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Management of positional plagiocephaly

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Plagiocephaly is a term derived from the Greek (plagios – “twisted” and kephale – “head”) and describes an asymmetric head shape. The potential causes of cranial asymmetry are multiple and the most important aspect in assessing any child with plagiocephaly is the need to exclude the possibility of craniosynostosis. Craniosynostosis is the premature fusion of one or more skull sutures and often leads to altered head shape; there may also be an associated intracranial hypertension and developmental delay. Premature closure may occur in a single suture or in multiple sutures, as is more commonly seen in syndromic craniosynostotic conditions such as Crouzon or Apert syndromes. Treatment involves assessment, multidisciplinary input from psychologists and speech therapists, and surgery. Positional or deformational plagiocephaly usually presents as occipital flattening present in the perinatal period, either as a unilateral or bilateral deformity and may be associated with changes to the anterior craniofacial skeleton.1 The purpose of this article is to summarise current concepts in the management of positional plagiocephaly and to highlight the present controversy concerning management of the condition with helmet therapy.

AETIOLOGY

Positional plagiocephaly is the most common type of cranial asymmetry, with a prevalence ranging from 5% to 48% in healthy newborns.2 It is distinct from the cranial moulding associated with childbirth, which usually resolves spontaneously in the first weeks of life. Positional plagiocephaly, by contrast, tends to be a postnatal condition which arises due to external forces acting on a flexible cranial skeleton. The worldwide increase in the presentation of positional plagiocephaly has been linked to the various paediatric “Back to Sleep” campaigns, which have recommended that infants be placed supine, to reduce the risk of sudden infant death syndrome (SIDS).3 The incidence of SIDS has been reduced by up to 40%, but at the same time a significant increase in positional plagiocephaly has been noted.4 It is assumed that this increase is due to repetitive sleep positions, with the infant generally resting on one side of the occiput in a comfortable but persistent position, eventually leading to skull moulding.

Positional plagiocephaly is also associated with a number of other risk factors, notably prematurity, motor delay and positional preference. The latter simply refers to a child’s preference in maintaining the head turned to one side and may be associated with torticollis, ocular conditions, scoliosis of infancy, limited abduction of the contralateral hip and foot abnormalities.5 The recognition of congenital muscular torticollis is particularly important, as the unilateral muscular imbalance leads to a persistent aberrant head position and significant plagiocephaly.

DIAGNOSIS

The distinction between synostotic and non-synostotic plagiocephaly (positional plagiocephaly) is made based on a thorough history and examination, with radiographic confirmation as required. As deformational plagiocephaly affecting the anterior cranium is relatively uncommon, anterior plagiocephaly raises the suspicion of coronal suture synostosis. Lambdoid synostosis is much rarer with an incidence of 0.003% to 5%,1 whereas non-synostotic or positional posterior plagiocephaly is becoming increasingly prevalent. The history should include details of the parental medical history and the antenatal and birth histories of the child and any complications in pregnancy. As already stated, positional plagiocephaly tends to develop in the first 3–6 months of life and this information can often be elicited from the history. Abnormal head shape present from birth is much more likely to indicate true craniosynostosis. Examination of patients with positional plagiocephaly reveals specific key characteristics, including occipital parietal flattening, contralateral frontal flattening and forward displacement of the ipsilateral ear. In patients with torticollis, there may be skewing of the neck and, in more severe cases, some minor facial asymmetry. The unilateral flattening is assumed to be related to the repetitive positioning of the skull and as the sutures of the cranial vault are open, structures on the affected side are displaced anteriorly explaining the forward displacement of the ear. When viewed from above, the outline of the cranial vault resembles that of a parallelogram (fig 1). These specific features help differentiate positional plagiocephaly from the more rare lambdoid synostosis, in which the growth restriction of the lambdoid suture causes compensatory overgrowth of the remaining patent sutures. This therefore leads to contralateral parietal bossing and ipsilateral mastoid bulging. There is also a posterior displacement of the ipsilateral ear, and often bony ridging can be felt over the fused suture. When viewed from above, patients with lambdoid synostosis have a trapezoid-shaped skull (fig 2) and from behind they have a “wind-swept” appearance (fig 3).

Clearly, the diagnosis of positional plagiocephaly is thus made primarily clinically, but some authorities6 suggest that radiographs should be taken in all infants with plagiocephaly. We suggest skull x rays if there is no improvement or worsening of head shape; however, the standard series of skull x rays can be difficult to interpret because of the very presence of the plagiocephaly,
and often lead to potentially misleading reports in which partial visualisation or partial absence of sutures is often stated (personal communication, P Anslow). This confusion can raise parental anxiety, and in such cases specialist referral will often be required. In patients with severe positional plagiocephaly, CT evaluation may be required to fully confirm the patency of all the sutures of the cranial vault and skull base.

**MANAGEMENT**

Having confirmed that an infant has non-synostotic positional plagiocephaly, the mainstay of treatment remains early recognition and behaviour modification. Parents should be given instruction about alternating head movements and avoiding repetitive positioning. Simple measures such as a rolled-up towel placed behind the child’s back will help to exert less pressure on the flattest part of the head. Cot adjustments and ensuring that an infant does not remain in the same position in car seats and buggies can often help in limiting positional plagiocephaly. In the United States, the promotion of a new “Back to Sleep and Round Again” message continues to acknowledge the importance of the “Back to Sleep” campaign but also encourages head-repositioning manoeuvres and “tummy time”, where infants are encouraged to spend more time on their abdomen. The earlier these habits are encouraged, the less likely it is that a child will develop a comfortable preferential position and this will ultimately reduce the severity of any positional plagiocephaly. The observation that few children initially placed in the “back to sleep” position present in later life with significant plagiocephaly suggests that these simple measures are all that is required and that positional plagiocephaly can be regarded as a self-correcting condition. This observation was confirmed in an internal review of a small number of patients with this diagnosis but managed conservatively in a British craniofacial unit. It must be emphasised that although the brain and cranium grow rapidly in the first 2 years of life, there is the potential for self-correction for many years beyond this. In patients presenting with congenital torticollis or positional preference and sternocleidomastoid shortening, neck exercises are vital and parents should be encouraged to gently turn the head in both directions on a regular basis. Active physiotherapy will often help correct the sternocleidomastoid imbalance, but patients should be monitored and considered for specialist referral if the torticollis is persistent beyond the age of 1.

**Helmet therapy**

The use of helmets remains extremely controversial. Some authorities suggest they be used in all cases of deformational plagiocephaly still present at 6 months, while others suggest that deformational plagiocephaly is totally self-correcting with time. The cranial orthotic devices are, broadly speaking, divided...
into those that facilitate passive cranial reshaping and those which are said to be dynamic or active devices. The principle of dynamic devices is that they apply pressure on the areas of deformity, limiting further growth, and encourage the relieved adjacent flattened areas to expand. Kelly et al. have suggested that the greatest benefit from these devices is derived when they are used before 1 year of age and ideally when initiated before 6 months of age. They argue that this allows the propulsive rate of brain growth during this period to drive outward expansion and remodelling of the cranium. It is accepted that the value of helmets beyond 1 year of age is less and that by the time a child is 18–24 months of age, the benefit of helmet therapy is very limited. The observation that the vast majority of head shapes due to deforming plagiopcephaly self correct suggests that although helmet therapy may expedite this improvement, such active treatment can be considered unnecessary.

Clarens et al. first reported the use of helmet therapy for plagiocephaly and congenital muscular torticollis. The number of patients in their study, however, was limited to 10, of whom only four actually underwent helmet therapy. Assessment was only for 6–9 months, with very little information on how the plagiocephaly was assessed. In 1996, Turk et al. reported on 52 patients with positional plagiocephaly. In 73%, simple head turning resolved the condition, but the authors recommended helmet therapy in those patients with increasing asymmetry. Four patients underwent surgery for severe deformity, yet the authors failed to provide any details on how the severity was scored and they gave no criteria for those patients who received helmet treatment. Polack et al. reviewed 71 patients, 34 of whom were offered helmet therapy after 2–3 months of positional therapy. The criteria for deciding that this group of patients should undergo helmet therapy are unclear and the method of assessing improvement in 29 of the treated patients is also not stated. Similar improvements with helmet therapy have been reported by Kelly et al., Teichgraeber et al. and Mulliken et al., but all these studies have potential weaknesses relating to inclusion criteria, severity assessment and length of follow-up. To date we were unable to find any controlled, randomised trial confirming the benefits of helmet therapy in deforming plagiocephaly.

In addition to the lack of clear evidence, helmets, to be effective, have to be worn for at least 25 h a day and it is our observation along with reports in the popular press that they may be associated with contact dermatitis, pressure sores and skin irritation. Infants in helmets may suffer social, and potentially psychological, stigma. Helmets are at present not routinely funded on the UK National Health Service, so there are parental cost implications. The findings of Boere-Boonekamp et al. and of Hutchinson et al. also bring into question the value of helmet therapy. Both these studies, with 2-year follow-up, suggest that with time most cases of positional plagiocephaly resolve spontaneously and these findings are consistent with the lack of children presenting in later life with skull deformity. Currently, in the United Kingdom the four designated craniofacial units (Oxford, Birmingham, Great Ormond Street and Liverpool) do not feel there is sufficient evidence to support the use of helmet therapy. The consensus is that the mainstay of management should remain early recognition and repositioning strategies, with physiotherapy for those cases associated with torticollis. Specialist referral should be considered when craniosynostosis is suspected or where parental reassurance is required for severe positional plagiocephaly.

Competing interests: None.

REFERENCES