Minimally Invasive Surgical Techniques with Postoperative Assistive Remodeling for the Treatment of Early-Diagnosed Craniosynostosis. Have they become a New Standard of Care?

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Abstract

Craniosynostosis is the congenital premature closure of sutures in the cranium that affects approximately 1 in 2500 live births [1,2]. Treatment of craniosynostosis is primarily surgical and warranted not only for cosmetic purposes, but also for the prevention of negative psychosocial consequences and developmental delay associated with potentially increased intracranial pressure resulting from sutural fusion-restricted skull bone expansion that otherwise parallels brain growth. Historically, the first attempts at surgical repair of craniosynostosis were by suturectomy. However, the poor outcomes associated with these late nineteenth century repairs led surgeons to abandon the suturectomy method for several decades. Ensuing advances in surgical technique, anesthesia, and blood transfusions allowed surgeons to resume their efforts at surgically repairing craniosynostosis, culminating in refined techniques at open cranial vault remodeling that dominated the latter half of the twentieth century. Nonetheless, efforts at minimizing intraoperative blood loss, postoperative hospital stay, complications, and incisional length have given rise to several important, minimally invasive surgical techniques for craniosynostosis repair over the last quarter-century. These include endoscopic suturectomy with postoperative orthosis, spring-assisted cranioplasties, and distraction osteogenesis. While selection of appropriate surgical technique remains the subject of substantial debate among surgeons, available data suggests that the minimally invasive suturectomy technique assisted with postoperative external remodeling devices for the treatment of craniosynostosis has clear benefits for young infants over open reconstructive surgery in terms of lessened operative blood loss, procedural length, hospital stay, and even cost. When and wherever the conditions and resources are available, the minimally invasive suturectomy technique assisted with postoperative external remodeling devices should be considered the standard of care.

Keywords

- Craniosynostosis
- Suturectomy
- Orthosis

INTRODUCTION

Craniosynostosis is the congenital premature closure of sutures in the cranium that affects approximately 1 in 2500 live births [1,2]. It results from premature ossification and closure of the fibrous joints between the bones of the skull, which disrupts the otherwise naturally symmetric expansion of the infant’s skull that regularly takes place with brain growth, as the bones cannot expand perpendicularly to the closed suture. The resulting abnormal head shape depends on which suture has fused. Single-suture fusion is the most common and can occur with an unknown etiology (non-syndromic) or with a syndromic etiology.

Non-syndromic craniosynostosis is often further classified as complex or simple, depending upon whether it affects multiple sutures or a single suture. It is usually identified by the type of suture fusion: sagittal, metopic, bi- or uni-coronal, bi- or uni-lambdoidal synostoses. These specific fusions lead to the development of the following abnormal skull shapes: scaphocephaly, trigonocephaly, brachycephaly, and anterior or posterior plagiocephaly, respectively. The sagittal suture is most commonly affected, (accounting for more than 50% of cases), followed by the metopic (approximately quarter of cases), and then uni-coronal (estimated 18% of cases) sutures [1-3].

Syndromic craniosynostosis accounts for approximately 25-30% of all cases [1], and manifests with additional anomalies and developmental delays; monogenic mutations or chromosomal
defects are identified in 80% of syndromic patients [3], the majority being associated with mutations in genes encoding fibroblast growth factors (including FGFR-1, -2, and -3) and the transcription factors TWIST and MSX-2 [2,4-7]. Non-syndromic craniosynostosis has been associated with additional gene mutations, including TCF12, ERF, TWIST, ALX4, RUNX2, FREM1, within interconnected signaling pathways [1]. Certain prenatal exposures, such as to maternal tobacco smoking and other hypoxemia-inducing agents, have been associated with isolated craniosynostosis [8].

The purpose of this review is to present the existing accumulated knowledge regarding minimally invasive surgical techniques for the treatment of early-diagnosed craniosynostosis and discuss whether these techniques should be considered the new standard of care, compared to the accepted open cranial vault reconstruction (CVR).

METHODS

We conducted a literature review on the topic of surgical treatment options for craniosynostosis, focusing on the historical evolution of treatment and the comparison between open-cranial vault reconstruction and minimally invasive treatment options in the context of surgical indications, outcomes, complications, and cost-benefit analysis. We specifically searched PubMed for permutations combining the following key words with “craniosynostosis:” cranial vault reconstruction, endoscopic suturectomy, suturectomy, strip craniectomy, sagittal, metopic, coronal, plagiocephaly, cranial orthosis, external remodeling, spring assisted cranioplasty, distraction osteogenesis, and cost-value-benefit.

RESULTS

History

Recognition of the importance of the skull sutures and their relationship to head shape dates back centuries to the times of such investigators as Hippocrates, Galen, and Celsus. In 1791, Von Sommering was the first known to explicitly document the origin of calvarial growth at the suture lines and recognize that premature suture closure led to restriction of growth perpendicular to the affected suture. In 1851, Virchow first introduced the term ‘craniosynostosis,’ [9] and his theories served as the basis for ensuing surgical treatment of craniosynostosis with the goal of removing the affected suture and thereby permitting resumed symmetric growth.

In 1890, Lannelongue performed the first documented attempt at surgical correction of craniosynostosis in France, describing bilateral strip craniectomies for the treatment of sagittal synostosis [10]. Lane followed in the United States in 1892; unfortunately, Lane’s patient died shortly after surgery from anesthesia-related complications [11]. In 1894, Jacobi reported on a review of 33 open strip craniectomies on craniosynostosis patients with astoundingly high morbidity and mortality rates due to procedural blood loss [10]. Following Jacobi’s highly publicized review, there was a paucity of such procedures performed secondary to concerns over poor outcomes. Decades after, Mehner began the revival of corrective surgeries, reporting on the successful completion of a craniectomy for removal of a fused suture [12]. In 1927, Faber and Towne likewise reported their results for extensive craniotomies and removal of fused sutures, reporting significantly lower rates of morbidity and mortality than documented by Jacobi [13]. In the decades that followed, enormous advances in anesthesia, blood transfusion, and surgical techniques ensued, enabling surgeons to achieve substantially better outcomes for a wide array of procedures including correction of craniosynostosis. A variety of new procedures were pioneered. These included subtemporal decompressions on one or both sides, elevation of large bone flaps, circular craniectomies, removal of several plaques of bone, creation of artificial sutures, and morcellation of the entire calvarium [14].

In 1967, Paul Tessier published his experience remodeling and repositioning the frontal bone and orbits and established surgery as the acceptable treatment for craniosynostosis with a focus on achieving high-quality cosmetic outcomes [15]. Different techniques were developed in the 1970s [16].

In 1982, the premier focus on cosmetic improvement as a driving force for surgery was altered. At this time, Renier and colleagues published their landmark report on the association between untreated craniosynostosis and elevated intracranial pressure (ICP). They demonstrated that one third of patients with untreated multi-suture synostosis had intracranial hypertension (IC-HTN) and an inverse relationship between ICP and cognitive status, effectively highlighting the link between skull deformity and the non-cosmetic consequences [17]. Since then, the correction of the skull deformity has been done not only for the purpose of cosmetic improvement and prevention of negative psychosocial consequences, but for the prevention of potentially developing intracranial hypertension and the concomitant brain injury resulting from structural and physiological connectivity associated impairments [18].

Beyond Renier’s work, seven additional studies with postoperative measurement of ICP with invasive monitors have been published, including a total of 762 patients. These studies have shown that 5 and 4% of patients operated on for sagittal or all other types of simple craniosynostosis respectively had persistent intracranial hypertension of >15 mmHg after surgery. In addition, a recent study of 10 patients older than three years of age with non-treated craniosynostosis revealed high rates of debilitating headaches, developmental delays, head shape anomalies, and comorbid Chiari malformations [19].

Re-emergence of the suturectomy

Surgery is the indisputably warranted treatment option for patients with craniosynostosis. However, with multiple surgical techniques available for repair, the selection of appropriate surgical technique remains hotly debated. Significant factors tipping the scales in favor of one technique over another include ideal age at the time of surgery, length of the surgery, surgery-related blood loss and need for blood transfusion, length of postoperative hospital stay, morbidly, mortality, endurance of the
reconstruction over time, and cost-effectiveness.

As mentioned, open cranial vault reconstruction has been the most popular and accepted method employed for the treatment of craniosynostosis since the suturectomies were abandoned. However, during the last two decades, there has been a growing trend to revive the more minimalistic repair surgeries, where only the fused suture is removed for cases of non-syndromic craniosynostosis. One of the primary concerns that prompted surgeons to think about alternatives was the high rate of reoperation required over time with open cranial vault reconstruction.

In 1979, McCarthy and colleagues first reported a 13.5% rate of reoperation for simple craniosynostosis and 36.8% revision rate for complex craniosynostosis [20]. Surgeons then learned to over-correct the reconstructions in order to compensate for the subsequent lagging growth of the craniotomized bone fragments, and the frequency of required reoperation dropped to 2-7.2% [21,22].

The second look into the historical suturectomy or strip craniectomy was possible secondary to a number of important factors and circumstances such as the advent of new technologies like high speed drills and craniotomes, endoscopes and ultrasonic scalpels, in tandem with improved pediatric anesthesia, and intensive care units, increased availability of hemostatic materials and medications, and the innovative utilization of external remodeling devices such as cranial orthosis, springs, and external distractors. Furthermore, evidence pointed at the fact that long-term neuro-cognitive outcomes were better when surgery was performed before 6 months of age [23,24]. So, the classic timing for surgery at ages 6-9 months favored by surgeons for performing a cranial vault reconstruction, was challenged, as it was also well established that when open cranial vault reconstructions were performed before the age of 6 months, rates of surgical revisions and blood loss related morbidity and mortality increased [25].

**Suturectomy with cranial orthosis**

In 1987, Ham and Meyer reported the first case where a skull molding cap was used in conjunction with craniosynostosis surgery [26]. Then, in 1998, Jimenez and Barone developed and promoted the endoscopic technique for performing a strip craniectomy of fused sutures followed by a period of external remodeling with a cranial orthosis. Jimenez and Barone [27], first performed the procedure for patients younger than 3 months, and provided the foundational platform for the advent and adaptation of various other modified techniques, which have trended towards further minimalism, and sparing the use of the endoscope by some surgeons.

Specific to the sagittal craniosynostosis, authors have analyzed whether the addition of lateral barrel-stave osteotomies to the strip craniectomy improved morphological outcomes, and showed that similar excellent outcomes were achieved without them and therefore they are not warranted in surgery [28]. The suturectomy width has also been studied and the conclusion has been that narrower, 2 cm sagittal suturectomies, produced nearly identical clinical results as wider, 4-6 cm suturectomies with barrel stave osteotomies, while requiring fewer procedural steps and significantly less operative time [29]. Compared to posterior cranial vault reconstructions for sagittal craniosynostosis, suturectomies assisted with cranial orthosis have been reported to yield comparable enduring improvements in cranial volume and shape, and to reduce operative times to as low as 46.3 minutes with a corresponding mean length of stay as short as 1.35 days [30]. As with sagittal craniosynostosis, endoscopic suturectomy has successfully treated the majority of patients with uni or bilateral coronal and metopic craniosynostosis, with equal short procedure times of around 56-73 minutes, low blood loss volumes (36-40 ml) with no intraoperative blood transfusions and brief hospital stays (mean 1.2 days) [22,23,31]. On the whole, there is sufficient data to support the notion that sagittal, coronal and metopic craniosynostosis can be safely and effectively treated with suturectomy assisted with postoperative cranial orthosis therapy.

Compared to CVRs, the minimally invasive suturectomy technique with postoperative orthosis has been proven to be associated with less operative blood loss (30ml or 5% vs 25-500% of estimated blood volume), shorter length of surgery (1 vs 4-7 hours), reduced postoperative lengths of hospital stay (1 vs 4-7 days) and higher surgical endurance with lower rates of needing repeated reconstructive surgery (2% vs 5%) [27,30,32,33]. This has been our personal experience as well when performing both procedures. In addition, as opposed to the open cranial vault reconstructions, where the growth of the craniotomized bones never parallels that of the non-cut skull, and thus, the need for overcorrection, the bones in children undergoing minimal invasive techniques grow in parallel and in a more physiological way to the rest of the skull.

From an economic perspective, the endoscopic suturectomy, assisted with postoperative cranial orthosis, has been reported to reduce the cost (and morbidity) when compared with traditional open cranial vault remodeling (CVR) procedures, incurring in less costs in nearly all categories, including overall 1-year costs, physician services, hospital services, supplies/equipment, medications/intravenous fluids, and laboratory and blood bank services, despite having higher immediate postoperative costs because of the cost associated with orthotic services and indirect patient costs for travel and lost work. One-year median costs were $55,121 for CVR and $23,377 for endoscopic suturectomy with orthosis [34,35].

Whereas suturectomy has been demonstrated to be effective and enduring for most types of craniosynostosis, including more recently some syndromic cases as long as the external remodeling device is used, standalone suturectomies without postoperative remodeling assistance have been shown to be inefficient and insufficient [31-34].

**Spring-assisted cranioplasty**

Spring-assisted cranioplasty represents an additional surgical technique employed for the treatment of craniosynostosis.
Spring-assisted cranioplasty was initially performed by Lauritzen in 1997 [36], following demonstrated success in a rabbit model by Persing and colleagues [37]. This technique involves a suturectomy followed by placement of specialized tension springs across the osteotomy sites and potential placement of compressive springs along areas of compensatory growth. The implanted springs are usually maintained in place for a period of 4 to 7 months after the operation and then require a second surgery to remove them [38].

Advantages of spring-assisted cranioplasty as opposed to open cranial repair include its minimally invasive nature, reduced morbidity and hospital stay. Potential drawbacks, especially when compared to suturectomy assisted with a head orthosis, include the need for a second procedure for removal of the springs, longer surgical times (97-215 minutes) and length of stay in the hospital (1-2 days), higher blood loss during surgery, larger incisions, and slightly higher complication rate [39].

Rodgers and colleagues presented a single institution experience of 100 consecutive cases using a novel spring design. The series documented a 9% transfusion rate, 2% rate of CSF leak, and 1% rate of spring-related infection. Moreover, 5% required further calvarial remodeling surgery [40]. This requirement for repeat surgery is more than double the 2% rate published by Han and colleagues for 144 non-syndromic sagittal synostosis patients who underwent endoscopic suturectomy with post-operative orthosis [41]. Hence, these data tend to favor suturectomy with orthosis over spring-assisted cranioplasty. However, fewer studies exist in the literature for the spring-assisted technique and the future publication of additional cases will enable a more comprehensive comparison between the two techniques.

Distraction osteogenesis

Similar to spring-assisted cranioplasty, distraction osteogenesis represents a third venue of minimally invasive treatment techniques for craniosynostosis. This technique involves osteotomies at the sites of synostosed sutures and placement of internal or external distraction devices. The distraction devices are activated 3 to 5 days post-operatively for several weeks until the desired is achieved. The devices are then removed after an additional consolidative period of 2-3 weeks [42,43]. Given the scarcity of large studies or reports within the literature, it is not yet possible to adequately compare this minimally invasive technique to the others or to open cranial vault reconstruction in terms of efficacy or safety.

DISCUSSION

Proponents of reconstructive surgery argue that open surgery gives the surgeon the most control over the mechanics of repair, and popular objections to suturectomy assisted with an external remodeling device such as the head orthosis include the presumption that molding helmets are overly burdensome for the treated child and family. Delve and colleagues effectively dismissed this presumption, demonstrating that, for a series of 111 consecutive cases of endoscopic suturectomy followed by orthosis therapy, the procedure was associated with satisfying cosmetic results and the burden of the helmet therapy for the child and family was deemed very low [44,45].

Despite the described advantages of the minimally invasive suturectomy with postoperative orthosis, open cranial vault remodeling is still widely practiced and needed as many children are diagnosed with craniosynostosis at older ages, after the age of 4-6 months, precluding offering the suturectomy surgery; in addition, resources and available timely, non-urgent operating time are not at the reach of many surgeons and patients, even if diagnosed promptly.

One study, pooling the preferences of 180 pediatric neurosurgeons and 102 craniofacial plastic surgeons demonstrate that the suturectomy with remodeling assistance has not been fully adopted yet by the community for the treatment of non-syndromic craniosynostosis [46]. However, as more data on outcomes associated with suturectomy and remodeling orthosis come to the surface, the minimally invasive technique is likely to increase in popularity among providers.

We believe that the existing literature clearly favors suturectomies assisted with cranial orthosis over CVRs in all analyzed aspects, whereas related to morbidity and mortality [47], to endurance, need for blood transfusions, length of surgery and hospital stay and overall costs, and thus, we think that when the diagnosis of craniosynostosis is done early, and the surgical availability and resources are at hand, it should be considered the standard of care. Future work should be focused on whether this minimal invasive technique can be recommended for older infants between the ages of 6 and 12 months old.

CONCLUSION

The minimally invasive suturectomy technique assisted with postoperative external cranial remodeling for the treatment of craniosynostosis has clear benefits for young infants over open reconstructive surgery. When and wherever the conditions and resources are available, it should be considered the standard of care.

REFERENCES


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