Craniosynostosis

- Biomechanical study of an additively manufactured NiTi patient-specific device for the treatment of craniosynostosis
- The Current State of Prospective Registries for Congenital Craniofacial Anomalies: A Review
- Assessing Parents' Perceived Stress, Psychological Distress, and Health-Related Quality of Life Before and One Year After Their Child's Craniosynostosis Surgery
- Evaluation and Potential Improvement of Patient Attendance at a Multidisciplinary Clinic
- Identification and characterization of short-chain dehydrogenase/reductase 3 (DHRS3) deficiency, a retinoic acid embryopathy of humans
- Quantitative analysis of intracranial and intraorbital volume changes following craniosynostosis surgery: a retrospective imaging study
- Cranial Bone Changes Associated With Intracranial Hypertension in Apert Syndrome: Insights for Early Surgical Intervention
- Spontaneous Correction of Facial Torsion in Nonsyndromic Unicoronal Craniosynostosis: Does Time Heal All?

Definition

Craniosynostosis (CSO) was originally called craniostenosis, and is the premature ossification of a cranial suture.

Epidemiology

Incidence: ≈ 0.6/1000 live births.

The most common form of craniosynostosis is synostosis of sagittal suture called scaphocephaly.

Classification

Craniosynostosis classification.

Etiology

Craniosynostosis Etiology.

Pathogenesis

Once the suture ossifies, the normal growth of the skull perpendicular to the suture terminates and tends to proceed parallel to the suture.

Primary CSO is usually a prenatal deformity. Etiologies of secondary CSO include: metabolic (rickets, hyperthyroidism…), toxic (drugs such as phenytoin, valproate, methotrexate…), hematologic (sickle cell, thalassemia…) and structural (lack of brain growth due to e.g. microcephaly, lissence- phaly, micropolygyria…). CSO is rarely associated with hydrocephalus (HCP).11 The assertion that CSO may follow CSF shunting for HCP is unproven.

Treatment is usually surgical. In most instances, the indication for surgery is for cosmesis and to prevent the severe psychological effects of having a disfiguring deformity. However, with multisutural CSO, brain growth may be impeded by the unyielding skull. Also, ICP may be pathologically elevated, and although this is more common in multiple CSO,12 elevated ICP occurs in ≈ 11% of cases with a single stenotic suture. Coronal synostosis can cause amblyopia. Most cases of single suture involvement can be treated with linear excision of the suture. The involvement of multiple sutures of the skull base usually requires the combined efforts of a neurosurgeon and craniofacial surgeon and may need to be staged in some cases. Risks of surgery include blood loss, seizures, and stroke.

Examples of synostoses include: craniosynostosis being an abnormal fusion of two or more cranial bones.

Craniosynostosis causes significant cranial deformity in the pediatric population.

In 1791, S. Soemmering, a German physiologist, anatomist, and anthropologist, admitted that premature fusion of the cranial suture played a significant role in pathogenesis of craniosynostosis.

Virchow R., (1859) defines the craniosynostoses as early closure of the sutures followed by secondary skull deformities that follow a law that says “the normal bone growth is inhibited on the orthogonal direction relative to the closed suture; a compensatory bone growth develops in parallel with the closed suture”.

Virchow’s law partly maintains its validity today. Virchow’s craniosynostose classification chart is still of reference today. However, the early closure of a suture may not always result in a compensatory bone growth. On this ground, the early closure of the sutures should not be defined by the secondary deformity but by the suture or the sutures that were affected [1)].

Notably, craniosynostosis was earlier considered to be an incurable disease and was not of no interest for surgeons. Currently, taking into account the modern methods for diagnostics and treatment, the tactics for this pathology have been significantly changed. There is a clear trend for early diagnosis; earliest possible correction of craniosynostosis is considered to be crucial. Over than a century of surgical treatment of this pathology, multiple surgical techniques have been proposed for different types of craniosynostosis; some of them have been proposed by Russian surgeons. Several of these methods are still used in clinics.

Epidemiology

Craniosynostosis has an incidence of 1 in 2000 to 2500 live births [2)].

Increased ICP

Evidence of increased ICP in the newborn with craniosynostosis include:

1. radiographic signs (on plain skull X-ray or CT)

2. failure of calvarial growth (unlike the non-synostotic skull where increased ICP causes macrocrania in the newborn, here it is the synostosis that causes the increased ICP and lack of skull growth)

3. papilledema

4. developmental delay

Pathophysiology

Craniosynostosis Pathophysiology.

Diagnosis

Craniosynostosis diagnosis.

Evaluation

Cephalic index

Treatment

Craniosynostosis treatment.

Systematic reviews

Craniosynostosis systematic reviews.

Case series

Craniosynostosis case series.

1)

Virchow R. Virchow’s Arch. Path. Anat. 1859;13:323.

2)

Shillito J Jr, Matson DD. Craniosynostosis: a review of 519 surgical patients. Pediatrics. 1968 Apr;41(4):829-53. PubMed PMID: 5643989.