How ultrasonography can contribute to diagnosis of craniosynostosis

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Comment l'échographie peut aider au diagnostic de craniosténose

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Abstract

Introduction. The aim of this article was to provide an overview of ultrasound (US) techniques for the investigation of cranial sutures in infants.

Material and methods. We first describe a high-resolution sonography technique and its limitations. We then analyze the reliability, effectiveness and role of ultrasonography in routine practice using a PubMed literature review.

Results. Ten studies reported excellent correlations between ultrasonography and 3D-CT. Cranial US for the diagnosis of a closed suture had 100% sensitivity in 8 studies and 86-100% specificity before the age of 12 months. Negative findings mean imaging investigation can be stopped. If ultrasonography confirms diagnosis, neurosurgical consultation is required. Thus, 3D-CT can be postponed until appropriate before surgery.

Conclusion. Cranial suture ultrasound is an effective and reliable technique for the diagnosis of craniosynostosis. It has many advantages: it is fast and non-irradiating, and no sedation is required. It should be used as first-line imaging in infants below the age of 8-12 months when craniosynostosis is clinically suspected.

Keywords: Craniosynostosis; suture; infant; ultrasound; skull

1. Introduction

Several studies have shown the effectiveness of ultrasound (US) in craniosynostosis, but it is not used for routine screening in many centers, as this depends on the preferences of referring physicians. The present study first discusses US techniques for the exploration of cranial sutures; we then review the literature, and finally propose a decision-tree for suspected craniosynostosis in infants.

2. Ultrasonography examination of cranial sutures

The sonographic aspect of normal major cranial sutures, in neonates and infants, was first described by Soboleski et al. [1]. High-resolution sonography of abnormal cranial sutures was described 1 year later by the same author [2]. A high frequency linear transducer with a minimum 12.5 MHz (and ideally >17.5 MHz) should be used, with parameters adjusted depending on the device. The sonographic examination may begin at the anterior fontanelle, with a coronal orientation. A transverse orientation, perpendicular to the plane of the long axis of the suture, has to be maintained along the length of the suture. Coronal, sagittal, lambdoid and metopic sutures must be followed along their whole length.

A normal suture is defined as a hypoechoic gap between two hyperechoic bony plates. The sagittal suture usually has an end-to-end appearance; the lambdoid and coronal sutures usually have a beveled or overlapped appearance [1] (Fig. 1). There was no correlation between suture width or thickness and patient age between 0 and 5 months in Soboleski et al.'s study [1]. A synostosed suture shows loss of the echo-poor fibrous gap between bony plates [2]. An osseous ridge may be observed, in addition to the loss of the normal hypoechoic gap (Fig. 2). Closure may be partial or complete (Fig. 3).

Ultrasound imaging is often described as an "operator-dependent" technique. Although education of radiologists and a short training module is recommended, cranial suture US is relatively easy to perform. Furthermore, it has been shown that there is no interobserver variability with this method [3]. Analysis of coronal sutures may be a little more difficult because of the overlap and the difficulty of maintaining a transverse orientation to the suture while scanning.

3. Validity of the technique

A comprehensive literature review of the National Library of Medicine (PubMed) database was performed to identify relevant studies. It was conducted in February 2019 and analyzed articles published up to that date, with no starting date limit. We used the following key-words: "craniosynostosis diagnosis ultrasound sensitivity" and "cranial suture ultrasound diagnosis craniosynostosis". Studies were selected if their Abstracts were written in English and reported an original article comparing US to a gold standard examination (CT and/or X-ray and/or clinical follow-up) for exploring cranial suture patency. Ultrasound sensitivity and specificity were required to be reported in the study.

The effectiveness and reliability of US examination in children with suspected craniosynostosis, in comparison with radiography, CT or clinical follow up, has been demonstrated in several studies [4–13], showing high sensitivity and specificity (Table 1). In addition, the capacity to determine complete or incomplete closure of the sutures also reaches 100% [13]. Inconclusive US examination is very infrequent: it may be due to poor cooperation in some studies [10] or to relatively advanced age (12-18 months) [11,12]. The less conclusive results in Krimmel's study [7] can be explained by the inclusion of inconclusive US findings as false-positive and false-negative results in their statistics. Alizadeh et al. [8] had one false

negative case in a 7 month-old boy, but did not provide an explanation for this. Pogliani et al. [11] reported 2 false positives during study start-up, and 1 false negative case of bi-temporal suture closure missed by US: they did not evaluate temporal sutures either for technical difficulties or for the lack of definitive literature data about their significance in true craniosynostosis.

4. Role of US in diagnosis strategy (Fig.4)

Diagnosis of positional plagiocephaly can be assessed on physical examination, based on typical features [9] such as trapezoidal head shape, mastoid bulge, skull base tilt and pushed-forward ear position. However, clinical differentiation between isolated unilateral lambdoid and coronal synostosis can be difficult, especially for general practitioners or pediatricians. Rozovsky et al. [10] recommended an algorithm for diagnosis or exclusion of non-syndromic craniosynostosis by pediatricians or general practitioners: when craniosynostosis is clinically suspected, a cranial US examination should be performed first. If negative, there is no need for further imaging studies. If cranial US confirms diagnosis or is indecisive, then neurosurgical consultation is required.

The main advantages of US examination are that it is a fast non-irradiating technique which does not require sedation. It is easy to carry out, facilitating early diagnosis. With US as first-line imaging procedure, 3D-CT may be performed subsequently in case of uncertain US clinical situations. In case of positive US findings in preoperative planning, 3D-CT may also be postponed before surgery at the surgeon's discretion. Considering the potential side-effects of ionizing radiation, sedation risks and costs, some authors even suggest that operative correction of isolated sagittal synostosis can proceed without any radiological investigations, unless clinical examination exhibits atypical features [14,15]. In our opinion, diagnosis still needs confirming,

and cranial US can be a good alternative to 3D-CT. Clinical examination may occasionally be insufficient even for neurosurgeons. For example, some patients have a non-synostotic elongated cranium that suggests scaphocephaly [13,16]; these children should not be operated on and, if they are treated early by molding helmet therapy (at < 12 months of age), head shape may become normalized. Also, breech babies are known to have a mild skull deformation (dolichocephaly) known as “breech head”, which may mimic scaphocephaly [17]. Finally, and regarding these last two points, when craniosynostosis is clinically suspected we consider it necessary to confirm diagnosis systematically on imaging.

According to our experience and the literature, US is limited by age, with the upper limit around 8-12 months [3]. Reliability decreases with age because of hair growth, skull growth and thinner sutures. As most children with cranial deformity are usually seen in the first month of life, we recommend carrying out US examination as soon as possible when diagnosis is suspected, and ideally before the age of 8 months. Finally, and given that physiological closure occurs much earlier in the metopic suture than in the others (normally between the age of 3 and 9 months [18]), complete fusion of the metopic suture does not necessary mean trigonocephaly. Typical triangular frontal deformation and hypotelorism should also be associated with complete fusion of the metopic suture for trigonocephaly to be diagnosed with certainty.

5. Conclusion

US examination of cranial sutures has been shown in the literature to be an effective and reliable technique. Cranial US has many advantages: it is fast and easy to implement and non-irradiating, and infants do not need to undergo sedation. It may serve as first-line imaging when a clinical diagnosis of craniosynostosis is suspected. It allows the 3D-CT examination, if required, to be postponed to an appropriate time before surgery. Given the virtual impossibility of

assessing suture patency after a certain age (around 8-12 months), the earlier the US scan is performed the more reliable the examination.

Disclosure of interest

The authors declare that they have no competing interests.

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Figure legends

Figure 1: Ultrasonographic appearance of normal sutures. A = appearance of normal sagittal sutures (thin arrow). The scalp is indicated by the thick arrow outside the suture. B = appearance of normal coronal suture. Note the difficulty of visualizing the hypoechoic gap of the normal coronal suture (thin arrow) in the lower image because of advanced age (11 months)

Figure 2: Ultrasonographic appearance of synostosed sutures. 3D-CT confirms scaphocephaly (A) and trigonocephaly (C). The corresponding US image at the level of the blue line on the 3D-CT scan shows the synostosed sagittal (B) and metopic (D) sutures with continuous hyperechoic bone and no hypoechoic gap.

Figure 3: Ultrasonographic appearance of sutures (A) and lateral 3D-CT view (B) in a 3 month-old boy with an atypical craniosynostosis (scaphocephaly plus left plagiocephaly). The corresponding US image at the level of the orange line on the 3D-CT scan shows an open metopic suture (1), a sagittal suture partially fused on its posterior part (2-3-4) and a synostosed left coronal suture (5).

Figure 4: Decision-tree for diagnosis of craniosynostosis.