American Academy of Orthotists and Prosthetists
Third Consensus Conference

DRAFT

Orthotic Management of Deformational Plagiocephaly

Held at: Texas Scottish Rite Hospital
Dallas, Texas

April 7 – 9, 2004

Organizing Committee

Douglas G. Smith, MD *Primary Investigator* Associate Professor of Orthopedic Surgery University of Washington Medical Director, Amputee Coalition of America Seattle, WA	John Michael, CPO, FAAOP *Primary Investigator* CPO Services, Inc. Private Consultant Portage, IN
Don Katz, CO, FAAOP *Primary Investigator* Texas Scottish Rite Hospital for Children Program Director Dallas, TX	

Acknowledgements

This conference was made possible by a grant from the U.S. Department of Education and Project Quantum Leap (PQL) of the American Academy of Orthotists and Prosthetists (AAOP). Without such funding efforts, the advancement of medical science and patient care programs would not be possible. The dedication and participation of the conference participants is one step toward the development of clinical standards of practice that will enhance treatment procedures and outcomes for infants with plagiocephaly. It is their collective expertise combined with the available scientific research that makes this Consensus Report possible.

The whole of science is nothing more than the refinement of everyday thinking.
– Albert Einstein: Physics and Reality

The Organizing Committee and participants of the Consensus Conference would also like to take this opportunity to recognize the staff at the American Academy of Orthotists and Prosthetists national office for their support to this project. In addition, we recognize the contribution of Texas Scottish Rite Hospital for Children in Dallas, Texas for their assistance in the organization and hosting of this event.

Table of Contents

Key Reviewers and Expert Discussants

Bill Barringer, CO University of Oklahoma 1122 NE 13th Street, Medical Center Oklahoma City, OK 73117-1026	Wendy Biggs, MD Clinical Assistant Professor Michigan State University College of Human Medicine Assistant Director Midland Family Practice Residency Midland, Michigan
Deanna Fish, MS, CPO Orthomerica Products, Inc. 1508 West Elm Hill Circle Taylorsville, UT 84123	Alex Kane, MD Washington University School of Medicine St. Louis Children's Hospital One Children's Place, Suite 11 W 7 St. Louis, MO 63110
Charles Kuffel, CPO Pongratz Orthotics & Prosthetics, Inc. 2530 E. Thomas Road Phoenix, AZ 85016	Jeff Larsen, CO Gillette Children's Specialty Healthcare 200 University Ave. E. St. Paul, MN 55101-2598
Dulcey Lima, CO, OTR/L Orthomerica Products, Inc. 21 W. 272 Coronet Road Lombard, IL 60148	Tim Littlefield, MS Cranial Technologies, Inc. 1395 West Auto Drive Tempe, AZ 85284
Colleen Coulter-O'Berry, PT Children's Healthcare of Atlanta 5445 Meridian Marks Road NE, Suite 200 Atlanta, GA 30342-4755	Ammanath Peethambaran, CO University of Michigan Orthotics & Prosthetics 2850 S. Industrial Hwy, Suite 400 Ann Arbor, MI 4810406768
Laura Plank, CO Children's Healthcare of Atlanta 5445 Meridian Marks Road NE, Suite 200 Atlanta, GA 30342-4755	Aaron Sorensen, CPO Restorative Health Services, Inc. 311 18th Ave. N Nashville, TN 37203
Joe Terpenning, CO Eastern Cranial Affiliates 1600 Wilson Blvd., Suite 200 Arlington, VA 22209	

List of Participants by Profession

Orthotist	Therapist
Bill Barringer, CO Deanna Fish, MS, CPO Charles Kuffel, CPO Jeff Larsen, CO Dulcey Lima, CO, OTR/L Ammanath Peethambaran, CO Laura Plank, CO Aaron Sorensen, CPO Joe Terpenning, CO	Colleen Coulter-O'Berry, PT
Physician	**Engineer**
Wendy Biggs, MD Alex Kane, MD	Tim Littlefield, MS

Introduction Background Information

> *"Without a scientific basis for the assessment and measurement process, we face the future as independent practitioners unable to communicate with one another, unable to document treatment efficacy, and unable to claim scientific credibility for our profession."*
>
> *J.M. Rothstein*

Clinical standards of practice are systematically developed guidelines to assist clinicians in making decisions about patient care practices and procedures. Specific patient populations, diagnoses and treatments are identified within the guidelines. Clinical practice guidelines reflect the available research on a particular topic, evaluate the strength of the scientific evidence, and also promote improved clinical decision-making. Guidelines must also be dynamic in nature to reflect the constantly changing strides made in medicine. They must also be multidisciplinary to address the many members of the health care medical treatment team. Finally, these guidelines also improve the quality and efficiency of health care with safe reductions in costs.

The purpose of the "Clinical Standards of Practice on the Orthotic Management of Deformational Plagiocephaly" (CSOP #3) was to establish and define parameters that would guide clinical decision-making and treatment protocols of clinicians involved in the care of infants with deformational plagiocephaly. Practice guidelines help to reduce the variability in clinical procedures and improve the quality of care. Demonstrating measurable effects and improvements in patient care promote practice guidelines as efficacious and efficient clinical tools. Ultimately, the value of such guidelines is evidenced by the impact made on patient care practices – specifically, quality of care and improved patient satisfaction.

Deformational Plagiocephaly

Symmetry is present in nature, art, mathematics, architecture, engineering and in the human body. It corresponds to an arrangement and balancing of the parts or elements of a whole in respect to size, shape, and position on opposite sides of an axis or center[1]. In the case of the human skeleton, many paired bones and extremities exist and those that are not paired, like the sternum, are positioned in the middle of the body. For example, symmetry is manifested in the face where there are two eyes and ears, but only one nose and mouth that are precisely centered.

The skull has paired frontal, parietal, temporal, and sphenoid bones, with one occipital bone positioned midline. This midline and paired orientation aligns the head in relation to the rest of the body, placing it in the optimal position to send and receive olfactory, visual, and auditory information. A disruption in this midline position through trauma or deformation can affect the processing of sensory stimulation and lead to visual disturbances[2-4]. It has also been shown that the mandibles are asymmetrical in infants with skull and facial asymmetry[5], resulting in feeding difficulties and the potential for temporal mandibular joint (TMJ) dysfunction over time.

For thousands of years, people around the world have used various means to change the shape of infants' heads. Meticulously wrapped string, bark, woven skullcaps, and backboards with wooden forehead restraints have been used by many different cultures attempting to create a symmetrical shape and a specific proportion. In 1979, Sterling Clarren, M.D., published a paper in the Journal of Pediatrics entitled "Helmet Treatment for Plagiocephaly and Congenital Muscular Torticollis"[6]. Cranial molding orthoses were introduced as ways to achieve cranial symmetry and proportion, and correction of the congenital muscular torticollis was also noted. Once these initial results were published, many medical centers across the United States began using cranial molding orthoses to treat infants with skull and facial asymmetry rather than perform invasive and costly surgeries to correct these deformations.

Cranial asymmetry is referred to as plagiocephaly (from the Greek word "oblique"), and the incidence has been steadily increasing over the last ten years. This increase coincides with two factors that occurred in the early 1990's. The first contributing factor relates to infant sleep position. Babies have been placed in a supine position (i.e. on their backs) rather than prone (i.e. on their tummies) since 1992 when the American Academy of Pediatricians (AAP) and other physicians around the world initiated the "Back to Sleep" program. This recommendation was directly related to the fact that infants sleeping in a prone position had a higher incidence of Sudden Infant Death Syndrome (SIDS)[7]. This very successful campaign has resulted in a 40% drop in the incidence of SIDS[8]. Concurrently, a dramatic increase in deformational plagiocephaly also developed.

A second factor relating to deformational plagiocephaly is the extensive use of infant seats and carriers. In the early 90's, these products were redesigned for convenient use in a variety of places with greater ergonomic efficiency for the parents. The multifunction seats that can be used in the car, stroller, high chair, etc. have become extremely popular in today's mobile society. As a result, infants are often placed in them for extended periods of time throughout the day, putting continuous pressure on the back of the baby's head. The soft bony plates of the infant's skull are easily shifted and misaligned with continuous pressure over time, and deformational plagiocephaly can occur in a very short period of time. Combined with supine sleep positions, cranial flattening has become a common occurrence in young infants. Despite the best efforts of parents to provide a variety of positions while the infant is awake and supervised, many infants will continue to develop progressive craniofacial asymmetry.

There are many additional causes of deformational plagiocephaly including: intra-uterine constraint, prematurity, trauma during the birth process, breech presentation, early descent into the maternal pelvis, congenital cervical disc problems, and congenital muscular torticollis. In previous years, head shape deformities often resolved after birth, most likely because the infant was in many different positions throughout the course of the day and night. With the Back to Sleep program and the popularity of infant carriers, however, head deformities are less likely to correct without specific intervention(s) because the deforming forces are continuous. In addition, babies that sleep supine do not acquire motor milestones as quickly as prone sleeping babies, and therefore it takes infants longer to acquire the skills to roll, sit, creep, crawl and pull to stand[9,10]. In the first six months of life, the infant skull is very moldable and even a few weeks delay in rolling can have a significant effect on skull shape.

Deformational plagiocephaly is also associated with torticollis and is estimated to occur in about 1 of every 300 live births[6]. Torticollis is identified by an imbalance of the cervical muscles,

which may include the sternocleidomastoid, upper trapezius, scalenes and levator scapula. When the two conditions occur together, the deformational forces can be quite severe and result in rotational and translational changes in the skull and facial bones. When recognized in the first few months of life, parents are taught massage, repositioning, and handling techniques that include both passive and active alignments. The infant is monitored closely for symmetry, neck strength, trunk control and range of motion in all directions. Therapy is indicated for moderate to severe torticollis that does not resolve with parental repositioning, and if often prescribed before, during and after orthotic management for deformational plagiocephaly. If the torticollis is severe and does not resolve with therapy, surgery may be considered to lengthen the contracted muscle.

Repositioning programs are most effective during the first three to four months of life. If the infant's head has not become sufficiently symmetrical or proportional during that time, further evaluation and/or diagnostic tests are undertaken to determine if the deformity is due to craniosynostosis, some other anomaly such as a hemivertebrae, or deformational forces. It is often the parent that brings the infant's head deformation to the attention of the pediatrician. Diagnostic tests may include x-ray, magnetic resonance imaging (MRI), or three-dimensional computed tomography (CT scan), and a referral to a craniofacial clinic may be necessary. If there is no evidence of craniosynostosis, the infant is referred to an orthotist for further evaluation and design of a cranial molding orthosis.

Contraindications to cranial molding orthoses include young infants (under three months of age) who can benefit from intensive repositioning and handling programs. Infants with unmanaged hydrocephalus must be stabilized prior to beginning treatment with a cranial orthosis, and infants with craniosynostosis are not candidates for orthotic intervention until the fused suture(s) has been surgically removed. Infants older than 18 months do not benefit as much from cranial molding orthoses because peak growth periods have already transpired. However, if the deformity is moderate to severe, and there is remolding potential remaining, young children into their second year may still experience some correction[11].

Cranial molding orthoses are now used as the primary treatment method for infants older than 3 months of age with moderate to severe deformational plagiocephaly and other skull deformities that have not benefited from repositioning and handling programs[12-15]. The cranial molding orthosis provides total contact over the bossed or prominent areas of the infant's head, and allows space over the flattened areas to provide a pathway for growth to occur. The rapid growth of the infant's brain in the first 12 months of life expands and pushes against the plates of the skull in the areas of least resistance. Consequently, the synergy between the inherent symmetrical brain growth and the skull's natural flexibility and plasticity in these early months creates dynamic correction and improves cranial shape. In some cases, improvement in the alignment of facial structures is also noted.

Head shapes are classified according to a specific pattern of deformity. The term deformational plagiocephaly refers to a head shape that has unilateral occipital flattening, anterior progression of the ipsilateral ear, and varying degrees of ipsilateral forehead and contralateral occipital bossing. In effect, the continuous pressure on one side of the cranium causes all bones on the same side to progress forward creating an asymmetrical skull shape.

The term deformational brachycephaly refers to a disproportional head shape that is short and wide. The occiput is flattened to a similar degree on both sides, and the dimension of the width and length is abnormal. Infants with this head shape often have a prominent or bossed forehead and increased cranial vault[16]. Normal anthropometric data for six-month old infants indicates that the cephalic index (ratio of the cranial width to the cranial length) is about 78%. Infants with deformational brachycephaly often exhibit ratios greater than 85% for girls and 90% for boys (approximately two standard deviations above the norms).

Another head shape seen in infants with cranial deformities is called deformational scaphocephaly. (Scaphocephaly and dolichocephaly are used interchangeably throughout the literature and in various medical dictionaries. There was no consensus as to the proper terminology to describe the non-synostotic head shape and therefore the term scaphocephaly will be used throughout this document.) These infants have a very long and narrow head that often develops as a result of extended time spent in a side-lying position. Side lying is the position of choice in neonatal intensive care units due to the need to monitor the infants. The cephalic index in scaphocephalic infants is very low, and the ratio of the cranial width-to-length measurement is significantly less than 78%. Premature infants are particularly vulnerable to the development of this deformity because the cranial structures are even softer than the flexible heads of full term infants. Premature infants tend to have delayed head control related to their prematurity, and the long narrow head shape places the neck muscles at an additional mechanical disadvantage. Breech positioning will also produce this head shape even with cesarean deliver.

The design and application of a cranial molding orthosis does not alter the magnitude of intrinsic brain growth but merely its direction[13,17]. Symmetrical growth is directed by consistent evaluation and adjustment of the orthosis based upon the infant's head shape and growth pattern. Many different orthotic designs have been developed over the last 24 years to effectively address this patient population. The basic principle of all cranial molding orthoses is to create a pathway for symmetrical and/or proportional growth to occur.

The outcomes of orthotic treatment are dependent upon a definitive diagnosis, available brain/cranial growth, the age of the infant at the beginning of treatment, and compliance with the treatment protocols. Cranial molding orthoses for infants with deformational plagiocephaly, brachycephaly and scaphocephaly use the rapid period of infant brain growth, particularly in the first year, to create greater facial, cranial, and core symmetry. Therapy and orthotic treatment can improve the infant's cranial alignment and shape, visual tracking, overall postural alignment, and provide a more symmetrical head position of acquiring developmental skills. Early intervention, team networking, and consistent care are the key components of an effective treatment program with measurable outcomes.

Previous AAOP Consensus Conferences

The American Academy of Orthotists and Prosthetists recognizes the need for the continued advancement of the field of orthotics in regards to defining, measuring, and improving the quality of care provided by orthotic clinicians. Consensus conferences serve as a resource for evidence-based material on specific patient populations and clinical treatment programs. They also serve to identify limitations in clinical knowledge and areas in need of further investigation and research. Two previous AAOP consensus conferences have been held:

(February 2002) Orthotic Treatment of Idiopathic Scoliosis and Scheuermann's Kyphosis

(May 2003) Major Lower Limb Amputation Post-Operative Strategies

A fourth consensus conference on care of the diabetic foot is scheduled for August of 2004.

Conference Summary

The scope of the Consensus Conference was to develop guidelines covering the comprehensive care of infants with deformational plagiocephaly and post-operative craniosynostosis. The identification, diagnosis and medical management of these infants require a multidisciplinary approach. It was determined that the following topics would provide the foundation for critical assessment of the literature and expert discussion:

- General identification and diagnosis
- Orthotic management
- Therapeutic management
- Surgical management

The conference objectives were multifold.

1. Identify the patient population and formulate the questions.
2. Develop and execute plan to gather a multidisciplinary panel of experts.
3. Critically review and rank the available scientific literature.
4. Analyze and interpret the data; draw relevance to current clinical practices.
5. Identify gaps in the scientific literature where further research is necessary.
6. Generate manuscripts on specific topics for future publication.
7. Discuss critical questions that may or may not have been answered in the available literature.
8. Achieve consensus on clinical practice guidelines that would reduce variability of care and increase quality of care.
9. Document and report findings of conference to AAOP and the U.S. Department of Education
10. Make findings accessible to the medical field via online publications, journal articles, lectures and other educational materials.

In advance of the conference, the organizing committee determined a budget. A conference chair, vice chair and panel of experts were then selected. Participants represented the following medical fields: orthotics, therapy, obstetrics/gynecology, plastic surgery and engineering. Each participant was assigned a topic of study and required to write a research paper, present that paper at the conference, develop critical questions and also participate in the group discussions.

In general, a series of two to three papers were presented and each presentation was followed by a brief question and answer session. At specific intervals, lecture sessions were halted and group discussions of the critical questions were undertaken. Great effort was made to reach consensus and create recommendations for clinical guidelines. Final revisions to the manuscripts were undertaken following the conference, and these manuscripts will be available electronically in their entirety through the AAOP website. Further, edited versions of the manuscripts will be published in a future supplement of the Journal of Prosthetics and Orthotics.

The Chair and Vice Chair are listed below:

Deanna Fish, MS, CPO *Chair* Orthomerica Products, Inc. Clinical Education Manager Salt Lake City, UT	Dulcey Lima, CO, OTR/L *Vice Chair* Orthomerica Products, Inc. Clinical Education Manager Lombard, IL

Conference Agenda

Tuesday, April 6, 2004

7:00pm	Welcome dinner and introductions	

Wednesday, April 7, 2004

7:30am	Shuttle to Texas Scottish Rite Hospital	
8:00 – 8:05am	Welcome	D. Katz
8:05 – 8:10am	Introduction	D. Fish
8:10 – 8:15am	AAOP Education Grant Synopsis	J. Michael
8:15 – 8:40am	(1) The "Epidemic" of Deformational Plagiocephaly and the American Academy of Pediatrics' Response	W. Biggs
8:40 – 8:45am	Questions / comments	
8:45 – 9:15am	(2) Overview of Treatment Results for Children With Deformational Plagiocephaly and Other Head Shape Deformities	D. Lima
9:15 – 9:20am	Questions / comments	
9:20 – 9:50am	(3) Clinical Evaluation Processes and Procedures for the Orthotic Treatment of Infants with Plagiocephaly	D. Fish
9:50 – 9:55am	Questions / comments	
9:55 – 10:10am	Break	
10:10 – 11:10am	Syndicate Session with Discussion of Critical Questions and Consensus Agreement for Topics 1 – 3	All
11:10 – 11:40am	(4) The Classification of Mild, Moderate and Severe in Deformational Plagiocephaly	L. Plank
11:40 – 11:45am	Questions / comments	
11:45 – 12:00noon	Open discussion	All
12:00 – 1:00pm	Lunch	
1:00 – 1:30pm	(5) Obtaining a Positive Model for Craniofacial Deformities: An Empirical Review of Casting Procedures	A. Sorenson
1:30 – 1:35pm	Questions / comments	
1:35 – 2:35pm	Syndicate Session with Discussion of Critical Questions and Consensus Agreement for Topics 4 – 5	All
2:35 – 3:05pm	(6) Modification Procedures of the Positive Model	A. Peethenbaram
3:05 – 3:10pm	Questions / comments	
3:10 – 3:20pm	Break	
3:20 – 3:50pm	(7) Orthotic Cranioplasty: Material and Design Considerations	J. Terpenning
3:50 – 3:55pm	Questions / comments	
3:55 – 4:45pm	Syndicate Session with Discussion of Critical Questions and Consensus Agreement for Topics 6 – 7	All
4:45pm	Shuttle to Melrose Hotel	
6:00pm	Reception and dinner at Melrose Hotel	

Thursday, April 7, 2004

7:30am	Shuttle to Texas Scottish Rite Hospital	
8:00 – 8:30am	(8) Orthotic Modeling of the Developing Skull	C. Kuffel
8:30 – 8:35am	Questions / comments	
8:35 – 9:05am	(9) Orthotic Treatment Protocols for Plagiocephaly	J. Larsen
9:05 – 9:10am	Questions / comments	
9:10 – 10:10am	Syndicate Session with Discussion of Critical Questions and Consensus Agreement for Topics 8 – 9	All
10:10 – 10:20am	Break	
10:20 – 10:50am	(10) An Overview of Craniosynostosis	A. Kane
10:50 – 10:55am	Questions / comments	
10:55 – 11:25am	(11) The Use of Post-Operative Cranial Orthoses in the Management of Craniosynostosis	Wm. Barringer
11:25 – 11:30am	Questions / comments	
11:30 – 12:30pm	Syndicate Session with Discussion of Critical Questions and Consensus Agreement for Topics 10 – 11	All
12:30 – 1:30pm	Lunch	
1:30 – 2:00pm	(12) Identification and Treatment of Congenital Muscular Torticollis in Infants	C. Coulter-O'Berry
2:00 – 2:05pm	Questions / comments	
2:05 – 2:20pm	(13) Team Members and Roles	D. Fish
2:20 – 2:25pm	Questions / comments	
2:25 – 2:55pm	(14) Deformational Plagiocephaly: Recommendations for Future Research	T. Littlefield
2:55 – 3:00pm	Questions / comments	
3:00 – 3:10pm	Break	
3:10 – 4:10pm	Syndicate Session with Discussion of Critical Questions and Consensus Agreement for Topics 12 – 14	All
4:10 – 4:25pm	FDA Regulation of Cranial Molding Devices	T. Littlefield
4:25 – 4:45pm	Closing Comments	J. Michael, D. Katz, D. Fish, D. Lima
4:45pm	Shuttle to Melrose Hotel	
6:00pm	Dinner at Nuevo Leon	

Literature Review

For the purpose of this review, the term deformational plagiocephaly will be used to describe head shape deformities whose etiology is mechanical in nature and is not caused by premature synostosis. This review will discuss the literature as it relates to specific treatment options and protocols for infants with deformational plagiocephaly. Articles presenting data on potential long-term effects of untreated deformational plagiocephaly will be reviewed and critiqued for the strength of the scientific rigor and studies using statistical analysis will be identified. A table containing a synopsis of the articles related to orthotic, surgical, and repositioning management regimes is attached in Appendix 1.

Terminology

It is apparent that no consistent terminology exists to define and describe the various head shape deformations of young infants. Clarren used the term "deformational plagiocephaly"[18]; Bruneteau and Mulliken proposed the term "deformational frontal plagiocephaly"[14]; and Hellbusch et al used the term "deformational occipital plagiocephaly" while describing a repositioning program[19]. In Pollack et al's article in 1997, the term "posterior plagiocephaly" was used generally to describe posterior flattening that was either synostotic or non-synostotic[13]. Later in the same article the terms deformational molding and deformational plagiocephaly were also used to describe infants whose head shape deformities were non-synostotic. Panchal et al use "plagiocephaly without synostosis" and “occipital nonsynostotic plagiocephaly” in an article published in 2001[20]. Newspaper publications frequently use the term "flat head syndrome" which is both derogatory and a misnomer since this head shape deformity is not a syndrome. Clearly, there is a need for consistent terminology to describe this deformity that results from the application of external mechanical forces.

Incidence

The lack of consistent terminology continues to thwart efforts to establish the incidence of deformational plagiocephaly. In addition, there has been no research that establishes the point at which normal asymmetry leaves off and deformational plagiocephaly begins. A study by Boere-Boonekamp et al sought to establish the incidence of positional head preference in a 2001 study involving 7,609 infants under the age of 6 months[21]. Neck asymmetry range of motion and the shape of the infant's head were noted and compared to matched samples. Children were followed at six months, one year, and two years after the initial contact. The prevalence of positional preference in infants below the age of 6 months was 8.2%. In infants younger than eight weeks of age, the prevalence was 10%, decreasing to 3% by 26 weeks. The prevalence of flattening of the occiput was calculated at 9.9% of the total number of infants and was much higher than the authors expected. At the two- to three-year follow up, skull asymmetry was still present in 45% of the infants who exhibited asymmetry in the first six months of life.

Peitsch et al published their findings of the incidence of cranial asymmetry in healthy newborns in 2002[22]. In a sample of 201 healthy newborns, researchers found a 13% incidence of localized cranial flattening and other "anomalous" head shapes in 11% of the infants that were singletons. In twins, there was a much higher incidence of localized flattening of 56%. Neonates with areas of occipital flattening had a mean of 4.74 mm of difference in their transcranial measurements. Infants classified as normal had a mean transcranial difference of

.88 mm. No follow-up measurements were included in this study as the intent was to document the incidence of deformation present at birth.

Etiology

There are several risk factors identified with the development of deformational plagiocephaly with high agreement among researchers. These include multiple births[14,21-26], unusual birth position[14,21-25], first-born[14,21-23,26], and male gender[15,21-30]. Additional factors found to be significant include neck tightness including torticollis[3,12,13,18,19,21,25,26,29-34], premature birth[14,23,25,26,35], supine positioning[15,21,23,26,27,36,37], developmental delay[13,18,20,26,38], and prolonged labor[21-23] among other less common causes.

Long-Term Consequences of Untreated Deformational Plagiocephaly

Studies discussing the long-term consequences of deformational plagiocephaly are limited. The increase in this deformity has largely been in the last decade since placing infants on their backs for sleeping and the widespread use of carriers as daytime positioning devices have become standard parenting practices. Long-term studies with large sample sizes are just now becoming possible as this first group of infants introduced to these childcare practices reaches middle school and younger children reach school age. It is only now that suspected problems like higher than normal numbers of children requiring orthodontic procedures, treatment for scoliosis, visual disturbances, auditory problems can be identified and studied because there are large numbers of children to assess. School services should be another area of investigation as researchers determine if children with deformational plagiocephaly that did not received treatment have any higher incidence of learning problems or social issues than a norm-matched group of children.

Developmental Issues

Several authors identified developmental delay as a factor contributing to deformational plagiocephaly but it is not known whether the developmental delay is the cause or the effect of the head deformation[13,18,20,26,38]. Miller et al published a paper in 2000 researching whether there was an increased rate of developmental delay in school-aged children diagnosed with deformational plagiocephaly as infants[38]. He compared the number of school services needed by these children compared to their siblings and to the statistics from the state of Washington listing the percentage of children in each grade requiring special services in 1997. A significant difference was found between the need for special education services in the group with deformational plagiocephaly compared to their siblings. Services needed by the children with deformational plagiocephaly included speech therapy, occupational therapy and physical therapy. The researchers did not determine whether deformational plagiocephaly was an early sign of subtle brain dysfunction or whether early brain molding can lead to subtle brain dysfunction.

Panchal et al studied neurodevelopment in 21 infants diagnosed with "plagiocephaly without synostosis"[20] through the use of a mental developmental index (MDA), psychomotor developmental index (PDI) and compared it with a standardized Bayley Scale of Infant Development-II (BSID II). The infants were untreated for their plagiocephaly, and were found to be significantly different from the standardized distribution expected. 82.6% of the infants scored within normal limits, but the percent of infants scoring with significant delays (8.7%) was significantly higher than expected (normal =2.7%). No infants were identified in the accelerated range of functioning. Authors suggest that further study is needed to determine if

these results persist over time, and to determine if children will score differently if they are treated for their deformational plagiocephaly with a cranial remolding orthosis.

A presentation at the American Cleft Palate and Craniofacial Association Meeting in 2004 by Patel et al reported additional problems with development related to visual field problems[39]. Further research needs to identify whether these areas of developmental dysfunction are significant enough to effect the normal development of a infant, or whether the dysfunction represents a benign problem.

Studies with Random Control Groups or Cohort Studies

There is no study published with a random control group. In the United States, the use of cranial molding orthoses and repositioning are prescribed frequently for deformational plagiocephaly and it is questionable whether it is ethical to withhold treatment of either modality. Both types of treatment are mentioned in the majority of studies published about deformational plagiocephaly[12-15,18-20,23,25-28,31,35,40], which may indicate that it has become the standard of care for this deformity. It is possible that a randomized study with a control group may be done as a multi-center study comparing current practice in a facility using cranial remolding orthoses and/or repositioning in the United States to management of infants with deformational plagiocephaly in another country that does not treat the deformity.

There are several cohort studies and these have been documented in the appendix[12,13,25,41,42,44]. These included four different studies by Pollack, Mulliken, Loveday and Vles who compared the results of two groups of patients: one group treated with positional therapy and the other group treated with a cranial molding orthosis[12,13,31,42]. Another cohort study by Teichgraeber compared the orthotic treatment results of infants with deformational plagiocephaly to those with deformational brachycephaly[44].

Surgical Management of Deformational Plagiocephaly

Few studies advocate the use of surgery to treat deformational plagiocephaly unless the head shape deformity was severe and the infants failed to respond to more conservative treatment such as repositioning or orthotic management[12-15,24,31,36,37,40,41,44-46]. In a study of 204 patients over a 16-year period of time in Australia, David et al reported that 9.3% patients had surgery to correct their severe plagiocephaly after trying repositioning as the only treatment modality[47].

Studies Using Subjective Measures

Researchers of deformational plagiocephaly sometimes use parent satisfaction or other subjective measures to assess improvement after treatment. Vles et al used a rating scale pre and post treatment[42]. Pollack's group questioned parents about whether they considered their infant's craniofacial features: normal or nearly normal, abnormal but acceptable, or unacceptable and in need of further intervention. Members of the craniofacial team used the same scale and at the end of treatment the scaled assessment was repeated to assess change. David et al simply asked parents if they considered their infant to have an acceptable improvement in their head shape[47].

Referral Criteria

There is widespread agreement that it is important to identify head shape deformities early. This is best described in the article by the American Academy of Pediatrics Clinical Report entitled "Prevention and Management of Positional Skull Deformities in Infants"[46]. This article

advises the pediatrician and other allied health providers involved in newborn care practices to educate parents on ways to decrease the risk of deformational plagiocephaly. Tummy time while the infant is supervised and awake is encouraged to prevent flattening and facilitate development of the shoulders and trunk. They discussed alternate positioning including sleep positioning with parents, in addition to changing the infant's orientation to the surroundings. Evaluation of cranial symmetry is recommended at each visit for health care services. Evaluation should include and document examination from the top of the head including the head shape, ear position, and cheekbone symmetry from this angle. The health care provider should also examine the face and the neck for signs of asymmetry, and have an understanding of how to differentiate deformational plagiocephaly from craniosynostosis.

This report goes on to suggest that x-rays are other diagnostic tests are not necessary in most situations. If craniosynostosis is suspected, the infant should be referred to a specialist for further evaluation. If therapeutic positioning and exercise as advised by the health care provider does not improve the skull shape after 2 to 3 months, the AAP report suggests that the physician refer the infant to a pediatric craniofacial specialist. The specialist can provide further evaluation and make recommendations that may include skull molding helmets or surgery. The specialist may also recommend physical therapy if the neck range of motion does not improve with exercises and repositioning within 2-3 months.

The AAP report notes that the best response for helmets occurs in the age range of 4 to 12 months due to malleability of the infant's skull and rapid growth during this period. These recommendations are substantiated by studies involving repositioning and/or the use of cranial molding orthoses as treatment modalities for this population. Researchers overwhelmingly recommend that orthotic treatment be initiated by 6 months of age to receive optimal results[3,13,18,19,25,27,28,31,35,37,40,41].

Conclusions

The increased research about deformational plagiocephaly in the last decade has been in direct response to the increased incidence of this condition seen across the world. The etiology is well documented and includes both pre- and post-natal factors. Researchers agree that excessive time spent in the supine position has escalated the incidence of deformational plagiocephaly. They also agree that early identification and parent education about tummy time and alternate positions while the infant is awake and supervised can decrease deformity if this treatment is initiated early. In cases where the deformity is still significant after a repositioning trial, and prior to six months of age, the infant will often benefit from a cranial molding orthosis.

Further information about these articles pertaining can be found in Appendix 1.

Rekate has published a critical review of the literature and excerpts are reprinted below:

> Occipital Plagiocephaly: A Critical Review of the Literature. Harold L. Rekate, M.D. Journal of Neurosurgery, 89:24-30, 1998.
> *The objective of this review was to determine what information is available on the incidence, pathophysiology, late complications, and treatment paradigms for occipital plagiocephaly (OP) based on a critical review of the literature obtained from recognized databases in peer-reviewed scientific publications.*

The content of this article is based on a critical review of the literature, and when discussing treatment options, classification of those articles with respect to the strength of the recommendations they contain.

Using standard computerized search techniques, databases containing medical literature were queried for key words related to occipital plagiocephaly beginning in 1966. Key words used for this search were: lambdoid, craniosynostosis, cranial sutures, facial asymmetry, torticollis, and plagiocephaly. Titles of all articles were scanned for relevance. Copies of all potentially relevant articles published in the English language were obtained and received at least a cursory review. Several articles not captured by these methods were found to be important when referenced in the articles obtained. Articles discussing treatment were divided into Class I, Class II, and Class III data for the purpose of deciding on their applicability to the development of a potential consensus for the treatment of this controversial condition.

Using the aforementioned key words, there were 4308 articles identified with potential relevance: scanning by title excluded all but 89. Of the 89, those with on-line abstracts were scanned; the remainder were obtained via interlibrary loan when needed for scanning of the article itself. The actual incidence of occipital plagiocephaly is unknown and there are no population-based studies of its incidence or prevalence. The reported incidence of lambdoid craniosynostosis ranges from 3 to 20% with the differences primarily due to differences in diagnostic criteria. With the possible exception of a lambdoid suture, which is replaced throughout its entire course by a dense ridge of bone, there are no other diagnostic criteria upon which there is agreement. There are no Class I and only one Class II studies in which a group of patients treated with one form of therapy is compared with another form of therapy or an untreated group. Treatment options that are recommended include observation only, mechanical treatments such as exercises, positioning, remodeling helmets, and a wide variety of surgical techniques. Very few reports accessed through the aforestated methodology report patients suffering any significant late effects of occipital plagiocephaly, although it may be morphometrically evident in as many as 14% of adults.

Controlled clinical trials will be needed before any form of intervention can be generally recommended. If surgery, which is expensive and potentially dangerous, is to continue to play a role in the management of this condition, efforts should be made to ascertain from the general population, which patients who have not been treated have suffered from this lack of treatment.

Rekate further states:

Summary of Treatment Options:

A. Standards: There are insufficient data to support a treatment standard for OP.

B. Guidelines:

1. Mechanical means such as propping of the child and neck stretching exercises are useful in preventing worsening of OP and may help reverse some of the deformity;

2. *In patients with moderate-to-severe degrees of OP, helmet or band therapy will result in improvement in the observed and measurable asymmetry. The earlier it is applied the more quickly and more completely the correction can be accomplished. The degree of deformity necessary to require the use of this form of treatment cannot be determined from currently available studies;*
3. *There are insufficient data to support guidelines for the surgical management of OP.*

C. *Options:*
   1. *Neck stretching exercises and lateral propping of the baby's head may be sufficient to correct OP if they are begun early in life and performed consistently;*
   2. *Head band or helmet therapy should be utilized in babies in whom the asymmetry persists despite other mechanical forms of intervention;*
   3. *Head band or helmet therapy should be utilized prior to considering surgical intervention in patients in whom the condition is recognized within the first year of life;*
   4. *For patients with severe residual deformity following head band or helmet therapy or for those in whom referral occurs beyond one year surgical intervention may be considered;*
   5. *If surgery is to be performed for OP there are several techniques, which have been reported to be effective. There are no studies showing compelling reasons to choose one over another.*

Rekate concludes:

> *Forms of therapy available to treat this condition include mechanical reversal of the deforming forces, mechanical redirection of those forces using helmet therapy, and surgical intervention. Because little information exists about the true risk of leaving this condition untreated, careful consideration is necessary in determining the role of each of these modalities in the management of the patient with OP.*

General Recommendations

General recommendations (R) are good "practice points". They are supported by clinical practice, but may not be supported in the literature. Many of these points should be used as a basis for further research.

Recommendation: Cranial molding orthoses should be considered in the management of deformational plagiocephaly.

Discussion: Cranial molding orthoses have been used medically since 1979 to correct head shape deformities. This is well documented in the literature, which consistently supports the use of cranial molding orthoses for infants with moderate to severe cranial deformations.

Recommendation: Repositioning techniques and therapy are viable treatments for infants with deformational plagiocephaly.

Discussion: Every article published about infants with deformational plagiocephaly also mentions repositioning as an important component of the treatment program. Repositioning is most effective between birth and 4 months, before the infant is able to roll and "self" position. Repositioning can sometimes be the sole treatment for infants with deformational plagiocephaly if the infant is young enough and the head shape deformity is apparent but not significantly abnormal.

Therapy should be considered when the infant is not meeting developmental targets, lacks active range of motion in the neck and shoulders, is unable to hold the head in midline after 16 weeks of age, or demonstrates other neuromuscular delays.

Recommendation: Allied health professionals should be aware of their role in the identification and prevention of deformational plagiocephaly.

Discussion: Health care providers have a key role in identifying infants at risk to develop deformational plagiocephaly. Staff in maternity wards, primary care offices, health clinics, therapy centers and other health related offices see infants at very young ages and are positioned to educate parents about the importance of prone positioning and frequent position changes when the baby is awake and supervised. Staff should also train parents to visually inspect the baby's head on a consistent basis to identify early signs of head shape deformity.

Recommendation: Allied health care providers should be educated on the indications for referring infants for a cranial molding orthosis.

Discussion: It is important for allied health care staff to know when to continue monitoring a deformity, and when it is advisable to refer for the infant for treatment with a cranial molding orthosis. It is also advisable to have a referral network and a relationship with specialists including a trained orthotist who can further evaluate the infant. At this time referral recommendations are based on subjective measures of severity. Future research should define Clinical Pathways and provide a severity scale that can be used easily by allied health care providers to define infants that require treatment with a cranial remolding orthosis.

Recommendation: Qualified orthotists should be included in the team management of deformational plagiocephaly.

Discussion: Orthotists trained to evaluate and treat infants with deformational plagiocephaly can provide other allied health professionals and parents with specific anthropometric measurements and other evaluative information that will help the team decide the best course of treatment. Orthotists will recommend monitoring for some infants and treatment for others based on the infant's age, developmental skills, and severity of the head shape deformity.

Recommendation: Scientific research on deformational plagiocephaly is needed to advance the understanding and treatment of this condition.

Discussion: Most of the research on deformational plagiocephaly has been undertaken in the last 10 years although the first paper was published in 1979. Research is still sketchy, and conclusions are often based on subjective measures, or even on anthropometric measures that do not reflect the 3-dimensional qualities of the deformities. Presently, it is difficult to define which infants should be referred for treatment because there is no baseline of normal asymmetry, and no evidence that referral of infants with a particular level of severity is more appropriate compared to infants with less severe conditions. Additional studies should be undertaken to answer these questions.

Recommendation: Scientific literature on the natural course of untreated deformational plagiocephaly is lacking.

Discussion: No Class I studies with random control groups exist in the literature. These studies are difficult to undertake since they involve non-treatment and raise ethical questions. There is, however, a large sample of infants in the United States and other countries with deformational plagiocephaly that have not received treatment with a cranial molding orthosis or received intervention with repositioning. Primary care providers may be in the best position to identify those infants so they can be evaluated to help determine the natural history of deformational plagiocephaly. This study should include a long-term follow-up study to document the types of developmental issues, school problems, intellectual, visual, auditory, psychological, or cosmetic issues that may arise for the infant. The research should also assess the cost of non-treatment of this condition.

Recommendation: Terminology must be standardized.

Discussion: It is impossible to determine the prevalence of deformational plagiocephaly when different terminology is used to describe the condition. Authors refer to deformational plagiocephaly as occipital plagiocephaly, frontal plagiocephaly, flat head syndrome, etc. Consistent definition and usage of the terms may also encourage the Center for Disease Control to track deformational plagiocephaly in the United States and would also facilitate comparison of treatment results and other findings. The consensus conference recommended the use of the terms deformational plagiocephaly, deformational brachycephaly, and deformational scaphocephaly.

Recommendation: Parents should learn about the potential for head shape deformities in prenatal and postnatal information provided at the hospital.

Discussion: Early parent education may help parents aggressively treat babies born with some skull deformation, and/or prevent deformity development later. Information at prenatal classes and publications intended for parent reading prior to delivery provide the information at a time when the parent may have more time to concentrate on the content prior to the birth of the infant. This preventative and repositioning information should also be included in the information received at the hospital in printed, verbal, and demonstration formats. Hospital staff should be educated in supporting the "Back to Sleep" and the "Tummy to Play" recommendations by the AAP. This information should also be provided at state health departments and pediatrician's offices and reinforced by all health professionals.

Recommendation: There are many cranial orthoses in a variety of designs. Most are based on the same biomechanical principles.

Discussion: Cranial orthoses can be designed with a side opening, top opening, closed top, air holes, over the ears, ears open, etc. These designs all have the common feature of curbing growth in the areas of total contact where the head is prominent and allowing growth in the areas of flattening where growth is desirable. The variety of designs is due to the particular strategy of the orthotist and physician working together to create the best orthosis with their treatment regimen. The treatment protocols may differ from one design to the other because of the inherent properties of the design, modification, and follow-up process. Some designs may be selected because they are most appropriate for a particular head shape, severity of the deformity, or age of the infant.

All cranial remolding designs are considered "passive" in nature since they have no moving parts that are "active". The active vs. passive nature of the treatment program is related to specific protocols that either requires frequent and ongoing modifications to the orthosis by the orthotist to direct head growth, or occasional follow-up visits. Orthoses that require less frequent follow up incorporate design features that use pre-defined growth into the original cast modifications. In all cases, treatment is supervised, and growth is monitored to ensure that it is proceeding in the proper direction and magnitude.

Recommendation: Therapists treating developmental delay and/or torticollis should be aware of deformational plagiocephaly and request a referral for a cranial molding orthosis is necessary.

Discussion: Therapists often see infants in the first six months of life when concerns about delayed development or torticollis arise. Either of these conditions, and especially torticollis, can lead to deformational plagiocephaly and other head shape deformities. As an early intervention health care provider, the therapist can educate, monitor, and refer infants for cranial molding orthoses when the head shape deformity requires treatment. Further research is necessary to substantiate the point at which referral is appropriate, but even in mild cases, it may be appropriate to refer the infant to an orthotist for baseline anthropometric measurements and orthotic evaluation.

Recommendation: Orthotists should be aware of the tendency for infants with deformational plagiocephaly to also have developmental delays and neck asymmetry, and refer the infant for physical therapy.

Discussion: There is a strong link in the literature between deformational plagiocephaly and torticollis. There is also clinical evidence to support the idea that there is a link between deformational plagiocephaly and delays in development although this is still being researched. Orthotists should include a simple developmental and active neck range of motion assessment in their patient evaluation process and request a referral for physical therapy if infants do not score within normal limits.

Recommendation: Documentation and maintenance of complete records is essential to good treatment.

Discussion: The health care team managing the infant with deformational plagiocephaly must establish baseline information about the deformity. This may be done by a variety of health care providers and may include anthropometric measurements, active neck range of motion, cephalic index, and a description of the head shape. Re-evaluation is likely to be performed on a monthly basis and may also include a head tracing or three-dimensional scan of the head that can be used to monitor head shape changes as treatment progresses. Any difficulties tolerating the orthosis, skin issues or modifications made to the orthosis, or time out of the orthosis due to illness or fit should be documented along with any parental concerns or pertinent comments.

Points of General Agreement among Participants

Q: What is the prevalence of deformational plagiocephaly in the American pediatric population?

Conclusion: The actual prevalence is unknown due in part to a lack of agreement on how deformational plagiocephaly is identified and defined. It is clear that the presentation of deformational plagiocephaly has exponentially increased due to sustained supine positioning during the day and night.

Q: What is the natural history of untreated deformational plagiocephaly?

Conclusion: The natural history is also unknown due to the lack of an untreated control group. Through growth, there appears to be a slowing of the progression of the deformity. After skull mineralization is near complete, further progression or regression is unlikely. The deformity is not limited to the calvarium and may translate to the facial and mandibular structures.

Q: What are the long-term consequences of untreated and treated deformational plagiocephaly?

Conclusion: Long-term consequences of untreated deformational plagiocephaly are unknown since large samples of these infants are now reaching their pre-teen years. Concern regarding oromaxillary, facial, ophthalmologic, torticollis, movement asymmetry, visual field deficits, musculoskeletal and other neurodevelopment issues have been raised. Psychosocial issues may exist, not excluding the caregivers of the infants.

Q: What factors determine an infant to be at high-risk for deformational plagiocephaly and what intervention(s) should be done to prevent this condition?

Conclusion: Risk factors for deformational plagiocephaly include but are not limited to: in-utero constraint, large gestational size, multiple births, premature births, early descent into the maternal pelvis, difficult deliveries, generalized hypotonicity, sustained supine positioning, congenital muscular torticollis, cervical abnormalities, and birth trauma. Early identification and intervention with repositioning and handling programs may have an impact on the severity of the condition. Specific methods for prevention of deformational plagiocephaly are not known at this time.

Q: What is the appropriate referral sequence for patients with deformational plagiocephaly who are not improving with repositioning?

Conclusion: There is significant regional variation in regards to the awareness and diagnosis of deformational plagiocephaly. The current referral pattern appears to be primary care providers to craniofacial specialists and/or centers. Appropriately trained primary care providers may not need to refer to craniofacial specialists, however, the parameters of appropriate training have not been developed.

Q: Is there adequate evidence to support the use of caregiver repositioning programs alone for infants with deformational plagiocephaly?

Conclusion: To date, most reports are qualitative and anecdotal. Improvement in overall head shape has been observed in very young infants prior to the development of head and neck

control. Improvement may not be as significant in infants with abnormal neuromuscular development. Improvements noted by repositioning alone are less significant than those resulting from more structured physical therapy and orthotic management programs, and those reported to benefit as much from repositioning as orthotic management have tended to be milder deformities.

Q: How is the efficacy of a cranial molding orthosis determined?
Conclusion: Head shape change has not been sufficiently defined due to inconsistencies of quantifiable measures in studies. Many different indices of two-dimensional and complex three-dimensional measures have been advocated yet it is not know if any of these measures adequately represent craniofacial growth. The need for inexpensive, simple, reproducible and reliable measurements is identified.

Q: What recommendations are made to improve evidence-based practices and integration into orthotic clinics?
Conclusion: Multi-center studies of orthotic efficacy should be developed and based on defined measurements. Collaboration among multidisciplinary professional groups (e.g. AAP, AAFP,etc.) would produce the most meaningful information.

Q: What is the relationship between treatment success and age at initiation of orthotic management?
Conclusion: The exact relationship is unknown due to diversity of research methods, measurements, treatment groups, design features, and many other confounding factors. Multiple factors influence treatment success such as biomechanics of skull growth, mineralization of bony plate, neuromotor maturation, etc. Chronologic age may not be the defining factor but further research is needed to determine its impact on treatment success. It is the consensus and experience of the group that early orthotic intervention leads to improved outcomes.

Q: Can a randomized control study be performed with this patient population?
Conclusion: Control groups without any intervention raise ethical and legal issues. Pseudo-control groups are attainable by being untreated through choice or circumstances, but may be biased. Comparing treated vs. untreated groups is difficult, as any advice on repositioning for the untreated group should be considered an intervention. International studies may provide an opportunity to compare intervention versus non-intervention groups.

Q: When and were should families learn about deformational plagiocephaly?
Conclusion: Deformational plagiocephaly should be discussed during prenatal and parenting classes prior to the birth of the infant, as well as during the post-partum education of the mother. As a consequence of sustained supine positioning, deformational plagiocephaly should be discussed in conjunction with the AAP's Back to Sleep program and information on SIDS. Education should be undertaken as a multidisciplinary effort and introduced as early as possible in an infant's life. Pediatricians and family practice physicians should continue to reinforce this information during well-child visits.

Q: Who is responsible for first identifying and documenting abnormal head shapes in young infants?

Conclusion: Initial health care providers such as nurses and obstetricians are the first to observe the infant's head and provide information on preventative measures. Pediatricians and family physicians should be charged with further evaluation and documentation of head shapes not responding to repositioning programs. Other allied health care professionals and parents are also responsible for bringing the abnormal head shape to the attention of an appropriately trained physician.

Q: What anthropometric measurement should be taken to document changes in infant head shape that are clinically feasible, reproducible and reliable?

Conclusion: Measurements are a common research tool and need to be developed to identify simple, reproducible, and reliable anthropometric measurements that can be obtained in a variety of clinical settings. Essentially, qualitative observable features need to be translated to quantifiable measures of asymmetry and disproportion. Still, measurements in isolation do not capture the three-dimensional quality of the deformity.

Q: What kind of diagnostic tests (e.g. x-ray, CR scan, MRI), if any, are necessary to diagnose deformational plagiocephaly or can diagnosis be made by patient history and physical examination?

Conclusion: An experienced and appropriately trained examiner can make the clinical diagnosis based on patient history and physical examination. While this may be sufficient for the initial screening, there is not enough evidence to come to consensus on a specific protocol for definitive diagnosis.

Q: What are the clinical definitions of mild, moderate and severe deformational plagiocephaly?

Conclusion: There is insufficient evidence to standardize the classifications of mild, moderate and severe. It is agreed that the development of such defining parameters would be potentially useful but must be objective, and must also consider the three-dimensional aspects of head deformation. Classifications of severity create a continuum and two scales are necessary to address various head shape deformations: (1) asymmetry, and (2) disproportion. Such classification must also consider the infant's age as relative asymmetry and disproportion change with circumferential growth factors.

Q: What clinical factors also relate to the classification of severity and decision to prescribe a cranial molding orthosis?

Conclusion: Many other factors should be considered but are not limited to: asymmetry, disproportion, chronological age, level of motor development, presence or absence of torticollis, and progression or regression of the deformity.

Q: Is subjective visual rating a factor in determining severity?

Conclusion: A validated subjective scale of visual rating is a desired goal. It may be useful to look at the literature on cleft lip and palates for examples.

Q: Are there sub-types of head shapes within the classification of deformational plagiocephaly, brachycephaly and scaphocephaly?

Conclusion: Combinations of asymmetry and disproportion may occur but have not yet been clearly defined.

Q: How is the accuracy of the infant's head, negative and positive molds assessed?

Conclusion: A wide variety of casting and scanning techniques are used to obtain negative impressions of the infant's head. Confirming mold accuracy is a basic clinical competency for the treating orthotist. There is no widely accepted method of verifying accuracy of the negative or positive mold but would be a valuable process as it relates directly to potential orthotic outcomes.

Q: Do specific casting procedures create more accurate molds?

Conclusion: Casting procedures may vary in relation to the modification procedure, orthotic design, head shape, severity of deformation, age of the infant, etc. Future research may reveal the accuracy of various techniques.

Q: How can stress to the infant and caregivers be reduced?

Conclusion: It is important to acknowledge that stress may be greater for the caregiver than the infant during the casting procedure. Educational materials are useful in reducing stress along with family participation during the procedure. It is generally accepted that shorter procedures are potentially less stressful for everyone.

Q: Is repeat casting at the end of treatment more valuable than anthropometric measurements?

Conclusion: There is no consensus regarding casting versus measurements at the end of treatment. Data recording at the initiation and end of treatment are potentially useful although the reliability of linear measurements has not yet been determined. Development of a consistent method of data recording may be a valuable research project.

Q: How does the degree of severity influence cast modifications?

Conclusion: The targeted degree of correction is a product of the severity of head deformation at the initiation of the orthotic treatment program and specific orthotic design. Modification is an orthotist-specific process at this time and is often based upon the anticipated growth rate of the infant's skull. The primary goal(s) of symmetry and proportion are considered with modifications designed to direct skull growth toward a more normal shape.

Q: Is force/pressure actually applied to the infant's skull or does total contact eventually provide a resistant force?

Conclusion: Most orthotic designs apply some initial force as evidenced by erythema of the skin in those areas of observable skin contact. All orthoses eventually provide a resistant force if head shape change is to occur. The basic premise of cranial molding orthoses is that growth will be constrained in the areas of contact and growth will be encouraged into areas of voids within the orthosis contours.

Q: How are modifications verified upon completion of the rectification process?

Conclusion: Modifications can be verified by visual inspection, hand measuring techniques or scanning technology.

Q: Do cranial molding orthoses address facial and/or mandibular asymmetry in deformational plagiocephaly?

Conclusion: There is evidence to suggest that facial and mandibular asymmetry exists in infants diagnosed with deformational plagiocephaly. Subjective evidence suggests that correction of the calvaria results in some correction of the facial and mandibular structures. However, no quantifiable data is available to support these subjective observations.

Q: What is the magnitude of the force of growth of the infant skull, and can this be measured and quantified?

Conclusion: No current evidence exists to address this question. This topic needs further exploration and studies may be currently in process.

Q: Are there specific clinical criteria that would make an infant a better candidate for a (1) helmet or band, or (2) lined or unlined style of cranial molding orthosis?

Conclusion: The variability of designs make the terms "helmet" and band" difficult to apply. Cranial molding orthosis (CMO) is the term generally accepted by consensus. No studies have been conducted to compare the effectiveness of different CMO styles but should be an area of future research.

Q: What is the definition of "active/dynamic" versus "passive", "band" versus "helmet", and "corrective" versus "protective" designs?

Conclusion: By design, all CMOs are static. The term dynamic refers to the process of management that is determined by a feedback loop dependent upon the growth of the infant's head and appropriate modifications made to the orthosis. All CMOs are corrective if a positive change in head shape occurs.

Q: How do materials and material strength relate to orthotic management?

Conclusion: The efficacy of material use in CMOs has not been proven and additional studies should be undertaken. No studies exist comparing one material to another when used in the design of a cranial molding orthosis.

Q: How would a research model be designed to accurately assess the correction provided by different polymers, considering the addition of liners may skew the performance of a polymer?

Conclusion: The development of an animal model for study of CMOs may be useful however it may be difficult to accomplish due to an inherent bias relative to proprietary issues of manufacturing and distribution of various styles of cranial molding orthoses.

Q: When an infant presents with deformational plagiocephaly, are the frontal and occipital areas of flattening addressed separately or simultaneously?

Conclusion: Most CMOs provide two voids to address both areas simultaneously. Indications for alternative approaches are dependent upon the individual presentation as well as additional parental or clinician concerns. The occipital region of the skull usually corrects first, followed by the frontal region.

Q: What is the prime period of time for orthotic remodeling and when to remodeling efforts become significantly less effective and ultimately ineffective?

Conclusion: There is a closing window of opportunity to achieve effective symmetry and/or proportion in the rapidly growing infant. The ideal age range for optimal correction is three to eight months of age, and it is ideal to have orthotic treatment completed by 12 months of age. Improvement is clearly possible after 12 months of age with longer treatment times and reduced correction.

Q: How does correction of the flattened areas occur?
Conclusion: Growth is not a uniform process. The orthosis generally guides growth from the periphery of the void. The exact biomechanical progression of the remolding process has not been documented.

Q: Which head shape responds the best to remolding procedures?
Conclusion: Significant symmetry is achieved with the orthotic management of plagiocephalic head shapes. Orthotic management of scaphocephalic and brachycephalic head shapes results in improved proportion. Improvements in overall head shape can be obtained is all three deformational presentations. Plagiocephaly is often considered "easier" to correct and remold to within normal limits of symmetry.

Q: Are there different norms in cranial shapes for various ethnic group?
Conclusion: There are definitely different norms although scientific documentation is scarce. Cultural patterns of infant sleep positions and caregiver handling techniques may influence these difference. It is clear that a need exists for new documentation of norms of head shape for various racial groups.

Q: Does unresolved torticollis have an effect on length of orthotic treatment?
Conclusion: Unresolved torticollis does affect the outcome and increases the length of orthotic treatment programs. Upon completion of the remolding process, regression of head deformation may occur if the torticollis is untreated or not yet resolved.

Q: How do orthotic treatment protocols different when addressing moderately flat versus severely flat areas?
Conclusion: Treatment protocols are largely age dependent. Treatment strategies for severe head deformations are challenging and may require multiple orthoses to obtain a satisfactory head shape. The potential adjustability of the CMO is important in the treatment of severe head deformations.

Q: What are the clinical indications for the use of multiple orthoses?
Conclusion: The need for multiple orthoses depends on the input of physicians and family regarding outcomes. There is an infrequent need for multiple orthoses; most infants complete effective treatment programs with a single cranial molding orthosis. Very young and very old infants may require multiple orthoses, along with infants with severe head deformations. Ultimately, multiple CMOs may be indicated when the redirection of corrective forces cannot be achieved within the original orthosis.

Q: What are general treatment protocols for the management of infants with deformational plagiocephaly and cranial molding orthoses?
Conclusion: The ideal wear time is 22 – 23 hours per day for optimum outcomes. Exceptions to the recommended wearing schedule include skin sensitivity, excessive heat, febrile illness or

other factors. In these instances, removal of the CMO for short periods of time may be necessary. It should be noted that extended time out of the orthosis, including isolated night time wear, may impact fit and function of the CMO, extend treatment time, impact treatment outcome, and/or result in the necessity of an additional orthosis to achieve desired results.

Q: When is orthotic management discontinued?

Treatment is discontinued when agreement is reached between the parents and the medical care team in the context of expected improvement and biological induced constraints such as age, rate of cranial growth, and level of cranial skeletal maturity.

Q: What is the most commonly accepted theory of the pathogenesis of craniosynostosis?

Conclusion: There is not one commonly accepted theory.

Q: How often are protective or remolding cranial orthoses indicated after surgery?

Conclusion: No data exists to answer this question. It is largely the preference of the surgeon whether an orthosis is used post-operatively. The type of surgery may determine whether the post-operative orthosis is protective, remolding or both.

Q: What are the clinical signs that contraindicate continued post-operative remolding efforts with a CMO?

Conclusion: Regression of head shape deformation is a warning sign that further evaluation of the cranial structures is needed.

Q: What role do cranial orthoses have in the post-operative treatment of craniosynostosis?

Conclusion: The roles of the post-operative CMO are to protect the infant's head from environmental factors, stabilize the surgical reconstruction, and/or to produce continued improvement in overall head shape.

Q: Does the type of surgical procedure influence the use and design of post-operative cranial orthoses?

Conclusion: The type of surgery may influence the application of corrective forces provided by the post-operative orthosis and therefore communication between orthotist and surgeon is recommended.

Q: What factors best determine the length of the post-operative treatment program?

Conclusion: The protocols of the treatment team determine the length of post-operative orthotic management. Factors may include but are not limited to the type of surgery performed, pre- and post-operative head shape, age of the infant, as well as other medical conditions.

Q: What specific casting procedures should be followed for post-operative orthotic management?

Conclusion: It is generally necessary to wait two to three days for the post-operative edema to subside before casting the infant's head. Care must be taken to protect the incision from contaminants.

Q: Should infants with combined congenital muscular torticollis (CMT) and plagiocephaly comprise a subgroup of children with CMT? Should infants with deformational plagiocephaly who develop secondary impairments of the cervical muscles be included in the diagnosis of CMT?

Conclusion: Deformational plagiocephaly is often accompanied by neck muscle tightening or imbalance, making the orthotic management more difficult. Infants with neck tightness and/or asymmetry combined with deformational plagiocephaly should be referred for therapy evaluation.

Q: How is CMT distinguished from other types of torticollis?

Conclusion: Classic CMT is characterized by a tilt to one side and rotation in the opposite direction. It is usually synonymous with sternocleidomastoid (SCM) tightness. Other presentations of torticollis involve muscles in the neck, shoulder and trunk.

Q: Currently there are no age-dependent norms for cervical range of motion or validity of measurement tools. How much asymmetry is considered abnormal?

Conclusion: The development of age-dependent norms for cervical range of motion would be beneficial in the evaluation and treatment of infants with torticollis and deformational plagiocephaly.

Q: How are infants with deformational plagiocephaly identified who would most benefit from a formal physical therapy and/or occupational assessment and treatment?

Conclusion: Indications for referral to physical therapy include but are not limited to:

- Persistent positional preference not responsive to repositioning efforts
- Persistent head tilt or turn despite home range of motion regimen
- Developmental delay and/or hypotonia
- Asymmetry of nuchal folds or erythema in the neck folds may be an indication of torticollis
- Significant limitations in active and passive range of motion in spite of home neck exercises

It is generally accepted that earlier intervention leads to improved results.

Q: What factors influence the decision to refer for an orthotic evaluation?

Conclusion: Physical factors influencing the referral for orthotic evaluation include but are not limited to:

- Failure of expected improvement
- Obvious ear misalignment in the coronal plane
- Progression of head shape deformity
- Facial asymmetry
- Obvious mandibular asymmetry in position or motion
- Obvious malar prominence
- Obvious anteroposterior orbital dystopia

- o Obvious frontal asymmetry
- o Obvious occipital asymmetry
- o Obvious proportional deviation

Q: What is the quality of evidence that deformational plagiocephaly is associated with neuropsychological and/or functional deficits, and if treatment with remolding orthoses prevents later trauma and restores function?

Conclusion: Significant opportunities exist for evidence-based studies as the literature is scarce regarding neuropsychological and/or functional deficits in relation to deformational plagiocephaly. Quality of life and psychosocial issues should also be researched.

Q: What recommendations are made for Standardization of Terms to describe both the condition and the devices used to treat it? How can we define/classify a deformity in a non-subjective manner?

Conclusion: Recommendations for terminology include deformational plagiocephaly, deformational brachycephaly, and deformational scaphocephaly. It is important to note that these entities are not always seen in isolation as combinations are possible and frequent. No attempt was made to define these terms. Consideration of the American Medical Association's (AMA) definitions for cosmetic and reconstructive procedures will be useful in defining these conditions and treatment protocols.

Q: How can we best measure treatment outcomes?

Conclusion: This is a critical research priority as insufficient data exists at this time.

Q: How do we identify and recruit untreated infants in order to document the natural history of this condition?

Conclusion: Collaboration with primary care providers should be explored to document the natural history of untreated deformational plagiocephaly. Consultation with an epidemiologist is also recommended.

Standards of Care

Identification

Some infants are born with skull deformation secondary to intrauterine positioning, crowding, long or difficult labors, or method of extraction from the birth canal. After the deforming force is removed after birth, many skulls assume a more symmetrical shape within the first six weeks as the infant is held and placed in a number of different positions. If for some reason the infant is continuously placed in the same position, the skull deformity is not likely to disappear and the infant's head will tend to rest on the previously flattened area. When this occurs, the head shape deformity will generally stay the same or get worse. Occasionally, there are medical reasons for maintaining the infant in a specific position. Premature infants are placed in side-lying positions in ICU to have best access for monitoring devices. Infants with reflux, chronic ear infections or other problems may require a semi-reclined position to reduce feeding, breathing, or other difficulties. In addition, infants with developmental delay often do not move as freely as other babies and develop head shape abnormalities secondary to the consistency of supine positioning.

Parents are often the first to identify the unusual skull shape and bring it to the physician's attention. When identified in the first three months, the AAP recommends that physicians discuss repositioning strategies with parents and assess and monitor the infant's head shape at every visit. After three months, if the repositioning efforts have not been effective and the head shape deformity involves secondary changes such as ear misalignment or forehead asymmetry, the physician may refer the infant to a craniofacial specialist for further evaluation. A primary care physician may also refer the infant directly to an orthotist for evaluation and treatment with a cranial molding orthosis after ruling out the possibility of craniosynostosis. If the infant has neck muscle asymmetry that is not corrected with repositioning and stretching exercises, or if the baby has delays in the development of midline head control or acquisition of other developmental skills, the physician should refer the infant for physical or occupational therapy.

Optimal Age for Treatment

Infants less than three months old benefit the most from repositioning and stretching exercises. If the infant has a deformity that is mild, only involves the posterior quadrant without secondary changes, or if the cephalic index is less than two standard deviations above or below the mean, the head shape should continue to be monitored and the repositioning program extended another month. If the head deformity remains mild or improves by the time the infant is able to achieve midline head control, roll, or sit independently then no further treatment is warranted.

If after three months the head deformity progresses in spite of the repositioning and exercise program, the infant should be referred for a cranial molding orthosis. Moderate to severe head shape deformities of a proportional nature (deformational scaphocephaly or brachycephaly) have a cephalic index this is two or more standard deviations above or below the mean. Moderate to severe plagiocephaly is present if there is flattening in the posterior quadrant and secondary changes such as ear misalignment, forehead bossing or prominence on the same side as the posterior flattening. Facial asymmetry may also be involved in moderate to severe cases. Orthotic treatment is most effective from three to eight months of age when cranial growth is about 1 cm per month. It is recommended that physicians refer infants by six months

of age to capitalize on this rapid growth period and to facilitate the completion of treatment by the time the infant is a year old. Although skull growth slows after 12 months of age, head shape changes can still occur until about 18 months of age. Orthotic management after 12 months takes longer due to reduced growth and a more rigid skull, and treatment outcomes may not be as significant.

Orthotic Prescription

When an orthotist receives the prescription for a cranial remolding orthosis, the appointment is scheduled as quickly as possible since treatment is time-sensitive. Parents are encouraged to check with their insurance provider prior to the appointment to determine if there is coverage for cranial molding orthoses. Insurance companies may require that the family obtain treatment at an orthotic facility in network or request specific anthropometric measurements prior to approving a cranial molding orthosis. It is likely that insurance companies will request a letter of medical necessity from the referring physician before approving the claim. Parents may choose to pursue treatment at their own expense if the insurance company does not approve orthotic management for a cranial molding orthosis.

Parents may schedule an appointment with the orthotist prior to beginning treatment to ask questions, request an evaluation of their infant's head shape deformity, and determine if they want to proceed with the treatment program. After choosing an orthotist, a formal evaluation and assessment of the predisposing factors leading to the head shape deformity is undertaken. The orthotist performs a physical exam of the skull and notes any ridging, areas of flattening and bossing. Anthropometric measurements are acquired to obtain a baseline of information. Scanning technology may replace hand measurements. Clinical photographs of frontal, posterior, right and left sagittal, and vertex views are obtained to qualitatively document the head shape.

Casting or Scanning Process

The orthotist will acquire a cast or scan of the infant to create a negative model of the patient's skull. The preparation and casting process takes about 15-30 minutes, and the scan is usually acquired in less than one minute. Either procedure may upset the infant and/or parent, but are time limited and do not hurt the infant. The cast will be poured and/or the scan sent to an FDA cleared facility for the fabrication of a cranial molding orthosis. Once the positive model is created, it is modified to provide total contact in the areas where growth is to be curbed and provide space in the areas where growth is desirable. A custom molded cranial orthosis is fabricated from the positive cast.

Orthotic Design

Designs of cranial orthoses vary according to the preference of the physician who refers the infant and the design favored by the orthotist. The design is also chosen based on its ability to address the specific head shape deformity of the infant. Regardless of the design, the orthosis should be fit within two weeks of the casting or scanning process.

Follow-Up Appointments

Fitting and follow-up protocols are design and age-specific and vary from weekly to monthly follow up visits. If the design has a thick inner liner, the orthotist monitors the infant's head growth and ensures that there is adequate space for growth in the areas of flattening. The liner is ground away to direct growth into the flattened areas. Some orthoses have no inner

liner, and the plastic shell can be heated to allow more space for growth if growth occurs that has not been modified into the orthosis prior to fabrication.

At intervals throughout treatment, the orthotist documents head shape changes using follow up photographs, anthropometric measurements, and other significant indexes. This data helps to guide the treatment process, and determine when the treatment program should be discontinued. The orthotist sends the referring physician periodic updates about the infant's skull shape changes.

Treatment Termination

The average length of orthotic treatment is three to four months. Treatment is usually shorter for younger infants and longer for older infants. Very young infants who have not developed midline head control, rolling, or sitting, may require a second orthosis to prevent regression of the head shape. Older infants may also require longer treatment programs because their heads are growing slowly and their skulls are thicker and more resistant to change. Generally, the medical team determines when treatment can be safely discontinued, and this team includes the parents, orthotist, therapist, and the referring physician. By reviewing the documentation, the team can assess progress and determine if the treatment goals have been obtained. Treatment is discontinued as the infant's head approaches improved symmetry and/or proportion. If the orthosis is still fitting properly, nighttime only wear may be recommended to maintain the correction and encourage additional growth.

Outcomes

Treatment is considered successful when the head shape is acceptable to the parents and team members. Full symmetry may not always be possible, and for these infants, treatment is discontinued when the infant's head shape is considered mild or considerably improved and does not merit further orthotic intervention. Clinical photographs and all anthropometric measurements and indexes are repeated at the end of treatment. A report is sent to the referring physician, and the orthotist documents the treatment using the pre- and post-treatment data.

Follow-up visits ensure that the head shape is not reverting to its pre-orthotic management state. Regression is very rare but it is possible when treatment is completed before the infant is able to roll, sit, and reposition itself independently. A one-month follow-up appointment is a good way to affirm the efficacy of the orthotic results or determine if additional treatment is needed to promote greater symmetry.

Standards for Assessing Outcomes

A variety of techniques for measuring change have been published, including the acquisition of measurements using bony landmarks, mathematical equations comparing pre- and post-treatment linear measurements of a particular cross section of the skull, three-dimensional scans, and parent surveys. Currently, no standards exist for assessing treatment outcomes for infants with deformational plagiocephaly.

Linear Anthropometric Measurements

The method used most often in the literature to assess outcomes is a series of linear, anthropometric measurements. These include measurements between facial and skull landmarks to assess upper facial asymmetry, cranial vault asymmetry, and cranial base asymmetry. In the hands of a trained clinician, these measurements are reproducible and can document change. A shortfall of this measurement process is that deformational plagiocephaly is a three-dimensional deformity, and linear measurements are two-dimensional unless they are plotted using a method that is rather complex and time consuming. Linear measurements also do not clarify the relationship of the measurements to the size of the infant's head. A measurement of 6 mm of difference is more significant in a tiny newborn baby's head than in the larger head of a 14-month-old child. There was consensus among those present at the CSOP conference that linear anthropometric measurements serve as a good documentation tool but future research (i.e. with three-dimensional scanners) should further define the complexity of head shape deformations and efficacy of orthotic treatment programs.

Cranial Index

One type of proportional anthropometric measurement is the cranial index. This index is the maximum width of the head divided by the maximum length of the head. This mathematical formula results in a ratio that can be changed to a percentage and compared at the beginning and end of the treatment program. This ratio also provides a method of comparing the treatment results of one infant to another. The cranial index is most appropriate for measuring outcomes in infants with proportional deformities such as brachycephaly and scaphocephaly. This index is consistently reproducible and there was consensus that since this is a index rather than a linear measurement, it is a useful measurement for assessing outcomes when the goal of orthotic management is to change the proportion of the infant's head shape.

Clarren's Method of Measuring Cranial Vault Asymmetry

In this measurement method, a photograph is taken of the vertex or top view of the infant's skull. Lines are drawn to define the anterior-posterior axis, the minimal axis and maximal axis of the skull. The angles where the minimal axis and maximal axis cross the anterior-posterior axis are compared and the difference in the angle is noted. The lengths of the minimal and maximal lines are compared to each other in the form of a ratio. Over the course of treatment it is expected that the difference between the angles will decrease and the percentage of difference between the minimal and maximal axes will increase. This method assesses change at one particular cross-section and can be used for comparison. It is limited in its ability to assess the three-dimensional deformity, and the measurements are difficult to reproduce reliably.

Loveday and De Chalain's method of Measuring Cranial Vault Asymmetry

A flexicurve ruler is used to obtain the shape of the infant's skull at the equator with markings indicating the placement of the inion, nasion and ears. The inside contour of the flexicurve is traced on a piece of paper as are the anatomical markings. The midline is recorded and lines are diagonal lines are drawn at 30 degree angles from the midline. The lengths of the lines are measured and compared in the form of a ratio. Over the course of treatment it is expected that the difference will decrease and approach 0. This method captures anterior and posterior asymmetry at one cross-section. It can be used for comparison, but is still a two-dimensional representation of a three-dimensional deformity.

Survey Method of Assessing Change

A number of studies have been published using parent and physician surveys to document change. The premise of these surveys is that since the deformity is visually observable, the subjective assessment of the degree of deformity before and after treatment is a useful method of assessing change. The visual survey method lacks objective measures and does not provide an assessment tool that can be used for evidence-based practice. It also makes it difficult to assess treatment results from one infant to another.

Three-Dimensional Laser Scanning

Loveday, Teichgraeber, and others mention three-dimensional imaging as a likely tool for measuring head shape deformity and change. In the past, infants were unable to stay still for the amount of time it took to scan their heads. But recent advances in scanning technology have decreased the time of the scanning process, or even allowed the infant to move while being scanned. There was consensus that capturing a three-dimensional image of the infant's head shape and being able to compare images at the beginning and end of treatment represents the most viable way to assess change over time.

At this point, no studies have been published using three-dimensional scanner technology although a poster exhibit was presented at the American Cleft Palate and Craniofacial Association meeting in 2003 using the STARscanner to document treatment results. Lectures were also given at the Association of Children's Prosthetic and Orthotic Clinics in 2003 and 2004 to present research conducted using the STARscanner to document change[43]. A 3DMD scanner capable of capturing a three-dimensional image using digital photography is being used to provide a high-resolution image imposed on the three-dimensional shape. A hand-held scanner by Polhemus uses a remote tracking device to scan and track the infant's head even when the infant is moving. Scanners are already being used to acquire scan data to fabricate cranial orthoses and track head shape changes. This technology is already in place at many hospitals and orthotic and prosthetic clinics across the United States and Europe, and may facilitate multi-center research studies using the same equipment and documentation.

Not every facility treating infants with deformational plagiocephaly has access to a scanner since the equipment is still too expensive for many orthotic and prosthetic facilities. But it is important that facilities with the technology conduct scientific studies to address many of the questions that have remained unanswered for many years. Scientific studies will promote better treatment techniques and understanding of this condition.

Research Recommendations

Clinical practice involves the evaluation, prediction, intervention and control of the individual presentations and/or problems presented for orthotic management. Scientific method also allows clinicians to describe, predict and explain clinical observations. This large patient population has increased significantly over the last 10 years and, as a result of this rapidly growing demand, has been poorly studied. Treatment programs have been developed and adapted to meet the needs of parents seeking effective, non-surgical treatment for their infants. Little is known about the natural history of untreated deformational plagiocephaly or any long-term functional limitations associated with non-treatment. Measurement of treatment outcomes is often qualitative and subjective and lacks solid scientific foundation. It is clear that much research needs to be performed on this patient population to provide clear guidelines and indications for various treatment methods.

The following research recommendations are excerpts from "Deformational Plagiocephaly: Recommendations for Future Research" by Timothy R. Littlefield, MS and Kevin M. Kelly, PhD.

> *In comparison to other topics in medicine the research available on deformational plagiocephaly is relatively sparse. Among these articles are a wide variety of studies that range from simple case reports and clinical observations to well designed cohort studies. Unfortunately, the large variations between these studies with respect to purpose, study design, population size, and terminology can often make it difficult to draw conclusions from the existing literature.*
>
> *Considering that the majority of articles on the subject have been written in just the past decade, significant progress has been made in understanding the etiology of this condition, differentiating it from the more severe yet rare forms of craniosynostosis, and the success of various treatment modalities. A true milestone in these efforts was the 1997 Skull Molding Symposium in which the countries' leading craniofacial and pediatric neurosurgeons met to develop a consensus regarding the diagnosis and management of this condition.[48] While a formal consensus was never reached, from this meeting came some very significant findings. This included work on the differential diagnosis; the identification that a diagnosis could be made through detailed clinical observation of the head shape and that CT scans and X-rays were not required unless patency of the sutures remains in question; the laying to rest of the concept of the 'sticky suture' (not fused but acting as it is); and the recognition that surgery is not indicated for the vast majority of these infants. At the time, the belief was that conservative interventions, such as repositioning and neck exercises, were all that was required for successful management of this condition. The use of molding helmets was not yet widely endorsed as many specialists still felt that this condition would round out on its own. Furthermore, molding helmet studies that had been done at that time had small sample sizes and were poorly controlled. The natural history of untreated plagiocephaly, also failed to be available.*
>
> *In the mid 1990's, it was also felt that all cases of abnormal head shape needed to be referred to a specialist as the pediatrician, family practice, and general practitioner were not qualified to make a differential diagnosis between synostotic and non-synostotic*

plagiocephaly. This is a trend which has significantly changed in recent years. First, as the specialists have become overrun by the sheer number of infants presenting with this condition, and second as more efforts have been made to educate the previously mentioned disciplines.

Even with many of these advances, there remain many critical issues that need to be addressed. Many issues are still debated within the medical community, and research needs to be conducted to address concerns not only of physicians, but also of treatment providers and third-party payers. Therefore, in the remainder of this paper we discuss areas for future research which we believe are critical to the subject of deformational plagiocephaly. These include:

- ***Standardization of terminology*** *- Significant confusion exists because of the wide variety of terms used to describe this condition.*

- ***Documentation of the natural history*** *- To date there have been no studies documenting the true natural history of untreated plagiocephaly.*

- ***Investigation of functional issues associated with plagiocephaly*** *- To date there has been no conclusive evidence that demonstrates a relationship between functional problems and untreated plagiocephaly.*

- ***Well-controlled treatment studies*** *- The criticism remains that many of these studies are not 'controlled'.*

- ***Demonstration of the cost effectiveness of treatment*** *- If cranial remodeling devices are to be continued to be covered by third party payers, evidence that treatment of plagiocephaly prevents future functional problems will also need to be demonstrated.*

- ***Establishment of treatment guidelines*** *- The significant number of infants requiring intervention calls for some form of guidelines to be developed and endorsed by a reputable medical organization.*

Appendices

1. Literature Review on Deformational Plagiocephaly
2. Literature Review on Repositioning
3. References

Literature Review – Appendix 1

Appendix 1
Efficacy of Cranial Remolding Orthoses

Author	***Year***	***Study Type/Level of Evidence***	***Quality Rating***	***Population***	***Intervention***	***Outcomes Measured***
Clarren, Smith & Hanson	*1979*	*Descriptive study Level of evidence: VI*	*	*10 infants with deformational plagiocephaly*	*Cranial remolding orthosis*	*Visual assessment*
Clarren	*1981*	*Before and after Level of evidence: V*	*C*	*25 infants with deformational plagiocephaly 3 non treated*	*Cranial remolding orthosis*	*Cranial vault asymmetry (computed the difference in the angles where the min/max axes intersected the mid-sagittal line)*
Pattisapu, Walker, Myers & Cheever	*1989*	*Descriptive study Level of evidence: VI*	*C*	*24 infants with deformational plagiocephaly*	*Repositioning and/or cranial remolding orthosis*	*Family graded cosmetic changes as good, fair, poor, and no change.*
Ripley, Pomatto, Beals, Joganic, Manwaring & Moss	*1994*	*Retrospective before and after Level of evidence: V*	*C*	*72 infants with deformational plagiocephaly*	*Cranial remolding orthosis*	*Cranial vault asymmetry, skull base asymmetry, orbital tragial depth asymmetry*
Argenta, David, Wilson & Bell	*1996*	*Before and after Level of evidence: III*	*C*	*51 infants with deformational plagiocephaly*	*Repositioning or prefabricated soft helmet*	*Visual assessment*
Loveday & De Chalain	*1996*	*Retrospective cohort Level of evidence: III*	*C*	*74 infants with deformational plagiocephaly*	*Repositioning or cranial remolding orthosis*	*Two-dimensional head tracings, cranial index, cranial vault asymmetry*
Pomatto, Littlefield, Manwaring & Beals	*1994*	*Before and after Level of evidence: VI*	*C*	*3 triplets with deformational plagiocephaly*	*Cranial remolding orthosis*	*Cranial vault asymmetry, skull base asymmetry, orbital tragial depth asymmetry*

Author	Year	Study Type/Level of Evidence	Quality Rating	Population	Intervention	Outcomes Measured
Pollack, Losken & Fasick	*1997*	*Prospective cohort Level of evidence: III*	*B*	*69 infants with deformational plagiocephaly*	*Repositioning or cranial remolding orthosis*	*Parents were asked to rate their satisfaction with their child's appearance and whether the craniofacial features were normal or nearly normal, abnormal but acceptable, unacceptable and in need of further intervention.*
Littlefield, Pomatto, Beals & Joganic	*1997*	*Retrospective before and after Level of evidence: V*	*C*	*285 infants with deformational plagiocephaly*	*Cranial remolding orthosis*	*Cranial vault asymmetry, skull base asymmetry, orbital tragial depth asymmetry to determine their stability over time.*
Moss	*1997*	*Prospective Cohort Level of evidence: III*	*C*	*72 infants with deformational plagiocephaly*	*Cranial remolding orthosis or non-treatment*	*Cranial vault asymmetry, skull base asymmetry, orbital tragial depth asymmetry*
Kelly, Littlefield, Pomatto, Manwaring & Beals	*1999*	*Retrospective case study Level of evidence: V*	*C*	*504 infants with deformational plagiocephaly*	*Cranial remolding orthosis*	*Cranial vault asymmetry, skull base asymmetry, orbital tragial depth asymmetry, cranial circumference, cranial breadth, cranial length, impact of age and severity on treatment length and correction*
Kelly, Littlefield, Pomatto, Ripley, Beals & Joganic	*1999*	*Retrospective Before and after Level of evidence: V*	*C*	*258 patients with deformational plagiocephaly*	*Cranial remolding orthosis*	*Age at initiation of treatment, total treatment time, overall asymmetry*
Mulliken, VanderWoude, Hansen, LaBrie & Scott	*1999*	*Cohort study Level of evidence: III*	*B*	*114 infants*	*Repositioning or cranial remolding orthosis*	*Oblique cranial measurements, measured % changed and age at beginning of therapy.*
Littlefield, Pomatto & Kelly	*2000*	*Case study Level of evidence: VI*	*C*	*4 infants with deformational plagiocephaly*	*Cranial remolding orthosis*	*Cranial vault asymmetry, skull base asymmetry, orbital tragial depth asymmetry, cranial circumference*

Author	Year	Study Type/Level of Evidence	Quality Rating	Population	Intervention	Outcomes Measured
Vles, Colla, Weber, Wilmink & Kingma	*2000*	*Cohort study Level of evidence: III*	*B*	*105 infants with deformational plagiocephaly*	*Cranial remolding orthosis or non-treatment*	*Parent severity scale pre and post treatment with helmet or repositioning.*
Terpenning	*2001*	*Before and after Level of evidence: V*	*C*	*12 infants with deformational plagiocephaly*	*Cranial remolding orthosis*	*Cranial vault asymmetry, skull base asymmetry, orbital tragial depth asymmetry*
Teichgraeber, Ault, Baumgartner, Waller & Messersmith	*2002*	*Before and after cohort Level of evidence: III*	*B*	*125 infants with deformation plagiocephaly*	*Analyzed whether age at the beginning of treatment was related to treatment outcomes*	*18 anthropometric measurements Correlation between age at start of treatment and treatment outcomes (none)*
Coulter-O'Berry, Hylton-Plank, Lima & Fish	*2003*	*Cohort study Unpublished poster Level of evidence: III*	*B*	*169 infants*	*Repositioning/PT only, PT/cranial remolding orthosis, cranial remolding orthosis, or non-treatment*	*Cephalic index, overall symmetry ratio, posterior symmetry index, cranial vault asymmetry index (Loveday)*
Teichgraeber, Seymour-Dempsey, Baumgartner, Xia, Waller & Gateno	*2002*	*Retrospective Cohort Level of evidence III*	*B*	*23 infants with deformational brachycephaly and 71 infants with deformational plagiocephaly*	*Cranial remolding orthosis*	*Cephalic index, cranial vault asymmetry, age*

***Evidence Statement**: There is insufficient evidence on the basis of treatment effects in this group of studies. The relationship between deformational plagiocephaly and the various treatment regimens of self-correction, repositioning, therapy and orthotic modeling has not been well substantiated in the literature.*

***Grade of Recommendation**: The assigned grade relates to the quality of the statistical evidence and scientific rigor. This body of scientific evidence receives an overall grade of C.*

Literature Review – Appendix 2

Appendix 2
Studies Related to Repositioning

Author	*Year*	*Study Type*	*Quality Rating*	*Population*	*Intervention*	*Outcomes Measured*
Hellbusch, Hellbusch, Bruneteau	*1995*	*Retrospective before and after Level of evidence: V*	*C*	*25 infants involved in an active counter-positioning program to manage deformational plagiocephaly*	*Active counter-positioning program*	*Parent's rated their baby's plagiocephaly before and after treatment with a rating of severe, moderate-severe, moderate, mild-moderate, mild, and normal*
Pople, Sanford, Muhlbauer	*1996*	*Retrospective before and after Level of evidence: V*	*C*	*100 consecutive infants*	*Altered sleep position and surgery*	*Parents and physician rated the child's degree of flattening and facial symmetry as improved, not changed, or worse.*
O'Broin, Allcutt, Earley	*1999*	*Retrospective before and after Level of evidence: V*	*C*	*39 infants with posterior plagiocephaly*	*Sleep posture modification and therapy*	*Parents were asked to score the deformity at the first visit and 1 year later.*
David, Menard	*2000*	*Retrospective, descriptive Level of evidence: V*	*C*	*204 patients with occipital plagiocephaly over 16 years*	*183 patients responded to head positioning and therapy. 21 underwent surgery to correct their deformities*	*94% of parents noted "acceptable improvement".*

***Evidence Statement**: There is insufficient evidence on the basis of treatment effects in this small group of studies. The relationship between deformational plagiocephaly and repositioning and/or surgery has not been well substantiated in the literature.*
***Grade of Recommendation**: The assigned grade relates to the quality of the statistical evidence and scientific rigor. This body of scientific evidence receives an overall grade of C.*

References – Appendix 3

1. The New International Webster's Comprehensive Dictionary of the English Language, Trident Press International p. 1281, 1996.
2. Goodman CR, Chabner E, Guyton DL. Should early strabismus surgery be performed for ocular torticollis to prevent facial asymmetry? J Pediatr Ophthalmol Strabismus 32:162-166, 1992.
3. Frederick DR, Mulliken JB, Robb RM. Ocular manifestations of deformational plagiocephaly. J Pediatr Ophthalmol Strabismus 30:92-95, 1993.
4. Limon de Brown E, Monasteries FO, Feldman MS. Strabismus in plagiocephaly. J Pediatr Ophthalmol Strabismus 25(4):180-190, July-Aug 1998.
5. Kane AA, Lo LJ, Vannier MW, Marsh JL. Mandibular dysmorphology in unicoronal synostosis and plagiocephaly without synostosis. Cleft Palate-Craniofac J 33(5):418-423, 1996.
6. Clarren SK, Smith DW, Hanson JW. Helmet treatment for plagiocephaly and congenital muscular torticollis. J Pediatr 94(1):43-46, 1979.
7. American Academy of Pediatrics Task Force on Infant Positioning and SIDS. Pediatrics 89:1120-1126, 1992.
8. American Academy of Pediatrics Task Force on Infant Sleep Position and Sudden Infant Death Syndrome. Changing concepts of Sudden Infant Death Syndrome: Implications for infant sleeping environment and sleep position. Pediatrics 105(3):650-656, 2000.
9. Davis BE, Moon RY, Sachs HC, Ottolini MC. Effects of sleep position on infant motor development. Pediatrics 102:1135-1140, 1998.
10. Jantz JW, Blosser CD, Fruechting LA. A motor milestone change noted with a change in sleep position. Arch Pediatr Adolesc Med 151:565-568, 1997.
11. Littlefield, TR, Pomatto JK, Kelly KM. Dynamic orthotic cranioplasty treatment of the older infant. Neurosurg Focus 9:1-4, 2000.
12. Mulliken JB, Vander Woude DL, Hansen M, LaBrie RA, Scott RM. An analysis of posterior plagiocephaly: Deformational versus synostotic. Plast Reconstruc Surg 103(2):371-380, 1999.
13. Pollack IF, Losken HW, Fasick P. Diagnosis and management of posterior plagiocephaly. Pediatrics 99:180-185, 1997.
14. Bruneteau RJ, Mulliken JB. Frontal plagiocephaly: Synostotic, compensational, or deformational. Plast Reconstruc Surg 89(1):21-30, 1992.
15. Argenta LC, David LR, Wilson JA, Bell WO. An increase in infant cranial deformity with supine sleeping position. J Craniofacial Surg 7(1):5-11, 1996.
16. Kolar JC, Salter EM. Adapted from Craniofacial Anthropometry. Springfield, IL. Charles C. Thomas Publisher, 1997.
17. Kelly KM, Littlefield TK, Pomatto JK, Manwaring KH, Beals SP. Cranial growth unrestricted during treatment of deformational plagiocephaly. Pediatric Neurosurg 30:193-199, 1999.
18. Clarren SK. Plagiocephaly and torticollis: Etiology, natural history and helmet treatment. J Pediatr 98(1):92-95, 1981.
19. Hellbusch JL, Hellbusch LC, Bruneteau RJ. Active counterpositioning treatment of deformational plagiocephaly. Neb Med J 80:344-349, 1995.
20. Panchal J, Amirshebani H, Gurwitch R, Cook V, Francel P, Neas B, Levine N. Neurodevelopment in children with single-suture craniosynostosis and plagiocephaly without synostosis. Plast Reconstr Surg 108(6):1492-1498, 2001.

21. Boere-Boonekamp MM, Van der Linden-Kuiper LT. Positional preference: Prevalence in infants and follow-up after two years. Pediatrics 107(2):339-343, 2001.
22. Peitsch WK, Keefer CH, LaBrie RA, Mulliken JB. Incidence of cranial asymmetry in healthy newborns. Pediatrics 110(6):1-8, 2002.
23. Littlefield TR, Kelly KM, Pomatto JK, Beals SP. Multiple-birth infants at higher risk for development of deformational plagiocephaly. Pediatrics 103(3):565-569, 1999.
24. Kane AA, Mitchell LE, Craven KP, Marsh JL. Observations on a recent increase in plagiocephaly without synostosis. Pediatrics 33(5):702-704, 1986.
25. Teichgraeber JF, Ault JK, Baumgartner J, Waller A, Messersmith M, Gateno J, Bravenec B, Xia J. Deformational posterior plagiocephaly: Diagnosis and treatment. Cleft Palate Craniofac J 39:582-586, 2002.
26. Hutchison BL, Thompson JMD, Mitchell EA. Determinants of non-synostotic plagiocephaly: A case-control study. Pediatrics 112(4):e316-e322, 2003.
27. Littlefield TR, Pomatto JK, Beals SP, Joganic EF, Manwaring KH, Ripley CE. Efficacy and stability of dynamic cranial orthoplasty: An eight-year investigation. In Whitaker ed. Craniofacial Surgery VII: 109-111, 1997.
28. Kelly KM, Littlefield TR, Pomatto JK, Ripley CE, Beals SP, Joganic EF. The importance of early recognition and treatment of deformational plagiocephaly with orthotic cranioplasty. Cleft Palate Craniofac J 36:127-130, 1999.
29. Golden KA, Beals SP, Littlefield TR, Pomatto JK. Sternocleidomastoid imbalance versus congenital muscular torticollis: Their relationship to positional plagiocephaly. Cleft Palate Craniofac J 36(3):256-261, 1999.
30. State K, Posnick JC, Armstrong D, Buncic JR. Cervical spine subluxation associated with congenital muscular torticollis and craniofacial asymmetry. Plast Reconstr Surg 92(7):1187-1195, 1993.
31. Loveday BPT, De Chalain TB. Active counterpositioning or orthotic device to treat positional plagiocephaly. J Craniofac Surg 12(4):308-313, 2001.
32. Hollier LH, Kim J, Grayson BH, McCarthy JG. Congenital muscular torticollis and the associated craniofacial changes. Plast Reconstr Surg 105(3):827-835, 2000.
33. Jones BM, Hayward R, Evans R, Britto J. Occipital plagiocephaly: An epidemic of craniosynostosis. British Med J 315(7110):693-694, 1997.
34. Ferguson JW. Surgical correction of the facial deformities secondary to untreated congenital muscular torticollis. J Cranio-Maxillo-Facial Surg:137-142, 1993.
35. Ripley CE, Pomatto JK, Beals SP, Joganic EF, Manwaring KH, Moss SD. Treatment of positional plagiocephaly with dynamic orthotic cranioplasty. J Craniofac Surg 5(3):150-159, 1994.
36. Chadduck WM, Kasy J, Donahue DJ. The enigma of lambdoid positional molding. Pediatr Neurosurg 26:304-311, 1997.
37. O'Broin ES, Allcutt D, Earley MJ. Posterior plagiocephaly: Proactive conservative management. Br J Plast Surg 52:18-23, 1999.
38. Miller RI, Clarren SK. Long-term developmental outcomes in patients with deformational plagiocephaly. Pediatrics 105(2):1-5, 2000.
39. Patel S, Panchal J, Shams Z, Ahad W, Gurwitch R, Francel P. Developmental delays in children with developmental plagiocephaly: An updated report. 2004 ACPA Conference.
40. Hansen M, Mulliken JB. Frontal plagiocephaly: Diagnosis and treatment. Clin Plast Surg 21(4):543-553, 1994.
41. Moss SD. Nonsurgical, nonorthotic treatment of occipital plagiocephaly: What is the natural history of the misshapen neonatal head? J Neurosurg 87:667-670, 1997.

42. Vles JSH, Van Zutphen S, Hasaart T, Dassen W, Lodder J. Supine and prone head orientation preference in term infants. Brain Dev 13(2):87-90, 1991.
43. Coulter-O'Berry C, Hylton-Plank L, Lima D, Fish D. Coordinating physical therapy, a parent-infant training program and orthotic management using the STARscanner data acquisition system to document head shape deformity and progress. Unpublished poster exhibit, American Cleft Palate and Craniofacial Association and the Association of Children's Prosthetic and Orthotic Clinics, 2003.
44. Teichgraeber JF, Seymour-Dempsey K, Baumgartner J, Xia J, Waller A, Gateno J. Molding helmet therapy in the treatment of brachycephaly and plagiocephaly. J Craniofac Surg 15(1):118-123, 2004.
45. Pattisapu JV, Walker ML, Myers GG, Cheever J. Use of helmets for positional molding. Concepts Pediatr Neurosurg 9:178-184, 1989.
46. Persing JA, James H, Swanson J, Kattwinkel. Prevention and management of positional skull deformities in infants. Pediatrics 112(1):199-202, 2003.
47. David DJ, Menard RM. Occipital plagiocephaly. Br J Plast Surg 53:367-377, 2000.