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Surgical correction of craniosynostosis. A review of 100 cases

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ABSTRACT

Purpose: Provide outcome data for open cranial vault reshaping at a single institution by a single craniofacial surgeon treating 100 patients.
Methods and subjects: A total of 100 patient records were reviewed. Criteria for selection included patients less than three years of age undergoing primary surgery with open cranial vault reshaping and a minimum follow up time of 2 years.
Results: Of the 100 patients (27 female, 73 male) treated 6 were syndromic and 94 nonsyndromic. Average age and weight were 8.9 months and 9.51 kg, respectively. The oldest child was 30 months and the youngest 5 months at the time of surgery. The estimated blood volume lost was 42.7% of total calculated blood volume ranging from 16.6% to 336%. Average surgical time was 216.7 min. Complications included 2 hematomas, 2 wound infections, 1 subgaleal abscess, 6 dural tears, 3 patients requiring reoperation for residual deformity, 4 cases requiring coronal scar revision, 1 sagittal sinus bleed, and 1 intraoperative death.
Conclusions: Our review of 100 open repairs of patients with craniosynostosis demonstrates good long-term results with an overall low complication rate. The outcome data will assist in developing future prospective studies aimed at improving the multidisciplinary care of these patients.

1. Introduction

Craniosynostosis, the premature fusion of cranial sutures, occurs in approximately 3.5–4.5 out of 10,000 live births worldwide (Cohen and MacLean, 2000). Although Samuel Thomas von Soemmerring first realized the association between premature cranial suture fusion and dysmorphic cranial growth in 1791, Virchow is credited with coining the term craniosynostosis and defining the growth principles of cranial dysmorphology resulting from synostosis in 1851. While there have been many advances in the treatment of craniosynostosis, the pathogenesis is only recently beginning to be understood. Virchow suggested that the primary abnormality is at the cranial suture. This is in contrast to Moss (1959) who popularized the concept of an underlying cranial base malformation that propagated via the dura through key ligamentous attachments, resulting in restricted cranial growth. Recent research shows that craniosynostosis results from abnormalities in the equilibrium between proliferation and differentiation of osteoprogenitor cells of the cranial sutures due to gene mutations in fibroblast growth factor receptors and the *Msx* homeobox among others. In general, these mutations lead to defects in signaling and tissue interactions, ultimately resulting in abnormal suture maturation and cranial malformation (Slater et al., 2008).

Craniosynostosis can affect one or multiple sutures, occur as an isolated defect or be associated with a craniofacial syndrome. Nonsyndromic craniosynostosis presents more commonly than syndromic craniosynostosis. Single suture craniosynostosis results in head shape deformities with classic presentations depending on which suture is involved. Sagittal suture fusion results in scaphocephaly, the most common synostosis abnormality in the United States. Unilateral coronal suture fusion, more commonly, or lambdoidal suture fusion, less commonly result in plagiocephalic head shapes, and bilateral coronal or lambdoidal fusions present with brachiocephalic head shape deformities (Ghali et al., 2002).

Intracranial hypertension, visual impairment, limitation of brain growth and neuropsychiatric disorders are associated with craniosynostosis; generally with greater functional disturbance in proportion to the number of sutures involved (Bristol et al., 2004; Kordestani et al., 2006; Siatkowski et al., 2005; Magge et al.,

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2002; Panchal et al., 2001). In surgically correctable cases, where abnormal skull growth is not related to underlying brain growth abnormalities, various approaches have been described. Historically, all repairs were performed via open transcranial approaches. Recently, interest in minimally invasive techniques, such as endoscopic suture release, spring assisted surgery, and distraction osteogenesis have been studied in an effort to potentially reduce surgical morbidity (Jiminez and Barone, 2010; David et al., 2010; Kim et al., 2008; Tellado and Lema, 2009).

While inherent risks of open cranial vault reshaping exist, the past two decades have enjoyed advances in resorbable fixation, imaging modalities, and perioperative medical management. The purpose of this retrospective review is to provide a single surgeon's experience in management techniques, types of craniosynostosis, perioperative data and complication rates for open cranial vault reshaping at Louisiana State Health Center in Shreveport.

2. Materials and methods

2.1. Patient selection and study design

The Institutional Review Board at Louisiana State Health Sciences Center in Shreveport, Louisiana (LSUHSC-S) approved this retrospective case review. The inclusion criteria were patients less than three years of age undergoing primary surgery with open cranial vault reshaping and a minimum of 2 years follow up. Based on inclusion criteria, 100 patients with craniosynostosis, treated with surgical correction between 1997 and 2011 by the institutional craniofacial team (Departments of Oral and Maxillofacial Surgery and Neurological Surgery) were included. The surgical procedure of choice was single stage open transcranial vault reshaping with barrel-staving and orbital bandeau advancement as needed for existing fronto-orbital dysmorphology. Resorbable plates and screws were used exclusively, based on their success and safety in pediatric craniofacial surgery (Eppley et al., 2004).

All cases were performed at LSUHSC-S by a single craniofacial surgeon (GEG), two pediatric neurological surgeons (BW) (CN), and rotating anesthesiologists and pediatric intensivists assigned to the craniofacial team. Patient medical records were used to assess the length of surgery, estimated blood loss, postoperative complications, and average length of hospital stay.

2.2. Preoperative assessment

A complete history and physical exam were performed along with computed tomography utilizing three-dimensional reconstruction for pre-surgical planning.

2.3. Anesthesia considerations

Standard monitoring using temperature probes, electrocardiography, capnography, and pulse oximetry were employed. Induction was achieved with sevoflurane in most cases. The standard protocol included central venous access and an arterial line placed by a pediatric general surgeon, hypotensive anesthesia, and packed red blood cell transfusions given at key portions of each case to correspond with anticipated blood loss (Sinn et al., 1997; Hilley et al., 1992). No antifibrinolytic agents, such as aminocaproic or tranexamic acids, were used.

2.4. Patient positioning

Patients undergoing anterior cranial vault reshaping for metopic or coronal suture synostosis were placed supine in the pediatric horseshoe headrest. The endotracheal tube was secured to the chin using 2-0 silk suture. Temporary tarsorrhaphy sutures were placed for protection of the globes. Those undergoing surgery for posterior or total cranial vault reshaping were placed prone with the neck slightly extended to allow access to the entire cranial vault. In prone cases, extra care in the way of foam padding was used to protect the globes.

2.5. Surgical approach

All procedures employed a coronal incision with Raney clips for hemostatic assistance. Dissection was carried out in a subperiosteal plane to expose the necessary area for reshaping (Fig. 1).

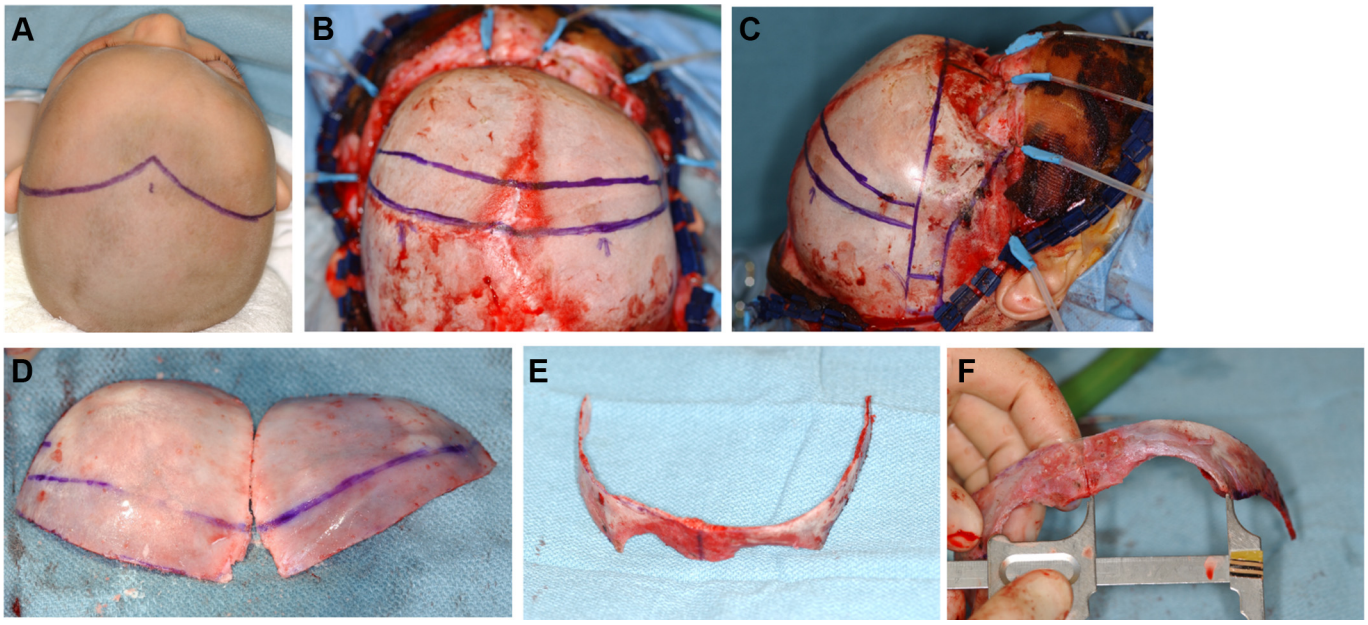

Fig. 1. Patient with anterior plagiocephaly secondary to right coronal synostosis. (A) Proposed coronal incision marked. (B) Coronal suture osteotomies marked. (C) Orbital bandeau osteotomies marked. (D) Frontal bone with midline osteotomy to facilitate reshaping. (E) Orbital bandeau showing obvious deformation. (F) Orbital bandeau after osteotomies (midline and bitemporal) and reshaping to achieve symmetrical superior orbital rim position and length.