Beyond Cosmetic Concerns
Functional Deficits Associated with Deformational Plagiocephaly

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Functional Deficits Associated with Deformational Plagiocephaly

After discussing the rationale, timelines, and costs of cranial remolding orthoses, many parents of infants with plagiocephaly ask, "What happens if we don't correct this head shape?" As common as this question is, the literature fails to provide an unequivocal answer. For most families, recognizing that the prescribed intervention will improve the cosmetic concerns of an asymmetrical head shape is sufficient justification for the treatment. However, the essence of the original question remains in the minds of parents and practitioners alike. While the literature really doesn't tell us what happens in the absence of corrective treatment of plagiocephaly, it has begun to inform us about the various functional deficits that appear to be associated with the presentation of deformational plagiocephaly. These deficits, which will be discussed below, include apparent compromises in the functions of the ears, eyes, and jaw along with elevated risk of cognitive and motor delays. The purpose of this article is to bring clinicians up to speed on the documented deficits associated with plagiocephaly beyond the more obvious cosmetic concerns.

Otitis Media

Most parents are well acquainted with the frequency of ear infections or *otitis* media during the first year of a child's life. Running from the nasopharynx to the middle ear, the eustachian tubes serve to equilibrate middle-ear pressures and to drain the secretions of the middle ear into the nasopharynx. When the eustachian tubes fail to protect the middle ear from the secretions of the

nasopharynx, pathogens may enter the middle ear and lead to otitis media. Children have shorter, wider, more horizontal eustachian tubes, making them more prone to middle-ear infections. This vulnerability decreases with age as the tubes naturally shift from a horizontal to a more vertical position.

However, in children with plagiocephaly, these improvements in the geometries of the eustachian tubes are impeded, a supposition that is easily verified through computerized axial tomography (CT) scan.[1] There has been speculation for years that the altered cranial anatomy associated with plagiocephaly may further exacerbate the risk of otitis media. A recent study performed by researchers at Wake Forest University, Winston-Salem, North Carolina, provides a clearer picture about the extent to which an altered cranial anatomy affects the well-being of the middle ear.

Drawing from a convenience sample of patients seen and diagnosed with positional plagiocephaly at the North Carolina Center for Cleft & Craniofacial Deformities at the Wake Forest University Baptist Medical Center, the authors documented the status of the middle ear through the tympanometry technique, in which a pulse is emitted in the ear canal to assess pressure values. Of the 124 subjects tested, 121 (98 percent) had readings indicative of active, developing, or resolving otitis media, suggesting consistent compromised function of the eustachian tubes in draining the middle ear. Furthermore, parental reports of ear infections were more common among those children with more severe presentations of deformational plagiocephaly.[1]

Visual Defects

Two studies have been published that report on the prevalence of visual disturbances in children with deformational plagiocephaly. In the first of these, Gupta et al. affirmed that patients with deformational plagiocephaly do not appear to have a higher prevalence of *strabismus* (aka being "cross-eyed") or astigmatism.[2] However, the presence of deformational plagiocephaly does appear to be associated with other visual field defects.[3] Researchers from the University of Oklahoma, Norman, assessed the breadth of the horizontal visual fields of infants with deformational plagiocephaly, comparing the values of the left and right visual hemifields against one another and against normal controls. For the purposes of their reporting, two abnormal conditions were identified: hemifield asymmetries, in which the breadth of the left and right hemifields differed by more than 20 degrees; and constricted hemifields, in which a given hemifield was at least 20 degrees less than those established for normal controls of the same age.

Both conditions were observed with some frequency. Constriction of one or both hemifields occurred in 35 percent of patients, and hemifield asymmetries were identified in 17.5 percent of the tested subjects.[3] In addition, the data from the study population suggests a generally delayed progression of visual-field breadth compared with standard curves.[3] Interestingly, there was no statistical correlation between the side of the visual-field defects relative to the side of the occipital flatness. However, for the majority of the 16 infants for whom these comparisons were investigated, the visual defects were ipsilateral to the flattened occiput. A correlation between the severity of the hemifield constriction and the severity of the cranial asymmetry was also observed.

While these observations are certainly thought provoking, they do not speak to the natural history of visual-field development in infants with deformational plagiocephaly or to the possible effects of helmet therapy on these fields. They also do not speak to whether these defects are a product of plagiocephaly itself or the result of a more global developmental delay, a question that will recur with several of the other deficits that will be discussed below.

Mandibular Symmetry

Also relevant to the conversation are observations made at the Washington University School of Medicine, St. Louis, Missouri, shortly after the Back to Sleep campaign—which was introduced in 1994—lead to a rapid increase in the prevalence of deformational plagiocephaly.[4] At that time, CT scans were commonly obtained in this population to gain additional insights into this newer presentation. Scans of 23 such patients were available to analyze the shape and volume of the mandible, which, although not directly affected by positional forces, appeared to be affected by the altered shape of the cranium. Various landmarks were identified, along with associated distances and angles between them. This analysis found that the volume of the mandible on the affected side (i.e., the side of the occipital flatness) was roughly 4 percent larger than on the contralateral side.[4] Similarly, small but consistently significant differences were observed in several other defined linear measurements between the affected and non-affected sides of the mandible.[4] Unfortunately, given the preliminary nature of the article, neither the possible effects of these mandibular asymmetries on oral functions nor their response to interventions such as remolding orthoses were investigated.

Event-Related Potentials

One of the techniques used to predict the developmental outcomes of infants is the electrophysiological measurement of event-related potentials (ERPs). These electrical events within the brain are triggered by external stimuli and appear as small voltage changes on an electroencephalography (EEG), providing a quantifiable measure of neural information processing that can be safely obtained from infants.[5] In a preliminary Finnish study, the ERPs of ten infants diagnosed with plagiocephaly were recorded and compared to those of normal controls. The peak amplitudes of the measured ERPs were significantly larger in the controls, suggesting that infants with plagiocephaly were already experiencing compromised brain function.[5] More specifically, the lowered ERP amplitudes were suggestive of "depressed cortical sound processing," indicating an auditory processing dysfunction. Of the ten infants with plagiocephaly, only three demonstrated ERPs in the normal range. Interestingly, the ERPs were significantly smaller on the right side, which was the side of the posterior flattening for eight of the ten infants.[5] The study authors acknowledged that spontaneous recovery of these deficits may occur in the early years but stressed that these infants appeared to be at a greater risk of auditory processing disorders that may persist into the school-age years.[5]

Developmental Delay

A series of investigations have been published suggesting a relationship between both cognitive and motor delays in patients with plagiocephaly. Importantly, none of these papers attempt to

establish a causative relationship. As with the earlier observations regarding visual deficits, it is not known whether plagiocephaly causes these delays or whether the delays encourage the development of the cranial asymmetries. To the extent that correlations exist between the two, screening for developmental delays in children with positional plagiocephaly may be justified to facilitate the implementation of additional interventions where appropriate. For the purposes of this review, they will be divided according to the ages of the subjects assessed: infants, toddlers, and school-aged children.

Delays Observed During Infancy

Following up and expanding upon an earlier, similar study,[6] researchers from the University of Oklahoma reported their findings on 110 infants with deformational plagiocephaly.[7] Their assessment tool was the Bayley Scales of Infant Development, second edition (BSID-II), scoring system. This standardized assessment ultimately yields a mental and a psychomotor index. Normative values of both indices are well established, with mean scores of 100 and standard deviations (SD) of 15 and 16 points, respectively. As a result, the scores of a given infant can be readily classified as accelerated (greater than 1 SD above the mean), normal (between 1 SD above and 1 SD below the mean), mildly delayed (between 1 and 2 SDs below the mean), and severely delayed (more than 2 SDs below the mean). Normative population scores are also established, such that the percentages of infants from a random sample that will perform in each of these ranges are known.

Compared to the normative values, infants with deformational plagiocephaly were found to have significant differences in the mental and psychomotor developmental indices. Beginning with motor index, 90 percent of the infants with plagiocephaly were "normal" compared to the 69 percent that would be expected from random sampling. However, whereas random sampling would anticipate 16.5 percent of the tested infants to be "accelerated," none of the tested infants achieved an "accelerated" score. The frequencies of "mild" and "severe" delays, expected at 12.5 and 2 percent respectively, were observed at 7 and 3 percent respectively. So while mental delays were reduced, there were no cases of accelerated mental performance.

The observations of psychomotor development were more striking. Though a normal population sampling would anticipate a 15 percent prevalence of "accelerated" psychomotor performance, there were no such scores among the tested infants with plagiocephaly. "Normal" scores, expected at 73 percent, were observed in 74 percent. "Mild" and "severe" delays, anticipated at 11 and 2 percent respectively, were observed at 19 and 7 percent respectively in the infants with plagiocephaly. Thus, while there appears to be an association between deformational plagiocephaly and mental delay, there may be an even stronger association between plagiocephaly and psychomotor delay.[7]

A second, related investigation was recently published through a collaboration between the University of Washington, Seattle, and the Seattle Children's Hospital.[8] In this report, 235 infants with plagiocephaly were evaluated using the expanded Bayley Scales of Infant and Toddler Development, third edition (Bayley-III), which yields cognitive, language, and motor development scores. These were compared against scores obtained from a control group of demographically similar infants. As with the previous study, the infants with plagiocephaly fared

worse across all developmental domains. Where language and motor delays were observed in 10 and 9 percent respectively in the control group, they were identified in 19 and 20 percent respectively among infants with plagiocephaly. Cognitive delays were less common, occurring in 4 percent of the children with plagiocephaly and in none of the control population.[8] The most striking difference between the two cohorts was identified in the motor scores, where the infants scored, on average, ten points below the matched controls. Recalling that the SD on this index is 15 points, the implications are that as a cohort, the infants with plagiocephaly approached the categorization of mild motor delay.[8]

Delays among Toddlers

The same cohort discussed in the previous study were reevaluated at 18 months of age to determine to what extent the delays observed in infancy persisted with aging and development.[9] Across the three domains, children with plagiocephaly scored lower than their non-affected peers. However, the mean values for both groups approached the normalized means, with the control group scoring slightly above them and the affected group scoring just below them. Interestingly, the delays in motor development, which were more prominent in infancy, were slightly attenuated among the toddlers. By contrast, the differences in cognitive and language skills were as large or larger than those observed in the same children in infancy.[9] Thus, while the delays observed in infancy due to plagiocephaly appear to persist in toddlers, both affected and non-affected children score within the average range of normal scores at this age.

Delays among School-Aged Children

The only examination of developmental delays in school-aged children who had plagiocephaly as infants was co-authored by Sterling K. Clarren, MD, who first described the use of remolding orthoses in an earlier publication in 1979.[10] In this 2000 review, telephone interviews were attempted with the parents of infants who were evaluated with plagiocephaly between 1980 and 1991.[11] Of the 63 families who agreed to be interviewed, 40 percent reported that their children had subsequently received special help in grade school, including special education assistance, physical, occupational, or speech therapies, or children who otherwise required Individualized Education Plans (IEPs). This led the authors to conclude that the presence of deformational plagiocephaly in infancy may be a marker for early disorganization of the central nervous system that may later manifest itself during the school years. However, the authors were careful to note that the population in question developed plagiocephaly prior to the introduction of the Back to Sleep campaign. They continued to point out that children who are routinely placed on their backs and develop abnormal head shapes as a result are a different population than those children who spontaneously restricted their movements prior to the Back to Sleep campaign.[10] Unfortunately, the prevalence of delays in school-aged children who were diagnosed with infantile plagiocephaly following the introduction of Back to Sleep has not been reported.

Summary

The strongest argument in favor of cranial remolding orthoses remains the obvious aesthetic benefits. However, deficits in middle ear function, jaw proportion, and visual abilities have all

been reported. Furthermore, compromised electrophysiological brain events and motor delays have been reported during infancy, which appear to persist to some degree into toddler and school ages. Critically speaking, these deficits have only been associated with plagiocephaly; no causative relationships have been established. While plagiocephaly may cause delays, it is as likely that the delays themselves may predispose an infant toward the development of plagiocephaly. Future research will undoubtedly continue to clarify these relationships. Regardless, there is sufficient evidence to suggest that the deficits associated with plagiocephaly should not be considered purely cosmetic.

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