Surgical treatment of single-suture craniosynostosis: an argument for quantitative methods to evaluate cosmetic outcomes

A review

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The traditional reasons for surgical intervention in children with single-suture craniosynostosis (SSC) are cosmetic improvement and the avoidance/treatment of intracranial hypertension, which has been thought to contribute to neurocognitive deficits. Despite considerable work on the topic, the exact prevalence of intracranial hypertension in the population of patients with SSC is unknown, although it appears to be present in only a minority. Additionally, recent neuropsychological and anatomical literature suggests that the subtle neurocognitive deficits identified in children with a history of SSC may not result from external compression. They may instead reflect an underlying developmental condition that includes disordered primary CNS development and early suture fusion. This implies that current surgical techniques are unlikely to prevent neurocognitive deficits in patients with SSC. As such, the most common indication for surgical treatment in SSC is cosmetic, and most patients benefit from considerable subjective cosmetic normalization following surgery. Pediatric craniofacial surgeons have not, however, agreed upon objective means to assess postoperative cranial morphological improvement. We should therefore endeavor to agree upon objective craniometric tools for the assessment of operative outcomes, allowing us to accurately compare the various surgical techniques that are currently available. (DOI: 10.3171/2010.5.PEDS09313)

Key Words • craniosynostosis • single suture • craniometrics • intracranial hypertension • intracranial pressure

Single suture craniosynostosis most commonly occurs in the absence of syndromic disease and affects approximately 1 in 2000 children.31,32 The condition involves the premature fusion of one of the 6 major cranial sutures, with the sagittal suture being the most commonly affected and the lambdoid being exceptionally rarely involved. It is widely recognized that children with SSC, regardless of previous surgical treatment, are at increased risk of developing subtle neurocognitive deficits as they progress through childhood.6,9,25,31,33,38,40 Historically, surgical intervention for the treatment of SSC has been intended to improve cosmesis and to prevent or treat intracranial hypertension. There is little debate that surgery is indicated to improve the cosmetic deformities associated with SSC and to prevent the potential emotional sequelae of such deformities.5,7 The role of surgical intervention for the treatment of intracranial hypertension has been somewhat more controversial for a number of reasons. Although multiple reports demonstrate that cranial vault remodeling can alleviate elevated ICP,1,21,42,44,54 the prevalence of elevated ICP in patients with SSC remains unknown. Additionally, there is a paucity of evidence connecting elevated ICP to the neurocognitive deficits that are seen in these patients during childhood. Likewise, there is a concurrent lack of evidence correlating surgical intervention with a decrease in the risk of developing neurocognitive deficits.

Neuropsychological30,31,51 and anatomical literature2,4,20 has suggested that, rather than being the cause of underlying brain injury, the cranial morphological changes associated with SSC result from an underlying

Abbreviations used in this paper: ICP = intracranial pressure; SSC = single-suture craniosynostosis.
primary developmental condition, which may also produce the neurocognitive symptoms associated with SSC. This theory implies that current surgical techniques are of limited value in preventing the neurocognitive deficits that affect these children. Although some studies have demonstrated postsurgical improvements in development, a causal link has not been established. This does not call into question the need for surgical treatment for these children, but rather helps surgeons focus on the fact that the primary indication in the majority of cases is cosmetic. Normocephaly not only improves the parent/child relationship but also protects the child against the potentially severe emotional sequelae that may result from cranial deformity. These facts, when considered in the context of the various options available for the surgical treatment of SSC, should strongly influence pediatric craniofacial surgeons to agree upon objective methods for the assessment of the cosmetic goals of SSC treatment.

Challenges in Determining the Prevalence of Elevated ICP in SSC

Multiple authors have reported studies using invasive monitoring to determine the prevalence of elevated ICP in the SSC population. In a thorough review of the literature regarding findings such as papilledema, intracranial volume measurement, Pittman concluded that, although there is a subpopulation of children with SSC who harbor elevated ICP, it is difficult to determine the exact size of this group or identify individuals with intracranial hypertension in the absence of clinical symptoms. Among the major reasons why this group of patients remains elusive are that there is no universally accepted definition of elevated ICP in infants and young children and there has been considerable variation in the selection criteria used to identify subjects. Although efforts using clinical and/ or radiographic parameters to identify SSC patients with elevated ICP have been unable to identify a tool or group of tools that is both sensitive and specific, estimates of the prevalence of preoperative intracranial hypertension range from 4.0% to 24%, indicating that a minority of SSC patients harbor elevated ICP. This supports the assertion that the primary operative goal for the vast majority of infants who undergo surgical treatment for SSC is improved cosmesis.

Neurocognitive Deficits, Elevated ICP, and Surgical Treatment

The prevalence of neurocognitive deficits in children with SSC, regardless of prior treatment, has been established through multiple studies, which were reviewed by Speltz et al. and Kapp-Simon et al. Despite having normal measures of global intelligence, school-aged children with SSC have a 35%–50% risk of harboring subtle neurocognitive deficits, which is 3–5 times the rate seen in unaffected children and is higher than any estimate of the prevalence of elevated preoperative ICP. In infants and young children, these deficits most frequently manifest as delays in mental or psychomotor development, while language deficits emerge as children progress to school age. Studies by the group from Paris and the group from Paris were unable to identify a correlation between elevated ICP and neurocognitive outcomes in children with SSC, indicating that chronic elevations in ICP are unlikely to be the causative agent for these delays.

Although there are conflicting reports, the preponderance of current evidence does not indicate that early surgical intervention results in improved neurocognitive outcomes in children with SSC. Starr and colleagues completed pre- and postoperative neurodevelopmental testing in 168 infants with SSC and 115 controls who did not undergo surgery. The authors found that infants with SSC were more likely than controls to score in the delayed range on measures of mental and psychomotor development both pre- and postoperatively. A smaller study comparing SSC patients who did or did not undergo surgical treatment found no difference in developmental scores, implying that early surgical correction did not improve neurocognitive function. Speltz et al. also reported a small study that was unable to confirm a positive effect of surgery on neurocognitive outcomes. Becker and colleagues reported a 39%–61% incidence of speech or psychological abnormalities at 5.1 years of age in children with SSC who had undergone previous surgical treatment. However, a group of studies did report an inverse correlation between the age of surgery and developmental scores. It is, however, difficult to draw definitive conclusions from these studies due to an absence of control groups or a lack of statistically significant results. Taken as a whole, the current literature suggests that cognitive impairment in children with SSC occurs despite early surgical intervention.
Craniofacial abnormalities of major brain structures could contribute to the subtle cognitive dysfunction observed in some of these patients. Furthermore, in their analysis of long-term outcome of a large surgical series Fearon and colleagues also suggest that disordered postoperative growth patterns following SSC correction may be symptomatic of an underlying primary process.

Craniofacial Appearance

As previously stated, the ability of surgical intervention to correct the morphological disturbance associated with SSC is well established. Direct anthropometric techniques, such as calculation of the cranial index ([maximum cranial width \( \times \) maximum cranial length]) have been the most commonly employed methods of quantitatively assessing results. These methods are objective, reproducible and avoid the use of ionizing radiation. Additionally, the existence of normative databases allows comparison of surgical outcomes. However, simple anthropometric techniques, such as the cranial index, fail to characterize the dysmorphic state with sufficient detail to be a standard for pre- and postsurgical measurement. More recently described techniques for quantitative craniofacial analysis can generally be divided into those that are based on radiographic data and those that use direct measurement or other measurement methods that do not involve ionizing radiation. While avoiding exposure to ionizing radiation is preferable in young children, the precision, availability, and temporal comparability of current CT-based measurement techniques are superior to alternative methods.

Plank and colleagues described their application of the STARscanner system (Orthomerica Products, Inc.) to quantify the 3D cranial contours in 224 infants with deformational plagiocephaly. Images were obtained in less than 2 seconds using a nonionizing laser-based system. In this study, the reconstructed images were used to create custom cranial orthoses for the treatment of deformational plagiocephaly. Advantages of this method of quantification include rapid acquisition time, absence of ionizing radiation, and the ability to acquire 3D data. The primary limitation is lack of access to the imaging system, which is currently available in approximately 28 medical centers. Van Adrichem and colleagues proposed the use of plagiocephaly as a reliable means of obtaining 2D skull measurements in infants and young children without the use of ionizing radiation or the need for anesthesia. Plagiocephalmometry involves the placement of a thermoplastic strip (Thermo Extradform, Non-Perfo, GeniMedical) around the widest circumference of the child's head and marking of the position of the ears and nose. The strip retains its shape following removal from the patient's head. The authors reported excellent inter- and intrarater reliability and correlation with CT-based imaging. Advantages of this method are the low cost and ease of repeated application. Disadvantages are that measurements are limited to 2 dimensions and the device requires approximately 2 minutes in position to maintain its shape.

Marcus and colleagues used reformatted CT data to apply a technique called “mid-sagittal vector analysis.” The authors established a midline point at the apex of the dorsum sellae and from there created a series of vectors projecting radially to the outer table of the skull. The length of each vector was then measured. The authors subsequently used this technique to establish a normative database against which to compare data obtained in SSC patients who have undergone surgical correction. Using the normative database as a standard, surgeons could potentially directly compare the results of various surgical techniques. Frühwald and colleagues and Danelson and colleagues demonstrated that CT data can be reliably converted into stereolithographic models of the skulls of children with craniofacial anomalies. These models could potentially be used for postoperative assessment. An example of this concept was demonstrated by Amm and Denny, who used postoperative CT to generate 3D digital models, allowing them to assess the evolution of each patient's skull shape over a 3-year period. Smartt and colleagues used basic craniometric analysis derived from reformatted 3D surface imaging to quantify the pre- and postoperative ear position of 9 patients with isolated unilateral lambdoid synostosis. Ruiz-Correa and colleagues used mathematical modeling techniques to classify synostotic head shapes based on CT slices. The obvious disadvantage of this group of CT-based methods is the requirement for postoperative exposure to ionizing radiation, although low-dose CT protocols may somewhat diminish the associated risk as would the need for only a limited number of primary images.

Conclusions

The surgical treatment of SSC has traditionally endeavored to achieve cosmetic goals and prevent the neurocognitive decline ostensibly resulting from chronic elevations in ICP. Improvement of the cosmetic deformity associated with SSC not only eases parental anxiety but facilitates early parent-child bonding and will, in many cases, prevent damage to the growing child's self image. In the majority of cases, these goals represent the primary indication for surgical intervention. As such, pediatric craniofacial surgeons are obliged to assess their outcomes with regard to this parameter in an objective manner. The measurement techniques discussed above represent potential means through which this could be achieved. Until a satisfactory craniofacial method or group of methods is established, it will be difficult to meaningfully compare the outcomes of the myriad operative techniques currently available for the treatment of SSC.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Hankinson, Anderson, Feldstein. Acquisition of data: Hankinson, Fontana. Analysis and interpretation of data: Hankinson, Anderson, Feldstein. Drafting the article: Hankinson. Critically revising the article: Anderson, Feldstein. Reviewed final version of the manuscript and approved it for submission: all authors. Administrative/technical/material support: Hankinson.
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Treatment of single-suture craniosynostosis

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Manuscript submitted July 14, 2009.

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