fused in a fashion similar to that of other forms of craniosynostosis in all 4 children with true craniosynostosis.

Analysis. This study was stimulated by an increased number of referrals for the management of posterior plagiocephaly, interpreted as the result of major awareness