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Technical Note

Technical Advances in Craniosynostosis Surgery

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Abstract

Introduction: Craniosynostosis is the premature closure of calvarial sutures, resulting in characteristic skull deformations. Surgery has evolved much in this field especially due to technical advances.

Methods: We performed a critical review based in literature about technical advances in craniosynostosis surgery.

Results: Few studies have addressed microscopic approaches in craniosynostosis treatment. Advocated advantages include less operative time, blood loss, and hospitalization. Endoscope-assisted procedure became another important advance warranting less invasive surgeries. The endoscope-assisted repair with postoperative helmet molding is cost-effective and with less operative risk and minimal postoperative morbidity. Absorbable materials are applied in cranial osteosyntesis since 1971. Most materials applied are glicolyc acid and copolymer. They are not associated with increased morbidity, have no interference with normal growth, promote reossification and keep head shape. One of the most limitant factors in early craniosynostosis surgery is blood loss. Many protocols have tried to standardize management of such blood losses. Most try to perform preoperative hemoglobin stimulus combined with intraoperative blood recycling devices.

Conclusion: Craniosynostosis surgical treatment evolved in the last years, mostly due to advances in biotechnology of synthetic materials, surgical techniques and pediatric intensive care measures.

Keywords: Surgery; Craniosynostosis; Technique; Materials

Introduction

Craniosynostosis is the premature closure of calvarial sutures, resulting in characteristic skull deformations. It occurs in approximately 1 in 2500 births. In bilateral coronal, metopic, and lambdoidal nonsyndromic craniosynostosis, there is no observed gender predilection. However, in sagittal synostosis males outnumber females in a ratio of 4:1; in unilateral coronal synostosis, females outnumber males in a ratio of 3:2 [1-5].

Several genetic mutations involving transcription factor, growth factor receptor, and cytokine expression have been identified in association with premature suture fusion in nonsyndromic craniosynostosis [1-3,6,7].

Mounting evidence point to treatment of craniosynostosis instead of conservative approach. The consequences of craniosynostosis may be elevated intracranial pressure, which can result in optic atrophy, blindness and developmental delay making its detection and prevention a priority in treating patients with craniosynostosis [6,7].

Timing for treatment

General indications for surgical intervention include presence of cosmetic deformity and/or functional impairment, such as intracranial hypertension or optic atrophy. Most reference centers delay surgery until a child is at least 3 months old and able to withstand the physiologic stresses of surgery, particularly bleeding. Still, ideal timing for surgical correction of craniosynostosis has been debated, and evidence supporting both early and late intervention (defined as surgery at greater than 1 year of life) exists. The timing of surgical intervention is influenced by surgeon preference, timing of referral to a specialist, and preferred surgical technique. For example, endoscopic techniques are best performed as early as possible, preferably at age less than 3 months. Timing is critical, as the endoscopic technique requires several months of molding helmet therapy postoperatively to optimize results. In contrast, open techniques do not require postoperative helmet molding and can be done later, as the bones are surgically placed in, not molded to, the desired position [7,8].

Preoperative Evaluation

In addition to a thorough medical history and physical examination, ophthalmologic screening should be performed in affected infants, as visual abnormalities commonly accompany craniosynostosis. Imaging techniques such as plain radiographs, CT, magnetic resonance imaging, and ultrasound have been used in the diagnosis of and preoperative planning for craniosynostosis. It has become the standard of care, therefore, to obtain a CT scan with three-dimensional reconstruction. However, with recent concern over the ill effects of ionizing radiation, particularly during infancy, along with the associated cost of such scans, the need for CT in preoperative evaluation of patients with craniosynostosis has been questioned. Fearon et al showed in a prospective multicenter outcome study that the diagnostic accuracy of physical examination alone was 98% in cases of single sutural synostoses. Furthermore, the majority of surgeons poled in this study reported CT scans were not useful to them during surgery [7,8].

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Preoperative imaging has utility in confirming diagnosis and is still performed at most institutions. Imaging is also indicated when the surgeon must evaluate for changes in brain parenchyma, signs of hydrocephalus and ventriculomegaly, presence of tonsillar herniation, or in preoperative planning for cases in which calvarial bone graft will be needed [7,8].

Surgery

It must be stressed that there is no consensus on the optimal surgical techniques for skull reconstruction in any form of craniosynostosis. There are many techniques and modifications that have been described and/or presented. The techniques advocated are dependent on surgeon preference and experience alone, without comparative trials or agreed-upon aesthetic outcomes [9-11].

Sagittal

Surgical approaches for correction of scaphocephaly in sagittal synostosis range from synostectomy (either endoscopic or open), a Pi procedure that involves more extensive strip craniectomy for anteroposterior shortening, to near-total cranial vault reconstruction for children [7,9,10].

Coronal

The correction of unicoronal and bicoronal synostosis requires a frontal reconstruction that addresses the superior and lateral periorbital skeleton as well as the forehead, classically described as frontoorbital advancement. Most surgeons advocate an open technique that is performed as a bifrontal orbital advancement [7,9,10].

Metopic

Surgical correction for metopic craniosynostosis also requires a frontal reconstruction that addresses the superior and lateral periorbital skeleton as well as the forehead. This procedure is preferably done between 8 and 12 months of age. Most surgeons choose an open approach that allows for complete frontoorbital advancement. A bifrontal craniotomy is performed. The frontal bandeau is removed with bilateral temporal extensions. The entire lateral orbital rims are included in the bandeau as C-shaped osteotomies. Trigonocephaly results in a narrow frontal bandeau, even when it is flattened at the glabella. Therefore, it is important to widen the bandeau. This can be done by splitting the bandeau in the midline and inserting 5- to 8-mm wide bone graft (usually parietal bone) that is secured into place with resorbable plates and screws. The orbital bandeau is fixed in place with advancement and twist maneuvers to optimize brow projection [7,9,10].

Lambdoidal

Correction of either unilateral or bilateral lambdoidal synostosis requires bilateral occipital and parietal reconstruction. Posterior vault reconstruction is performed between 3 and 6 months of age in prone position. Bilateral posterior parietal-occipital craniotomies are made. The anterior extent of the parietal cuts is made anterior to the compensatory bulging where the skull resumes normal morphology. The entire posterior parietal-occipital bone can be removed as a single piece or in two pieces with an occipital bandeau, depending on the comfort of the neurosurgeon [7,9,10].

Surgical Complications and Outcomes

Acute complications following open surgical repair of craniosynostosis include bleeding, infection, CSF leak, meningitis, stroke, and even death. Reported postoperative complications include infection, failure of reossification, contour irregularity, and need for reoperation **[7,9,10].**

Technical Advances

Microscopic versus conventional surgery

Few studies have addressed microscopic approaches in craniosynostosis treatment. Advocated advantages include less operative time, blood loss, and hospitalization. On the other hand, there is still few centers experienced enough to perform such surgeries. In those centers, microscopic approach is the treatment of choice in nonsyndromic patients with sagittal and lambdoidal craniosynostosis [11].

Endoscopy

Endoscope-assisted procedure became another important advance warranting less invasive surgeries. However, results must be complemented by helmet use. The endoscope-assisted repair with postoperative helmet molding is cost-effective and with less operative risk and minimal postoperative morbidity. Due to considerably less blood loss, such surgeries may be performed in children under 6 months, especially near 3 months, contributing to prevent the development of associated ventriculomegaly and Chiari I malformation [12-15].

Although most experience of encoscopic technique is to treat sagittal craniosynostosis, it may also be applied in cases of coronal and metopic repair [12-15].

Helmet

Helmet may be applied in adjuvant treatment of endoscopic assisted surgery or may also be used in positional plagiocephaly or brachicephaly. Evidence is lacking to prove effectiveness of helmet therapy compared with the natural course of positional skull deformation [12-15].

Absorbable materials

Absorbable materials are applied in cranial osteosyntesis since 1971. Most materials applied are glicolyc acid and copolymer. They are not associated with increased morbidity, have no interference with normal growth, promote reossification and keep head shape [13].

The application of prefabricated templates in cranio-orbital reshaping is highly useful for accurate preoperative planning, reproducible and efficient intra-operative correction of dysmorphology and objective surgical outcomes assessment [14].

Assessment of blood loss

One of the most limitant factors in early craniosynostosis surgery is blood loss. In young children, total blood volume may be approximately 250ml. In such cases, small perioperative losses might impair surgical results, with need of transfusion and increased rates of infection and hospitalization. Many protocols have tried to standardize management of such blood losses [16].

Most try to perform preoperative hemoglobin stimulus combined with intraoperative blood recycling devices. The CHoR protocol was instituted in November 2008, with the following 3 components; 1) the use of preoperative erythropoietin and iron therapy, 2) the use of an intraoperative blood recycling device, and 3) acceptance of a lower level of hemoglobin as a trigger for transfusion (< 7 g/dl) [16]. This proposal decreased transfusion utilization and length of stay, being suggested as an adequate option [16]. Further analysis are still imperative to disclose complete advantages and disadvantages.

Conclusion

Craniosynostosis surgical treatment evolved in the last years, mostly due to advances in biotechnology of synthetic materials, surgical techniques and pediatric intensive care measures. Such advances have warranted safe surgeries in younger children, resulting in better results and less complications.

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