A recent article in the British journal, Archives of Diseases in Children, proclaimed that promotional literature for skull molding helmets (orthoses) for positional plagiocephaly contained “an element of commercial skullduggery.” But does the clinical science actually support this cynical point of view?

CASE VIGNETTES
Case 1
The 7-month-old infant girl is in your office for a routine checkup (see Figures 1A and 1B). She has been growing and thriving well, with each growth parameter of length, weight, and head circumference in the 60th to 70th percentiles. The baby is sitting calmly on the mother’s lap throughout your physical examination. However, as you finish examining the normal tympanic membranes, something bothers you about the infant’s facial appearance — you note that her face is asymmetric around the posterior skull, jaw-line, and forehead. You also observe a mild facial tilt to the left as well. You have already documented that her anterior fontanel is still patent; but as you assess the range of motion of the neck, you discover that the left sternocleidomastoid muscle is quite tight and thickened. In addition, her ability to flex the skull to the right shoulder is severely limited, whereas flexion to the left shoulder is normal.

Case 2
The 5-month-old infant male presents to your office with a runny nose and “ear tugging.” (See Figure 2A, page 498). He has been fussy at nights, with a poor appetite and low-grade fever. As you examine him, you are immediately alarmed by the facial and skull asymmetry, the tilting of his head to the right side and the downward displacement of right eye along with compression of the right side of the face.

You are somewhat reassured by the vertex view of the skull with the mild flattening of the left occiput (see Figure 2B, page 498). However, a more dorsal tangential view of the left occiput reveals marked flattening (see Figure 2C, page 498). Surprisingly, the lateral, anterior, and rotation of the neck flexion are all within normal ranges. His head circumference, height and weight growth parameters are following the same average growth curve since birth. You also confirm he has an incidental left acute otitis media.

Which of the two children (see Figures 1A and 2A, page 498) do you think warrants further evaluation for facial asymmetry? Does either child need radiographic imaging of the skull, helmet orthoses or referral to a physical therapist?
LONG-TERM EFFECTS OF POSITIONAL PLAGIOCEPHALY

A large longitudinal study of 7,609 Dutch infants, published in 2010, showed a persistence of occipital asymmetry in nearly 5% of all children by the third year of life. In addition, mild positional plagiocephaly (PP) and minor craniofacial asymmetry may persist into adolescence and adulthood, respectively.

By contrast, PP is not usually associated with developmental delays. Although vision defects and mandibular asymmetry are often observed in children with PP, they have not yet been causally linked to it. Another study longitudinally evaluated 161 children with PP who were treated with “repositioning” only. By preschool, only 61% of children achieved normal skull contour, and 4% still had severe residual deformities (see Figure 3).

INCREASED PP INCIDENT RATE

As a practicing general pediatrician for over 30 years, I have been amazed at the increasing incidence of malformed skulls and PP in the last decade. These children rarely present with the deformation within the first 2 months of life. PP usually peaks by 4 months old, and starts regressing by 6 months old. PP is the most common malformation of the skull with a reported 10-fold increase in prevalence since 1992. Although estimates of PP range from 3% to 48% depending on diagnostic criteria, the policy statement from the American Academy of Pediatrics (AAP) has settled on the alarming rate of 13% among healthy singleton infants.

PP is also known as deformational plagiocephaly or nonsynostotic plagiocephaly, because no cranial sutures are prematurely fused in the infant with PP. Most milder PP and many moderate cases of PP will spontaneously improve or resolve by 12 to 18 months old. However, it is the moderately severe or severe cases of PP, and cases of PP which are associated with significant torticollosis, that must be diagnosed early and more carefully managed by the general pediatrician.

In fact, at each well visit in the first 6 months of life, you should routinely take a few seconds to assess the gestalt of the facial symmetry, lateral neck flexion (for torticollosis), and a vertex view of the scalp for PP (see Sidebar 1, page 499).

During the first 2 years after birth, brain volume quadruples and brain size increases to 75% of its adult volume. PP must be differentiated from a premature unilateral coronal or lambdoidal synostosis (posterior synostoses), both of which will require surgical vault correction of the fused suture. Fortunately, these synostotic conditions only occur in about 1 in 10,000 children, and are actually an unlikely cause of plagiocephaly that you will routinely see. Vigilance is still key, however, particularly if the child’s skull is brachycephalic (see Figures 4A-C, page 499).

RISK FACTORS FOR PP

The major risk factors for PP are congenital torticollosis (see Figure 1A, page 497), male gender, multiple births, primiparity, breech births, low birth weight, and most importantly, the recent emphasis on infant supine sleeping with the “Back to Sleep” campaign. Congenital torticollosis due to a tight sternocleidomastoid muscle must be managed independently of the skull defect.

DIAGNOSIS OF PP

The diagnosis of PP is primarily based on clinical examination. When any significant or severe deformation of the skull is encountered, I recommend a specific evaluation of the skull (see Sidebar 1, page 499) on a monthly basis until...
you and the parent are satisfied with the skull improvement.

When PP is compared with posterior synostosis, the vertex geometric view of the skull appears to be more like a parallelogram (see Figures 5A and 5B, page 500), rather than a rhomboid. In PP both the ear and the anterior forehead appear displaced or “sheared” anteriorly on the ipsilateral side of the occipital deformation. PP tends to occur predominantly on the right occiput. Also, in PP, torticollis is often coincidentally present and sutural ridging of synostosis is absent. If torticollis is present, it must be managed as well.

Although rarely needed, according to the AAP policy statement, if you are unsure of the diagnosis of synostosis, plain radiologic films of the skull will usually be diagnostic, by showing whether or not the posterior sutures are patent. This is important because posterior synostoses may be associated with increased intracranial pressure and inhibition of brain growth, developmental delays, and poor self esteem and social isolation.

I think that one of the more complex decisions for any of us is how to manage the child with brachycephaly (see Figure 4). The entire skull shape is compressed in the anterior-posterior dimension, with marked widening of the coronal axis. These children warrant a skull radiograph, and prompt referral to a craniofacial team for evaluation for synostotic conditions (eg, Crouzon’s syndrome) and possible early orthotics, preferably by 4 or 5 months old.

**PREVENTION OF PP**

Because most mild and moderate PP will spontaneously resolve, during at least one of the routine visits at age 2 to 4 months, I recommend to parents that they give their infant “tummy time” for about 60 minutes or more when awake, several times daily. As early as 3 months, parents could consider placing the child in a “helmet orthosis” to correct their child’s head shape.

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**SIDEBAR 1.**

*EVALUATION OF THE SKULL FOR DIAGNOSIS OF POSITIONAL PLAGIOCEPHALY*

1. Head circumference.
2. Palpation of the anterior fontanel and each cranial suture for ridging.
3. Careful examination of the facial symmetry including the mandible, ears, eyes and forehead.
5. Viewing the skull from the vertex and more dorsal posterior views, particularly noting ear axis symmetry and any skull flattening (see Figures 2B and 2C, page 498).

*Source: Block SL*

**SIDEBAR 2.**

*HELMET ORTHOSES VS. ‘REPOSITIONING’*

**Helmet Orthoses**

- Should be instituted before 9 months, but deformational outcomes are likely much better if it is started before age 6 months.
- Is expensive: averaging about $3,000 for just the orthoses, plus requiring multiple office visits to both the orthotic supplier and the subspecialists. In addition, commercial insurance companies rarely cover costs of the helmet orthoses, except for post-synostoses surgery. Medicaid coverage is variable.
- May cause scalp and skin rashes, including seborrhea capitis.
- May make the parents self-conscious about their child’s problem.
- Require the child to wear the orthoses for about 23 hours daily for about 2 to 4 months at minimum.

**Repositioning**

- Is inexpensive.
- Requires at least 60 minutes of awake active “tummy time.”
- Requires dedicated parents who are willing and able to proactively and continuously alter their child’s head position and room logistics (daycare attendance will be a major hurdle).
- May arguably be more acceptable for females, who can rely on customarily longer hair as they age to cover their deformity if the PP does not improve.

*Source: Block SL*
in a “Jumperoo” (Fisher-Price) or a bouncy seat while awake, thus spending much less time on the back. I also recommend counter-positioning the infant so that he or she has to turn the head to the opposite side of the PP in order to see interesting stimuli in the room or crib. The layout of the child’s room should have the infant and the crib placed parallel to and directly against the far wall.

Other recommendations include avoiding prolonged time in the car and infant seats, and otherwise strategically placing the car seat in the room where the infant is forced to turn contralateral to the PP for the room’s activity. All feedings and cuddling the infant should avoid the flattened portion of the posterior skull as well.

ORTHOSES VS. ‘REPOSITIONING’

In fact, you were so alarmed by the severity of posterior PP in the child of Case 2, that you obtained skull radiographs, which were normal. You felt some urgency to rectify the PP in both Case 1 and Case 2, because of their “older” age, and because the earliest appointment to have the children seen by your routine referral cranial subspecialist was nearly 2 months later. You have read that the earlier the application of the cranial orthoses, the more robust the correction towards a cosmetic symmetry.\(^3,8\) Thus, you undertake the referral directly to the orthotic supplier for skull measurements and helmet orthoses, and begin follow-up yourself until your specialist can see the infants.

Yet you are perplexed by your own observations that most cases of significant PP do not really manifest until 4 months (probably linked to the infant Back to Sleep Campaign, my guess), and that over the next 2 or 3 months you are still supposed to observe the infants for the usual spontaneous improvements in PP. This approach however, may be suboptimal for maximum correction of PP, particularly if your subspecialist has a full schedule.

Once you encounter a child younger than 9 months with obvious PP, you must make a complex decision, which is lifelong for the child, as to whether the child merely needs active repositioning or helmet orthoses. Each therapy has its pros and cons (see Sidebar 2, page 499). But first, determine whether the child has any element of torticollis. Daily concomitant physical therapy of the neck muscles will be essential too, since it often improves not only the neck muscle tightness, but it also commonly improves the PP as well. Importantly, many children with more severe PP and concomitant torticollis will probably need helmet orthoses.

Lipira et al\(^2\) showed that for infants enrolled by a mean age of about 5 months, duration of treatment was significantly shorter for helmet vs. repositioning (3.1 months vs. 5.2 months). Mean reduction of cranial vault asymmetry was also somewhat better for the helmet compared with repositioning (4.5 mm vs. 3.4 mm). Head growth was equivalent in both groups. However, when helmet therapy was instituted even earlier in infants (< 4 months old) and compared with repositioning, the mean reduction in cranial vault asymmetry was even more robust: from baseline 11 mm to 3.5 mm vs. from baseline 9 cm to 8 cm, respectively.\(^8\) This is in distinct contrast with the previous conventional wisdom that recommended helmet therapy could be instituted even as late as 12 months. This approach was thought to allow spontaneous improvement to occur during 6 to 11 months of age, allowing the family to avoid the expensive and cumbersome helmet orthoses.

Thus the evidence now shows that the earlier the better for helmet orthoses therapy. It appears that the point of
no return, or lack of “moldability,” may not actually be 12 months. I surmise that waiting longer than 9 months of age may notably reduce effectiveness.

This will really complicate the PP decision process for us practitioners who generally utilize “watchful waiting” and “repositioning,” and then reevaluating cautiously for 2 to 6 months before deciding whether to: institute active repositioning alone; perform skull radiographs; refer to plastic surgery or cranial team for helmet orthoses; or initiate helmet orthoses ourselves, with referral for nonresponders. (see Sidebar 2, page 499). Yet, the cost of these orthoses is prohibitive for many families.

The goal of any PP therapy is to achieve a satisfactory cosmetic and aesthetic outcome for the child’s skull deformity and facial symmetry (see Figure 6A and 6B, page 500). But we still lack definitive studies that define: what constitutes pathologic head asymmetry that should be treated; how much more durable over years is the better improvement observed with orthosis intervention vs. repositioning; and what improvement threshold is acceptable for either approach.

TREATMENT OF TORTICOLLOSIS

Early correction with physical therapy of the neck muscular structure may alleviate some of the PP as well. Not only will many infants need a therapist, but the parents must be instructed to perform neck exercises at home three to four times a day, and many parents are bothered by hearing their child cry during this exercise. Stretching exercises of the trapezius and sternocleidomastoid muscles include rotating the head in three directions: 1) lateral rotation of the neck: chin to shoulder; 2) lateral flexion of the neck: ear to shoulder; and 3) neck flexion: chin to chest.

Each time, the head is held in these positions bilaterally for 10 to 15 seconds in sets of 3. If helmet therapy has been concomitantly instituted, these exercises are continued throughout helmet therapy.

Although rarely discussed, this physical therapy is expensive: it could require therapist visits 2 to 3 times weekly for 3 to 5 months, which often consists of 25 to nearly 60 visits; it could cost $2,000 to $4,000 out of pocket.

CONCLUSION

During the era of the “Back to Sleep” campaign for infants, positional plagiocephaly has recently become increasingly common (> 10%) in routine healthy infants during the first 6 months of life. Watchful waiting, aggressive use of “repositioning,” increased “tummy time,” and use of a bouncy seat is appropriate for nearly all infants with mild to moderate PP.

However, helmet therapy is not “skull-duggery,” because recent data suggest that for infants with severe PP (or PP associated with torticollis), helmet orthoses for 2 to 4 months provides more robust cosmetic improvement than does intense repositioning alone, especially if initiated before 6 months of life, but I have seen infants who do respond to helmets up to 9 months of age.

Months of expensive but necessary physical therapy of the lateral neck muscle is needed for the child with torticollis. Most adverse effects of helmet orthoses are minor and transient, except for the costs. Better algorithms for the evaluation, timing, management and referral pattern for moderately severe and severe PP are needed, particularly evaluating the cost effectiveness of therapy. Better longitudinal data of PP effects on dental, visual, self-esteem, and psychiatric problems are needed too. Helmet therapy, if deemed necessary, apparently should be considered by 6 months old and initiated by at least 9 months old; by 10 to 12 months old it will likely be less beneficial.

REFERENCES
