A Functional Aesthetic Approach to Correcting the Sequelae of Sagittal Synostosis

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Abstract

Keywords

- craniosynostosis
- occipital
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- temporal

Sagittal synostosis is the most commonly treated form of craniosynostosis exhibiting an incidence of 1:5000 births.¹ Early closure of the sagittal suture is the cause of the classic phenotypic presentation of an elongated head, which is characterized by a narrow and low occiput, a saddle-shaped parietal region, and noticeable frontal bossing. Early surgical intervention is the treatment of choice and surgical options range from simple suture excision to total cranial vault remodeling. Noting that a significant proportion of patients present with limited frontal bossing and asymmetry that is more pronounced in the posterior cranium, the author developed the functional aesthetic approach to reconstruction. This operative technique addresses the changes noted in the posterior two-thirds of the cranial vault, completely correcting the sequelae associated with early closure of the sagittal suture. As an added advantage, the reconstructive paradigm is designed to create a dynamic temporal-occipital component that will activate when a child rests in the supine position. This enhances widening of the posterior cranium thus improving long-term results.

Sagittal synostosis is the most commonly treated form of early suture closure noted in the growing infant exhibiting an incidence of 1:5000 births.¹ In general, the suture should be highly active through the first 22 to 24 months of life and any diminution in the growth capacity during this early period of rapid brain growth can and will have an effect on cranial shape and size. Although mechanical stress and genetic factors associated with mutations affecting fibroblast growth factor receptors (FGFRs) 1, 2, and 3² have been identified as possible contributors to early sutural closure, a singular cause of early sutural closure has yet to be identified. Unfortunately, the ability to manipulate FGFRs and/or manage the genetic factors that contribute to the demise of sutural growth is beyond our current capabilities. This leaves the treating surgeon with the task of fixing the result of the disease rather than addressing the cause. In this setting, it is imperative that the operating surgeon understands the full nature of the anomaly and visualizes the anatomical norm that should have been. The functional aesthetic approach presented herein is

one surgeon's attempt to restore the scaphocephalic cranium to its anatomical baseline.

Clinical Presentation of Scaphocephaly

Sagittal synostosis, like any of the synostotic phenomenon, is a process in which a cranial growth center is compromised in its ability to allow for further cranial growth in a direction perpendicular to the axis of the affected suture. The sagittal suture is the longest suture of the cranium, extending from the anterior fontanel to the confluence of the lambdoid sutures in the occipital region. This length creates a unique circumstance with respect to development of cranial vault asymmetries. Historical review of the clinical and computed tomography (CT) data of my last 70 patients indicates that most infants with sagittal synostosis will present with anomalies that have a predilection for being more pronounced in either the posterior or anterior cranium. I theorize, and radiographs seem to support, that early closure of the sagittal

Fig. 1 The classic appearance of a scaphocephalic cranium. The anterior cranium demonstrates frontal bossing while the midvault is saddle-shaped. The posterior cranium is remarkable for its lack of height and narrow, anvil-like shape.

suture starts in the area of the more severe cranial deformity and then progresses either anteriorly or posteriorly. One may liken the progression to a zipper closing with the most severely affected segment reflecting the starting point of the zipper effect. In my practice, I have noted that in greater than 52% of cases with early closure of the sagittal suture there is a propensity for closing in a back to front direction with the last closure spot being the anterior fontanel. This leads to a clinical presentation in which the child's cranial anomalies are more severe posteriorly than anteriorly. The anterior cranium may be rounded or minimally bossed, while the occiput takes on an anvil shape and the posterior skull is found to be low and narrow (►Fig. 1). As the sutural closure progresses through the midvault one sees the development of the characteristic saddle-shape deformity. In this setting (posterior to anterior closure pattern), it is the severity of the posterior changes that drives the methodology of the repair.

Clinical Management and Indications for Correction

Cranial growth is predicated on brain growth. Standard growth charts estimate that 70% to 80% of head growth occurs in the first 18 months of life.³ Thus, any factor that disrupts normal cranial growth during this critical time period will have a much more profound effect on final head shape than any other cranial growth disturbance.

The deformity associated with any synostotic process is a result of two distinct issues. First, there is the failure of growth and development that leaves the effected suture in a veritable state of growth stasis. Whatever day or moment that the suture stops growing reflects the final point of development for that particular spot on the skull. In essence, if the suture stops growing at 2 months of age, then that child is destined to have that cranial anatomy remain at the 2-month-old size. This constitutes the first anatomical problem the craniofacial surgeon faces. In developing a plan, he or

she must consider the restoration of the cranium to an age-appropriate size. As such, the surgical paradigm must contain a plan for "catch-up growth."

The second issue encountered when a suture closes is diametrically opposed to the first. Although closure of a suture will limit growth at the point of closure, compensatory overactivity by the remaining patent sutures leads to the secondary deformities that are characteristic of scaphocephalic skull. When evaluating these changes, it is helpful if the surgeon considers the directional growth potential of the remaining sutures. When this is done, it becomes readily apparent as to why the scaphocephalic head is elongated. The cause of this anomalous shape is the coronal orientation of the majority of the remaining active sutures: the coronals and the lambdoids. The metopic has a limited ability to add to the compensation-based growth deformity for two reasons. First, it is a small and rather brief suture, and second it is designed to close earlier than the rest of the sutures and as such its contributions with regards to compensatory growth are limited.⁴

Understanding the Deformity

Having discussed the factors that create the skull anomalies associated with sagittal synostosis, it is prudent to assess the effect of these divergent processes. As previously stated, it is my observation that a significant number of sagittal sutures begin their premature closure at the junction of the sagittal suture and the lambdoid sutures in the parieto-occipital region. The closure then marches forward leading to a presentation in which the posterior two-thirds of the skull is more severely affected than the anterior cranium (►Fig. 2). This, in turn, leads to the characteristic head shape described earlier in this treatise. In this form of sagittal synostosis with scaphocephaly, there are four issues that must be addressed if the surgeon is to create a symmetric and anatomical three-dimensional cranial shape (►Fig. 3).

Fig. 2 A 3-dimensional computed tomography scan demonstrating sagittal suture closure and a scaphocephalic cranial shape. Note the extreme narrowness of the occipital region and the ridging of the sagittal suture. Bony lacunae are present posteriorly, indicating that this region of the skull has been subject to intense cerebral pressure over time. The anterior cranium is broad and lacks lacuna. These findings are physiologically consistent with the posterior to anterior “zipper closure theory.”

1. The normally broad occipital region is narrowed, assuming an “anvil” or cone shape. The occiput is longer in the anteroposterior (AP) plane than a standard infant skull and this creates a posterior shelf that protrudes over the neck. The posterior cranial bone is positioned lower than is anatomically correct when referenced to a horizontal plane based on the positioning of the mid- and/or anterior cranium. The surgical plan designed to ameliorate this

Fig. 3 Lateral photo highlighting the anatomical issues that are characteristic of posterior to anterior closure of the sagittal suture. (1) The narrow, anvil-shaped occiput. (2) The lack of bioccipital prominences. (3) Saddle-shaped midvault. (4) Frontal bossing is absent.

anomaly requires a shortening of the AP length, a flattening and widening of the occipital cone, and a means of elevating the occiput to a height that better matches the horizontal norm created by the anterior cranium.

2. The natural front to back widening of the skull is absent. The cranium gets increasingly narrow as it progresses in a posterior direction. The bioccipital prominences, usually the highest and widest points on the head, are low and narrow. Intervention must be designed to restore both the height and width of the bioccipital prominences so that they assume a slightly wider and more elevated position as compared with the anterior skull. This widening must match seamlessly with the changes that will be made to the anvil-shaped occiput.
3. The midcranium, directly over the sagittal suture, has its own unique anatomical pathology. In this area, the major deformity is related to the scooped or saddle-shaped appearance of the central cranium. There is decreasing height and increased narrowing to the parietal skull when referenced to standard AP cranial development. The width and height issues are addressed according to number 2 above, whereas the scalloped midvault requires an intervention that allows for flattening of the saddle shape.
4. If sagittal synostosis is identified and treated early on its course, the anterior cranium may show little ill effect from the forces driving compensatory growth. However, there are cases that present late in the disease process and there are those cases that follow a suture-closure pattern that demonstrates an anterior to posterior progression. In these cases, compensatory growth associated with the anterior cranium leads to frontal bossing and temporal narrowing. If the presentation is moderate in severity, then posterior release and the utilization of a molding helmet may be sufficient therapy. However, severe cases warrant a standard frontal cranial vault remodeling focused on widening the temporal region and flattening the bulbous frontal bone.

Reconstruction

A multitude of reconstructive paradigms have been applied to remedy the scaphocephalic skull shape that arises from sagittal synostosis. These include strip craniectomy,^{5,6} strip craniectomy with morselization, endoscopic suture release⁷⁻⁹ with head-banding/helmeting, and suture removal followed by direct reshaping via osteotomy and bone manipulation.¹⁰⁻¹³ Recently, new approaches have endeavored to reshape the cranium through the application of active forces using distraction or spring-assist devices.¹⁴⁻¹⁷ Each approach has its advocates and though many yield satisfactory results, the diversity of approaches indicates that controversy still swirls around the question as to what constitutes the best approach. Clearly, every surgeon should and must factor in the age of the patient and the severity of the deformity when he or she is selecting the most appropriate treatment pathway. If a child is below the age of 4 months and the cranial deformity is mild to moderate, then minimally invasive techniques may be suitable to correct the pathology at

hand. Outside of these limited parameters, it is my opinion that a child presenting with scaphocephaly is best served by an open procedure in which planned osteotomies and direct reconstruction of the cranial form are employed. To this end, over the past 16 years I have utilized a functional aesthetic approach to cranial vault reconstruction in those patients who present with scaphocephaly secondary to the posterior to anterior “sagittal zipper” suture-closing pattern. This technique has consistently yielded superior aesthetic results and high rates of patient satisfaction.

The Functional Aesthetic Approach to Manage Scaphocephaly

Having been an early advocate of the single-stage, total cranial vault (TCV) remodeling approach in the management of sagittal synostosis, I have been able to review the success and failure of this “one and done” approach to the scaphocephalic head. It is noteworthy that TCV results were generally good, but it is clear that there are limits to the “total” nature of this approach. By definition, positioning of the patient for a TCV will necessarily limit the surgeon’s ability to comprehensively expose and reshape the entire cranium. In essence, the ability to reshape the frontal bandeau will limit the amount of posterior exposure/intervention and vice-versa. Some centers have tried to overcome this limitation by performing a single-sitting two-stage procedure where one half of the vault is corrected, after which the patient is turned, repped/draped and the other section of skull is addressed.¹⁸ We have not pursued this approach due

to the perceived increase in operative risk associated with the addition of a second stage. These risks include increased blood loss, coagulopathy due to fluid replacement and cooling, and the specter of postoperative infection.

Understanding the previously outlined issues associated with a total cranial vault remodeling, a two-step approach was developed for the management of sagittal synostosis. In stage 1, the most severely deformed portion of the cranium (commonly the posterior two-thirds of the skull) undergoes open reconstruction after which any secondary interventions are addressed on a case by case basis.

Operative Technique

The patient is placed in a prone position on an outrigger headholder and care is taken to protect the eyes and to secure the endotracheal tube. The proposed incision line is injected with 1% Lidocaine with epinephrine 1:100,000, and a standard zig-zag incision is made through the scalp. Cautery is used to cut through the galea and a scalp flap is turned to the level of the lambdoid sutures where the pericranium is incised. At this point, the pericranium is scored in the midline and laterally based pericranial flaps are created. The posterior scalp and retro lambdoid pericranium is taken down to a suboccipital level and dissection is stopped at the point where the neck musculature becomes apparent. This is generally below the lateral sinuses. With exposure complete, a trapezoid is drawn outlining the parietal bone to be removed by the neurosurgeon. The trapezoid is shaped such that the anterior portion is wider than the posterior portion of bone. The

AB= Posterior-Lateral Triangles

Fig. 4 The parietal bones are removed in a trapezoidal shape after which two triangles are removed from the posterior lateral aspect of the temporal bone. The anterior boundary of the triangles is the point where the parietal bone turns downward (A). The posterior boundary is the lambdoid suture (B).

Fig. 5 After the parietal bone is removed, it is step cut and expanded to fit the new construct. The triangular osteotomies and the C-shaped occipital bone flap are delineated. The inferior occipital tang is left as the base for reconstruction.

design is predicated upon the concept that the posterior temporal bone and the residual parietal bone left attached to the posterior temporal bone will serve as the osseous framework needed to raise this portion of the skull upwards as we work to create the appropriate height of the bioccipital prominences. According to plan, extra bone that is left in situ will be bent from horizontal to vertical in an effort to balance anterior/posterior height discrepancies. The trapezoidal pa-

rietal bone flap extends from the coronal sutures to the lambdoid suture. With the parietal bone removed, the neurosurgeon has a clear view under the occiput and into the lateral temporal areas where the transverse sinus resides (> Fig. 4). The next key planning point involves the identification of the two triangular wedges that will be removed from the posterolateral temporal bone. I begin by noting where the posterior skull begins to narrow and angle

Fig. 6 After removal the C-shaped occipital bone, the flap is rotated 180 degrees after which it is attached to the occipital tang and the posterior temporal bone in such a fashion as to widen the occipital form. The triangular flaps will be rotated 60 degrees and inset below the C-shaped flap. Note the relaxing scroll osteotomy that facilitates temporal bone widening.

Fig. 7 The C-shaped occipital flap in place, holding the temporal bones in a wider, more raised position.

inferiorly. This is a critical point in that it represents the narrowest point of the harvested parietal bone flap and as such the future bioccipital prominence. A vertical osteotomy from this point to a point posterior to the mastoid at the junction of the squamosal and lambdoid suture frees the anterior portion of the temporal bone triangle. The posterior margin of the triangle is denoted by the lambdoid suture (► **Figs. 4 and 5**). The suture is dissected under direct vision and then the triangles are removed using a saw or bone scissors. The space created by the removal of these triangles will allow the surgeon to foreshorten the skull and correct the inappropriate length that is characteristic of scaphocephaly.

The occipital bone is dissected from the underlying dura. Care is taken to safely manage the transverse sinuses and the confluens sinus. A C-shaped osteotomy is designed to remove the occipital bone. The anterior portion of the C is denoted by the lambdoid sutures and the inner void of the C is due to the designed approach in which a tongue of centrally located occipital bone is left behind as the mounting plate for the reconstruction. The lower arms of the C arise as straight line osteotomies that run parallel to the cranial base from the retro-mastoid region to the central tongue of bone. The entire occiput is taken off as a single unit. With the main osteotomies complete, relaxing osteotomies are performed on the in situ temporal bone (► **Fig. 6**). Symmetric, bilateral scroll type osteotomies are made in the posterior temporal bone to allow the temporal bone to flair outward. This gives new width to the previously narrowed occipital region. If the temporal bone does not flair easily (minimal digital pressure) then short vertical osteotomies are made just posterior to the coronal sutures. Generally, this added relaxation will be sufficient to generate supple temporal bone mobility.

Reconstruction begins with careful manipulation of the bony elements using a Tessier bone bender. First, the in situ temporal bone is coaxed to a more lateral position and the posterior temporal parietal area is recurved into a more vertical position. This sets the highest posterior point or the bioccipital prominence. The bone benders are then used to flatten the occipital C. This effectively removes the anvil cone and widens the arms of the C. In thicker bone, small

Fig. 8 Digital pressure applied to the occipital construct demonstrates the functional/dynamic capabilities of the operative technique. Note the difference in the ruler markings from ► **Figure 7**. With pressure the occiput widens almost 2 cm more than baseline.

incomplete relaxing osteotomies may be needed to diminish the risk of fracture and facilitate bending. The occipital bone is then turned 180 degrees and attached to the in situ occipital tang that serves as the base of the reconstruction. KLS resorbable plates and rivets (KLS-Martin, Jacksonville, FL) have greatly enhanced the stability of this construct, but one may use a 2.0 PDS resorbable suture (Ethicon, Somerville, NJ) in a figure-of-8 fashion if plates are not available. It is important to note that at times, the occipital tang may assume a posteriorly angled position. Careful use of Tessier bone benders will allow for increased AP shortening via anterior repositioning of this osseous segment. With the inferior portion of the occipital bone in place, the arms of the C are now attached to the temporal bone using KLS resorbable plates or 2.0 PDS sutures. Having relaxed the temporal bone and flattened the occipital bone, the joining of these two osseous segments effectively flairs the posterior aspect of the temporal bone thus widening the posterior cranium. This

Fig. 9 The completed reconstruction.

Fig. 10 A vertex view of the reconstruction at the end of the procedure.

Fig. 12 Vertex view 3 months postoperatively. The occiput is broad and round. The anteroposterior length of the head is aesthetically normal.

maneuver recreates the normal cranial aesthetic and creates a dynamic functional reshaping mechanism that will serve to improve shape and width over time. This dynamic functionality arises due to the fact that any future pressures placed on the occiput (i.e., when a child is at rest) will cause the occiput to push forward. This in turn will cause the arms of the C and the posterior elements of the temporal bone to flair, further widening the posterior cranium (►Figs. 7 and 8).

With the occiput situated in a new foreshortened and elevated position, it is notable that the space previously occupied by the two lateral temporal triangles is no longer present in the upper temporal parietal skull area. Instead, two spaces have been created inferior and posterior to the arms of the occipital bone. The triangle remnants that had been previously removed are brought to the table and trimmed to fit the defects. Inset is accomplished using 2.0 PDS resorbable sutures.

The final step in the reconstruction consists of shortening the AP length of the parietal bone and then step-cutting and widening the bone so that it will fit the new construct. The tongue-in-groove step cut is created in an anterior to posterior line. Usually, one or two tongues will suffice. Once the appropriate width is obtained, the parietal bone is inset and the tongue and groove expansions are maintained at an

appropriate width utilizing 2.0 PDS sutures (►Fig. 9). The reconstruction complete, the pericranial flaps are resuspended. The entire construct is then irrigated with antibiotic impregnated saline and a two-layered closure is achieved (►Figs. 10 and 11).

A word about the anterior cranium: In children with moderate to severe frontal bossing and/or bitemporal narrowing, I take the time to educate parents with respect to the potential need for a second surgical intervention. They understand that a second procedure would be used to address any pronounced frontal cranial anomalies. Recently, in an effort to obviate the need for a second intervention, I have utilized a postoperative molding helmet as a means of modifying forehead shape. Enlisting the skills of my orthotist colleagues, a series of patients with moderate frontal deformity have been placed in a postoperative molding helmet for 3 months. All patients in the series demonstrated a positive response to therapy and none warranted further surgical intervention. In the now rare case that a second surgery is required, it is usually performed 3 to 6 months after the first intervention. A standard frontal cranial vault remodeling with widening of the bandeau at the temples accompanied by flattening and derotation of the frontal bone is performed. This is done in an effort to compensate

Fig. 11 Side-by-side later images preoperative versus postoperative. Note the rounding and increased bioccipital height.

Fig. 13 Side-by-side lateral postoperative images. The anteroposterior length has been reduced and the anvil-shape of the occiput has been corrected.

for the bulbous forehead and temporal hollowing that is characteristic of the anterior changes associated with advanced scaphocephaly.

Conclusion

All surgical interventions must be patient/problem specific, thus no one solution will fit every case. However, as the field of craniofacial surgery and specifically the management of craniosynostosis continue to advance, it behooves the practitioner to entertain surgical solutions that effectively restore and enhance form and function. With a baseline aesthetic predicated on “normal,” a successful outcome is one in which the patient and the family feel that the final result yields a head shape that is not discernibly different from any of the child’s surrounding peers (► **Figs. 12 and 13**). The functional aesthetic approach with its anatomical reconstruction and its functionally adaptive capabilities offers the craniofacial surgeon an effective, reproducible means by which he or she can deliver consistently excellent results that will enhance the physical, psychological, and social development in children who present with sagittal synostosis.

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