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On the Current Incidence of Deformational Plagiocephaly: An Estimation Based on Prospective Registration at a Single Center

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In 1992, the American Academy of Pediatrics (AAP) recommended supine sleeping to reduce the risk of sudden infant death syndrome. Although the incidence of deformational plagiocephaly is unknown, the consensus is that it has increased since this recommendation was made. To estimate the current incidence of plagiocephaly, we examined 342 infants for signs of deformational plagiocephaly, including occipital flattening, ear misalignment, frontal bossing, and facial asymmetry. Noticeable occipital flattening was documented in 15.2% of the infants (95% confidence interval, 11.6% to 19.5%); 1.46% had significant cranial deformities that also affected the skull base and face. Significant cranial asymmetry, defined as occipital flattening with concomitant skull base involvement and facial asymmetry, was observed in almost 1 in 68 infants. Adding to a growing body of evidence, our findings suggest significant increases in clinical deformational plagiocephaly since initiation of the AAP's "Back to Sleep" campaign.

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Deformational plagiocephaly (from the Greek *plagio* = oblique, *kephale* = head) refers to a condition in which

an infant's head becomes deformed as the result of external forces applied to the malleable cranium. This deformity can occur prenatally or postnatally. Prenatally, congenital deformities are associated with a restrictive intrauterine environment, including a small maternal pelvis, abnormal uterine structure, large or multiple fetuses, paucity or excess of amniotic fluid, or increased abdominal or uterine muscle tone. I-IO Postnatally, cranial deformation has been attributed to a supine sleeping position, 11-16 congenital muscular torticollis, 17-19 neurologic or cervical deficits, 20,21 premature birth, 9,10,21,22 and prolonged periods in car seats and infant carriers. 16,23

The condition is characterized by right or left occipital flattening, with anterior advancement of the ipsilateral ear and forehead. Although recently a topic of frequent discussion, plagiocephaly has been a documented clinical entity for many years. 24-31 In 1921, Greig defined the condition as a "deforma-

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tion of the skull in which one frontal region protrudes unduly forwards while the opposite occipital region projects unduly backwards."31 In 1961, Walker described the condition as an "oblique skull."32 Vulliamy is credited as the first to use the term "parallelogram skull" to describe the deformity?3

Although the historic prevalence of deformational plagio- cephalo cannot be known with certainty,34 it has been eStimated to occur in 5% to 48% of otherwise healthy newborns.28,29 In 1974, Dunn provided the most frequently cited incidence, esti- mating that deformational plagiocephaly and congenital muscu- lartorticollis occur in about 1 in 300 infants (0.33%)?5 In recent years, however, the incidence of plagiocephaly has been increas- ing, primarily related to a switch from prone to supine sleeping in the United States. 1 1-14,16,36,37 Many independent craniofacial centers have observed this increase, but the actual incidence of the condition is still unknown. The goal of the present investi- gation was to estimate the current incidence of plagiocephaly.

Patients and Methods

Between January and December 1999, infants and their par- ents were enrolled in a study of deformational plagiocephaly. All infants were less than 10 months old at the time of eval- uation and were seen with their parent during regular well- baby clinics at Pediatric Medical Associates in Mesa, Arizona. Of the 346 infants meeting these criteria, the parents of 342 agreed to panicipate; only 4 parents declined. The study was

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fully explained to the parents, and informed consent was obtained before their evaluation.

Participating infants were evaluated for signs of deforma- tional plagiocephaly, including occipital flattening, ear mis- alignment, frontal bossing, and facial asymmetry. To limit interobserver variability, all evaluations were conducted by a single physician (N .M.S.). Before the interview and examina- tion, the parents completed a detailed questionnaire on their child's prenatal and postnatal history that included (1) de- mographic data (age, race, sex), (2) perinatal data (gestational age, birth weight and length, plurality, mode of delivery, presentation at delivery, primiparity, forceps, suction, epi- duraVspinal), (3) potential etiologic factors (special in utero conditions, sleeping orientation, neck involvement, congen- ital or cervical anomalies, neurological deficits), and (4) the presence of plagiocephaly.

Similar demographic, perinatal, and maternal information was gathered for the state of Arizona from the Arizona De- partment of Health Services,38 and a ¥ analysis was per- formed to determine how representative the sample was of the Arizona population. Differences were considered signifi- cant if the probability level (ie, P value) was < 0.05.

Categorization of Plagiocephaly

A physical examination was perfonned on each infant to detennine the presence of plagiocephaly. The condition was ranked in severity from none to severe. The general criteria used by the evaluating physician (NMS) were as follows:

- .Mild: On clinical evaluation, some asymmetry or flatten- ing of the back of the head could be identified. An infant with mild plagiocephaly had minimal or no discrepancy between the location of the ears and no involvement of the face (prominence of the forehead, facial asymmetry).
- .Moderate: On clinical evaluation, asymmetry or flatten- ing of the occiput was immediately evident, and a dis- crepancy of half an inch or more existed with respect to the locations of ears. An infant with moderate plagio- cephalo may have some minimal facial asymmetry or forehead involvement.
- .Severe: On clinical evaluation, the infant had a notable and significant flattening of the occiput, ear misalign- ment of an inch or more, protrusion of the forehead on the affected side, and facial asymmetry.

Results

The sample consisted of 342 infants with a mean age of 3.4 months (standard deviation, 2-:2 months). Males were slightly overrepresented at 50.5%. Low binh weight < 2500 g) and very low binh weight < 1500g) were docu- mented in 6.4% and 0.6% of the infants, respectively. Pre- maturity < 37 weeks gestation) was documented in 10.8% of the cases.

Mothers under age 19 years represented 1.2% of the pop- ulation; mothers over age 30 years, 44.2 % . The infant was the mother's first-born children in 43.6% of cases and was deliv- ered by cesarean delivery in 23.4% of cases. Forceps and

Table 1 Summary of Classification of Infants With Deforma- tional Plagiocephaly

Severity	Number of Infants (%)
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290 (84.8)	45 (13.1)
2 (0.6)	
4 (1.2)	
1 (0.6)	
0 (0)	

suction were used in 2.6% and 4.4% of the deliveries, respectively. An epidural or spinal anesthetic was administered in 74.9% of cases. Plurality included four sets of twins, representing 2.3% of the study population.

Analysis of the cross-tabulated data showed no significant difference between the sample population and the Arizona population with respect to race, gender, gestational age, birth weight, plurality, primiparity, and mode of delivery. However, the sample population included significantly fewer mothers under age 19 years (1.2% vs 12.1%) ($P < 0.001$) and significantly more mothers over age 30 years (44.2% vs 32.6%) ($P < 0.001$).

In this study, the infants were reported to have slept in the following orientations: supine (34.2%), prone (5.3%), side(s) (51.8%), and a combination of positions (8.7%). At the time of this study, the overall sleeping position was highly non-prone. However, only 34.2% slept exclusively in the supine position. This low incidence suggests that an earlier recommendation from the American Academy of Pediatrics to sleep infants *only* on their back, rather than on their back and/or sides, may not yet have been practiced in the general population at the time of this study (1999).^{36,39-41}

In total, 52 of the 342 infants (15.2%) had some cranial asymmetry, occipital flattening, or both on clinical evaluation (Table 1). Forty-five (13.1%) were classified with mild asymmetry, whereas two (0.6%) were categorized with mild to moderate asymmetry. The parents were advised to reposition these infants off the existing flattened region and were shown neck-stretching exercises to resolve any neck involvement. Five infants (1.5%) had significant cranial asymmetry, defined as occipital flattening with concomitant skull base involvement and facial asymmetry. Four infants were classified with moderate deformity (1.2%), and one infant was diagnosed with a moderate to severe deformity (0.3%). The parents of these infants were referred to a specialist for further evaluation or directly referred for orthotic management of their infant's plagiocephaly.

Discussion

After the American Academy of Pediatrics (AAP) published the recommendation to place infants on their back to reduce the risk of sudden infant death syndrome (SIDS) during sleep, craniofacial centers around the United States began to observe an increase in the number of infants with positional plagiocephaly. By 1996, several studies had documented the apparent cause-and-effect relationship between supine sleep-

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ing and the development of plagiocephaly.¹¹⁻¹⁴ The condition became so prevalent that a special meeting of craniofacial and pediatric neurosurgeons was convened in 1997 to draft a position statement on the management of this condition.⁴² The most recent statements from the AAP Task Force on Infant Sleeping Position and SIDS has now acknowledged occipital plagiocephaly as a complication of a nonprone sleeping position.^{16,43}

To our knowledge, the present investigation is the first study to attempt to estimate the current incidence of deformational plagiocephaly in the United States. Previous studies have based their estimates on increased numbers of patients reponing to craniofacial clinics.^{11,12,14} In contrast, our findings derive from a prospective series of infants examined during routine pediatric visits. In a similar, recently published study from the Netherlands, the investigators observed occipital flattening, "the most common sign of deformational plagiocephaly," in almost 10% of their study population.⁴⁴ We observed occipital flattening in 15.2% of the infants evaluated, and like the Dutch authors, we found the incidence to be far greater than anticipated.

But is it appropriate to state that the current prevalence of deformational plagiocephaly is 10% to 15%? Many would argue that mild occipital flattening does not warrant classification as plagiocephaly and that including these cases inappropriately inflates the estimate. Consequently, we chose to estimate the incidence by focusing only on infants whose occipital flattening was associated with some degree of concomitant skull base and facial asymmetry. We believe that this subset more accurately reflects infants seen in craniofacial clinics across the United States. Given that restriction, the estimated prevalence decreases to a somewhat more realistic value of 1 in 68 (1.5%). Some may argue that this inclusion criterion is too liberal or too restrictive. However, the 95% confidence interval for this estimate ranges from a low of 1 in 158 infants to a high of 1 in 30 infants—a range well above the frequently cited incidence of 1 in 300. Thus, despite the restrictive definition, the current study adds to a growing body of evidence supporting a marked increase in the incidence of plagiocephaly.

The suggestion that supine sleeping is associated with an increased risk of deformational plagiocephaly should not be interpreted as criticism of this important recommendation. Since its inception, the AAP's "Back-to-Sleep" campaign has saved thousands of lives, and SIDS-related deaths have decreased by >40% since 1992.¹⁶ Given this dramatic reduction in SIDS, it is important that these recommendations continue to be followed. Therefore, pediatricians will likely continue to be confronted with a large number of infants with deformational plagiocephaly. As primary care providers, pediatricians can play a key role in reducing the incidence of plagiocephaly through early intervention and in educating parents about conservative preventive measures (eg, repositioning, physical therapy, supervised tummy time).

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