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Mini Review

Update on MRI in Craniosynostosis: A Mini Review

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Introduction

Craniosynostosis are congenital conditions characterized by the premature fusion of one or more of cranial sutures. Cranial sutures are the articulations between the plates of membranous bone and, once formed, constitute the major growth centers of the cranial vault, allowing the concurrent expansion and growth of brain and cranial vault. The premature pathologic fusion of cranial sutures causes characteristic skull deformities, facial asymmetry, and impairment of brain development, in relation to which and how many sutures are involved [1].

Craniosynostosis can be classified depending on whether they appear as a primary condition or secondary to other underlying causes (like metabolic, hematological, teratogenic, etc.); moreover, they are classified in non-syndromic forms, the prevalent form, and syndromic forms [2-4]. The most common non-syndromic affected suture is the sagittal one (40-60% of cases) [3]; the most common syndromes in which craniosynostosis is one of the findings are Apert syndrome, Pfeiffer syndrome and Crouzon syndrome [5]. Syndromic craniosynostosis are often associated to many and various anomalies affecting central nervous system, like Chiari I malformation and polymicrogyria [6,7].

The diagnosis of craniosynostosis is achieved by physical examination and medical imaging [1]. Radiological assessment is especially important in cases of compound synostosis, syndromic forms and when a surgical treatment is planned, radiology allows diagnosis, pre-operative planning and post-operative follow-up [1,8,9]. Radiological imaging is very challenging in infants; many techniques use ionizing radiations, and are therefore perceived as harmful and avoided, and there is usually the need of sedation/anesthesia, raising the risk of adverse effects in this delicate category of patients [6.8].

Computed Tomography (CT) scans have been unavoidable in some complex, syndromic, or complicated types of craniosynostosis [10,11]. Among all the other possible radiological tools that are available nowadays, Magnetic Resonance Imaging (MRI) is gaining more and more attention. In fact, MRI avoid the use of ionizing radiations of CT and the risk of late radiation effects, maintaining many diagnostic advantages. The advent of MRI opened new possibilities for studying brain anomalies associated with craniosynostoses like hydrocephalus or tonsillar herniation, and, thanks to technological progress, for recognizing and distinguishing cranial bones from cranial sutures [6,8,10,12].

Historically, MRI was considered only a complementary technique used to study cerebral and craniofacial soft tissue anomalies in craniosynostosis; one of the more common and worrying brain anomaly associated to craniosynostosis is the concomitant Dural venous abnormalities, caused by abnormal Dural sinus maturation, venous hypertension, and veno-occlusive disease [13,14]. In this kind of patients, MRI venogram allow to understand the location, shape and size of venous sinuses and to provide valuable information to the surgeon.

One of the first papers about MRI and suture analysis was the one of Cotton et al., published in 2005 [15], in which the appearance of cranial sutures and craniometric points were analyzed in adult population. In their imaging protocol, sutures appeared as areas of signal void. In later years, Eley et al. [16] also investigated the potential role of MRI in sutures, applying this interest in pediatric population investigating suture patency. Their protocol consisted of axial T1w, axial T2w, coronal fluid–attenuated inversion recovery (FLAIR), axial short tau inversion recovery (STIR), and sagittal T1; in the axial T2w images, patent sutures were characterized by a signal void, a finding that is also present in other structures like blood vessels. The need to have a specific sequence to provide more detailed information about sutures motivated the same group of authors [17] to develop a new sequence, the so-called "Black Bone" MRI (BB MRI), in which contrast between soft tissue and bones is really improved; BBMRI allowed the visualization of the calvarian bone, sutures, and the craniofacial skeleton.

The technical characteristics of this sequence are 3D volume acquisition, short TE, short TR, and low flip angle. As a result, imaging acquisition times are short, with an average of 4 minutes to acquire craniofacial skeleton axial images. Coronal and sagittal planes are acquired in post-processing. On BB-MRI, the cranial sutures appear as areas of increased signal intensity, well demarcated from the surrounding ipointensity of the bone. In children with pathological suture premature fusion, the patent sutures remain with an increased signal intensity while this feature is absent at the site of synostosis [17].

Recently, black-bone imaging has also been used to obtain 3d images and reconstructions that are very useful to neurosurgeons in the surgical planning [18]. Suchyta et al. [19] also demonstrated the potential of this MRI sequence in producing virtual surgical planning for calvarial vault reconstruction. BB MRI can also help in differentiating between posterior synostotic plagiocephaly and positional plagiocephaly [20].

The most significant limitation of this technique is its intrinsic difficulty in evaluating areas in which there is an air-bone interface, like paranasal sinuses or mastoid region [6].

Discussion

CT scans provide information about skeletal phenotype, with excellent resolution of the sutures and evaluation of skull deformities but has a poorer diagnostic role in the study of brain. On the other hand, MRI is considered the modality of choice to study the brain anatomy and its alterations, any associated brain diseases, like intracranial herniations or hydrocephalus, veno-occlusive disease, and extra-cranial involvement, having though a very marginal role in the assessment of bone and sutures.

Children with craniosynostosis need an accurate evaluation on skull and sutures but also on brain parenchyma, focusing on all the possible CNS anomalies that are frequently concomitant in these patients.

As the use of ionizing radiations is worrying in pediatric patients, other radiological tools are often elected to evaluate pediatric pathologies. In craniosynostosis, MRI with a specific study protocol, including this innovating interesting Black-Bone sequence, allow to study both brain, sutures and bones, to obtain 3d reconstructions and have all the information requested by the surgeon. Moreover, it allows to perform a radiation-free follow-up, avoiding prolonged exposition to ionizing radiations throughout the course of time.

Conclusion

MRI protocol with the BB-MRI seems to be a very promising alternative to CT when imaging patients with craniosynostosis. This protocol provides information on diagnosis and differential diagnosis of skull deformities and allow to recognize intra- and extra-cranial involvement; it covers all critical aspects in preoperative planning in complex syndromic cases and enable to perform a safe follow-up.

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Conflicts of Interest

The authors declare no conflicts of interest.

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