Abstract

Positional cranial deformities are a group of disorders that result in asymmetrical cranial vault due to the application of prolonged exterior pressure on the skull. These can be classified as deformational plagiocephaly, deformational brachycephaly and deformational scaphocephaly, in order of frequency. Clinical awareness of the condition has increased due to its increased incidence resulting from sleep practices advocated by the American Academy of Pediatrics to reduce sudden infant death syndrome. Therapy is directed towards cosmetic concerns, and treatment options include observation, active head repositioning, physical therapy, cranial orthoses and surgical treatment in a few very severe cases. In this paper, the efficacy of helmet use to treat positional cranial deformities is reviewed.

ABBREVIATIONS

PCD: Positional Cranial Deformity; DP: Deformational Plagiocephaly; DB: Deformational Brachycephaly; DS: Deformational Scaphocephaly; TCD: Transcranial Difference; CVAI: Cranial Vault Asymmetry Index

INTRODUCTION

Definition of positional cranial deformity and types

Positional cranial deformity (PCD) is defined as skull asymmetry due to external forces unrelated to the premature fusion of one or more of the cranial sutures [1]. PCD is a common problem of infancy. With the implementation of the 'back to sleep' campaign promoted by the American Academy of Pediatrics in 1992, incidence has increased by up to 46% [2]. The campaign resulted in decreased incidences of sudden infant death syndrome with the promotion of supine sleeping in infants but consequently increased the number of parents seeking medical attention for head deformities. PCDs can be classified as deformational plagiocephaly (DP), deformational brachycephaly (DB) and deformational scaphocephaly (DS).

Deformational plagiocephaly (DP)

DP is the most common form of non-synostotic cranial deformity, resulting from the exertion of persistent unidirectional force on the back of the head [3]. These results in unilateral flattening in the parieto-occipital region, which may be accompanied by a compensatory volume shift towards the ipsilateral frontal bone and on the bi-parietal plane, and it, may in turn lead to ipsilateral prominence of the maxillofacial structures and anterior displacement of the ipsilateral ear (Figure 1 and 2).

There are various risk factors of DP including sex (males have higher rates of DP), multiple pregnancy, prematurity, difficult labor, congenital anomalies and prolonged supine position. Some authors even postulate that antenatal factors may play part in disease etiology [4]. Many risk factors result in DP via limited self-mobilization of the infant, thereby rendering the patients head immobile in prolonged durations.

Among the clinical entities that may accompany DP are intellectual impairment, developmental delays, visual disturbances, otitis media, decreased motor tone and, occlusal problems [5].

Deformational brachycephaly (DB)

DB is the symmetrical flattening of the back of the head, which can lead to prominence of the temporal areas, making the head wider (Figure 3).

Deformational scaphocephaly (DS)

DS is the flattening of the parietal region without significant occipital asymmetry. DS is frequently seen in premature infants that have been left to sleep for prolonged periods in a lateral position (Figure 4).

Diagnosis of positional cranial deformities

Referrals to craniofacial surgeons and neurosurgeons have increased since awareness of PCD amongst the public and physicians’ has increased. Concern from parents and primary care pediatricians is of paramount importance for the early diagnosis of the condition. The most important fact to know is whether the infant has a head positional preference, which, is commonly seen as an early manifestation of congenital muscular...
assessments tools for the diagnosis and follow-up of patients with DP [5,6]. TCD defines the absolute difference between two oblique measurements of the head, whereas the CVAI can be calculated via the following formula: (longer oblique measurement − shorter oblique measurement)/shorter oblique measurement x100. A CVAI of 0% indicates perfect symmetry whereas a CVAI higher than 3.5% denotes DP [5]. Severity stratification of the deformity can be conducted using a CVAI and the classification scheme reported by Wilbrand et al., [6]. The deformity can be further classified into three categories using the following CVAI ratios: 3-7% as mild, 7-12% as moderate and >12% as severe [6].

A study led by Couture et al., showed that Argenta classification is another effective stratification method based on morphological parameters [1]. In Argenta classification, deformity is classified into five types according to morphological parameters like forehead asymmetry, ear asymmetry, occipital bumps and diagonal difference [7].

DB and DS are almost always quantified using a cephalic index (CI), which is the maximum width (biparietal diameter) of the head divided by its maximum length (occipitofrontal diameter).

**Treatment of positional cranial deformities**

The primary objective of treatment is cosmetic improvement. It is very doubtful whether PCD leads to neuro developmental delays, although vice versa is thought to be reasonable through limited head repositioning by the affected infant. Treatment options for PCD include observation, active head repositioning, physical therapy, pre-fabricated or individualized orthosis and in severe cases surgery [1]. Head repositioning in infants with a mild deformity or infants at high risk of developing deformity is a reasonable measure in the first few months (<4 months of age) as older children fail to respond to manipulation [5]. Another conservative measure is encouraging 'tummy time' to decrease the time spent in a supine position.

**Helmet therapy for positional cranial deformities**

Inpatients refractory to conservative measures, helmet therapy can be instituted. Helmet therapy was first utilized by Clarren et al., in 1979. Some authors have set a TCD of 10 mm or above in DP and 9 mm or above in DB as thresholds for helmet therapy [5].

The rationale behind helmet therapy is based on the fact that skull enlargement will proceed towards the area with the least resistance. Cranial orthoses can be classified as active or passive depending on whether pressure is applied to the protruding parts of the skull. Active cranial orthoses work by applying firm pressure to the bulging parts of the skull, whereas in passive helmets, space is left for the effected region, and the other parts of the helmets are in close proximity to the skull without applying pressure. In other words, passive orthoses allow room for growth in the flattened areas and apply minimal pressure to areas with bossing, whereas active orthoses apply compression to the bossed areas, possibly resulting in a more rapid deformity correction [8].

Helmet therapy indications are not clear, and decisions are based either on clinical grading systems or anthropometric measurements [6,8]. Parents are generally advised that the
helmet should be worn 23 hours a day, although shorter durations have been published [7]. It is unclear when to stop treatment; in addition to clinical grading and anthropometric measurements and parents’ satisfaction levels must be taken into account [5]. The cost of customized cranial orthoses can be significantly high, up to 3000 USD, and the reluctance of insurance companies to reimburse this cost is a difficult for parents.

**Efficacy of helmet therapy for positional cranial deformities**

In a study based on 1050 PCD cases, a correction rate of 81% was achieved using helmet therapy, irrespective of deformity severity [1]. A mean CVAI of 9.8% was seen to decrease to 5.4% in a study of 213 patients, with the highest rate of correction on younger patients than 24 weeks [3]. In another study, performed with 62 infants on the timing of helmet therapy, it was shown that the early therapy was instituted, the better the correction rate in a shorter duration. Infants were classified into two groups depending on the age when helmet therapy was initiated, either younger or older than 6 months. Improvements to asymmetry were found to be significantly better in patients in the helmet therapy group than 6 months [9]. Our clinical experience accords with those results (Figure 5 and 6).

In a study comparing helmet therapy to conservative therapy in 171 infants, both groups showed significant reductions in deformity, but the reduction in the helmet group was significantly better [7]. Kwon assessed the efficacy of helmet therapy using sonographic examinations on 26 patients and reported a CVAI reduction from 9.3% to 3.5% [10]. Lee et al., investigated the effects of helmet therapy on mid-facial deformity correction and reported significant deformity even in patients who started helmet therapy at the mean age of 22.5 months; however, they claim that patients who started helmet therapy earlier had significantly better results [11]. Contradicting the majority of published data, the only randomized clinical trial on the issue found no difference in deformity correction with helmet therapy compared to the natural course in a cohort of 84 patients aged 5-6 months [12].

Complications of helmet therapy include scalp issues, either due to direct pressure or hypersensitivity, non-fitting orthoses, and psychological distress associated with unsolicited attention directed towards helmet users; complications reported in around 26% of patients [3,13]. Complications of helmet therapy in patients with combined DP and DB were found to be significantly higher compared to those with isolated deformities, but overall complication rates were found irrespective of deformity severity [6].

**REFERENCES**
