Deformational Plagiocephaly, Brachycephaly, and Scaphocephaly. Part II: Prevention and Treatment

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Abstract: Cranial deformation is the most common cause of abnormal head shape. Intentional and unintentional alterations of cranial form are associated with the application of external pressure to the growing infant head, and such changes have been recorded throughout man's history. Recent changes in Western sleeping practices, instituted to reduce the incidence of sudden infant death syndrome, have led to a dramatic rise in cranial deformation and renewed interest in this subject. This 2-part review presents a pragmatic clinical approach to this topic including a critical review of the literature as it applies to each aspect of this common diagnosis: historical perspective, terminology, differential diagnosis, etiopathogenesis and predisposing factors, and prevention and treatment.

Key Words: Deformational plagiocephaly, deformational brachycephaly, deformational scaphocephaly, treatment, prevention, cranial orthotic, torticollis, head rotation preference

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PREVENTION AND TREATMENT

Infants at Risk: Early Identification and Intervention

Early identification of at-risk infants offers the best opportunity to prevent deformational flattening. The most important fact to know is whether the infant has a head positional preference—the earliest manifestation of congenital muscular torticollis (CMT). This simple finding is the key to identifying most vulnerable infants. I recommend that pediatricians inquire about this tendency during the first well-baby visit. Most parents are cognizant of this trait but do not understand the significance. The simple question, "Does your infant have a favorite or preferred head position when lying?" on the new baby intake form would help to identify most at-risk infants. Cervical range of motion can be easily evaluated with the neonate lying supine. A significant difference in rotation to either side is indicative of cervical imbalance or torticollis (Figs. 1A, B). Any infant who has a notable difference in head rotation to one side, has a positional preference, is premature, or has a medical condition that might delay neuromuscular development should be managed proactively. Traditional recommendations for infants who appear at risk for deformational plagiocephaly (DP) or deformational brachycephaly (DB), or have just started to develop a flat spot, include physical therapy to address the cervical muscular contracture and repositioning. Data to support these recommendations are sparse.[1] Nevertheless, a recent study showed that early physical therapy protocol for infants with a positional preference lowered the incidence of "severe" DP relative to those who had no intervention.[2] The reported reduction was 46% at 6 months of age and 57% at 12 months of age. Because the authors used specific cutoff measurement (oblique diameter difference index, >104%), and not average change in each group, the degree of improvement after this regimen is unknown. Repositioning is possible only in very young and immobile infants (<4 months of age) and, if done consistently, may serve to prevent or limit further deformation. This method is extremely difficult to use in older, mobile infants[3,4] and is generally ineffective at correcting established flattening.[5,6] In addition, the Food and Drug Administration (FDA) has raised concerns over the safety and effectiveness of positioning devices.

Another method to prevent or treat early plagiocephaly is to alter the shape of the infant's sleeping surface so that the occiput rests against a concave and not a flat surface. These devices act to redistribute and diffuse the contact pressure between the infant's head and the resting surface. Consequently, there is little redistribution of volume as the head grows. Alternative sleeping surfaces do not require altering the infant's head or body positioning, and the infant remains supine at all times. In this sense, alternative resting surfaces are distinctly different than repositioning devices such as wedges. In a case-control study, our group compared treatment of early plagiocephaly using a custom-fabricated foam cup to repositioning and physical therapy for the torticollis.[6] At the end of the 2-month treatment period, improvement in the average transcranial difference was significantly different between the groups ($P = 0.000$): 11.2 to 3.5 mm in the cup group and 9 to 8 mm in the group who received physical therapy and repositioning. The design has been upgraded to include removable liners of increasing size so that parents can adjust the fit as the infant grows (Figs. 2A, B). We currently recommend this orthotic for all infants younger than 3 months who have evidence of torticollis (ie, head preference), are premature, or have early established DP. Because the cup design cannot control lateral (biparietal) cranial growth, it is generally ineffective for established DB.

There are many commercial repositioning devices and alternative sleep surfaces on the market that claim to prevent or treat early DP. With the exception of the orthotic described above, none of these have been shown to be effective in a controlled clinical trial. Some manufacturers have sought FDA approval as a means to legitimize their product, but this is an attestation to safety and not effectiveness. These 510(k) type approvals require the manufacturer to demonstrate that their device is "substantially equivalent" to others that have already been approved. Clinical trials are not necessary for a 510(k) application, and manufacturers are not permitted to make any claims about the effectiveness of their product. Unfortunately, such restrictions are rarely followed.

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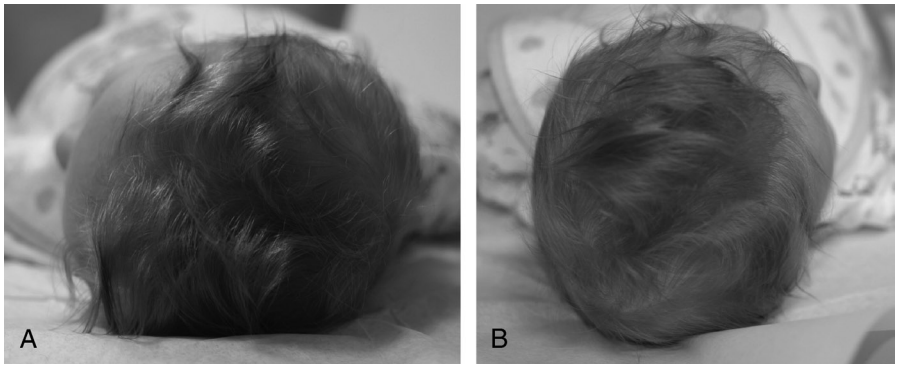

FIGURE 1. Head rotational difference demonstrated in a neonate with congenital muscular torticollis (CMT) and poor head control. The supine infant is stimulated to look each way by a light or sound. A, Head rotation to the left, full. B, Head rotation on the right, limited. The angular difference in rotation to each side reflects the degree of cervical imbalance.

Older Infants

By age 3 months, most infants are beginning to develop head control.[7] They become mobile enough to avoid further flattening. Thus, it is rare to see progression of flattening after 4 months of age (in a term child). Most infants with torticollis will have improved to a point where their head tilt may be subtle, intermittent, or completely absent. Even so, a modest but measurable difference in head rotation to either side is typical (Figs. 3A, B). Minor rotational differences or intermittent head tilting often reflects a persistent minor weakness in the SCM muscle opposite the tilt, and physical therapy is not indicated. The usefulness of off-the-shelf orthotic devices or repositioning to address cranial flattening after this age is very limited. Most infants can easily move off of wedges and other such devices and rarely stay in the position in which they were placed. The difficult issue is deciding if treatment is warranted. Unfortunately, the literature offers little guidance in this area because of a lack of consensus about how to define and measure DP or DB.[1,8] Some authors have proposed strict definitions[9,10] and standardized measurement techniques,[10–12] but these recommendations have largely gone unheralded. Consequently, the indications for treatment are quite variable, and most clinicians make this decision based on subjective impression,[13–20] a clinical grading system,[21–23] or some method of anthropometric measurement.[2,5,9,18,24–28]

Methods of Assessing Severity

Subjective impression is inherently inaccurate and strongly influenced by observer bias. When deciding whether to treat an affected infant, it is important to remember that the parents, and not the clinician, will live with the outcome. Not infrequently, I am asked to consult with distraught parents of older children with unresolved DP or DB. They lament that they had sought early care for their child only to be told by a clinician that the baby's head "looked fine" and "does not require treatment." Assured by the "professional" opinion, they continue to observe the deformity until the child is too old for treatment. Consequently, in my opinion, the only subjective impression worth considering in the treatment decision is that of the parents.

Direct or indirect anthropometry offers some objective standard on which to predicate treatment. Direct measurements can be easily obtained using an anthropometric caliper (Fig. 4). Nevertheless, methods of obtaining data and the interpretation of such measurements are quite variable.[1,8] Deformational brachycephaly and deformational scaphocephaly (DS) are almost always quantified using cephalic index (CI), which is the maximum width of the head divided by the anteroposterior length (Fig. 5). This measurement is standard, and normative data are available. Nevertheless, the indications for treatment are still somewhat arbitrary because normal CI varies significantly among different cultures[29] and has increased in North America after the "Back to Sleep Campaign." Deformational plagiocephaly is quantified by the degree of asymmetry using either absolute measurements (eg, transcranial difference; the difference between 2 oblique head measurements) or cranial ratios (eg, oblique cranial length ratio; ratio of the longer cross-diagonal to the shorter one). We prefer absolute measurements obtained with an anthropometric caliper for assessing DP. The validity and reliability of this technique have been established.[30,31] Different landmarks have been proposed, and this makes it difficult to compare studies. Regardless of the technique used, consistency among measurements is critical. We use fixed landmarks that have been described previously[32] (Fig. 6). No method of two-dimensional anthropometry can accurately quantify the three-dimensional volume

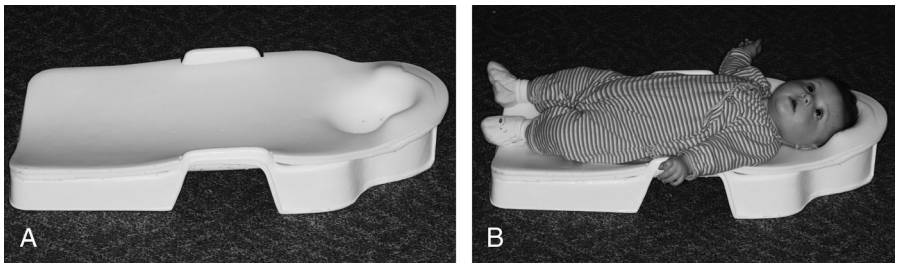

FIGURE 2. The PlagioCradle (PlagioPrevention, LLC, Boston, MA). A, Progressively enlarging foam liners allow the cranial recess to enlarge as the infant's head. This ensures normal cranial shape throughout early growth. This can be used in infants from birth (prevention) to 4 months of age. B, Infant comfortably lying in the PlagioCradle.

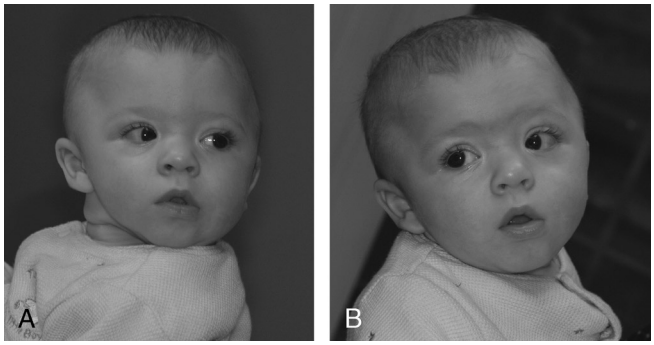

FIGURE 3. Head rotational difference demonstrated in an older child with resolving CMT. A, Overrotation of the head to the left reflects a weak sternocleidomastoid muscle on that side. This is the last feature of CMT to resolve and often manifests in older infants as an intermittent head tilt. B, Minor underrotation of the head to the right from residual sternocleidomastoid tightness.

loss that occurs in DP. Furthermore, standardized anthropometry using fixed anatomic landmarks is not accurate for all variations of plagiocephaly. For example, patients with DS typically have flattening of the parietal region but no significant occipital asymmetry. A standard transcranial measurement may fail to detect a difference, even though there is a volume deficit on one side of the cranium. Despite these limitations, anthropometry is an easy and helpful assessment tool. In the absence of an anthropometric caliper, one can easily estimate the degree of flattening. With the infant's head centered, fingertips are placed on opposite sides of the occiput in parallel lines posterior to the anterior globe. Typical finger width is 8 to 11 mm, and this can be used to estimate the offset between the sides of the posterior cranium (Fig. 7). While this is imprecise, it is a quick and easy method to screen infants.

Some authors prefer to discuss cranial vault asymmetry in terms of cranial ratios. The primary problem with these measurements is that any fixed asymmetry will seem to improve merely because the head becomes larger. Based on these calculated measurements, some researchers have mistakenly concluded that DP naturally gets better,[18,24] even though the absolute asymmetry in DP does not decrease significantly. This type of "relative" improvement is relevant only in infants and young children who are typically viewed from the top of their heads. In these patients, increased head size will make a fixed asymmetry look less impressive relative to the total size of the head. A useful analogy is comparing the appearance

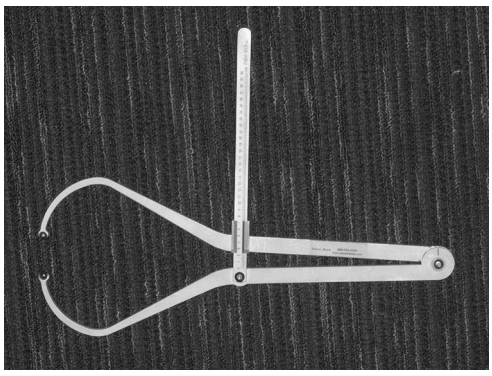

FIGURE 4. Anthropometric caliper.

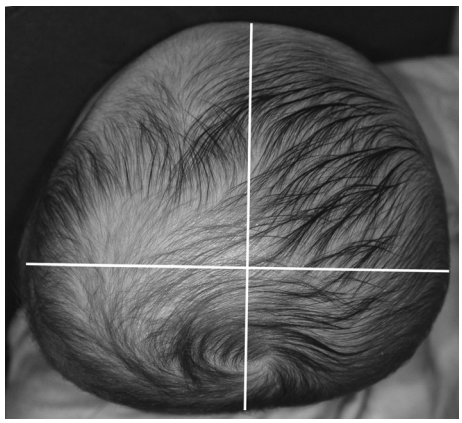

FIGURE 5. Cephalic index is maximal biparietal width divided by anteroposterior length. This is the best measurement for DB and DS.

of a 1-cm indentation on a baseball with the same-size depression on a soccer ball. Of course, the smaller object looks more severely impacted. Nevertheless, cranial ratios are meaningless in adolescents and adults, because the top of their head is rarely visualized by other persons. The cranium of older, taller persons is seen only from a posterior or side view. Thus, the difference in projection between the 2 sides of the occiput will ultimately determine how the asymmetry appears. This difference is best captured by absolute measures such as transcranial difference.

Helmet Therapy

Helmet orthosis is a useful way to improve moderate and severe deformational flattening (Figs. 8A, B). I rarely "recommend" helmets to parents, as the decision to treat flattening should be left to the parents once they are properly educated on what to expect from each alternative. In my practice, I discuss the option of a helmet when the cranial shape meets certain minimum measurement criteria (transcranial difference: ≥10 mm in DP, ≥0.90 in DB), is clearly

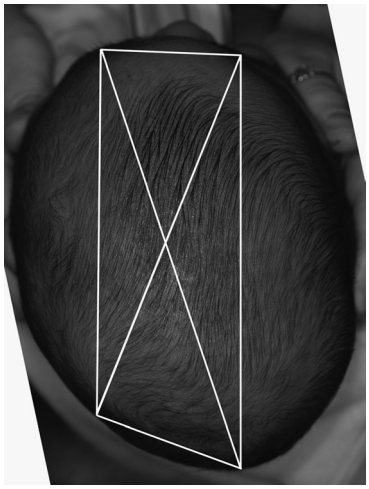

FIGURE 6. Asymmetry can be measured by subtracting the shorter transcranial oblique from the longer oblique (crossed lines). The difference is called transcranial difference (TCD). The anterior and posterior landmarks should be consistent and reproducible.

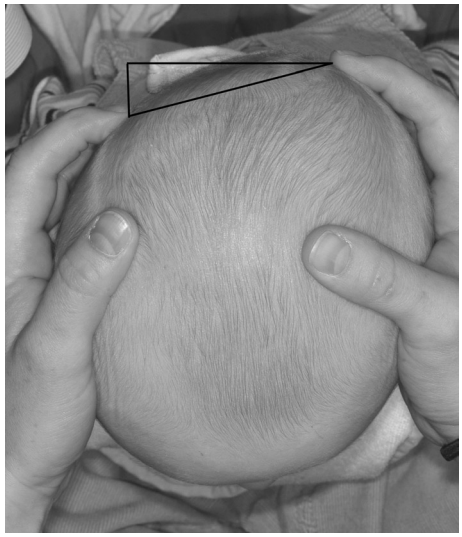

FIGURE 7. Bimanual estimate of cranial asymmetry.

visible from the posterior and side view, and is of concern to the parents. These devices have an excellent track record of safety and effectiveness.[4,5,13,18,33–36] Some skeptics are quick to note the lack of type I evidence (randomized controlled studies) to support the effectiveness of this method,[3] although lower levels of evidence do exist.[1] Others have reported some minor relapse with longer follow-up.[36] The first helmets were made and tested in 1979 by Sterling Clarren.[37,38] His design concept was simple: "If the pressure of a rapidly growing brain against a flat surface would flatten the skull, then pressure [relief] over the concave surface should round it back again." A common misconception is that the helmet is actively "molding" or squeezing the cranium and will cause discomfort. Helmets do not function in this manner and act more like a brace to redirect remaining cranial growth toward the flattened areas. When parents ask how much pressure is being applied to the head with these devices, I remind them how little pressure was required to develop the flattening in the first place—the weight of their newborn's head against an unyielding surface was enough to redirect cranial growth. Therefore, the amount of contact pressure required to correct the shape is negligible and similar to the pressure applied to the head by a well-fitted hat. If compressive force was applied to the head, as some skeptics suggest, the skin would begin to break down within a matter of days. It is analogous to wearing a pair of tight shoes—the skin will become red and irritated in a few hours, begin to blister after a day, and eventually break down if the pressure persists. The same outcome can be expected with a tight or constricting helmet orthotic. This is one reason why the manufacture of these devices is regulated by the FDA, and a skilled orthotist is necessary to monitor the fit. Certain proprietary helmets are touted as "active orthotics" or bands and are said to apply pressure over bony prominences to provide a better or faster correction of shape. As mentioned above, the scalp (like any soft tissue of the body) does not tolerate sustained pressure well and any device that applies sufficient external pressure to move or "push in" a bony prominence would soon cause skin breakdown. Thus, the "advantages" suggested by these manufacturers are more marketing than fact.

Proprietary claims aside, all helmets generally work in the same fashion. To allow the desired changes, the helmet has a foam liner that is selectively cut away in the area where increased growth is desired. The remaining foam limits growth in areas where there is already excessive expansion (Fig. 9). The direction of growth is altered, but the overall cranial growth continues normally and there is no restriction of brain expansion.[39] Over time, excellent correction of flattening can be obtained (Fig. 10). Helmets are custom made and are tolerated very well by most infants. Sleep patterns are rarely affected. Helmets are effective as long as there is cranial growth remaining, and the rate of correction is proportionate to the rate of growth. Consequently, younger patients will correct much faster than older ones. Children as old as 18 months of age can still have some correction of flattening, but the process may take as long as 6 to 8 months. In contrast, a 4-month-old with a moderate-to-severe asymmetry can often be corrected in 6 to 8 weeks. The effect of age at treatment has been well described.[4] Orthotic correction of DP is generally faster and more effective than that of DB, and this outcome difference has been attributed to slow growth of the cranial base.[40] However, a more plausible explanation is that correction of DB requires growth of the entire occipital region, whereas DP requires expansion of only one half. Hence, it will take considerably more time to correct DB than DP.

Some reports suggest that helmet therapy may improve facial asymmetry and ear alignment,[4,35] but this has been refuted by other studies.[41] Fortunately, the face grows steadily well into adolescence. As a result, small difference in facial projection that appears quite noticeable on a small infant will disappear with growth. These early

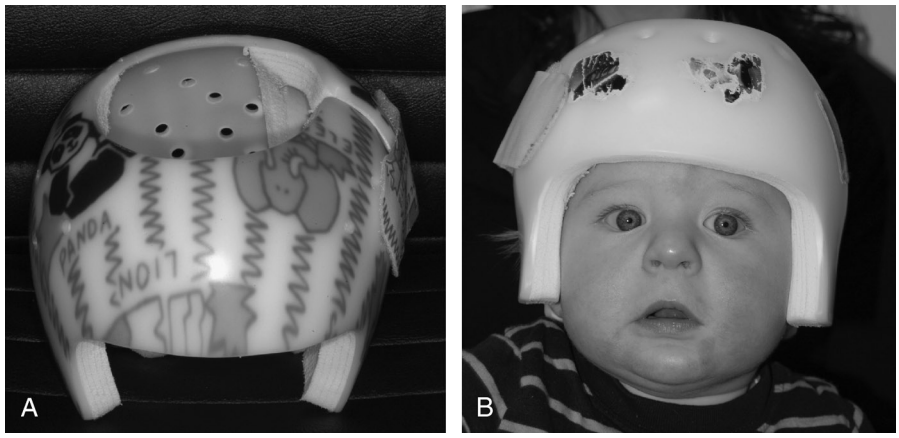

FIGURE 8. Cranial helmet orthosis. A, A light thin plastic shell with foam liner. The posterior foam is cut away to allow cranial expansion in area of flattening. B, Helmet is comfortable, and infants adapt readily.

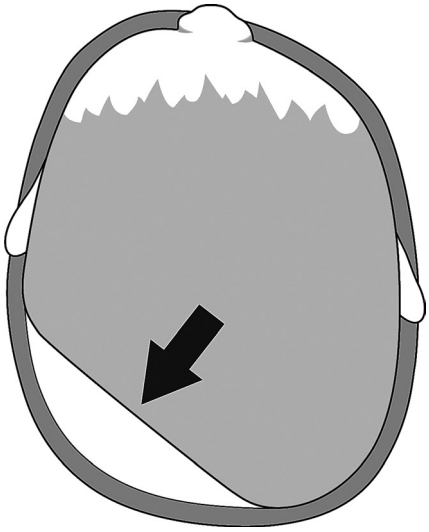

FIGURE 9. Helmet (darker edge) acts as a brace to redirect growth (arrow) to region of flattening. There is no compression, merely contact.

changes are rarely evident in adult, even if there is still significant cranial asymmetry.

OBSERVATION OF ESTABLISHED FLATTENING

By age 4 months, most developmentally normal infants have achieved head control, and they can sit with assistance. Consequently, many infants with torticollis typically have marked improvement in head rotation and will be unlikely to have further flattening. It has been suggested that once the infant begins to sit up or roll over, the flat spot will spontaneously "pop out." This rather absurd notion implies that the cranium can grow differentially, that is, speeding up on the flat side while slowing down of the round side. There is no physiological mechanism by which this can occur. Once the infant can reposition the head, the sustained obstruction to growth is removed, and each side of the occiput will grow at a similar rate. Thus, the degree of flattening remains unchanged. Nevertheless, many observers have stated that the flattening looks less noticeable as the child's head grows. These seemingly conflicting observations are based on 2 factors. First, the head continues to expand even after there is no further flattening. Thus, relative to overall head size, the flattening becomes less pronounced. Compare an indentation on an apple with the same size indentation on a cantaloupe. The flat spot appears less obvious on the larger, wider fruit. It is no coincidence that studies that used either subjective visual assessment[13,16,17,19,20] or cranial ratios[18,24] typically reported improvement of DP over time. In contrast, other studies that used absolute differences (transcranial difference) show only minor improvement.[5,6] Numerous other studies have also documented "incomplete" correction with growth[7,14,34,37,38,42–44] Cranial growth is finite and nearly complete in early childhood. Therefore, in severe cases, there may not be enough remaining growth left to normalize appearance (Figs. 11A, B). One follow-up study found that 58% of parents still noticed residual asymmetry in their older children (mean age, 7.1 years), although only a small percentage reported being teased by peers.[31] The brain is fully grown by this age, and unfortunately, further improvement through intracranial expansion is impossible.

The second important factor that makes cranial shape look better with growth is the change in an observer's perspective. Cranial asymmetry is more obvious when viewed from the top of the head than from the back or sides. This is because size difference between 2 objects (including the 2 sides of the occiput) is easier to quantify when they are viewed in a plane perpendicular to the axis in which the discrepancy occurs. For example, if one is looking directly down from an airplane on the top of the Sears Tower, it would be difficult if not impossible to determine the difference in height between it and the adjacent shorter buildings. Nevertheless, the contrast in building height is easy to visualize from the ground. Similarly, cranial asymmetry (which occurs in the sagittal plane of the head) is much more obvious when viewed from the top of the head (axial plane). As a child grows taller, this perspective is almost irrelevant. With the exception of perhaps shoe salesmen, the vertex of an adult's head is rarely seen by casual observers. In most instances, the cranium will be viewed from the back or the sides, both of which are much more forgiving for those with established asymmetry (Figs. 12A, B). In my experience, occipital asymmetry of 10 to 12 mm and greater is obvious in older children and adults with very short hair. As a general rule, if the contrast between the sides of the occiput is easily seen from a posterior or side view in infancy, it is likely that it will be even after growth.

Hair growth can also provide some camouflage for persistent cranial deformation. It is commonly suggested by parents and pediatricians alike as the remedy for residual flattening.[24] This forces one to adopt a longer hairstyle throughout life to compensate for the asymmetry or flattening. Even with longer hair, the shape of the head can be visualized if the hair is long and straight, is worn on top of the head, or if it is wet. Additionally, hair does little to cover the increased head width observed in children with DB. These children are often referred to by their parents as having a "big head," when in fact their overall head size may be normal. In general, hair growth should not be factored into the decision to treat cranial flattening or asymmetry.

IMPLICATIONS OF UNTREATED CRANIAL DEFORMATION

Although most authorities believe that the only potential long-term effect of deformational flattening is altered cranial shape, some reports have suggested that DP can have medical consequences, such as intellectual impairment or developmental delays[27,45–48] visual disturbances,[49] otitis media,[50] decreased motor tone,[51] and occlusal problems.[36] These studies suffer from methodological flaws and have been justifiably criticized.[1,52] Although infants with developmental delay may be more prone to develop DP (presumably due to poor early mobility), few investigators believe that the process of flattening actually causes these delays.[1,52,53] In our experience, the

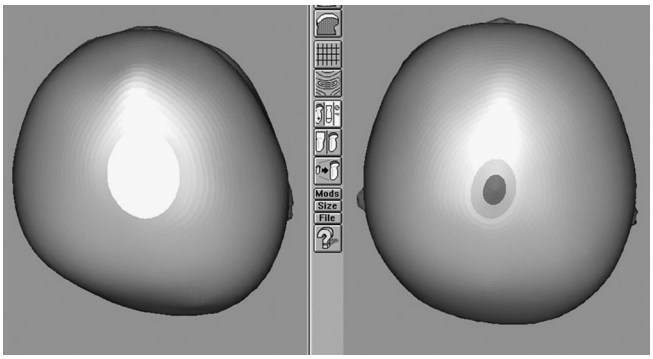

FIGURE 10. Three-dimensional surface laser image of infant with right DP. Left, Before helmet treatment, age 5 months. Right, Correction after 3 months of helmet therapy.

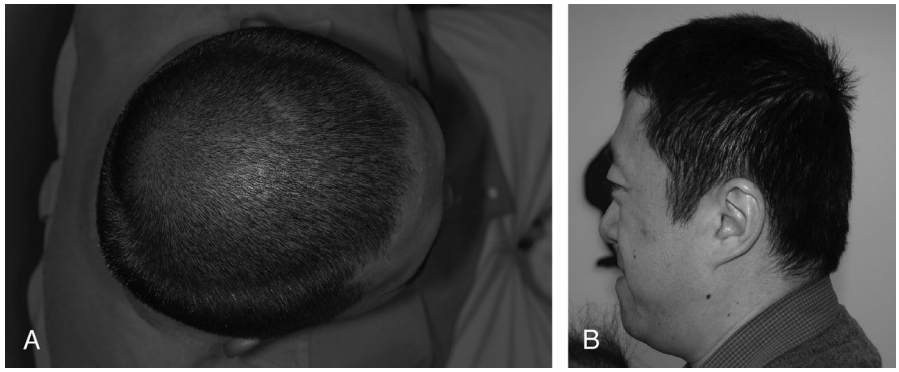

FIGURE 11. Uncorrected deformational flattening. A, A 30-year-old man with moderate uncorrected left DP (TCD = 14 mm). B, A 38-year-old man with severe DB (CI = 1.0).

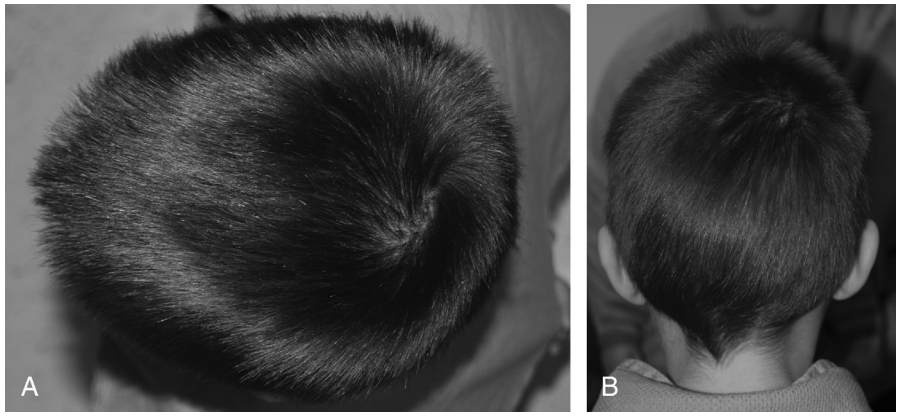

FIGURE 12. A 9-year-old with uncorrected mild (TCD = 11 mm) left DP. A, Asymmetry is easily noticed from vertex view. B, The asymmetry is almost imperceptible from the back. This illustrates how cranial asymmetry can appear ''resolved'' merely by changing the viewer's perspective.

overwhelming majority of infants who develop DP or DB have no identifiable developmental delay, and in those who do, there is a clear comorbid condition that accounts for such a finding.[26,32] The facial asymmetry that is often associated with DP or DS and seems to improve with growth. The face, unlike the cranium, continues to grow well into adolescence. Consequently, facial asymmetries that may be noted in an infant with DP or DS tend to become progressively less obvious over time. The improvement is relative and not absolute, and severe degrees of sagittal (anterior) shifting of auricular position can affect the fit of glasses later in life.

CONCLUSIONS

Deformational cranial flattening is a benign process that typically occurs over the first few months of life as a result of poor infant head mobility. Although many risk factors have been identified, nearly all have some impact on the ability of the newborn to reposition his/her head early in life. Cranial flattening stabilizes in early infancy. Shape changes that are present in infancy are generally fixed, but are mitigated by subsequent cranial growth (proportionate improvement), hair growth, and the angle from which the flattening is viewed (top-down in infants; posterior and side in adults). Most affected infants demonstrate a "preferred" head position, which is the earliest evidence of cervical tightness or imbalance. These infants should be considered at risk, and proactive preventative measures should be instituted. We prefer the adjustable concave resting surface for infants 3 months or younger. Older children who have more pronounced flattening that is conspicuous from a posterior or side view may be considered for helmet therapy. There is little evidence that untreated cranial flattening has cognitive or medical implications.

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