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#### **REVIEW ARTICLE**

# Diagnosis of infant synostotic and nonsynostotic cranial deformities: a review for pediatricians

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KEYWORDS Craniofacial abnormalities; Craniosynostosis; Diagnosis; Pediatricians

#### Abstract

*Objective:* To review the current comprehensive care for nonsyndromic craniosynostosis and nonsynostotic cranial deformity and to offer an overall view of these craniofacial conditions. *Data source:* The review was conducted in the PubMed, SciELO, and LILACS databases without time or language restrictions. Relevant articles were selected for the review.

*Data synthesis*: We included the anatomy and physiology of normal skull development of children, discussing nuances related to nomenclature, epidemiology, etiology, and treatment of the most common forms of nonsyndromic craniosynostosis. The clinical criteria for the differential diagnosis between positional deformities and nonsyndromic craniosynostosis were also discussed, giving to the pediatrician subsidies for a quick and safe clinical diagnosis. If positional deformity is accurately diagnosed, it can be treated successfully with behavior modification. Diagnostic doubts and craniosynostosis patients should be referred straightaway to a multidisciplinary craniofacial center.

*Conclusions*: Pediatricians are Q3in the forefront of the diagnosis of patients with cranial deformities. Thus, it is of paramount importance that they recognize subtle cranial deformities as it may be related to premature fusion of cranial sutures.

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PALAVRAS-CHAVE

Anormalidades craniofaciais; Craniossinostose; Diagnóstico; Pediatras Diagnóstico das deformidades cranianas sinostóticas e não-sinostóticas em bebês: uma revisão para pediatras

#### Resumo

*Objetivo:* revisar o atendimento integral atual de craniossinostose não-sindrômica e deformidade craniana não-sinostótica e oferecer uma visão global dessas condições craniofaciais. *Fonte de dados:* A revisão foi realizada nas bases de dados PubMed, SciELO, LILACS e sem

restrições de tempo ou idioma. Artigos relevantes foram selecionados para a revisão. *Síntese dos dados:* Foram incluídas a anatomia e fisiologia do desenvolvimento normal do crânio em crianças, discutindo nuances relacionadas à nomenclatura, epidemiologia, etiologia e tratamento das formas mais comuns de craniossinostose não sindrômica. Também foram discutidos os critérios clínicos para o diagnóstico diferencial entre deformidades posicionais e craniossinostose não sindrômica, dando aos pediatras subsídios para um diagnóstico clínico rápido e seguro. Se deformidades posicionais forem diagnosticadas com precisão, elas podem ser tratadas com sucesso através da modificação do comportamento. Dúvidas de diagnóstico e pacientes portadores de craniossinostose devem ser encaminhados imediatamente a um centro multidisciplinar craniofacial.

*Conclusões:* os pediatras estão na vanguarda do diagnóstico de pacientes com deformidades cranianas. Assim, é de suma importância que reconheçam deformidades cranianas sutis, pois elas podem estar relacionadas à fusão prematura das suturas cranianas.

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#### Introduction

Cranial deformities are a common complaint in pediatric units, since 25% of infants of single pregnancies and 50% of multiple pregnancies have some degree of skull deformity at birth. In general, parents usually recognize these changes in the first weeks or months of life.<sup>1</sup> However, in some scenarios the diagnosis may be overlooked by the family that tends to deny the problem. In these cases, pediatricians must be aware of these issues and counsel the family for the importance to seek a craniofacial team. In addition, it is of fundamental importance that pediatrician be prepared at first consultation to make the differential diagnosis between a positional deformity and craniosynostosis, considering that children born with a positional deformity does not need to be exposed to the ionizing radiation of a computed tomography (CT), apart from the costs of the procedure and the sedation risks to achieve it.<sup>2</sup>

In this report, we review the anatomy and physiology of normal skull development of children, discussing nuances related to nomenclature, epidemiology, etiology, and treatment of the most common forms of craniosynostosis. The clinical criteria for the differential diagnosis between positional deformities and craniosynostosis are also presented, allowing the pediatrician subsidies for a quick and safe clinical diagnosis.

#### Method

The present study is a literature review, with a descriptive approach. We performed a literature review by searching the Medline (PubMed), SciELO, and LILACS databases without time or language restrictions. The final literature review was performed on July 2015. To identify all relevant articles (review articles, clinical trials, and cohort studies) about the current comprehensive care for nonsyndromic craniosynostosis and nonsynostotic cranial deformity the following search terms were used: ''nonsyndromic craniosynostosis', ''nonsynostotic cranial deformity'', ''positional deformity''. ''nonsynostotic posterior plagiocephaly'', and ''positional plagiocephaly''. Each relevant study was individually reviewed to identify information concerning normal skull development of children, nomenclature, epidemiology, etiology, diagnosis, and treatment of the most common forms of nonsyndromic craniosynostosis and nonsynostotic cranial deformity.

#### **Cranial anatomy**

The skull of a newborn is composed of multiple bones and sutures that make it malleable and subject to external forces that deform it, to enable its passage through the birth canal and to accommodate the encephalon, since the brain volume is quadrupled in the first two years of life.<sup>3</sup>

The skull is composed of four major sutures (metopic, sagittal, coronal, and lambdoid), three secondary sutures (frontonasal, temporal squamosal, and frontosphenoidal), and four main bones (temporal, frontal, parietal, and occipital). The metopic suture separates the frontal bones from each other, the sagittal suture from the parietal bones; the coronal suture from the parietal and frontal bones, and the lambdoid suture from the parietal and occipital bones.<sup>3</sup> Besides the bones and sutures, soft and membranous space separating the skull bones are named fontanelle, being of great importance are the anterior or bregmatic (bounded by

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