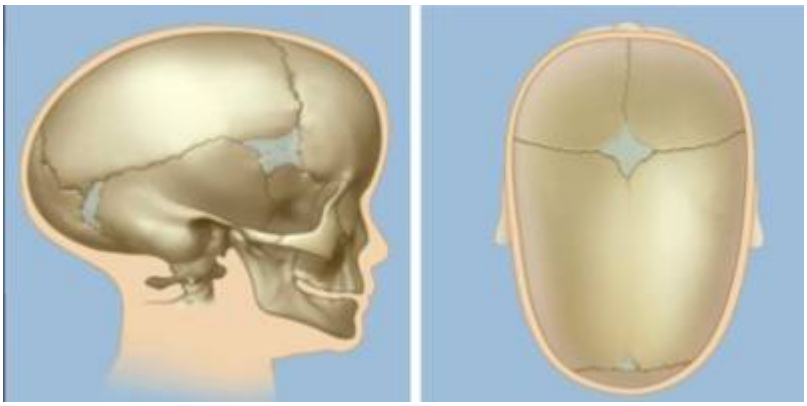
Isolated sagittal synostosis

V.Fernández Cornejo; I.Verdú-Martinez; J.Sales-Llopis

Neurosurgery Department, General University Hospital Alicante, Spain

Latest PubMed sagittal synostosis related Articles

- Augmented Reality Integration in Surgery for Craniosynostoses: Advancing Precision in the Management of Craniofacial Deformities
- Extended Phenotype of Bilateral Coronal Craniosynostosis Due to Novel TCF12 Mutation
- Quantitative analysis of intracranial and intraorbital volume changes following craniosynostosis surgery: a retrospective imaging study
- Spontaneous Correction of Facial Torsion in Nonsyndromic Unicoronal Craniosynostosis: Does Time Heal All?
- Syndromic Craniosynostosis: The Hidden Burden of Comorbidities on Surgical Outcomes
- Laryngeal and pleural ultrasound and acoustic radiation force impulse elastography in dogs with brachycephalic obstructive airway syndrome
- Translation and evaluation of the Infant Characteristics Questionnaire in a sample of Swedish patients with craniosynostosis
- Perioperative complications in surgery for craniosynostosis in children: a systematic review and meta-analysis



Sagittal craniosynostosis, a rare but fascinating craniofacial anomaly, presents a unique challenge for both diagnosis and treatment. This condition involves premature fusion of the sagittal suture, which alters the normal growth pattern of the skull and can affect neurological development. Sagittal craniosynostosis is characterized by a pronounced head shape, often referred to as scaphocephaly. Asymmetry of the face and head, protrusion of the fontanel, and increased intracranial pressure are common clinical manifestations. Early recognition of these features is crucial for early intervention, and understanding the aetiology is, therefore, essential. Although the exact cause remains unclear, genetic factors are thought to play an important role. Mutations in genes such as FGFR2 and FGFR3, which disrupt the normal development of the skull, are suspected. Environmental factors and various insults during pregnancy can also contribute to the occurrence of the disease. An accurate diagnosis

is crucial for treatment. Imaging studies such as ultrasound, computed tomography, magnetic resonance imaging, and three-dimensional reconstructions play a crucial role in visualising the prematurely fused sagittal suture. Clinicians also rely on a physical examination and medical history to confirm the diagnosis. Early detection allows for quick intervention and better treatment outcomes. The treatment of sagittal craniosynostosis requires a multidisciplinary approach that includes neurosurgery, craniofacial surgery, and paediatric care. Traditional treatment consists of an open reconstruction of the cranial vault, where the fused suture is surgically released to allow normal growth of the skull. However, advances in minimally invasive techniques, such as endoscopic strip craniectomy, are becoming increasingly popular due to their lower morbidity and shorter recovery times. This review aims to provide a comprehensive overview of sagittal craniosynostosis, highlighting the aetiology, clinical presentation, diagnostic methods, and current treatment options [1].

Test

“Isolated sagittal synostosis” and “sagittal synostosis” are terms that are often used interchangeably to describe the same condition. Both refer to a specific type of craniosynostosis where the sagittal suture in an infant's skull closes prematurely, leading to a long, narrow, and keel-shaped appearance of the skull. So, whether you hear “isolated sagittal synostosis” or just “sagittal synostosis,” they are typically referring to the closure of the sagittal suture. The term “isolated” may be added to indicate that the sagittal suture is the only suture affected, as opposed to multiple sutures being prematurely closed.

Sagittal craniosynostosis or scaphocephaly is the premature fusion of the suture at the top of the head (sagittal suture) that forces the head to grow long and narrow, rather than wide.

Not ever isolated absence of the sagittal suture does not produce a scaphocephalic skull shape [2].

Epidemiology

While sagittal synostosis is the most common craniosynostosis, long-term follow-up of these patients is lacking.

The data in the literature show that scaphocephaly has an incidence of 0.4 out of 1000 new-born, it has male preponderance M/F = 3,5/1 and familial case occurrence – rare [3], being 40–60% of all nonsyndromic forms [4] [5].

Relationship to gender is observed: 70–90% of all cases are found in boys [6].

If differential progressive deformity occurs between the vault and base, this might affect decision making regarding the timing of surgical intervention.

Types

Atypical sagittal craniosynostosis can be detected initially or occur with a delay in apparently

standard SCS. Leptocephaly is a specific entity. Because of the implications on the management and risk for the patient, preoperative evaluation of patients with SCS with CT scanner and prolonged follow-up are necessary [7].

Association of achondroplasia with sagittal synostosis and scaphocephaly in two patients is reported [8].

Etiology

The etiology is often described as premature fusion of the sagittal suture.

The study of Sakamoto et al., observed wave-like deformations in abnormally long and narrow skulls typical of scaphocephaly, and these deformations were divided into two types: type I deformations (one wave) and type II deformations (two waves). However, the pathogeneses of these deformations are unknown. Computed tomographic data sets were retrospectively analyzed from 18 patients with scaphocephaly who were admitted to the hospital between 2000-2010. Using three-dimensional reconstructions of the computed tomographic images, the relationship was analysed between the wave deformation types and the state of the sutures and fontanelles. The results demonstrate that the type of wave deformation was dependent on the location of the sagittal suture closure. Specifically, the premature closure of the posterior half of the sagittal suture caused a type I deformation, while total closure resulted in a type II deformation ($p < 0.001$). It is hypothesized that restricted growth of the fused suture causes billowing, which results in a waving deformation. The deformities that are often observed in sagittal synostosis can be explained more accurately [9].

Sinus pericranii association [10].

Diagnosis

Sagittal synostosis diagnosis.

Radiomics

Calandrelli et al. investigated the potentialities of radiomic analysis and develop radiomic models to predict the skull dysmorphology severity and post-surgical outcome in children with isolated sagittal synostosis (ISS).

Preoperative high resolution CT scans of infants with ISS treated with surgical correction were retrospectively reviewed. The sagittal suture (ROI_entire) and its sections (ROI_anterior/central/posterior) were segmented. Radiomic features extracted from ROI_entire were correlated to the scaphocephalic severity, while radiomic features extracted from ROI_anterior/central/posterior were correlated to the post-surgical outcome. Logistic regression models were built from selected radiomic features and validated to predict the scaphocephalic severity and post-surgical outcome.

A total of 105 patients were enrolled in this study. The kurtosis was obtained from the feature selection process for both scaphocephalic severity and post-surgical outcome prediction. The model predicting the scaphocephalic severity had an area under the curve (AUC) of the receiver operating

characteristic of 0.71 and a positive predictive value of 0.83 for the testing set. The model built for the post-surgical outcome showed an AUC (95% CI) of 0.75 (0.61;0.88) and a negative predictive value (95% CI) of 0.95 (0.84;0.99).

The results suggest that radiomics could be useful in quantifying tissue microarchitecture along with the mid-suture space and potentially provide relevant biological information about the sutural ossification processes to predict the onset of skull deformities and stratify post-surgical outcome [11].

Treatment

see Sagittal craniosynostosis treatment.

Outcome

see Sagittal craniosynostosis outcome.

Case series

see Sagittal craniosynostosis case series.

Case reports

Rao et al. reported a patient with development of sagittal craniosynostosis after birth, which we term postnatal sagittal craniosynostosis (PSC). This is a rare occurrence in which management considerations are critical but are not well discussed. A 3-year old boy presented with concerns of a metopic ridge. Work-up revealed metopic ridging and an open sagittal suture. The patient later developed signs of increased intracranial pressure (ICP) and repeat CT scan 14 months later identified a newly fused sagittal suture. The patient underwent open posterior cranial vault expansion, resulting in resolution of symptoms. PSC is a rare condition and should be considered in otherwise unexplained increases in ICP among pediatric patients. Open posterior cranial vault expansion represents a safe and effective method to treat this condition [12].

Case reports from the HGUA

Q11573

A 6-month-old female patient was admitted for a scheduled surgical intervention related to sagittal craniosynostosis, specifically scaphocephaly.

Patient History:

No reported adverse reactions to medications (RAMs) or food intolerances. Obstetric history: Controlled and normal evolution of pregnancy, eutocic birth. Personal history: Up-to-date vaccinations, no current medication, and no prior interventions or anesthetic issues.

Current Condition:

The patient, with dolichocephaly, presents for a planned surgical cranial remodeling due to scaphocephaly.

Physical examination reveals a normal developmental status for her age, with an open anterior fontanelle.

Diagnostic Findings:

A cranial CT scan indicates a dolichocephalic calvaria morphology with bilateral frontal bulging. Complete closure of the sagittal suture with the presence of a crest along its entire length. Wide anterior fontanelle. Other sutures are patent, with slight thinning in the middle segment of both coronal sutures.

Surgical Justification:

The infant exhibits cranial deformity with an increased anteroposterior diameter and premature closure of the sagittal suture. Surgical intervention is deemed necessary before the age of one to facilitate cranial remodeling.

Surgical Approach and Technique:

Prone position with a soft U-shaped headrest in a sphinx-like position. Bicoronal zig-zag incision approximately 3 cm posterior to the coronal suture. Periosteal dissection, biparietal craniectomy, removal of the sagittal suture bilaterally, and corrective osteotomies. Frontal radial osteotomies to reduce frontal bossing. Fixation of osteotomies with absorbable material to allow cranial remodeling and increase the biparietal cranial diameter. Closure by layers with subcutaneous and skin sutures.

Materials Used:

Motor Absorbable bone plates

Expected Duration and Complications:

No anticipated extension or specific time estimation was provided. Possible complications listed include hematoma, dural tears, injury to the superior sagittal sinus, deficit in cranial correction, and wound dehiscence.

References

1)
Spazzapan P, Velnar T. Isolated Sagittal Craniosynostosis: A Comprehensive Review. Diagnostics (Basel). 2024 Feb 16;14(4):435. doi: 10.3390/diagnostics14040435. PMID: 38396475; PMCID: PMC10887665.
2)
Padmalayam D, Tubbs RS, Loukas M, Cohen-Gadol AA. Absence of the sagittal suture does not result

in scaphocephaly. Childs Nerv Syst. 2013 Apr;29(4):673-7. doi: 10.1007/s00381-012-2005-4. Epub 2012 Dec 21. PubMed PMID: 23263744.
3)
Shillito J Jr, Matson DD. Craniosynostosis: a review of 519 surgical patients. Pediatrics. 1968 Apr;41(4):829-53. PubMed PMID: 5643989.
4)
Komotar R.J., Zacharia B.E., Ellis J.A., Feldstein N.A., Anderson R.C. Pitfalls for the pediatrician: positional molding or craniosynostosis? Pediatr Ann 2006; 35: 5: 365—375.
5)
Shuper A., Merlob P., Grunebaum M., Reisner S.H. The incidence of isolated craniosynostosis in the newborn infant. Am J Dis Child 1985; 139: 1: 85—86.
6)
Komotar R.J., Zacharia B.E., Ellis J.A., Feldstein N.A., Anderson R.C. Pitfalls for the pediatrician: positional molding or craniosynostosis? Pediatr Ann 2006; 35: 5: 365—375.
7)
Vinchon M, Pellerin P, Guerreschi P, Baroncini M, Dhellemmes P. Atypical scaphocephaly: a review. Childs Nerv Syst. 2012 Sep;28(9):1319-25. doi: 10.1007/s00381-012-1807-8. Epub 2012 Aug 8. Review. PubMed PMID: 22872243.
8)
Accogli A, Pacetti M, Fiaschi P, Pavanello M, Piatelli G, Nuzzi D, Baldi M, Tassano E, Severino MS, Allegri A, Capra V. Association of achondroplasia with sagittal synostosis and scaphocephaly in two patients, an underestimated condition? Am J Med Genet A. 2015 Mar;167(3):646-52. doi: 10.1002/ajmg.a.36933. PubMed PMID: 25691418.
9)
Sakamoto Y, Nakajima H, Tamada I, Miwa T, Kishi K, Yoshida K. New pathogenesis and the classification in scaphocephaly. J Plast Surg Hand Surg. 2014 Feb;48(1):24-7. doi: 10.3109/2000656X.2013.793602. Epub 2013 Apr 30. PubMed PMID: 23627629.
10)
Kimiwada T, Hayashi T, Sanada T, Shirane R, Tominaga T. Surgical treatment of scaphocephaly with sinus pericranii. Neurol Med Chir (Tokyo). 2013;53(2):121-5. PubMed PMID: 23438666.
11)
Calandrelli R, Boldrini L, Tran HE, Quinci V, Massimi L, Pilato F, Lenkowicz J, Votta C, Colosimo C. CT-based radiomics modeling for skull dysmorphology severity and surgical outcome prediction in children with isolated sagittal synostosis: a hypothesis-generating study. Radiol Med. 2022 May 10. doi: 10.1007/s11547-022-01493-6. Epub ahead of print. PMID: 35538388.
12)
Rao V, Ali R, Roussel LO, Crozier JW, Svokos K, Woo AS. Postnatal Sagittal Craniosynostosis: A Novel Presentation and Considerations in Diagnosis and Management. J Craniofac Surg. 2021 Mar 1. doi: 10.1097/SCS.0000000000007599. Epub ahead of print. PMID: 33654043.