

Head Measurements for the Diagnosis of Craniosynostosis

As medidas cranianas no diagnóstico das craniossinostoses

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Abstract

Objective Craniosynostosis is a group of growth disturbances of the skull, which can result in serious consequences for the children who suffer from it. Early diagnosis provides treatment at the correct time. Most of the time, the pediatrician uses the head circumference (HC) as a parameter for the diagnosis, but the HC does not appear to be changed in this disease, leading to incorrect interpretations. The anteroposterior distance (APD) and biauricular distance (BAD) appear to be more accurate. The aim of this study is to analyze the value of the HC and the ratio between the measurements of the APD and the BAD for this diagnosis.

Methods We analyzed the data from 129 children who had already undergone an operation for craniosynostosis at an institution in Southeastern Brazil. These data were compared with the normal standards of the population and statistically analyzed to establish their alterations.

Results The HC did not change significantly in cases of craniosynostosis, neither when it was considered as a single disease, nor when considering the different subtypes. The APD/BAD ratio changed significantly in the group.

Conclusion The HC does not appear to be useful in the diagnosis of craniosynostosis as an individual parameter. The APD/BAD ratio appears to help in the diagnosis.

Keywords

- ▶ child care
- ▶ diagnostic recommendations
- ▶ craniosynostosis and head circumference

Resumo

Palavras-chave

- ▶ cuidado infantil
- ▶ recomendações de diagnóstico
- ▶ craniossinostoses e perímetro cefálico

Objetivo Craniossinostoses são alterações do crescimento do crânio que podem resultar graves consequências para a criança. O diagnóstico precoce propicia o tratamento correto. O perímetro cefálico (PC) é habitualmente usado como parâmetro para o diagnóstico. Porém, como na maioria das vezes há alteração na forma e não no perímetro, sua utilização isolada pode levar a detecção tardia. As medidas do diâmetro ântero posterior (DAB) e bi auricular (DBA) parecem ser mais precisas. O objetivo do trabalho é analisar o valor do PC no diagnóstico, bem como o da razão entre as medidas DAP/ DBA.

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Métodos Foram analisadas dados de 139 crianças já operadas no Biocor Instituto, em Minas Gerais. Os dados foram comparados com os parâmetros normais já conhecidos da população.

Resultados O PC não se alterou de maneira significativa nas craniossinostoses, seja como um grupo único ou nos subtipos. A razão das medidas DAP/ DBA alterou-se.

Conclusão O PC isolado parece não ser útil para o diagnóstico das craniossinostoses. A razão das medidas DAP/ DBA parecendo ter utilidade.

Introduction

Craniosynostosis is a well-known group of changes to the shape of the skull, secondary to the early fusion of the sutures. Those changes may cause restrictions in the development of some regions of the head, which are compensated for by abnormal growth in other areas.

The diagnosis of craniosynostosis is essentially clinical, based on the shape of the skull. Anthropometric data should be measured routinely by pediatricians during consultations to help the diagnosis. But, in the daily practice, it is common to use only the head circumference (HC) as a parameter, suggesting that, in the presence of a craniosynostosis, these data would be reduced because the skull does not grow satisfactorily. However, in most cases there is compensation in skull growth, resulting in a dysmorphic perimeter, but a normal HC. The measurement of the anteroposterior distance (APD) and the biauricular distance (BAD) could be more accurate, reflecting the changes in the shape of the head.

The aim of this work is to demonstrate the hole of those measurements in the diagnosis of the disease.

Methods

We analyzed the data from 129 children who had undergone surgical treatment for craniosynostosis at the Biocor Instituto, which were stored in the database of the surgical team. The data collected were age in months, sex, and anthropometric variables (HC, APD, and BAD) measured in centimeters by the traditional semiological technique.¹ The children were classified into craniosynostosis subtypes, depending on the clinical presentation. The subtypes considered were trigonocephaly (closure of the metopic suture), scaphocephaly (closure of the sagittal suture), brachycephaly (closure of the coronal sutures bilaterally), plagiocephaly (closure of the coronal or lambdoid sutures unilaterally), oxycephaly (potential closure of all sutures) or mixed (closure of more than one suture).

The data were subjected to analysis using the Statistical Package for the Social Sciences (SPSS, IBM Corp., Armonk, NY, US) software. A comparison of the percentage of patients was performed with a frequency table and the chi-square test.²

The HC was regarded as decreased, normal or enlarged following the World Health Organization (WHO) criteria (http://www.who.int/childgrowth/standards/hc_for_age).

The APD/BAD ratio was considered normal or abnormal based on a theoretical assumption that the ratio of the measurements would be equal to 1.

Results

Most of the children were male. The mean age was 10.5 months, with a median of 7 months.

Trigonocephaly was the most frequent subtype (49 cases), followed by scaphocephaly (31 cases), plagiocephaly (25 cases), brachycephaly (12 cases), oxycephaly (10 cases) and mixed (2 cases) (►Fig. 1).

The statistical analyses showed that there was a significant predominance of patients with a normal HC (►Fig. 2). Children with decreased or increased values of HC had the same statistical expression (►Table 1).

The HC in the subtypes of craniosynostosis was also analyzed according to the parameters of normal and abnormal (►Table 2). Although there were numerical differences in the expression of normal or altered HC in the different subtypes, the number of children with decreased or increased values of HC was not significantly different (►Fig. 3).

Data showed that the frequency of patients with an abnormal APD/BAD ratio increased (►Table 3).

Discussion

Craniosynostosis is a group of changes to the shape and growth of the skull, secondary to the early partial or total fusion of one or more sutures of the calvaria and skull base.³⁻⁵ Generally, changes in the function of the sutures cause restrictions in the development of some regions of the skull, which are compensated for by abnormal growth in other areas.⁶ According to the involvement of the sutures, several specific subtypes may develop.⁷

The etiology is still debated. Teratogenesis and environmental factors are known, but genetic involvement appears to be the most prevalent cause.^{6,8-10} As a consequence, the disease has a precocious presentation.

Craniosynostosis can result in serious consequences for the child. Changes in the shape of the skull can lead to aesthetic impairments with a variable intensity. Rejection of their appearance, social dumping and lack of emotional commitment are known secondary phenomena of these alterations.^{11,12}

Dysmorphisms may affect the cranial orbits, the base of the skull and the face, and lead to functional impairment.¹³

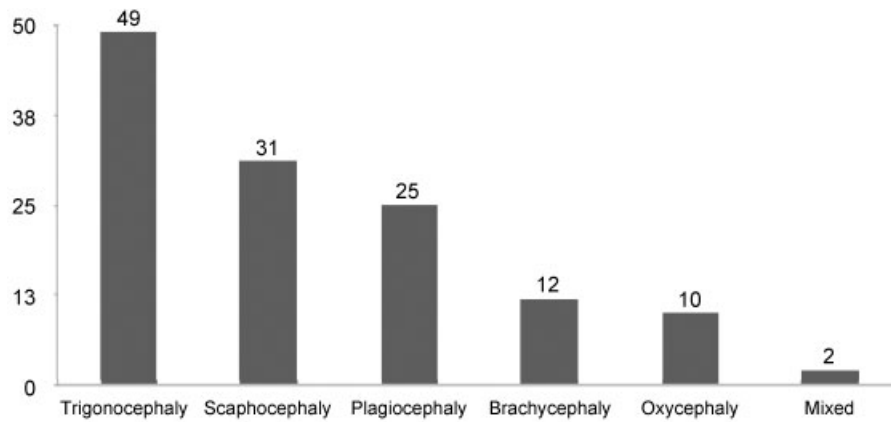


Fig. 1 Distribution of the subtypes of craniosynostosis.

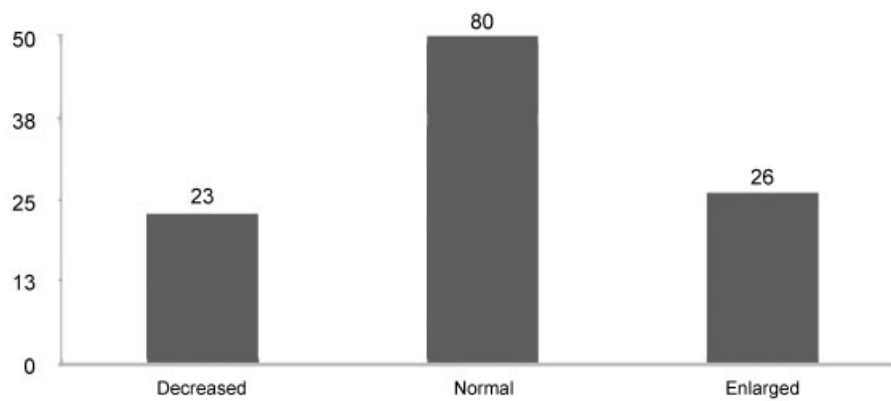


Fig. 2 Distribution by head circumference (HC).

Table 1 Statistical frequency of cases by head circumference

Head circumference			Total	p-value
Decreased	Normal	Enlarged		
23 (17.8%)	80 (62.0%)	26 (20.2%)	129 (100%)	< 0.001

Note: Chi-square test.

Table 2 Distribution by subtypes and head circumference

Craniosynostosis	Head circumference		Total
	Altered	Normal	
Trigonocephaly	15 (30.6%)	34 (69.4%)	49 (100)
Scaphocephaly	18 (58.1%)	13 (41.9%)	31 (100)
Plagiocephaly	6 (24.0%)	19 (76.0%)	25 (100)
Brachycephaly	6 (50.0%)	6 (50.0%)	12 (100)
Oxycephaly	3 (30.0%)	7 (70.0%)	10 (100)
Mixed	1 (50.0%)	1 (50.0%)	2 (100)
p-value	0.086	—	—

Note: Chi-squared test.

Hypo- or hypertelorism, airway involvement and localized brain compression, especially in the posterior region, may follow these manifestations.¹⁴⁻¹⁶

There might also be restrictions to the growth of the skull. During the first year of life, such growth occurs significantly, following the exponential increase in the size of the brain. The restriction of this growth by the inefficient action of the sutures can lead to decreased cranial volume, with consequent chronic intracranial hypertension and injury, in varying degrees, to the infant brain.^{7,17,18}

Surgical treatment is indicated for a significant portion of children with craniosynostosis to avoid the aforementioned consequences. The treatment should be performed early, even in the first months of life, to promote better cosmetic and functional results, and to prevent compression of the brain.^{6,19-22} In addition, the procedure is more aggressive, has more morbidity, and sometimes is less effective when performed later.^{22,23} However, for an early treatment, an even earlier diagnosis is necessary.

The diagnosis of craniosynostosis is essentially clinical, based on the shape of the skull. Pediatricians see the children in consultations in the first months, and have the opportunity to make an early diagnosis of the disease. Anthropometric data such as the HC, the APD and the BAD should be

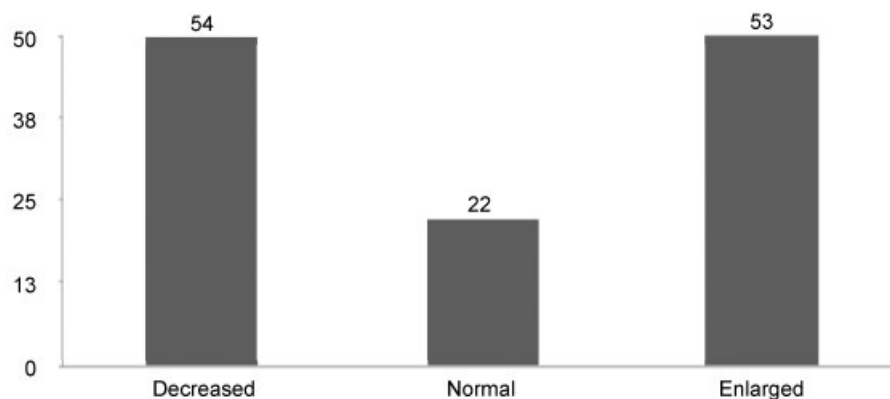


Fig. 3 Distribution by anteroposterior distance (APD)/biauricular distance (BAD) ratio.

Table 3 Comparison of the percentage of patients with normal APD/BAD ratios

	Ratio APD/BAD		Total
	Normal	Altered	
	22 (17.0%)	107 (83.0%)	129 (100%)
<i>p</i> -value	0.196	—	—

Abbreviations: APD, anteroposterior distance; BAD, biauricular distance. Note: Chi-squared test.

measured routinely to help the diagnosis. However, in the daily practice, pediatricians commonly use the HC as a parameter, believing that in the presence of a craniosynostosis, these data would be reduced because the skull does not grow satisfactorily. However, in most cases there is compensation in the skull growth toward the closed suture, resulting in a dysmorphic perimeter, but a normal HC.²⁴ This measurement also does not represent the intracranial volume.^{21,25,26}

The cranial dysmorphism resulting from asymmetric growth could indeed be confirmed by a change in the APD/BAD ratio.^{17,27} Those measurements, not universally used by the pediatricians, could be more precise for the diagnosis.

The analysis of the data showed that the gender of the children reflected the traditional knowledge, with craniosynostosis being more prevalent in boys. The age of the sample revealed the timing of the treatment, when the disease has become well manifested.

The incidence of the subtypes of craniosynostosis draws attention. Classically, scaphocephaly is the most frequent subtype. However, in our sample, trigonocephaly had the highest number of cases. This phenomenon has been previously observed in several services throughout the world, and has been considered a confirmed fact for some authors.²⁸

The analysis of the data seems to show that there are distinct behaviors between them and the craniosynostosis. While the HC does not change, the APD and the BAD are altered in this disease.

The data related to the HC were analyzed considering craniosynostosis as a single disease, and considering each

subtype separately. The HC is normal in the majority of cases, which is in contrast with the classical belief about craniosynostosis. There was a significant similarity in the number of children with increased and decreased HCs. This finding is also in contrast with the perception that the HC is decreased in craniosynostosis.

When analyzing the subtypes, we noted a predominance of HC changes in cases of scaphocephaly and brachycephaly, but such changes were not significant. Therefore, a normal HC is prevalent in all subtypes of craniosynostosis that were studied.

With the APD and the BAD, the perception is the opposite, as these measurements appear to be altered in craniosynostosis. The number of patients with increased or decreased APD/BAD ratio changed in the majority of cases. The statistical analysis of the data showed that these values were significant. Thus, the APD/DBP ratio was altered in craniosynostosis.

Conclusions

The analysis of the data suggests that in craniosynostosis, considered as a single disease, there is no significant change in the HC. The measurement does not change significantly, even when considering the subtypes individually.

Thus, this measurement should not be considered in isolation for the diagnosis of this disease.

Additionally, our study suggests that the APD/BAD ratio is altered in craniosynostosis. Therefore, the analysis of this ratio appears to be useful as a parameter for the diagnosis of this illness.

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Conflict of Interest

All authors have no conflicts of interest to disclose.
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