

# The 'Epidemic' of Deformational Plagiocephaly and the American Academy of Pediatrics' Response

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Historically, cradleboards and other external pressure devices have been used to intentionally mold children's skulls into desirable shapes to signify high social status, ethnicity, or both.<sup>1</sup> In the last 10 years, several tertiary care centers in the United States have noticed a marked increase in the incidence of unintentionally flattened asymmetric infant skulls.<sup>2,3</sup> Multiple terms have been used to describe this posterior skull deformation: functional synostosis,<sup>4</sup> plagiocephaly without synostosis,<sup>3</sup> deformational plagiocephaly,<sup>5</sup> positional plagiocephaly,<sup>6</sup> lambdoid positional molding,<sup>7</sup> occipital plagiocephaly,<sup>8</sup> and positional skull deformity.<sup>9</sup> Whatever the term used, the infants all had varying degrees of unilateral occipital flattening, forehead protrusion, facial asymmetry, anteriorly displaced ears, and almost all slept on their backs.<sup>2</sup>

## IDENTIFICATION

Determining whether the infant's occipital flattening is the result of deformation or craniosynostosis is the most important task. Several clinical features could distinguish between the two ( Table 1 ).<sup>10</sup> A "parallelogram" skull shape strongly suggests deformational plagiocephaly ( Figure 1 ). More facial asymmetry may occur with deformational plagiocephaly resulting from the frontal protuberance on the ipsilateral side of the occipital flattening.<sup>11</sup>

Sign	Indicates
Examination	
Palpate lambdoidal suture	Palpable ridge suggests synostosis
Inspect by aerial view	"Parallelogram-shaped" head suggests DP
Check ear position	Ear on flattened side more posterior or inferior than the other ear suggests synostosis
	Ear on flattened side displaced more anterior than the other ear suggests DP
Assess facial symmetry	Forehead protruding on the side of the flattening suggests DP
Observe unilateral "bald spot"	A unilateral bald spot suggests DP; physician should palpate occiput for synostosis

Adapted with permission from Biggs W. Diagnosis and management of positional head deformity. *Am Fam Phys* 2003;67:1953-1956

Table 1. Signs distinguishing deformational plagiocephaly (DP) versus synostotic lambdoid suture.

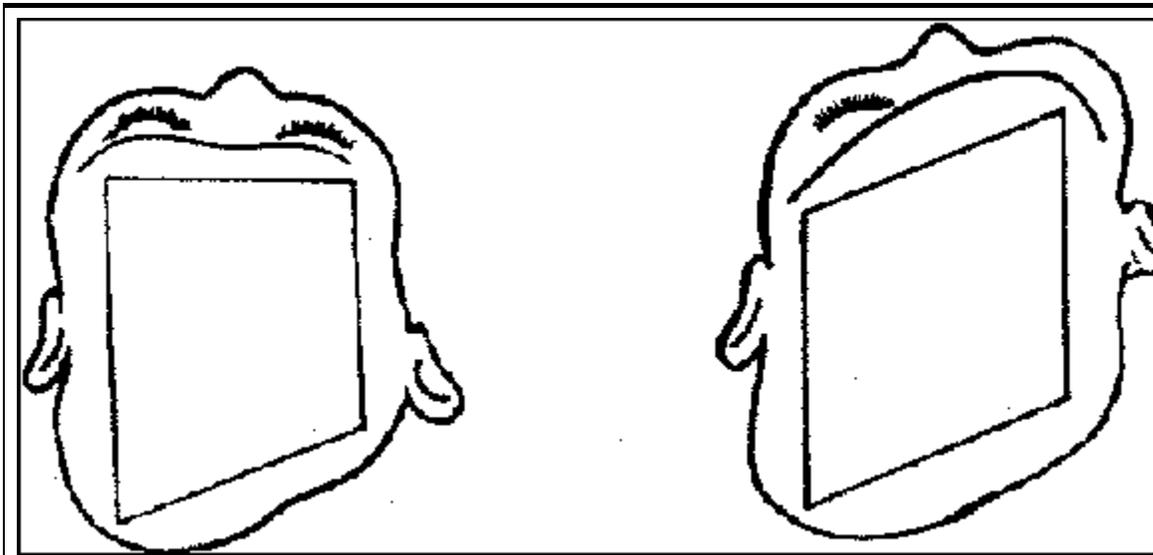


Figure 1. The head shape of lambdoid suture synostosis (A). The anterior protrusion of the forehead forms the characteristic parallelogram shape of the head of deformational plagiocephaly (B).

## INCIDENCE AND PREVALENCE

The increase in posterior deformational plagiocephaly since 1992 is presumed to be related to the American Academy of Pediatrics' recommendation to place infants in a supine sleep position to decrease the risk of sudden infant death syndrome (SIDS). The campaign was very successful with the incidence of SIDS dropping more than 40%.<sup>12</sup> However, an analogous increase in the posterior deformational plagiocephaly occurred. Several tertiary care centers with craniofacial specialists published data showing cases of deformational plagiocephaly increasing exponentially after 1992.<sup>3,13</sup> One study demonstrated a fivefold increase in posterior plagiocephaly comparing 1990–1992 with 1992–1994, and all infants, retrospectively, were found to be supine sleepers<sup>3</sup> (Figure 2). Because the U.S. data emanate from academic medical centers, the true incidence and prevalence of deformational plagiocephaly in America could be even larger than reported. These populations could reflect only cases geographically near the tertiary care centers or severe enough to be referred. The U.S. healthcare system is not conducive for an evaluation of deformational plagiocephaly of a large number of children presenting for routine visits. In The Netherlands, The Infant Health Care Program, their national system for well child care, provides for the evaluation and counseling for well child visits. In September 1995, the prevalence of deformational plagiocephaly presenting in 7609 children for well child examinations was noted to be 9.9% in infants less than 6 months of age.<sup>14</sup>

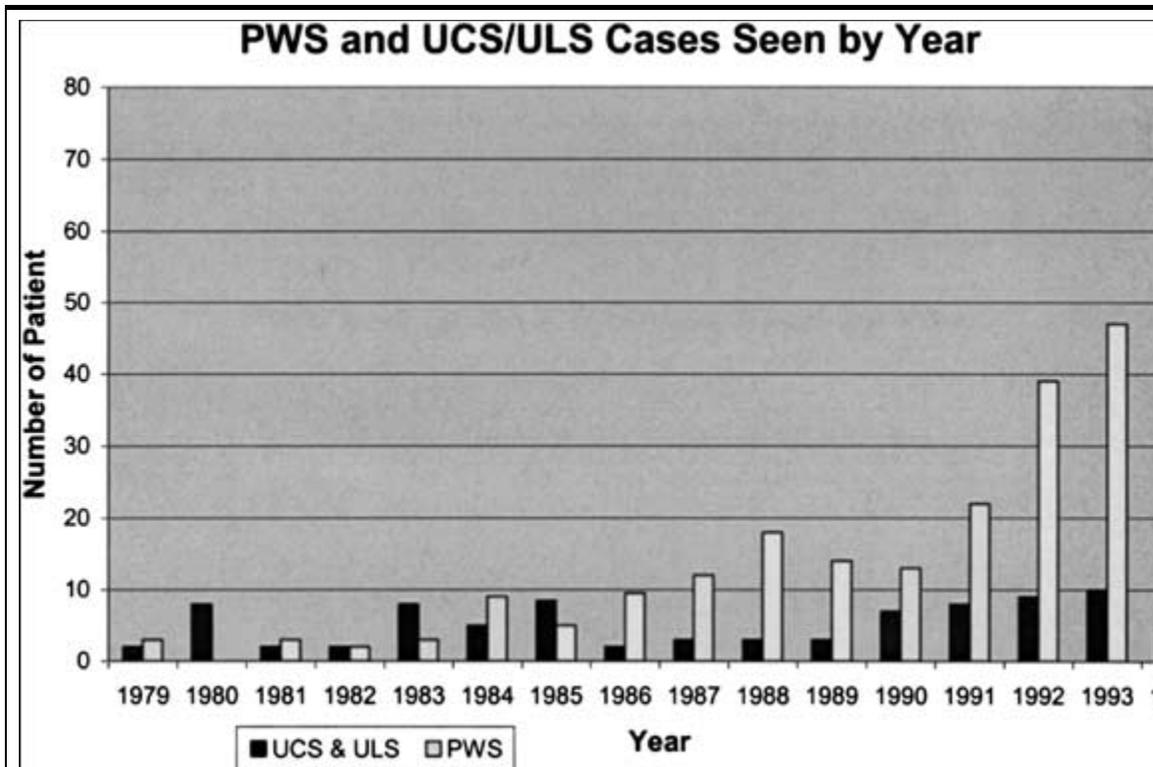


Figure 2. Increase in cases of deformational plagiocephaly (plagiocephaly without synostosis [PWS] versus unilateral coronal synostosis [UCS] and unilateral lambdoidal synostosis [ULS]) from Kane et al. <sup>3</sup> For each year, the bar on the left represents UCS/ULS and the bar on the right represents PWS. Observations on a recent increase in plagiocephaly without synostosis. *Pediatrics* 1996;97:877-885.

## CAUSES

Although pressure on the occiput of the growing infant's skull is the cause of deformational plagiocephaly, the origin for the side preference remains unclear. Multiple factors have been associated, encompassing intrauterine, neonatal, and environmental forces.

### INTRAUTERINE

Some have postulated that the occipital flattening develops in utero. Most fetuses lie in the left occiput anterior position. The pressure of the maternal pelvic bones on the fetal head could begin the unilateral flattening. <sup>15</sup> Infants from multiple births (twins, triplets, and so on) with in utero crowding are at increased risk for deformational plagiocephaly. In one study examining newborns 24 to 72 hours after birth, the incidence of localized cranial flattening in singleton infants was 13%, whereas in twins, the incidence was 56%. <sup>16</sup> Oligohydramnios similarly could cause intrauterine crowding and pressure on the developing head. <sup>17</sup>

### NEONATAL

Factors that develop during birth or in the neonatal period also increase the risk for deformational plagiocephaly. Assisted or prolonged labor or birth in a primiparous mother was associated with an increased incidence of localized cranial flattening within the first 72 hours of birth. <sup>15</sup> The presence of a cephalohematoma also predisposed the infant to develop a side preference when lying supine. <sup>15</sup> If a side

preference is determined prenatally, plagiocephaly could worsen during the first few weeks of life.<sup>17</sup> Posterior plagiocephaly has also been noted to occur more frequently in premature, low-birth-weight, and hypotonic infants, presumably as a result of weakened neck muscles affecting the infant's ability to reposition his or her head.<sup>18</sup> Similarly, the extra weight of an enlarged head in infants with hydrocephalus could predispose them to deformational plagiocephaly.<sup>7,17</sup>

Torticollis could be both a cause and effect of deformational plagiocephaly. If the infant is born with congenital muscular torticollis, the turn and tilt of the head from the shortened sternocleidomastoid muscle initiates the side preference and the occiput flattens correspondingly.<sup>19</sup> In the prevalence study done in The Netherlands in 1995, however, torticollis accounted only for 7.6% of positional preference found in infants less than 6 months of age.<sup>14</sup> Alternatively, the child could first develop a preference for head positioning, and the persistence of the head rotated in one direction could cause chronic shortening of the sternocleidomastoid muscle resulting in a torticollis.<sup>11</sup>

## **ENVIRONMENTAL**

The supine sleep position has been strongly associated with deformational plagiocephaly. When demographics are compared in studies, the majority of infants with deformational plagiocephaly sleep supine.<sup>2,3</sup> As the infant spends more time supine or reclining with his or her head on a hard surface such as in a carseat or swing, the occiput is more likely to deform.<sup>20</sup> Bottle-fed infants in which the bottle is always held in the same hand appear to develop a positional preference and a greater tendency to have deformational plagiocephaly.<sup>14</sup>

## **OTHER CHARACTERISTICS**

Posterior plagiocephaly has consistently been shown in many studies to be more common in male infants, with male:female ratios ranging from 1.58:1 to as high as 3:1.<sup>3,13</sup> The occipital flattening also tends to be more common on the right than the left, often at a 2:1 ratio.<sup>3,21</sup> Some congenital malformations such as congenital hip dislocation and congenital scoliosis are associated strongly with deformational plagiocephaly.<sup>17</sup>

## **NATURAL HISTORY**

The natural history of deformational plagiocephaly is unknown, because no studies have examined untreated populations of infants and children.<sup>8</sup> One study followed a population of children with deformational plagiocephaly until they were 2 to 3 years old.<sup>14</sup> The only intervention reported for positional deformity for this study was counseling parents on repositioning of the infant. At 7 to 14 months of age, 55% of children still showed signs of cranial or facial asymmetry, and at 2 to 3 years old, 53% still demonstrated some asymmetry. Unfortunately, what constituted "asymmetry" was not described or quantified. One study focused on conservative management of deformational plagiocephaly. With only sleep position modification and physical therapy, nearly all patients ( $n = 39$ ) improved. It was felt that most children would retain a mild deformity unlikely to resolve as they grew.<sup>21</sup>

## **OTHER CONSEQUENCES**

Consequences other than continued asymmetry of the face or head are less studied and are more controversial. Supinesleeping infants showed delayed motor milestones in gross motor movements requiring upper body strength (roll, sit, pull to stand) in comparison to a prone-sleeping group. In one study, the milestones were delayed but did not fall outside the range of normal,<sup>22</sup> whereas another study found the differences at 6 months of age to be significant.<sup>23</sup> In each, no difference was found between the groups for the gross motor milestones using lower body strength such as walking. At 18 months, there were no significant differences in skills between the groups.<sup>22,23</sup>

More controversial is whether some intellectual differences can be detected between children with deformational plagiocephaly and those without cranial deformity. One study demonstrated that 39% of children with persistent deformational plagiocephaly received special educational services versus 7.7% of their siblings (controls).<sup>5</sup> Male children with abnormal head shapes at birth associated with uterine

constraint were at highest risk.<sup>5</sup> Posterior cranial deformity could correct in 6 to 12 weeks with treatment, but the facial asymmetry could correct over a more prolonged period, up to 18 months.<sup>2</sup> No study, however, has examined children with deformational plagiocephaly to discover if facial asymmetry persists into adolescence. If the facial asymmetry persists, it could have adverse psychosocial consequences, similar to strabismus, which has been shown to hamper an adolescent's interpersonal relationships.<sup>24</sup>

## **MANAGEMENT**

Three treatment options exist for deformational plagiocephaly: 1) observation with or without minor physical intervention, 2) aggressive physical therapy with or without a static or dynamic cranial orthosis, and 3) surgery. Before the escalating incidence of deformational plagiocephaly after 1992, most mild and moderate cases were treated with observation or positioning, with surgery offered for severe cases. Torticollis was treated with aggressive physical therapy. A cranial orthosis was first introduced in 1979 to help remodel the skull.<sup>25</sup> Dynamic orthotic cranioplasty was also shown to be an effective skull remodeling method.<sup>26</sup> Different subjective and objective methods have been used to describe degrees of deformational plagiocephaly, thus it is difficult to compare results among studies. All methods appear to be effective to some degree and depend on the severity of deformation and intervention used.

## **AMERICAN ACADEMY OF PEDIATRICS' RESPONSE**

Academic craniofacial centers suggested that the increased prevalence of deformational plagiocephaly coincided with the 1992 American Academy of Pediatrics (AAP) recommendation that increased the number of infants placed on their "back to sleep." The AAP responded with the guide for the pediatric clinician in July 2003: the clinical report "Prevention and Management of Positional Skull Deformities in Infants."<sup>9</sup>

The diagnosis of deformational plagiocephaly is made primarily by history and physical examination. The clinician should assess any risk factors regarding the child's birth and the parents' assessment of head shape since birth. The clinician is urged to view the infant's head from the top at each health supervision visit up to 1 year of age. The "parallelogram" shape, with flattened occiput and the bulging ipsilateral forehead, is pathognomonic for deformational plagiocephaly (Figure 1). The location of the ears should be examined and compared. Anterior displacement of the ear ipsilateral to the occipital flattening suggests deformational plagiocephaly. Evaluation for torticollis is encouraged. The report states that "because the diagnosis of deformational plagiocephaly is made on the basis of history and findings on physical examination, imaging studies are unnecessary in most situations" (p. 200).<sup>9</sup> If imaging studies are obtained, skull radiographs are recommended for initial assessment. If equivocal, computed tomography (CT) scans could be helpful to rule out lambdoid suture craniosynostosis.

For management of deformational plagiocephaly, the clinical report focuses on prevention, mechanical adjustments or exercises, referral, and skull-molding helmets. Clinicians are recommended at the 2- to 4-week well child examinations to advise caregivers on alternating sleeping sites using prone positioning while the infant is awake and minimizing the infant's time in carseats and swings. If the child appears to be developing deformational plagiocephaly, the report instructs the clinician on exercises the caregivers can do simply to diminish torticollis and on positioning maneuvers. If, after a 2- to 3-month interval, the skull shape is not improving significantly, the clinical report recommends referral to a specialist in either pediatric neurosurgery or craniofacial surgery. The purpose of the surgeon's involvement would be to confirm the diagnosis, possibly suggest a "helmet," and facilitate a referral to physical therapy. The helmets (head orthoses) can be used in moderate to severe deformational plagiocephaly to improve head shape and facial asymmetry. Surgery is rarely indicated. The clinical report confirms that most cases of deformational plagiocephaly can be recognized and managed by primary care clinicians with simple interventions and counseling.

## **CONCLUSION**

An increased incidence of deformational plagiocephaly was noted after 1992. Although deformational plagiocephaly is associated with other factors such as torticollis, uterine constraint, and hypotonia, the large increase was attributed to the AAP recommendation of the "Back to Sleep" campaign in 1992. More infants were placed supine to sleep and, as a result, more infants unintentionally developed the unilateral occipital flattening of deformational plagiocephaly. Thus, the "almost epidemic eruption of posterior plagiocephaly" (p.

164)<sup>19</sup> demonstrates that the general population did follow the "Back to Sleep" recommendation. Hopefully, the public response will be similar for the AAP's clinical report "Prevention and Management of Positional Skull Deformities in Infants."<sup>9</sup> The report encourages clinicians to incorporate prevention counseling into the well child health examinations. Clinical diagnosis is clearly defined and some simple interventions described. The discussion regarding treatment focuses on conservative management with positioning and physical therapy. For moderate or severe deformity or deformity that is unchanged with physical therapy, a cranial molding orthosis could be used. Surgery is rarely needed. Hopefully, as clinicians and parents increase their awareness of deformational plagiocephaly, more cases will be prevented or treated conservatively sooner.

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