CONGRESS OF NEUROLOGICAL SURGEONS SYSTEMATIC REVIEW AND EVIDENCE-BASED GUIDELINES FOR THE MANAGEMENT OF PATIENTS WITH POSITIONAL PLAGIOCEPHALY: INTRODUCTION AND METHODOLOGY

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This clinical systematic review and evidence-based guideline was developed by a physician volunteer task force and serves as an educational tool that reflects the current state of knowledge at the time of completion. The presentations are designed to provide an accurate review of the subject matter covered. This guideline is disseminated with the understanding that

the recommendations by the authors and consultants who have collaborated in its development are not meant to replace the individualized care and treatment advice from a patient's physician(s). If medical advice or assistance is required, the services of a physician should be sought. The proposals contained in this guideline may not be suitable for use in all circumstances. The choice to implement any particular recommendation contained in this guideline must be made by a managing physician in light of the situation in each particular patient and on the basis of existing resources.

ABSTRACT

Background: Positional plagiocephaly is a common problem seen by pediatricians, pediatric neurologists, and pediatric neurosurgeons. Currently, there are no evidence-based guidelines on the management of positional plagiocephaly. The topics addressed in subsequent chapters of this guideline include: diagnosis, repositioning, physical therapy, and orthotic devices.

Objective: To evaluate topics relevant to the diagnosis and management of patients with positional plagiocephaly. The rigorous systematic process in which this guideline was created is presented in this chapter.

Methods: This guideline was prepared by the Plagiocephaly Guideline Task Force, a multidisciplinary team comprised of physician volunteers (clinical experts), medical librarians, and clinical guidelines specialists. The task force conducted a series of systematic literature searches of the National Library of Medicine and the Cochrane Library, according to standard protocols described below, for each topic addressed in subsequent chapters of this guideline.

Results: The systematic literature searches returned 396 abstracts relative to the 4 main topics addressed in this guideline. The results were analyzed and are described in detail in each subsequent chapter included in this guideline.

Conclusion: Evidence-based guidelines for the management of infants with positional plagiocephaly will help practitioners manage this common disorder.

Keywords: Evidence-based medicine; methodology; infants; plagiocephaly, non-synostotic; positional plagiocephaly; practice guideline

INTRODUCTION

Accurate diagnosis and treatment of infants with positional plagiocephaly is important, as it is a common finding seen by multiple pediatric specialties, including general pediatricians, neurosurgeons, neurologists, plastic surgeons, and physical therapists. With almost 4 million births in the United States in 2013, it is estimated that up to 20% of infants experienced some degree of positional deformation.[1] Deformational plagiocephaly is an acquired flattening of the infant head, seen as a result of pressure, usually from laying on that area for periods of time. It is not a result of premature closure of the sutures.

While the treatment of patients with plagiocephaly is non-surgical, neurosurgeons are frequently consulted. Neurosurgeons have 2 primary objectives when evaluating patients with positional plagiocephaly: ruling out craniosynostosis and determining whether the patient requires intervention, such as physical therapy or a molding helmet. Distinguishing patients with positional plagiocephaly from patients with true craniosynostosis is almost always straightforward and does not require imaging. Helmet therapy, as discussed in greater detail in Chapter 4, remains controversial.

It is important to realize that the natural history of positional plagiocephaly is benign. Nevertheless, positional plagiocephaly and craniofacial asymmetry cause considerable anxiety among patients' families and their pediatricians. There is often fear or concern that the infant may have craniosynostosis. Parents may also be concerned about facial asymmetries that are noted by their family members. Certainly, for infants with severe positional plagiocephaly and craniofacial asymmetry, parental concerns about cosmetic outcome and the potential social or psychological effects on their child are valid and very real.

Two reviews on the treatment of positional plagiocephaly had been published prior to the development of this systematic review and evidence-based guideline. Goh and associates conducted a systematic literature review, evaluating 42 articles on the use of helmet orthosis in the treatment of plagiocephaly.[2] In a second systematic review, Shweikeh et al also evaluated existing literature on the prevention and management of plagiocephaly, including 15 relevant articles.[3] Even though the authors of these studies conducted systematic literature reviews and evaluated the existing literature on positional plagiocephaly, updated evidence-based recommendations regarding the prevention and management of plagiocephaly were not provided

and spurred the efforts to develop guidelines specifically for the diagnosis and treatment of this condition frequently seen in infants.

The development of these guidelines was initiated by the Congress of Neurological Surgeons (CNS) and the Section on Pediatric Neurosurgery in response to members' concerns about the variation in the diagnosis and treatment paradigms being utilized. A multidisciplinary team comprised of physician volunteers (clinical experts), a clinical guidelines expert, and medical librarians was convened to conduct a systematic search of the literature and prepare clinical guidelines on the topic of pediatric plagiocephaly. After initial discussions, the members of the Plagiocephaly Guideline Task Force (hereinafter referred to as "the task force") decided, a priori, that the 4 major sub-topics would include: imaging modalities in the diagnosis of plagiocephaly, repositioning, physical therapy, and molding orthoses (helmet therapy).

The task force collaborated with medical librarians to conduct systematic literature searches (see below summary, by topic). As a result of these searches, the task force reviewed a total of 396 abstracts on 4 topics. After abstracts were reviewed by authors in each topic subcommittee, 110 articles were selected for full text review. Sixty were found to have relevant evidence that met the inclusion and exclusion criteria, which are described below. These sources were used to create the 10 guideline recommendations. In the body of each paper, the task force describes the rational for exclusion of articles selected for full-text review that were not included in the evidence tables.

Searches were done using MESH terms of plagiocephaly and brachycephaly for each of the 4 major topics. Posterior plagiocephaly occurs when there is unilateral flattening of the parietooccipital region, resulting in a rhomboid-like shift of the calvarium with an anterior shift of the ipsilateral ear and bulging or bossing of the ipsilateral forehead. The second, less common variant is sometimes called brachycephaly, in which there is flattening of the entire occipital region, resulting in a foreshortened head in the anterior-posterior dimension. However, the term "brachycephaly" is also used in children with craniosynostosis. Henceforth, we will refer to non-synostotic calvarial positional deformity as plagiocephaly.

Review of the abstracts revealed 2 important features. First, results were similar in the searches for both plagiocephaly and brachycephaly for imaging and treatment. Some authors published their series of the 2 problems separately. All authors acknowledged that positional molding was the cause of the 2 conditions. The treatments for the 2 conditions were identical and

included repositioning, physical therapy, and cranial orthotics, such as helmets and bands. Furthermore, abstracts from the searches that included brachycephaly were far more likely to include true craniosynostosis as a result, and, therefore, many of the papers from those searches were automatically excluded. In addition, a recent paper from Meyer-Marcotty et al used cephalic index measurements to discern the overlap between the 2 conditions and declared that they represent a continuum.[4] As a result, for the purposes of this set of guidelines, the evidence for brachycephaly was incorporated into the positional plagiocephaly guidelines. The term "plagiocephaly," in these guidelines inclusively, is used to describe positional skull deformity causing asymmetry (plagiocephaly) and brachycephaly (occipital flattening).

METHODS

Potential Conflicts of Interest

All guideline task force members were required to disclose all potential conflicts of interest (COIs) prior to beginning work on the guideline, using the COI disclosure form of the Joint Guidelines Committee of the American Association of Neurological Surgeons (AANS) and the CNS (hereinafter referred to as the Joint Guidelines Committee). The CNS Guidelines Committee and the task force chair reviewed any disclosures and either approved or disapproved the nomination and participation on the task force. The CNS Guidelines Committee and the task force chair may approve nominations of task force members with possible conflicts and restrict the writing, reviewing, and/or voting privileges of that person to topics that are unrelated to the possible COIs.

Literature Search

The task force worked with medical librarians to determine appropriate search terms and to create search strategies for each guideline chapter. The National Library of Medicine and the Cochrane Library were searched for literature published between 1966 and October 2014. Task force members used the article inclusion/exclusion criteria described below to screen abstracts and provide a list of relevant articles for full-text review. Task force members were blinded to the selection of abstracts provided by other task force members. CNS staff compiled lists of manuscripts for full-text review and approval by all of the task force members, and these full-text articles were review by all task force members. In addition, task force members also screened the

bibliographies of relevant systematic reviews for potentially relevant articles. The topic-specific search strategies can be found within the appendices of each chapter.

Article Inclusion Criteria

Included articles must have met certain criteria, as detailed below. To reduce bias, these criteria were specified before conducting the literature searches. To be included in our review, an article had to meet the following criteria:

- Studies had to investigate pediatric (<18 years of age) patients with non-synostotic plagiocephaly or brachycephaly.
- Studies with mixed patient populations and that combined the results of these patient groups must have enrolled ≥80% of pediatric patients with plagiocephaly or brachycephaly.
- The study was a full article report of a clinical study.
- Studies had to have appeared in a peer-reviewed publication or a registry report.
- Studies had to enroll at least 10 patients (5 per treatment arm) for each distinct outcome measured. If it was a comparative study, a minimum enrollment of 5 patients per treatment arm for each outcome was necessary.
- The study involved humans.
- The study was published in or after 1966.
- The study presented results quantitatively.
- The study did not involve "in vitro," "biomechanical," or results performed on cadavers.
- The study was published in English.

Systematic reviews, meta-analyses, or guidelines developed by others were not considered as evidence to support this guideline. The task force screened the bibliographies of these publications to ensure the accuracy and comprehensiveness of the literature search results used for this guideline.

Rating Quality of Evidence

The quality of evidence was rated using an evidence hierarchy developed by the Joint Guidelines Committee for each of the 4 different study types (ie, therapeutic, diagnostic, prognostic, and clinical assessment). Additional information regarding the hierarchy

classification of evidence can be located here: https://www.cns.org/guidelines/guideline-procedures-policies/guideline-development-methodology.

Strength of Recommendations Rating Scheme

The task force used the methodologies endorsed by the Joint Guidelines Committee (JGC) to assign a strength of recommendation for each recommendation included in this guideline. Linking evidence to recommendations, through the utilization of evidentiary tables, has been endorsed by the American Medical Association (AMA), the CNS, and the AANS. This process validates and supports the relationship between the strength of evidence and the strength of recommendations.

Demonstrating the highest degree of clinical certainty, Class I evidence is used to support recommendations of the strongest type, defined as Level I recommendations. Level II recommendations reflect a moderate degree of clinical certainty and are supported by Class II evidence or strong consensus of Class III evidence. Level III recommendations denote clinical uncertainty supported by inconclusive or conflicting evidence or expert opinion.

Voting on the Recommendations

The task force used voting among its members to approve the final recommendations, language, and strength of recommendations. The voting was used to ensure that the language of each recommendation accurately reflected the evidence and the strength of the evidence. All the recommendations in this review were approved following the first round of voting, and no further discussion was needed to finalize the recommendations described below. The voting technique is referred to as the nominal group technique, as described in an article by Murphy et al.[5] During the course of editing and finalization of the document, changes were made to allow recommendations to conform to the rules of evidence and language as described above. When this occurred, the changes were reviewed and approved by the group.

Guideline Panel Consensus and Approval Process

Topic teams were created from the task force based on expertise of the task force members with respect to each topic addressed within the review. Each group took part in literature selection, review of the literature, creation of the evidence tables, creation of the

guideline, editing, and final review. The final draft of the guideline was then circulated to the entire task force for feedback, discussion, and ultimately approval.

Following task force approval, drafts of the completed guidelines were presented to the JGC for peer review and, ultimately, recommendation of endorsement by the CNS and the AANS. The reviewers for the JGC were vetted by the editorial staff of the journal *Neurosurgery*. During the review process, the peer reviewers were blinded to the identities of the task force members. As part of the evaluation process, reviewers could provide input on the content and the methodologies used to create the systematic review. Development of this systematic review and set of guidelines was editorially independent of the funding agencies (CNS and the Section on Pediatric Neurosurgery). See Figure 1 for an outline of the key steps in the process of developing this systematic review and set of evidence-based guidelines.

Revision Plans

In accordance with the Institute of Medicine's standards for developing clinical practice guidelines and criteria specified by the National Guideline Clearinghouse, the task force will monitor related publications following the release of this document and will revise the entire document and/or specific sections "if new evidence shows that a recommended intervention causes previously unknown substantial harm; that a new intervention is significantly superior to a previously recommended intervention from an efficacy or harms perspective; or that a recommendation can be applied to new populations."[6] In addition, the task force will confirm within 5 years from the date of publication that the content reflects current clinical practice and the available technologies for the treatment of pediatric positional plagiocephaly.

CONCLUSION

Evidence-based guidelines for the treatment of positional plagiocephaly are necessary and important to deal with this common disorder. Systematic literature searches were conducted according to standard protocols, identifying a total of 396 abstracts on 4 topics relevant to positional plagiocephaly. After the topic sub-groups reviewed these abstracts, the task force groups selected articles for full text review to create the evidence tables. The entire task force reviewed the full text articles and evidence tables to ensure uniformity of criteria and classification throughout the document. Sufficient evidence was found to create 11 recommendations, 3 of which were Level I.

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Disclosures

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Figure 1 Overview of Guideline Development Process