

Deformational Plagiocephaly: Recommendations for Future Research

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Compared with other topics in medicine, research on deformational plagiocephaly is relatively sparse. Among the published articles are a wide variety of studies ranging from simple case reports to well-designed cohort investigations. Unfortunately, variations in study design, methodology, sample size, and terminology often make it difficult to draw conclusions from this body of work.

As we look to the future of research in this field, we recognize that many issues still need to be addressed. However, before we can move forward, we must first understand what work has already been completed. Therefore, we first give a brief overview of the literature and then provide recommendations for future research.

SUMMARY OF THE LITERATURE

Finding articles written on the subject of deformational plagiocephaly can be difficult because terminology has been inconsistently used throughout the medical literature. The term plagiocephaly has been generically used to describe distortion of the cranium that occurs from both premature fusions of the cranial sutures (synostotic plagiocephaly) as well as from external molding forces (deformational plagiocephaly). Only recently has a clear differentiation between these two conditions been made, and it is now recognized that many of the articles previously reporting surgical intervention for craniosynostosis were really cases of misdiagnosed deformational plagiocephaly.¹⁻⁷

The deformational form of plagiocephaly has also been referred to in the medical literature under many different names, including positional plagiocephaly, nonsynostotic plagiocephaly, plagiocephaly without synostosis (PWS), occipital plagiocephaly, posterior plagiocephaly, benign position molding, functional lambdoid synostosis, skull molding, and flat head syndrome. Additionally, many early studies mention cranial deformation that occurred as a consequence of congenital muscular torticollis, scoliosis, neurologic issues, and so on, but never specifically use the term plagiocephaly. A Medline search will typically not identify these studies, and only careful review of cited references has turned up these older, yet relevant, articles.

Review of the literature will confirm that the majority of articles focus on either the etiology⁸⁻³⁷ (see also "[The 'Epidemic' of Deformational Plagiocephaly and the American Academy of Pediatrics' Response](#)" in this issue) or various treatments of plagiocephaly^{9,22,30-37} (see also "[The Management of Deformational Plagiocephaly: A Review of the Literature](#)" in this issue). A few studies discuss how to differentiate deformational plagiocephaly from craniosynostosis^{20,38,39} (see also "[An Overview of Craniosynostosis](#)" in this issue). The smallest group of studies examines the potential functional, developmental, and neurologic issues that could be associated with this condition.^{40,41}

FUTURE RESEARCH

The paucity of articles on plagiocephaly can be explained by the fact that until the mid-1990s, plagiocephaly was a relatively rare condition affecting only one in 300 infants.⁴² Today, conservative estimates predict the incidence to be as high as one in 50, representing a sixfold increase from the pre-1990 levels.¹⁸ It is no surprise then that roughly two thirds of the articles written on the subject have been published in just the last 10 years.

Considering the majority of articles have been written in the past decade, significant progress has been made in understanding the etiology, differentiating it from the more severe yet rare forms of craniosynostosis and the success of various treatments. A true milestone in these efforts was the 1997 Skull Molding Symposium in which the country's leading craniofacial and pediatric neurosurgeons met to develop a

consensus regarding the management of this condition.⁴³ Although a formal consensus was never reached, several significant findings were reported:

- Work on the differential diagnosis;
- A study describing how a diagnosis could be made by detailed clinical observation of the head shape and that computed tomography scans and x-rays were not required unless patency of the sutures was in question;
- Laying to rest of the concept of the "sticky suture" (not fused but acting like it is); and
- General agreement that surgery was not indicated for the majority of infants.

At the time of the symposium, the belief was that conservative interventions (repositioning and neck exercises) were all that was required for successful management of this condition. Use of molding helmets was not yet widely endorsed, because many specialists believed that this condition would round out on its own. Furthermore, molding helmet studies that had been done at that time had small sample sizes and were poorly controlled. The natural history of untreated plagiocephaly also failed to be available.

In the mid-1990s, it was also felt that all cases of abnormal head shape needed to be referred to a specialist because the pediatrician, family practice professional, and general practitioner were not qualified to make a differential diagnosis between synostotic and nonsynostotic plagiocephaly. This is a trend that has significantly changed in recent years, first, because the specialists have become overrun by the sheer number of infants presenting with this condition, and second, because more efforts have been made to educate the previously mentioned disciplines.

Even with many of these advances, there remain many critical issues to address. Many issues are debated within the medical community, and research needs to be performed to address concerns not only of physicians, but also of treatment providers and third-party payers. We present areas for future research that we believe are critical to the subject of deformational plagiocephaly.

STANDARDIZATION OF TERMS

The first issue to be addressed is the standardization of terminology. As discussed previously, significant confusion exists because of the wide variety of terms used to describe this condition. We do not favor the designations of occipital, posterior, or frontal plagiocephaly; these attempt to describe the deformity by where it occurs on the cranium. Deformational plagiocephaly is a multiplanar, three-dimensional deformity rarely isolated to a single location on the cranium. We also do not advocate designations that refer to this condition by what it is not such as nonsynostotic plagiocephaly, plagiocephaly without synostosis, or functional lambdoid synostosis. Moreover, we object to terminology that implies that the condition is insignificant as in "benign positional molding" or that suggest the condition is a syndrome as coined by the media in "flat head syndrome."

In the past, we have used the term positional plagiocephaly but now shy away from this because of how it has been misinterpreted by insurance providers. Positional plagiocephaly suggests that the deformity is caused *only* by postnatal positioning, and therefore it is the parents' responsibility to reposition the child off the flattened occiput.

We do agree with the arguments first put forth by Dunn,⁴² and promoted by Graham,⁴⁴ Bruneteau and Mulliken,⁹ and Mulliken et al.,²² that this condition should be referred to as deformational plagiocephaly, which clearly identifies it as a deformational process, not a congenital malformation. This deformational process can occur either prenatally (eg, restrictive intrauterine environment) or postnatally (eg, supine sleeping position), and thus this term does an excellent job of covering most known risk factors for development of plagiocephaly.

NATURAL HISTORY

As previously mentioned, to date, there have been no studies documenting the natural history of untreated plagiocephaly. There are still those who believe that deformational plagiocephaly will spontaneously resolve with normal growth and development. Conversely, arguments against spontaneous resolution have also been made. In 1981, Clarren³⁰ argues that "the Pacific Northwest Indians, who deliberately deformed their infants' skulls, discontinued treatment between 6 and 8 months of age because permanent deformity was

assured by that time" (p. 94). In 1999, Mulliken²² writes "we do not think there is significant 'self-correction' of deformational posterior plagiocephaly in childhood. The basis for this statement is the relatively minor slope of the cranial growth curve after infancy. In addition, we have found helmet therapy to be minimally effective after 1 year, and not effective after 16 to 18 months. Furthermore, we have measured several children who first presented at 14 to 18 months with major cranial asymmetry (12 to 18 mm difference), despite the fact that their parents had conscientiously used crib positioning" (p. 379). Regardless of where you stand on this issue, it is evident that the natural history of untreated plagiocephaly needs to be properly documented.

FUNCTIONAL PROBLEMS

Tied closely to the issue of natural history is the debate of whether there are any functional or developmental issues associated with untreated plagiocephaly. A strong motive for pursuing treatment is the concern that functional problems (ocular, auditory, temporomandibular joint, and so on) could occur later in life as the result of residual cranial deformity. Despite these concerns, there is no conclusive evidence demonstrating a relationship between functional problems and untreated plagiocephaly. Until functional problems are documented, insurance providers will deny treatment on the basis that plagiocephaly is a "cosmetic" issue.

TREATMENT OUTCOMES

Although many studies documenting the effectiveness of molding helmets have been performed, the criticism remains that many of these studies are not controlled, and the argument can be made that these heads would have rounded out in time without intervention. If improvement does occur with normal growth and development, it will be documented in the studies investigating the natural history as discussed earlier.

With respect to studies designed to document the effectiveness of cranial remodeling devices, future researchers would be advised to include a nontreated control arm if possible. Although this would provide the most conclusive evidence, it could be difficult to accomplish. The reason is that it requires some infants to be placed in an untreated category and therefore exposes the referring physician to both ethical issues and potential litigation should the untreated infants not improve. It must also be recognized that this research needs to be directed by physicians and not treatment providers, because treatment providers do not have the authority to consign patients to a nontreatment control arm.

We also emphasize the need for outcomes to be reported with both qualitative and quantitative evidence. As previously discussed (see ["The Management of Deformational Plagiocephaly: A Review of the Literature"](#) in this issue), many studies report success only subjectively, based on the satisfaction of either the physician or parent. Others have performed quantifiable studies using anthropometric measurements, yet fail to show any photographic evidence of the improvement reported. These articles are often difficult to interpret because most individuals do not have an intuitive feel for what 15 mm of cranial vault asymmetry is or what an improvement of 6 mm means.

The best solution is to supplement any quantitative study with solid photographic records. Alternatively, the most conclusive evidence will likely come from the development of three-dimensional digitization systems.^{45,46} However, much work still remains to prove that the data acquired by these systems are accurate and reliable.

COST-EFFECTIVENESS

In today's healthcare environment, demonstration that a device is safe and effective is not enough to ensure availability in the marketplace. Although clearance from the U.S. Food and Drug Administration (FDA) is a critical first step, reimbursement by third-party payers is an increasingly important factor. Higher standards exist for obtaining reimbursement and are typically set by the Center for Medicare and Medicaid Services (CMS). CMS determines which products and services will be covered, and other third-party payers frequently adopt these coverage decisions.

Specifically, manufacturers need to show not only that their device works, but also that their device provides improved outcomes or a cost reduction over existing therapies. Therefore, a medical device needs to be not

only safe and effective, but also reasonable and necessary.⁴⁷⁻⁴⁹ Suffice it to say, not every drug or device approved by the FDA as "safe and effective" is considered "reasonable and necessary" by CMS.

If cranial remodeling devices are to be covered by thirdparty payers, evidence that treatment of plagiocephaly prevents future functional problems might need to be provided. Until this time, insurance providers will argue that deformational plagiocephaly is a cosmetic issue with no long-term ramifications. Without evidence that the untreated condition persists into adulthood, third-party payers will also continue to argue that plagiocephaly spontaneously improves in time and will promote repositioning well beyond when it is considered effective (< 5 months of age) because this is the one treatment alternative that costs them nothing.

TREATMENT GUIDELINES

The objective of the 1997 Skull Molding Symposium was to develop a consensus regarding the management of plagiocephaly. Unfortunately, a consensus was not reached as a result of lack of information regarding the natural history of plagiocephaly, as well as evidence demonstrating success of various treatment options. Today, we still struggle with many of the same issues.

Nonetheless, the significant number of infants requiring intervention calls for some form of guidelines to be developed and endorsed by a reputable medical organization. These guidelines should recommend a course of action dependent on the age of the infant and severity of the condition and will need to include the full spectrum of treatment modalities from simple observation to repositioning, molding helmets, and even surgery.

Although evidence is not available to conclusively address each and every controversy, significant clinical experience from experts who have spent years treating these infants should provide guidance that is both reasonable and appropriate. The hope that a consensus will be reached among the various medical professions (craniofacial surgeons, neurosurgeons, pediatricians, family practice professionals) is not realistic because each profession views the problem from its own unique perspective. Additionally, the guidance established today based on current knowledge of plagiocephaly can be revised as additional evidence from future research becomes available.

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