Deformational Plagiocephaly: A Review

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Deformational plagiocephaly (DP) is a common condition and the leading cause of head shape abnormalities in infants. It is characterized by asymmetrical distortion of the skull resulting from external forces on the back of the head. DP has become more prevalent in infants and has increased dramatically since the introduction in 1994 of the “Back to Sleep” campaign. Management of this condition is not often covered by insurance, and can be costly and lengthy; however, DP can be prevented. Early recognition and management can make a significant difference in patients’ outcomes and reduce the cost of treatment. Nurses play a key role in recognition and prevention, are instrumental in educating parents and caregivers, and have a significant impact on preventing and reducing the risk of DP. Current systematic approaches to clinical assessment, diagnosis, and management strategies of DP can help nurses who care for infants and their families understand DP and take the necessary steps to prevent it.

Key Words: Deformational plagiocephaly, sudden infant death syndrome (SIDS), cranial helmet, deformational posterior plagiocephaly, positional plagiocephaly, posterior plagiocephaly, occipital plagiocephaly, nonsynostosis plagiocephaly, plagiocephaly without synostosis.

Deformational plagiocephaly (DP) is the leading cause of head shape abnormalities in infants, and it is characterized by an asymmetrical distortion of the skull resulting from external forces (Rogers, 2011a, b). A persistent or prolonged resting head orientation or supine position may expose the infant’s rapidly growing and malleable cranial bones to external forces. Over time, this position will lead to cranial deformation and flattening of one side of the skull, resulting in DP (Pogliani, Mameli, Fabiano, & Zuccotti, 2011). A common condition, DP has been referred to by many names, such as deformational posterior plagiocephaly, positional plagiocephaly, posterior plagiocephaly, occipital plagiocephaly, nonsynostosis plagiocephaly, or plagiocephaly without synostosis (Flannery, Looman, & Kemper, 2012; Laughlin, Lueressen, & Dias, for the Committee on Practice and Ambulatory Medicine Section on Neurological Surgery, 2011; Looman & Flannery, 2012; Shweikeh, Nuño, Danielpou, Krieger, & Drazin, 2013).

Background

DP has become more prevalent in infants and has increased since the introduction of the “Back to Sleep” 1994 campaign, which recommended placing healthy infants on their back to sleep, based on the evidence that the supine position might reduce the incidence of sudden infant death syndrome (SIDS) (American Academy of Pediatrics [AAP] Task Force on Sudden Infant Death Syndrome, 2011). A study conducted at a large, 1,004-bed, tertiary medical center located in North Carolina reported a 390% increase in DP referrals, from 9 to 316 patients, between 1996-2007 (Branch et al., 2015). The incidence of DP was previously reported at 8.2% (Boere-Boonekamp & van der Linden-Kuiper, 2001). However, a study by Mawji, Vollman, Hatfield, McNeil, and Suavé (2013) indicated the incidence of plagiocephaly was estimated at 46.6%. In Texas, Sheu, Ethen, Scheuerle, and Langlois (2011) documented and described a dramatic increase in plagiocephaly of more than nine-fold, from 3.0 cases to 28.8 cases per 10,000 live births, which was equivalent to an average annual increase of 21.2% per year from 1999 to 2007. Within the pediatric population with DP, prevalence varies with age. It is highest in infants at four months of age (Hutchison, Hutchinson, Thompson, & Mitchell, 2004).

Previously, DP was considered a minor and purely cosmetic problem by many practitioners (Pogliani et al., 2011; Speltz et al., 2010), but it causes parental anxiety, and can require costly and lengthy management (McKinney, Cunningham, Holt, Leroux, & Starr, 2009). Additionally, in recent years, potential association with learning disabilities, language disorders, virtual-perceptual problems, motor delays, and problems with attention span have been assessed in regard to DP (Pogliani et al., 2011).

Motor development has been of interest. Kennedy, Majnemer, Farmer, Barr, and Platt (2009) compared motor development between infants with and without DP, and suggested that infants who have decreased exposure to prone positioning may not only have higher incidence of DP, but also may be at risk for a delay in the acquisition of certain motor skills. Speltz et al. (2010) assessed the neurodevelopment of 235 infants with DP and 237 infants without DP using the Bayley Scales of Infant Development III, and found that DP is a marker of elevated risk for delays and seems to be associated with early neurodevelopmental disadvantage in motor functions. Kordestani, Patel, Bard, Gurwitch, and

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Learning outcome, instructions for completing the evaluation, and statements of disclosure can be found on page 63.
Panchal (2006) studied neurodevelopmental delays in children with DP when compared with a standardized population, using the Bayley Scales of Infant Development II scoring system, and found infants with DP to have psychomotor and mental developmental indexes significantly different from those of the standardized population ($p < 0.0001$).

Development outcomes were the focus of Steinbok, Lam, Singh, Mortenson, and Singhal (2007), who studied 65 children at least 5 years of age who had been diagnosed in infancy with DP. Altogether, 33% of these children had received learning assistance, and 14% of them were in special needs classes. Similar results were obtained by Miller and Clarren (2000), who studied long-term developmental outcomes in 63 school-age children who presented as infants with DP without obvious signs of delay at the time of initial evaluation. Of these children, 39.7% received special help in primary school, including special education assistance, physical therapy, occupational therapy, or speech therapy, generally through an individual education plan, while only 7.7% of their siblings had required similar services.

Siatkow ski et al. (2005) studied visual field defects in infants with DP. They found 35% of the infants had constriction of one or both hemifields by at least 20 degrees, and 17.5% of the infants had hemifield asymmetry of 20 degrees or more. This indicates that DP may affect visual field development, but neither the laterality nor the severity of skull deformity is predictive of the severity of visual field defects.

The relationship between DP and developmental delay appears to be undefined (Kordestani et al., 2006), and the evidence of association between DP and health outcomes is limited (Pogliani et al., 2011; Rogers, 2011b). However, infants with DP should be screened and monitored for motor development and developmental delays or deficits (AAP Task Force on Sudden Infant Death Syndrome, 2011; Pogliani et al., 2011).

Nurses are not only integral primary healthcare providers who should be familiar with and understand DP because it is the most common type of cranial asymmetry in infancy (Pogliani et al., 2011), but they are also the primary educators of new parents before childbirth. It is well documented that supine sleeping positioning has significantly increased the incidence of DP; however, it is vital that the supine sleeping position should be maintained for SIDS protection. Thus, it is very important for nurses who care for infants and interact with families to be aware of positioning, whether during sleep or during supervised awake periods (Chizaw sky & Scott-Findlay, 2005), and provide care, modeling, and education to avoid DP.

**Etiology**

Many studies have identified risk factors associated with DP (see Figure 1) (Bialocerkowski, Vladusic, & Wei Ng, 2008; Dec & Warren, 2011; Joganic, Lynch, Littlefield, & Verrelli, 2009; Losee, Mason, Dudas, Hua, & Mooney, 2007; McKinney et al., 2009; Oh, Hoy, & Rogers, 2009; Rubio et al., 2009; van Vlimmeren et al., 2007).
Diagnosis

Early recognition, diagnosis, and treatment are essential to achieving the best outcomes (Kluba, Kraut, Reinert, & Krimmel, 2011). Early and accurate diagnosis, especially differentiating between synostotic (craniosynostosis) and deformational (non-synostotic) plagiocephaly, is very important because the diagnosis directs the course of treatment (Dec & Warren, 2011; Looman & Flannery, 2012; Pogliani et al., 2011). Synostotic plagiocephaly or craniosynostosis refers to a rare condition caused by the premature fusion of one or more cranial sutures. If the diagnosis of synostotic plagiocephaly is confirmed, surgical correction is required (Kabbani & Raghuveer, 2004); however, surgery for correction of DP is rare (Robinson & Proctor, 2009; Shweikeh et al., 2013).

The diagnosis of DP in infancy is made primarily on the basis of history, and it is confirmed by physical examination that should be performed at birth and at each health supervision visit up to one year of age (see Figures 2 and 3). In each patient with a skull deformity, a complete head-to-toe examination focusing on the face and skull, and examining the infant from anterior, posterior, and vertex views, should be performed. Neck examination is essential in infants with suspected DP; range of motion and head movement should be tested. Clinically, infants with DP most often present with ipsilateral frontal bossing, ipsilateral occipitoparietal flattening, contralateral occipital bossing, and ipsilateral displacement of the ear anteriorly. Computed tomography (CT) scanning can be used selectively to make the diagnosis in complex cases and differentiate DP from synostotic plagiocephaly (Looman & Flannery, 2012).

Management Strategies

Common management options for DP include repositioning therapy, helmet (orthotic) therapy, or a combination of these (Flannery et al., 2012; Paquereau, 2013). Early treatment of DP is essential (Flannery et al., 2012; Kluba et al., 2011). The age at the initial visit and the severity of DP are important considerations in determining whether infants should be treated with repositioning or helmet therapy (Kluba et al., 2011; Xia et al., 2008).

Repositioning therapy is the most conservative intervention (Flannery et al., 2012) and involves efforts to prevent the child from turning to the flattened side. Infants with mild or moderate DP who are younger than four months of age can be treated with repositioning therapy (Dec & Warren, 2011; Rogers, 2011b). Research indicates that the earlier infants with DP present for repositioning therapy, the more likely they are to improve compared with those who present later (Hutchison, Stewart, de Chalain, & Mitchell, 2010).

A helmet is commonly used and referred to by many names, such as a cranial helmet, cranial orthotic device, cranial orthosis, cranial band, orthotic, orthotic helmet, molding helmet, or pneumatic orthotic cranial molding helmet (Goh, Bauer, Durham, & Stotland, 2013; Kluba, Kraut, Calgeer, Reinert, & Krimmel, 2014; Paquereau, 2013; Seruya, Oh, Taylor, Sauerhammer, & Rogers, 2013). Helmet therapy is usually recommended for infants who present with moderate to severe DP, signs of significant anterior craniofacial deformities, or developmental delay, and for infants who do not improve with repositioning therapy (Graham, Kreutzman et al., 2005; Kluba et al., 2011, 2014). Research suggests that starting helmet therapy for infants with DP between five to six months of age is important and leads to a significantly better outcome in a shorter treatment time (Kluba et al., 2011).
However, the challenge with helmet therapy is that it requires strict compliance with wearing the helmet 23 hours per day, carries an additional financial cost, and requires frequent modifications of the helmet to accommodate for head growth and correction of the deformity over a period of two to six months of therapy, depending on the age of the infant and severity of the DP (Dec & Warren, 2011; Poglani et al., 2011). Additionally, the cost of a helmet is high (approximately $1,500 to $3,000 each) and often is not covered by insurance (Lee et al., 2010) or is considered with reluctance by third-party payers (Robinson & Proctor, 2009). Moreover, at present, the Centers for Medicare and Medicaid Services (CMS) (2016) does not have a national coverage determination for helmets for treatment of DP.

Controversy exists about diagnosis and management of DP (Shweikeh, 2013; Xia et al., 2008). Management of this condition has generated much research and debate (Seruya et al., 2013) because there is no gold standard for evaluating and quantifying the deformity (Paquereau, 2013), and there is no standardized management protocol for the treatment of DP (Lee et al., 2010). Additionally, a lack of defined standards for initiating therapy or monitoring change of the DP and a lack of standardized tools to measure treatment outcomes impedes prompt diagnosis and treatment (Goh et al., 2013; Lee et al., 2010). It is believed that treatment decisions are influenced more strongly by referral and physician bias than by medical evidence (Lee et al., 2010).

A recent research study reported helmet therapy had no added value in the treatment of DP as compared with conservative repositioning treatment. The authors concluded that the use of a helmet as a standard treatment should be discouraged (van Wijk et al., 2014). In the United Kingdom and New Zealand, helmets are not readily available, and treatment has concentrated on positioning recommendations and reassurance that it will improve over time (Hutchison et al., 2010; McGarry et al., 2008). This approach was supported by the studies of children without helmet therapy by Hutchison, Stewart, and Mitchell (2011). They compared head shape measurements, parental concerns about head shape, and developmental delays in children between the ages of three to four years who were diagnosed in infancy with DP and were treated without helmet therapy. They found that 61% of head shape measurements reverted to the normal range, parental concerns were reduced from 85% to 15%, and 41% of children who initially had one or more delays decreased to 11%.

There remains a lack of high quality randomized controlled trials according to a current systematic review by Goh et al. (2013). The authors concluded that there is a lack of evidence supporting the use of helmet therapy and there are controversies surrounding the use of helmet therapy, such as appropriate use, cost, use in older children, and long-term outcomes. High-quality randomized controlled trials are needed to evaluate the efficacy of helmet orthotic therapy versus repositioning therapy.

Prevention

Prevention is paramount. DP can be and should be prevented (AAP Task Force on Sudden Infant Death Syndrome, 2011; Laughlin et al., 2011). The most effective way to eradicate DP is to educate parents and other caregivers about the importance of proper infant positioning to prevent abnormal appearance of head shape, provide postural support, and facilitate normal growth and development. Parental education is one key to prevention and treatment of DP, and it needs to be instituted by primary caregivers when infants are at a very early age (Rogers, 2011b).

It is well documented that supine sleeping position has significantly increased the incidence of DP; however, it is vital that the supine sleeping position should be maintained for SIDS protection. From birth, it is essential to address the importance of the supine sleeping position, but equal weight should be given to educating parents about changing the head position frequently, and the use of prone and upright time when awake and under supervision (AAP Task Force on Sudden Infant Death Syndrome, 2011; Dec & Warren, 2011; Laughlin et al., 2011). “Tummy time,” when children are prone, should start from the first day at home from the hospital. Parents and caregivers should be encouraged to provide supervised tummy time for infants while they are awake, at least two to three times per day (three to five minutes each time), increasing the amount of time as the infant shows he or she enjoys the activity (AAP Task Force on Sudden Infant Death Syndrome, 2011; Laughlin et al., 2011) to reduce the incidence and severity of DP.

At the first sign of occipital flattening, repositioning and tummy time guidelines should be promptly initiated to correct the deformity. The development of DP may indicate that parents are not providing their infants with tummy time (Graham, Gomez et al., 2005). Additionally, parents should be instructed to a) avoid using the car seat when infant is not in a car, b) limit seating that maintains the supine position, and c) promote tummy time while infants are awake and being supervised. Moreover, parents should be made aware that although most DP cases improve spontaneously by two years of age, some anomalies in head shapes do not revert to normal without intervention; therefore, parents should be very attentive to positioning strategies while the infant is still very young (Rogers et al., 2011b).

Healthcare providers must teach parents to position infants for sleep, play, and development once discharged (Waitzman, 2007). Nurses represent the largest group of healthcare providers (Kurtzman, Dawson, Johnson, & Sheingold, 2010) and play a vital role throughout the continuum of care (Wilson, Whitaker, & Whitford, 2012). Therefore, nurses should have a significant impact on preventing and reducing the incidence of DP. Recent literature indicates that nurses can significantly reduce the incidence of DP (8.5% vs. 25.6%, p < 0.05) (Lennartsson, 2011). However, Koren, Reece, Kahn-D’angelo, and Medeiros (2010) indicated that only 55% of postpartum mothers and 26% of mothers at two months after birth received information regarding how to position the infant when the infant was awake. Results indicated that a lack of clarity and education for both parents and providers might have failed to communicate the importance of tummy time because it was limited in terms of frequency and duration. There is a need for parental education about positioning infants during infants’ sleep and awake times. There should be ongoing, systematic educational programs for nurses that can improve nurses’ knowledge and early recognition of DP, and parental
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2. Evaluations must be completed online by April 30, 2019. Upon completion of the evaluation, your CNE certificate for 1.3 contact hour(s) will be mailed to you.

Learning Outcome

After completing this learning activity, the learner will be able to discuss deformational plagiocephaly and the necessary steps to prevent it.

Learning Engagement Activity

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References


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