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THE "EPIDEMIC" OF OCCIPITAL FLATTENING: WHY IT IS HAPPENING AND WHAT TO DO ABOUT IT

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There is a temporal association between the increase in plagiocephaly without synostosis and the "Back to Sleep" campaign. The vast majority of cases of mild and moderate occipital flattening resolve spontaneously. Referral is reserved for those cases in which

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

Since 1992, an unusual trend has been noted in pediatric neurosurgical and craniofacial clinics: an increasingly large number of infants are referred for occipital flattening.[1] Flattening of the posterior cranium (posterior plagiocephaly) is the hallmark of lambdoid synostosis, a condition in which there is premature fusion of the parietal and occipital skull bones. Many infants were thus thought to have lambdoid synostosis and underwent surgical repair. ln retrospect, it is now clear that the vast majority of these infants do not have lambdoid synostosis, but rather a condition that has become known as plagiocephaly without synostosis (PWS). This condition can be treated non-surgically. Since its recognition, PWS has been highlighted in the public media, including national prime time television reports, newspapers and discussion groups on the internet.[2] Coincidentally, evidence was mounting in the medical literature to support a temporal relationship between the increased incidence of cranial deformation and infant supine sleep position.[1]

Sudden Infant Death Syndrome (SlDS) is the leading cause of early infantile deaths in the United States.[3] Approximately 6,000 infants die of this syndrome each year, with an incidence 1.2 per 1000 live births.[4] After a critical review of epidemiological studies, the American Academy of Pediatrics issued a report in 1992 recommending that healthy infants be placed to sleep on their sides or backs.[5] This recommendation was followed in June of 1994 by a national public educational campaign, "Back to Sleep," supported by a coalition which included the US Public Health Service, the SIDS Alliance, and the Association of SJDS Program Professionals.[3] There are reports in the literature that suggest that this, as well as national campaigns in other countries, have resulted in a statistically significant reduction in the incidence of SIDS.[6] Most pediatricians now recommend that healthy infants be placed wholly on their backs.

A causal relationship between the "Back to Sleep" campaign and PWS remains presumptive and is largely based on their temporal relationship. Although the issue of how the supine sleep position adversely affects the lambdoid suture remains unclear, it is reasonable to assume that the mechanism is related to a disturbance of sutural function and skull growth. Craniofacial anomalies secondary to suture pathology demonstrates the complex relation- ships that exists between skull, brain and facial development. Alterations in the normal growth pattern of one component can produce significant changes in others. In the case of synostosis or retardation of growth of the lambdoid suture, as 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 that shape of the skull deformations were predictable in cases of craniosynostosis, depending on which suture was prematurely fused (7). He noted that not only was the skull growth retarded parallel to the abnormal suture, but that skull growth was enhanced in a perpendicular plane. Thus, in cases of lambdoid synostosis or untreated PWS, cranial growth is exaggerated in a frontal direction on the same side (ipsilateral frontal bossing) and in a posterior direction on the opposite side (contralateral occipital bossing) (Figure 1). These changes are the result of normal brain growth that exerts a force on the pliable cranium of the infant.[7]

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 which results in the frontal bossing, also results in forward displacement of the petrous bone. This bone houses the ear and the tempromandibular joint. Infants with PWS frequently have f01ward 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.

lnfants 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 that influence the infants head position and should be excluded during the initial evaluation -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 %) 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, on rare occasion, bracing. Strabismus, especially when secondary to a IVth cranial nerve palsy, may be intermittent and difficult to detect clinically. Parents may report intermittent disturbances of ocular position. If strabismus is detected, 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 in itself is not of cosmetic significance as it is usually well masked by early childhood with hair coverage, thickening of the scalp and further cranial growth that tends to minimizes the asymmetry. However, associated facial asymmeuies 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 suggest that the vast majo1ity of cases of mild and moderate occipital flattening resolve spontaneously A minority of cases, particularly those that demonstrate progressive deformities, may require surgical intervention. We therefore 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, and 4) parental anxiety.

The treatment of PWS is dependent on two main factors; the infants 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, non-surgical treatments. Skull X- rays are not routinely obtained as they are not sufficiently sensitive or specific w 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. The parents are instructed on how to keep the child off the flattened region by utilizing various repositioning maneuvers. The parents should be warned that the infant will initially resist these attempts. ln the majority of cases, repositioning the infant on the opposite occipital region or in a lateral position is successful with some coaxing after several days. This is usually all that is required. The head assumes a more normal sym-metric shape with further cranial growth. Infants that do not respond to repositioning or have moderate to severe deformity at greater than 7 months of age are placed in a cranial molding helmet A cranial molding helmet is a custom fitted device, similar to a football helmet , that is designed to apply continuous pressure to the cranium. This device has proved to be very effective in allowing the growing brain to reshape the cranium (l). It is applied continuously and removed only for bathing. The helmet is usually used for 2-3 months. Because of increasing rigidity of the skull, after one year of age molding helmets have little value.

Infants with PWS require close follow-up, usually at 2- month intervals. Response to conservative treatment, either by repositioning or placement in a 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 of choice is a head CT scan. This very sensitive study can detect subtle, isolated fusion of the sutures. If synostosis is present and the deformity severe, surgery is considered. We conclude that 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, non-surgical treatments. Education of the parents regarding the importance of head rotation should result in a reduction in positional PWS.

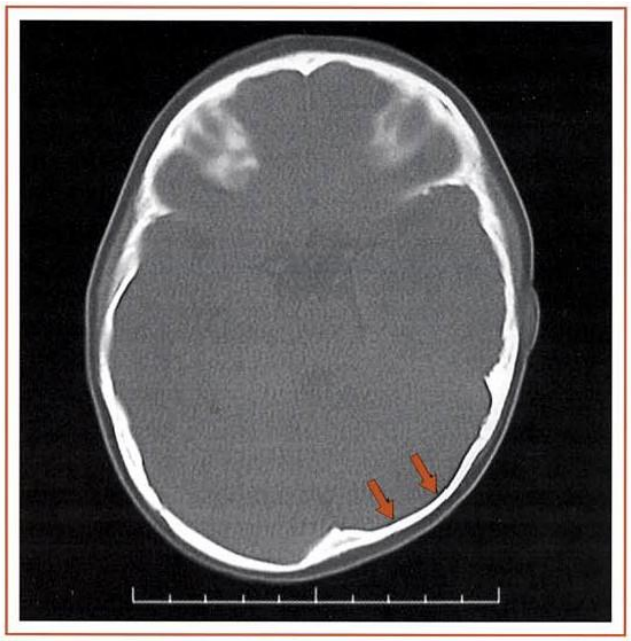

Figure 1. Bone windows of an axial CT scan. Arrows point to flattened left occipital. With occipital plagiocephaly the head assumes a trapezoid shape (in this case with the left side being in front of the right).

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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.

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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:

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