



Plagiocephaly – more questions than answers

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There has been a striking increase in referrals for plagiocephaly without synostosis (PWS) to neurosurgical and plastic surgical units in New Zealand and overseas.^{1,2} It appears that this increase is related to the increasing acceptance of the SIDS-protective supine sleeping position, although greater awareness and better recognition of the problem also play a part. Prior to 1991, when the National SIDS Prevention Campaign in New Zealand first promoted non-prone sleeping, and at a time when most Western babies were sleeping on their tummies or sides, 'plagiocephaly' meant frontal head flattening; now, there is an almost universal association with occipital flattening. The effects of gravity on a soft, rapidly growing infant cranium lying on a flat surface are such that moulding can occur if the resting position is constantly the same. And, if that is combined with a neck-muscle dysfunction that impairs head rotation, the stage is set for the typical cephalic configurations of a parallelogram-shaped head or brachycephaly or both.

The prevalence of PWS is unclear and has historically been clouded by misclassified lambdoid synostosis.^{1,3,4} A population-based Dutch study found a prevalence of 9.9% in infants under the age of six months; however, the deformity was visually assessed and not quantified.⁵ Head shape varies between the perfectly symmetrical to the severely abnormal, and thus prevalence will vary depending upon the criteria used to define normal and abnormal. However, there are difficulties in quantifying PWS. Although a subjective visual assessment is important, a quick, reliable, objective and noninvasive measure of severity would be useful in order to quantify the abnormality and to follow the deformity over time.³ Methods described in the literature include callipers,^{6,7} 3-D CT scans,^{8,9} photographs,¹⁰ articulated rulers,¹¹ and manual tracings made from flexible strips pressed around the circumference of the infant's head.¹² The fact that there are many different methods suggests that none is ideal.

The natural history of PWS is unknown; few adults have such a deformity, suggesting that it is self-correcting or masked by hair growth. Probably most cases routinely improve with time, but there are no good long-term studies that address the issue of how much time. The effects of true non-treatment are unknown and difficult to evaluate because most parents will try to correct the condition using counter-positioning. There seems to be a group of infants with persistent and severe PWS, but as yet we do not know how to identify these children early in order to institute early treatment strategies.

Primary prevention advice includes gently varying the head position at each sleep until the infant can do it alone, changing the cot environment to encourage looking around, and giving supervised tummy time for play from an early age to encourage upper-body strength.¹³ Although we have conducted a case-control study that gives some support to these recommendations, a larger prospective study is needed to confirm the integrity of this advice.¹⁴

The two fundamental principles underlying treatment of PWS are early recognition and keeping the infant from lying on the flat spot. Positioning advice for the treatment of PWS has ranged from simply staying off the flattened occiput whilst asleep,² to 'active counter-positioning',¹² which not only advises keeping off the flat spot but actively applies pressure to prominent areas by positioning the infant on the bossed side of the occiput. Upright time and supervised tummy time also help to avoid pressure on the occiput. Some authors¹⁵ argue that repositioning is effective only before four months of age, because after that infants tend to reposition themselves, and because longer trials of repositioning only serve to delay treatment, thus making positive outcomes more difficult. Treatment options for severe cases include helmets, which constrain the bossed areas and provide room for growth over the flat areas. More invasive treatment options are cranial surgery and botulinum toxin treatment for severe torticollis contributing to head deformity.

The plagiocephaly literature contains a few references to devices designed to keep the infant positioned off the flat part of the occiput. One author has advocated tying a large knot in a stocking cap, to be positioned over the flat area,¹⁶ others have suggested 'positioning rolls',² sandbags¹⁷ or foam wedges;⁶ none appears to have been assessed for safety. Soft sleeping helmets with a block or cone attached to the flattened side have also been used to keep the plagiocephalic infant from turning to a favoured postural position. To our knowledge, there are three commercially available positioning devices for sale in New Zealand at present. They are a foam-wedge system called the Sleep-EzTM; a sloping foam block with a saucer-shaped indentation for the head to rest in called the Occ-Block; and a fabric sleep wrap known as the Safe-T-Sleep[®]. The lower portion of the Safe-T-Sleep[®] device is fastened around the mattress while the upper portion wraps around the infant's chest and abdomen to maintain sleep position.

If a positioning device is to be used it needs to not only maintain the position but be absolutely safe. De Chalain has attempted to address the difficulty of recommending a safe and effective system of keeping infants off a flat occiput by testing the Safe-T-Sleep[®] device in the monitored environment of a hospital setting. His findings are published in this issue of the NZMJ.¹⁸ The infants tested were sick children. In this study, the head position was maintained in the desired position for 85% of the observed hours. Would the results be any different in healthy and possibly more active infants at home in the care of busy parents? It is not clear how often the nursing staff elected to use the supine position and the semi-supine position, and whether this position was changed during the night; one would imagine that maintenance of head position would be easier in the semi-supine posture. In addition, it would be interesting to know whether the clothing was always pinned to the Safe-T-Sleep[®]. We are of the opinion that fastening the Safe-T-Sleep[®] to the infant's clothing would be the only way to prevent an active, strong-minded baby from rolling to prone were they determined to do so.

It is uncertain from this report as to the seriousness of the adverse events encountered. Safety would be difficult to assess in such a study, as rare events would not be detected. The two adverse events in this trial involving 31 babies are summarised as 'Device too loose – unwanted movement'. This suggests a very real possibility that untrained caregivers could have the same problem were the device not secured correctly *every time*. However, the manufacturer reports that no deaths or serious

adverse effects have been reported with over 70 000 units sold (personal communication, M Rutherford, 2003).

Early awareness and recognition are vital for the management of plagiocephaly. The authors are now recommending the Safe-T-Sleep® for the treatment of plagiocephaly, but have not subjected this to any formal trial. Although they have found that counter-positioning is as effective as treatment with a helmet, this might indicate that neither changes the natural history of the disorder. There is a need for a randomised controlled trial of sleep-positioning devices versus an education programme for the treatment of mild to moderate PWS.

Unfortunately, there are more questions than answers. There is no quick fix for a misshapen infant head, and the old maxim 'prevention is better than cure' is very relevant. Because of parental and health professional concern about plagiocephaly, recommendations need to be made, even though the evidence for these recommendations is limited. Prevention programmes should raise awareness that not just sleeping, but holding and playing positions and the use of car seats may be important. Watching for a preferential head orientation and encouraging turning of the head both ways are prudent. Perhaps we could do away with the expense of and reliance upon positioning aids except in the more severe cases.

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