

POSITIONAL PLAGIOCEPHALY: PATHOGENESIS, DIAGNOSIS, AND MANAGEMENT

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Positional plagiocephaly is a deformation resulting from intrauterine constraint or postnatal positioning leading to asymmetrical cranial growth. There has been a steady increase in referrals for positional plagiocephaly following the release of the American Academy of Pediatrics recommendation of supine infant sleeping position to prevent Sudden Infant Death Syndrome (SIDS) in 1992, largely because of poor parent education on the risks of prolonged occipital pressures. While this deformity is fairly easy to manage when diagnosed early, treatment can become more difficult and complicated with prolonged course. Because of this, it is essential that primary care physicians and parents be educated on recognition of positional plagiocephaly, prevention strategies, and treatment options. In milder cases, where diagnosis is made early, the deformation can be managed by stretching exercises and regular prone positioning, while in more severe cases molding helmets may be needed. Following appropriate treatment, success rates for acceptable cranial shape may be as high as 92%.

INTRODUCTION

In order to accurately diagnose and manage an infant presenting with a flattened occiput, it is essential to determine the cause of the asymmetry. The condition is known as plagiocephaly, literally "oblique head."¹ Most cases of plagiocephaly are presumably the result of the natural pressures applied to the infant skull as it passes through the birth canal, but other forces contribute to both the etiology and persistence of the problem. Most recently, the Sudden Infant Death Syndrome (SIDS) prevention regimen known as the "back to sleep" program has played a role in the dramatic rise in incidence. Another contributing factor in the promotion of plagiocephaly is the role of the neck musculature on the dynamic molding of the cranium. These neck forces are involved both in maintaining the head position on the flat area and, if left untreated, in prevention of skull base remodeling.

Despite conservative treatment, some cases of plagiocephaly persist. Persistent occipital asymmetry may be caused by premature fusion of skull sutures, particularly the lambdoid suture (craniosynostosis), craniofacial synostosis syndromes, metabolic bone disorders, depressed skull fractures, excessive intracranial volume (hydrocephalus), and diminished intracranial volume (microcephalus). Positional plagiocephaly, also known as deformational plagiocephaly and plagiocephaly without synostosis (PWS), does not fit into any of the above categories. It is formed following intrauterine constraint, postnatal positioning and torticollis, which leads to asymmetrical cranial growth.²

Both prenatal and postnatal pressures can cause positional plagiocephaly. Prenatal causes include uterine compression, intrauterine constraint, and extrauterine compression. A common historical feature is prolonged labor with the head resting low in the pelvis. The infant's sleeping position, congenital muscular torticollis (CMT), neurological and cervical defects, as well as premature birth, are postnatal causes of positional plagiocephaly.³

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If the head is repositioned most cases will spontaneously reverse. A more severe condition develops if the infant continues to apply pressure to the flattened occipital surface. In this situation, gravitational forces act on the already flattened surface causing the skull to assume a parallelogram appearance.⁴ In severe cases, the ear ipsilateral to the flattened occiput is displaced anteriorly, the contralateral occiput shows bossing, and there may be significant forehead asymmetry. Prolonged unidirectional positioning of the neck, as may occur in these patients, may lead to shortening of the sternocleidomastoid muscle leading to sternocleidomastoid muscle imbalance.¹

In 1992 the American Academy of Pediatrics (AAP) recommended, after noting an increased risk for SIDS with prone and side sleeping positions, that all healthy infants be placed to sleep supine.⁵ Prior to the AAP supine sleeping recommendation, at least 70% of infants were placed to sleep prone. Since the recommendation in 1992, only 20% of infants are placed in the prone position, and the SIDS rate has decreased by more than 40%.⁶ A complication of the recommended supine sleeping position, or "back to sleep campaign," is prolonged pressure on the infant's occiput, leading to an increase in cases of positional plagiocephaly. A year following the initial AAP recommendation, a 30% increase in referrals for positional plagiocephaly was observed, as well as continued increases in subsequent years. These increases were not noted in referrals for other craniofacial anomalies.² While positional plagiocephaly is a serious complication of the AAP recommendation, it is important to note that the supine position is still the favored position for healthy infants as it does successfully decrease the risk for SIDS.¹

PREVENTION

Prevention of permanent plagiocephaly requires early intervention. Early intervention actually prevents the fixed deformity with the resultant psychosocial consequences of the “funny looking head.” The diagnostic clues of positional plagiocephaly are often detectable at birth or within the first month. With this in mind, the education of parents and caregivers is focused on two areas. The first is early diagnosis, which makes early intervention more effective due to the plastic nature of the neonatal cranium. The second area is the intervention itself, which will be reviewed under the management section.

DIAGNOSIS

The diagnosis of positional plagiocephaly is made by focusing on two areas of the cranium. The first is the skull base. The appearance of the child with skull base asymmetry is ear auricle asymmetry. This is measured concretely by the subnasal-to-ear tragus distance (see Figure 1). A difference of 10mm or more is considered significant. If this distance is equal, it supports that the skull base is not involved, but does not always reflect the status of the upper cranium. The appearance of the upper cranium is another determinate of positional plagio-

cephaly, which reveals occipital flattening either bilaterally or most often unilaterally. With further growth, the cranium assumes a warped appearance in the configuration of a parallelogram. The cranial measurement to document this is the frontozygomatic-to-contralateral occipital (euryon) distance (see Figure 1).¹⁰ A difference of greater than 10mm is considered significant.

The abnormality of the neck in this diagnosis is either a result of the cranial deformity or is the etiology for the positional plagiocephaly. In neonates, neck motion is minimal. If unilateral positional plagiocephaly is present, the cranium rests on the flat area maintaining the asymmetric length of the sternomastoid muscle, which is maintaining the asymmetric force of the muscle on the skull base. This in turn contributes to the asymmetry of the cranium. The clinical diagnosis that is the result of plagiocephaly is called sternomastoid muscle imbalance, which perpetuates the unilateral plagiocephaly. The neck disorder that most commonly causes plagiocephaly is called torticollis. Both of these neck problems respond to physical therapy in most cases. In some cases of torticollis, surgical release is required.

MANAGEMENT

Capitalizing on the plasticity of the neonatal cranium is the essence of intervention. The first intervention is based on repositioning. The AAP-recommended supine positioning is for sleeping only. Parents and caregivers are taught to alternate the side on which the infant is laid to sleep. This is done before the child is able to reposition itself during sleep. This can be performed at set intervals or alternate sides based on days of the week. In terms of awake repositioning, prone positioning, commonly referred to as “tummy time,” is the cornerstone of therapy. It is important both for shoulder girdle development, which is essential for motor progression, and pressure prevention on the flat surface. This position is frustrating for the child due to low upper body strength, but diminishes with improved strength. When the

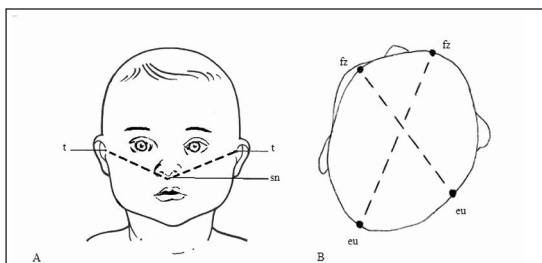


Figure 1. Frontal (A) and bird's-eye (B) views of key landmarks used in evaluating abnormal head shape. Calculation of cranial vault asymmetry involves measuring the difference between the measurements of left frontozygomatic (fz) to the right euryon (eu) and the right frontozygomatic (fz) to the left euryon (eu). Calculation of skull base asymmetry involves the difference between the measurements of the subnasal to the right and left tragi (t), respectively.

child's neck musculature is strong enough to maintain head elevation, the use of sitting devices are utilized.

In regards to the neck, if a tight sternomastoid muscle is detected, the parents are instructed on a home neck-stretching program. This regimen consists of simply stretching the neck towards the nonflat side, slowly bringing the chin to the shoulder. This position may take time to achieve depending on the degree of sternomastoid tightness. The duration of the stretch is 5-10 seconds and is performed at each diaper change. This maneuver is performed by placing the child in the supine position holding one hand on the upper chest and the other hand turning the head to align the chin with the shoulder. A second component of this stretching is tilting the head to bring the ear towards the shoulder. If the tightness is severe or a diagnosis of torticollis is made, a physical therapist is utilized to manage the therapy. In more severe cases, screening radiographs are obtained to rule out cervical anomalies.⁶

The use of cranial molding orthotics is utilized when repositioning therapy has failed and the deformity remains severe. If the age of presentation is delayed and the deformity severe, proceeding to a molding helmet is appropriate because waiting for the response to repositioning therapy would eliminate the molding helmet option. The window of opportunity for cranial molding is best at 4-6 months of age, but remains effective to some degree later, especially if the child is premature or is developmentally delayed. The success of cranial molding therapy is based on the principal of rapid brain growth during the first year as well as the plasticity of the youthful cranial bone. The helmet places a restraint on the round areas of the cranium while brain growth forces the flat areas to round out. The average duration of treatment is 3 months. The helmet is worn for 22 hours a day until an acceptable shape is achieved.

OUTCOME

While estimations of the incidence of positional plagiocephaly vary between 1 in 300 live births

before the back to sleep campaign,^{7,8} to between 1 of 68 and 1 of 72 live births after the AAP recommendations,⁹ it is clear that it is much more common than the more severe lambdoid craniosynostosis with an incidence of around 1 in 100,000 live births.⁷ Several factors contribute to making actual calculations of prevalence quite difficult. For example, in the majority of patients, the diagnosis is not detected at birth even in children managed in a neonatal nursery for other postnatal complications. Another confounder is the fact that many families do not recognize this as a deformity and are not alerted to this problem by their pediatrician.

In our practice, we evaluate an average of 210 children per year as a tertiary referral service and refer an average of 48% for cranial molding therapy. The overall success rate is 92%, which is based on an acceptable cranial shape. The success rate drops to 87% for acceptable skull base symmetry with fairly normalized ear symmetry. In closing, the financial burden of this clinical disorder is now shifted to the family due to strategic wording in health insurance policies. This denial contributes to the underlying stress for many families already anxious about their newborn.

CONCLUSIONS

Successful management of positional plagiocephaly requires early intervention. It is essential that primary care physicians be educated on detecting positional plagiocephaly, in addition to understanding appropriate management options. Physicians should educate parents on the AAP back-to-sleep campaign, as well as the measures needed to prevent positional plagiocephaly as a consequence of prolonged supine positioning. If the condition is detected in the first weeks of life and parents implement appropriate strategies to halt progression of the deformity, the need for helmets and other molding devices as well as the possibility of surgery are highly unlikely.¹¹

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