Congress of Neurological Surgeons

GUIDELINES

CONGRESS OF NEUROLOGICAL SURGEONS SYSTEMATIC REVIEW AND EVIDENCE-BASED GUIDELINE FOR THE DIAGNOSIS OF PATIENTS WITH POSITIONAL PLAGIOCEPHALY: THE ROLE OF IMAGING

Sponsored by

Congress of Neurological Surgeons (CNS) and the Section on Pediatric Neurosurgery

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Joint Guidelines Committee of the American Association of Neurological Surgeons (AANS) and the Congress of Neurological Surgeons (CNS) and American Academy of Pediatrics (AAP)

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This clinical systematic review and evidence-based guideline was developed by a physician volunteer task force 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 recommendations 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: No evidence-based guidelines exist for imaging of patients with positional plagiocephaly.

Objective: To answer the question: Is imaging necessary for infants with positional plagiocephaly in order to make a diagnosis?

Methods: The National Library of Medicine Medline database and the Cochrane Library were queried using MeSH headings and keywords relevant to imaging as a means to diagnose plagiocephaly. Abstracts were reviewed and an evidentiary table was assembled summarizing the studies and the quality of evidence (Classes I-III). Based on the quality of the literature, a recommendation was rendered (Level I, II, or III).

Results: A total of 42 full-text articles selected for review. Of these, 10 were eliminated. There were 32 full-text manuscripts selected. There was no Class I evidence, but 2 Class II and 30 Class III studies were included. Three-dimensional cranial topographical imaging, ultrasound, skull x-rays, computed tomography, and magnetic resonance imaging were investigated.

Conclusion: Clinical examination is most often sufficient to diagnose plagiocephaly (quality, Class III; strength, Level III). Within limits of this systematic review, the evidence suggests that imaging is rarely necessary and should be reserved for cases where the clinical examination is equivocal. Many of the imaging studies were not designed to address the diagnostic utility of the imaging modality, and authors were actually assessing the utility of the imaging in longitudinal follow-up, not initial diagnosis. For this reason, some of the studies reviewed were downgraded in Level of Evidence. When needed, 3-dimensional cranial topographical photo, skull x-rays, or ultrasound imaging is almost always sufficient for definitive diagnosis. Computed tomography scanning should not be used to diagnose plagiocephaly, but it may be necessary to rule out craniosynostosis.

Short Title: Guideline for the Diagnosis of Patients with Positional Plagiocephaly: The Role of Imaging

Key Words:

imaging; infants; diagnosis; plagiocephaly, non-synostotic; positional plagiocephaly;

RECOMMENDATIONS

1. Clinical examination is recommended for the diagnosis of plagiocephaly and imaging is rarely necessary, except in cases in which clinical diagnosis is equivocal.

Strength of recommendation: Level III—low clinical certainty

2. In cases in which the clinical examination is equivocal, skull x-rays or ultrasound imaging of the suspect suture is recommended.

Strength of recommendation: Level II—moderate clinical certainty

3. In cases in which the clinical examination is equivocal, surface imaging (computer-based topographical scans) or stereophotogrammetry is recommended for the assessment of infants with plagiocephaly without synostosis.

Strength of recommendation: Level III—low clinical certainty

4. Only for infants in whom x-rays or ultrasound are non-diagnostic, a CT scan is recommended for definitive diagnosis.

Strength of recommendation: Level III—low clinical certainty

INTRODUCTION

Infants may present with abnormal head shape any time after birth. The reason for referral is commonly "to rule out craniosynostosis." The diagnosis of true craniosynostosis is important because this condition is amenable to surgical correction, whereas positional, posterior plagiocephaly without synostosis (PWS) is adequately treated with repositioning, physical therapy, or, in moderate to severe cases, a cranial molding helmet.[1,2]

It has been the experience of many craniofacial specialists, including those on the Plagiocephaly Task Force (hereinafter referred to as the "task force"), that most infants with plagiocephaly can be adequately diagnosed through a detailed clinical examination. Three dimensional (3-D) topographical scanning may be useful for diagnosis and baseline assessment of severity. In those rare cases in which the clinical examination was equivocal, skull x-rays or an ultrasound of the suture in question could be used to rule out craniosynostosis. Only if those

radiological studies are equivocal, should a computed tomography (CT) scan of the head be performed.

METHODS

The Congress of Neurological Surgeons (CNS) and the Section on Pediatric Neurosurgery initiated a systematic review of the literature and evidence-based guideline relevant to the management of positional plagiocephaly. Additional information about the methods utilized in this systematic review is provided below and within the introduction and methodology chapter of the guideline.

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. 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 guideline 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 members collaborated with medical librarians to search the National Library of Medicine/PubMed database and the Cochrane Library for the period from 1966 to October 2014 using the MeSH subject headings and PubMed search strategies provided in Appendix A. Manual searches of bibliographies were also conducted.

Our searches resulted in 204 abstracts. The task force selected 42 full-text articles for review. Of these, 10 were rejected for not meeting inclusion criteria or for being off-topic (Figure 1). For example, some studies were not performed on human subjects, not limited to infants, or were mainly reporting study results of infants with craniosynostosis. Thirty-two articles were selected for systematic review (Table 2).

Rating Quality of Diagnostic Evidence

For diagnostic-type papers, evidence classification had definitions targeted toward diagnosis. The issues addressed by papers on diagnosis are related to the ability of the diagnostic test to successfully distinguish between patients who have and do not have a disease or pertinent

finding. This speaks to the validity of the test and is illustrated in Table 1. Additional information regarding the hierarchy classification of evidence can be located here: https://www.cns.org/guidelines/guideline-procedures-policies/guideline-development-methodology.

Many of the imaging studies were not designed to address the diagnostic utility of the imaging modality, and authors were actually assessing the utility of the imaging in longitudinal follow-up, not initial diagnosis. For this reason, some of the studies reviewed were downgraded in Level of Evidence.

DISCUSSION

Clinical examination is recommended for the diagnosis of plagiocephaly and imaging is rarely necessary, except in cases in which clinical diagnosis is equivocal. Within the limits of this systematic review, we found that imaging is rarely necessary and should be reserved for when the clinical examination is equivocal. In a Class III, 2013 study done by Kuang and Bergquist, the effectiveness and safety of pediatric nurse practitioners for the diagnosis of infants with plagiocephaly was evaluated.[3] They completed a retrospective review of the electronic medical records of 1228 infants seen in a craniofacial clinic from 2005 to 2011. The authors concluded that a nurse practitioner was able to effectively and safely diagnose and treat plagiocephaly without the need of imaging.[3] However, in their study, it was the surgical team who ruled out craniosynostosis by clinical examination alone in 325 of the 590 infants that the nurse practitioners believed to have craniosynostosis.[3] It was the craniofacial surgeons who made the correct diagnosis of plagiocephaly in most of the infants. The nurse practitioners in the Kuang study each had 12-20 years of prior experience working with the pediatric neurosurgeon on the craniofacial team.

In another report by Linz, 269 infants with abnormal head shape were evaluated clinically, resulting in 258 diagnosed with deformational plagiocephaly, and 8 were “definitely” diagnosed with craniosynostosis; in 3 infants, the diagnosis of plagiocephaly was suspected but could not be made definitively.[4] All 269 had cranial ultrasounds and 8 had x-rays of the skull performed. Of the 261 infants with plagiocephaly, in 258 the correct diagnosis was made by clinical examination alone.[4] Of the 3 infants whose diagnosis was uncertain and clinical examination could not definitely rule out craniosynostosis, ultrasounds did confirm open sutures. Skull x-rays were ordered for 8 infants suspected of having craniosynostosis, and in those 8

infants, craniosynostosis was confirmed on x-ray.[4] This protocol, whereby skull x-rays were ordered only in equivocal cases, was followed by many authors.[4-6]

There are well-described cranial shape differences between infants with PWS and craniosynostosis of the lambdoid suture.[3,4,7] From the posterior coronal view, infants with PWS had ipsilateral occipital-parietal flattening and contralateral occipital prominence but an essentially “normal” head shape from behind. However, in infants with true unilateral lambdoid synostosis, skull growth persists from either end of the fused suture, leading to ipsilateral occipital-mastoid bossing and contralateral parietal bossing, forming a diamond shape from behind that is wider posteriorly than anteriorly, with skull base tilt.[4,7] From the aerial vertex view, in PWS there is posterior flattening with ipsilateral frontal prominence leading to an aerial parallelogram-shaped head. In patients with true unilateral lambdoid synostosis, frontal bossing is contralateral instead, though sometimes minimally prominent, which, when combined with occipital flattening on the synostotic side, forms an aerial-view trapezoid with ipsilateral ear pulled posteriorly.[3,4,7]

In a Class III, retrospective analysis of 287 infants seen at their craniofacial clinic, Hutchison et al reported that only 2% (n = 7) were referred for CT scanning and that clinical examination and head shape measurements were usually sufficient for diagnosis.[8] Of those 7 infants in whom the diagnosis could not be made on clinical examination alone, 4 had confirmed craniosynostosis, 1 had an abnormal "extra" suture, and 2 had severe positional plagiocephaly without true craniosynostosis. In the Mulliken et al study, only 11/115 infants required CT scans.[5] In most studies, CT scanning is not indicated for all infants with plagiocephaly, only those in whom the clinical diagnosis is not apparent and skull x-rays or other imaging is equivocal or non-diagnostic.[5,6,8]

Anthropometric measurements have been used as part of the clinical examination in several studies.[6,9-11] In 1 Class III study, the repeatability and reliability of intra-rater measurements were found to be +/- 1 mm.[9] A total of 14 measurements were recorded using spreading and sliding calipers and a linen measuring tape, at the initial assessment and at various intervals thereafter. Intra-rater and inter-rater caliper-based anthropometric measurement reliability was compared to measurements made from stereophotographs and were found to be accurate. There was low variability in the variance component analysis in the study done by Shaaf.[10]

3D Surface Imaging and/or Stereophotogrammetry

In cases in which the clinical examination is equivocal, surface imaging (computer based tomography scans) or stereophotogrammetry is recommended for the assessment of infants with plagiocephaly without synostosis.

In 2012, there was a prospective, non-randomized study done by Collett et al that utilized clinical assessment and cranial topographical scanning to follow infants with and without PWS.[12] Even though the study was a prospective design, the lack of blinded or randomized comparison between the utility of topographical scanning versus independent clinical examination made this Class III data. Three-dimensional (3D) topographical scanning is not performed in the radiology department but is often done in the clinic or practice setting and does not expose the infant to radiation. Topographical scanning, as mentioned in several studies, usually involves a light source and recorder that utilizes data obtained from the reflection of light at various angles from the infant's head to construct a computer-generated 3D image.[12-18] There is no exposure to ionizing radiation. Although the Collett and Heike study demonstrated improvement in symmetries, there was no blinded or randomized comparison between the utility of topographical scanning versus independent clinical examination, and for this reason we downgraded this to Class III evidence.[12] This was true in the Katzel, Thompson, and Moghaddam studies as well, making the overall recommendation Level III.[13,17,18] Most authors found that 3D topogrammetry provided better objectivity, but often the significance, sensitivity, and specificities for diagnosis were not reported or compared to independent clinical observation for various reasons.

In a Class III study by Kluba et al, clinical examination and 3D stereophotogrammetry demonstrated equipoise in assessment of head shape asymmetry, but 3D topographical scanning was superior in the assessment of "ear shift" seen in positional plagiocephaly.[14] Stereophotogrammetry involves multiple camera systems that provide 3D imagery.[14,16,19] Three-dimensional cranial topography of infants with severe positional plagiocephaly was performed before and after cranial molding therapy.[10,14,16,19] The cranial vault asymmetry index (CVAI) and ear shift were measured and statistically compared.[14] Three craniofacial surgeons also evaluated the 3D stereophotogrammetry results independently and reported their assessment with a standard questionnaire. The results were compared with the three-dimensional, anthropometric measurements, and there existed a good correlation.[10,14] Other authors have utilized 3D topographical scanning and stereophotogrammetry for the analysis of initial head shape and the

clinical follow-up of infants with plagiocephaly, as well.[15,16,19] Other groups have used digital photography to record, evaluate, and follow infants with plagiocephaly.[11]

Skull X-ray or Ultrasound

In cases in which the clinical examination is equivocal, a skull x-ray or ultrasound imaging of the suspect suture is recommended.

When the clinical examination is equivocal, skull x-rays or ultrasound imaging is recommended for definitive diagnosis.[20-23] In well-done Class II studies with prospective clinical comparisons, Krimmel et al and Sze et al evaluated the value of high-resolution ultrasound (US) in the differential diagnosis of scaphocephaly and occipital plagiocephaly.[20,23] In Krimmel's study, radiological data from 54 infants under 12 months of age with plagiocephaly were presented. The 2 comparison groups were 47 infants with simple positional plagiocephaly and 7 infants with true craniosynostosis. Under the premise that the inconclusive US findings are regarded as false-positive and false-negative results, the US method had at least a sensitivity of 71.4% (95% confidence interval: 35.5%, 100%), a specificity of 95.7%, a positive predictive value of 71.4%, and a negative predictive value of 95.7%.[20] In 96% (45/47) of infants with positional plagiocephaly, the ultrasound images did confirm patent sutures. In 4% (2/47) of the infants studied, the ultrasound was inconclusive. In 5 out of 7 infants with craniosynostosis, the ultrasound did show closed cranial sutures. In only 2 of the 7 infants with craniosynostosis, the ultrasound findings were inconclusive and were followed by a CT in 1 patient and a skull x-ray in the other. In conclusion, the authors felt that ultrasound effectively distinguished between open and closed sutures.[20] Sze et al reported in 2003 that ultrasonography of the lambdoid sutures shows "excellent promise as a screening test of lambdoid sutural patency."[23] Their group prospectively evaluated ultrasound as a screening test of suture patency using CT as the gold standard. The reported mean sensitivity and specificity of cranial ultrasound in distinguishing a patent from fused suture by 3 blinded pediatric radiologists was 100% and 89%, respectively. This was a Class II study used to support ultrasound as a Level II recommendation as a study that can be used for infants in whom clinical diagnosis is uncertain. In a prospective Class III study done by Linz et al, 269 infants with plagiocephaly without synostosis (PWS, n = 261) and 8 infants with lambdoid synostosis were clinically examined to outline the specific clinical features of true positional deformity vs craniosynostosis.[4] After clinical examination, cranial ultrasound of the sutures was performed. Ultrasound revealed either a lambdoid synostosis or a patent

lambdoid suture in cases of PWS. In 96% (258/269) of PWS infants, clinical examination was able to confirm the diagnosis. Of note, in 3 infants who were initially diagnosed with PWS, a lambdoid synostosis was found on ultrasound. In all true lambdoid synostosis cases, ultrasound did support the clinical diagnosis. Their conclusion was that CT scan was not necessary in the diagnosis of true lambdoid synostosis and that there are quite distinctive clinical features apparent in infants with lambdoid synostosis when compared to infants with PWS. Additionally, the group concluded that ultrasonography done in infants ≤12 months can be used to confirm the diagnosis of synostosis.[4] These findings have been supported by additional studies.[22]

Many authors followed protocols whereby infants with plagiocephaly were evaluated with skull x-rays, and only if the skull x-rays were non-diagnostic would CT scans be performed.[6,21,24] In the study by David et al, skull x-rays done for 204 infants with posterior plagiocephaly showed 202 infants with patent sutures. Less than 1% (2/204) of the infants were found to have true synostosis. The authors concluded that clinical examination and skull x-rays were sufficient for diagnosis, and CT imaging should be reserved for only those patients in whom both the clinical exam and radiological skull x-ray results are equivocal.[24]

Computed Tomography

Only for infants in whom x-rays or ultrasound is non-diagnostic, a computed tomography scan is recommended for definitive diagnosis.

It is our opinion that rarely is CT ever needed for the diagnosis of plagiocephaly or to rule out craniosynostosis. Furthermore, children, particularly infants, should be spared unnecessary radiation. In many clinical studies, the utility of CT has been investigated in relationship to the differential diagnosis of PWS versus craniosynostosis.[5,8,21,22,25-27] In a Class III study conducted by Fisher et al, the authors pointed out that only in equivocal or severe cases of skull deformity could confusion with true synostosis arise, and the diagnosis must be confirmed to correctly identify infants with true craniosynostosis.[25] The diagnosis can be determined by clinical examination of the head shape and, only if absolutely necessary, established with a CT scan.[25] However, for purposes of our study, the study by Fisher et al was Class III study supporting a Level III recommendation because, as the authors pointed out, the purpose of their study was 2-fold: (1) to evaluate the incidence of true metopic synostosis in infants with plagiocephaly and (2) to examine the morphology of the metopic suture, forehead,

and interorbital distance among the positional plagiocephaly group and to compare it with the morphologies of the suture and forehead and interorbital distances of infants with classic metopic synostosis and trigonocephaly. Additionally, there was no comparison of CT findings to clinical observations or physical examination, or even skull x-rays as diagnostic modalities. Losee et al published a Class III study evaluating CT scanning as a modality to diagnose lambdoid synostosis.[27] Thirty-three infants with posterior plagiocephaly were imaged; 26 did not have craniosynostosis, and 7 infants did have lambdoid synostosis diagnosed after 10 suture characteristics were evaluated.[27] Although this was a well-done study, it was a Class III study because there was no comparison of imaging modalities or comparison to clinical examination alone.

In another Class III retrospective review of 204 patients with occipital plagiocephaly, all of whom had clinical examination and skull x-rays, 2 CT scans were done in cases in which the diagnosis was in question.[24] In a Class III retrospective analysis of 287 infants seen at their craniofacial clinic, Hutchison et al reported that only 2% (n = 7) were referred for CT scanning and that clinical examination and head shape measurements were usually sufficient for diagnosis.[8] In a similar Class III retrospective analysis of 287 infants seen at their craniofacial clinic, Hutchison et al reported that only 2% of their patients (n = 7) were referred for CT scanning and that clinical examination and head shape measurements were usually sufficient for diagnosis.[8] Of those 7 infants in whom the diagnosis could not be made on clinical examination alone, 4 had confirmed craniosynostosis, 1 had an abnormal “extra” suture, and 2 had severe positional plagiocephaly without true craniosynostosis. In the authors’ opinion, CT scanning is not indicated for all infants with plagiocephaly, only those in whom the skull deformation is severe and the suture is “ridged.”[8]

While CT scans are not necessary in most infants with plagiocephaly, CT scanning may be useful for differential diagnosis in difficult or equivocal cases.[5,21,22,26,28,29] In a Class III retrospective review of 102 CT scans performed on infants with skull asymmetry, of whom 82 had deformational plagiocephaly and 20 had craniosynostosis, skull base asymmetries were evaluated and compared.[29] Although there were obvious and different asymmetries recorded, the value of these measurements over a simple clinical examination was not established, and there was no consideration of radiation exposure to these children.[29] Similar findings were published

by Lo et al and Netherway et al[26, 30] CT scans of infants with plagiocephaly and craniosynostosis did show differences in external perimeter analysis, cranial fossae symmetries, and cranial midline angulation; however, in most of these studies, the absolute needs for CT imaging for diagnosis and comparison to expert clinical examination were not done.[26,30] Differences in 3D anatomy that are made obvious on imaging studies have allowed clinicians to follow improvement in cranial symmetry over time. Although some studies have documented better symmetry in head shape in infants with PWS treated with cranial orthotics, the absolute need for 3D CT imaging for "follow-up" was never demonstrated.[2,30] Mulliken et al reported that only 11/115 infants required CT scans.[5] In most studies, CT scanning is not indicated for all infants with plagiocephaly, only those in whom the clinical diagnosis is not apparent, and skull x-rays or other imaging is equivocal or non-diagnostic.

In a Class III, retrospective comparative study of 202 infants, 66 with PWS and CT data, Abbott et al demonstrated that intracranial volumes are similar to the intracranial volumes of "normal" infants.[1] Intracranial volume calculated for the 66 infants with deformational plagiocephaly was not statistically different compared to "normal," age-matched infants.[1] This information was validated in another study by Bruner et al, who prospectively studied 3D CT data and calculated intracranial volume in infants with PWS treated with cranial molding therapy.[2] In several studies, 3D scans were found "useful" for evaluating the results of helmet therapy for infants with deformational plagiocephaly.[2,30] However, the purpose of the Bruner study was to use 3D CT to assess the efficacy of helmet therapy. Their analysis was not undertaken in order to compare clinical examination for diagnosis to CT imaging, and so, for our purposes, we did downgrade the Bruner study to Class III evidence. Criticisms of the study include that (1) no mention was made of radiation dose exposure to infants, (2) only 34/69 infants completed the study, and (3) no comparison was made to initial or clinical assessment of the severity of deformity/plagiocephaly. Although other studies have also demonstrated that 3D CT scanning does show differences in craniofacial skeletal development in infants with plagiocephaly or unilateral coronal synostosis as compared to normal infants, this does not prove the need for CT scanning.[28,30,31] In fact, in most of these studies, there was no consideration or discussion of radiation dose or exposure, and there was no evidence to suggest that CT scans

were necessary for diagnosis. Risk of radiation exposure in infancy is of obvious concern, and unnecessary CT scans should be avoided.

Magnetic Resonance Imaging

The utility of magnetic resonance imaging (MRI) for plagiocephaly has also been investigated.[32] No recommendation can be made for MRI, however, because there was only a single study, and the study did not assess the true need for MRI in the diagnosis of craniosynostosis.

In a Class III, prospective, non-randomized study, Collett and Aylward assessed brain volume and shape in infants with deformational plagiocephaly.[32] The authors compared 2 cohorts of infants: 20 infants with deformational plagiocephaly and 21 "normal" infants. This was a magnetic resonance imaging (MRI) study of various intracranial measurements and volumes in infants with or without deformational plagiocephaly. The authors concluded that: (1) the shape of the brain is affected or controlled by the skull shape and (2) degree of asymmetry is associated with neurodevelopmental outcomes. Criticisms include that the authors were unable to establish whether plagiocephaly develops because of developmental delays or vice versa. Additionally, although there were 78 infants with plagiocephaly identified, only 50 consented to the study, and only 30 MRI studies were attempted, and of those, only 20 completed the MRI study successfully. The cost and time involved in obtaining the MRI studies were not calculated, and there was no comparison between clinical assessment of head asymmetry and findings on MRI.[32]

CONCLUSION

Within the limits of this systematic review, we found that imaging is unnecessary as a first step in infants with plagiocephaly and should be reserved for when the clinical examination is equivocal. In these situations, skull x-rays or ultrasound are almost always sufficient for definitive diagnosis. CT is the gold-standard but should be used sparingly, always making sure the benefit of making a diagnosis is worth the radiation exposure. MRI plays no role.

Recommendations

1. Clinical examination is recommended for the diagnosis of plagiocephaly and imaging is rarely necessary, except in cases in which clinical diagnosis is equivocal.
   Strength of recommendation: Level III—low clinical certainty

2. In cases in which the clinical examination is equivocal, skull x-rays or ultrasound imaging of the suspect suture is recommended.

Strength of recommendation: Level II—moderate clinical certainty

3. In cases in which the clinical examination is equivocal, surface imaging (computer-based topographical scans) or stereophotogrammetry is recommended for the assessment of infants with plagiocephaly without synostosis.

Strength of recommendation: Level III—low clinical certainty

4. Only for infants in whom x-rays or ultrasound are non-diagnostic, a CT scan is recommended for definitive diagnosis.

Strength of recommendation: Level III—low clinical certainty

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TABLES

Table 1: Classification of Evidence on Diagnosis

Class I Evidence Level I (or A) Recommendation	Evidence provided by 1 or more well-designed clinical studies of a diverse population using a "gold standard" reference test in a blinded evaluation appropriate for the diagnostic applications and enabling the assessment of sensitivity, specificity, positive and negative predictive values, and, when applicable, likelihood ratios.
Class II Evidence Level II (or B) Recommendation	Evidence provided by 1 or more well-designed clinical studies of a restricted population using a "gold standard" reference test in a blinded evaluation appropriate for the diagnostic applications and enabling the assessment of sensitivity, specificity, positive and negative predictive values, and, when applicable, likelihood ratios.
Class III Evidence Level III (or C) Recommendation	Evidence provided by expert opinion or studies that do not meet the criteria for the delineation of sensitivity, specificity, positive and negative predictive values, and, when applicable, likelihood ratios.

Table 2: Evidentiary Table

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Abbott AH, Netherway DJ, et al (1998)	Computer Tomography Determined Intracranial Volume of Infants With Deformational Plagiocephaly: A Useful "Normal"?	Retrospective review of 202 infants with deformational plagiocephaly. An intracranial volume (ICV) was calculated for 66 infants based on data available from their computed tomography (CT) scans.	Class III—Retrospective review. No delineation of sensitivity, specificity, positive or negative predictive values.	Intracranial volume calculated through CT imaging for the 66 infants with deformational plagiocephaly was not statistically different compared to "normal," age-matched infants from previously reported studies. Although CT scan may have some role in imaging, calculation of ICV was not useful.
Bruner TW, David LR, et al (2004)	Objective Outcome Analysis of Soft Shell Helmet Therapy in the Treatment of Deformational Plagiocephaly	Prospective, non-randomized, single arm study evaluating the results of helmet therapy for infants with deformational plagiocephaly. Three-dimensional (3D) reconstructed CT scans were used to calculate ICV. Pre-treatment CT scans and CT scans obtained 6 months after initiation of treatment with a soft shell helmet were compared.	Class III—Prospective, single cohort. No delineation of sensitivity, specificity, positive or negative predictive values.	This group concluded that 3D CT scans are a useful method of comparing head shapes before and after therapy. Criticisms include that (1) no mention was made of radiation dose exposure to infants, (2) only 34/69 infants completed the study, and (3) no comparison was made to initial or clinical assessment of the severity of deformity/plagiocephaly.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Captier GN, Leboucq, et al (2003)	Plagiocephaly: morphometry of skull base asymmetry	Retrospective review of 102 CT scans performed on infants with skull asymmetry, of which 82 had deformational plagiocephaly. Twenty had craniosynostosis. Skull base asymmetries were evaluated and compared.	Class III—Retrospective review of CT scans.	Both groups had skull base asymmetries as characterized by CT scanning. Criticisms: (1) Again, no mention made of radiation exposure. (2) There was no comparison of this methodology to clinical assessment, ie did CT scanning add anything to the diagnosis?

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Collett BR, Heike CL, et al (2012)	Longitudinal, Three-Dimensional Analysis of Head Shape in Children With and Without Deformational Plagiocephaly or Brachycephaly	Prospective, non-randomized, study of 3 cohorts of infants with clinically diagnosed plagiocephaly or brachycephaly, non-diagnosed "normal head-shaped" infants, and non-diagnosed infants with asymmetries appreciated only after 3D topographical imaging. Imaging and measurements were done at enrollment and at 18 months. All groups demonstrated improvement in asymmetries.	Class III—Prospective, non-randomized without blinded or randomized comparison. Adjusted z-score differences in head shape at different times during treatment or observation were reported. 3D surface scans of children with diagnosed plagiocephaly or brachycephaly, confirmed on 3D topographical imaging (all cases); children with brachycephaly, but not plagiocephaly (isolated brachycephaly); children without diagnosed plagiocephaly or brachycephaly, with some dysmorphology (affected controls); and children without previously diagnosed plagiocephaly or brachycephaly and confirmed absence of cranial dysmorphology on 3D imaging (unaffected controls) were evaluated and significance and CI values were reported.	All groups demonstrated improvement in head shape as measured with 3D topographical imaging. Infants treated with cranial molding orthotics demonstrated better improvement than those who did not receive molding therapy. 3D topographical imaging provided a useful tool for actualized and documenting improvement in cranial asymmetries. Criticism: there was no comparison between clinical assessment of severity and actual measurements, so could improvement have been realized without 3D topography?

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Collett BR, Aylward EH, et al (2012)	Brain volume and shape in infants with deformational plagiocephaly	Retrospective, non-randomized, cohort comparison of 20 infants with deformational plagiocephaly and 21 without.	Class III—Prospective, non-randomized. This was a magnetic resonance imaging (MRI) study of various intracranial measurements and volumes of infants with or without deformational plagiocephaly. Adjusted group differences in brain shape for children with confirmed deformational plagiocephaly (DP; cases) versus children with confirmed absence of DP (unaffected controls) were reported with confidence intervals and significance of measurements.	The authors concluded that: (1) the shape of the brain as noted through MRI is affected or controlled by the skull shape, and (2) degree of asymmetry is associated with neurodevelopmental outcomes. Criticisms include that although there were 78 infants with plagiocephaly identified, only 50 consented to the study, and only 30 MRI studies were attempted and 20 completed successfully. The cost and time involved in obtaining the MRI studies were not calculated, and there was no comparison between clinical assessment of head asymmetry and findings on MRI.
David DJ and Menard RM	Occipital plagiocephaly	Retrospective review of 204 patients with occipital plagiocephaly, all of whom had skull x-rays and only 2 had true synostosis. CT scans were done in cases in which the diagnosis was in question.	Class III—Evidence from a single center experience.	The authors concluded that clinical examination and skull x-rays were sufficient for diagnosis, and CT imaging should be reserved for only those patients in which both the clinical exam and radiological skull x-rays results are equivocal.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Fisher DC, Kornrumpf BP, et al (2011)	Increased Incidence of Metopic Suture Abnormalities in Children with Positional Plagiocephaly	Retrospective review of the CT scans of 2 groups of patients: those with deformational plagiocephaly and those with metopic synostosis. Of the 4754 patients diagnosed with DP over a 10-year period, 291 had a CT scan performed. There were 41 infants treated for metopic synostosis.	Class III—Retrospective review of 2 cohorts of CT scans; 1 group of infants with DP and 1 group with metopic synostosis. Anterior-interorbital, lateral-orbital, lateral-temporal, and mid-orbital distance measurements of infants with positional plagiocephaly were reported and compared to normal and to infants with true metopic synostosis. Standard deviations and confidence intervals were calculated.	The authors found that there was a high degree of metopic suture abnormalities associated with DP but that these abnormalities do not cause trigonocephaly, which is typically seen in true metopic synostosis. Criticisms of this manuscript include that there was no comparison of clinical assessment with CT based findings and actual measurements. While CT scans were useful in documenting the degree of asymmetries, there was no comparison to clinically based assessment of severity.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Huang MH, Gruss JS, et al (1996)	The Differential Diagnosis of Posterior Plagiocephaly: True Lambdoid Synostosis versus Positional Molding	Retrospective review of 102 infants seen from 1991 to 1994 in a large multi-disciplinary center at the Children's Hospital and Medical Center in Seattle Washington with posterior plagiocephaly.	Class III—Retrospective review; single cohort.	Of the 102 infants, 98 were diagnosed with positional plagiocephaly based on clinical examination. Only 4 were deemed to have craniosynostosis. There were 4 criteria described for the clinical diagnosis of plagiocephaly. CT scans were done only "when indicated;" it is not clear how many did receive CT scans.
Hutchison BL, Stewart AW, et al (2009)	Characteristics, head shape measurements and developmental delay in 287 consecutive infants attending a plagiocephaly clinic	Retrospective review of all children who attended the craniofacial clinic of the department of Pediatrics, the University of Auckland, New Zealand, from May 2005 to August 2007.	Class III—Retrospective review of 287 consecutive infants with plagiocephaly.	Of the 287 infants evaluated, 7 (2%) were suspected of having craniosynostosis and so were referred for CT. In the author's opinion, CT scanning is not indicated for all infants with plagiocephaly, only those in whom the skull deformation is severe and the suture is "ridged."

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Kane AA, et al	Mandibular dysmorphology in unicoronal synostosis and plagiocephaly without synostosis	Retrospective analysis of CT data of 20 infants with plagiocephaly without synostosis (PWS) who had pre-treatment CT data available.	Class III—Evidence from a comparative clinical study evaluating the CT findings of infants with PWS vs normal infants and infants with true synostosis.	The authors concluded that 3D CT did show significant differences in mandibular morphology and was a useful study. The authors' primary interest was to investigate whether or not dentoskeletal abnormalities existed in PWS and in coronal synostosis and to determine if those abnormalities were significantly different from "normal."

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Katzel EB, Koltz PF, et al (2011)	Treatment of Plagiocephaly with Helmet Molding Therapy: Do Actual Results Mimic Perception?	Retrospective analysis of parent opinion/assessment vs topographical analysis of 61 infants with plagiocephaly and 91 age-matched infants with topographical scans before and after treatment.	Class III—Retrospective comparison of parents' opinion about improvement in head shape and the topographical data from infants treated with cranial molding therapy.	The authors evaluated parents' opinions about their infants' head shapes before and after cranial molding therapy. The topographical data from 91 age-matched infants were studied. These were not the same infants assessed by their parents, so the correlation with parent opinion and actual improvement cannot be made. The only true findings included that, in general, parents were happy with the outcome after cranial molding therapy and that topographical scanning can be used as an objective measure of head shape.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Kluba SR, Schreiber, et al (2012)	Does Helmet Therapy Influence the Ear Shift in Positional Plagiocephaly?	Prospective longitudinal, single arm study of a single cohort. 3D stereophotogrammetry of 80 infants with positional plagiocephaly was accomplished before and after helmet therapy. Of those 80, 60 were found to have an ear shift. Separately, 3 surgeons were asked to evaluate these 80 infants before and after cranial molding therapy.	Class III—Evidence from a prospective, single case series	There was good correlation in clinical assessment of cranial asymmetry, whereas clinical assessment of ear position asymmetries did not correlate to data assessment through 3D stereophotogrammetry. The authors had 2 main conclusions: (1) clinical assessment of cranial asymmetry, but not ear position, does correlate well with 3D stereophotogrammetry data, and (2) helmet treatment does improve ear position in infants with plagiocephaly, as documented with 3D stereophotogrammetry. One may conclude, then, that both clinical assessment and 3D stereogrammetry are both valid and useful in the assessment of infants with plagiocephaly.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Krimmel MB, Will, et al (2012)	Value of high-resolution ultrasound in the differential diagnosis of scaphocephaly and occipital plagiocephaly	Prospective clinical comparison.	Class II—Clinical, ultrasound, and radiological data from 54 infants under 12 months of age with plagiocephaly were presented. The 2 groups described were 47 infants with solely positional plagiocephaly and 7 infants with true craniosynostosis. Under their premise that the inconclusive US findings are regarded as false-positive and false-negative results, the US method had at least a sensitivity of 71.4% (95% confidence interval: 35.5%, 100%), a specificity of 95.7%, a positive predictive value of 71.4% and a negative predictive value of 95.7%.	In 45/47 infants with positional plagiocephaly, the ultrasound images did confirm patent sutures. In 2/47 studies, the ultrasound was inconclusive. In 5/7 with craniosynostosis, the ultrasound did show closed cranial sutures. In 2/7 infants with craniosynostosis, the ultrasound was inconclusive and was followed by a CT in 1 patient and a skull x-ray in the other. In conclusion, the authors felt that ultrasound effectively distinguished between open and closed sutures.
Kuang AA, Bergquist C, et al (2013)	Effectiveness and Safety of Independent Pediatric Nurse Practitioners in Evaluating Plagiocephaly	Retrospective review of the electronic medical records of all patients (N = 1228) seen in a craniofacial clinic from 2005 to 2011.	Class III—Retrospective review of a single series.	The authors concluded that clinical examination by a skilled craniofacial team was able to effectively and safely diagnose plagiocephaly and rule out craniosynostosis without the need of imaging.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Linz C, et al	Occipital plagiocephaly: unilateral lambdoid synostosis versus positional plagiocephaly	A prospective study of 269 children with plagiocephaly without synostosis (PWS, n = 261) and 8 infants with lambdoid synostosis were clinically examined to outline the specific clinical features of both true positional deformity versus craniosynostosis. After clinical examination, ultrasounds (US) were performed. US revealed either a lambdoid synostosis or a patent lambdoid suture in cases of PWS. In 258 of the 269 PWS infants, clinical examination was able to confirm the diagnosis, so that in 3 infants who were initially diagnosed with PWS, a lambdoid synostosis was found on US. In all true lambdoid synostosis cases, US did support the clinical diagnosis. Their conclusion was that CT scan was not necessary in the diagnosis of true lambdoid synostosis.	Class III—Evidence from a prospective, non-randomized, comparative clinical study evaluating the clinical and radiological–ultrasound findings of infants with plagiocephaly, with or without synostosis.	The authors concluded that there are quite distinctive clinical features apparent in infants with lambdoid synostosis when compared to infants with PWS. Additionally, the group concluded that ultrasonography done in infants ≤12 months can be used to confirm the diagnosis of synostosis.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Lipira AB, Gordon S, et al (2010)	Helmet Versus Active Repositioning for Plagiocephaly: A Three-Dimensional Analysis	Retrospective data analysis of 70 infants with plagiocephaly prospectively entered into a non-randomized study with 2 treatment arms: helmet versus positional therapy. 3D topographical analysis was performed before and after treatment. Infants treated with positional therapy (n = 35) were matched for severity of deformity.	Class III—Evidence from a well-designed, case-controlled, comparative clinical study, in which infants with plagiocephaly and treated differently were compared. Cases were matched for severity of deformity. Clinical outcomes were assessed in an objective manner using 3D stereophotogrammetric analysis. 3D topographical imaging was found to be useful for initial assessment and follow-up of infants with PWS.	Using 3D stereophotogrammetric analysis, the authors demonstrated that the infants treated with cranial molding therapy did show a larger reduction in cranial asymmetry as compared to the non-helmeted group. Whole head surface topogrammetry was a useful, objective tool in assessing and determining the cranial asymmetry in infants with plagiocephaly. Of note, comparison of clinical assessment versus efficacy or objectivity of topographical scanning was not the primary aim of this study.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Littlefield TR, Beals SP, et al (1998)	Treatment of Craniofacial Asymmetry with Dynamic Orthotic Cranioplasty	Retrospective analysis of an Excel database of 759 patients with positional plagiocephaly treated between 1988 and 1995 with a Dynamic Orthotic Cranioplasty (DOC).	Class III—Evidence from a single case series of 759 patients with plagiocephaly treated with a cranial molding helmet. Data for 285 (37.5%) were complete and available for analysis.	Photographs and anthropometric measurements were used to assess and evaluate cranial asymmetry before, during, and after treatment. The authors concluded that cranial molding therapy did reduce cranial asymmetry and that anthropometric measurements are reliable and useful.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Lo LJ, et al. (1996)	Plagiocephaly: differential diagnosis based on endocranial morphology	A retrospective study of CT scans done on 234 infants with skull deformity, of whom 170 had PWS, 60 had coronal synostosis, and 4 had lambdoid synostosis. CT data was available in 32 PWS, 27 UCS, and 4 LS. Four radiologists were asked to diagnose unilateral coronal synostosis, true lambdoid synostosis, or positional plagiocephaly (PWS). They found that quantitation of anteroposterior fossae midline angulation assisted in the correct diagnosis of PWS versus lambdoid synostosis.	Class III—Retrospective review of 3 cohorts of infants' CT scans; 1 group of infants with PWS and 2 groups with synostosis.	The authors concluded that CT was useful for the differential diagnosis of clinical plagiocephaly. CT imaging showed significant differences in the external perimeter analysis, cranial fossae symmetry and shape, and the cranial fossa midline angulation. Criticisms include that there was no comparison with sensitivities and specificities of clinical examination and that there was no consideration of radiation dose or exposure.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Losee JE, Feldman E, et al (2005)	Nonsynostotic Occipital Plagiocephaly: Radiographic Diagnosis of the "Sticky Suture"	Retrospective analysis of 3D CT data from 33 infants with posterior plagiocephaly. There were 26 infants without craniosynostosis and 7 infants with classic lambdoid craniosynostosis.	Class III—Evidence from a comparative clinical study evaluating the CT findings of infants with lambdoid synostosis vs those infants without true or classic synostosis. The differences in (1) suture sclerosis and narrowing, (2) endocranial ridging, (3) focal fusions, (4) change in normal overlapping suture morphology to end-to-end orientation, (5) perisutural thickening and thinning, and (6) increases in ipsilateral frontal subarachnoid spacing were reported and significance was determined.	CT was found to be helpful in the differential diagnosis of lambdoid synostosis versus simple occipital plagiocephaly or a "sticky suture." The need for CT scanning for diagnosis was not compared to skull x-rays or clinical examination.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Meyer-Marcotty P, Bohm H, et al (2012)	Head orthosis therapy in infants with unilateral positional plagiocephaly: an interdisciplinary approach to broadening the range of orthodontic treatment	Prospective, longitudinal designed, IRB-approved, case-controlled study of 20 infants with positional plagiocephaly and 20 age-matched controls without cranial asymmetry. Any patient with craniosynostosis diagnosed clinically or confirmed with cranial ultrasound was excluded. A 3D topographical scan was constructed using 5 synchronized cameras.	Class III—Evidence from a well-designed comparative study of infants with and without positional plagiocephaly. The utility of topographical scanning for diagnosis and follow-up of infants with PWS was reported.	The authors found the 3D stereophotogrammetry very useful.
Moghaddam MB, Brown TM, et al. (2014).	Outcome analysis after helmet therapy using 3D photogrammetry in patients with deformational plagiocephaly: the role of root mean square	Retrospective analysis of 40 infants with positional plagiocephaly, between 4 and 10 months of age. The authors felt that clinical assessment by the parent or clinician and anthropometric measurements are subjective, too variable, and time-consuming. They developed a clinical protocol with Root Mean Square (RMS) that is a measure unique to 3D photogrammetry that takes into account changes in shape and volume over time.	Class III—Evidence from a case series of 40 infants under 10 months with positional plagiocephaly. All had 3D stereophotogrammetry before and after treatment with a cranial molding orthotic.	The authors concluded that 3D stereophotogrammetry was an effective and useful tool for the diagnosis and treatment of plagiocephaly. The RMS application was useful in following head shape changes over time.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Mulliken JB, et al.	Analysis of posterior plagiocephaly: deformational versus synostotic.	In a prospective study of 115 infants with posterior plagiocephaly, only 1 infant had lambdoid synostosis and 114 infants had PWS. Some children (n = 54) had skull x-rays and only 11 had CT scans done. CT scans were able to confirm the diagnosis of PWS when there was a clinical question.	Class III—Evidence from a single case series of 114 infants with PWS.	The authors concluded that clinical examination of infants is usually sufficient for the diagnosis of PWS, but in some cases (54/114) skull x-rays were needed. In 11/54 equivocal skull x-rays, a 3D CT confirmed the diagnosis of PWS. Only 1 infant had true lambdoid synostosis.
Netherway DJ, Abbott AH, et al (2006)	Three-Dimensional Computed Tomography Cephalometry of Plagiocephaly: Asymmetry and Shape Analysis	Retrospective study of 21 children with positional plagiocephaly and 20 with craniosynostosis.	Class III—Evidence from a series of infants with plagiocephaly from craniosynostosis or positional deformity. A 3D CT scan was done for each child. All but 1 child were under 27 months. Seventy-eight different osseous landmarks were identified and used to measure cranial asymmetry.	The authors concluded that 3D CT scanning and the measurement of cranial skull vault asymmetry based on the acquired images are useful in the diagnosis and treatment of plagiocephaly in children.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
O'Broin ES, Allcutt D, Earley MJ	Posterior plagiocephaly: proactive conservative management	A retrospective case review of 39 infants with PWS who underwent a clinical examination, orthopometric measurements, photographs, skull x-rays, and 4 had CT scans done.	Class III—Evidence from a single case series of 39 infants with plagiocephaly.	The authors found that clinical examination was usually useful for diagnosis and skull x-rays were difficult to interpret. For cases in which the skull x-ray results and clinical examination were equivocal, 3D CT scans were recommended.
Pollack IF, Losken HW, et al (1997)	Diagnosis and Management of Posterior Plagiocephaly	Retrospective analysis of 71 infants treated prospectively with a clinical management protocol for posterior plagiocephaly. All had skull x-rays. When skull x-rays were not diagnostic, a CT scan was performed.	Class III—Forty out of 71 skull x-rays clearly showed open and patent cranial sutures. CT scans of the head were then obtained in 31/71, and 29 CT scans did show patent, open sutures. In 2 children, true lambdoid synostosis was diagnosed and confirmed.	The authors concluded that their management protocol provided satisfactory clinical improvement in 64/ 69 patients with posterior plagiocephaly. Fifty-six percent (40/71) of skull x-rays were diagnostic and obviated the need for CT scans. In 31 infants (43%), CT scans were necessary.

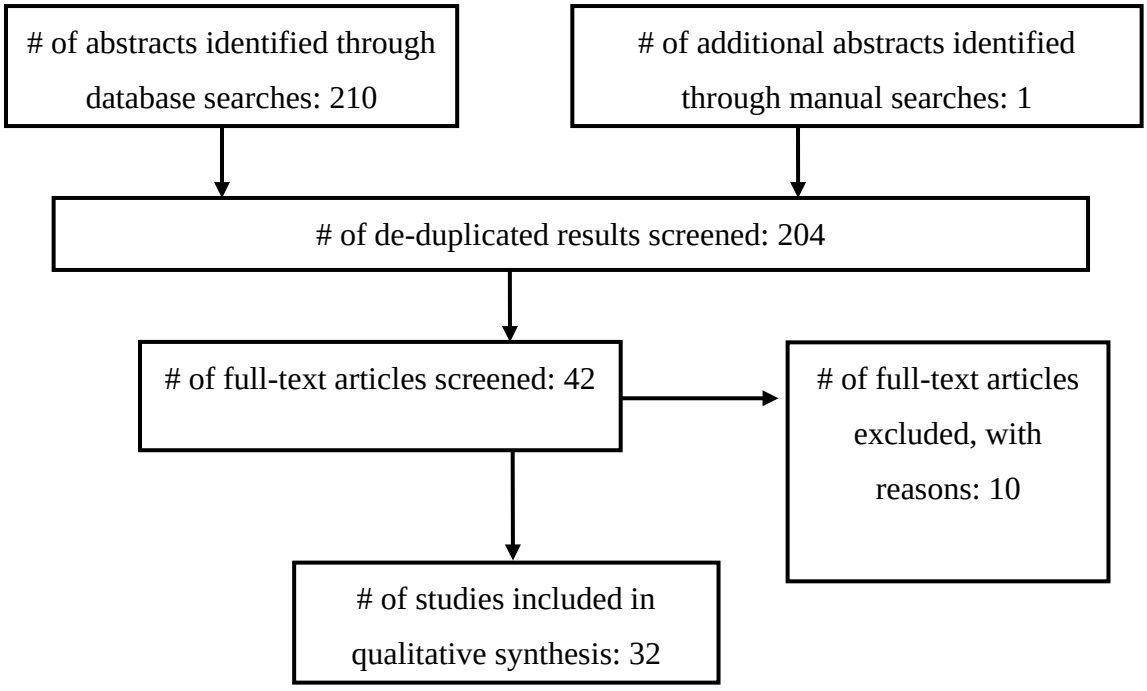
AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Schaaf H, Malik CY, et al (2010)	Three-Dimensional Photographic Analysis of Outcome After Helmet Treatment of a Nonsynostotic Cranial Deformity	Prospective analysis of 181 infants with positional plagiocephaly. All infants were subjected to a 3D stereophotogrammetric analysis of head shape before and after treatment.	Class III—Evidence from a single series of infants with positional plagiocephaly treated with cranial molding therapy. Head shape imaging was acquired on all using 3D stereophotogrammetric analysis. Cranial vault asymmetry index, cranial vault symmetry, and cranial index were assessed and compared before and after treatment.	The authors concluded that 3D stereophotogrammetric analysis provided useful and accurate information for the diagnosis and treatment of positional plagiocephaly.
Schaaf H, Pons-Kuehnemann J, et al (2010)	Accuracy of Three-Dimensional Photogrammetric Images in Non-Synostotic Cranial Deformities	Retrospective, non-consecutive, single case series of 100 infants under 20 months of age, randomly chosen, with deformational plagiocephaly. All infants underwent clinical assessment using caliper measurements and 3D stereophotogrammetric analysis, by 5 clinicians. Each clinician re-measured the 3D stereophotogrammetry pictures 5 times.	Class III—These infants were assessed for cranial vault asymmetry using 2 different methodologies: caliper measurement and 3D stereophotogrammetry. The inter- and intra-rater agreements of the 3D stereophotographs had a low variability in the variance component analysis.	The authors concluded that 3D stereophotogrammetry is a safe and effective method to diagnose and treat infants with plagiocephaly. There was very little variation between the actual caliper measurements and the measurements done using 3D stereophotographs.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Schweitzer T, Bohm H, et al (2012)	Avoiding CT scans in children with single suture craniosynostosis	Retrospective analysis of 137 infants who were being evaluated for craniosynostosis or positional deformity during 2008-2009. There were some infants with single-suture craniosynostosis (n = 110) and some diagnosed with positional plagiocephaly (n = 27).	Class III—Evidence from a single center experience.	The authors concluded that CT scanning is rarely necessary in the differentiation of plagiocephaly versus craniosynostosis. In 133 (97%) of the 137 infants, the diagnosis was made on clinical examination only. Two infants had diagnostic ultrasounds, and only 2 needed CT scan confirmation of the diagnosis.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Sze RW, et al	Ultrasound screening of the lambdoid suture in the child with posterior plagiocephaly	Prospective study of 41 infants who were referred for CT scans of the head. Of those infants, 29 were referred for plagiocephaly and suspected synostosis, of whom 2 had lambdoid fusion. Twelve infants had been referred for indications not related to head shape and were found to have a normal study. Ultrasound imaging and evaluation of the lambdoid sutures were performed. Radiologists interpreting the ultrasound images were blinded to the CT results. The authors reported that in their study, the mean sensitivity and specificity of ultrasound for distinguishing an open from a closed, fused lambdoid suture by 3 blinded pediatric radiologists was 100% and 89%, respectively.	Class II—Evidence from a comparative clinical study evaluating the US findings of infants (n = 27) with PWS versus "normal" infants (n = 12) and 2 with true lambdoid synostosis. The mean sensitivity and specificity of ultrasound in the differential diagnosis of an open from a fused lambdoid suture by 3 blinded pediatric radiologists were 100% and 89%, respectively.	The author concluded that sonography of the lambdoid sutures shows excellent preliminary promise as a screening test of lambdoid suture patency and is helpful in the diagnosis of PWS.

AUTHORS	TITLE	STUDY DESCRIPTION	DATA CLASS/QUALITY AND REASONS	RESULTS AND CONCLUSIONS
Thompson JT, David LR, et al (2009)	Outcome Analysis of Helmet Therapy for Positional Plagiocephaly Using a Three-Dimensional Surface Scanning Laser	Retrospective review of 175 infants; 59 were excluded for failure to follow up or other craniofacial conditions. The remaining 116 infants were included.	Class III—Evidence from single series of infants evaluated using a 3D laser scanning system.	The authors concluded that the laser scanning system was a useful method for objectively measuring outcomes in infants being treated for plagiocephaly with cranial molding helmets.
Vu HL, Panchal J, et al (2001)	The Timing of Physiologic Closure of the Metopic Suture: A Review of 159 Patients Using Reconstructed 3D CT Scans of the Craniofacial Region	Retrospective series of 84 infants with plagiocephaly and 75 infants with trauma who received a CT scan. All CT scans were reviewed for patency of the metopic suture.	Class III—Evidence from a series of children with plagiocephaly who received a 3D CT scan to assess suture patency.	The authors concluded that based on their findings, 3D CT scans did have a role in the evaluation and treatment of plagiocephaly.
Zonenshayn M, Kronberg E, et al (2004)	Cranial index of symmetry: an objective semi-automated measure of plagiocephaly	Retrospective review of 16 infants with plagiocephaly. Diagnosis and treatment was facilitated with a digital camera, a special headband, and measurements made from the digital images.	Class III—Evidence from a single case series of infants with plagiocephaly.	The authors concluded that their system with the headband and digital photography was useful and efficient and reduced radiation exposure.

Figure 1 Flow diagram showing the selection of studies for inclusion in the systematic review



APPENDIX A

PubMed—Plagiocephaly

1. "Plagiocephaly, Nonsynostotic"[Mesh terms]
2. "nonsynostotic plagiocephaly" OR "Positional plagiocephaly" OR "deformational plagiocephaly" OR "flat head" OR “posterior plagiocephaly” OR “positional posterior plagiocephaly” OR “deformational posterior plagiocephaly” OR “occipital plagiocephaly” OR “nonsynostotic plagiocephaly” OR “non-synostotic plagiocephaly”
3. “Plagiocephaly” [All Fields]
4. 1 OR 2 OR 3
5. 4 AND "skull x-ray" OR CT OR "computed tomography" OR MRI OR "magnetic resonance imaging" OR imaging

Limits: “NOT animals”, English language, NOT Comment [publication type], NOT Letter [publication type]

PubMed—Brachycephaly

1. brachycephaly[tiab] OR brachiocephaly OR brachycephalic[tiab] OR brachycephalies[tiab]
2. 1 AND "skull x-ray" OR CT OR "computed tomography" OR MRI OR "magnetic resonance imaging" OR imaging

Limits: “NOT animals”, English language, NOT Comment [publication type], NOT Letter [publication type]

Cochrane Library

1. MeSH descriptor: [Plagiocephaly, Nonsynostotic] explode all trees
2. Title, Abstract, Keywords: “positional plagiocephaly” OR “deformational plagiocephaly” OR “nonsynostotic plagiocephaly” OR “flat head”
3. Title, Abstract: “brachycephaly”
4. 1 or 2 or 3

Limit to English, Humans