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ABNORMALITIES OF HEAD SHAPE IN AN INFANT

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Since the early nineties, an unusual trend among pediatric neurosurgical and craniofacial clinics has been noted: an increasingly large number of infants are referred for occipital flattening.[1] Flattening of the posterior cranium (posterior plagiocephaly) was considered the hallmark of lambdoid synostosis, a condition caused by a premature fusion of the parietal and occipital skull bones.

Recently, it is clear the vast majority of these infants do not have lambdoid synostosis, but rather a condition known as plagiocephaly without synostosis (PWS). This condition can be treated nonsurgically. Since its recognition, PWS has been highlighted in the public media, including national prime time television reports, newspapers and Internet discussion groups.[2] Coincidentally, evidence is mounting in medical literature to support a relationship between the increased incidence of cranial deformation and infant supine sleep position.[1]

Sudden infant death syndrome (SIDS) is the leading cause of early infantile deaths in the United States.[3] Approximately, 6,000 infants die of this syndrome each year, an incidence of 1.2 per 1,000 live births.[4] After a critical review of epidemiological studies, the American Academy of Pediatrics (AAP) issued a report in 1992, recommending healthy infants be placed to sleep on their sides or backs.[5] This recommendation was followed in June 1994 by a national educational campaign, Back to Sleep, supported by a coalition, which included the U.S. Public Health Service, the SIDS Alliance and the Association of SIDS Program Professionals.[3] The literature suggests this, as well as national campaigns in other countries, has resulted in a statistically significant reduction of the incidence of SIDS.[6] Now, most pediatricians recommend healthy infants be placed wholly on their backs.

A causal relationship between the Back to Sleep campaign and PWS remains presumptive and largely based on a temporal relationship. Although, the issue of how the supine sleep position adversely affects the lambdoid suture remains unclear; it is reasonable to assume the mechanism is related to a disturbance of sutural function and skull growth. Craniofacial anomalies, secondary to suture pathology, demonstrate the complex relationship that exists between skull, brain and facial development.

Alterations of the normal growth pattern of one component can produce significant changes to others. In the case of synostosis or slow growth of the lambdoid suture, as it is thought to occur in PWS, skull growth perpendicular to the dysfunctional suture is impeded. The occipital region on the side of the cranial suture becomes flattened. This is not a new observation.

In 1851, Virchow recognized the shape of skull deformations were predictable in cases of craniosynostosis, depending on which suture was prematurely fused.[7] He noted not only that the skull growth was retarded parallel to the abnormal suture, but the skull growth was enhanced in a perpendicular plane. Thus, in cases of lambdoid synostosis on untreated PWS, cranial growth is exaggerated in a frontal direction on the same side (ipsilateral frontal bossing) (Figure 1). These changes are the result of normal brain growth exerting a force on the pliable cranium of the infant.[7]

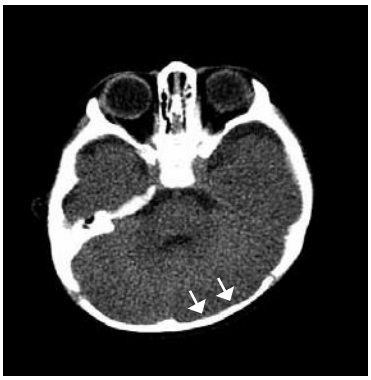

Figure 1. Bone windows of an axial CT scan. Arrows point to flattened left occipital. With occipital plagiocephaly the head assumes a trapezoid shape (in this case with the left side being in front of the right).

The clinical features of an infant with PWS are best appreciated when the child is examined from above, looking down on the vertex of the head. From this view, the skull has a trapezoid or parallelogram shape. Occipital flattening, contralateral occipital bulging and ipsilateral frontal bossing are the most prominent features. This asymmetry may not be readily appreciated when the child is examined face on. The anterior compensatory cranial growth results in the frontal bossing, also results in forward displacement of the petrous bone. This bone houses the ear and the temporomandibular joint. Infants with PWS frequently have forward and inferior displacement of the ear on the same side as the occipital flattening and deviation of the chin to the opposite side. An area of alopecia on the occiput is frequently noted and marks the site of continuous head positioning.

Infants with PWS have normal neurologic examinations, no signs of raised intracranial pressure and developmental milestones appropriate for age.[1, 8] Further, the head circumference is typically within the normal range.

There are two other conditions influencing an infant's head position that should be excluded during the initial evaluations— torticollis and strabismus.[1] Torticollis is characterized by a head tilt and/or restriction of neck movements. If torticollis is untreated and the head position unchanged, the resultant cranial deformity may progress. The majority of infants with restricted neck movements will respond to physiotherapy alone. A minority (< 5 percent) will require release of the sternocleidomastoid muscle. True torticollis must be distinguished from poor head control secondary to muscle weakness, which is treated with physiotherapy and, rarely, bracing. Strabismus, especially when secondary to IVth cranial nerve palsy, may be intermittent and difficult to detect clinically. Parents may report intermittent disturbances of ocular position. If strabismus is detected, then an ophthalmic consultation should be obtained.

We subjectively grade the degree of cranial deformity as mild, moderate or severe. Mild deformity consists of slight occipital flattening. A moderate deformity involves more occipital flattening and some facial and ear asymmetry. Severe cases are characterized primarily by greater frontal bone distortions and asymmetry of the face. Occipital flattening itself is not of cosmetic significance, as it is usually well-masked by early childhood hair coverage and thickening of the scalp. Further cranial growth tends to minimize the asymmetry. However, associated facial asymmetries are of concern, as they may become fixed, cosmetically significant disfigurations for life.

The natural history of PWS is unknown.[8] However, our anecdotal experience, and that from other centers, suggests the vast majority of cases of mild and moderate occipital flattening resolve spontaneously. A minority of cases, particularly those demonstrating progressive deformities, may require surgical intervention. Therefore, we recommend referral to a neurosurgeon or plastic surgeon when:

1) There is severe occipital flattening.
2) The occipital flattening progresses despite infant repositioning.
3) There is associated frontal asymmetry and disfigurement of the face.
4) There is parental anxiety.

The treatment of PWS is dependent on two main factors: the infant's age and the degree of cranial distortion. The younger the infant and the less severe the skull asymmetry, the more likely the child will respond to conservative, nonsurgical treatments. Skull X-rays are not routinely obtained, as they are not sufficiently sensitive or specific to detect sutural abnormalities, i.e. the initial management of PWS is based on clinical, rather than radiographic criteria.

Young infants (<3 months) who have occipital flattening are treated with repositioning only. Parents are instructed about how to keep the child off the flattened region by utilizing various repositioning maneuvers. Parents should be warned—the infant will initially resist these attempts. In the majority of cases, repositioning the infant on the opposite occipital region or a lateral position is successful with some coaxing after several days. This is usually all that is required. The head assumes a more normal symmetric shape with further cranial growth. Infants not responding to repositioning or who have moderate to severe deformity at more than seven months of age should wear the cranial molding helmet. A cranial molding helmet is a custom-fit device, similar to a football helmet, which is designed to apply continuous pressure to the cranium. The device has proved effective, allowing the growing brain to reshape the cranium.[1] It is applied continuously and removed only for bathing. The helmet is usually used for two to three months. Due to the increasing rigidity of the skull after age 1, molding helmets have little value.

Infants with PWS require close follow-up, usually at two month intervals. Response to conservative treatment, either by repositioning or the molding helmet, is usually evident within the first few months. If there is no significant improvement, then diagnostic evaluation to rule out synostosis is undertaken. The diagnostic test choice area is a head computed tomography (CT) scan. This sensitive study can detect subtle, isolated fusion of the sutures. If synostosis is present and the deformity severe, then surgery is considered.

We conclude there is a strong temporal association between the increase in PWS and the introduction of the Back to Sleep campaign. PWS is preventable and the vast majority of cases respond to early intervention with conservative, nonsurgical treatments. Education of the parents regarding the importance of head rotation should result in a reduction in positioning PWS.

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