**Cranial Molding Deformity:**

**Evidence for Prevention and Treatment in the NICU**

**Introduction**

Cranial molding deformity (CMD) is an alteration in the normocephalic shaping of the skull due to an external mechanical force.1 CMD occurs in up to 48% of newborn infants and is generally mild and reversible in nature.1,2 Infants are particularly susceptible to this condition due to malleability of the skull; however, this property combined with a rapid rate of head growth during the first year of life is also what allows for reversal of deformity through the application of corrective forces.1 This review discusses current available literature related to the types of CMD as described by cranial shape, the various etiologies and risk factors for CMD, and the evidence for conservative interventions with a particular focus on premature infants in the neonatal intensive care unit (NICU).

**Abnormal Cranial Shapes**

Various abnormal cranial shapes can develop based on the area of the skull to which pressure is applied over time, the most common of which are plagiocephaly, brachycephaly, and dolichocephaly.

**Plagiocephaly**

The term plagiocephaly comes from the Greek for “oblique head,” and this condition is defined as unilateral flattening of the occiput that results in an asymmetrical head shape.3,4 The peak prevalence of this cranial molding deformity occurs at 4 months of age.5 Plagiocephaly has been described in the literature for several decades, though it was not widely studied prior to the initiation of the “Back to Sleep” campaign in 1994 due to a relatively low prevalence of 1 in 300 children.6,7 In addition to unilateral occipital flattening, the characteristic presentation of plagiocephaly may also include ipsilateral frontal bossing, contralateral occipital bossing, contralateral frontal flattening, anterior displacement of the ipsilateral ear, and prominence of the ipsilateral cheekbone.3,7 Clinically, plagiocephaly can be diagnosed by observation, with the vertex view typically being most advantageous, as well as by the objective measure of Cranial Vault Asymmetry Index (CVAI).8 CVAI is calculated by anthropometric measurements of the head in millimeters as indicated in the following equation:8

$$CVAI= \frac{diagonal A-diagonal B}{diagonal A} x 100$$

These measurements can be obtained through the use of a cranial caliper, and diagonal A represents the greater length compared to diagonal B.9 Diagonals are measured beginning at 30\* from the center of the nose.9 A CVAI of less than 3.5 is indicative of cranial symmetry that is within normal limits, whereas a value of 3.5 or greater suggests that a repositioning program or more intensive form of treatment is required in order to correct the asymmetry.9

**Brachycephaly**

 In contrast to plagiocephaly, brachycephaly is a symmetrical head deformity that is characterized by bilateral flattening of the posterior cranium.4 This pattern of deformity often results from spending a prolonged period of time with pressure against the back of the head, which can occur from a car seat, a baby bouncer, or supine sleeping in a position of minimal neck rotation.1 Infants with brachycephaly typically present with a skull that appears wider than the normocephalic skull, and this condition is less prevalent in full-term and premature infants than both plagiocephaly and dolichocephaly.5,10 Since brachycephaly is a symmetrical deformity, CVAI is not an appropriate measure with which to identify this condition. Instead, Cranial Index (CI) is often utilized, which is a ratio of cranial width to length as indicated in the following equation:8

$$CI= \frac{cranial width}{cranial length} x 100$$

Cranial width is measured as the greatest biparietal diameter of the head, and cranial length is measured from the glabella anteriorly to the opisthocranion point posteriorly.5 Though there is discrepancy in the literature in terms of cranial index interpretation, Hutchinson et al conducted a widely cited case-control study which suggests that a CI of greater than or equal to 93% is indicative of brachycephaly.11 Brachycephaly and plagiocephaly can also occur simultaneously as a combined deformity in which CI and CVAI are both abnormal.8

**Dolichocephaly**

The term dolichocephaly comes from the Greek for “long head,” and this condition presents as flattening of both sides of the skull that results in a decreased cranial width and increased cranial length.12 Also known as scaphocephaly, this head shaping deformity occurs more frequently in premature infants than in full-term infants, and breech delivery has also been identified as a factor that commonly leads to this condition.1 Due to poor cervical strength, the head of a premature infant typically falls to the side when placed in a supine position, contributing to pressure against the side of the head.12 In addition, prone and side-lying positions are more frequently utilized with this patient population in order to mitigate certain medical issues such as bradycardia, apnea, and reflux.12 Furthermore, preterm infants have greater malleability of the skull compared to full-term infants, thereby increasing the susceptibility to CMD due to the influence of gravity.1,12 In terms of cranial measurements, a cranial index of less than 76% is indicative of dolichocephaly.12

**Etiology of CMD: Supine Sleeping Position**

In 1992, the American Academy of Pediatrics (AAP) introduced the recommendation that infants be placed on their back or side to sleep for the first 6 months of life in order to decrease the risk of sudden infant death syndrome (SIDS).13 This proposal arose from the identification of prone sleeping as a primary modifiable risk factor for SIDS.13 The United States Public Health Service further advocated for this recommendation in 1994 by coordinating the “Back to Sleep” campaign in order to disseminate this information to parents.14 Prior to the origination of this recommendation, approximately 74% of infants in the United States slept in the prone position, and this percentage had decreased to 20% by the year 2000.13,15 In 2011, the AAP expanded its safe infant sleep guidelines through the “Safe to Sleep” campaign.16 A few of the recommendations provided in this update include always sleeping in a supine position, using a firm sleep surface, and avoiding soft bedding materials.16 Overall, reducing the prevalence of prone sleeping has been successful in decreasing the occurrence of SIDS as well; however, this change in sleeping position also resulted in a significant increase in cases of cranial molding deformity due to the pressure of a firm, flat surface against the occiput for a prolonged period of time.15

**Etiology of CMD: Torticollis**

A second factor that has been noted as a potential contributor to CMD is torticollis.1 Torticollis is a condition characterized by unilateral tightness of the neck musculature, namely the sternocleidomastoid muscle, which results in a resting head position of ipsilateral side bending and contralateral rotation. This head positioning causes a particular area of the cranium to experience the greatest amount of pressure when the infant is lying down due to the difficulty or inability to provide distribution of pressure through alternate head positions.1 Although there is a known association between torticollis and CMD, it is unclear as to which condition typically presents first and leads to the other.5 Rogers et al suggest that cervical imbalance leads to head position preference which presents as torticollis, and flattening of the head develops subsequently.17 However, there may also be cases in which CMD leads to head position preference, eventually resulting in tightness of the neck musculature and the development of torticollis. Despite the uncertainty regarding which condition typically presents first, torticollis should always be identified and addressed as part of a comprehensive plan of care as it can limit the effectiveness of efforts to correct CMD.18

**Etiology of CMD: Additional Risk Factors**

There are numerous risk factors for CMD that have been noted in the literature, many of which are specific to certain patterns of deformity. Risk factors for plagiocephaly include male gender,5,19,20 first-born rank,5,19 supine position,5 prematurity,10 neck problems,5 intrauterine constraint,5 assisted delivery,5,19 multiple pregnancy,19 and gestational diabetes.19 Fewer risk factors have been associated specifically with brachycephaly, and these include supine position20 and intrauterine constraint.19 For dolichocephaly, noted risk factors include female gender,10 prematurity,10 extended hospitalization at birth,10 and breech position.1

**Prematurity**

Prematurity is a significant risk factor for CMD due to increased malleability of the skull at a younger age.10 In addition, preterm infants are more susceptible to the effects of gravity due to poor muscle tone, which contributes to positional deformity of the skull.1 Ifflaender et al conducted a recent study to determine the prevalence of cranial deformity in preterm infants.10 In this particular study, “very preterm” infants were born at less than 32 weeks gestational age, and “late preterm” infants were born between 32 and 36+6 weeks (36 weeks, 6 days) gestational age.10 The researchers found that 38% of very preterm infants demonstrated plagiocephaly at their term-equivalent age (TEA), while 73% of this group presented with dolichocephaly at TEA.10 In addition, 28% of late preterm infants exhibited dolichocephaly at TEA.10 Each of these values is significantly greater than the prevalence of these deformities in infants who are born full-term.10 The results of this study indicate the need for CMD prevention and treatment interventions that are designed specifically for premature infants.

**Treatment of CMD: Repositioning and Helmet Therapy**

 In the vast majority of CMD cases, complete correction of deformity can be achieved via conservative treatment consisting of repositioning therapy.2 The goal of repositioning therapy is to avoid prolonged periods of pressure on the flattened area of the head through the use of positioning devices, prone play, exercises to stretch and strengthen cervical musculature, and periodic repositioning of the infant’s head and body.2 For more severe deformities or if initial repositioning therapy is unsuccessful, orthotic therapy with a customized cranial molding helmet to be worn 23 hours per day is typically the subsequent recommendation.2,8 These orthotic devices are designed with openings over the flat spots of the head to eliminate pressure in these areas, while also providing external pressure to the remainder of the skull in order to direct growth towards the flattened areas. Practitioner oversight and parent education about the importance of compliance are essential components to both of these therapies in order to ensure that they are being carried out as prescribed.

**Treatment of CMD: Current Practice in the Neonatal Intensive Care Unit**

While the combination of repositioning and orthotic therapy has demonstrated effectiveness in correcting head shape for greater than 90% of infants with CMD, there remains the problem of how best to prevent and treat CMD in the neonatal intensive care unit since helmet therapy is not to be utilized until at least 4 months of age.2,8 This issue is particularly challenging since premature infants in the NICU are at increased risk of developing CMD than their full-term peers.10 In an attempt to remedy this situation, numerous positioning devices have been developed with the goal of preventing or correcting CMD in this patient population. The following is a list of the most widely utilized head positioning devices in NICUs around the United States based on a recent survey of NICU clinicians:21

* Blanket Rolls
* Frederick T. Frog (Philips)
* DandleROO, DandleROO2, DandleROO Lite, or DandleWrap (Dandle-LION)
* Sundance Fluidized Full-body Mattress or Tube, a.k.a. “Zflo” (Sundance Solutions)
* Sundance Fluidized Small or Medium Utility Pillow (Sundance Solutions)
* Gel-E Donut (Philips)
* Tortle Midliner and/or Tortle Air (Tortle Products)

At this point in time, however, only three of these positioning devices have been mentioned in the literature: the DandleROO, the Gel-E Donut, and the Sundance Fluidized Utility Pillow. The DandleROO is a containment device made of stretchable cotton that creates a womb-like environment by allowing the infant to extend the extremities while providing a gentle force to return the head and body to a position of midline flexion.22 In a survey of neonatal nurses’ and therapists’ perceptions of positioning for preterm infants, the DandleROO was identified as the “ideal method of neonatal positioning” by 62% of nurses and 86% of therapists.22 Furthermore, this survey also indicated that 44% of nurses and 57% of therapists found the DandleROO to be the easiest method of positioning to utilize in the NICU.22 This study, however, did not specifically address the DandleROO’s capacity to prevent or treat CMD.

The Gel-E Donut is a gel-filled pillow that is designed to provide pressure relief when placed beneath the occiput.23 In a study comparing interface pressures of various support surfaces in the pediatric population, a foam overlay alone and in conjunction with the Gel-E Donut was found to produce the lowest occipital pressures in infants less than two years of age.23 The Gel-E Donut alone was less effective at decreasing occipital pressure than when used in combination with the foam overlay.23 Other surfaces compared in this study that demonstrated higher occipital pressures were a standard crib mattress and a low-air-loss bed.23

 Overall, there is currently a great need for research that assesses the safety and effectiveness of each of these positioning devices for the prevention and treatment of cranial molding deformity, specifically for preterm infants in the neonatal intensive care unit.

**Treatment of CMD: Cranial Cup**

 One additional positioning device that is not yet widely utilized in clinical practice is the cranial cup. This device was originally created to treat plagiocephaly in full-term infants, but Michele DeGrazia, a neonatal nurse practitioner, worked with the designers of the cranial cup to transform it into an appropriate size for preterm infants. The cranial cup is an orthotic device made up of a plastic base overlaid with up to four layers of polyethylene foam that can be removed to adjust the fit of the device as the infant grows.24 It contains a concave portion for the head in order to encourage normal cranial development, and it supports the infant’s body in both supine and semi-side-lying positions.24 Prone positioning is contraindicated while utilizing this device, and it is also not designed for infants weighing less than 1000 grams.25 A recent randomized trial was conducted by DeGrazia et al to determine the effectiveness of utilizing the cranial cup along with a moldable positioner (Sundance Fluidized Utility Pillow) for the prevention of CMD in at-risk, hospitalized infants.25 While the experimental group rotated between the cranial cup and the moldable positioner with position changes every 3 to 4 hours, the control group used only the moldable positioner with position changes every 3 to 4 hours.25 The moldable positioner is defined as a pillow-shaped, fluidized device that supports the infant’s head, neck, and shoulders while in the supine, side-lying, or prone position.25 Specific details regarding positions changes were not provided in the article; however, nurses were instructed that infants in the experimental group should be positioned on the cranial cup for at least 12 hours per day.25 Results of this study showed that infants who utilized the cranial cup were less likely to develop CMD than those who utilized the moldable positioner alone.25

 Another recent study was conducted by Knorr et al to determine the effectiveness of the cranial cup for correcting head shape in hospitalized premature infants with visible CMD.24 This study was a prospective descriptive design in which all subjects utilized an adjustable cranial cup device for the management of their head shaping condition.24 Nurses were instructed that infants should spend a minimum of 12 hours per day on the cranial cup with position changes being performed every 3 to 4 hours.24 While not utilizing the cranial cup, infants were placed on another modifiable positioner, a gel pillow, a bed mattress, or were held by a caregiver.24 Again, specific details regarding position changes were not provided in the article. The results of this study suggest that the cranial cup is effective for normalizing head shape in hospitalized premature infants, although there is no way to know whether use of the device can account for the improvements in head shaping due to the lack of a control group for comparison.24

 Overall, more research of higher quality and methodological rigor are needed in order to ensure the safety and effectiveness of the cranial cup; however, this device demonstrates the potential to be effective for both the prevention and treatment of CMD in premature infants in the NICU.

**Conclusion**

Cranial molding deformity is a widespread condition that is typically reversible with conservative intervention. However, the evidence is lacking in regards to positioning devices that are commonly utilized with premature infants in neonatal intensive care units around the country. In future research on this topic, additional high quality randomized controlled trials and systematic reviews should be conducted in order to increase the level of evidence that exists regarding cranial molding interventions for the hospitalized, preterm infant. Studies with larger sample sizes are needed as well in order to decrease the variability of results. Another implication for further research is that increased specificity of intervention protocols is needed in order to improve the clinical relevance and utility of future studies. In general, current evidence does not allow for precise replication of treatment interventions in the clinical setting due to a lack of detailed procedures regarding how these interventions were implemented during the study. Lastly, long-term follow up of study participants would be another beneficial component of future research in order to determine the effects of cranial molding interventions over time.

**References**:

1. Bronfin DR. Misshapen Heads in Babies: Position or Pathology?. *The Ochsner Journal*. 2001;3(4):191-199.
2. Steinberg J, Rawlani R, Humphries L, et al. Effectiveness of Conservative Therapy and Helmet Therapy for Positional Cranial Deformation. *Plastic and Reconstructive Surgery*. 2015;135(3):833-842. doi:10.1097/prs.0000000000000955.
3. Persing J, James H, Swanson J, Kattwinkel J. Prevention and Management of Positional Skull Deformities in Infants. *PEDIATRICS*. 2003;112(1):199-202. doi:10.1542/peds.112.1.199.
4. Feijen M, Franssen B, Vincken N, van der Hulst R. Prevalence and Consequences of Positional Plagiocephaly and Brachycephaly. *Journal of Craniofacial Surgery*. 2015;26(8):e770-e773. doi:10.1097/scs.0000000000002222.
5. Looman W, Kack Flannery A. Evidence-Based Care of the Child With Deformational Plagiocephaly, Part I: Assessment and Diagnosis. *Journal of Pediatric Health Care*. 2012;26(4):242-250. doi:10.1016/j.pedhc.2011.10.003.
6. Clarren S. Plagiocephaly and torticollis: Etiology, natural history, and helmet treatment. *The Journal of Pediatrics*. 1981;98(1):92-95. doi:10.1016/s0022-3476(81)80549-5.
7. Guillaume D. Positional Plagiocephaly in Children Homepage. *International Society For Pediatric Neurosurgery*. 2017. Available at: http://ispn.guide/book/The%20ISPN%20Guide%20to%20Pediatric%20Neurosurgery/Congenital%20Disorders%20of%20the%20Nervous%20System/positional-plagiocephaly-homepage. Accessed March 1, 2017.
8. Wilbrand J, Schmidtberg K, Bierther U. Clinical Classification of Infant Nonsynostotic Cranial Deformity. *The Journal of Pediatrics*. 2012;161(6):1120-1125.e1. doi:10.1016/j.jpeds.2012.05.023.
9. *Plagiocephaly Severity Scale*. Atlanta, GA: Children's Healthcare of Atlanta; 2015. Available at: https://pediatricapta.org/special-interest-groups/HB/ORTH\_961942\_PlagiocephalyScale\_BWInfo.pdf. Accessed March 1, 2017.
10. Ifflaender S, Rüdiger M, Konstantelos D, et al. Prevalence of head deformities in preterm infants at term equivalent age. *Early Human Development*. 2013;89(12):1041-1047. doi:10.1016/j.earlhumdev.2013.08.011.
11. Hutchison B, Hutchison L, Thompson J, Mitchell E. Quantification of Plagiocephaly and Brachycephaly in Infants Using a Digital Photographic Technique. *The Cleft Palate-Craniofacial Journal*. 2005;42(5):539-547. doi:10.1597/04-059r.1.
12. McCarty D, Peat J, Malcolm W, et al. Dolichocephaly in Preterm Infants: Prevalence, Risk Factors, and Early Motor Outcomes. *American Journal of Perinatology*. 2016;34(04):372-378. doi:10.1055/s-0036-1592128.
13. Changing Concepts of Sudden Infant Death Syndrome: Implications for Infant Sleeping Environment and Sleep Position. *PEDIATRICS*. 2000;105(3):650-656. doi:10.1542/peds.105.3.650.
14. Turk A, McCarthy J, Thome C, Wisoff J. The “Back to Sleep Campaign” and Deformational Plagiocephaly. *Journal of Craniofacial Surgery*. 1996;7(1):12-18. doi:10.1097/00001665-199601000-00006.
15. Argenta L, David L, Wilson J, Bell W. An Increase in Infant Cranial Deformity with Supine Sleeping Position. *Journal of Craniofacial Surgery*. 1996;7(1):5-11.
16. SIDS and Other Sleep-Related Infant Deaths: Expansion of Recommendations for a Safe Infant Sleeping Environment. *PEDIATRICS*. 2011;128(5):1030-1039. doi:10.1542/peds.2011-2284.
17. Rogers G, Oh A, Mulliken J. The Role of Congenital Muscular Torticollis in the Development of Deformational Plagiocephaly. *Plastic and Reconstructive Surgery*. 2009;123(2):643-652. doi:10.1097/prs.0b013e318196b9be.
18. Van Vlimmeren L, Helders P, Van Adrichem L, Engelbert R. Torticollis and plagiocephaly in infancy: Therapeutic strategies. *Pediatric Rehabilitation*. 2006;9(1):40-46. doi:10.1080/13638490500037904.
19. Aarnivala H, Valkama A, Pirttiniemi P. Cranial shape, size and cervical motion in normal newborns. *Early Human Development*. 2014;90(8):425-430. doi:10.1016/j.earlhumdev.2014.05.007.
20. *Deformational Plagiocephaly & Cranial Remolding In Infants*. Alexandria, VA: APTA Section on Pediatrics; 2007. Available at: https://pediatricapta.org/includes/fact-sheets/pdfs/Plagiocephaly.pdf. Accessed March 5, 2017.
21. McCarty DM, Eberbach M, Peat J. Therapeutic Practices for Cranial Molding Deformity in the Neonatal Intensive Care Unit: A National Survey. *Manuscript in preparation.* 2017.
22. Zarem C, Crapnell T, Tiltges L, et al. Neonatal Nurses' and Therapists' Perceptions of Positioning for Preterm Infants in the Neonatal Intensive Care Unit. *Neonatal Network: The Journal of Neonatal Nursing*. 2013;32(2):110-116. doi:10.1891/0730-0832.32.2.110.
23. McLane K, Krouskop T, McCord S, Fraley J. Comparison of Interface Pressures in the Pediatric Population Among Various Support Surfaces. *Journal of Wound, Ostomy and Continence Nursing*. 2002;29(5):242-251. doi:10.1097/00152192-200209000-00007.
24. Knorr A, Gauvreau K, Porter C, Serino E, DeGrazia M. Use of the Cranial Cup to Correct Positional Head Shape Deformities in Hospitalized Premature Infants. *Journal of Obstetric, Gynecologic & Neonatal Nursing*. 2016;45(4):542-552. doi:10.1016/j.jogn.2016.03.141.
25. DeGrazia M, Giambanco D, Hamn G, Ditzel A, Tucker L, Gauvreau K. Prevention of Deformational Plagiocephaly in Hospitalized Infants Using a New Orthotic Device. *Journal of Obstetric, Gynecologic & Neonatal Nursing*. 2015;44(1):28-41. doi:10.1111/1552-6909.12523.