Surgical Management of Complex Craniosynostosis, Case Reports Interdisciplinary Experience

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ABSTRACT

Craniosynostosis, represents the fusion, ossification and sclerosis of one or more cranial sutures, either of the vault or of the base, which cause different degrees of cerebral compression, intracranial hypertension, cognitive deterioration and visual alterations. They can be accompanied by various types of craniofacial dysmorphism, are usually congenital and most require surgery.

Materials and methods: 7 cases of complex craniosynostosis were chosen from formal random, where interdisciplinary management was proposed between the child neurosurgery and maxillofacial surgery team. In this article and / or manuscript we show the satisfactory experience with a positive impact on the aesthetic and functional aspect.

Keywords: Craniosynostosis, Syndromic and Non-Syndromic, Craniosynostosis Surgical Techniques

Introduction

Craniosynostosis, also called craniostenosis, represents the fusion, ossification and sclerosis of one or more cranial sutures, either of the vault or of the base, which cause different degrees of cerebral compression, intracranial hypertension, cognitive deterioration and visual alterations (Mundinger *et al.*, 2016). They can be accompanied by various types of craniofacial dysmorphism, are usually congenital and most of them, require surgery. Craniosynostosis can be non-syndromic or be associated with some syndrome; they are classified as primary and secondary. Primary craniosynostosis occurs in the prenatal period due to developmental disorders. 85% of primary craniosynostosis are non-syndromic (isolated in normal individuals), while 15% are syndromic (belong to poliformative syndromes) (Flaherty *et al.*, 2016; Pérez *et al.*, 2016; Garrocho-Rangel *et al.*, 2018).

70-85% of non-syndromic craniosynostosis are single (affect one suture) and 20-25% are multisutural (Flaherty *et al.*, 2016)

Different classifications of craniosynostosis are used depending on the mechanism, the presence of other disorders or the number of fused sutures. For example, if a craniosynostosis develops due to a primary defect of the ossification process, it is called primary craniosynostosis (Cohen *et al.*, 2017).

On the other hand, secondary craniosynostosis is the result of known systemic diseases, with hematological and/or metabolic dysfunction, such as rickets and hypothyroidism. Secondary craniosynostosis can also develop in newborns with microcephaly due to failure of brain growth or after placement of a shunt in children with hydrocephalus (Cohen *et al.*, 2017).

In addition, craniosynostosis can be classified as syndromic, for example, those that came from Apert, Crouzon or Pfeiffer syndrome, and the most common are non-syndromic craniosynostoses, where it develops as an isolated disorder (Morris, 2016).

As objective of the present study, in Table 1 we grouped the classification of craniosynostosis proposed by Thompson and Hayward (de León, 2011).

Sagittal Synostosis

Affected suture: sagittal

Resulting deformity: Scaphocephaly.

Incidence: 1 in 2,000 (40% of all non-syndromic craniosynostoses)

Epidemiology: male to female ratio is 4: 1

Physical examination: palpable suture of the midline with narrowing of the skull in a biparietal and bitemporal dimension. Frontal and occipital protuberances are present.

Metopic Synostosis

Affected suture: metopic

Resulting deformity: Trigonocephaly.

Incidence: 1 in 2,500 to 15,000 (10-20%; Recent studies have shown an increase in metopic synostosis up to 40% of all non-syndromic craniosynostosis)

Epidemiology: male to female ratio is 3: 1

Physical examination: can occur in a spectrum of deformities ranging from a palpable suture in the middle of the forehead to a severe forehead associated with bitemporal narrowing, biparietal prominence and hypotelorism.

Table 1. Thompson and Hayward classification of craniostenosis.

Type	Suture	Syndromic	Name
Primary	Single suture	Non-syndromic	Scaphocephaly
		Non-syndromics	Plagiocephaly
			Trigonocephaly
			Brachycephaly
			Oxycephaly
	Multiple sutures	Syndromics	Crouzon
			Apert
			Pfeiffer
			Saethre- Chotzen
Secondary	To storage disorders of mucopolysaccharides		Hurler
			Morquio
	To metabolic disorders		Rickets
			Hypothyroidism
	To hematological disorders		Polycythemia vera
			Thalassemia
	To medication intake		Retinoic acid
			Diphenylhydantoin

The types of craniosynostosis

Unicoronal Synostosis

Affected suture: coronal

Resulting deformity: anterior plagiocephaly.

Incidence: 1 in 10,000 (20% of all non-syndromic craniosynostoses)

Epidemiology: male to female ratio is 3: 2

Physical examination: Ipsilaterally, there is a widening of the palpebral fissure, elevation and anterior displacement of the ear, deviation of the nasal base towards the affected side, a prominent malar eminence and an elevation of the supraorbital recess.

Contralaterally, there is a frontal protuberance and the chin is usually deflected to the unaffected side.

Images: the harlequin orbit is present due to the lack of descent of the greater wing of the sphenoid. This deformity consists in the loss of the lateral orbital height with an elevation of the supraorbital recess.

Bicoronal Synostosis

Affected suture: coronal

Resulting deformity: turribrachicephaly

Incidence: 1 in 10,000 (20% of all non-syndromic craniosynostoses)

Epidemiology: male to female ratio is 1:1

Physical examination: the skull typically shortens, anteroposterior diameter and tends to have a widened forehead with hypoplastic supraorbital arches

Lambdoid Synostosis

Affected suture: Lambdoidea

Resulting deformity: posterior plagiocephaly.

Incidence: Rare, less than 3% of all non-syndromic craniosynostoses

Epidemiology: male to female ratio is 2: 1

Physical examination: Ipsilaterally, there is flattening in the occipital region, the ear moves downwards and backwards. Contralaterally, there is occipital prominence (Cohen *et al.*, 2017).

The surgical management of craniosynostosis has evolved significantly over the years. At the end of the 19th century, craniectomies were performed, where the first procedures for craniosynostosis were reported, the objective was to release the fused suture and allow the restriction area to expand with the brain that was growing. In the 1970s (Hu *et al.*, 2017), the objectives changed and not only surgical procedures were performed to release the areas of restriction, but also the aesthetic aspect of the skull was addressed. Cranial vault reconstruction techniques are introduced, using long incisions with extraction and reorganization of large bony flaps to create a normal head form. These were prolonged surgical procedures with greater blood loss, but achieved excellent

results. In recent decades, less invasive surgical procedures have been introduced by adding new techniques and applications to achieve the same results obtained by more aggressive cranial vault reconstructions with less blood loss and shorter scars (Chesler *et al.*, 2018; Pagnoni *et al.*, 2014).

Among the surgical indications for the management of craniosynostosis, are (1) the correction of cranial deformity and (2) the treatment and/or prevention of increased intracranial pressure in order to prevent negative impacts on neurocognitive development and/or the visual disability of patients.

The surgical moment depends of multiple factors that include the type and severity of the synostosis, the surgical technique, the co-morbidities and also the preference of the surgeon. Most surgeons agree that surgical intervention is ideally done before 12 months of age (Brandel *et al.*, 2018).

In general, early surgical intervention occurs between 2 and 3 months of age with minimally invasive procedures. Surgeries at an early age take advantage of the fact that the cranial bones are thinner and more flexible (Prada-Madrid *et al.*, 2017). Surgical intervention in the first months of age could result in a less severe compensatory deformity and early and timely treatment of elevated intracranial pressure. A surgical intervention at more advanced ages (6 to 12 months of age) has the advantage that the cranial bones are harder to support surgical reconstruction, which allows a lower relapse (Jeong *et al.*, 2017)

Many variables come into the decision-making process about the type of surgical intervention for non-syndromic craniosynostosis. They include the age of the patient, the location and degree of the deformity, the general health of the patient and the possible need to repeat the surgery (Joly *et al.*, 2019)

In general, approaches such as endoscopic or linear craniectomy, are usually reserved for selected younger patients (less than 6 months of age) with mild deformities that affect only one suture (Balaji, 2017). More invasive approaches (remodeling of open cranial vault with fronto-orbital advancement) are generally used in older children (older than 6 months) who have more severe deformities that affect one or more sutures.

Surgical procedures often involve the removal of the affected bones, with osteotomies and adjustments of these bone flaps which are remodeled, are placed again on the exposed dura in a more anatomical position and stabilized using different methods (suture, wire, plates, screws). The modern trend of fixation involves resorbable plate systems that are beneficial in that regard, because first, it is not necessary to remove them, as they are reabsorbed over time and second, have less probabilities

than titanium plates to interfere with facial growth. The main features of any operation for craniosynostosis include: release of the fused sutures and remodeling of the hypoplastic and compensatory anomalies of the craniofacial skeleton (de León, 2011).

The surgical techniques commonly used are: reconstruction of the anterior, posterior and total cranial vault, fronto-orbital advancement, minimally invasive techniques, endoscopic craniectomy with the use of a helmet, assisted craniectomy or with the use of a spring, distraction of the cranial vault by osteogenesis.

Each of these techniques are used for the correction of craniosynostosis, there are techniques that can be used more frequently depending on the type of craniosynostosis, for the surgical management of complex craniosynostoses, recommendation is directed to perform an interdisciplinary management, since the approach is holistic with decreased risks as well as better functional and aesthetic results (Kajdic *et al.*, 2018).

Next, the interdisciplinary surgical management that was performed in patients with various cases of craniosynostosis will be related.

Case #1 (premature synostosis unilateral coronal suture)

Frontal-orbital advancement unilateral

Images

Skull CT plus 3D reconstruction

Planning

Positioning and marking

Approach

Result

Case #2 (Brachicephaly)

Planning

Skull CT plus 3D reconstruction

Positioning

Approach

Case # 3 (Saethre Chotzen syndrome)

Coronal flap

Osteotomy lateral wall of the orbit

Orbital osteotomy

Craniofacial disjuncture

RED distractor placement

First week of distraction

Third week of distraction/ consolidation

Distractor removal

Case # 4 (Pfeiffer syndrom)

Monobloc advance osteogenic distraction

Craniotomy

Craniofacial disjuncture

Stage of distraction (craniofacial advance)

Before / After

Case # 5 (Cranio-facial cleft of Tessier)

Approach

Case # 6 (Kabuki syndrome)

Case # 7 (Kleeblattschädel syndrome)

Preoperative CT in first time

Skull CT plus 3D reconstruction

CT postoperative in first time

Positioning in second time

Approach

Discussion

The craniosynostosis, are complex and broad entities in their approach, which not only hinders their diagnosis but also their management; At present, and as it has been previously referenced, surgical management is not only performed for aesthetic purposes, but also functional, since it is demonstrated that adequate surgical management and at the proper time, decreases the appearance of neurological sequelae secondary to late management of craniosynostosis

Within the objectives of surgical management are: correct anomalies (functional and aesthetic), restore normal spatial relationships (of the skull, brain structures and vascular structures), correct alterations of cerebrospinal fluid dynamics and venous circulation and finally, reorientate normal growth vectors.

Therefore, from our experience, we can leave some messages for those interested in the study of childhood neurosurgery:

1. The surgical decision is not easy
2. Is necessary to always have support from the interdisciplinary team
3. Parental support in these cases is essential to make decisions to define expectations that they have in surgery
4. Develop processes for patient follow-up and thus generate: a. institutional memory b. institutional experience c. fertile ground for residents
5. Most appropriate surgical technique will depend on: a. neurosurgeon experience b. interdisciplinary team experience c. therapeutic and social synergism to improve the impact on surgery patients.

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