Nonsyndromic Craniosynostosis and Deformational Head Shape Disorders

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KEYWORDS

• Craniosynostosis • Deformational plagiocephaly • Positional plagiocephaly
• Nonsyndromic craniosynostosis • Treatment of craniosynostosis • Cranial vault reconstruction
• Minimally invasive • Helmet therapy

KEY POINTS

- The incidence of infant head shape abnormalities is increasing. Physicians should be able to identify these patients and distinguish between deformational plagiocephaly and craniosynostosis.
- Deformational plagiocephaly does not have a known negative impact on the brain; however, early diagnosis and treatment is needed to correct the dysmorphic head.
- Most craniosynostoses are nonsyndromic and include sagittal, metopic, coronal, lambdoid, and multisuture synostosis. Surgeons should be able to make a diagnosis based on clinical findings.
- Surgical indications for craniosynostosis are to correct the abnormal craniofacial appearance, prevent negative effects of increased intracranial pressure on the brain and optic nerves, and protect the globe of the eye.
- Cranial vault reconstruction immediately corrects both the fused suture and cranial abnormality; minimally invasive procedures allow for improvement of the cranial shape over time.

INTRODUCTION

A persistent abnormal head shape is a concerning finding in an infant and can be from craniosynostosis (intrinsic) or deformational plagiocephaly (DP; extrinsic) causes. It is imperative the treating physician can differentiate between the 2 abnormalities to initiate appropriate treatment and avoid any long-term sequela. *Craniosynostosis* is the premature fusion of 1 or more cranial sutures, causing an abnormal head shape. This early fusion of the cranial sutures restricts normal skull growth, causing not only a dysmorphic head shape but also possible increased intracranial pressure leading to neurocognitive impairment. *Deformational plagiocephaly*, in contrast, is an atypical head shape caused by extrinsic forces pushing on the soft, malleable skull bones. The cranial sutures remain open and functional, with no risk for increased intracranial pressure causing impairment to the developing brain.

DEFORMATIONAL PLAGIOCEPHALY

Also termed *positional plagiocephaly* or *nonsynostotic plagiocephaly*, this asymmetric head shape abnormality has increased in incidence over the past 2 decades and is the leading cause for atypical head shapes.[1–5] The cause of DP is asymmetric external forces on the soft infant calvarium, which are typically created by gravity pushing the infants head against the crib mattress

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with an equal but opposite force pushing back on the head (**Fig. 1**). In 1992, the American Academy of Pediatrics initiated the Back to Sleep Campaign in which infants were placed on their backs during sleep to reduce the risk of sudden infant death syndrome.[6–10] This campaign dramatically deceased the incidence of sudden infant death syndrome, but resulted in a significant increase in DP.[4,5] A recent study in Canada found that nearly 47% of infants between 7 and 12 weeks of age had some degree of DP.[11] Children with torticollis are at increased risk for DP, because they have limited head movement and lie with their head turned to only 1 side. Any preference of head position should be viewed as an early manifestation of torticollis and a high risk for DP.[12] Other risk factors for DP include prematurity, developmental delay, multiple gestation pregnancy, male gender, assisted delivery, primaparity, uterine abnormalities, oligohydramnios, and breech presentation. Each of these factors cause intrauterine deformation and/or increase the risk of limited head movement during the first months of life.[5,7,13] Deformational changes of the skull can lead to various head shapes, depending on what part of the head is positioned against the surface of the bed (**Box 1**).

Treatment of Deformational Plagiocephaly

Parental education about the etiology of DP and the importance of alternating the sleeping position by placing the infant supine and turning the head to either side will help to prevent the deformity from developing.[5,9,13] Infants with torticollis, or any evidence of head position preference, should undergo physical therapy to allow full range of motion of the neck.[13] The underlying goal of repositioning therapy is to keep the infant from lying on the flat part of the head. This includes monitored tummy time during the day, avoiding a car seat when not in a vehicle, and encouraging free and spontaneous movements of the infant.[12] If the infant is young (<3 months) and the deformity is mild, repositioning of the infant to prevent lying on the flattened portion of the head may be all

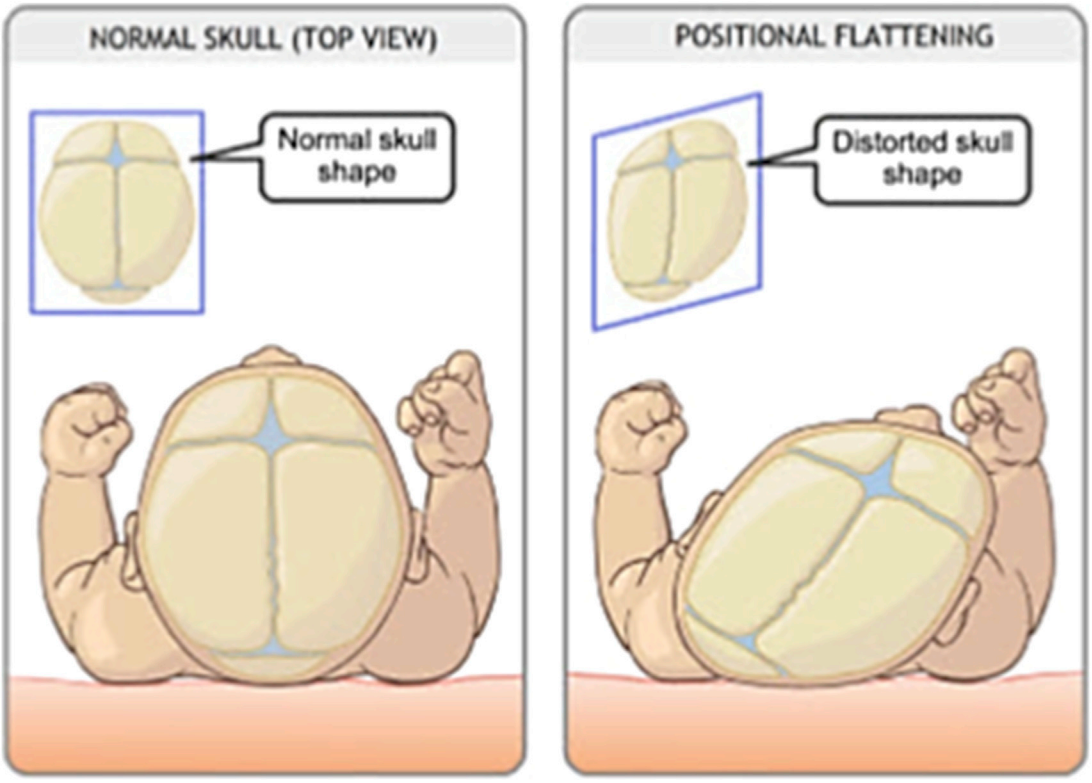

Fig. 1. Normal head shape versus deformational plagiocephaly. This figure depicts the changes that occur to the soft cranium with deformational plagiocephaly. In the illustration on the right, there is flattening of the right occipital region and forward advancement of the right frontal region. (*From* Mortenson P, Steinbok P, Smith D. Deformational plagiocephaly and orthotic treatment: indications and limitations. Childs Nerv Syst 2012;28(9):1408; with permission.)