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Atlas of the
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Preface

Ramon L. Ruiz, DMD, MD Jogi V. Pattisapu, MD
Guest Editors

Craniomaxillofacial surgery has advanced significantly in recent years, due to contributions from various experts in the field. We have learned much from each other in caring for patients with complex facial disorders. In this volume of the *Atlas of the Oral/Maxillofacial Surgery Clinics of North America*, we compiled several concepts and approaches to the surgical management in a variety of situations. The authors have dedicated themselves to improving the techniques and have offered their expertise by sharing from their collective fund of knowledge.

Surgical techniques have been advanced over the past century in an effort to solve problems affecting the complex anatomy of the craniomaxillofacial region. Maneuvers such as the application of rigid internal fixation for facial fractures, the creation of osteotomies for repositioning of skeletal segments, and the use of bone grafts for reconstruction of defects were developed for the treatment of conditions affecting the lower face. These original techniques of maxillofacial surgery were subsequently applied to correct congenital deformities and injuries of the upper face, orbit, and cranial vault and the subspecialty of craniofacial surgery was created. This close collaboration between the craniomaxillofacial surgeon and neurosurgeon allows safe transcranial access to the upper face and allowed us to treat congenital deformities or complex injuries, yielding better outcomes. In addition, modifications in craniomaxillofacial techniques have offered new opportunities in managing complicated skull base and intracranial pathologies. The combined approaches have improved our ability to reach various anatomical regions and increased our effectiveness while decreasing associated morbidity.

We hope this issue offers the reader new ideas and surgical approaches, with vivid examples of variations and the indications for them in caring for these individuals. The editors are grateful to the many authors who, despite their own heavy professional commitments, spared their time, efforts, and energies to submit the articles on time. We are conscious of the time factor and delays that arise in a multi-authored publication. To those who submitted articles promptly, we offer our apologies for such delays.

We have invited a wide range of experts from Neurosurgery, Oral and Maxillofacial Surgery, Otolaryngology/Head and Neck Surgery, and Plastic and Reconstructive Surgery to present "state-of-the-art" reviews on each of the important aspects of procedures involving a combined craniomaxillofacial and neurosurgical approach. The authors have contributed key topics covering the spectrum of craniomaxillofacial surgery, which reflects current standards in the field. Recent progress over the years has been given more importance, and it is inevitable that some concepts may soon be outdated. Every effort has been made to keep these to a minimum. It is also inevitable that some author's opinion may differ from others, along with overlap between articles. Rather than avoid this, the editors feel it is more profitable for the readers to have available both differing and converging viewpoints.

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The team approach to such complex issues is reflected in these articles, and most centers of excellence use such a system to deliver the best care for patients. We sincerely hope this publication will be an important asset to those caring for these disorders.

As editors of this volume, we have been intrigued by the growing number of opportunities to treat complex craniomaxillofacial, skull base, and intracranial problems that have resulted from the close collaboration between our specialties. At our own institution, patients benefit from the synergy of an interdisciplinary surgical team delivering coordinated care. Collaboration between our craniomaxillofacial and neurosurgical services has proven invaluable in establishing evidence-based treatment philosophies, evaluating rationales for surgical intervention, and formulating well-thought-out surgical treatment plans for the benefit of our patients.

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Atlas of the Oral and Maxillofacial Surgery Clinics

The Coronal Scalp Flap: Surgical Technique

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Use of incisions that minimize visible scars is an important principle of craniomaxillofacial surgery. To accomplish this goal, the coronal incision and scalp flap are frequently used during combined craniomaxillofacial and neurosurgical procedures.

Prior descriptions of the coronal scalp flap include an incision just behind the hairline with the lateral components of the approach created in the pre-auricular areas: the hairline of the patient being a primary consideration in the placement of the incision for the flap. Although anterior extension of the midportion of the coronal flap was considered to enhance flap retraction and access to the midface, the resulting scar may subsequently become obvious with male pattern baldness. Over time, this scalp flap technique has been refined with the introduction of a postauricular location. An important technical point is for the surgeon to understand that a more posterior placement of the incision will not limit access to the orbital structures. The amount of exposure of the cranio-orbital skeleton is related proportionately to the inferior extent of the incision and not necessarily to a more anterior position. The incision is placed from one auricular area to the other and the degree of skeletal exposure required for a given procedure dictates the inferior extent of the incision. When access to the zygoma and infraorbital rims is necessary, the incisions must be extended further inferiorly. The incision is outlined within the postauricular region to more effectively conceal the postsurgical scar. The authors' preference is to place the incision behind the top of the vertex of the head rather than carrying it toward the forehead (Fig. 1). The use of a postauricular coronal incision eliminates visible scars in the preauricular area and decreases risk to the frontal branch of the facial nerve in reoperated patients. The postauricular placement of the incision is also desirable in children because of the tendency for the coronal incision scar to migrate forward with subsequent ongoing growth of the cranial vault.

When secondary operations are performed in cases where a previous coronal scalp flap has been used, it is preferable to reincise thru the original scar. Although it may be tempting to place the incision in a different location, consideration must be given to the effect of the previous scar on flap perfusion and wound healing.

Use of a zigzag (Stealth modification) incision avoids a straight-line scar and is particularly useful in camouflaging the scar within the surrounding hair-bearing scalp. The authors favor a variation of this technique using a curvilinear incision design. Unless meticulous attention is paid to hemostasis during the initial dissection, additional blood loss associated with a greater incision length may offset the benefit.

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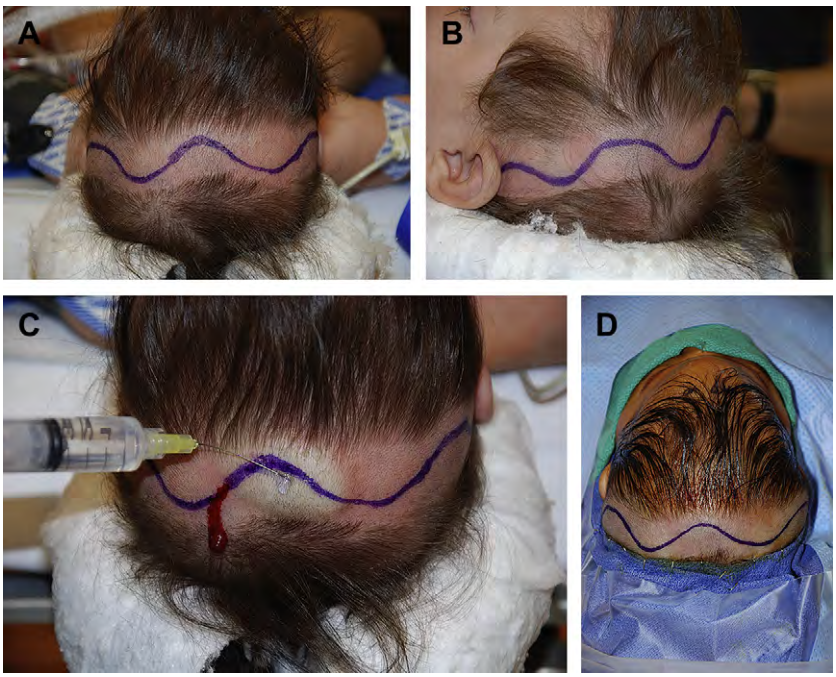

Fig. 1. (*A*–*D*) The patient is positioned on a Mayfield horseshoe-shaped headrest. In patients with longer hair, a strip of hair may be clipped along the proposed incision line. The authors prefer a curvilinear incision design placed within the postauricular area, which conceals the residual scar most effectively. The face and head are prepped with providone iodine and draped in sterile fashion.

Surgical technique

The patient's head is positioned on a Mayfield horseshoe-shaped headrest. Removal of hair is not mandatory. The authors will frequently clip a small strip of hair, approximately 1.5 cm in width along the location of the coronal scalp incision (see Fig. 1). This makes the initial dissection and subsequent closure easier with less interference from longer hair. In infants, no hair removal is necessary. When

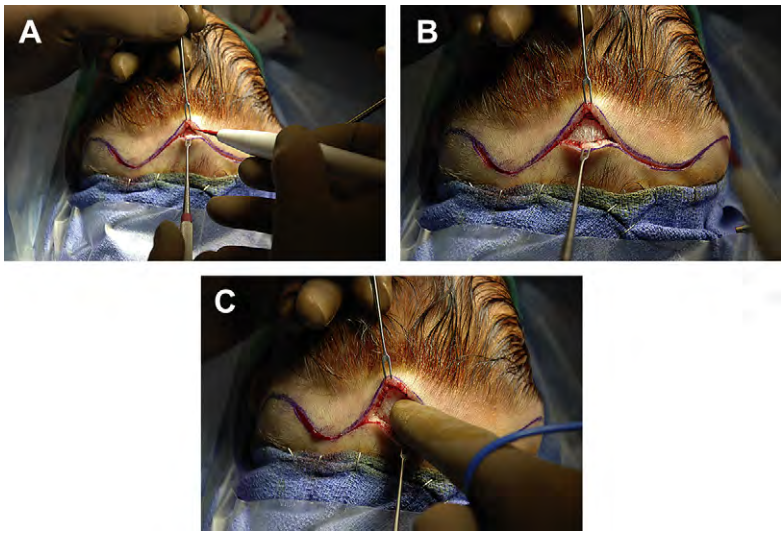

Fig. 2. (*A*) The initial incision is created with a #15 surgical blade and is carried thru skin and dermis. Double-ended skin hooks are then placed and the Colorado Needle (Stryker, Kalamazoo, MI, USA) monopolar electrocautery set on low energy (cut 8, coag 10) is then used to divide the subcutaneous tissues and galea aponeurotica. (*B*) Superior and outward traction is applied to the skin flaps using the skin hooks and the loose connective tissue beneath the galea is encountered. (*C*) Gentle finger dissection can be used to divide the loose connective tissue, exposing the intact pericranium and further developing the subgaleal dissection plane.

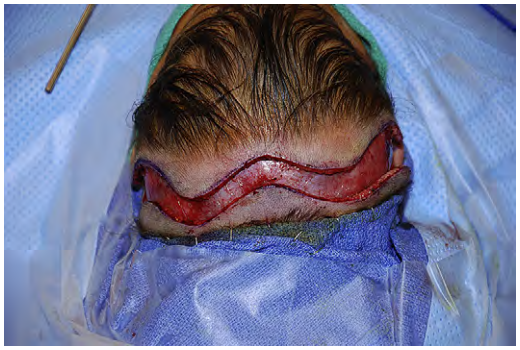

Fig. 3. Completed coronal scalp incision and initial flap development. Dissection thru the skin and galea is performed using the #15 blade for the initial incision and the fine electrocautery needle to divide the subcutaneous tissues and galea. This process (skin incision, traction using double-ended skin hooks, and fine cautery dissection to divide the subcutaneous tissues through galea) is performed in segments beginning at the vertex of the head and then extending along the right and left temporal regions toward the back of each ear. By completing the initial dissection in small segments, bleeding is easily controlled. Note the minimal blood loss associated with a completed coronal incision and excellent hemostasis without the need for scalp clips.

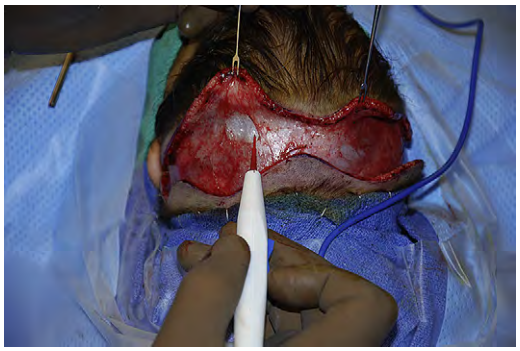

Fig. 4. A skin hook is used and anterior traction on the flap is applied. The monopolar electrocautery can be used to divide subgaleal connective tissue.

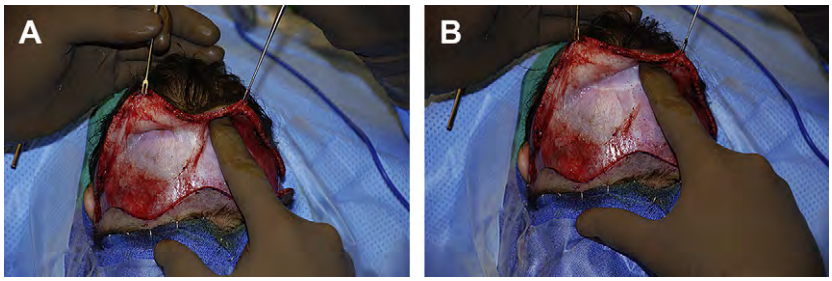

Fig. 5. (*A, B*) Blunt dissection thru the loose areolar connective tissue of the scalp can also be performed manually. Dissection proceeds rapidly and bloodlessly in the subgaleal-suprapericranial plane and the flap is turned anteriorly.

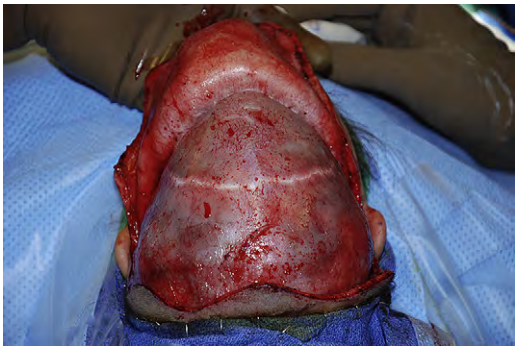

Fig. 6. Elevation of coronal scalp flap in the subgaleal-supraperiosteal plane.

hair removal is performed, it should be done with sterile, disposable clippers. Shaving should be avoided, as it produces increased epithelial damage to the skin and contributes to an increased incidence of infection.

A curvilinear incision is outlined with a surgical marker and the patient's head and face are prepped and draped in sterile fashion. The proposed incision line is injected with a dilute solution of 0.5% lidocaine with 1:200,000 epinephrine to aid with hemostasis. The injection is limited to the subgaleal region along the incision line. This reduces bleeding and helps initiate dissection along the subaponeurotic plane. The scalp has a rich vascular supply and so dissection is performed in segments with meticulous attention to hemostasis. Adequate hemostasis is especially important in infants and young children because of the potential loss of their blood volume. The conventional approach to minimizing blood loss during opening has been to apply hemoclips to the edge of the flap. Bipolar electrocautery is used to obtain hemostasis; this has a minimal effect on the adjacent peripheral hair follicles.

The use of monopolar electrocautery was previously contraindicated because of the increased risk of destroying regional hair follicles, which results in alopecia and a more visible scar. The authors' preferred approach involves the use of a #15 surgical blade to create the initial skin incision. Further dissection through the subcutaneous tissues and galea aponeurotica is performed using the monopolar electrocautery with a fine-tipped Colorado needle on low-energy settings (Cut 8 and Coag 10) (Fig. 2). Because dissection with the Colorado needle is done below the level of the skin and dermis, there is no damage to hair follicles. This technique also dramatically minimizes blood loss and obviates the need for the use of vascular clips (Fig. 3). The authors frequently open an entire coronal scalp flap incision using this technique with less than 5 mL of blood loss.

Once the pericranium is identified, a plane of dissection is established above it. Skin hooks are placed and upward and outward traction is applied to reach the loose areolar connective tissue between the aponeurosis and pericranium. Dissection proceeds rapidly and bloodlessly toward the forehead in this subgaleal-supraperiosteal plane (Figs. 4–6). Finger pressure is often enough to easily continue dissection in this plane.

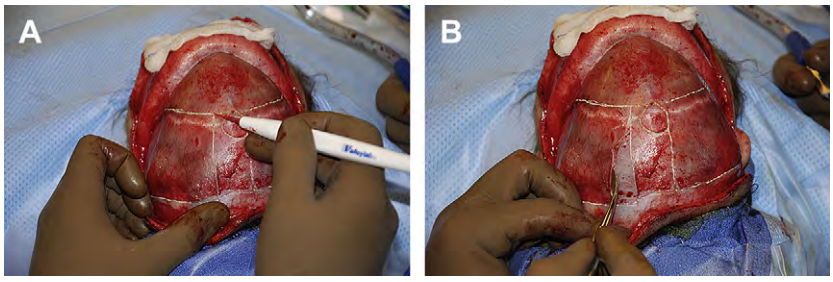

Fig. 7. (*A, B*) Elevation of pericranium as a separate layer. The electrocautery is used to incise the pericranium and an elevator is then used to carry out dissection for exposure of the cranial vault and orbital structures in the subperiosteal plane. Bleeding from venous lakes within the bone is quickly eliminated using sterile bone wax and collagen sponges (gelfoam) soaked in thrombin.

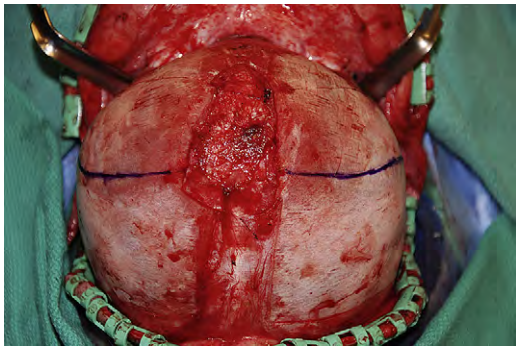

Fig. 8. Open fontanelle within the anterior cranial vault. Care must be taken to avoid injury to the underlying dura during elevation of the coronal scalp flap. Initial development of the flap can be done easily and safely in the subgaleal plane while leaving the pericranium intact over the open fontanelle region. Elevation of the pericranium over the fontanelle involves careful blunt dissection to avoid dural tear. The authors preferred approach is to elevate the initial skin flap (in the subgaleal plane) and then leave the periosteum over the fontanelle in place while elevating the surrounding pericranium.

Approximately 2 cm posterior to the superior orbital rim, an incision is made through the pericranium and the dissection is then continued subperiosteally to expose the facial skeleton (Fig. 7). It is critical to remain within the subperiosteal plane during dissection over the facial skeleton to avoid injury to the facial nerve. Bleeding from vessels perforating the cranium can be controlled with sterile bone wax or thrombin-soaked gelfoam (collagen sponge material). In infants and young children, care must be exercised when dissecting over open fontanelles and sutures, especially midline, to avoid venous sinus hemorrhage and injury to the meninges (Fig. 8). Care must also be exercised when establishing a plane of dissection over the temporalis muscle. The natural plane of dissection is subgaleal. Within the region over the temporalis muscle, the plane should be deepened to the level of the muscle fascia (superficial layer of the temporal fascia). The temporoparietal fascia, which is superficial to the fascia of the temporalis muscle and is an extension of the superficial musculoaponeurotic system (SMAS), invests the temporal branch of the facial nerve. Deepening the dissection to the level of the temporal (temporalis muscle) fascia avoids the nerve and leads to subperiosteal dissection of the facial skeleton (Fig. 9). The supraorbital nerves sometimes restrict flap mobility and dissection of the periorbita. Removal of the bony floor of the foramina using a small osteotome or bone-cutting forceps is often required to release the supraorbital neurovascular bundles and permit further mobility of the flap.

Most previously described approaches to the facial skeleton using a coronal scalp flap involve a separate incision thru the pericranium approximately 2 cm behind the superior orbital rims and the plane of dissection is converted from supraperiosteal to subperiosteal. An alternative approach would

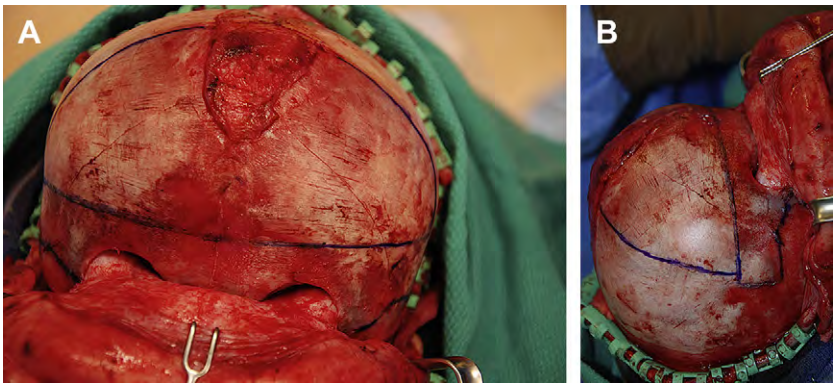

Fig. 9. (*A, B*) Exposure of anterior cranial vault, nasal complex, and orbital structures through a coronal scalp flap. This approach allows for broad subperiosteal exposure of the upper facial skeleton while concealing the skin incision within the hair-bearing scalp. In most cases, separate incisions within the lower eyelids are not required for adequate access to the inferior orbital rims.

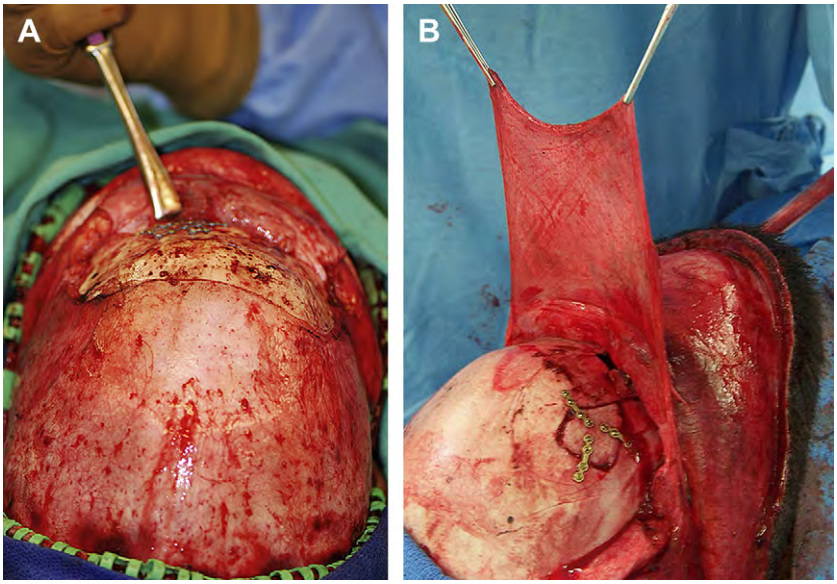

Fig. 10. Development of peri-cranial flap for use in anterior cranial base reconstruction. (*A*) Coronal flap elevated for repair of anterior table of frontal sinus fracture. Most of the pericranium is left in place and subperiosteal dissection is limited to the anterior cranial vault for exposure of the fracture. (*B*) Anteriorly based pericranial flap used for reconstruction of the anterior skull base in a teenaged child who presented with a more extensive frontal sinus fracture involving the posterior table with cerebrospinal fluid leak. The coronal scalp flap is elevated initially, as described previously, in the subgaleal-suprapericranial plane. The pericranium is incised and elevated as a separate layer. A large soft tissue flap can be developed for use in reconstruction of the skull base, closure of dural defects, and separation of the aerodigestive tract from the intracranial cavity.

be to elevate the coronal scalp flap in the subgaleal—suprapericranial plane initially and then design and elevate an anteriorly based pericranial flap as a separate step. An anteriorly based pericranial flap provides an abundant source of vascularized soft tissue for use in anterior skull base reconstruction (Fig. 10).

Closure of the incision in layers, even after facial advancement exceeding 15 mm is usually not a problem. The lateral canthus is resuspended to the fascia over the temporalis muscle or to the lateral orbital rim in an upward and posterior direction. Dissection of the posterior scalp in the subgaleal plane is sometimes necessary to facilitate closure. Although not considered critical, reapproximation of the pericranium should be performed using resorbable sutures as an initial step. The galea and subcutaneous tissues are reapproximated using inverted interrupted sutures. Skin closure can be completed using surgical staples or sutures. The authors prefer resorbable suture material in infants and very young children and nonresorbable suture material in older children and adults (Figs. 11 and 12).

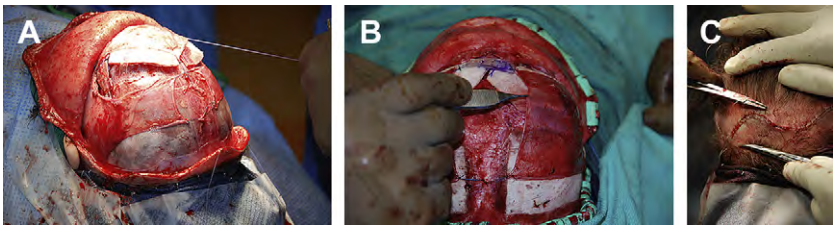

Fig. 11. (*A, B*) Closure of the coronal scalp flap. Although not critical, the authors prefer to reapproximate the periosteum. In the example shown, the anterior, right, and left pericranial flaps are reapproximated loosely following a fronto-orbital advancement procedure in an infant with craniosynostosis. (*C*) Closure of the skin flap begins with the placement of 3-0 vicryl sutures in inverted interrupted fashion to repair the galea and subcutaneous tissues. The skin is closed with a continuous suture. In infants, resorbable suture material (4-0 vicryl rapide or monocryl) is preferred. In older children and adults, nonresorbing suture (4-0 prolene) is used.

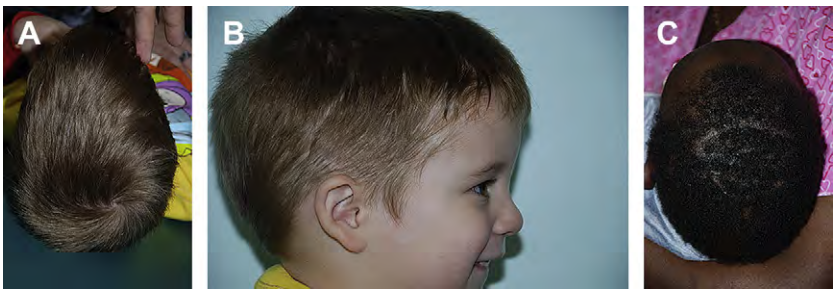

Fig. 12. (*A, B*) Well-healed coronal scalp incision in a child shown 6 months following cranial vault reconstruction. Scar is minimally detectable and curvilinear design helps conceal the incision line within the surrounding hair. (*C*) Postoperative bird's eye view of a well-healed coronal scalp incision almost 3 years after surgery. Although the appearance of the incision scar remains acceptable, there has been forward migration of the scar itself over time.

Summary

The coronal scalp flap is a versatile and cosmetically acceptable approach for access to the cranial vault, cranial base, forehead, nose, upper middle face, and orbits. With the use of this approach, inferior eyelid or transconjunctival access to the orbit is not necessary in most cases. In addition, relatively straightforward modifications of the surgical technique allow the surgeon to harvest vascularized pericranial flaps for use in combined craniomaxillofacial and neurosurgical procedures.

Further readings

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Atlas of the Oral and Maxillofacial Surgery Clinics

Craniosynostosis: Diagnosis and Surgical Management

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The infant brain develops at a rapid rate during the first few months of life. The cerebral volume doubles during the first 6 months and doubles again by the first year of life. By age 2 years, the average child has attained about 80% of the adult brain size, and this significant growth over the first few months of life requires a dynamic yet protective skull. Under normal conditions, the brain volume triples within the first year of life and by the second birthday, the cranial capacity is 4 times that at birth.

Cranial sutures represent a form of articulation between plates of membranous bone by a thin layer of fibrous tissue. Six major sutural areas constitute the cranial vault (Fig. 1), which allow head deformation during vaginal delivery. During postnatal development, cranial vault sutures allow rapid skull expansion and accommodate the significant brain expansion. Minimal pressure (approximately 5 mm Hg) from the growing brain is required to stimulate new bone deposition at the margins.

Craniosynostosis is defined as a *premature fusion* of the cranial vault suture or sutures, which often occurs during fetal development. Because it is an intrauterine event, a more accurate description may be *congenital absence* of the cranial vault sutures. Craniosynostosis results in fusion of the adjacent bones with arrested sutural growth in the affected region. Virchow's classic theory states that premature fusion of a cranial vault suture results in limited development of the skull perpendicular to the fused suture and a compensatory "overgrowth" through the sutures that remain open. The result is a characteristic dysmorphology related to the affected suture, with potential neurologic consequences if underlying brain compression occurs. Most forms of craniosynostosis represent random, nonsyndromic malformations limited to the cranial vault and orbital regions.

In this article, current diagnostic and surgical treatments for nonsyndromic craniosynostosis are presented, and perioperative considerations with specific surgical maneuvers used to treat different types of craniosynostosis are outlined with clinical examples. Management of these patients requires a combined neurosurgical and craniofacial team approach for thorough evaluations of the cranial abnormality. Well-planned surgical procedures are required to release the involved suture and reshape the dysmorphic skeletal components, and these timely interventions restore the skeletal architecture, allowing unrestricted brain growth.

Functional consequences of craniosynostosis

During the first 2 years of life, the cerebral volume increases at a rapid rate, while growth of the visceral structures (ie, brain and eyes) at the same time causes growth of the skeletal structures

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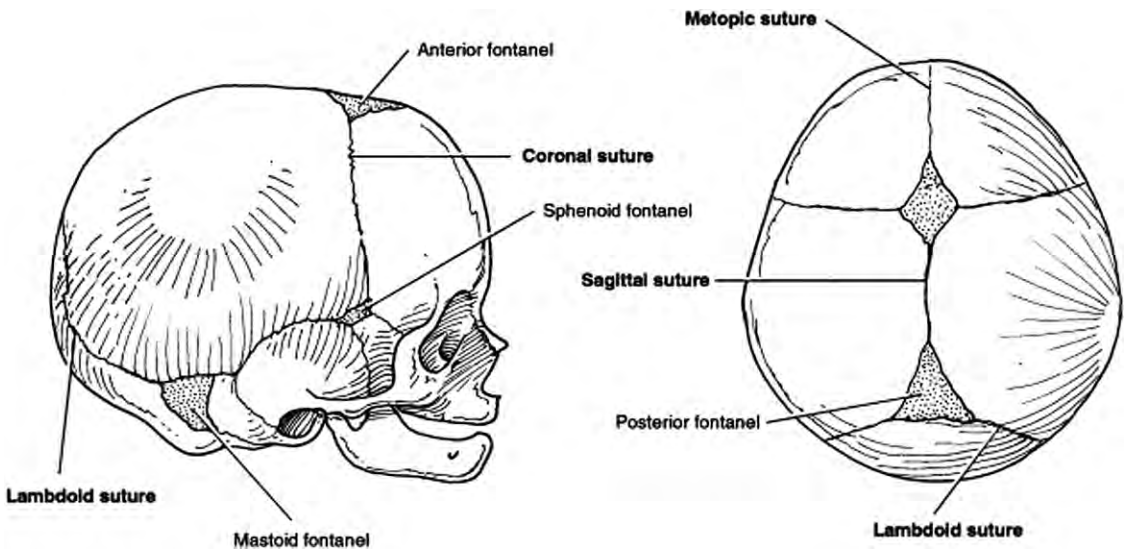

Fig. 1. During infancy, the cranial vault sutures remain open to allow for normal brain growth. Major cranial vault sutures include the metopic, right and left coronal, sagittal, and right and left lambdoid. (*From* Ruiz RL, Ritter AM, Turvey TA et al. Nonsyndromic craniosynostosis: diagnosis and contemporary surgical management. Oral Maxillofacial Surg Clin N Am 2004;16:447−63; with permission.)

(cranial vault and orbits). A complex of flexible cranial bones created by patent cranial vault sutures allow the growing brain to push them outward. At these open regions, cranial growth occurs in a perpendicular direction to each suture and by ectocranial deposition of bone (and simultaneous endocranial resorption).

Intracranial Pressure

Growth along an affected cranial suture is diminished when it is fused, and the compression of a rapidly expanding brain within the limited intracranial volume may result in increased intracranial pressure (ICP, usually >15 mm Hg). This sequence of events is traditionally used to explain the theoretical relationship between craniosynostosis and brain insult, but it does not adequately explain why only certain patients develop elevated ICP. Previous investigations have determined that approximately 14% of children with untreated single-suture craniosynostosis demonstrate increased ICP. When 2 or more sutures are fused, the likelihood of elevated ICP increases to 42%, the functional consequence of craniosynostosis of most concern, because it may adversely affect brain function.

Children with untreated craniosynostosis who develop increased ICP may exhibit several neurologic symptoms, including headaches, vomiting, sleep disturbances, feeding difficulties, behavioral changes, and diminished cognitive functioning. However, such symptoms related to ICP are difficult to detect before one year of age, often presenting with a slow, gradual onset and, left untreated, may be irreversible. In some instances, early recognition of developmental anomalies is difficult because of the lack of proper testing measures.

Hydrocephalus

Hydrocephalus is encountered in approximately 10% of children with multiple-suture craniosynostosis and often seen in the craniofacial dysostosis syndromes. In contrast, hydrocephalus is not usually observed in patients with nonsyndromic single-suture craniosynostosis, but it may occur independently and not necessarily as a consequence of this condition. For cases in which the infant presents with hydrocephalus and craniosynostosis, careful consideration must be given to the exact timing and sequence of the cranial vault reshaping and ventriculoperitoneal (VP) shunt placement. Often, the placement of the VP shunt should be performed as a separate surgery, after the cranial vault reconstruction when possible (using a temporary ventricular drain for the surgical procedure if necessary). Constant cerebrospinal fluid diversion via a VP shunt may decrease brain expansion, reduce the physical support to the bone segments, and, in some instances, alter the skull shape.

Ophthalmologic Effects

The optic nerve is at risk in patients with craniosynostosis and associated elevated ICP. Papilledema, optic nerve atrophy, and eventual loss of vision (possibly complete) may occur with prolonged, untreated ICP elevations. Visual function and eye motility are often impaired when orbital deformity occurs. For example, orbital dystopia secondary to unilateral coronal synostosis can result in disturbances of extraocular muscle movement (ie, strabismus), upper eyelid ptosis, and poor binocular vision. Decreased orbital volume (as seen in cases of syndromic or nonsyndromic bilateral coronal craniosynostosis) causes proptosis, corneal exposure, and increased risk of direct ocular trauma.

Diagnostic approach to abnormal head shape

When an infant presents with a cranial vault asymmetry, the examiner must be alert to the possibility of prematurely fused sutures. However, abnormal head shape may be produced by various causes, which must be distinguished from craniosynostosis. Most commonly, external forces applied to a flexible cranium with open sutures causes a deformation of the skull. The baby's early descent into the true pelvis (often a narrow birth canal) may result in deformational plagiocephaly involving the anterior and posterior cranial vault. Postnatal and external forces from repetitive sleep positioning

may cause these self-limiting skull shape abnormalities. In 1992, the American Academy of Pediatrics recommended that infants sleep on their backs to reduce the risk of sudden infant death syndrome. An increased number of referrals for nonsynostotic posterior plagiocephaly has resulted because of this "Back to Sleep" campaign and the increased awareness of cranial vault deformities within the pediatric community.

Proper identification of cranial vault deformities resulting from nonsynostotic causes (ie, positional plagiocephaly) and those associated with a true absence of a cranial vault suture (ie, craniosynostosis) is critical for diagnosis and appropriate treatment. The importance of a correct diagnosis is underscored by the dramatically different management approaches for each infant. Craniosynostosis requires surgical treatment, whereas benign positional skull molding requires conservative nonsurgical management using custom-made cranial orthotic devices and is not associated with neurologic or developmental sequelae.

A careful history and clinical examination are the primary basis for diagnosis of craniosynostosis, because each form produces a unique head shape. Because the condition is an intrauterine event, parents often report that the deformity was noted immediately after birth and without subsequent improvement. By contrast, cases of positional plagiocephaly have well-rounded head shape at birth, with asymmetry developing 3 to 6 months later (suggesting postnatal deformation of a flexible cranial vault). A consistent combination of arrested growth in one region and compensatory increase in others may explain unilateral sutural involvement causing a bilateral deformity or posterior suture fusion producing an anteroposterior (A-P) dimension deformity. An examiner familiar with these aberrant growth patterns and the characteristic dysmorphologies specific to each type of synostosis should consistently establish an accurate clinical diagnosis.

Craniosynostosis is primarily diagnosed by careful clinical examination but must be confirmed radiographically. Following clinical evaluation by a craniofacial team, a complete skull series of plain radiographs is obtained to evaluate the cranial vault sutures (usually not needed for the simple suture synostosis). Standard views of the cranial vault are often adequate to establish the absence of a cranial vault suture and confirm the diagnosis. For cases in which all the sutures cannot be adequately visualized using plain films, additional imaging using computed tomography (CT) is indicated. In addition to confirming the diagnosis, high-quality craniofacial CT scans provide more detailed 3-dimensional morphologic information, which is useful during the surgical planning phase. One-millimeter axial and coronal cuts with overlap and 3-dimensional reconstructions of the cranial vault, cranial base, orbits, and maxillofacial skeleton are recommended. Precise patient positioning is important in obtaining a diagnostic scan, and this may require sedation or general anesthesia.

Although technological advancements have dramatically increased the accuracy of standard radiographic studies and CT imaging, incorrect and outdated terminology is still occasionally used in describing cranial vault sutures. At some medical centers, radiologists and surgeons continue to use terms such as fibrous synostosis, suture dysfunction, and impending synostosis when describing cranial vault sutures that otherwise appear open. It is known that craniosynostosis is most often an intrauterine event that is usually recognized at birth. Therefore, a patent suture on a diagnostic-quality radiographic study negates the diagnosis of craniosynostosis. Although the postnatal fusion of cranial vault sutures has been described in the scientific literature, it is considered an extremely rare event that is specifically related to certain clinical and syndromic conditions. Some terms not consistent with contemporary knowledge about the condition may be confusing to parents and health care providers, perhaps resulting in inappropriate management.

Treatment of nonsyndromic craniosynostosis

General Considerations

The 2 primary objectives in the contemporary surgical management of nonsyndromic craniosynostosis are release of the involved (ie, fused) suture to allow brain growth in an unrestricted fashion and reconstruction of all dysmorphic skeletal components to achieve a more anatomically correct shape. The best results are often seen with collaboration between a well-organized craniofacial team experienced in managing these issues. Involvement of several other pediatric subspecialists, including geneticists, ophthalmologists, social workers, pediatric anesthesiologists, and pediatric intensivists is mandatory for a successful team.

Early surgical techniques used to treat craniosynostosis involved only the removal of involved sutures via a strip craniectomy. Generally, these limited craniectomy procedures would be performed by a neurosurgeon working independently. The theory behind this approach was that release of the suture would allow unrestricted brain growth and that the expanding brain would adequately recontour the bones without the need for formal craniofacial reconstruction. Although this approach does allow for cerebral decompression, a dysmorphic skull does not reshape itself, even in the presence of an expanding brain, and the result is a residual bony deformity.

Modern surgical management of craniosynostosis involves release of the involved sutures with a formal craniotomy performed by a neurosurgeon. Reconstruction requires the removal, dismantling, and reassembly of all dysmorphic skeletal components into a more appropriate anatomic position. Depending on which specific cranial vault suture is affected, the reconstruction may also involve the orbits. The exact surgical plan is formulated based on the extent of the skeletal deformity, the sutures involved, and the age of the patient at the time of diagnosis. In most cases of nonsyndromic single-suture craniosynostosis, one definitive surgical procedure is required to simultaneously release the suture and reshape the affected skull. This is more likely when the surgical procedure is performed during the first year of life and the patient has normal brain growth.

Timing of Surgery

Craniosynostosis diagnosed early does not represent a "surgical urgency," because the risk of developing increased ICP is minimal during the early growth period (while the fontanelles are open). In most patients with single-suture abnormality, elevated ICP is not frequently encountered and the cranial vault dysmorphology can be safely addressed in the first year of life. The specific, ideal age at which to proceed with craniosynostosis repair is controversial, leading to a wide range of recommendations among pediatric craniofacial and neurologic surgeons. Some advocate an early repair performed at 3 to 6 months of age, whereas others postpone surgery until after age 9 to 11 months. There are theoretical advantages and potential negative consequences associated with each approach.

Some surgeons recommended early surgical correction (3−6 months of age), suggesting that early suture release allows natural brain growth to mold the developing cranium. Such an approach minimizes the surgical bony remodeling and offers the theoretical advantage; however, this method does not obviate the need for formal skeletal reconstruction to establish desired anatomic configuration. Bone segments in younger infants are more malleable, which permits easier reshaping, and the growing brain does provide rapid elimination of extradural dead space and support for the newly constructed bony framework.

Delaying the surgical procedure until approximately 9 to 11 months of age permits a greater proportion of the child's cranial vault growth to occur before correction. This may translate into a more stable skeletal result with fewer postsurgical distortions related to subsequent growth. At this age, the bones are better ossified and harder, which results in less separation at suture lines (eg, frontozygomatic suture) and easier placement of rigid internal fixation devices.

When craniofacial reconstructive surgery is performed during infancy, complete healing of fairly large (1.5 cm) bony defects occurs without the need for additional bone grafting. This is due to the combined bony healing capacity of the pericranium and the dura, which is highly osteogenic during infancy. Most residual cranial vault defects heal completely when the surgery is performed during the first 2 years of life. When cranial vault surgery is undertaken between 2 and 4 years of life, complete healing of residual full-thickness defects is less predictable, and immediate grafting or a secondary procedure for repair may be indicated. After age 4 years, it is unlikely that even small full-thickness cranial vault defects would resolve without deliberate reconstruction or grafting at the time of the initial surgery.

Types of nonsyndromic craniosynostosis

Sagittal Suture Craniosynostosis (Scaphocephaly)

Sagittal suture craniosynostosis is the most common form of nonsyndromic single-suture synostosis, with a prevalence of approximately 1 in 5000 live births. More male children are typically affected, with a male/female ratio of 3:1 reported.

When the sagittal suture is prematurely fused, the result is a scaphocephalic skull deformity. Absence of the sagittal suture results in arrested development of the 2 parietal bone plates, with no growth perpendicular to the suture and a narrow bitemporal and biparietal dimension. The brain continues to expand, and there is compensatory growth at the major sutures that remain open (ie, lambdoid, coronal, and metopic). The overgrowth perpendicular to the coronal and lambdoid sutures causes an abnormal elongation of the cranial vault in the A-P dimension, and patients demonstrate frontal and occipital bossing as a result. This abnormal head shape often has a prominent midline suture with bony overgrowth causing a ridge, often described as a "keel." The exact dysmorphology varies greatly depending on the location of the sutural fusion (anterior, posterior, or the entire sagittal suture).

Surgical management of sagittal suture craniosynostosis requires biparietal craniotomy for release of the fused suture in combination with posterior cranial vault reshaping (Fig. 2). The dysmorphic skeletal components of the cranium are dismantled and reassembled to establish an anatomically correct head shape.

When the posterior region of the sagittal suture is absent, the authors' approach is to reshape the posterior two-thirds of the cranial vault. This reshaping is done with the patient in the prone position. A postauricular coronal scalp incision is made, and the initial dissection is carried posteriorly within the supraperiosteal plane. Once the coronal scalp flap is developed, additional incisions are created through the periosteum, and it is elevated as 3 separate flaps, right and left parietal and occipital. The coronal and lambdoid sutures are identified and burr-hole and osteotomy placement is marked with a sterile pencil. Formal biparietal and occipital craniotomy is then performed by the pediatric neurosurgeon and the dysmorphic bones are removed. Reshaping of the bone flaps is performed through the creation of additional osteotomies and use of bone bending instruments on a sterile back table. The goal of these maneuvers is to increase bitemporal and biparietal width and reduce A-P length of the cranial vault. The reshaped skeletal segments are then reinserted in an anatomically correct position and secured with rigid internal fixation consisting of bone plates secured with monocortical screws.

If only the anterior portion of the sagittal suture is involved, the dysmorphology may be characterized primarily by frontal bossing with less impact on the posterior cranium. In those cases, craniotomy and reshaping of the anterior cranial vault is performed with the patient in the supine position.

Some surgeons advocate a more aggressive total cranial vault reconstruction, which is performed as a single-stage operation for patients who present with fusion of the entire sagittal suture. Such extensive cranial reshaping is associated with disadvantages, including increased morbidity and blood loss, difficult intraoperative access to the entire cranial vault, and fewer points for anchorage of rigid internal fixation appliances. In the authors' experience, the use of total cranial vault reshaping procedures is rarely necessary. Instead, they prefer to manage patients who present with complete sagittal suture involvement and the associated cranial vault deformity (bitemporal and biparietal narrowing in combination with occipital and frontal bossing) with an initial surgical procedure in which only the posterior two-thirds of the cranium is reshaped. This procedure allows normalization of the bitemporal and biparietal width and improvement of the A-P length of the skull but does not address the problem of frontal bossing. If necessary, additional reshaping of the anterior cranial vault is then undertaken as a second phase of surgical treatment approximately 6 to 12 weeks later.

Coronal Suture Craniosynostosis (Anterior Plagiocephaly)

Craniosynostosis involving one of the coronal sutures is the second-most common type of sporadic, nonsyndromic synostosis, with a relative prevalence of approximately 1 in 10,000 children. Although the exact cause of coronal craniosynostosis remains unknown, recent work has suggested that a mutation within the fibroblast growth factor receptor 3 gene is implicated in certain individuals.

Congenital absence of the coronal suture results in the arrested development of the frontal and parietal bones. Clinically, there is anterior plagiocephaly with flattening of the forehead on the affected side. Other characteristic abnormal physical findings include orbital dystopia, with the orbit on the affected side displaced superiorly and posteriorly and with nasal asymmetry and ipsilateral zygomatic hypoplasia. At the same time, there is compensatory overgrowth along the cranial vault sutures, including the contralateral coronal suture, which remain open. The result is flattening of the

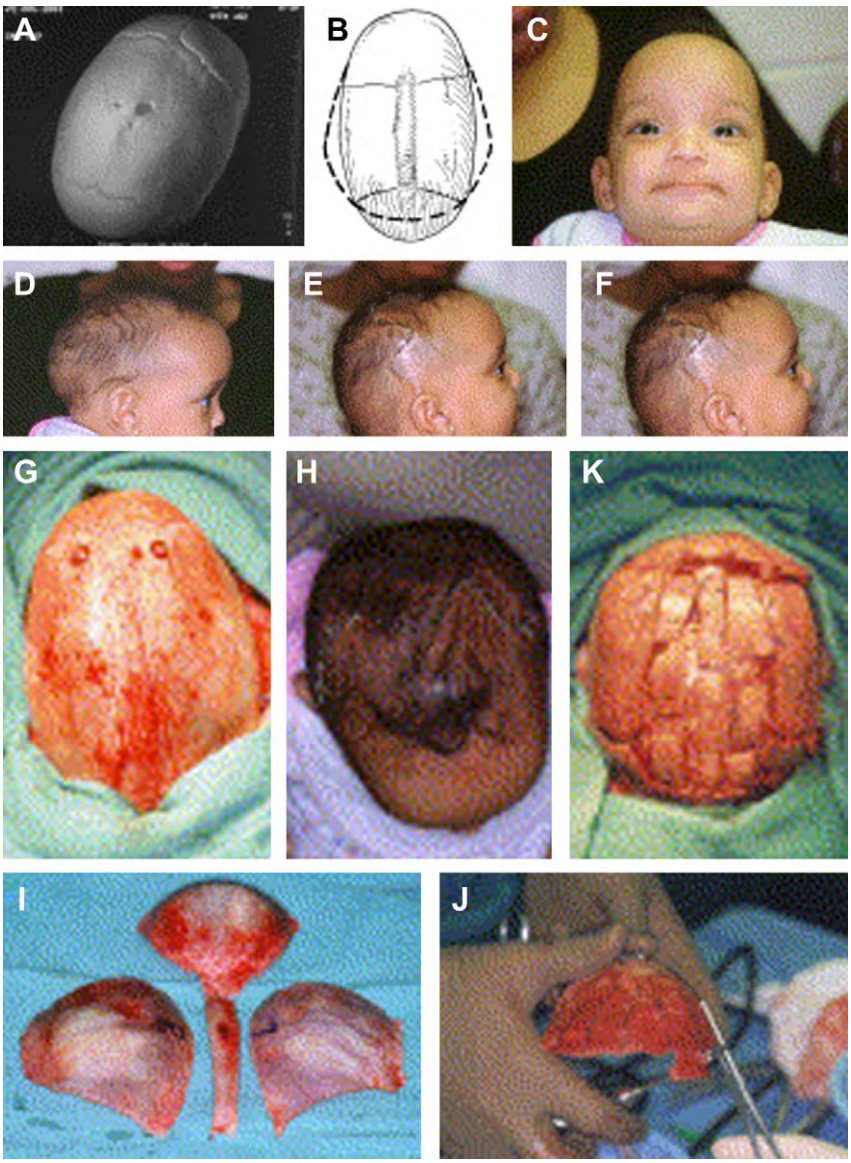

Fig. 2. A 6-month-old child with sagittal suture craniosynostosis and scaphocephalic skull deformity. She underwent posterior cranial vault reshaping. (*A*) Preoperative CT scan demonstrates conspicuous absence of the entire sagittal suture. (*B*) Figure illustrating the objectives of cranial vault reshaping; increased bitemporal/biparietal width and decreased A-P length. (*C*) Preoperative frontal view of the patient. Lack of growth along sagittal suture region results in narrow head shape. (*D*) Postoperative frontal view 3 weeks postsurgery. (*E*) Preoperative profile view demonstrates A-P elongation of the cranium. (*F*) Postoperative profile view. (*G*) Preoperative bird's-eye view. (*H*) Postoperative bird's-eye view. (*I*) Dismantled segments of the posterior cranial vault before reshaping. (*J*) Creation of osteotomies for reshaping of skeletal segments. (*K*) Insertion of bone segments following osteotomies and reshaping procedures. (*From* Ruiz RL, Ritter AM, Turvey TA, et al. Nonsyndromic craniosynostosis: diagnosis and contemporary surgical management. Oral Maxillofacial Surg Clin N Am 2004;16:447−63; with permission.)

forehead on the affected (ie, fused) side and frontal bossing on the unaffected (ie, patent) side of the anterior cranial vault. In this manner, a unilateral coronal synostosis produces a bilateral skull deformity.

In patients with coronal suture synostosis, the anterior skull base is also affected, with absence of the frontoethmoidal, frontosphenoidal, and sphenoethmoidal sutures. This results in a shortening of the anterior cranial base on the affected side and elevation of the sphenoid wing. A plain A-P skull radiograph of a patient with unilateral coronal synostosis demonstrates a characteristic "harlequin-eye" deformity created by the skull base distortion and the superiorly malpositioned orbital rim. Asymmetry of the entire cranial base is also common, with a shift in the anterior and posterior cranial

base to midsagittal plane angles of greater than 7° as a consistent finding on CT scan evaluation. Cranial base involvement also frequently produces some level of mandibular deviation and lower facial asymmetry.

The contemporary surgical correction of unilateral coronal suture synostosis consists of bifrontal craniotomy, orbital osteotomies with creation of an orbital bandeau, fronto-orbital advancement, and anterior cranial vault reshaping (Figs. 3 and 4). A postauricular coronal scalp incision is created and dissection is performed in the supraperiosteal plane. The pericranium and temporalis muscles are elevated separately. Bilateral circumferential periorbital dissection is then performed, with complete exposure of the orbits to the level of the inferior orbital rim and zygoma. This requires release of the lateral canthal ligaments. Care is taken to preserve the medial canthal attachments and nasolacrimal apparatus.

The authors outline the orbital bandeau, including the tenon extensions within the temporal bones, bilaterally using a sterile pencil before deciding on burr-hole placement. Typically, a bifrontal craniotomy is created by the pediatric neurosurgeon, with removal of the frontal bone unit. Next, the dura is dissected free of the inner table of the cranial vault and along the anterior skull base. A reciprocating saw with a small blade is used for the creation of osteotomies along the lateral orbital wall/rim, orbital roof, and superior-medial aspect of the orbital wall on each side of the bandeau. Care is taken to avoid injury to the temporal lobe of the brain during the sectioning of pterion bilaterally. The orbital osteotomies are carried one-half to three-quarters of the length of the orbit on each side. The final osteotomy cut is across the nasofrontal region joining both orbital osteotomies and allowing for mobilization of the orbital bandeau.

Recontouring frequently involves the creation of a vertical osteotomy in the midline to allow for advancement of the affected orbital segment. Additional osteotomies between the lateral orbital region and tenon extension may be used for additional reshaping of the bandeau. The newly constructed orbital unit is then inserted and secured to the temporoparietal bones bilaterally using bone plates and screws. The authors add a third point of fixation at the nasofrontal junction using 2-0 polydioxanone suture or stainless steel wires to avoid upward displacement of the bandeau when the coronal flap is repositioned. For cases in which severe flattening of the superior orbit is present, onlay bone grafts may be added on the affected side of the bandeau for additional contouring. The frontal bones are then osteotomized and used to reconstruct the anterior cranial vault region with reshaping as necessary. Closure begins with lateral canthopexies performed through the coronal access. The scalp flap is then closed in 2 layers.

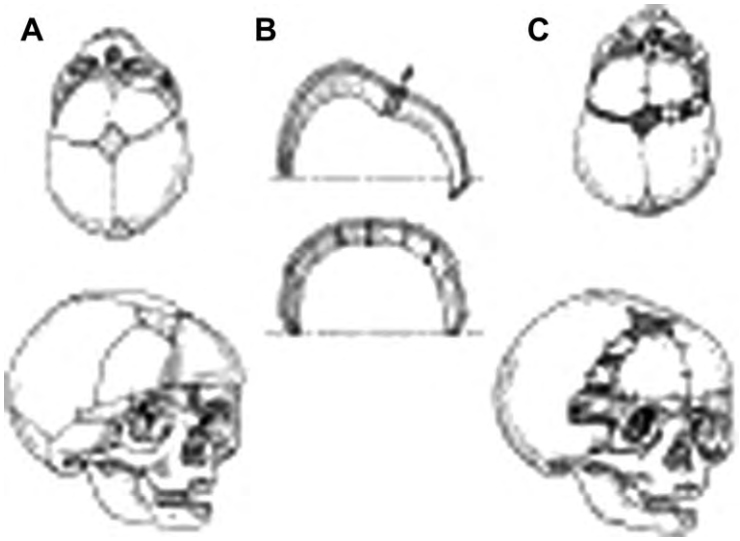

Fig. 3. Reconstructive procedure used to address the dysmorphology associated with unilateral coronal suture craniosynostosis. (*A*) Outline of the bifrontal craniotomy and orbital osteotomies. (*B*) The dysmorphic orbital bandeau is removed and vertical osteotomies are used for reshaping. (*C*) The reconstructed orbital bandeau is reinserted and anterior cranial vault reconstruction is performed. (*From* Ruiz RL, Ritter AM, Turvey TA, et al. Nonsyndromic craniosynostosis: diagnosis and contemporary surgical management. Oral Maxillofacial Surg Clin N Am 2004;16:447−63; with permission.)

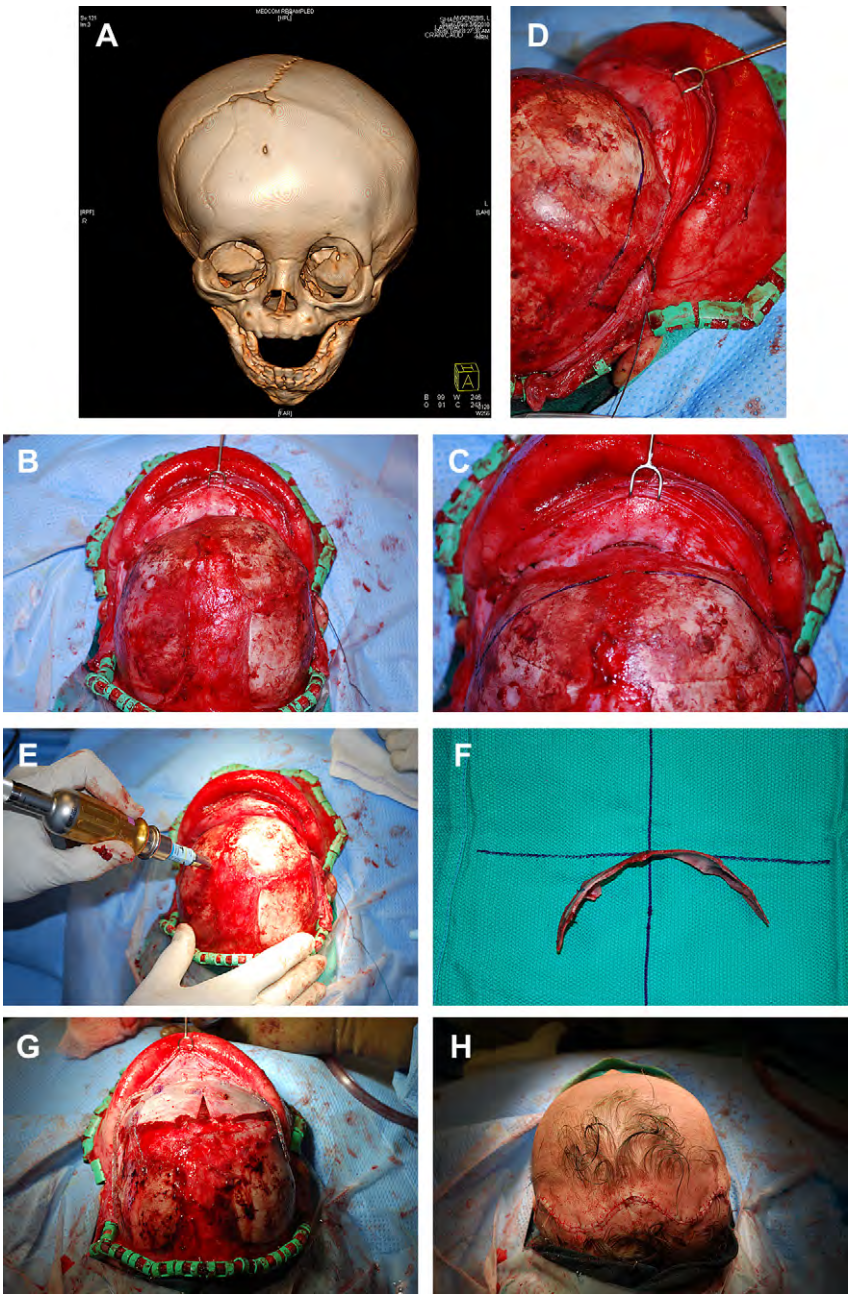

Fig. 4. A 7-month-old child with left-sided unilateral coronal suture craniosynostosis and anterior plagiocephaly. Surgical reconstruction consisted of bifrontal craniotomy, fronto-orbital advancement, and anterior cranial vault reshaping. (*A*) Three-dimensional Craniofacial CT scan reveals absence of the left coronal suture. Note the degree of cranio-orbital dysmorphology on the affected side. (*B—D*) Surgical exposure of the affected skeletal structures, including the fronto-orbital region and anterior cranial vault, through a coronal scalp flap. (*E*) Burr-hole placement allows for safe separation of the dura from the overlying cranial vault in preparation for bifrontal craniotomy. (*F*) Dysmorphic orbital segment (bandeau) on a sterile back table before segmentalization and reshaping. (*G, H*) Reconstructed bony segments in place with resorbable rigid internal fixation and closure of the soft-tissue flap. (*I*) Preoperative frontal view demonstrates a skeletal deformity characterized by flattening of the forehead on the affected side, orbital dystopia, and nasal asymmetry. (*J*) Postoperative frontal view 6 months post-surgery. (*K*) Close-up frontal view of fronto-orbital contour postoperatively. (*L*) Bird's-eye view preoperatively. (*M*) Bird's-eye view postoperatively.

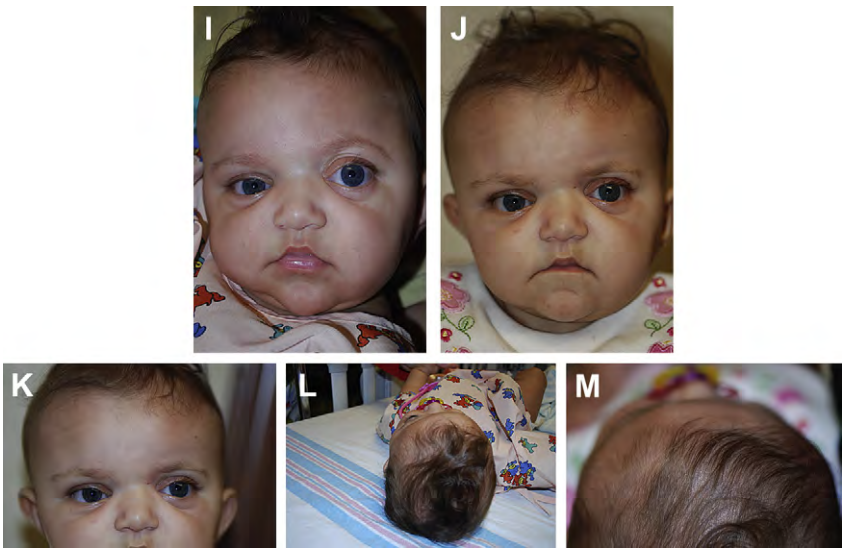

Fig. 4 (*continued*)

Metopic Suture Craniosynostosis (Trigonocephaly)

Metopic suture craniosynostosis is an uncommon form of single-suture craniosynostosis with a prevalence of approximately 1 in 15,000 children. An increased proportion of male children are affected. Unlike other forms of isolated, single-suture craniosynostosis, approximately 5% of patients with metopic fusion present with a positive family history.

Premature fusion of the metopic suture may be seen in nonsyndromic, otherwise healthy infants or as part of an identified syndrome. Previous reports have suggested that a third group of infants born with metopic synostosis demonstrate other congenital malformations, neuromuscular dysfunction, and/or neurodevelopmental delay, but without the delineation of a syndromic diagnosis. This observation is consistent with the authors' experience. At the University of North Carolina, a review of patients with metopic suture involvement, treated by the senior author (RLR), revealed a high rate of associated congenital malformations. Forty-three percent of patients seen with a diagnosis of

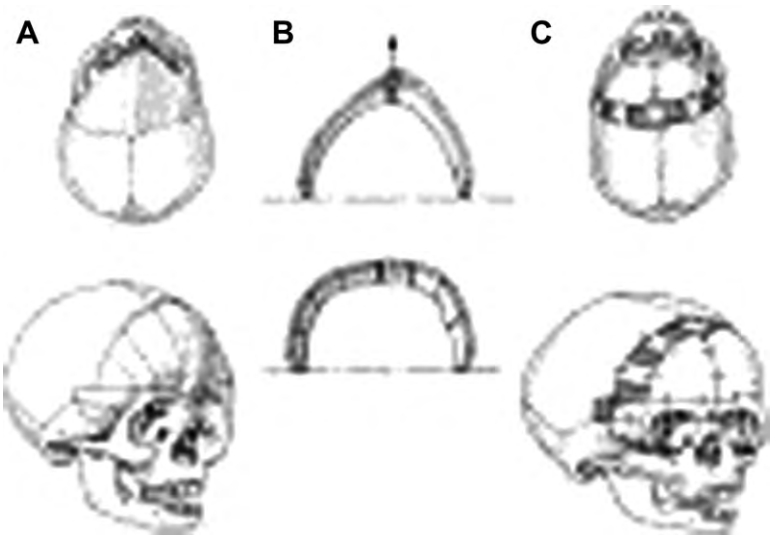

Fig. 5. Reconstructive procedure used to address the trigonocephalic skull deformity associated with metopic suture craniosynostosis. (*A*) Outline of the bifrontal craniotomy and orbital osteotomies. (*B*) The dysmorphic orbital bandeau is removed and vertical osteotomies are created for reshaping. (*C*) The reconstructed orbital bandeau is reinserted and anterior cranial vault reconstruction is completed. (*From* Ruiz RL, Ritter AM, Turvey TA, et al. Nonsyndromic craniosynostosis: diagnosis and contemporary surgical management. Oral Maxillofacial Surg Clin N Am 2004;16:447–63; with permission.)

metopic suture synostosis over the course of a 4-year period presented with associated malformations but no clear syndromic diagnosis. Malformations not previously described in association with metopic synostosis were observed, including in 3 patients with a tethered spinal cord. Whether the combination of metopic synostosis and associated anomalies represents an unknown syndrome remains unclear. Thorough medical and genetic evaluation of infants born with metopic craniosynostosis is indicated to prepare these children for craniofacial surgery, counsel parents, and determine long-term neurodevelopmental prognosis.

The combination of arrested growth of the frontal bones perpendicular to the midline (metopic) suture and continued growth at the coronal sutures creates a trigonocephalic (ie, triangle-skull) deformity. The dysmorphology is characterized by a triangular-shaped forehead, orbital hypotelorism, and horizontally retrusive lateral orbital rims. Also, clinical examination frequently reveals a pronounced vertical bony ridge within the center of the forehead.

Surgical intervention for metopic suture craniosynostosis is indicated for cases in which clinically significant dysmorphology is present. Not all patients require surgical intervention, because some present with mild ridging of the suture and only minimal morphologic deformity. This is especially true because the likelihood of a child developing increased ICP secondary to metopic synostosis is low.

The surgical approach to reconstruction of the trigonocephalic skull deformity associated with metopic synostosis is similar to the surgical procedure described for coronal synostosis. Bifrontal craniotomy is performed in conjunction with fronto-orbital advancement and anterior cranial vault reshaping (Figs. 5−7). Reshaping of the orbital bandeau is performed to advance the lateral orbital

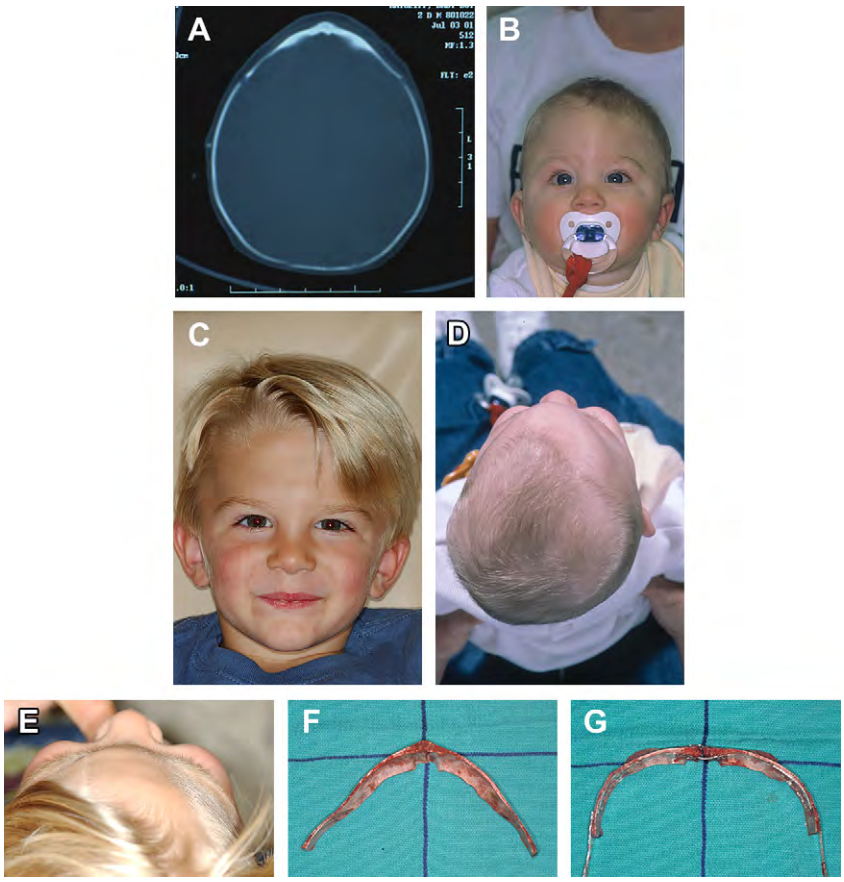

Fig. 6. A 7-month-old child with metopic suture craniosynostosis and trigonocephaly. He underwent craniofacial reconstruction via bifrontal craniotomy approach, fronto-orbital advancement, and anterior cranial vault reshaping. (*A*) Axial view CT scan reveals the triangular-shaped frontal bone complex with a prominent bony ridge in the region of the metopic suture. (*B*) In addition to the triangular-shaped forehead, the preoperative frontal view demonstrates hypotelorism and horizontal retrusion of the lateral orbital rims. (*C*) Postoperative frontal view 2 years after the surgical procedure. (*D*) Bird's-eye view preoperatively. (*E*) Bird's-eye view postoperatively. (*F*) Intraoperative view of the orbital bandeau before reshaping. (*G*) Intraoperative view of orbital bandeau after osteotomies, reshaping, and reassembly using resorbable bone plates and screws.

segments and create a more rounded forehead with improved anatomic contour. The creation of a gap at the vertical midline osteotomy within the bandeau and placement of autogenous bone graft may be performed in an effort to widen the orbits and address the orbital hypotelorism. Although this is a reasonable surgical maneuver, it usually has minimal impact on the hypotelorism encountered as part of the metopic synostosis deformity. This is due to the placement of a midline osteotomy with graft within the bandeau not addressing the patient's medial orbital wall-to-medial orbital wall distance.

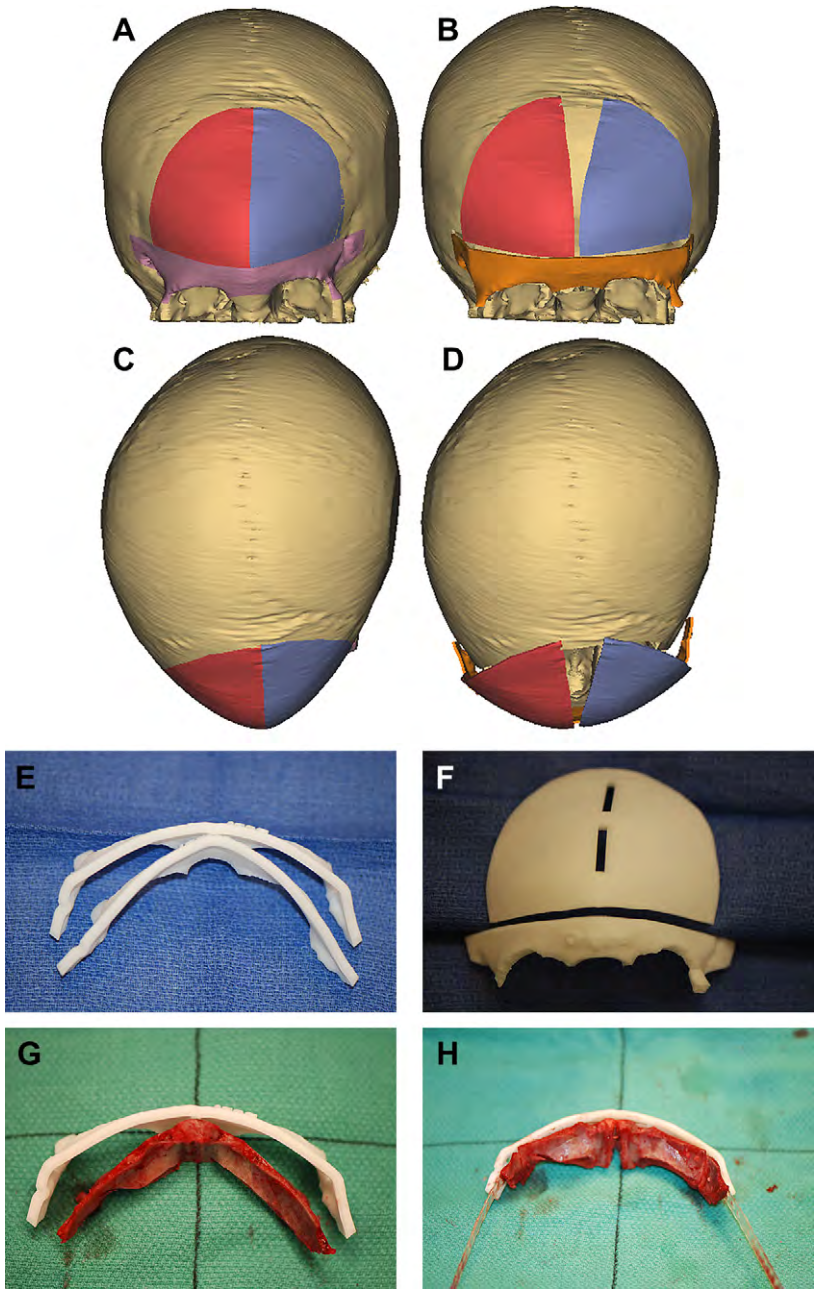

Fig. 7. Computer-aided treatment planning and intraoperative techniques. (*A*—*D*) Three-dimensional reconstruction of a CT scan with segmented fronto-orbital bandeau and bifrontal craniotomy bone flaps. Skeletal movements are planned. (*E, F*) Surgical templates are printed based on the proposed surgical movements. (*G, H*) Use of a surgical template in the reshaping of the orbital bandeau. Specific, preplanned changes in the morphology of the skeletal segments are executed with the assistance of templates based on computer-aided treatment plan. (*I*) Frontal view of patient with metopic craniosynostosis preoperatively. (*J*) Frontal view of patient 3 months after fronto-orbital advancement and anterior cranial vault reconstruction. (*K*) Bird's-eye view of patient preoperatively. (*L*) Bird's-eye view of patient postoperatively.

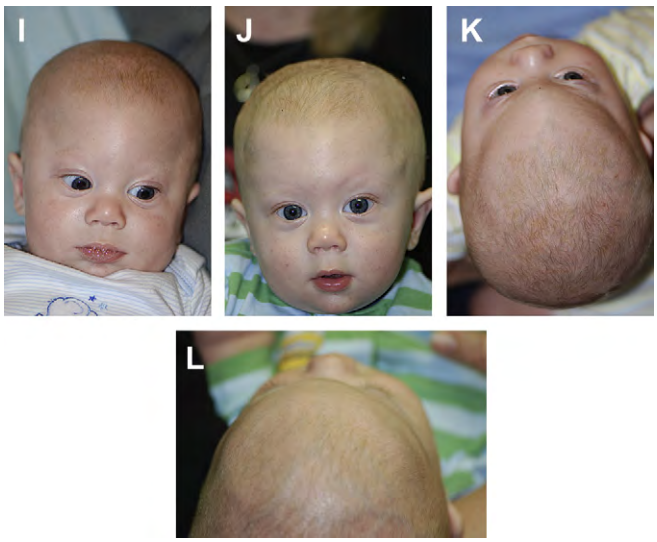

Fig. 7 (*continued*)

Lambdoid Suture Craniosynostosis (Posterior Plagiocephaly)

Posterior plagiocephaly secondary to true lambdoid suture craniosynostosis is a rare event with a prevalence of approximately 1 in 150,000 live births. Posterior plagiocephaly secondary to lambdoid synostosis is uncommon and is frequently confused with positional plagiocephaly. Therefore, the diagnosis must be confirmed with radiographic imaging before any surgical intervention is considered (Fig. 8).

When premature fusion of one lambdoid suture occurs, the most noticeable finding is often a severe flattening of the occipitoparietal region. Restricted growth of the ipsilateral parietal bone also produces retraction of the forehead on the affected side and superior-posterior displacement of the ear. By contrast, posterior positional plagiocephaly presents with a distinct morphologic pattern characterized by occipitoparietal flattening, ipsilateral frontal bossing, and forward displacement of the ear on the affected side.

Although surgeons may be tempted to perform a more limited procedure, such as strip craniectomy, to address unilateral lambdoid craniosynostosis, a combined craniofacial/neurosurgical approach remains the most appropriate treatment. The authors' preference is to carry out craniotomy with formal reshaping of the involved portions of the posterior cranium. The degree of skeletal dismantling and posterior cranial vault reshaping is determined by the extent of the dysmorphology.

Bilateral Coronal Suture Craniosynostosis (Brachycephaly)

When bilateral coronal craniosynostosis is seen in conjunction with midface deficiency, the possibility of a craniofacial dysostosis syndrome (eg, Apert, Crouzon, Pfeiffer, Saethre-Chotzen)

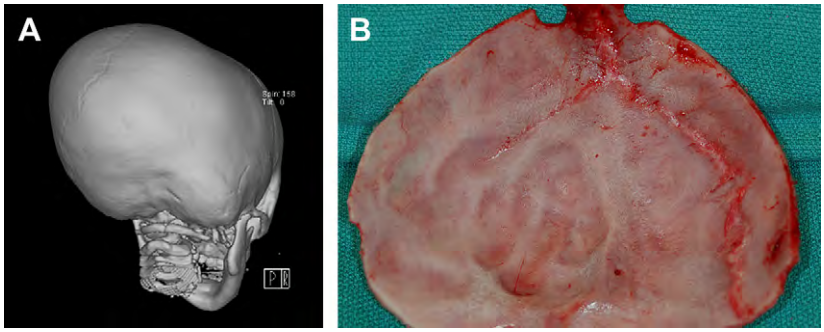

Fig. 8. (*A*) Three-dimensional CT scan of a patient with right-sided unilateral lambdoid suture synostosis. (*B*) Intraoperative view of the occipital bone reveals conspicuous absence of the lambdoid suture on the affected side.

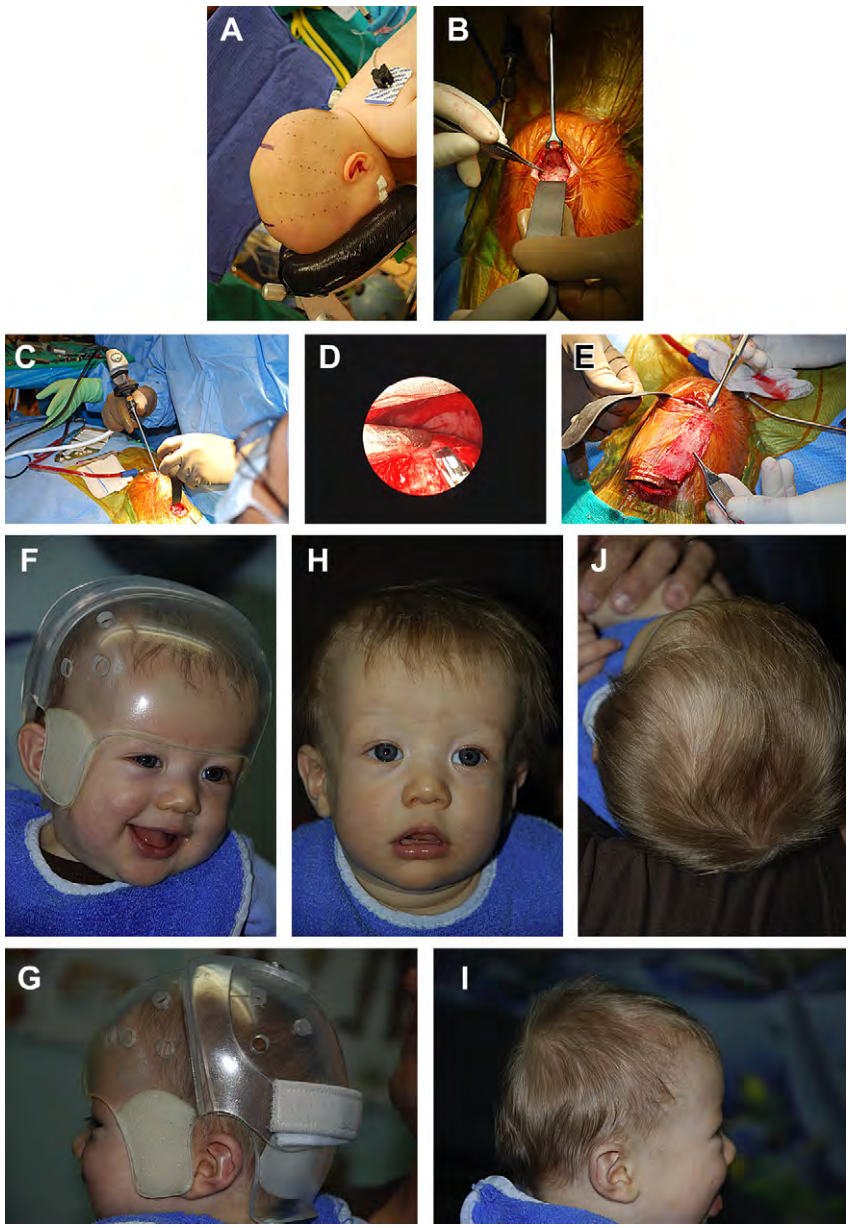

Fig. 9. Endoscopy-assisted craniosynostosis release via suture craniectomy. Three-month-old infant with sagittal suture craniosynostosis treated using and endoscopic approach. The fused cranial vault suture is released with using endoscopic visualization, allowing for normal (unrestricted) brain growth. Although the surgical procedure eliminates the risk of increased ICP, there is minimal immediate change in the cranial vault morphology of the infant. Reshaping of the dysmorphic skeletal structures is accomplished postoperatively using a custom-made cranial orthotic band. (*A*) Infant positioned prone. Meticulous care is taken to avoid any direct pressure over the eyes when working in the prone position. Proposed incisions at the anterior and posterior limits of the involved sagittal suture allow for adequate access. Other markings are made to outline additional barrel-staves and releasing osteotomies along the posterior two-thirds of the dysmorphic cranial vault. (*B*) Incisions are made, and dissection along involved suture in the subgaleal-supraperiosteal plane proceeds easily and bloodlessly. (*C, D*) Use of endoscopic visualization during release of involved sagittal suture, craniectomy, and creation of barrel-staves. (*E*) The entire sagittal suture and surrounding bone is removed. (*F, G*) Use of dynamic, custom cranial orthotic band for reshaping of the cranial vault following release of the involved suture. Most endoscopic craniosynostosis regimens require 8 to 12 months of reshaping/molding using the orthotic. (*H—J*) Postoperative appearance of patient after 8 months of therapy with custom orthotic helmet/band. Note well-healed incisions and correction of cranial vault shape (eg, decreased A-P length and increased bitemporal and biparietal width).

must be considered. However, the congenital absence of both coronal sutures is known to occur in patients without midface deficiency or syndromic diagnosis. Although these patients demonstrate what is thought to be a nonsyndromic form of bilateral coronal craniosynostosis, recent advances in molecular testing suggest that there may be some familial pattern with implication of mutations within the fibroblast growth factor receptor (1,2, and 3) genes.

Bilateral coronal suture fusion results in a characteristic craniofacial dysmorphology described as brachycephaly. This deformity includes shortening of the anterior skull base bilaterally resulting in a decreased A-P cranial dimension, widening of the skull secondary to compensatory growth at the sagittal suture, and horizontal deficiency of the lateral orbital rims. Also, some patients exhibit significant vertical elongation of the anterior cranial vault.

Because 2 cranial vault sutures are involved, the incidence of increased ICP rises to approximately 42%. Surgical intervention is undertaken to normalize intracranial and orbital volume and establish anatomic position and contour of the orbits and forehead. The standard surgical approach for repair of the deformity associated with bilateral coronal synostosis includes a more extensive bifrontal craniotomy than is used in unilateral cases, fronto-orbital advancement, and anterior cranial vault reconstruction.

Endoscopy-Assisted Craniosynostosis Repair

In recent years, the use of endoscopic instrumentation in the surgical management of craniosynostosis has been described and shows promise in the management of specific patients. An endoscopically assisted approach is used to carry out a strip craniectomy for release of the involved sutures and for creation of multiple "barrel-stave" osteotomies within the cranial vault. The primary advantage of this technique is a much smaller surgical incision. Other theoretical advantages include a lower incidence of perioperative blood transfusions, less morbidity, and decreased length of hospitalization (Fig. 9). Full-time molding using a custom helmet (ie, cranial orthotic) is often required for 8 to 12 months postsurgery.

Summary

Contemporary surgical management of nonsyndromic craniosynostosis requires the combined expertise of a pediatric craniofacial surgeon and pediatric neurosurgeon. The goals of surgical intervention are the release of the affected suture allowing for unrestricted development of the visceral components (eg, brain, eyes) and 3-dimensional reconstruction of the skeletal components establishing a more normal anatomic position and contour. Surgeons caring for infants with these cranial and orbital malformations must maintain a thorough understanding of the 3-dimensional anatomy, the characteristic dysmorphology associated with the different types of synostosis, and the complex interplay that exists between surgical intervention and ongoing skeletal growth.

Further readings

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Endoscopic Techniques for Craniosynostosis

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Craniosynostosis and the abnormalities associated with this clinical entity pose significant challenges to the treating craniofacial and neurosurgeons caring for patients with these conditions. Ever since Lannelongue [1] first introduced the concept of surgically treating this condition, surgeons have struggled to develop the best possible techniques [2–8]. At our craniofacial center we have also used many approaches extending from minimally invasive to extensive calvarial vault remodeling techniques [4,9–28]. Although technically rewarding at the time of surgery, the extensive calvarial vault remodeling techniques are almost universally associated with increased blood transfusion rates, massive facial swelling, bruising, and longer hospital stays. Furthermore, the results associated with these procedures are mixed at best and often times with poor outcomes when patients are followed longitudinally into their early adulthood.

About 15 years ago we decided to change our approach and began treating patients at the earliest age possible using endoscope-assisted minimally invasive procedures followed by helmet therapy for 1 year. The concept is simple and involves allowing the rapidly growing brain of an infant to reshape the craniofacial skeleton and correct the deformities associated with the prematurely closed suture. The corrective process is aided by the helmet once the stenosed suture is released, and the brain is allowed to resume its normal shape as it grows rapidly. The surgical techniques and the helmet therapy vary with each suture type and are described in detail in this article.

Sagittal suture

Because of predetermined genetic forces that lead the skulls of patients with sagittal synostosis to revert back to a scaphocephalic shape within a year after treatment with any procedure, we do not perform a single linear strip craniectomy. In essence, we remove a large portion of the calvarial vault extending from the anterior fontanelle to the lambda. The width of the craniectomy is inversely proportional to the child's age. In a several-weeks-old baby, the width of the craniectomy may be as much as 5 to 6 cm, whereas in patients aged 6 months or older, it may be 2 to 3 cm. In addition, we create bilateral barrel-stave type osteotomies behind the coronal suture and in front of the lambda. These osteotomies allow for immediate increase in the euryon-to-euryon distance and cephalic index (CI).

The patient is placed in the modified prone position in a padded beanbag with a viscoelastic pad. The first scalp incision, measuring between 2 and 3 cm, is made about 2 cm behind the anterior fontanelle, transversely across the sagittal suture. Subgaleal dissection is done with a needle tip bovie and the visualization of a zero-degree 4-mm rigid endoscope (Karl Storz, Germany). A rhinoplasty lighted retractor can also be used to develop the subgaleal plane. Care is taken not to disrupt or remove the pericranium because this leads to unnecessary blood loss and poor visualization. A second incision of similar length and direction is made in front of the lambda. In a comparable method,

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subgaleal plane dissection is undertaken until the entire area between the lambda and anterior fontanelle is developed.

A pediatric craniotome is used to create a 7-mm burr hole on one side of each incision. The dura is freed from the overlying bone using a #1 Penfield dissector across the stenosed suture. An osteotomy is created with a 5-mm angled Kerrison rongeur. Anteriorly, a wedge of bone extending to the anterior fontanelle is then removed. This maneuver allows for the introduction of a 4-mm rigid, 30°-angled endoscope under the bone. Separation of the dura from the overlying bone is done with the aid of an insulated suction tube and extends sequentially until the posterior osteotomy is reached. Gentle sweeping motion is used as the suction tube is used to separate the dura from the bone. Whenever a perforating vein of moderate to large caliber is encountered, it may be directly coagulated under full visualization, with bipolar forceps. Because the naturally occurring fibers connecting a patent suture to the dura are absent in craniosynostosis, separating these 2 structures under endoscopic visualization is a relatively easy maneuver.

Once the dura has been fully separated from the bone, a pair of parallel osteotomies are made using Mayo scissors (Teleflex Inc, PA, USA). Because of the patient's young age, the bone is relatively thin and easy to cut. In older patients with thicker cranium, bone-cutting scissors may be used to make the osteotomies. The bone may be folded or cut in half to remove it through the anterior scalpincision. The wedge osteotomies are done at the level of each incision and extended laterally and inferiorly toward the squamosal suture. Care is taken not to make the osteotomy too close to the coronal or lambdoid suture and risk a dural tear (Fig. 1); once the bony cuts are made, osseous hemostasis is achieved using a suction-cautery bovie unit (Valley Lab, Valley Forge, PA, USA).

This unit is set at 60 W and allows for rapid and efficient hemostasis. Often, multiple passes are needed until all of the bleeding and oozing stops, and the bone is cauterized until it turns black. No untoward effects have been found with this approach, and it has no effect on new bone regeneration. Further hemostasis is obtained using Surgiflo (Johnson & Johnson, New Brunswick, NJ, USA). Before skin closure, the surgical field is inspected with the endoscope to document complete hemostasis and look for dural tears. The galea is closed with 4-0 Monocryl (Ethicon, Inc, Somerville, NJ, USA), and the skin is sealed with DERMABOND (Ethicon, Inc, Somerville, NJ, USA).

Coronal suture

After induction of general anesthesia, the patient is intubated and placed in the supine position on a pediatric horseshoe head holder. Corneal protectors with ophthalmic lubricant are used to prevent corneal abrasions or damage. A 2-cm incision is made halfway (over the stephanion) between the anterior

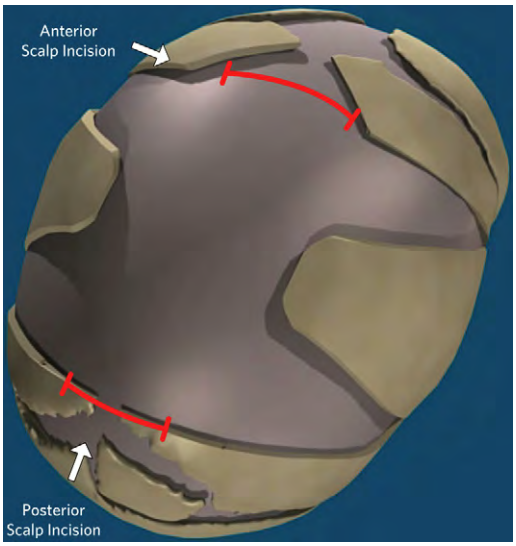

Fig. 1. The amount of bone removed for the treatment of sagittal craniosynostosis. The location of the incisions and their relationship to the anterior fontanele and the lambda.

fontanelle and the pterion on the side and across the stenosed suture. Subgaleal dissection is undertaken toward the anterior fontanelle with the aid of a rhinoplasty lighted retractor or a 0° rigid endoscope. The same maneuver is used to reach the area of the pterion and the squamosal suture on the affected side.

The pediatric craniotome is used to create a 7-mm burr hole at the incision. The burr hole is enlarged longitudinally along the suture axis using Kerrison rongeurs (5 mm). A 30° rigid endoscope is then inserted under the bone and first directed medially toward and up to the anterior fontanelle. The endoscope is then removed and redirected inferiorly and laterally toward and behind the pterion to reach the open squamosal suture. A 6- to 7-mm osteotomy is made with a combination of scissors and bone rongeurs, making sure that it extends all the way down to the squamosal suture (Fig. 2). Failure to do so prevents the orbitofrontal complex from moving forward and inferiorly. Although at the beginning of our series part of the sphenoid wing was removed, we have found this to be unnecessary and no longer do so. Osseous hemostasis is achieved using the suction-electrocautery unit as described in the sagittal section.

The dura overlying the coronal suture area seems to reossify at a different rate than the one over the vertex. Early on we made osteotomies as wide as 2 cm but found that it took much longer for the bone to fully reossify, and some patients were left with small areas of nonossification. In addition, the growing brain moves the frontal bone forward up to 2 to 2.5 cm, leaving a large area devoid of bone (Fig. 3). As such, we limit our osteotomy to the width of the bone or Kerrison rongeurs (6 mm). The surgical area is inspected with the endoscopes, and the galea is closed with Monocryl and the skin with DERMABOND; Surgiflo and liquid thrombin (Gen Trac Inc, Middleton, WI, USA) are used to obtain complete hemostasis.

Metopic suture

The patient is placed in the supine position on a cerebellar horseshoe head holder. Corneal protectors and ophthalmic lubricant are used as described earlier. The forehead and face are prepped with providone-iodine solution. A 2- to 3-cm incision is made across the metopic suture behind the hairline but in front of the anterior fontanelle. The incision is taken down through the galea, and the subgaleal plane is dissected with a needle tip bovie extending all the way to the nasion from the anterior fontanelle. This procedure can be done with malleable insulated scalp retractors and a 0° rigid endoscope. A pediatric craniotome is used to create a 7-mm burr hole, which is then enlarged longitudinally with 6-mm Kerrison rongeurs. A 30°-angled endoscope is inserted under the bone to develop the epidural space plane of dissection. About half way down the forehead, prominent

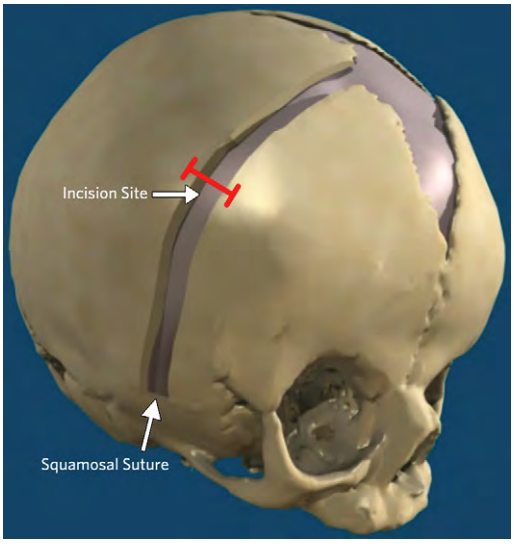

Fig. 2. Location of the osteotomy, which extends from the anterior fontanelle, incorporates the most proximal part of the stenosed coronal suture, and reaches the open squamosal suture behind the pterion.

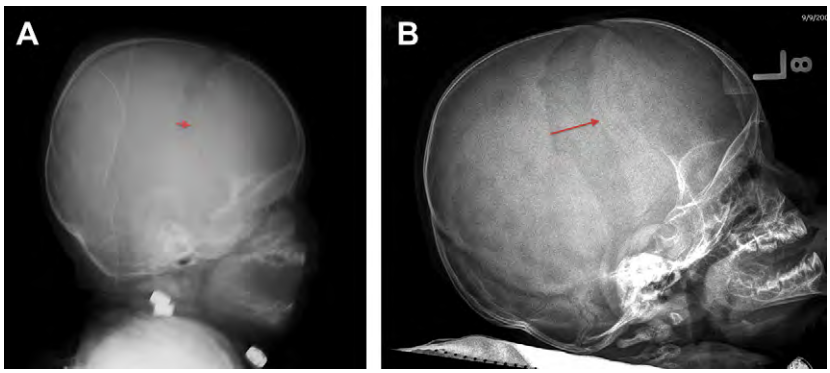

Fig. 3. (*A*) Lateral radiograph immediately after surgery demonstrates the osteotomy that measured 6 mm. (*B*) Radiograph 6 months after surgery shows that the osteotomy width has increased to almost 3 cm.

bridging veins are often found, which extend from the sagittal sinus to the bone. These veins can be a source of significant bleeding, if not promptly and properly cauterized. The generous use of Surgiflo and gelfoam soaked with thrombin can assist in achieving successful hemostasis. Care must be taken that the epidural plane is dissected all the way to the nasofrontal suture as well as to the anterior fontanelle. The osteotomy is then made with a combination of Mayo scissors and bone-cutting rongeurs and must extend all the way down to the nasofrontal suture (Fig. 4). Failure to do so leads to an unsuccessful result. Hemostasis is achieved as described earlier using the suction-electrocautery unit (set at 60 W), Surgiflo, and thrombin. The area is carefully inspected for bleeding bridging veins, oozing diploë, or dural tears. The galea and skin are closed as described earlier.

Immediate postoperative care

The patients are extubated and given intravenous morphine and rectal acetaminophen to help control postoperative pain. The patients are taken to the pediatric unit where they are observed overnight. Most patients experience various degrees of pain or discomfort during the first 8 hours after surgery. Soon afterward all patients settle down, begin oral intake appropriately and are ready

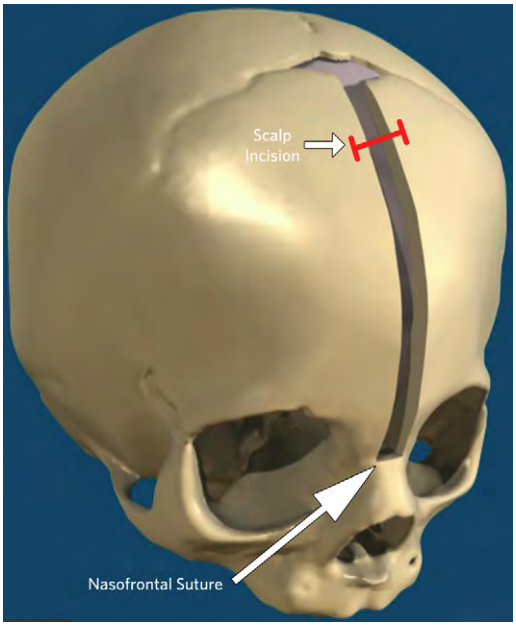

Fig. 4. Location and extent of the frontal osteotomy removed for the treatment of metopic synostosis. The incision is made behind the hairline but in front of the anterior fontanelle. The osteotomy extends down to the nasofrontal suture.

for discharge the following morning. If the patient is nursing, he/she is allowed to attempt to do so immediately after surgery, and many are able to do so without difficulties. The patients are placed on an alternative schedule of ibuprofen and acetaminophen every 3 hours for the next 3 days. Typically, only a small amount of swelling is seen at the surgical site, and bruising is almost never observed.

Helmets

The use of helmets is integral to the success of our treatment protocol. In the case of sagittal synostosis, the treatment can be divided into 3 phases. The first phase occurs immediately after surgery and lasts for 6 to 8 weeks. During this period, the CI of the head is allowed to increase markedly from preoperative levels. The aim of the second phase is to overcorrect the head shape and obtain a high CI (>80). The third phase is used to maintain the overcorrected shape and to counteract the naturally occurring tendency to return to a scaphocephalic shape. The total treatment lasts until the patient is about 18 months old. We have found that the head shape and CI of the patients remain stable over time once they reach the 18-month mark. This finding is certainly true for all of the patients who have been followed into their early teenage years. Any facial or scalp swelling is allowed to decrease during the first 72 hours, and then the patient is scanned using the infrared STARscanner (Orthoamerica, Orlando, FL, USA). The scanned data are forwarded to the manufacturing facility at Orlando, where the helmet is made and shipped back the next day. The helmets are manufactured with the commercially available resin Surlyn (DuPont, Wilmington, DE, USA), which is a thermoplastic ionomer resin and the random copolymer (ethylene-co-methacrylic acid) (Fig. 5). The properties of the resin make it an ideal material for molding cranial helmets. The helmet shell is made of Surlyn, and the inner padding is made of an ethylene acetate lining. Reston foam pads (3M, St Paul, MN, USA) are used to keep the helmet from shifting inappropriately. The orthotist makes the necessary adjustments, and then the patient starts wearing the helmet for a sequentially increasing duration until it is worn for about 23 hours per day. New helmets are made and fitted as the infant rapidly grows, and the head shape changes to reach the desired form (Fig. 6).

Results

Sagittal Suture

More than 250 patients have been treated during the past 12 years using the described techniques. Using CI as a criterion, the patients were divided into 3 categories: excellent: CI greater than 80; good: CI between 70 and 79; and poor: CI less than or equal to 69. Our results indicate that 87% of the patients obtained an excellent outcome, 9% a good outcome, and 4% a poor outcome. The patients with poor outcome were most likely to have parents who were noncompliant with helmet therapy or usage. Also in this group were those patients whose helmets were not properly

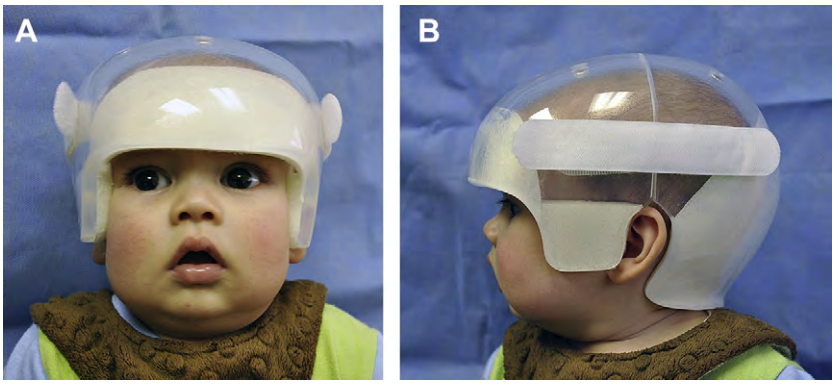

Fig. 5. (*A, B*) Surlyn helmet completely covers the cranial vault and helps to redirect brain growth rather than restrict it as shown in these anteroposterior and lateral photographs.

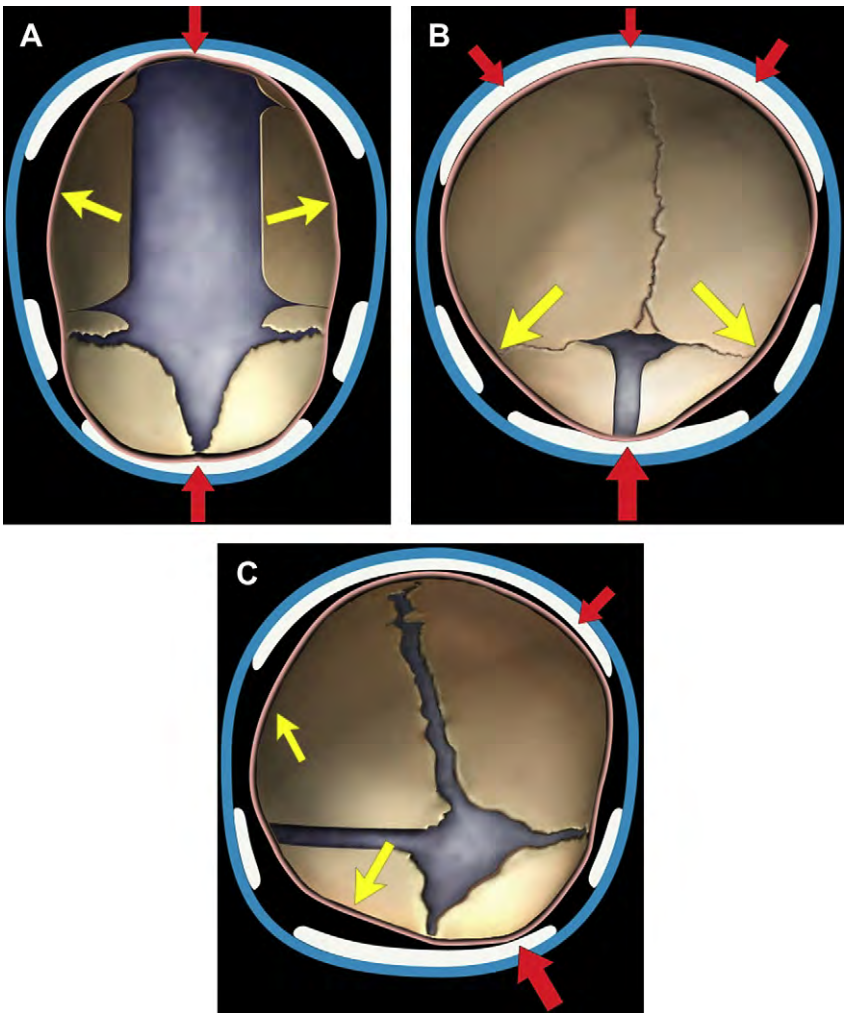

Fig. 6. (*A*) The helmet in sagittal synostosis allows for expansion of the temporal and parietal areas to achieve full correction of the scaphocephalic skull (*arrows*). (*B*) In the case of metopic synostosis, the helmet places pressure on the midforehead while allowing bitemporal and bifrontal expansions of the head (*arrows*). (*C*) For coronal synostosis, pressure is placed on the bossed contralateral frontal bone while allowing expansion of the recessed ipsilateral frontal area (*arrows*).

manufactured or fitted properly, leading to an inadequate head shape. The average patient wore 3 helmets for the duration of the treatment. About 95% of the patients stayed overnight and were discharged from the hospital next day. The overall blood transfusion rate was 8%, with only 2 patients receiving intraoperative blood transfusion. The mean surgical time was 57 minutes.

Case 1

A female infant aged 4 months and 3 weeks presented with sagittal synostosis, frontal bossing, and scaphocephalic features (Fig. 7). She underwent the endoscopic procedure as described earlier. Surgical time was 63 minutes. The estimated blood loss (EBL) was 15 mL. She did not undergo intra- or postoperative blood transfusion. She was discharged the morning after surgery and wore a total of 3 helmets over the next 12 months. Marked improvement was seen at her 7-month follow-up visit (Fig. 8). Her preoperative CI increased from 64 before surgery to 79 at 1.5 years after surgery (Fig. 9). Her CI at 4 years was 78. Her correction was maintained and can be appreciated at 4 years (Fig. 10) after the surgical intervention.

Case 2

A 4-week-old male infant was diagnosed with sagittal craniosynostosis and referred to our center for treatment (Figs. 11–15). He was a product of an uncomplicated pregnancy and delivery, who

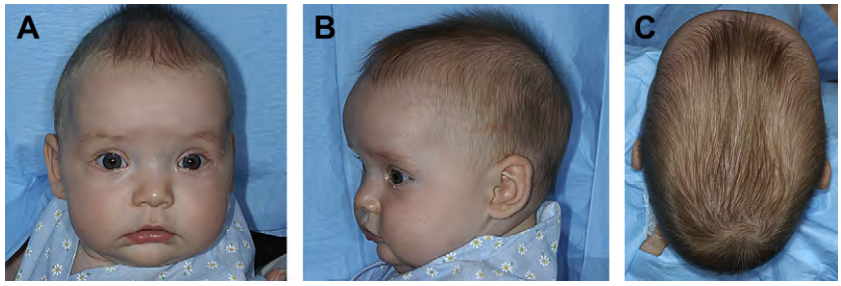

Fig. 7. (*A*) A 4-month-old female infant presented with bitemporal narrowing and frontal bossing as seen in this preoperative anteroposterior photograph. (*B*) Marked frontal bossing is seen in this preoperative lateral photograph. (*C*) Classical scaphocephalic shape as seen from above, with an elongated cranium and prominent forehead.

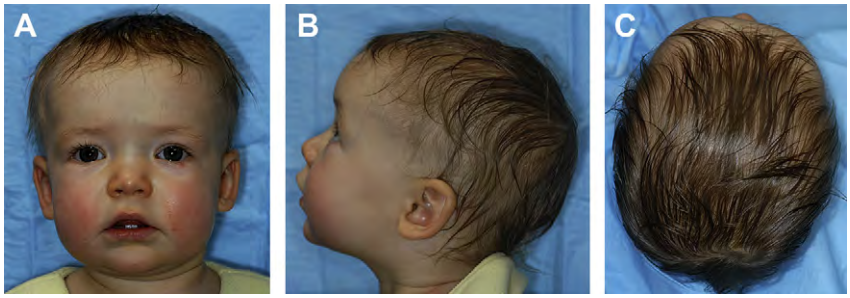

Fig. 8. (*A*) Seven months after surgery, the patient shows increased euryon-to-euryon distance and normalization of the facial features. (*B*) The forehead has recessed and the occipital area rounded to a more normal shape. (*C*) This topview photograph shows that the occipital is no longer pointed and elongated as in the preoperative images.

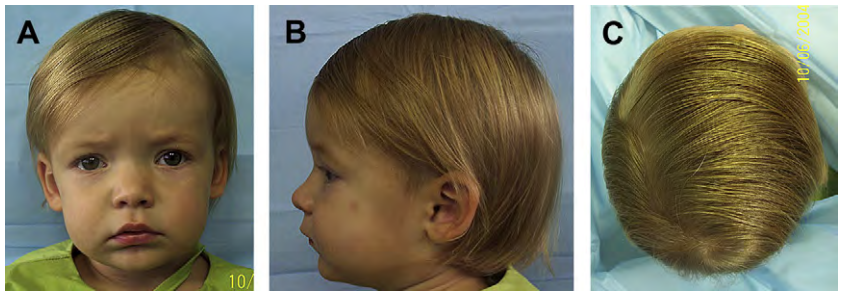

Fig. 9. (*A*−*C*) The patient has achieved a normal shape 1.5 years after surgery and is no longer scaphocephalic. Her CI is 79.

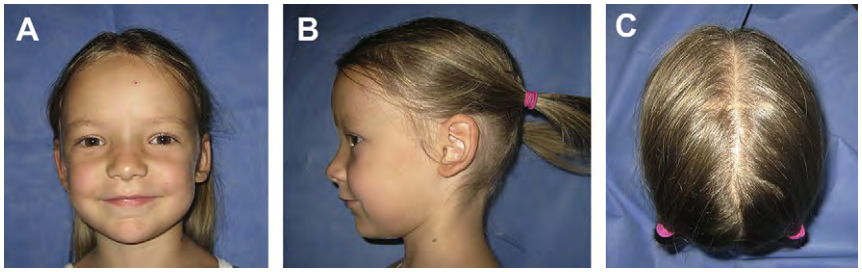

Fig. 10. (*A*−*C*) Four years after surgery the patient has maintained a normal shape and a CI of 78. No evidence of bumps, lumps, or indentations is seen as it is commonly encountered with the calvarial vault remodeling techniques.

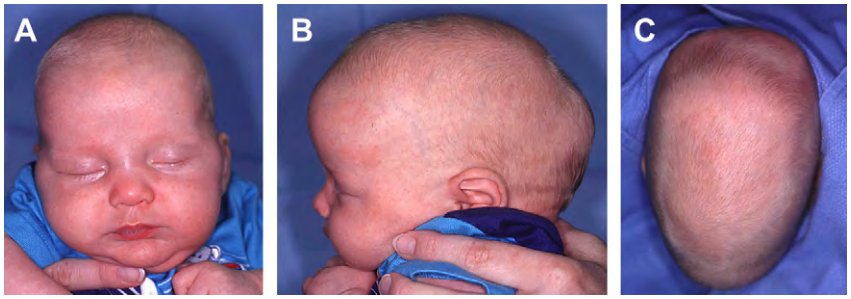

Fig. 11. (*A*) Preoperative anteroposterior photograph. (*B*) Lateral view shows marked forehead bossing with tip of forehead in line with patient's nose. (*C*) Top view shows significant scaphocephaly and head elongation.

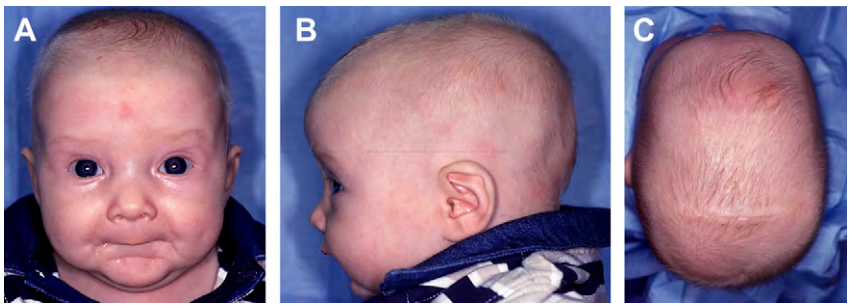

Fig. 12. (*A*−*C*) Early and prominent changes can be seen 5 weeks after surgery. Forehead recession as well as significant increase in the biparietal diameter is noted.

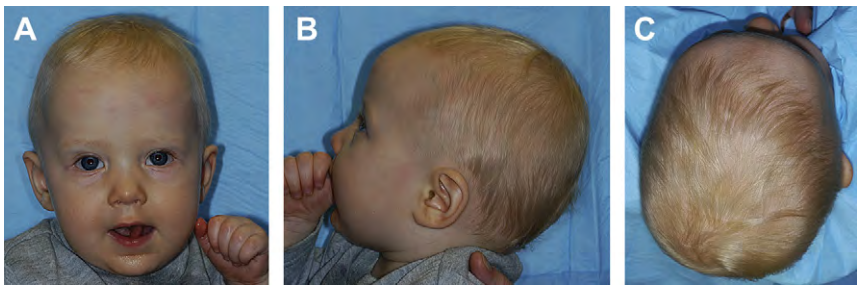

Fig. 13. (*A*−*C*) Continued improvement 8 months after surgery is demonstrated in these series of photographs.

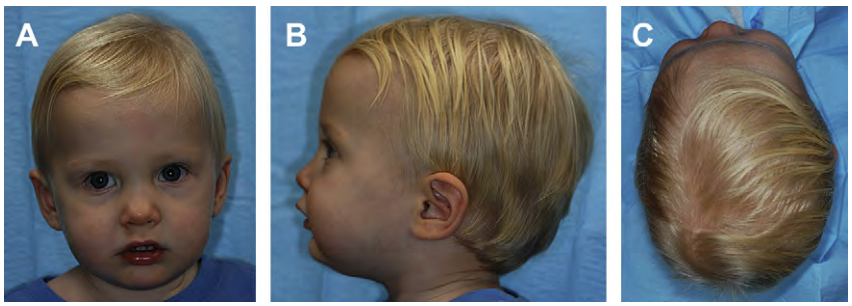

Fig. 14. (*A*−*C*) One and a half years after surgery persistent correction normalization of head shape has been achieved. Note the marked correction of the forehead prominence found before surgery.

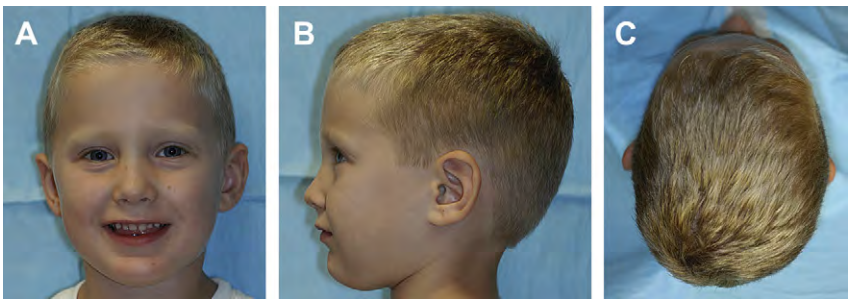

Fig. 15. (*A*−*C*) The patient looks normal and with no evidence of any preoperative deformities 5 years after surgery. The child's development is normal and shows no asymmetry or abnormality in the forehead or cranium.

presented with no significant medical or family history. The patient underwent the endoscope-assisted procedure as described at 6 weeks of age. Surgical time was 39 minutes and EBL was 20 mL. There was no evidence of venous air embolism, and the patient was discharged the morning after surgery. He did not require a blood transfusion and was treated with a helmet for 14 months.

Coronal Suture

More than 100 patients were treated, ranging in ages between 4 weeks and 10 months with a median age of 3.1 months. All patients presented with the classical plagiocephalic findings that included vertical dystopia, nasal deviation, craniofacial scoliosis, and ipsilateral forehead flattening. Cranial anthropometric measurements were used to quantify changes and included midsagittal axis deviation, vertical dystopia elevation, and angulation. Results indicate that 51% of the patients obtained full (100%) correction of the orbital dystopia and two-thirds obtained greater than 80% correction from preoperative baseline. The cranial scoliosis with sagittal imbalance and deviation of the cranial vault from midface and lower face showed full correction (100%) from preoperative baseline in 56% of the patients, with 80% of the patients achieving at least 80% correction. The average length of stay was 1 day. The mean EBL was 20 mL. One patient required an intraoperative blood transfusion (1.2%), and none needed postoperative transfusions. Mean operative time was 56 minutes.

Case 1

A 3-month-old female infant presented at 8 weeks with right coronal craniosynostosis. Preoperative photographs (Fig. 16) show right frontal plagiocephaly, vertical dystopia, and marked depression of the temporal and pterional areas. The procedure lasted 45 minutes, and she did not require blood transfusions (intra- or postoperatively). The patient was discharged the morning after the surgery and was treated with a helmet for 11 months. Photographs 1 year after surgery show that there has been a marked correction of the vertical dystopia, and although the right forehead is still recessed, it shows improvement (Fig. 17). The patient shows complete correction of the orbital dystopia 2 years after surgery and achieves bilateral supraorbital bar symmetry (Fig. 18).

Case 2

A 2-month-old male infant with the diagnosis of right coronal craniosynostosis presented to our craniofacial clinic for treatment. His preoperative photographs (Fig. 19) show vertical dystopia with marked exotropia and nasomandibular deviation to the contralateral side. In addition, the top view of the patient shows severe right frontal plagiocephaly, right ocular proptosis, and supraorbital rim recession and nasal deviation. The patient underwent endoscopic craniectomy at 2 months age. The width of the craniectomy was 4 mm and the length 12 cm. The EBL was 10 mL, and there was no need for intra- or postoperative blood transfusions. Total surgical time was 31 minutes, and the patient was discharged next morning. Helmet therapy was instituted as per protocol.

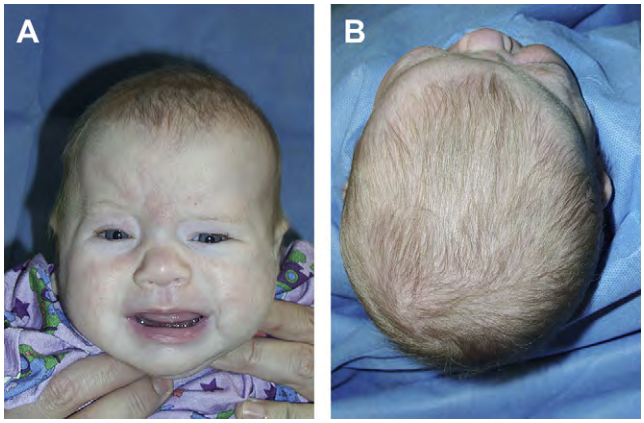

Fig. 16. (*A*) Preoperatively, this 8-week-old female infant presented with facial asymmetry, vertical dystopia, and elevation of right orbit and appreciable depression of the right temporal area. (*B*) Top view shows noticeable right forehead recession, ocular proptosis, and left forehead prominence.

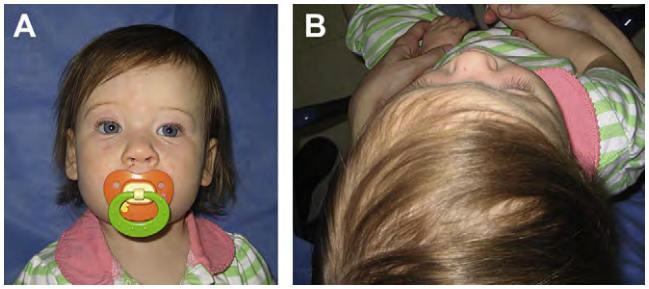

Fig. 17. (*A*) The temporal depression is no longer present and the dystopia significantly corrected 1 year after surgery. (*B*) Top view shows that there has been anterior movement of the right frontal bone and supraorbital rim complex. Still asymmetric, the forehead has improved, and proptosis is barely noticeable.

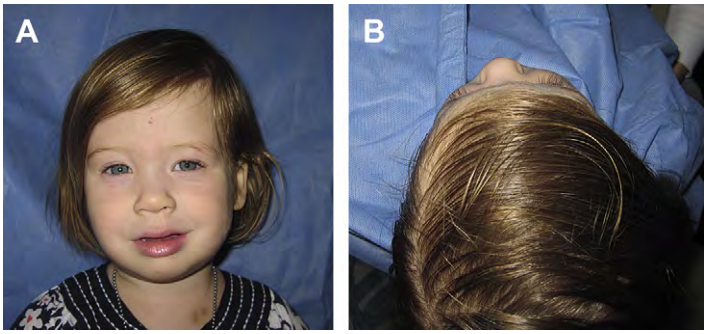

Fig. 18. (*A*) The patient continues to demonstrate a more normal appearance with barely noticeable asymmetry of the facial features 2 years after surgery. (*B*) Top view shows full correction of the forehead asymmetry. No longer proptotic, the right eye is symmetric with the left eye, as are the supraorbital rims.

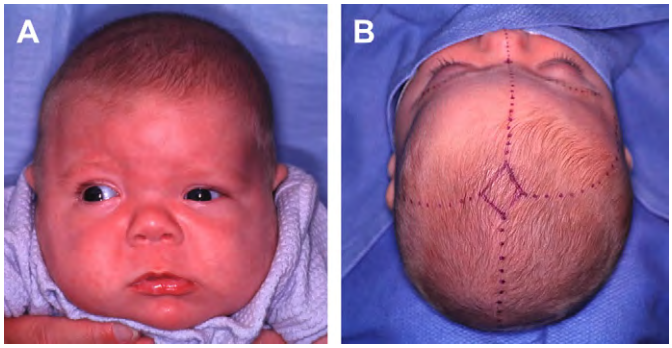

Fig. 19. (*A*) An 8-week-old male infant presented with right coronal craniosynostosis. Right exotropia as well as vertical dystopia and nasal deviation to the contralateral side is noted. (*B*) Right frontal plagiocephaly and ocular proptosis is seen preoperatively.

Photographs 3 years after surgery (Fig. 20) show that the patient's vertical dystopia and the exotropia had corrected without the need for ocular surgery. The face is realigned on the sagittal plane, and the frontal plagiocephaly has also fully corrected. The supraorbital rims show full symmetry, and the nose is no longer deviated. His kindergarten photograph shows a normal child with no evidence of residual craniosynostosis sequelae (Fig. 21).

Metopic Suture

During the last 12 years, 100 patients presenting with metopic suture craniosynostosis have been treated by our center using endoscope-assisted techniques as described in the previous section. All selected patients had trigonocephaly with bifrontal recession, a prominent midline ridge, and hypotelorism. Patients with the classical metopic ridge and with normal forehead contour and orbital position were excluded. There were 66 boys and 34 girls, and their ages ranged between 6 weeks and 10 months, with a mean of 3.8 months. The average surgical time was 57 minutes and the EBL was 30 mL. There was 1 intraoperative and 6 postoperative blood transfusions for a total transfusion rate of 7.1%. All of the patients, except for one, were discharged the morning after the surgery. All of the patients had increased interpupillary distance with orbital separation horizontally and correction of the preoperative hypotelorism. Most patients had correction of their forehead posterior angulation and

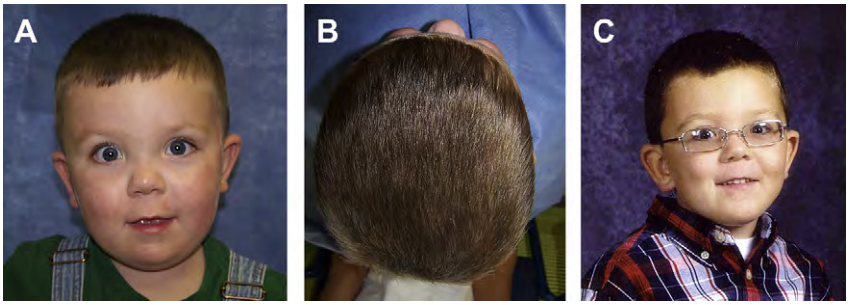

Fig. 20. (*A*) Postoperative photograph of a boy 3 years after undergoing the endoscopic craniectomy; his vertical dystopia has fully corrected as well as the exotropia, which did not require ocular surgery. (*B*) Top view shows forehead symmetry and malar alignment. (*C*) Kindergarten photograph shows persistent correction at 5 years of age.

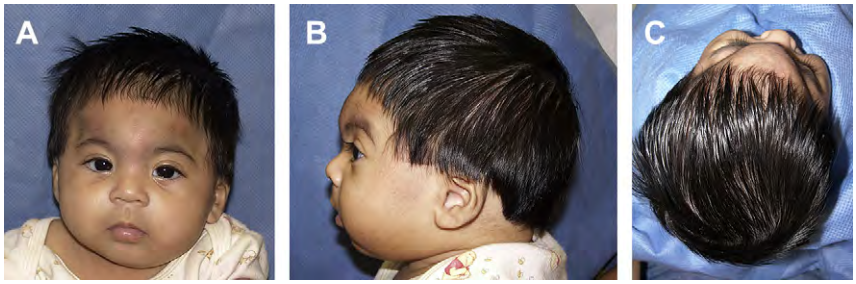

Fig. 21. (*A*) An anteroposterior preoperative photograph of a 5-week-old infant shows frontal recession associated with metopic synostosis. (*B*) Lateral view depicts prominent midline ridge. (*C*) Top view demonstrates severe trigonocephaly and bifrontal lateral recession.

recession. Unlike calvarial vault remodeling techniques, the patients did not develop turricephaly nor did they have forehead asymmetry as seen with indentations, depressions, or prominent bony ridges or palpable metal plates. Three patients developed subgaleal fluid collections at the surgical site, which required treatment with spinal drains or taps. None of the patients required reoperation to resolve the problem.

Case 1

A 5-week-old male infant was diagnosed with metopic craniosynostosis and trigonocephaly. He underwent surgery for an endoscopic craniectomy of the metopic suture and postoperative helmet therapy at 8 weeks of age. Surgical time was 47 minutes and EBL 30 mL; there was no need for intra- or postoperative blood transfusions. The patient was discharged on postoperative day 1. A helmet was worn for 10 months. Preoperative photographs (see Fig. 21) show hypotelorism, a prominent midline ridge, and evident trigonocephaly with recessed frontal bones. Postoperative photographs 2 years later (Fig. 22) show correction of hypotelorism and trigoncephaly.

Case 2

A 6-week-old male infant with metopic synostosis and trigonocephaly (Fig. 23) presented with constant irritability and crying in addition to his cranial deformities. He underwent surgery at 9 weeks of age. Surgical time was 58 minutes, and blood loss was 40 mL. There was no need for blood transfusion, and he was discharged the morning after surgery. Immediately after surgery, his crying and irritability began to decrease and had fully resolved within 2 weeks. On follow-up 6 years after surgery (Fig. 24), he showed normalization of his forehead abnormalities and had reached all milestones appropriately.

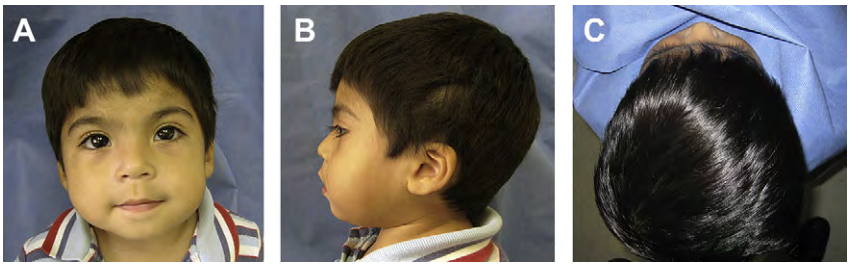

Fig. 22. (*A*) Two years after surgery, the patient shows normalization of the forehead and lateral orbital displacement. (*B*) Midline ridge is no longer present. (*C*) Forehead has a symmetric and rounded contour.

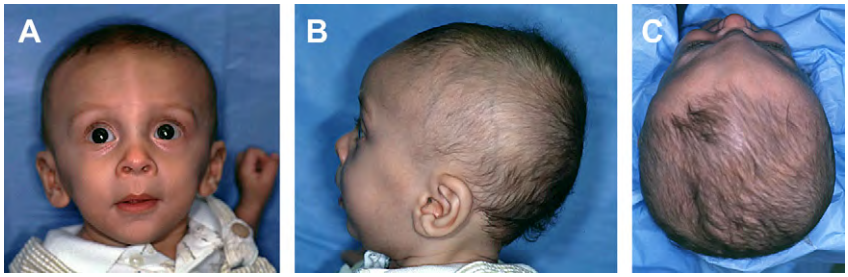

Fig. 23. (*A*) Midline ridge is noticed in this anteroposterior preoperative photograph of a 6-week-old infant with metopic synostosis. (*B*) Prominent forehead and midline ridge are visualized in a lateral view. (*C*) Midline ridge and trigonocephaly are shown in the top view.

Discussion

Review of our results in treating patients with minimally invasive endoscopy-assisted technique followed by helmet therapy and comparison with our results treating similar cohorts of patients using calvarial vault remodeling techniques show that the overall outcome of patients is much better with the former group. In all types of craniosynostosis, endoscopic techniques were associated with significantly less blood loss and blood transfusion rates. The same was the case with surgical times, anesthetic times, length of hospitalization, and postoperative facial swelling and bruising. Furthermore, long-term follow-up indicate sustained, long-lasting, and excellent results. Although somewhat fastidious and cumbersome at times, the use of molding helmets played, in our view, a significant role in achieving excellent results. Except for an occasional area of persistent redness of the skin, there were no problems encountered with the use of the helmets.

We believe that early treatment of these patients allows the treating surgeon to have better control of the outcomes. The rapidly growing brain is allowed to do most of the correction as it assumes normal morphology. Nowhere is this more evident than in our patients with coronal craniosynostosis. One of the problems that has plagued us in treating this condition at a later age (>9 months) is that the orbitocranial misalignment that sets in (vertical dystopia) is very difficult, if not impossible, to correct in an older child. Removal of the forehead and orbital bandeau does notcorrect the abnormally placed medial lateral and inferior orbital walls. Although excellent results are obtained at the time of surgery in terms of forehead symmetry, these benefits disappear over time as the child grows and commonly develops turricephaly and forehead asymmetry. We often treat teenagers with deviated noses that never corrected after a calvarial remodeling operation. Another significant benefit is that only 4 of 102 patients required ocular muscle corrective surgery to treat strabismus and exotropia. Realignment of the orbits by the growing brain precludes the need for ophthalmic surgery. These findings have been corroborated by MacKinnon and colleagues [29].

Fig. 24. (*A*−*C*) Six years after surgery, the patient shows correction of the abnormalities seen before the endoscopic surgery was done.

Summary

Careful analysis of our long-term review (>10 years) of our results treating patients with craniosynostosis at an early age using minimally invasive endoscope-assisted techniques indicate that it is a safe, effective, and efficient way to care for this patient population. These techniques are associated with significantly less blood loss, transfusion rates, hospitalization stay, pain, and discomfort. Although somewhat technically challenging at times (particularly in patients with metopic synostosis), any surgeon adept at using endoscopes should be able to master these techniques. In an era where informed consent is a must, these procedures should be provided as an option to the parents and ought to be considered by the treating surgeon.

Acknowledgments

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Craniofacial Dysostosis Syndromes: Evaluation and Staged Reconstructive Approach

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The term craniofacial dysostosis is used in a general way to describe syndromal forms of craniosynostosis. These disorders are characterized by sutural involvement that not only includes the cranial vault but also extends into the skull base and midfacial skeletal structures. In the past, craniofacial dysostosis syndromes have been described by Carpenter, Apert, Crouzon, Sathre-Chotzen, and Pfeiffer. Although the cranial vault and cranial base are believed to be the regions of primary involvement, there is generally significant effect on midfacial growth and development. In addition to cranial vault dysmorphology, individuals with these inherited conditions exhibit a characteristic but variable total midface deficiency that is syndrome specific and must be addressed as part of the staged reconstructive approach. Advances in molecular genetics now offer a more accurate understanding of the basic biology of these syndromes.

Genetic aspects

Fibroblast growth factor receptor (FGFR)-related craniofacial dysostosis syndromes include FGFR1-related craniosynostosis (Pfeiffer syndrome types 1, 2, and 3), FGFR2-related craniosynostosis (Apert syndrome, Beare-Stevenson syndrome, Crouzon syndrome), FGFR2-related isolated coronal synostosis (Jackson-Weiss syndrome, Pfeiffer syndrome types 1, 2, and 3), and FGFR3-related craniosynostosis (Crouzon syndrome with acanthosis nigricans, FGFR3-related isolated coronal synostosis, Muenke syndrome).

The 8 disorders considered as part of the *FGFR*-related craniosynostosis spectrum are Pfeiffer syndrome, Apert syndrome, Crouzon syndrome, Beare-Stevenson syndrome, *FGFR2*-related isolated coronal synostosis, Jackson-Weiss syndrome, Crouzon syndrome with acanthosis nigricans, and Muenke syndrome. All but Muenke syndrome and *FGFR2*-related isolated coronal synostosis generally present with bicoronal synostosis or cloverleaf skull anomaly.

The diagnosis of Muenke syndrome *(FGFR3*-related coronal synostosis) is based on identification of a disease-causing mutation in the *FGFR3* gene. The diagnosis of *FGFR2*-related isolated coronal synostosis is based on identification of a disease-causing mutation in the *FGFR2* gene. The diagnosis of the other 6 *FGFR*-related craniosynostoses is based on clinical findings; however, molecular genetic testing of the *FGFR1*, *FGFR2*, and *FGFR3* genes may be helpful in establishing the diagnosis of these syndromes in questionable cases.

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FGFR-related craniosynostosis is inherited in an autosomal-dominant manner. Affected individuals have a 50% chance of passing the mutant gene to each child. Prenatal testing is available; however, its use is limited by poor predictive value.

Molecular testing is necessary to establish the diagnosis for 2 of the disorders: Muenke syndrome and *FGFR2*-related isolated coronal synostosis. Individuals with Muenke syndrome may have unilateral coronal synostosis or megalencephaly without craniosynostosis; the accurate diagnosis depends on identification of a disease-causing mutation in the *FGFR3* gene. *FGFR2*-related isolated coronal synostosis is characterized only by uni- or bicoronal craniosynostosis; the accurate diagnosis depends on identification of a disease-causing mutation in the *FGFR2* gene.

Functional considerations

Restricted Brain Growth and Intracranial Pressure

If the rapid brain growth that normally occurs during infancy is to proceed unhindered, the cranial vault and skull base sutures must expand during phases of rapid growth, resulting in marginal ossification. In craniosynostosis, premature fusion of the suture(s) causes limited and abnormal skeletal expansion in the presence of continued brain growth. Depending on the number and location of prematurely fused sutures, the growth of the brain may be restricted. If surgical release of the affected suture(s) and reshaping of the involved skeleton to restore a more normal intracranial volume and configuration are not performed, decreased cognitive and behavioral function is likely to be the result.

Increased intracranial pressure (ICP) is the most serious functional problem associated with premature suture fusion. Radiographic findings that may suggest an increased ICP include the beaten-copper appearance along the inner table of the cranial vault seen on a plain radiograph or the loss of brain cisternae as observed on a computed tomographic (CT) scan. Although suggestive of increased ICP, these are considered soft findings.

Increased ICP is most likely to affect patients with great disparity between brain growth and intracranial capacity and is believed to occur in as many as 42% of untreated children in whom 2 or more sutures are affected. There is no agreement on what levels of ICP are normal at any given age in infancy and early childhood.

The clinical signs and symptoms related to increased ICP may have a slow onset and be difficult to recognize in the pediatric population. Although standardized CT scans allow for indirect measurement of intracranial volume, it is not yet possible to use these studies to make judgments as to who requires craniotomy for brain decompression. Comprehensive neurologic and ophthalmologic evaluation are critical components of the data gathering required to formulate definitive treatment plans in patients with one of the craniofacial dysostosis syndromes.

Vision

Untreated craniosynostosis with increased ICP may cause papilledema and eventual optic nerve atrophy, resulting in partial or complete blindness. If the orbits are shallow (exorbitism) and the eyes are proptotic (exophthalmus), as occurs in the craniofacial dysostosis syndromes, the cornea may be exposed and abrasions or ulcerations may occur. An eyeball extending outside a shallow orbit is also a risk for trauma. If the orbits are extremely shallow, herniation of the globe itself may occur, necessitating emergency reduction followed by tarsorrhapies or urgent orbital decompression.

Some forms of craniofacial dysostosis (eg, Apert syndrome) result in a degree of orbital hypertelorism, which may compromise visual acuity and restrict binocular vision. Divergent or convergent nonparalytic strabismus or exotropia occurs frequently and should be considered during the diagnostic evaluation. This condition may be the result of congenital anomalies of the extraocular muscles themselves.

Hydrocephalus

Hydrocephalus affects as many as 10% of patients with a craniofacial dysostosis syndrome. The risk of intracranial hypertension is greatest in Crouzon syndrome. Even if every medical complication

is managed promptly, a proportion of affected children develop congnitive delay and neurologic problems. Although the cause is often not clear, hydrocephalus may be secondary to a generalized cranial base stenosis with constriction of all the cranial base foramina, which affects the patient's cerebral venous drainage and cerebrospinal fluid (CSF) flow dynamics. Hydrocephalus may be identified with the help of a CT scan or magnetic resonance imaging to document progressively enlarging ventricles. Difficulty exists in interpreting ventricular findings as seen on a CT scan especially when the skull and cranial base are brachycephalic. The skeletal dysmorphology seen in a child with cranial dysmorphology related to craniosynostosis (eg, bicoronal synostosis) may translate into an abnormal ventricular shape that is not necessarily related to abnormal CSF flow. Serial imaging with clinical correlation and experienced neurologic judgment is required in making these assessments.

Effects of Midface Deficiency on Airway

All newborn infants are obligate nasal breathers. Many infants born with a craniofacial dysostosis syndrome have moderate to severe hypoplasia of the midface as a component of their malformation. They have diminished nasal and nasopharyngeal spaces, with resulting increased nasal airway resistance (obstruction). The affected child is thus forced to breath through the mouth. For a newborn infant to ingest food through the mouth requires sucking from a nipple to achieve negative pressure as well as an intact swallowing mechanism. The neonate with severe midface hypoplasia experiences diminished nasal airflow and is unable to accomplish this task and breathe through the nose at the same time. Complicating this clinical picture may be an elongated and ptotic palate (eg, Apert syndrome), and enlarged tonsils and adenoids. The compromised infant expends significant energy respiring and this may push the child into a catabolic state (negative nitrogen balance). Failure to thrive results unless either nasogastric tube feeding is instituted or a feeding gastrostomy is placed. Evaluation by a pediatrician, pediatric otolaryngologist, and feeding specialist with craniofacial experience can help distinguish minor feeding difficulties from those requiring more aggressive treatment.

Sleep apnea of central, obstructive, or mixed origin may also be present. If the apnea is found to be secondary to upper airway obstruction based on a formal sleep study, a tracheostomy may be indicated. In specific situations, early midface advancement may be performed to improve the airway, allowing for tracheostomy decannulation. Central apnea may occur from poorly treated intracranial hypertension as well as other contributing factors. If so, the condition may improve by reducing the ICP through brain decompression. This goal is accomplished with cranioorbital or posterior cranial vault expansion.

Dentition and Occlusion

The incidence of dental and oral anomalies is higher among children with craniofacial dysostosis syndromes than within the general population. In Apert syndrome in particular, the palate is high and constricted in width. The incidence of isolated cleft palate in patients with Apert syndrome approaches 30%. Clefting of the secondary palate may be submucous, incomplete, or complete. Delayed dental eruption should also be expected. Confusion has arisen about whether the oral malformations and absence of teeth that are often characteristic of these conditions are a result of congenital or iatrogenic factors (eg, injury to dental follicles associated with early midface surgery). The midfacial hypoplasia seen in the craniofacial dysostosis syndromes often results in limited maxillary alveolar bone to house a full compliment of teeth. The result is severe crowding, which often requires serial extractions to address it. An Angle class III skeletal relationship in combination with anterior open bite deformity is typical.

Hearing

Hearing deficits are more common among patients with the craniofacial dysostosis syndromes than among the general population. In Crouzon syndrome, conductive hearing deficits are common, and atresia of the external auditory canals may also occur. Otitis media is more common in Apert syndrome, although the exact incidence is unknown. Middle ear disease may be related to the presence of a cleft palate that results in dysfunction of the eustachian tube. Congenital fixation of the stapedial footplate is also believed to be a frequent finding. The possibility of significant hearing loss

is paramount and should not be overlooked because of preoccupation with other more easily appreciated craniofacial findings.

Morphologic considerations

General

Examination of the patient's entire craniofacial region should be meticulous and systematic. The skeleton and soft tissues are assessed in a standard way to identify all normal and abnormal anatomy. Specific findings tend to occur in particular malformations, but each patient is unique. The achievement of symmetry, normal proportions, and the reconstruction of specific aesthetic units is essential to forming an unobtrusive face in a child born with one of the craniofacial dysostosis syndromes.

Frontoforehead Aesthetic Unit

The frontoforehead region is dysmorphic in an infant with craniofacial dysostosis. Establishing the normal position of the forehead is critical to overall facial symmetry and balance. The forehead may be considered as 2 separate aesthetic components: the supraorbital ridge-lateral orbital rim region and the superior forehead. The supraorbital ridge-lateral orbital rim region includes the nasofrontal process and supraorbital rim extending inferiorly down each frontozygomatic suture toward the infraorbital rim and posteriorly along each temporoparietal region. The shape and position of the supraorbital ridge-lateral orbital rim region are a key element of upper facial aesthetics. In a normal forehead, at the level of the nasofrontal suture, an angle ranging from 90° degrees to 110° is formed by the supraorbital ridge and the nasal bones when viewed in profile. In addition, the eyebrows, overlying the supraorbital ridge, should be anterior to the cornea. When the supraorbital ridge is viewed from above, the rim should arc posteriorly to achieve a gentle 90° angle at the temporal fossa with a center point of the arc at the level of each frontozygomatic suture. The superior forehead component, about 1.0 to 1.5 cm up from the supraorbital rim, should have a gentle posterior curve of about 60°, leveling out at the coronal suture region when seen in profile.

Posterior Cranial Vault Aesthetic Unit

Symmetry, form, and adequate intracranial volume of the posterior cranial vault are closely linked. Posterior cranial vault flattening may result from either a unilateral or bilateral lambdoidal synostosis, which is rare, previous craniectomy with reossification in a dysmorphic flat shape, which is frequent, or postural molding because of repetitive supine sleep positioning. A short anterior-posterior cephalic length may be misinterpreted as an anterior cranial vault (forehead) problem when the occipitoparietal (posterior) skull represents the primary region of the deformity. Careful examination of the entire cranial vault is essential to defining the dysmorphic region so that when indicated appropriate cranial vault expansion may be performed.

Orbitonasozygomatic Aesthetic Unit

In the craniofacial dysostosis syndromes, the orbitonasozygomatic regional deformity is a reflection of the cranial base malformation. For example, in Crouzon syndrome when bilateral coronal suture synostosis is combined with skull base and midfacial deficiency, the orbitonasozygomatic region is dysmorphic and consistent with a short (anterior-posterior) and wide (transverse) anterior cranial base. In Apert syndrome, the nasal bones, orbits, and zygomas, like the anterior cranial base, are transversely wide from anterolateral bulging of the temporal lobes of the brain and horizontally short (retruded), resulting in a shallow hypertelloric reverse curved upper midface (zygomas, orbits, and nose). Surgically advancing the midface without simultaneously addressing the increased transverse width and reverse curve does not adequately correct the dysmorphology.

Maxillary-nasal Base Aesthetic Unit

In the patient with craniofacial dysostosis with midface deficiency, the upper anterior face is vertically short (nasion to maxillary incisor), and there is a lack of horizontal (anterior-posterior)

projection of the midface. These findings may be confirmed through cephalometric analysis that indicates an SNA angle below the mean value and a short upper anterior facial height (nasion to anterior nasal spine). The width of the maxilla in the dentoalveolar region is generally constricted, with a high arched palate. To normalize the maxillary-nasal base region, multidirectional surgical expansion and reshaping are generally required. The abnormal maxillary lip-to-tooth relationship and occlusion are improved through Le Fort I segmental osteotomies and orthodontic treatment as part of the staged reconstruction. The mandible and chin are frequently secondarily deformed and benefit from surgical repositioning as part of the orthognathic correction.

Surgical management

General Considerations

Philosophy regarding timing of intervention

In considering the timing and type of intervention the experienced surgeon should take several biologic realities into account: the natural course of the malformation (ie, is the dysmorphology associated with Crouzon syndrome progressively worsening or only a nonprogressive craniofacial deformity?); the tendency toward growth restriction of operated bones (aesthetic units) that have not yet reached maturity (ie, we know that operating on a cleft palate in infancy causes scarring and later results in maxillary hypoplasia in many individuals); and the relationship between the underlying growing viscera (ie, brain or eyes) and the congenitally affected and surgically altered skeleton (ie, in Crouzon syndrome if the cranial vault is not surgically expanded to decompress the brain by 1 year of life is brain compression likely to occur?).

In attempting to limit impairment and also achieve long-term preferred facial aesthetics and head and neck function an essential question the surgeon must ask is, "During the course of craniofacial development, does the operated-on facial skeleton of the child with craniofacial dysostosis tend to grow abnormally, resulting in further distortions and dysmorphology, or are the initial positive skeletal changes achieved (at operation) maintained during ongoing growth?" The proposed theory that craniofacial procedures performed early in infancy unlock growth has not been documented through the scientific method.

Incision placement

For exposure of the craniofacial skeleton above the Le Fort I level, the approach used is the coronal (skin) incision. This approach allows for a camouflaged access to the anterior and posterior cranial vault, orbits, nasal dorsum, zygomas, upper maxilla, pterygoid fossa, and temporomandibular joints. For added cosmetic advantage, placement of the coronal incision more posteriorly on the scalp and with postauricular rather than preauricular extensions is useful. When exposure of the maxilla at the Le Fort I level is required, a circumvestibular maxillary intraoral incision is used. Unless complications occur that warrant unusual exposure, no other incisions are required for managing any aspect of the reconstruction of the patient with craniofacial dysostosis. These incisions (coronal [scalp] and maxillary [circumvestibular]) may be reopened as needed to further complete the individual's staged reconstruction.

Management of the cranial fossa dead space and communication across the skull base after total midface advancement

Cranial reshaping in the patient with craniofacial dysostosis provides space for the compressed brain to expand into. After anterior cranial vault expansion and monobloc advancement an immediate extradural (retrofrontal) dead space is combined with the gap created by osteotomy across the skull base (connecting the anterior cranial fossa and the nasal cavity). This combination of factors may complicate the postoperative recovery (eg, CSF leakage, infection, bone loss, fistula formation). After frontofacial advancement the nasal cavity-cranial fossa communication is managed by being gentle to the tissues; good hemostasis; effective repair of any dural tears (dural grafting as needed); complete separation of dural and nasal mucosal tissue planes by interposing a combination of bone grafts, tissue sealants, and flaps; avoidance of pressure gradients across the opening while the nasal mucosa is healing; and prevention of over- or undershunting (when a shunt is in place).

The preferred way to manage the retrofrontal (lobes of the brain) dead space and the gap across the skull base osteotomy site (separating the cranial fossa and nasal cavities) after frontofacial advancement remains controversial but it is a critical aspect of the reconstruction.

In the patient with craniofacial dysostosis rapid filling of the surgically expanded intracranial volume (6−8 weeks) by the previously compressed frontal lobes of the brain has been documented after cranioorbital expansion in infants. It has also been shown to occur after frontofacial advancement in children and young adults when the volume increase remains in a physiologic range. These observations support the conservative management of the retrofrontal dead space in younger patients. More gradual and less complete filling of the space is believed to occur in older children and adults. If so, this process may be particularly troublesome when the anterior cranial fossa dead space communicates directly with the nasal cavity (ie, monobloc advancement, facial bipartition, intracranial Le Fort III) across the (open-gap) skull base interface. When feasible, closing off (sealing) the nasal cavity from the cranial fossa across the skull base osteotomy at the time of operation is preferred. Insertion of a pericranial flap or other fillers can help to separate the cavities. The use of fibrin glue to seal the anterior cranial base provides a temporary separation between the cavities, allowing time for the reepithelialization (healing) of the torn nasal mucosa. To reconstruct the defect across the skull base (gap) bone grafts of various types may also be used. Until the torn nasal mucosa heals, potential communication between the nasal cavity and cranial fossa may result in the transfer of air, fluid, bacteria, and nasocranial fistula formation. To facilitate nasal mucosa healing and limit a pressure gradient across the communication, postoperative endotracheal intubation may be extended for 3 to 5 days and/or bilateral nasopharyngeal airways may be placed after extubation. The avoidance of positive pressure ventilation, enforcement of sinus precautions, and restriction of nose blowing further limit reflux of air, fluid, and bacteria (nose to cranial fossa) during the early postoperative period. When anterior cranial vault procedures are performed and aerated frontal sinuses are present, management is by either cranialization or obliteration.

Aside from a learning curve in mastering the technical skills of completing the monobloc osteotomies and disimpaction, the surgical morbidity from these procedures primarily results from a combination of the anticipated retrofrontal dead space, unavoidable tears in the nasal mucosa, and management of nasocranial communication across the skull base gap with the potential for fluid, air, and bacteria contamination.

The achievement of a normal occlusion is rarely a treatment objective at the time of monobloc advancement. Accomplishing an ideal occlusion without creating enophthalmus requires a separate Le Fort I osteotomy to differentially advance the maxilla, often combined with maxillary segmentation and mandibular (sagittal split) osteotomies. To achieve the most favorable facial balance for the patient with craniofacial dysostosis an experienced clinician's aesthetic sense of the preferred morphology and focused technical expertise to alter the skeleton intraoperatively are essential. Several key technical aspects include:

The ability to remove, segment, and then reshape and stabilize (plates and screws) the anterior cranial vault

The ability to separate the orbits and midface as a unit (monobloc) from the skull base

The ability to further segment the monobloc (at the upper orbits) and reconstruct (with cranial grafts) as needed.

The ability to separate the monobloc into halves (facial bipartition) and then alter the 2 facial halves to achieve the most favorable morphology; this process often requires simultaneously increasing the maxillary transverse width and decreasing the upper face width to correct hypertelorism of the orbits, zygomas, nose, and bitemporal regions (eg, Apert syndrome); facial bipartition also provides the ability to correct transverse facial arc-of-rotation deformities (eg changing the Apert syndrome patient's concave facial arc of rotation toward a normal convexity is an essential aspect of the reconstruction)

Any potential advantage of limiting morbidity caused by infection across the skull base with the distraction osteogenesis (DO) technique should be considered in light of limitations to achieve the key technical and aesthetic aspects mentioned earlier. Added morbidity with the DO technique caused by pin tract infection/scarring/loosening requiring reapplication, the need for device removal, and dependence on a patient's, family's, and clinician's continued commitment to staying the course

for necessary outpatient DO procedures/adjustments to achieve an acceptable result must also be factored into the decision-making process.

When a patient with craniofacial dysostosis is to undergo intracranial volume expansion as part of the craniofacial procedure and they also require hydrocephalus management, the potential for morbidity increases. Complications may arise from excessive CSF drainage (overshunting). With overshunting there is decreased brain volume to fill any surgically created retrofrontal dead space. Frontofacial advancement and/or cranial vault expansion procedures should be carefully staged with ventriculoperitoneal (VP) shunting procedures. We believe that the presence or absence of a VP shunt is not in itself a major risk factor in the success of a frontofacial advancement procedure. An important aspect is satisfactory physiologic function of the ventricular system. The decision regarding the need for and sequencing of shunting is based on the patient's neurologic findings and the neurosurgeon's judgment. In a patient with a VP shunt in place before the surgery, experienced neurosurgical evaluation, including CT scanning of the ventricular system, is performed to confirm physiologic shunt function.

Soft-tissue management

A layered closure of the coronal (scalp) incision (galea and skin) optimizes healing and limits scar widening. Resuspension of the midface periosteum to the temporalis fascia may facilitate redraping of the soft tissues. Each lateral canthus should be reattached in a superior-posterior direction to the newly repositioned lateral orbital rim. The use of chromic gut on the skin in children may be used to obviate postoperative suture or staple removal.

Crouzon syndrome

Crouzon syndrome is a frequent form of craniofacial dysostosis. It is characterized by multiple anomalies of the craniofacial skeleton with an autosomal-dominant inheritance pattern. Its manifestations are generally less severe than those of Apert syndrome and there are no malformations of the extremities. Typically, the cranial vault presentation is premature synostosis of both coronal sutures with a resultant brachycephalic shape to the skull. Cranial vault suture involvement other than coronal may include sagittal, metopic, or lambdoidal involvement, either in isolation or in any combination. The cranial base and upper face sutures are variably involved, resulting in a degree of midface hypoplasia with an Angle class III malocclusion. The orbits are hypoplastic, resulting in a degree of proptosis with additional orbital dystopia that may produce a mild orbital hypertelorism and flatness to the (transverse) arc of rotation of the midface.

Staging of Reconstruction for Crouzon Syndrome

Primary brain decompression: cranioorbital reshaping in infancy

The initial treatment of any craniofacial dysostosis syndrome generally requires cranial vault suture release and simultaneous anterior cranial vault and upper orbital osteotomies, with reshaping and advancement for brain decompression (Fig. 1). In general, our preference is to carry this out when the child is 9 to 12 months of age unless clear signs of increased ICP are identified earlier in life. Reshaping of the upper three-quarters of the orbital rims, supraorbital ridges, and tenon extensions is geared to decreasing the bitemporal and anterior cranial base width, with simultaneous horizontal advancement to increase the anterior-posterior dimension. This strategy also increases the depth of the upper orbits, with some improvement of eye proptosis. The overlying forehead is then reconstructed according to morphologic needs. A degree of overcorrection is considered at the level of the supraorbital ridge when the procedure is performed in infancy. In our opinion, by allowing additional growth to occur before surgery (waiting until the child is 9−12 months old), the reconstructed cranial vault and upper orbital shape is better maintained, with less need for repeat craniotomy procedures in early childhood. Preoperative evaluation and monitoring confirm that waiting until this age (9−12 months) does not risk compression of the underlying brain.

The goals at this stage are to provide increased space in the anterior cranial vault for the brain, to increase the orbital volume (which allows the eyes to be positioned more normally for better protection from exposure), and to improve the morphology of the forehead and upper orbits.

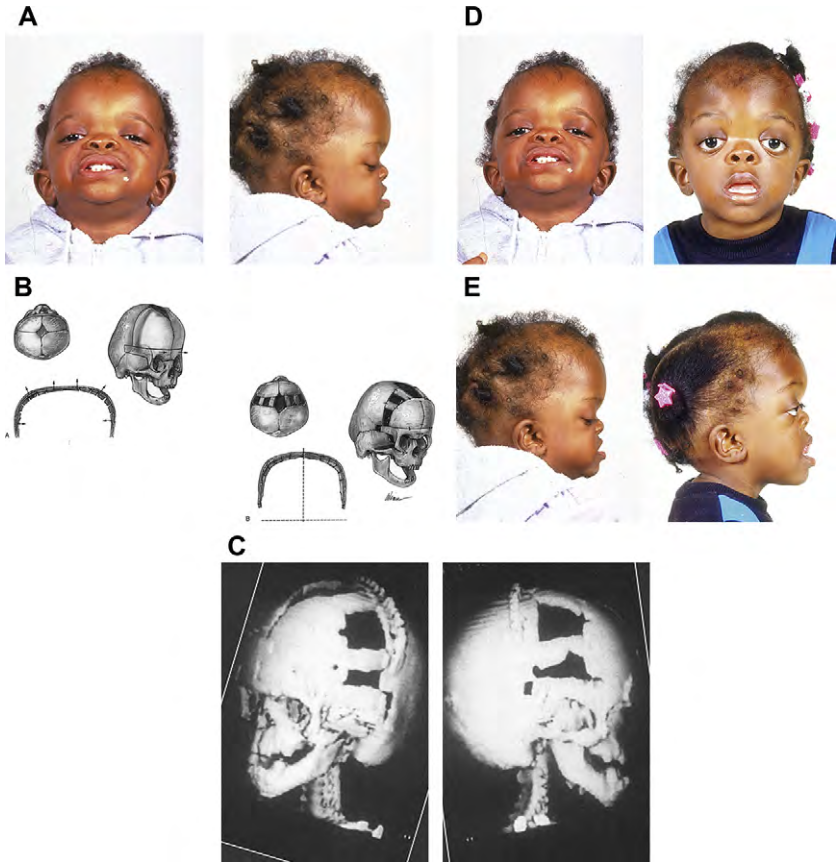

Fig. 1. An 18-month-old girl with brachycephaly and midface deficiency with a mild degree of papilledema was referred for evaluation. She was found to have bilateral coronal synostosis and midface hypoplasia without extremity anomalies. The diagnosis of Crouzon syndrome was confirmed. She underwent cranioorbital reshaping by the procedure described. Several months later, a ventriculoperitoneal shunt was placed for management of hydrocephalus. Further staged reconstruction will require a total midface advancement procedure later in childhood followed by orthodontic treatment and orthognathic surgery in the early teenage years. (*A*) Frontal and profile views before surgery. (*B*) Craniofacial deformity and planned operative procedure. (*C*) Three-dimensional CT scan views of craniofacial skeleton, 1 week after cranioorbital reshaping with advancement. (*D*) Frontal view before surgery and at 3 years of age, 1.5 years after undergoing cranioorbital decompression and reshaping. (*E*) Profile view before surgery and at 3 years of age, 1.5 years after undergoing first-stage cranioorbital decompression/reshaping. (*From* Posnick JC. Crouzon syndrome: evaluation and staging of reconstruction. In: Posnick JC, editor. Craniofacial and maxillofacial surgery in children and young adults, vol. 1. Philadelphia: WB Saunders; 2000. p. 271; with permission.)

A postauricular coronal (scalp) incision is made, and the anterior flap is elevated along with the temporalis muscle in the subperiosteal plane. Bilateral circumferential periorbital dissection follows, with detachment of the lateral canthi, but with preservation of the medial canthi and nasolacrimal apparatus to the medial orbital walls. The subperiosteal dissection is continued down the lateral and infraorbital rims to include the anterior aspect of the maxilla and zygomatic buttress. The neurosurgeon then completes the craniotomy to remove the dysmorphic anterior cranial vault. With retraction of the frontal and temporal lobes of the brain (remaining anterior to each olfactory bulb), direct visualization of the anterior cranial base and orbits at the time of osteotomy is possible.

The orbital osteotomies are then completed across the orbital roof and superior aspect of the medial orbital wall, laterally through the lateral orbital wall, and inferiorly just into the inferior orbital fissure. The three-quarters orbital osteotomy units, with their tenon extensions, are removed from the field. The orbital units are reshaped and reinset into a preferred position. Orbital depth is thereby increased and ocular globe proptosis is reduced. Fixation is generally achieved with interosseous wires at each infraorbital rim and with microplates and screws at the tenon extensions and frontonasal regions.

The removed calvaria is cut into segments, which are placed individually to achieve a more normally configured anterior cranial vault. The goal of reshaping is to narrow the anterior cranial base and orbital width slightly and provide more forward projection and overall normal morphology.

Further craniotomy for additional brain decompression: cranial vault expansion and reshaping in young children

After the initial suture release, brain decompression, and reshaping of the cranioorbital region are performed during infancy, the child is observed clinically at intervals by the craniofacial surgeon, pediatric neurosurgeon, pediatric ophthalmologist, and developmental specialist, and undergoes interval CT scanning. Should signs of increased ICP develop, urgent brain decompression with further cranial vault expansion and reshaping are performed. When increased ICP occurs, the suspected location of brain compression influences the region of the skull for which further expansion and reshaping are planned.

If the brain compression is judged to be anterior, then further anterior cranial vault and upper orbital osteotomies for expansion with reshaping and advancement are performed. The technique is similar to that described earlier (see Fig. 1). If the problem is posterior brain compression, expansion of the posterior cranial vault, with the patient in the prone position, is required (Fig. 2).

The repeat craniotomy performed for further brain decompression and cranial reshaping in the child with Crouzon syndrome is often complicated by brittle, irregular cortical bone (which lacks a diploic space and may contain sharp spicules piercing the dura), the presence of previously placed fixation devices in the operative field, and convoluted thin dura compressed against (or herniated into) the inner table of the skull. All of these problems result in a greater potential for dural tears during the calvarectomy than would normally occur during the primary procedure. A greater potential for morbidity should be anticipated when reelevating the scalp flap, dissecting the dura free of the inner table of the skull and cranial base, and then removing the cranial vault bone.

Management of total midface deformity in childhood

The type of osteotomies selected to manage the total midface deficiency/deformity and residual cranial vault dysplasia should depend on the extent and location of the presenting dysmorphology rather than on a fixed universal (tunnel vision) approach to the midface malformation. A main objective is to normalize the orbits, zygomas, and cranial vault, improving but not correcting the maxilla (at the Le Fort I level); correction of the maxillomandibular deformity requires orthognathic surgery. The selection of either a monobloc (with or without additional orbital segmentation), facial bipartition (with or without additional orbital segmentation), or Le Fort III osteotomy (intracranial vs extracranial) to manage the basic horizontal, transverse, and vertical upper and midface deficiencies/deformities in a patient with Crouzon syndrome depends on the patient's presenting midface and anterior cranial vault morphology. The observed dysmorphology is dependent on the original malformation, the previous procedures performed, and the effects of ongoing growth.

When evaluating the upper and midface in a child born with Crouzon syndrome if the supraorbital ridge is in good position when viewed from the sagittal plane (the depth of the upper orbits is adequate), the midface and forehead have a normal arc of rotation in the transverse plane (not concave), and the root of the nose is of normal width (minimal orbital hypertelorism), there is no need to reconstruct this region (the forehead and upper orbits) any further. In such patients, the basic residual midface deficiency/deformity is in the lower half of the orbits, zygomatic buttress, and maxilla. If so, the orbital-midface deformity may be effectively managed using an extracranial Le Fort III osteotomy (Fig. 3).

If the supraorbital region, anterior cranial base, zygomas, nose, lower orbits, and maxilla all remain deficient/deformed in the sagittal plane (horizontal retrusion), then a monobloc osteotomy is indicated (Fig. 4). In these patients, the forehead is generally flat and retruded and also requires reshaping and advancement. If upper midface hypertelorism (increased transverse width) and midface flattening (horizontal retrusion) with abnormal facial curvature (concave arc) are also present, then the monobloc unit is split vertically in the midline (facial bipartition), a wedge of interorbital (nasal and ethmoidal) bone is removed, and the orbits and zygomas are repositioned medially and the maxillary posterior arch is widened. A facial bipartition is rarely required in Crouzon syndrome but the monobloc is. When a monobloc or facial bipartition osteotomy is performed as the total midface procedure, additional segmentation of the upper and lateral orbits for reconstruction may also be required to normalize the morphology of the orbital aesthetic units.

In most cases, an error in judgment occurs if the surgeon attempts to simultaneously adjust the orbits and idealize the occlusion using the Le Fort III, monobloc, or facial bipartition osteotomy in isolation,

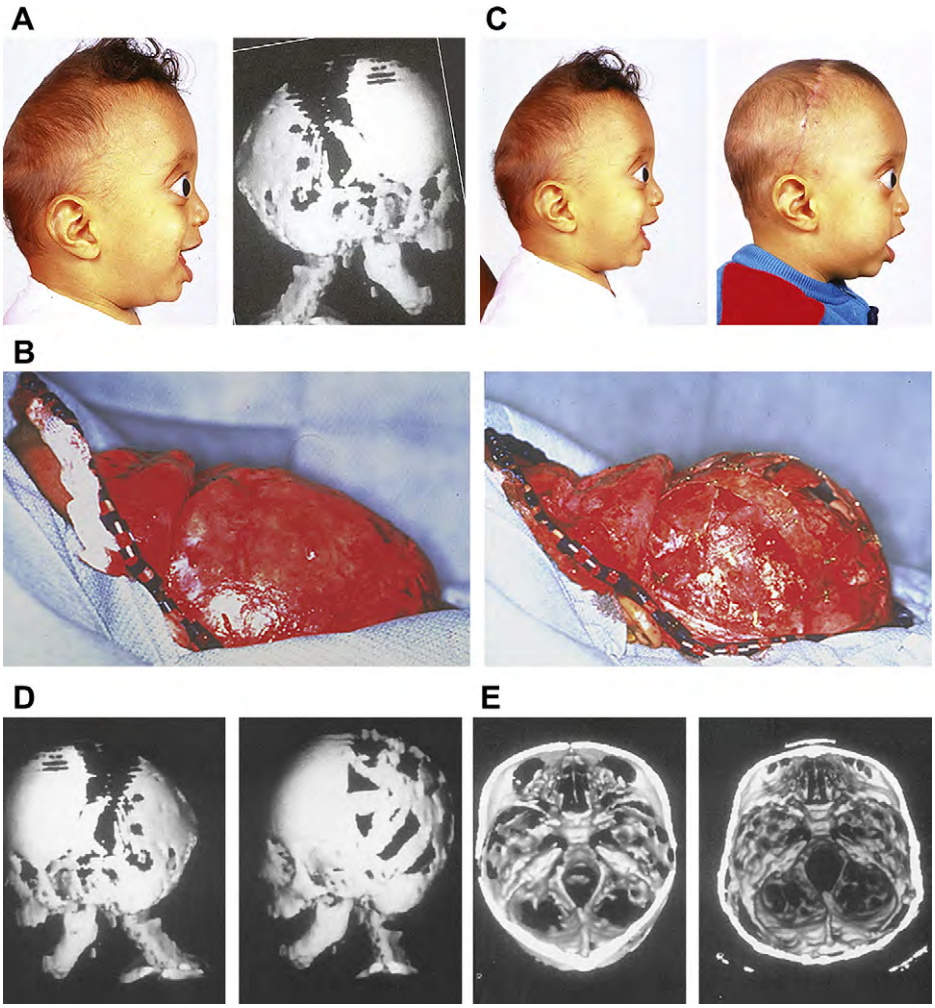

Fig. 2. A child with Crouzon syndrome is shown at 10 months of age. His deformities are characterized by mild bilateral coronal and marked bilateral lambdoid synostosis in combination with midface deficiency. He has diminished intracranial volume, resulting in brain compression. The orbits are shallow with resulting eye proptosis, and the midface is deficient with malocclusion. He is shown before and after undergoing posterior cranial vault decompression and reshaping to expand the intracranial volume. He later underwent placement of a ventriculoperitoneal shunt for management of hydrocephalus. He will require a total midface advancement with further anterior cranial vault reshaping after 5 years of age. This will be followed by orthognathic surgery in combination with orthodontic treatment in the teenage years. (*A*) Profile view and CT three-dimensional reconstruction before surgery. (*B*) lateral view of posterior cranial vault with scalp flap elevated and after posterior cranial vault decompression, reshaping and fixation. (*C*) Profile view before and after posterior cranial vault reconstruction (*D*) Comparison of three-dimensional CT scan views before and after reconstruction. (*E*) Comparison of three-dimensional CT scan views of cranial bases before and after reconstruction. (*From* Posnick JC. The craniofacial dysostosis syndromes: secondary management of craniofacial disorders. Clin Plast Surg 1997;24:429; with permission.)

without completing a separate Le Fort I osteotomy. The degree of horizontal deficiency observed at the orbits and maxillary dentition is rarely uniform. This further segmentation of the midface complex at the Le Fort I level is required to establish normal proportions. If a Le Fort I separation of the total midface complex is not performed and the surgeon attempts to achieve a positive overbite and overjet at the incisor teeth, overadvancement of the orbits with enophthalmos occurs. The Le Fort I osteotomy is generally not performed at the time of the total midface procedure. This procedure awaits skeletal maturity and then is combined with orthodontic treatment. Until then an Angle class III malocclusion remains. Occasionally, a teenager or adult presents requiring both total midface and orthognathic management. The procedures may be performed simultaneously (Fig. 5).

A major aesthetic problem specific to the Le Fort III osteotomy when its indications are less than ideal is the creation of irregular step-offs in the lateral orbital rims. This situation occurs when even

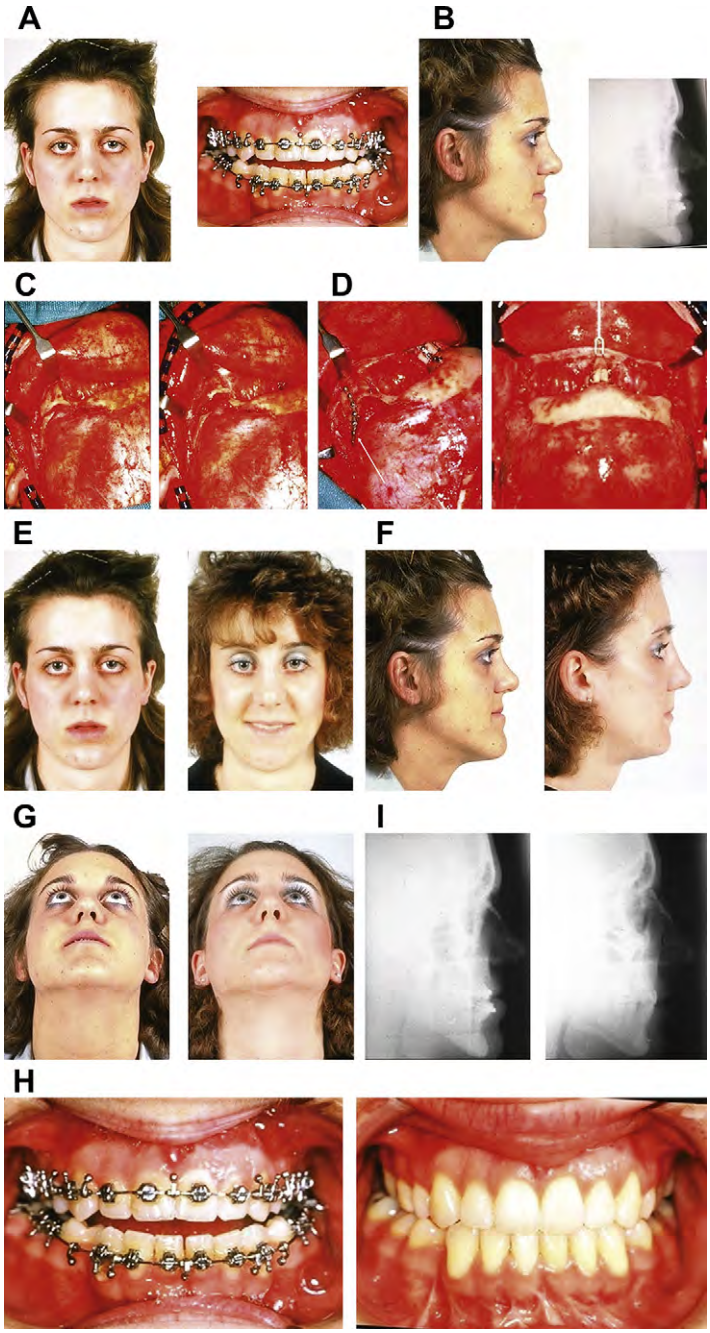

Fig. 3. A 15-year-old girl with Crouzon syndrome characterized by mild to moderate midface deficiency with retrusion of the infraorbital rims, zygomatic buttresses, and maxilla. The midface hypoplasia results in increased scleral show, nasal obstruction, and malocclusion. She is shown before and after undergoing a Le Fort III (extracranial) osteotomy with repositioning. Stabilization was with cranial bone grafts and plate and screw fixation. (*A*) Frontal and occlusal view before surgery. (*B*) Profile view and lateral cephalometric radiograph before surgery. (*C*) Intraoperative view of zygomatic complex just after osteotomies and after disimpaction. Note potential for unsightly step-off at lateral orbital rim. (*D*) Bird's-eye views of cranioorbital region after reshaping and stabilization; note cranial bone graft placed at frontonasal region. (*E*) Frontal views before and after extracranial Le Fort III osteotomy. (*F*) Profile views before and after extracranial Le Fort III osteotomy. (*G*) Worm's-eye views before and after extracranial Le Fort III osteotomy. (*H*) Occlusal view before and after reconstruction. (*I*) Comparison of preoperative and postoperative lateral cephalometric radiographs before and after reconstruction. (*From* Posnick JC. Craniosynostosis: surgical management of the midface deformity. In: Bell WH, editor. Modern practice in orthognathic and reconstructive surgery, vol. 3. Philadelphia: WB Saunders; 1992. p. 1888; with permission.)

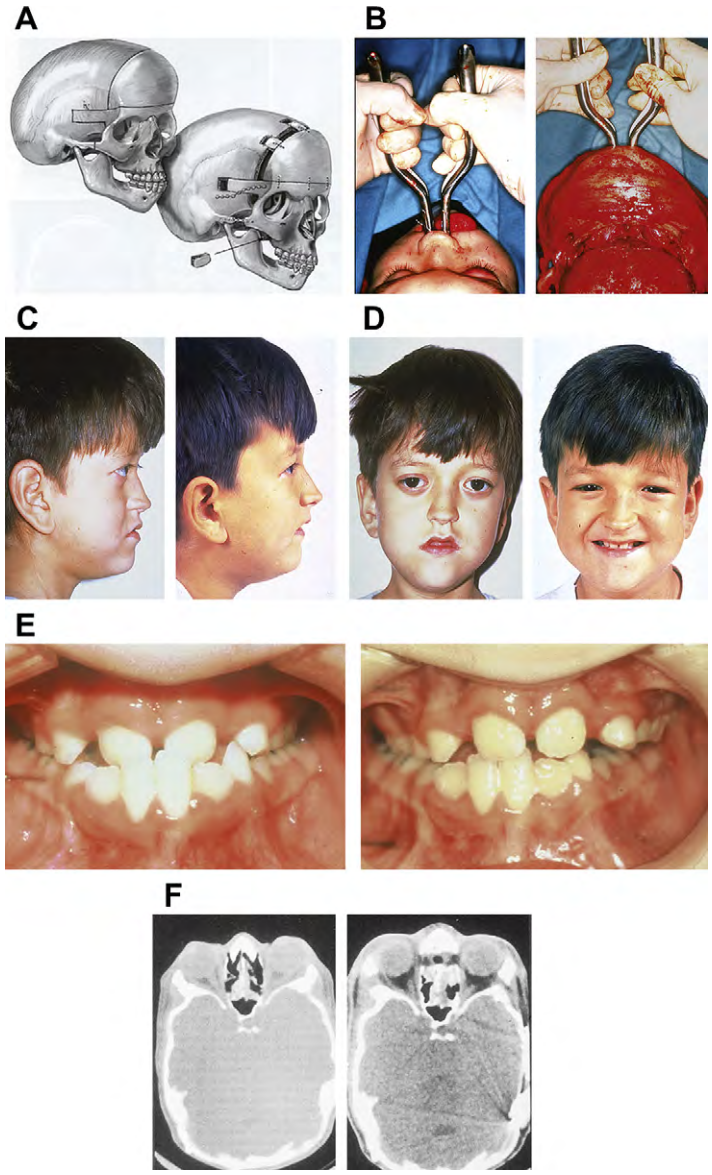

Fig. 4. An 8-year-old born with Crouzon syndrome who underwent a limited first-stage cranioorbital procedure at 6 weeks of age. He then underwent anterior cranial vault and monobloc (orbits and midface) osteotomies with advancement. (*A*) Craniofacial morphology before and after anterior cranial vault and monobloc osteotomies with advancement as performed. Osteotomy locations are indicated. Stabilization was with cranial bone grafts and miniplates and screws. (*B*) Intraoperative view of forceps in nose and mouth after monobloc osteotomy but before disimpaction, and same view with coronal incision turned down, indicating degree of advancement possible at supraorbital ridge level after disimpaction. (*C*) Profile view before and after monobloc advancement. (*D*) Frontal view before and after monobloc advancement. (*E*) Occlusal views before and after reconstruction. (*F*) Comparison of axial CT slices through midorbits before and after reconstruction, indicating resulting increased intraorbital depth and decreased proptosis achieved. (*From* Posnick JC. Craniosynostosis: surgical management of the midface deformity. In: Bell WH, editor. Modern practice in orthognathic and reconstructive surgery, vol. 3. Philadelphia: WB Saunders; 1992. p. 1889; with permission.)

a moderate (Le Fort III) advancement is performed. These lateral orbital step-offs are unattractive and are visible to the casual observer at conversational distance; surgical modification (revision) performed later is difficult, with less than ideal aesthetic results. Another problem with the Le Fort III osteotomy is the difficulty in judging an ideal orbital depth. A frequent result is either residual proptosis or enophthalmos. Simultaneous correction of orbital hypertelorism or correction of a midface arc-of-rotation problem is not possible with the Le Fort III procedure. Excessive lengthening of the nose, accompanied by flattening of the nasofrontal angle, also occurs if the Le Fort III osteotomy is selected when the skeletal morphology favors a monobloc or facial bipartition

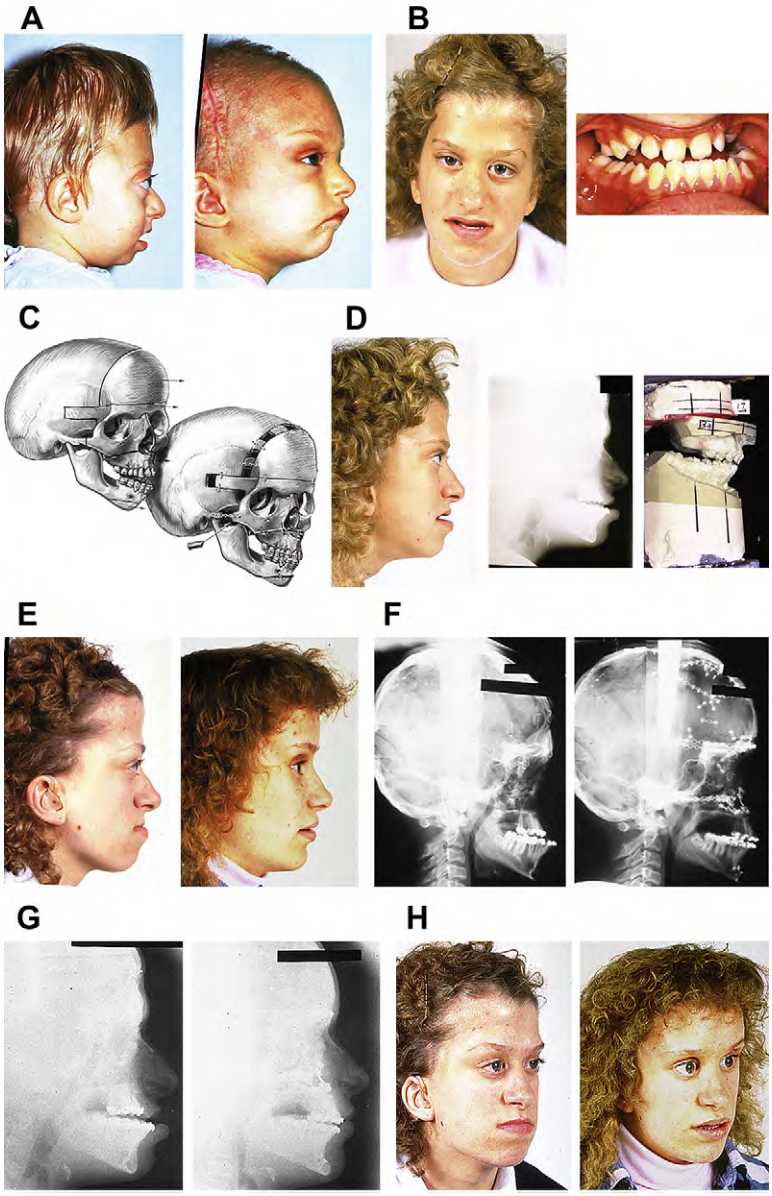

Fig. 5. A 14-year-old girl, born with Crouzon syndrome, underwent bilateral coronal suture release at 3 months of age. Additional craniotomy and cranial vault reshaping were completed when she was 9 months old. At 2 years of age, she underwent a Le Fort III (midface) osteotomy and forehead advancement procedure through an intracranial approach. She presented at the age of 14 years with residual deformity, for which she underwent simultaneous anterior cranial vault, monobloc, Le Fort I, and chin osteotomies with reshaping and three-dimensional repositioning. (*A*) Preoperative profile view at 1 year of age and profile view at 2 years of age following Le Fort III osteotomy. (*B*) Frontal and occlusal view at 14 years of age. (*C*) Planned anterior cranial vault, monobloc, Le Fort I, and chin osteotomies. (*D*) Preoperative profile view, lateral cephalometric radiograph and articulated dental casts after model reconstruction indicating that 17 mm advancement is required at occlusal level. The amount of advancement required at the supraorbital ridge level was only 12 mm. (*E*) Profile view before and after reconstruction. (*F*) Comparison of lateral cephalometric radiographs before and after reconstruction. (*G*) Comparison of soft-tissue lateral cephalometric radiographs before and after reconstruction. (*H*) Oblique view before and after reconstruction. (*I*) Occlusal views before and after reconstruction. (*J*) Comparison of axial-sliced CT scans through midorbits before and after reconstruction. (*K*) Comparison of axial CT slices through zygomatic arches before and after reconstruction. (*From* Posnick JC. Craniosynostosis: surgical management of the midface deformity. In: Bell WH, editor. Modern practice in orthognathic and reconstructive surgery, vol. 3. Philadelphia: WB Saunders; 1992. p. 1888; with permission.)

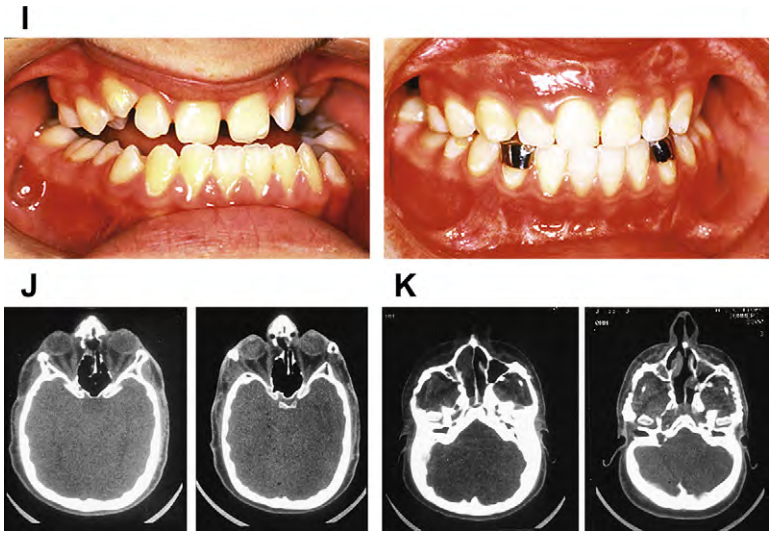

Fig. 5 (*continued*)

procedure. It is not possible to later correct the surgically created vertical elongation and flattened nasofrontal angle of the nose.

Final reconstruction of the cranial vault deformities and orbital dystopia in Crouzon syndrome can be managed as early as 6 to 9 years of age. By this age, the cranial vault and orbits normally attain approximately 85% to 90% of their adult size. Whenever feasible, waiting until the maxillary first molars are erupted and in occlusion is preferred. When the upper midface and final cranial vault procedure is performed after approximately 7 years of age, the reconstructive objectives are to approach adult dimensions in the cranioorbitozygomatic region, with the expectation of a stable result (no longer influenced by growth) once healing has occurred. No further horizontal growth of the midfacial skeleton should be anticipated after a midface advancement procedure. Psychosocial considerations also support the age 6 to 9 years time frame for the upper midface and final cranial vault procedure. When the procedure is performed at this age, the child may progress through school with a chance for satisfactory body image and self-esteem. Routine orthognathic surgery is necessary at the time of skeletal maturity to achieve an ideal occlusion and facial balance.

Orthognathic procedures for facial balance and definitive occlusal correction

Although the mandible has a normal basic growth potential in Crouzon syndrome, the maxilla does not. An Angle class III malocclusion, as a result of maxillary retrusion, is to be expected. A Le Fort I osteotomy (in segments) to allow for maxillary horizontal advancement, transverse widening, and vertical adjustment is generally required in combination with an osteoplastic genioplasty (vertical reduction and horizontal advancement) to further correct the lower face deformity. Secondary deformities of the mandible may also require correction through simultaneous sagittal ramus osteotomies. If correction of chronic nasal obstruction requires septoplasty and/or inferior turbinate reduction to improve the airway this is simultaneously completed. The elective orthognathic and intranasal surgery is performed in conjunction with orthodontic treatment planned for completion at the time of early skeletal maturity (approximately 13−15 years in girls and 14−16 years in boys).

Apert syndrome

Apert syndrome has previously been classified by its clinical findings. Postmortem histologic and radiographic studies have confirmed that skeletal deficiencies in the patient with Apert syndrome result from a cartilage dysplasia at the cranial base, leading to premature fusion of the midline sutures from the occiput to the anterior nasal septum. Mutations occur in the identical location as the *FGFR1*

mutation in Pfeiffer syndrome and the *FGFR3* mutation in Muenke syndrome. In addition, a component of the syndrome is 4-limb complex syndactylies of the hands and feet.

Staging of Reconstruction for Apert Syndrome

Primary brain decompression: cranioorbital reshaping in infancy

The initial craniofacial procedure for Apert syndrome generally requires bilateral coronal suture release and anterior cranial vault and upper three-quarter orbital osteotomies to decompress the brain and expand the anterior cranial vault and reshape the upper orbits and forehead. Our preference is to carry this out when the child is 9 to 12 months of age, unless signs of increased ICP are identified earlier in life. The main goals at this stage are to decompress the brain by providing increased space in the anterior cranial vault and to increase the orbital volume and decrease globe protrusion (Fig. 6). The frontoorbital surgical technique is similar to that described for Crouzon syndrome.

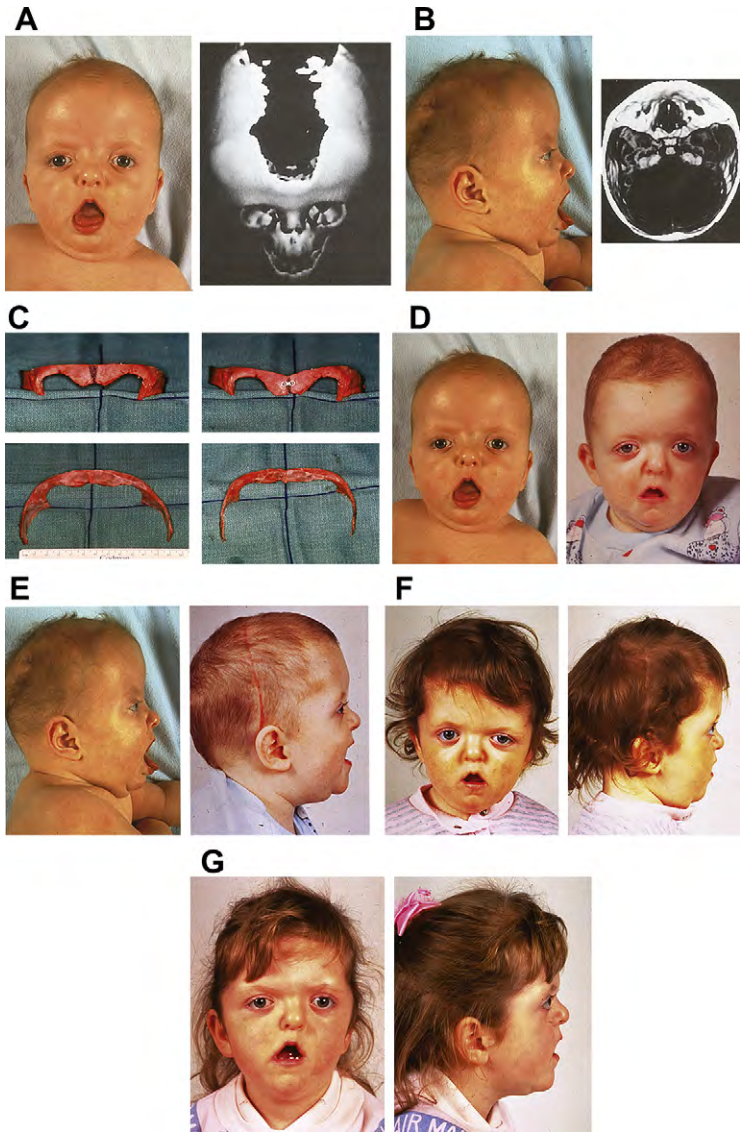

Fig. 6. A 6-month-old girl with Apert syndrome underwent anterior cranial vault and three-quarter orbital osteotomies with reshaping as described. (*A*) Frontal view and three-dimensional CT scan view of cranial vault before surgery. (*B*) Profile view and three-dimensional CT scan view of cranial base before surgery. (*C*) Frontal and bird's-eye views of orbital osteotomy unit before and after reshaping. (*D*) Frontal view before and 1 year after reconstruction. (*E*) Profile view before and 1 year after reconstruction. (*F*) Frontal and profile views 2 years after reconstruction. (*G*) Frontal and profile views 3 years after reconstruction. (*From* Posnick JC, Lin KY, Jhawar BJ, et al. Apert syndrome: quantitative assessment by CT scan of presenting deformity and surgical results after first-staged reconstruction. Plast Reconstr Surg 1994;93:489; with permission.)

Further craniotomy for additional brain decompression: cranial vault expansion and reshaping in young children

As described for Crouzon syndrome, the initial suture release, brain decompression, and cranioorbital reshaping are performed during infancy (9−12 months of age). The child is observed clinically at intervals by the craniofacial surgeon, pediatric neurosurgeon, pediatric ophthalmologist, and developmental pediatrician along with interval CT scanning. Should signs of increased ICP develop, further brain decompression with reshaping of the cranial vault to expand the intracranial volume is performed. In Apert syndrome the posterior cranial vault more commonly requires expansion at this stage. The technique is similar to what has been previously described (see Crouzon syndrome).

Management of the total midface deformity in childhood

In Apert syndrome, for almost all patients, facial bipartition osteotomies combined with further cranial vault reshaping permit a better correction of the abnormal craniofacial skeleton than can be achieved through other midface procedure options (ie, monobloc or Le Fort III). When using the facial bipartition approach a more correct arc of rotation of the midface complex is possible with the midline split (facial bipartition) (Fig. 7). This strategy further reduces the stigmata of the preoperative Apert "flat, wide, and retrusive" facial appearance. The facial bipartition also allows the orbits and zygomatic buttresses as units to shift to the midline (correction of hypertelorism), and the maxillary arch is simultaneously widened. A more correct horizontal positioning of the reassembled

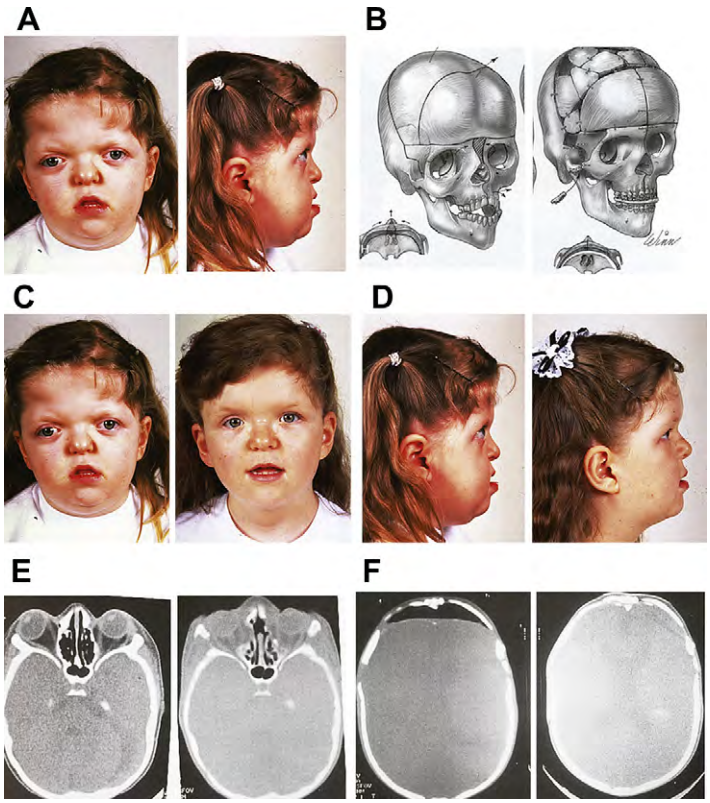

Fig. 7. A 5-year-old girl with Apert syndrome who underwent decompression and forehead/upper orbital reshaping at 6 months of age. She then presented with residual deformity requiring cranial vault and facial bipartition osteotomies with reshaping. She will require orthognathic surgery and orthodontic treatment later in the teenage years to complete her reconstruction. (*A*) Frontal and profile views before surgery. (*B*) Preoperative craniofacial morphology and planned and completed osteotomies and reshaping. Stabilization was achieved with cranial bone grafts and miniplate fixation. (*C*).Frontal view before and 2 years after reconstruction. (*D*) Profile view before and 2 years after reconstruction. (*E*) Comparison of axial-sliced CT scan views through midorbits before and after reconstruction, showing improvement in orbital hypertelorism and orbital depth and diminished eye proptosis. (*F*) Standard axial CT scan slices through cranial vault 1 week after facial bipartition (note dead space in the retrofrontal region), and at 1 year (notice that initial retrofrontal dead space has been resolved by brain expansion). (*From* Posnick JC. Craniofacial dysostosis: staging of reconstruction management of the midface deformity: craniofacial disorders. Neurosurg Clin N Am 1991;2:683; with permission.)

midface complex is then achieved, with corrected orbital depth and zygomatic length. The forehead is generally flat, tall, and retruded, with a constricting band just above the supraorbital ridge, giving the impression of bitemporal narrowing. Reshaping of the anterior cranial vault is simultaneously performed. A Le Fort III osteotomy is virtually never adequate for an ideal correction of the residual upper and midface deformity of Apert syndrome.

Orthognathic procedures for facial balance and definitive occlusal correction

The mandible has normal basic growth potential in Apert syndrome but may be secondarily deformed. The extent of maxillary hypoplasia/deformity results in an Angle class III malocclusion with severe anterior open bite deformity. A Le Fort I osteotomy (in segments) is required to allow for horizontal advancement, transverse widening, and vertical adjustment in combination with an osteoplastic genioplasty to vertically reduce and horizontally advance the chin, often combined with bilateral sagittal split osteotomies of the mandible. Septoplasty and inferior turbinate reduction are generally performed to improve nasal breathing. The orthognathic and intranasal surgery is performed in conjunction with detailed orthodontic treatment planned for completion at the time of early skeletal maturity (approximately 13−15 years of age).

Pfeiffer syndrome

In 1964, Pfeiffer described a syndrome consisting of craniosynostosis, broad thumbs, broad great toes, and occasionally partial soft-tissue syndactyly of the hands. This syndrome is known to have an autosomal-dominant inheritance pattern, with complete penetrance documented in all recorded 2- and 3-generation pedigrees. Variable expressivity of the craniofacial and extremity findings is common.

Carpenter syndrome

Carpenter syndrome is characterized by craniosynostosis often associated with preaxial polysyndactyly of the feet, short fingers with clinodactyly, and variable soft-tissue syndactyly, sometimes postaxial polydactyly, and other anomalies such as congenital heart defects, short stature, obesity, and mental deficiency. It was first described by Carpenter in 1901 and was later recognized to be an autosomal-recessive syndrome. In general, the reconstructive algorithm described for Crouzon syndrome can be followed.

Saethre-Chotzen syndrome

Saethre-Chotzen syndrome has an autosomal-dominant inheritance pattern with a high degree of penetrance and expressivity. Its pattern of malformations may include craniosynostosis, low-set frontal hairline, ptosis of the upper eyelids, facial asymmetry, brachydactyly, partial cutaneous syndactyly, and other skeletal anomalies. As part of the reconstruction, cranioorbital reshaping is almost certainly required and is similar to that described for Crouzon syndrome. Evaluation and management of the total midface deficiency and orthognathic deformities as described for Crouzon syndrome should be followed.

Cloverleaf skull anomaly

Kleeblattschadel anomaly (cloverleaf skull) is a trilobular-shaped skull, which is usually secondary to synostosis of the coronal, lambdoidal, metopic, and sagittal sutures. With pan synostosis the brain protrudes through the open anterior and parietal fontanelles, giving the characteristic cloverleaf skull appearance (Fig. 8). The cloverleaf skull anomaly is known to be both etiologically and pathogenetically heterogeneous. This anomaly is also nonspecific: it may occur as an isolated anomaly or together with other malformations, making up various syndromes (ie, Apert, Crouzon, Carpenter, Pfeiffer, and Saethre-Chotzen). The extent and timing of anterior cranial vault/upper

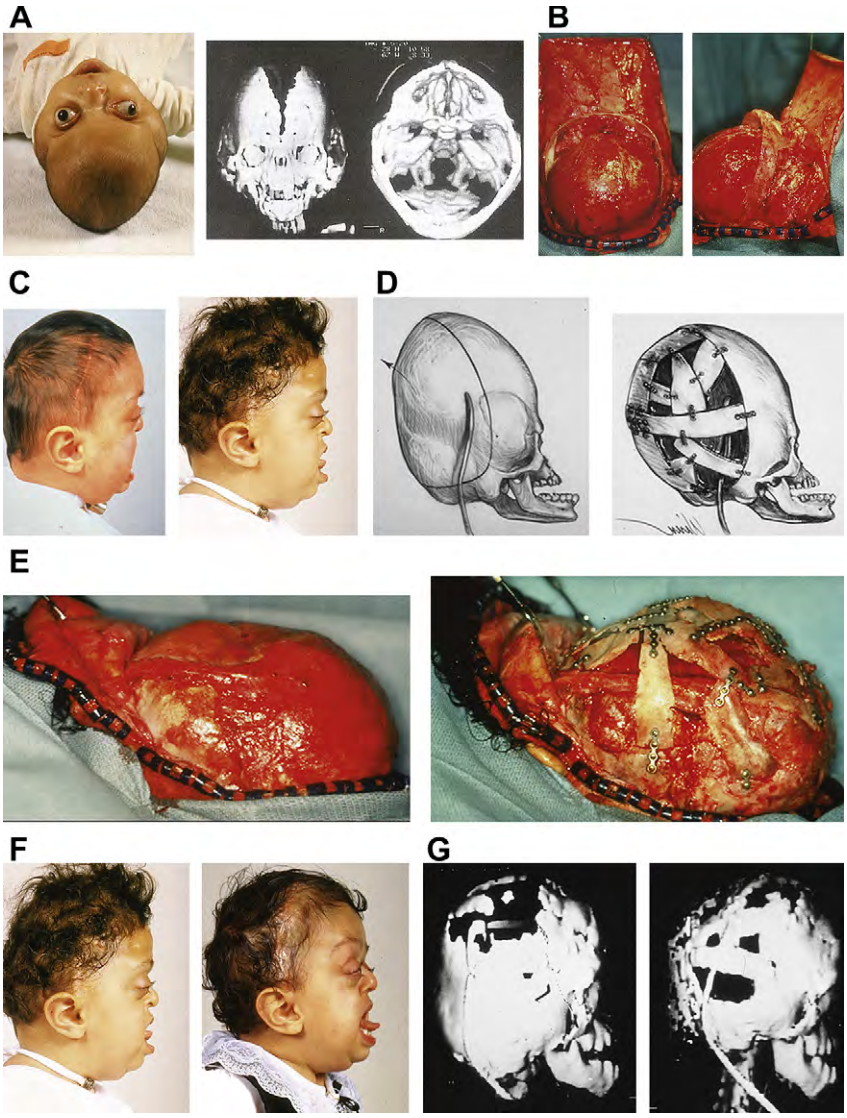

Fig. 8. A child with a severe form of cloverleaf skull anomaly. At 10 months of age she underwent first-stage cranial vault and upper orbital decompression with reshaping. She then required a ventriculoperitoneal shunt for management of hydrocephalus. At 3.5 years of age, posterior cranial vault decompression with reshaping to increase the intracranial volume was performed. At 4.5 years of age, facial bipartition osteotomies combined with anterior cranial vault reshaping was performed. After the cranial vault and facial bipartition procedure, nasal airflow improved, and it was possible to remove the tracheostomy tube. The cranial vault reshaping expanded the intracranial volume, providing more space for the brain. The midface advancement improved the proptosis and the ability to chew and articulate speech. As part of the staged reconstruction, she will require orthognathic surgery combined with orthodontic treatment at the time of early skeletal maturity. (*A*) Frontal view and craniofacial CT scan views at 10 months of age. (*B*) Intraoperative lateral and bird's-eye views at 10 months of age after craniotomy and removal of the upper orbits, followed by the construction of a bandeau with 3 cm advancement. (*C*) Profile view at 2.5 and 3.5 years of age with flattened posterior cranial vault and severe midface deficiency (*D*) Flattened posterior cranial vault. Note ventriculoperitoneal shunt in place and proposed cranial vault reshaping. (*E*) Intraoperative side view (patient in prone position) before and after craniotomy and reshaping and fixation of bone segments with miniplates and screws. Ventriculoperitoneal shunt remains intact and deep to the skull reconstruction. (*F*) Profile view at 3.5 years and 2 weeks after posterior cranial vault reconstruction. (*G*) Comparison of three-dimensional CT scan views before and just after posterior cranial vault reshaping. (*H*) Planned facial bipartition and cranial vault reconstruction. (*I*) Oblique views before and 6 months after facial bipartition osteotomies with reshaping and advancement. (*J*) Frontal views before and after facial bipartition reconstruction. (*K*) Profile view before and after facial bipartition reconstruction. (*L*) Comparison of axial CT slices through midorbits before and after reconstruction. (*M*) Comparison of axial CT slices through zygomatic arches before and after reconstruction. (*From* Posnick JC. The craniofacial dysostosis syndromes: secondary management of craniofacial disorders. Clin Plast Surg 1997;24:429; with permission.)

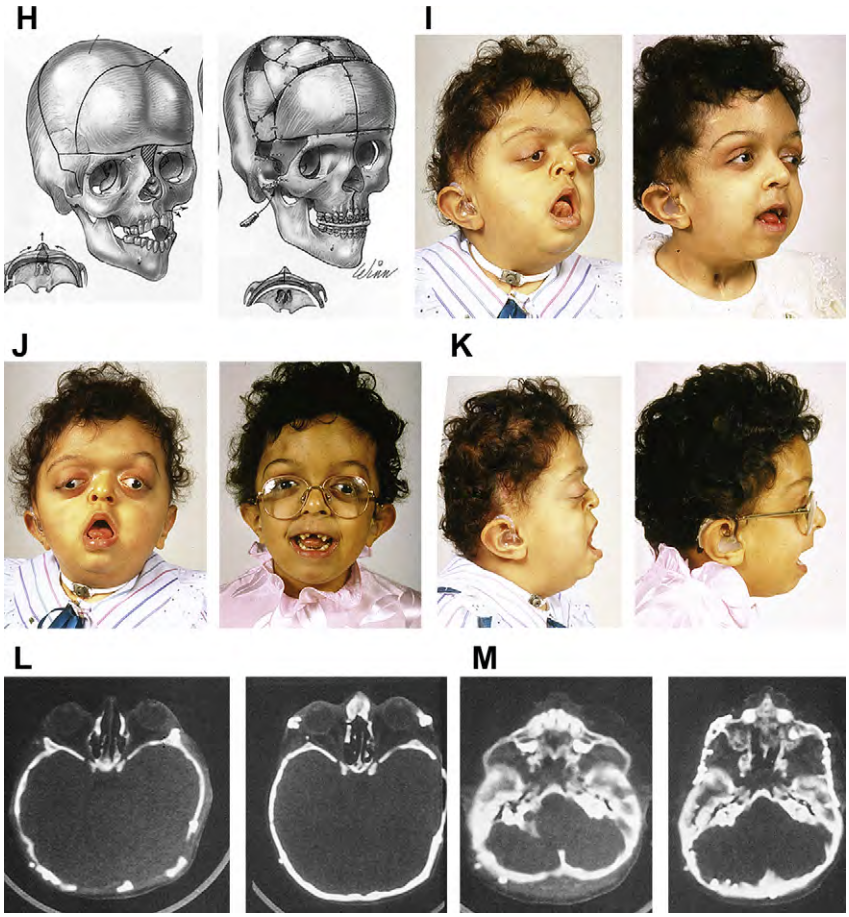

Fig. 8 (*continued*)

orbital, posterior cranial vault, midface, and orthognathic reconstruction are dependent on patient-specific malformations, functional needs, and any secondary surgical deformities. In general, the protocol described for Crouzon syndrome can be followed.

Summary

The preferred approach to the management of a patient with craniofacial dysostosis is to stage reconstruction to coincide with craniofacial growth patterns, visceral function (cognitive, vision, breathing, swallowing, speech, chewing, hearing), and psychosocial effects. Recognition of the need for a staged reconstruction approach serves to clarify the objectives of each phase of treatment of the surgeons, craniofacial team, and family. By continuing to define our rationale for the timing, techniques, and extent of intervention and objectively evaluating both head and neck function and morphologic (aesthetic) outcomes, the quality of life for children born with a craniofacial dysostosis syndrome is further improved.

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Reconstruction of Frontoethmoidal Encephalocele Defects

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… nasofrontal lesions are uncommon, frequently diagnosed incorrectly, and treated in a haphazard fashion.

—(Davis and Alexander, 1959)

Encephaloceles, historically termed cranium bifidum, were first documented in the medical literature in the sixteenth century. They are a result of a congenital defect in the skull base that allows the brain to herniate through the cranial floor. Encephaloceles are most commonly located in the posterior skull base (75%); however, anterior (sincipital) encephaloceles are considerably more deforming. The herniated cerebral contents may contain the meninges alone (meningocele), meninges and brain (meningoencephalocele), or in severe cases meninges, brain, and part of the ventricular system (hydroencephalomeningocele or meningoencephalocystocele).

There are many types of encephaloceles.

Classification of encephaloceles

- Frontoethmoid encephaloceles
  - Nasofontal
  - Nasoethmoidal
  - Nasoorbital
- Cranial vault encephaloceles
  - Interfrontal
  - Anterior fontanel
  - Interparietal
  - Posterior fontanel
  - Temporal
- Skull base encephaloceles
  - Transethmoidal
  - Sphenoethmoidal
  - Transsphenoidal
  - Frontosphenoidal
- Occipital encephaloceles
- Cranioschisis
  - Those associated with cranial/upper facial clefts
  - Those associated with basal/lower facial cleft
  - Occipitocervical clefts
  - Acrania and/or anencephaly.

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Suwanwela and Suwanwela developed a classification system for anterior encephaloceles based on their location of skull base herniation. Anterior encephaloceles are divided into 3 groups: frontoethmoidal, interfrontal, and those associated with craniofacial clefts. Frontoethmoidal encephaloceles are further divided into 3 groups: nasofrontal, nasoethmoidal, and nasoorbital. The nasofrontal type present with a herniation between the nasal and frontal bones. In the nasoethmoid type, the defect lies between the nasal bone and the nasal cartilages and the encephalocele herniates between the frontal and ethmoid bones. In the nasoorbital type, the neural contents herniate through the medial wall of the orbit between the lacrimal bone and the frontal process of the maxilla.

Suwanwela classification of anterior encephaloceles

- Frontoethmoidal
  - Nasofontal
  - Nasoethmoidal
  - Nasoorbital
- Interfrontal
- Encephaloceles associated with craniofacial clefts.

Presentation

Frontoethmoidal encephalocele have several hallmark findings on physical examination. A nasal mass located in the midline is the most common finding. It is often bluish in appearance, soft, compressible and often pulsatile as a result of its intracranial connection. They transilluminate and increase in size with valsalva, crying, or with compression of the internal jugular veins (Furstenberg test). Ortiz-Monasterio and Jackson independently popularized the terms long nose hypertelorism and long nose deformity to describe the nasal anatomy associated with frontoethmoidal encephaloceles. Nearly all patients present with a long, flat, wide nose that is more pronounced after the encephalocele is excised. In addition, there is a very specific orbital presentation in these patients. Telecanthus and interorbital hypertelorism are universal phenotypical findings associated with nasal encephaloceles but true orbital hypertelorism is rare. Some degree of deformational trigonocephaly is usually present. Other physical findings include medial eyebrow elevation, lacrimal obstruction, microencephaly, hydrocephalus, and anosmia.

Pathogenesis

To date, there is no known genetic mutation associated with the inheritance of the encephalocele trait. However, the incidence of frontoethmoidal encephaloceles is significantly higher in southeast Asia (1 in 6000 live births) compared with its incidence in Europe and North America (1 in 35,000 live births). One postulated theory for encephalocele formation is the primary failure of bone formation at the skull base allowing the brain to herniate. Alternatively, encephaloceles may result from adhesions between the brain, dura, and skin, which arrest bony development at the skull base. The most recent explanation for encephalocele development is increased intracranial pressure pushing the brain through the skull base and arresting bony development.

Embryologically, the facial primordial develops during the first 12 weeks of gestation. Between the third and the fourth week of fetal development, neural tube closure begins midembryo and propagates both cranially and caudally. As the neural tube closes cranially, neural crest cells migrate laterally and anteriorly into the frontonasal and maxillary processes. These cells are responsible for the mesenchymal structures such as the bones, cartilage, and muscles of the face. Several centers of mesenchymal structures fuse and ossify to form the skull base as well as the nasal and frontal bones. Potential spaces exist between these centers before their fusion and these spaces are important in the development of congenital midline nasal masses. These spaces consist of the fonticulus frontalis (space between the nasal and frontal bones), the prenasal space (between the nasal bones and the nasal capsule and cartilages/septum), and the foramen cecum (anterior neuropore at the anterior cranial vault floor). Abnormal development of these structures is believed to be responsible for the development of different midline nasal masses (Fig. 1).

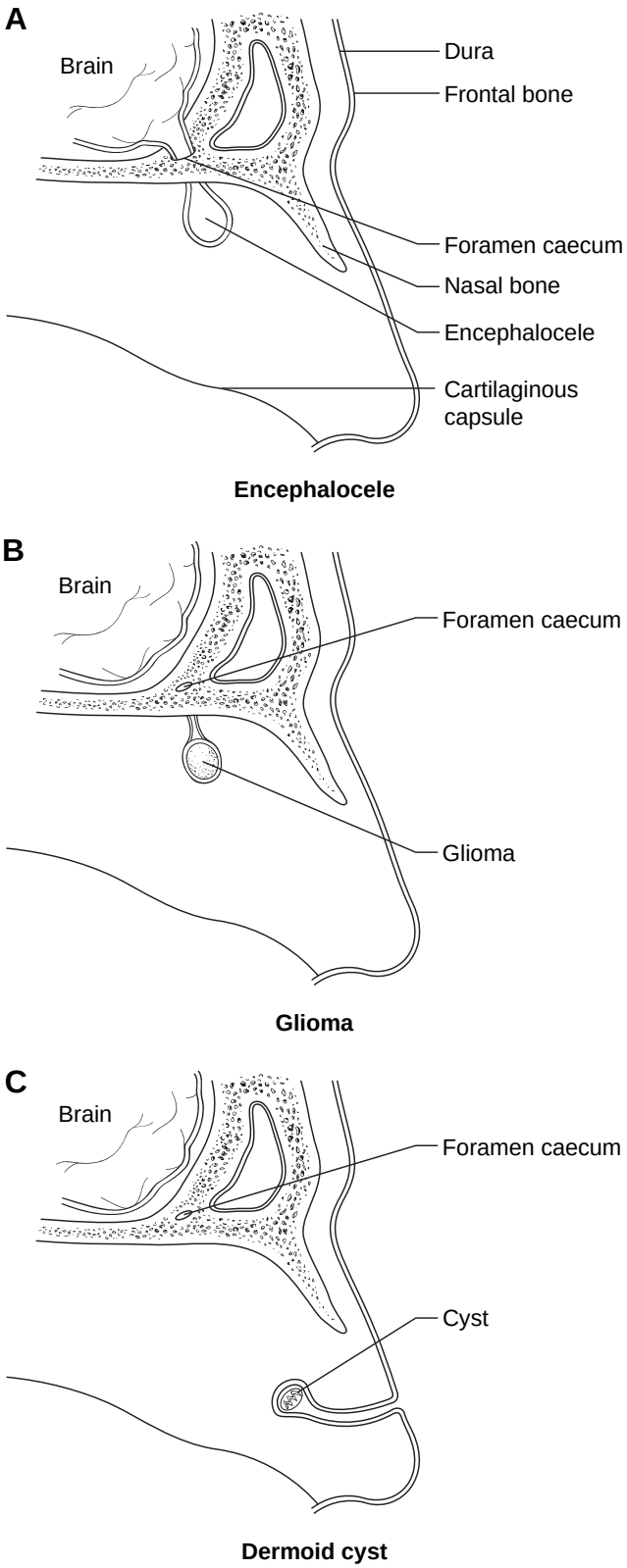

Fig. 1. Midline nasal mass pathophysiology. (*A*) Encephalocele: neural contents herniate through the foramen cecum. (*B*) Glioma: trapped neural contents with an intact skull base. (*C*) Dermoid: invagination of skin elements toward the skull base.

Differential diagnosis

Knowledge of embryology and craniofacial surgery is necessary to develop a comprehensive differential diagnosis and treat a child with a midline nasal mass. An understanding of how midline entities present is important in differentiating other midline nasal entities from encephaloceles. Nearly all are present at birth and are slow-growing masses at the nasal root. However, there are many differences among congenital midline masses that allow the correct diagnosis to be made before radiographic confirmation.

Nasal dermoid cysts are the most common midline nasal mass. They consist of ectodermal and mesodermal elements. Dermoids usually present at birth but are often not diagnosed until later in childhood. The hallmark of a nasal dermoid is a pit or punctum with a single hair located on the nasal dorsum. They can often become infected and drain sebaceous material. Intracranial connection is rare but cannot be ruled out on clinical examination. Therefore imaging is required to evaluate for intracranial extension.

A nasal glioma presents as a firm rubbery mass with a bluish or reddish appearance. They are composed of neurogenic material consisting of glial cells in a connective tissue matrix. The overlying skin usually contains capillary telengectasias and the mass often extends intranasally through the nasal bones. There is no cerebral spinal fluid (CSF) surrounding these masses as they do not connect with the subarachnoid space. Therefore, they are not pulsatile and do not transilluminate. Intracranial extension is uncommon but may occur through the cribiform plate or at the frontonasal suture.

Less common entities that may occur at the nasal midline include hemangioma, vascular malformation, teratoma, sebaceous cyst, neurofibromata, ganglioneuromata, nasal fibroma, adenoma, carcinoma, and chondroma. A thorough history and physical examination is required for the correct diagnosis. Imaging is used to confirm the diagnosis and to rule out intracranial extension. Biopsies before complete work-up are discouraged.

Surgical management

There are several principles in frontoethmoid encephalocele reconstruction. First, when present, any open skin defects must be debrided and closed immediately to prevent infection and/or desiccation of exposed brain tissue. Second, all nonfunctioning extracranial brain tissue herniating through the skull base must be debrided and/or reduced intracranially. Third, the dura should be repaired to provide a watertight closure around the viable cerebral contents. Fourth, the skull base defect is repaired to prevent future herniation and encephalocele recurrence. Fifth, craniofacial, nasal, and medial canthal reconstruction is performed to restore the appropriate premorbid anatomy.

Preoperative Assessment

A multidisciplinary craniofacial team is necessary for the appropriate management of these patients. Members of this team typically include a craniofacial surgeon, neurosurgeon, otolarynologist, geneticist, and general pediatrician. Often the diagnosis is clearly evident on physical examination and biopsy should be avoided. A fundoscopic examination should be performed to rule out papilledema caused by increased intracranial pressure as a result of hydrocephalus.

Computed tomography (CT) is the most useful imaging modality for assessing this abnormality. CT images should include both brain and bone windows in the axial, coronal, and sagittal planes as well as three-dimensional reconstructions of the cranium and skull base defect. Magnetic resonance imaging is also useful to evaluate nasal masses as it provides more detailed soft tissue information. Ultrasound may be used to assess ventricular size to rule out hydrocephalus, but is often redundant if a CT scan can be performed as part of the initial evaluation.

Surgical Positioning

The operation is performed in the supine position. A Mayfield headrest is used to secure the cranium. The pins of the headrest are positioned posteriorly so that they do not interfere with the

coronal flap or parietal bone graft harvest. The endotracheal tube is secured using a circummandibular 28-gauge dental wire. The entire head and face are prepared into the surgical field. A 6-0 silk tarsorraphy suture is used for eyelid closure and corneal protection.

Coronal Exposure

The transcranial portion of the case is performed via a coronal incision (Fig. 2). A wavy-line pattern is used to break up the scar, making it less noticeable when the hair is wet or short. Before incision the scalp is infiltrated with 0.5% to 1% lidocaine with 1:200,000 epinephrine. A #15 blade scalpel is used to make the skin incision. Dissection using a Colorado needle tip cautery is done deep to the dermis. This protects the hair follicles and alleviates the need for hemostasis clips on the coronal flap skin edges. The dissection proceeds through the pericranium to the cranial bones and the coronal flap is elevated anteriorly. A pericranial flap based on the supraorbital vessels should be preserved during the coronal flap elevation. This flap may be necessary to assist in dural closure after encephalocele resection. If the pericranial flap is necessary for dural closure, we prefer to raise it secondarily from the underside of the coronal skin flap.

Care is taken as the coronal flap is elevated over the anterior fontanel which may remain patent beyond the first year of life. Blunt dissection between the dura and pericranium is preferred and midline blood vessels are cauterized. These vessels often communicate with the underlying sagittal sinus and injury to these vessels can cause subdural bleeding. Laterally, we prefer to elevate the coronal flap deep to the temporalis muscle with the muscle elevated on the coronal flap in its anatomic position.

As the coronal flap elevation approaches the supraorbital rim, the supraorbital neurovascular bundle is identified and preserved and may require an osteotome to remove the neurovascular bundle from its foramen. At the glabella, the encephalocele sac is encountered and exposed to reveal its site of herniation. Care is taken to bluntly dissect the dura and cranial contents from the pericranium of the frontal bone and the periosteum over the nasal and orbital bones. The dura is malformed and fragile overlying the encephalocele sac. The periorbita is elevated circumferentially within the orbit and the lacrimal drainage system is preserved. The medial canthi are detached and must be repositioned and permanently secured later in the case.

Nasal Exposure

A skin incision at the encephalocele sac is necessary for its complete exposure and removal. The skin overlying the encephalocele is often thin and of different consistency from the surrounding tissues. It is of little use once the encephalocele is removed therefore it is left attached to the sac and removed when the sac is excised. A circumferential skin incision is made around the encephalocele stalk leaving enough tissue for skin closure once the sac is excised (Fig. 3). The sac is dissected from the skin toward the root of the nose and the origin of herniation. If an intranasal encephalocele component is present, a transcolumellar rhinoplasty approach is used for additional exposure.

Fig. 2. Surgical positioning. A wavy-line coronal incision is designed and the Mayfield headrest is positioned posteriorly.

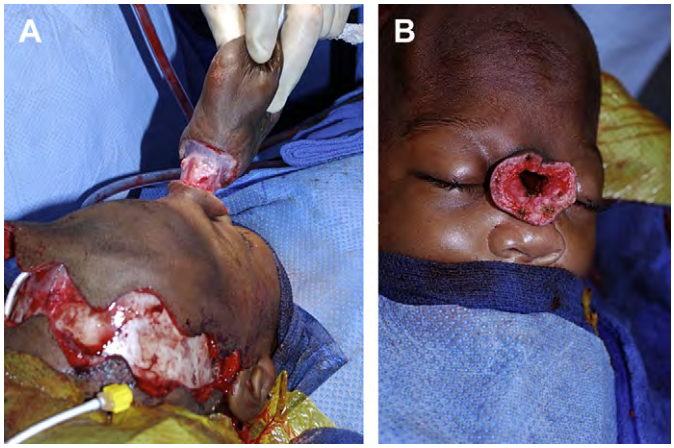

Fig. 3. Nasal exposure. (*A*) An incision is made circumferentially around the encephalocele stalk and the sac is dissected from the skin. (*B*) The encephalocele has been removed resulting in glabellar skin excess.

Frontal Craniotomy

Once exposure of the frontal bones, supraorbital rims, and nasal bones has been performed a frontal craniotomy is planned. A Marchac template is used to determine the appropriate position of the frontal craniotomy (Fig 4). A 1- to 2-cm frontoorbital bandeau is preserved above the supraorbital rims. The frontal bone is removed and placed on the back table. The dura is elevated from the anterior skull base and the bandeau is freed of its surrounding soft tissues. The bandeau is removed before the encephalocele resection. Once it is removed, the encephalocele sac is transected and the nonfunctioning neural tissue discarded. This creates a dural opening that must be repaired to prevent a cerebrospinal fluid leak. The dura is primarily repaired using Neurilon sutures. When the herniation occurs through the cribiform plate it is often difficult or impossible to achieve complete primary dural closure. In these situations, dural substitutes such as DuraGen or cadaveric dermis can be used. In addition, a pericranial flap can be elevated from the coronal skin flap and draped over the defect. This provides an additional layer of vascularized coverage at the site of tenuous dural repair. The bony skull base defect is reconstructed by the rotation and replacement of the bandeau. Occasionally, a small skull base defect persists requiring a calvarial bone graft onlay to prevent further herniation.

Cranial Remodeling

Once the encephalocele sac has been removed and the dura repaired, the cranial deformity must be corrected. This includes some degree of trigonocephaly and interorbital hypertelorism in every

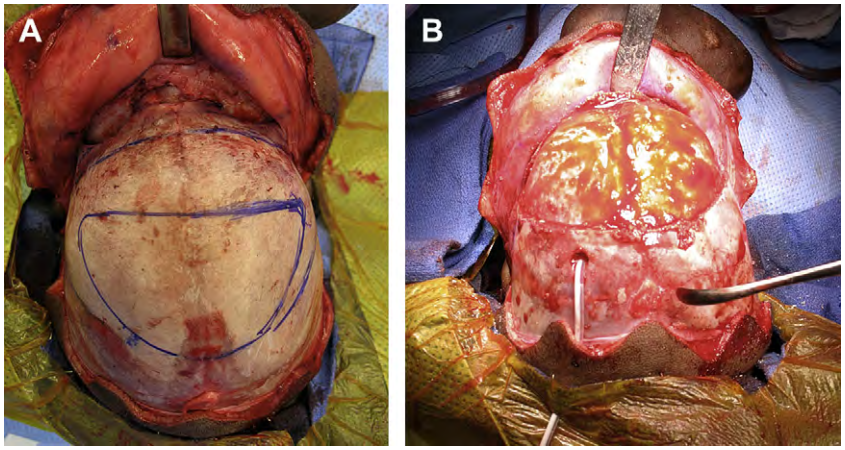

Fig. 4. Frontal craniotomy. (*A*) The Marchac template is used to find the correction location of the neofrontal bone craniotomy. A segment of bone exists between the frontal bone craniotomy and the bandeau that must be excised. (*B*) The frontal craniotomy has been performed and the bandeau removed.

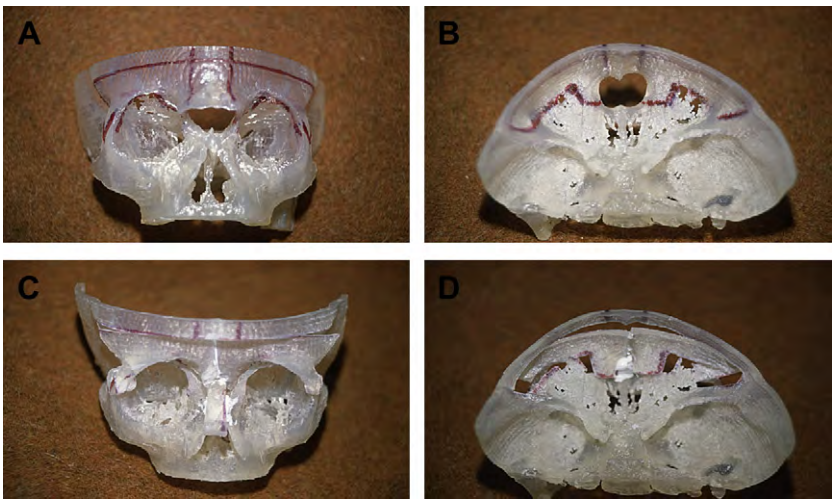

Fig. 5. Acrylic model surgical example. (*A*, *B*) Skull base defect caused by a midline anterior encephalocele. (*C*, *D*) The central segment of the bandeau is excised and the hemibandeaus are rotated medially, closing the cranial floor defect and reconstructing the orbital rims. (*Modified from* Holmes AD, Meara JG, Kolker AR, et al. Frontoethmoidal encephaloceles: reconstruction and refinements. J Craniofac Surg 2001;12(1):6−18; with permission.)

patient. When possible, a portion of the existing frontal bone is used for reconstruction. However, if severe trigonocephaly exists, the frontal bone may require replacement with a parietal bone graft to achieve an adequate reconstruction. In this situation, a parietal bone graft is harvested using the Marchac template and the donor site is reconstructed using the previously harvested frontal bone.

Cranial remodeling occurs on the back table as the neurosurgical repair is underway. The central segment of the bandeau is excised at the site of the encephalocele herniation, creating 2 hemibandeau segments. These hemibandeau segments are rotated and advanced medially to reposition the supraorbital rims appropriately and to close the skull base defect (Fig 5). The hemibandeau segments are secured to one another as well as to the neofrontal bone with suture (Fig. 6). We prefer 3-0 PDS suture fixation using drill holes placed in the cranial segments. The bandeau/neofrontal construct is placed in situ and fixated with resorbable plates after the nasal bone graft onlay is attached (Fig. 7). Any remaining calvarial defects are reconstructed with a combination of bone graft and bone mush harvested from the inner table of the neofrontal bone.

Nasal Reconstruction

Correction of the long nose deformity is an important component of the primary frontoethmoid encephalocele repair. Resection of the encephalocele makes this deformity more noticeable and its appearance is the stigmata of a poorly performed operation. A dorsal onlay graft is necessary to correct this abnormality and is constructed from calvarial bone or costochondral cartilage. It is contoured to provide the appropriate nasal dorsum shape and support. The graft is rigidly fixated as

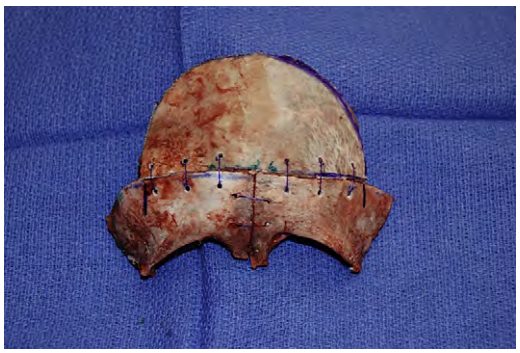

Fig. 6. Cranial remodeling. The neofrontal bone created by the Marchac template is suture fixated to the hemibandeaus. The orbital rims have been rotated medially and the central portion of the bandeau excised.

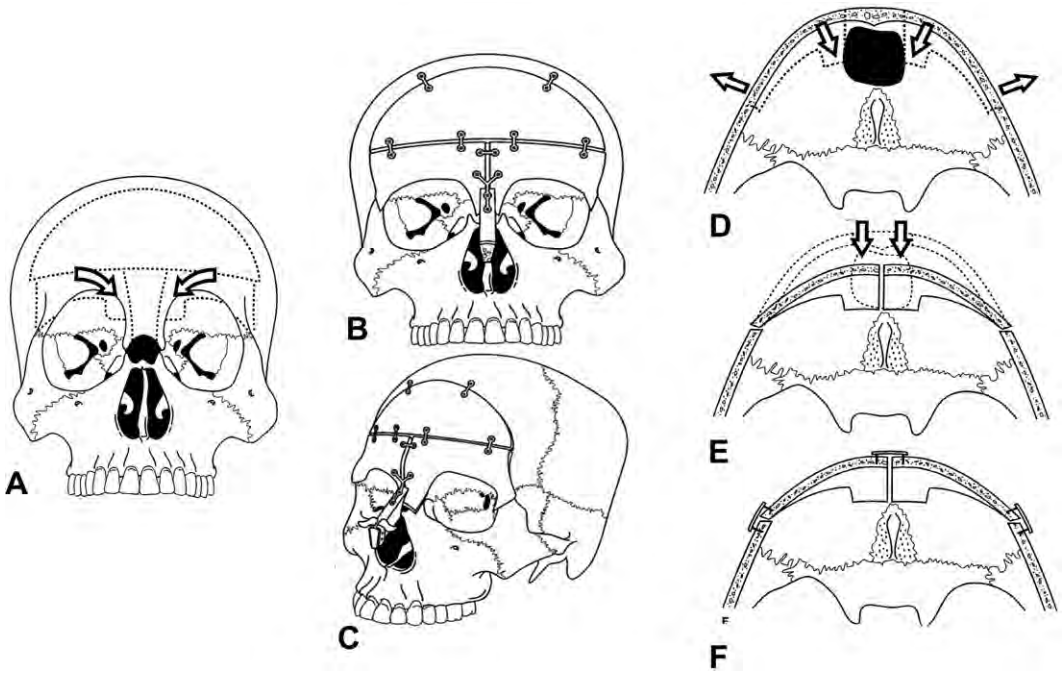

Fig. 7. Cranial remodeling. (*A*−*C*) The central segment of the bandeau is excised and the hemibandeaus are rotated medially. (*D*−*F*) The hemibandeaus are used to close the cranial base defect when rotated medially. (*Modified from* Holmes AD, Meara JG, Kolker AR, et al. Frontoethmoidal encephaloceles: reconstruction and refinements. J Craniofac Surg 2001;12(1):6−18; with permission.)

a cantilever using lag screws at the frontonasal junction of the bandeau/neofrontal construct (Fig. 8). We prefer calvarial bone grafts as they are easily harvested at the time of craniotomy. Critics of calvarial grafts state they are less malleable and their resorption is unpredictable and therefore advocate for the use of costochondral cartilage grafts. However, costochondral cartilage grafts require an additional operative site for their harvest and is unnecessary in our experience. Once the bandeau/neofrontal segment is placed in situ and secured, the lower lateral cartilages are often malpositioned. They often need to be elevated and suspended from the dorsal onlay graft to maintain their correct anatomic position.

Medial Canthal Repositioning

The medial canthi are inferiorly and laterally displaced by the encephalocele. Once the skeletal and nasal reconstruction has been performed, the medial canthal ligaments are identified. A medial canthoplasty is performed by transnasal wiring. Wiring is done by looping a 30-gauge wire through the canthi and joining the wires transnasally across the midline. Slight overcorrection is necessary as the canthal height will drift caudally and laterally as the swelling subsides and the surrounding tissues

Fig. 8. Nasal reconstruction. A cantilever calvarial bone graft is fixated as a dorsal onlay graft.

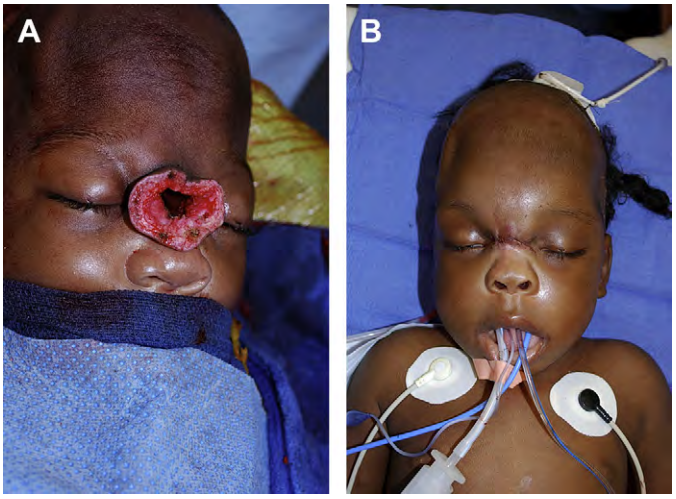

Fig. 9. Skin closure. (*A*) Skin excess exists in both the vertical and horizontal plane after the encephalocele is excised. (*B*) H-shaped closure of the defect.

relax. A nasal splint is placed at the end of the procedure to secure the repair as well as the position of the dorsal onlay graft.

Skin Closure

An external ventriculostomy is placed before skin closure. A closed suction drain is placed under the anterior skin flap but not at the site of dural repair. The coronal incision is closed in 2 layers to provide a watertight closure. Excess skin at the nasal dorsum is redraped and marked for excision. Correction of the medial eyebrow malposition is an important consideration at the time of nasal skin closure. An H-shaped excision is used to remove excess skin and reposition the medial brows with the horizontal excision located at the radix (Fig. 9). The vertical excision is performed along the nasal sidewall and extends superiorly to the upper medial brow. This allows the medial brow to be rotated inferomedially and positioned correctly at the supraorbital rim at the time of nasal closure. Z-plasties or W-plasties are aesthetically unacceptable at the radix and should be avoided when possible.

Postoperative Monitoring

A gentle compression dressing is placed around the head in the operating room before extubation. If physiologically stable, the child is extubated in the operating room and brought to an intensive care

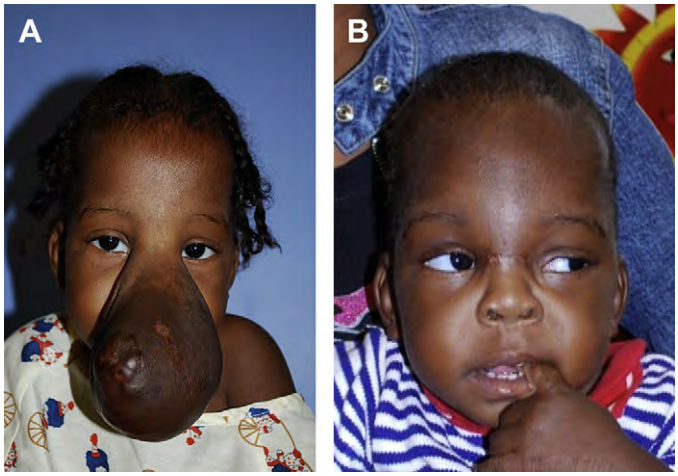

Fig. 10. Results of encephalocele reconstruction. (*A*) Preoperative photo; (*B*) postoperative results.

unit for observation overnight. Neurologic assessments are routinely performed postoperatively. The closed suction drain is monitored for potential CSF leak and is usually removed between day 3 and 5 postoperatively. The external ventriculostomy is left in place for 5 days and if necessary converted to an internal shunt should postoperative hydrocephalus arise. Periorbital swelling is universally present in the first 72 hours postoperatively. A 3-day steroid taper will help limit this swelling and eye ointment is used to prevent corneal abrasions. The child is discharged home between 3 and 5 days postoperatively and discharge is often dependent on the resolution of the periorbital edema.

Summary

Nasal encephaloceles are a rare and difficult problem requiring combined craniofacial and neurosurgical management. In experienced hands, a single-staged operation yields excellent functional and aesthetic outcomes (Fig. 10). Proper diagnosis and imaging are necessary for preoperative planning and a single-staged operation is performed. Intraoperative goals include resection of the encephalocele sac, dural closure, and repair of the skull base defect. Cranial remodeling is performed to correct the trigonocephaly and interorbital hypertelorism. Nasal reconstruction using a dorsal onlay graft is necessary to correct the long nose deformity and the medial canthi are repositioned. Skin closure is performed excising the excess skin at the nasal dorsum in an H-shaped pattern. Postoperative monitoring is done in an intensive care unit for the first 24 hours and the child remains hospitalized until the periorbital swelling has diminished and oral intake has resumed.

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Reconstruction of Skull Defects

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Cranial defects occur among all ages from a wide variety of causes. Trauma, infection, congenital malformations, pathology, and tumors and their surgical management can all lead to skull abnormalities and defects. Small defects that are covered in formidable soft tissue may not need repair. Other cranial defects can be immediately reconstructed when they are small or iatrogenically created for surgical access; most require secondary reconstruction.

Etiology

Cranial decompression has gained popularity in treating elevated intracranial pressure in traumatic brain injury. The mortality rate for this group of patients is high despite aggressive surgical and medical intervention. A full-thickness bone flap is removed and commonly stored in a freezer or subcutaneous abdominal pocket. The brain is often swollen and herniating out of the confines of the skull, and dural expansion is performed with autologous tissue or dural substitutes. After acute intracranial and neurosurgical issues are resolved, the autologous bone flap is normally replaced.

Complex or depressed skull fractures may require bone removal. If present, dural tears must be closed and elevated intracranial pressures treated. Current recommendations favor replacing the bone fragments at initial surgery. If the wound is contaminated, copious antibiotic irrigation should be used. The bone segments should also be soaked in antibiotic irrigation or betadine before reimplantation. Intravenous antibiotics are then continued for at least 48 hours. When the bone cannot be initially replaced, delayed skull reconstruction is indicated.

Skull osteomyelitis can arise from multiple sources, including extension of aerodigestive infection, progression of cutaneous wounds, and direct expansion of middle ear and mastoid disease. Each of these processes is associated with characteristic virulent organisms that can combine with patient susceptibility to produce a clinically significant infection. Although treatment with antibiotics is often successful, surgical drainage, debridement, and ultimately resection is sometimes necessary. Bone and tissue cultures should be taken to help guide length and type of antibiotic therapy. Unless frank osteomyelitis is noted at surgical drainage and debridement, the bone flap can be replaced, followed by placement of a subgaleal drain to eliminate fluid and infectious materials.

Encephaloceles are the most common congenital cause of cranial and skull base defects. Occipital lesions are normally diagnosed early in life because of their location. Midline, orbital, nasal, and skull base lesions may present in a delayed fashion as nasal obstruction, rhinorrhea, vision changes, recurrent meningitis, and facial deformity. Their treatment requires a combined neurosurgical and craniofacial approach for resection of redundant neurologic tissue, repair of dura, and separation of intracranial contents through reconstruction of the cranium and skull base.

Various other pathologic entities can lead to cranial defects that also require reconstruction. Extension of extracranial malignancies such as melanoma can necessitate skull removal as part of

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their surgical treatment. Osseous pathology such as fibrous dysplasia can also involve the cranium, and in severe cases may require contouring or resection of the affected area. Additionally, treatment of pathology with radiation and chemotherapy can lead to soft tissue fibrosis and atrophy and may require a soft tissue flap to cover the bony defect. Management of the primary pathology will dictate timing of the bony and soft tissue coverage.

Goals of treatment

The goal of secondary cranioplasty is restoration of permanent cerebral protection with both hard and soft tissue in a cosmetically acceptable fashion. Normalization of neurologic symptoms and cerebral physiology are also desired outcomes of successful reconstruction. Extreme examples of altered cerebral physiology that are associated with cranial defects are the syndrome of the trephined and paradoxically low intracranial pressure. Syndrome of the trephined involves the onset of new neurologic deficits from altered cerebral blood flow in brain parenchyma under the defect, whereas low intracranial pressure with herniation has been attributed to a negative pressure gradient between cranial and spinal compartments. Replacement of the bone flap can be curative in both conditions. Establishing a balance between waiting for resolution of intracranial pathology (eg, trauma, infection, bleeding, increased intracranial pressure, cerebrospinal fluid malabsorption) and the development of complications from the defect itself requires experience and good judgment.

Successful reconstruction necessitates the treatment and resolution of the cause. A healthy soft tissue envelope also must be present to accommodate an expanded cranial contour. Although numerous treatment protocols can be found in the literature, each patient requires a thorough physical examination and radiographic evaluation, including CT or MRI studies. The subsequent development of a thoughtful treatment plan must take into consideration the cause, timing, and resolution of the injury and intracranial sequelae, and the characteristics of the cranial defect.

Augmented treatment planning and custom implant fabrication

The availability of high-resolution imaging has combined with high-powered computer hardware, image manipulation software, and rapid prototyping to augment the reconstructive process. This process is facilitated by the use of stereolithographic biomodels, virtual surgical planning tools, and the production of custom fabricated craniomaxillofacial implants.

The evaluation of patients before cranioplasty includes CT imaging acquired at a maximum increment of 1.0-mm axial slices. Specific protocols are available from each implant or model manufacturer. These data are then converted to Digital Communications in Medicine (DICOM) standard files, which the radiology provider should make available on CD or DVD. The DICOM dataset can then, physically or electronically, be sent to companies that are approved by the US Food and Drug Administration to produce biomodels or custom craniomaxillofacial implants. If a model is desired, most manufacturers offer several different grades of material and levels of detail depending on the needs of the surgeon. These models can normally be fabricated and shipped within days. Surgical models are helpful for planning, and can also be used by the surgeon to locally fabricate or precontour implants.

Several manufacturers can facilitate surgeon-directed custom implant fabrication. This process involves DICOM data segmentation and the creation of a virtual skull model. Depending on the size, location, and complexity of the cranial defect, a virtual reconstruction can be performed to mirror or mimic normal anatomy. Subtracting the patient's virtual skull from the reconstructed skull then creates a virtual implant. The treatment plan can then be reviewed and modified by the surgeon. If necessary, a biomodel and implant prototype can be printed for physical evaluation and approval. If autogenous bone will be used for the clinical reconstruction, the virtual implant can be printed and used as a surgical template. Otherwise, the implant is fabricated, sterilized, and shipped to the surgeon for use at surgery.

Overall, this process allows for improved visualization of the cranial defect and helps optimize the treatment plan by allowing for multiple iterations of reconstruction. The fabricated implants are highly accurate and lead to improved cosmetic outcomes, reduction in dead space, and reduced

operative times. These benefits easily outweigh the increased costs associated with these techniques and materials that are commonly cited as a factor to justify exclusive use of traditional methods.

Skull defect reconstruction

Soft Tissue Preparation

The evaluation of the overlying soft tissue is a critical step before definitive cranial reconstruction. Consideration must be given to the health and thickness of the soft tissue, location of prior incisions, presence of scar tissue, anticipated expansion of the surface area, contour of the cranium after cranioplasty, and any history of radiation or infection. Depending on the size and nature of the soft tissue deficiency, staged soft tissue management may be required instead of simultaneous reconstruction. Regardless, placements of incisions away from the cranial defect, careful handling of tissues, and tension-free closure are all required.

Numerous local soft tissue rotation and advancement flaps are available for scalp resurfacing. Most designs involve local tissue excision and rearrangement with the use of back cuts and local undermining. These flaps can vary in size and range from simple to complex. If local tissue rearrangement is insufficient, regional or distant soft tissue recruitment requires careful consideration. The temporoparietal fasciocutaneous flap is the regional flap most commonly used in this area. It can be based on the anterior or posterior temporal arteries and has a great deal of flexibility in size and pedicle length. In some circumstances the skin flap overlying the cranial defect may be healthy but thin. Free fat grafting or the use of thin but well-vascularized fascial flaps, including the pericranium or temporoparietal fascia, may add additional bulk. If a significant amount of tissue is required or the area has a significant history of irradiation, microvascular free tissue transfer must be considered and may include either robust muscle flaps (ie, rectus abdominis, latissimus dorsi), which will atrophy to a more appropriate contour over time, or thinner tissues (ie, radial forearm, anterior lateral thigh) that are more consistent in appearance but provide less initial bulk.

Scalp expansion is an adjunctive technique that may be performed before hard tissue repair. Although specific protocols may vary, surgical implantation of tissue expanders is followed by serial hydraulic inflation. Resection of the damaged tissue occurs at skull reconstruction, with immediate coverage by the expanded scalp. The location of the expanders must be carefully chosen in areas with stable underlying cranium and healthy overlying hair-bearing scalp. Additional consideration must be given to the planned location of the incision for future exposure of the cranial defect, and the size of the planned scalp resection. Expanders are normally placed between the pericranium and galea, allowed to heal for 2 to 3 weeks, and then expanded on a weekly basis until the desired amount of expansion is achieved before final cranioplasty.

Neurosurgical Management

Outstanding neurosurgical issues can dictate success or failure of cranioplasty despite meticulous technique. Before skull reconstruction, imaging studies should be performed to assess brain pathology, rule out hydrocephalus or stroke, and ensure adequate brain relaxation to provide the optimum milieu for cranial repair. Many patients develop hydrocephalus after trauma or infection. If the cisterns around the brainstem and cervicomedullary area are patent, a lumbar drain or lumbar shunt can provide good cerebrospinal fluid (CSF) diversion. In most cases, however, a permanent ventriculoperitoneal shunt is needed. The risks of shunting include malfunction and infection (foreign body implant). Meticulous technique, thorough skin preparation (chlorhexidine or betadine), systemic antibiotics, and diligent postoperative wound care can minimize infection. If CSF leakage is present, it must be addressed through direct repair or diversion at or before cranial repair. Postoperative intracranial pressure should be maintained in the normal range (<20 mm Hg), because high or low pressures can contribute to bone resorption, poor healing, and other neurosurgical sequelae after cranioplasty.

General Surgical Technique

The patient is brought to the operating room and placed in a supine position for oral endotracheal intubation. The placement of ocular lubricant and tarsorrhaphy sutures is ideal. Depending on the

location of the planned reconstruction, the patient is placed on a horseshoe Mayfield headrest in a supine or prone position. If prone, the globes must be checked to ensure they are free from pressure. The head and body must be positioned so that the C-spine is in a passive and neutral position.

The planned incision is ideally marked over healthy tissue away from the area of reconstruction and should take into account the need for wide exposure, management of the soft tissue as previously discussed, a hidden position of the final scar, and tension-free closure. Depending on the location, not all prior incisions should be reused, and often a posteriorly positioned postauricular curvilinear coronal incision is the most appropriate. If desired, a small area of hair surrounding the incision line may be trimmed with electric hair clippers but should not be shaved.

Preoperative antibiotics should be used to cover skin flora unless intradural or aerodigestive tract communication is present. This treatment is continued for 48 hours for autogenous reconstruction, and for an additional 72 hours for alloplastic reconstruction. Preoperative steroids should be used to assist with soft tissue edema and continued for 24 hours unless contraindicated. A solution of 1% lidocaine with 1:100,000 epinephrine is used to inject the incision line and into the subgaleal plane for hydrodissection. The cutaneous incision is opened in segments with a knife. Dissection in the subgaleal plane is then performed bluntly, leaving the pericranium in place. The incision is then retracted with opposing skin hooks and a needle tip cautery device used to cut and coagulate the deeper tissues. This technique virtually eliminates blood and minimizes damage to the hair follicles. Raney scalp clips may be counted and applied if desired once the incision is completely opened.

Dissection is continued until the margins of the entire defect are delineated in a supraperiosteal plane. Further dissection of the scalp overlying the cranial defect may be difficult. It is best completed with a combination of sharp and blunt instrumentation with the goal of leaving a healthy overlying skin flap without violating the dura. The pericranium can then be incised around the margin of the defect, and pericranial flaps can be developed that are extremely versatile and may be used for dural patching, soft tissue bulk, and separation of the aerodigestive tract. Subperiosteal dissection is then performed and any bleeding from the cranium controlled with gelfoam and thrombin. The use of bone wax is minimized unless significant osseous bleeding occurs.

In the anterior region, the supraorbital neurovascular bundles should be identified and protected within the soft tissue flap. Laterally, the temporalis musculature should be left in place to minimize potential muscle atrophy and temporal hollowing. If temporal exposure is necessary, the temporalis can be taken up with the scalp flap or incised, leaving a small cranial margin of muscle to help with reapproximation, and then elevated as an independent muscle flap. If the zygomas or lateral orbits are to be exposed, dissection must be performed in a plane deep to the temporal branch of the facial nerve.

If dural repair is necessary, pedicled or free pericranium should be used over synthetic materials. This technique provides watertight coverage to prevent CSF leakage and brain herniation, and the pericranium itself has osteoinductive properties. Intracranial dead space is often left untreated to allow for any brain swelling that may occur. It is also reasonable to obliterate the space with epidural tacking sutures to prevent air or fluid accumulation and possible mass effect. Ventricular or lumbar shunt management is important because both can create negative intracranial pressure that can exacerbate intracranial dead space. Trapped air usually resolves on its own, but air under tension can be treated with high flow oxygen supplementation or, in extreme cases, aspiration.

The frontal and ethmoidal sinuses are often in continuity with the area requiring reconstruction. If a communication is encountered, any respiratory epithelium must be completely removed. The nasofrontal or ethmoidal recess should be packed with autologous bone, fat, muscle, dermis, or fascia, and pericranial tissue can then be brought down to provide an additional barrier between dura and aerodigestive tract. Consideration should be given to delaying further reconstruction until healing has occurred.

After wide exposure and management of the underlying dura, brain, and aerodigestive tract communication, the boney margins of the defect should be clearly delineated. Rotary instruments or rongeurs should be used to freshen the bone edges back to healthy bleeding bone with defined margins. If an autogenous bone graft is to be used for reconstruction, a template can be fashioned at this time, or a prefabricated template may be brought to the field and verified. The donor site is chosen and exposed, and the bone graft is harvested as outlined later. After the donor site is reconstructed, the graft should be adjusted to match the defect with minimal gaps or overlaps. Bone grafts may be bent, scored, or segmented to provide the best fit and contour. Large block grafts

may be fixated in one piece, whereas larger defects may require sequential reconstruction and fixation.

If a prefabricated implant is used, it should be soaked in antibiotic laden irrigation. After placement over the defect, it should be evaluated for bulk, contour, margin thickness, and position. Significant modifications are more readily made at a separate sterile table. Minor modifications may be completed in situ before or after application of fixation. The use of lag screws is ideal but require enough implant overlap and thickness to prevent material fracture. Otherwise, fixation should be applied with the fewest number of low-profile titanium plates and screws necessary for rigid stability.

In all cases the soft tissues should be redraped and the contour of the reconstructed skull evaluated for further irregularities. Over or under contour deformities may be corrected through repositioning of the principle bone graft or implant, additional onlay bone grafting, placement of bone cement, or ostectomy with rotary or hand instruments.

Once the reconstruction is complete, available pericranial flaps can be used to cover the implant edges and fixation hardware. If the temporalis muscles have been elevated, they must be reapproximated to the temporal line with permanent tacking sutures. If the dissection has included the zygomas or periorbital regions, the lateral canthi and supporting fascia of the face must also be resuspended using permanent tacking sutures to the skull, pericranium, or temporal fascia. Copious antibiotic laden irrigation should be used to ensure the wound is free of debris. Raney clips should be removed and counted. Closure is then performed in layers using Vicryl to close the underlying galea and subcutaneous tissue. The skin should be closed with permanent sutures to be removed in 7 to 14 days. Drains are not normally used, but meticulous attention must be given to hemostasis and layered closure to minimize the risk of hematoma and seroma. After closure, the head should be irrigated, cleaned, and covered with antibiotic ointment, fluff dressings, and a loose stocking.

Materials

Historically, numerous materials have been used for cranial reconstruction, including gold, silver, wine-soaked dressing, animal bone, and plaster. The ideal material should replace missing tissue in strength, weight, stability, and biocompatibility; it should be cost-effective, readily available without a donor, easy to custom fabricate and modify, resistant to infection, and have minimal impact on postsurgical radiation or imaging. Biotechnology will meet these goals in the future. Significant advances have been made in the use of stem cells and biocompatible scaffolds, recombinant bone morphogenetic proteins, and engineered tissues. However, widespread clinical application of these technologies has yet to be realized for craniofacial reconstruction.

The most commonly used materials include stored or harvested autogenous cranial bone, polymer implants, titanium mesh, and bone cements. Each may be applied to a specific clinical situation depending on their individual positive and negative qualities. The final selection of technique and material is made on an individual basis and may depend on the origin, defect, patient, surgeon preference, availability, and cost.

Autogenous Cranial Bone

The gold standard for skull reconstruction is autogenous cranial bone. In general, the use of autogenous bone is associated with low rates of infection and limited tissue reaction. Its use satisfies the reconstructive dictum of replacing like tissue with like tissue from an identical embryonic and intramembranous origin. Healing and integration occurs through osteogenesis, osteoconduction, and osteoinduction. It is readily available in the same surgical field and may provide an appropriate contour. In contrast, cranial bone is associated with additional donor site morbidity, has a limited volume of availability, can be difficult to shape, and may be associated with long-term resorption. In general, its use is supported for the replacement of previously stored iatrogenic bone flaps, the reconstruction of small to medium-sized defects, and pediatric cranioplasty.

Preoperative CT scans should be evaluated for the amount and quality of available cranial bone. In general, the most suitable portion of the cranium is the parietal region because of its thickness, hidden location, position in relation to the venous sinuses, and relationship to underlying intracranial

anatomy. The developmental age of the skull is also critical because a significant medullary separation of the cortical layers is rarely present until 3 to 5 years of age. In general, cranial bone can be harvested for reconstruction in full or partial thickness. Regardless of the technique used, the keys to successful bone graft harvest include wide subperiosteal undermining of the scalp and use of a template of the defect to outline the required graft.

Full-thickness harvest

Full-thickness graft harvest is most often indicated in cases that require a significant volume of bone for reconstruction (Fig. 1). Exposure through a coronal flap is ideal for both the cranial defect and the donor site. A formal craniotomy must be completed with the use of bur holes, wide dissection of the underlying dura, and a craniotome to delineate the bone flap. At a minimum, struts of stable cranium are left intact to acts as reference points for donor-site reconstruction. The bone is then removed and taken to a sterile table where it can be separated into an inner and outer layer with the use of a saw and osteotomes. Depending on the size and thickness of the bone flap, it may be more easily cut into strips and then divided into inner and outer cortices. The outer table is then used to immediately reconstruct the donor site, leaving the inner table split-thickness bone to be used for the cranioplasty. This bone is best stored in a sponge dampened with cool saline.

Split-thickness harvest

Split-thickness graft harvest is most often indicated in cases that require a smaller volume of bone for reconstruction (Fig. 2). The extent of exposure is determined by the amount of bone graft required. In this instance, a Steiger bur is used to outline the donor site through the outer cortex only. Depending on the size of the defect, the outer cortex may be harvested in one piece or again divided into smaller linear strips with the Steiger bur. A larger diameter bur is then used to bevel the skull adjacent to the donor site. The small Steiger bur is used to undermine the outer cortex into the cancellous space. A thin spatula osteotome is subsequently used under direct visualization to extend the undermining, with careful attention to the angulation of the osteotome to minimize the risk of inner cortical harvest or perforation. Once an adequate plane is developed from the periphery, a slightly larger osteotome is advanced further into the cancellous space. This process is completed circumferentially until the outer cortex is free, or sequentially to remove multiple strips. A curette may be used to harvest the remaining cancellous bone, and the inner table is then carefully

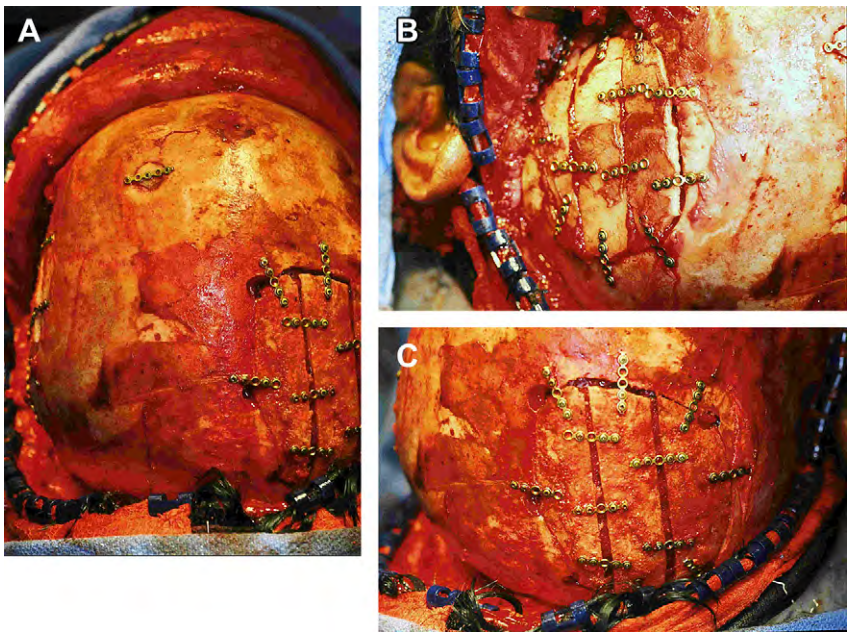

Fig. 1. Full-thickness bone graft harvesting and split insertion at donor and recipient sites. (*A*) Right parietal defect covered with graft secured by titanium plates. (*B*) Left temporal defect covered by harvested bone and metal plating system. (*C*) Parietal harvest site repaired with split bone graft.

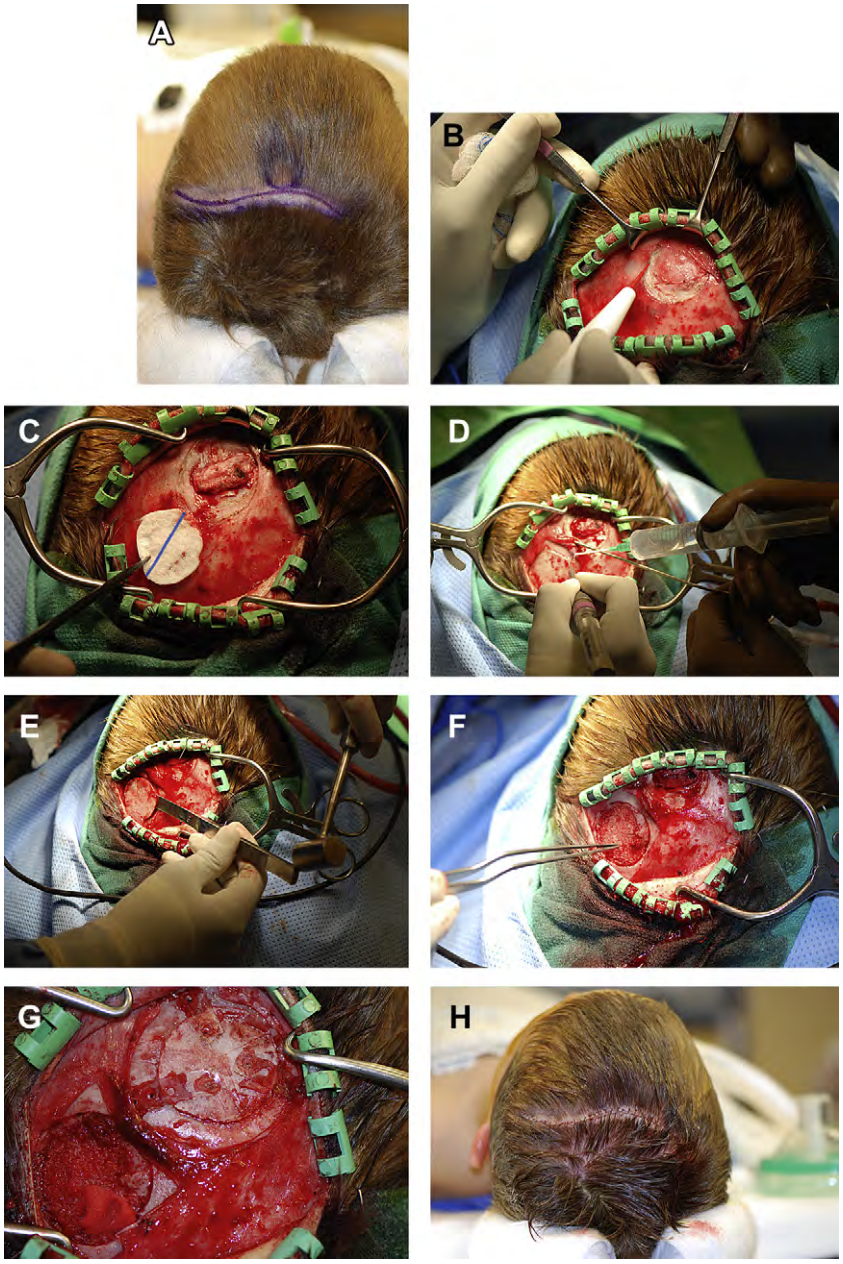

Fig. 2. In situ harvesting of external table for covering a small skull defect in a child. (*A*) Left parietal scalp incision at site of defect. (*B*) Elevation of pericranial flap, which is sometimes used for dural covering or reapproximated for better bone remolding. (*C*) Cotton pledge template from defect placed over donor site before marking. (*D*) Drilling of outer table along marked donor site. (*E*) Osteotome elevation of the outer table. (*F*) Donor site with intact inner table. (*G*) Bone graft secured in place using absorbable plating system. (*H*) Scalp incision closed in layers with absorbable suture material.

inspected for any areas of perforation. Bleeding should be controlled with the conservative application of bone wax.

Storage and replacement of iatrogenic bone flap

The replacement of a previously removed bone flap to the defect with or without the addition of fillers (eg, mesh, cement, autogenous particles) depends on the reason for craniectomy. When removed for trauma, the flap can be replaced when intracranial pressure issues have resolved (ie, swelling abated or hydrocephalus is addressed), not before. For defects incurred because of infection,

a trend is seen toward early cranial repair (as opposed to the long-held dogma of waiting up to 6 months). A preoperative scan should show resolved cerebral edema and an absence of enhancing fluid collections.

High resorption rates have been noted with replacement of bone in large defects after freezer storage. A relationship does not seem to exist between failure and time to replacement. Dividing the bone flap into two overlapping segments (or three) and placement in a subcutaneous abdominal pocket theoretically may reduce the risk of future infection. Other complications include poor wound healing, hematoma formation, sunken plate, seizures, and hydrocephalus, many of which can be prevented through waiting until intracranial issues resolve, the incision and flap are free of tension, and the patient is optimally medically managed.

After exposure, the bone flap can be replaced after soaking in a Betadine solution. The authors' preference is for replacement with an absorbable plating system, without leaving foreign material permanently. A meticulous dural repair with pericranium or dural substitute makes the return surgery for defect repair much easier and safer. A drain is left in the subgaleal space to prevent fluid collection formation around the flap that can become superinfected.

Alloplastic materials

Many different alloplastic materials can be applied to a variety of clinical situations. In general, the most desirable attribute is the elimination of a surgical donor site. In addition, many of these materials can be used in the presurgical fabrication of implants. These implants are completed either indirectly through the use of virtual surgical planning tools and rapid prototyping or directly with the use of custom fabricated stereolithographic models as previously described (Fig. 3).

Bone Cements

The latest generation of calcium phosphate craniomaxillofacial bone cements (ie, HydroSet, Stryker, Kalamazoo, Michigan; Norion, Synthes, West Chester, Pennsylvania) polymerize to form hydroxyapatite, the primary inorganic component of bone. After the exposure of the cranial defect and preparation of the margins, these products are mixed according to the specific package instructions and then immediately applied as putty or injected into smaller areas. Ideally, they rapidly set in a damp environment and quickly achieve a level of strength and stability so that they can be trimmed with hand or rotary instrumentation. Early manipulation is a common technical error that can lead to material microfracture and early breakdown.

These materials are useful for superficial cranial contouring and camouflaging partial thickness defects. They are commonly used to cover underlying titanium implants or as a "grout" surrounding

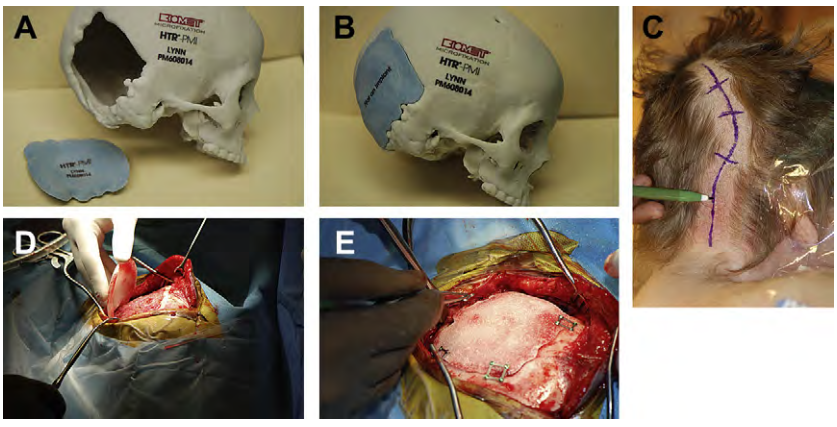

Fig. 3. Skull defect repair using alloplastic polymethylmethacrylate (PMMA) custom implant. (*A*) Skull mold with defect and custom-made PMMA implant (*blue*). (*B*) Excellent fit and complete coverage of the defect with implant (*blue*) in position. (*C*) Right parieto-occipital scalp incision (preexisting) over skull defect. (*D*) PMMA implant held in position for coverage and proper fit. (*E*) Implant secured in position using titanium plating system.

the edges of implants (Fig. 4). They are not useful for full-thickness defects and should not be placed in direct contact with the dura. Controversy exists in the literature regarding the ability of this material to act as an osteoconductive scaffold and eventually be replaced by host bone. If used in small volumes, the risk of infection is low; however, it is critical that they remain separate from the frontal sinus and the rest of the aerodigestive tract.

Polymers

A host of different polymers have been used over time to reconstruct cranial defects. In general, they provide adequate strength for the reconstruction of large full-thickness cranial defects. They are also radiolucent and nonferromagnetic, allowing for postoperative CT and MRI. However, they are associated with a risk of delayed infection and erosion. They are well suited for presurgical fabrication, and several North American companies offer virtual surgical planning and rapid prototyping services.

Polymethylmethacrylate (PMMA) is the oldest polymer that is still currently in use. The monomer is toxic and its polymerization is an exothermic process. These two attributes limit its use to indirect custom fabrication (ie, modified PMMA hard tissue replacement [HTR] computer aided design and computer-aided manufacturing [CAD/CAM] implants, Walter Lorenz, Jacksonville, Florida) or direct presurgical fabrication using stereolithographic models. Directly fabricated PMMA implants can be significantly less expensive because a prosthetic laboratory technician can fabricate them. The implants are strong and easily modified at surgery. They are normally surrounded by a fibrous capsule and do not integrate with the surrounding tissue.

Medpor (Porex Surgical Inc, Newnan, Georgia) is composed of high-density polyethylene that allows fibrous tissue ingrowth because of the size (>100 μm) and interconnectivity of its pore structure. This function is reported to increase stability and minimize likelihood of infection. Medpor cranial implants are available in several designs that can be sized and then modified with either hand or power instrumentation before or after implantation. Porex also provides a custom fabrication process for atypical or complicated defects.

Titanium

Titanium alloy is available as a mesh that can be used for cranioplasty. It can be presurgically cut and contoured to a stereolithographic model or adapted in situ. Titanium is light, relatively strong, biologically inert and nonferromagnetic. However, it causes some CT imaging interference and is prone to long-term erosion through the soft tissues. It can be used in conjunction with bone cements to improve contour and integration.

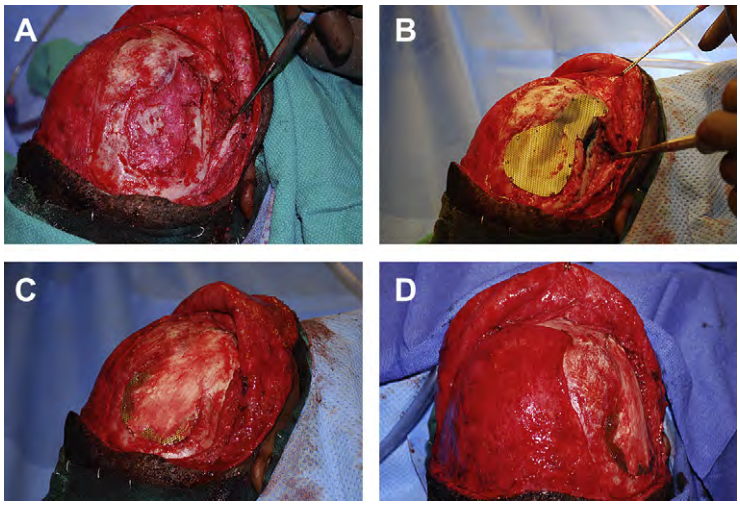

Fig. 4. Titanium mesh and calcium phosphate cement coverage of skull defect. (*A*) Right frontoparietal skull defect. (*B*) Titanium mesh secured into defect (note undercontouring for proper filling with cement). (*C*, *D*) Defect filled with calcium phosphate and contouring.

Complications

Complications include perioperative events and other short- and long-term sequelae. These complications have been studied extensively in the literature and correlate largely with the size and extent of the bony defect. The most significant perioperative complications relate to neurologic injury, including damage to the underlying brain, seizures, hydrocephalus, CSF leakage, and mass effect from swelling or bleeding. Often these can be attributed to the patient's original or unresolved brain pathology.

In the short term, seroma or hematoma collection places the reconstruction at risk. However, unless extremely large, they are most often managed expectantly with an additional period of systemic antibiotic coverage. Infection can be multifactorial and its occurrence must instigate a thorough local and systemic evaluation of the patient and surgical site. In most instances of infection, wound breakdown results and a partial or complete loss of the reconstruction or implant can occur.

Long-term follow-up is necessary to monitor for delayed sequelae. Implant exposure and scalp erosion, incomplete autogenous integration and bone resorption, reconstruction migration, and unacceptable cosmetic results are all encountered. These complications are more likely to occur in large complex defects with a compromised recipient site, and require a thorough reevaluation.

Special circumstances: the pediatric patient

Pediatric skull defect reconstruction requires the additional appreciation of normal growth and development of the craniofacial region. Brain growth is considered propulsive early in life, with brain and skull volumes quadrupling over the first 2 years. This rapid growth can cause the progression of an injury, such as the development of a growing skull fracture. In other situations, brain expansion is beneficial and simplifies the management of intracranial dead space, which quickly becomes obliterated in growing children without intervention. The cranium is 95% of its final size by 7 years of age.

The osteogenic and inductive potential of the pediatric dura and periosteum is extremely high. This characteristic allows for the regeneration of relatively large defects and may negate the need for reconstruction of smaller defects, depending on their location and soft tissue coverage. However, if a previously harvested bone flap is reimplanted, both the occurrence of growth and bone regeneration may cause difficulty when attempting to replace the flap.

The rapid growth and change in contour of the cranium limits the use of permanent alloplastic implants and titanium fixation. Because of continued skull growth an implant can be "outgrown," and the pseudo-migration of permanent fixation is well documented. Autogenous bone, resorbable cements, and biodegradable bone plates and screws should be used exclusively before 7 years of age. The development of the skull must also be considered for bone graft harvesting. The adult cranium is composed of an inner and outer cortex with interposed cancellous bone. The development of distinct layers does not occur until 3 to 5 years of age, which may complicate split-thickness calvarial bone harvesting early in life. If the skull is deemed to be of sufficient thickness (>5 mm), a full-thickness bone flap may be carefully split. Otherwise, small volumes of particulate bone graft may be harvested and used in conjunction with resorbable mesh or bone cement to reconstruct small cranial defects. Bone scrapers (ie, Mx Grafter, Maxilon Laboratories, Inc, Hollis, New Hampshire) can be used to scrape the outer cortex and collect the bone shavings. Another option is to use a rotary drill to perform a superficial ostectomy combined with the use of a suction trap (ie, Lukens Trap) to collect the bone particulate.

Summary

Secondary reconstruction of skull defects provides long-term restoration of both form and function. Resolution of the inciting pathology and residual neurosurgical issues should be followed by clinical and radiographic evaluation. Presurgical planning may be enhanced by the use of a stereolithographic models and virtual tools. Separation of the aerodigestive tract from the intracranial cavity and preparation of the soft tissue bed are both presurgical requisites. Wide exposure of the defect is followed by meticulous reconstruction with indicated materials and

placement of rigid fixation; gentle management of the soft tissues and careful layered closure help increase the likelihood of success. Long-term follow-up is the key to diagnosis of delayed complications and continued improvement in quality of care.

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Craniofacial Approach for Anterior Skull-Base Lesions

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In the field of craniofacial surgery, different surgical methods for treating skull-base lesions have evolved. A thorough understanding of the microsurgical and functional anatomy of the anterior skull base is crucial for optimal selection of surgical approach and minimizing complications. Frazell and Lewis' observation, "The entire cribriform plate can rarely be resected without creating cerebral complications" [1], reflects the therapeutic principles generally accepted in 1963. Since then, innovative and aggressive surgical techniques evolved because of the high rate of failure and complications following radiation therapy for nonresectable tumors in this area. Improved survival rates stemmed from the realization that en bloc resection of the tumor in the ethmoidal roof or cribriform plate area is possible only through a combined transcranial and facial approach [2].

Smith and colleagues [3] first described the combined facial and intracranial tumor resection for frontal sinus tumors. Dandy [4] and Ray and McLean [5] set the stage for surgical possibilities in the skull-base area when they described a combined intracranial orbital approach to posterior orbital tumors. In 1959, Malecki [1] described a combined approach for treatment of posterior frontal, sphenoid, and ethmoidal roof fractures [6]. In 1963, Ketcham and colleagues [7] emphasized the need for ethmoidal roof and cribriform resection of tumors in the paranasal sinuses. Derome [8] applied craniofacial resection techniques when developing transbasal surgery for regions of the sphenoid and clivus. In 1969, Terz and colleagues [9] described modifications for surgery of tumor extensions into the pterygoid fossa. The rate of complications associated with his modifications, however, was high. Over the next 3 decades, multiple publications established the usefulness of combined resection techniques for removal of tumors involving the anterior cranial floor, ethmoidal roof, cribriform plate, or orbital roof.

The risk of intraoperative hemorrhage, injury to intracranial structures, and cerebrospinal fluid (CSF) leaks can largely be obviated by the combined methods. Involvement of important structures, such as the cavernous sinus, internal carotid artery, and optic nerve, however, impose the primary limitations of craniofacial resection techniques for malignant anterior cranial-base lesions [2].

In the past decade, using endoscopy and endoscope-assisted microsurgery has evolved for the treatment of selected pathologies of the skull base, primarily in the sellar and suprasellar regions. The endonasal transsphenoidal approach permits the exposure and removal of sellar and selected suprasellar lesions without the need for brain retraction [10].

Craniofacial skull-base lesions

Microsurgery remains the preferred therapeutic modality for a myriad of disorders involving the craniofacial junction. These disorders may be classified as lesions arising extracranially, lesions arising from bones, and lesions arising intracranially but extending beyond the confines of the cranium (Table 1). Clinical behavior can be extremely variable. Fibrous dysplasia of the frontal bone may warrant surgery only when its local extension produces symptoms resulting from compression or

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Table 1
Classification of common pathologies involving the anterior cranial base

Arising Extracranially, May Extend Intracranially	Arising From Cranial Bones, May Extend Extracranially or Intracranially	Arising Intracranially, May Extend Extracranially
Cutaneous:	Chordoma	Meningioma
Basal and squamous cell carcinomas and melanoma	Osteoma	Neurofibroma
Orbital:	Fibrous dysplasia	
Neuroblastoma, optic nerve glioma and rhabdomyosarcoma	Chondroma	
Paranasal and nasal cavities:	Chondrosarcoma	
Adenocarcinoma and squamous cell carcinoma	Fibrosarcoma	
Salivary glands:	Osteosarcoma	
Adenocarcinoma and mucoepidermoid tumors		
Others:		
Esthesioneuroblastoma, nasopharyngeal angiofibroma, and sarcoma		

From Al-Mefty O. Surgery of the cranial base. Boston: Kluwer Academic; 1989; with permission.

expansion. Fibrosarcoma involving the same location, however, mandates immediate surgical excision. Radiographic features often help the surgeon make the proper differential diagnosis. CT scan and MR, with axial, sagittal, and coronal views, are complementary and are necessary for full delineation of the lesion and involvement of surrounding structures (Figs. 1–4). These factors are crucial in deciding operability and planning the surgical approach and its extensiveness. Adequate biopsies, however, are generally required for confirmation of the diagnosis and planning the surgical treatment. Biopsy is generally recommended to exclude tumors that can be treated with radiation or chemotherapy and do not require resection, such as lymphoma. Frequently, an adequate biopsy can be obtained via a transnasal, transantral, nasopharyngeal, or other extracranial approaches [2].

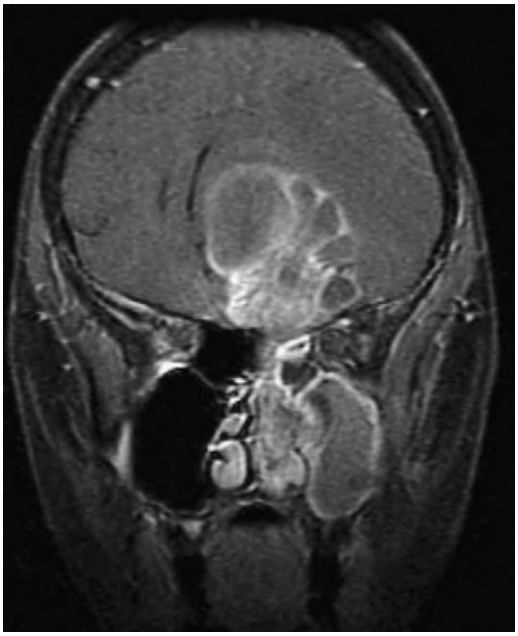

Fig. 1. Coronal MRI of Nasopharyngeal carcinoma extending intracranially. There is extension of tumor invasion into the frontal lobes and pterygomaxillary area.

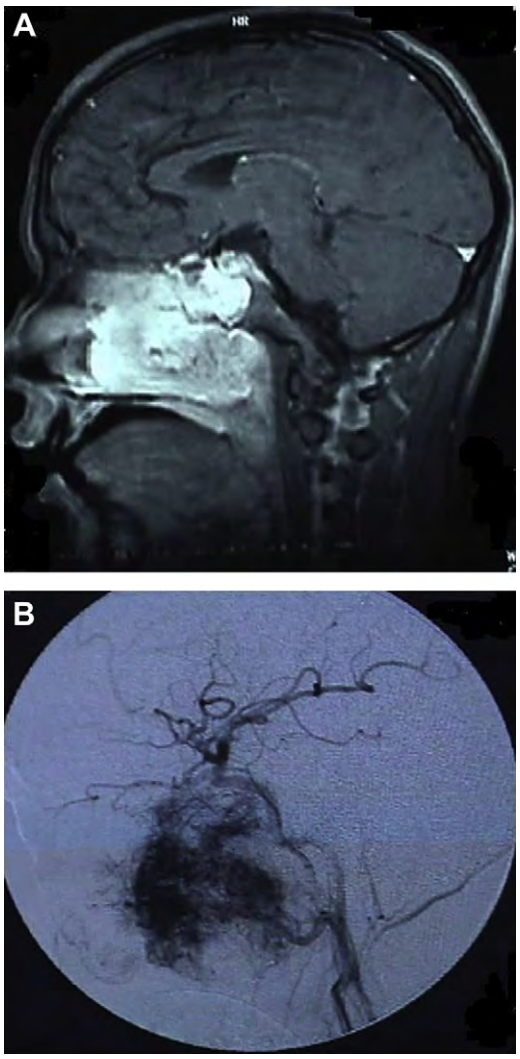

Fig. 2. Sagittal MRI and cerebral angiogram of Juvenile Nasopharyngeal Angiofibroma. Selective endovascular embolization of this vascular tumor can be essential prior to surgical resection (*A, B*).

Craniofacial resection of anterior skull-base lesions

Indications

This is a transcranial extradural approach to the anterior skull base through a unilateral or bifrontal craniotomy. It is primarily designed for sinonasal and nasopharyngeal tumors (see Fig. 1) involving the anterior skull base. Several variations of this approach with extensions involving osteotomies of the orbit and nasal bone allow further exposure of anterior and central skull-base structures in the midline [11]. This approach provides an excellent subfrontal access to midline and paramedian lesion but also for bilateral orbital and cavernous sinus lesions. An extended orbitofrontal approach is used for unilateral orbital and anterior middle fossa lesions.

This approach has many advantages: a cosmetically invisible incision; the ability to reconstruct the face, orbits, and nose, as well as the forehead; access to a large area of the skull base; and ready access to cranial bone for splitting to aid in the reconstruction. In addition, this approach under the frontal lobes minimizes brain retraction and allows both intradural and extradural access if necessary [12].

Operative Technique

The patient with an anterior skull-base lesion is placed in the supine position with the head in a horseshoe headrest or Mayfield head holder. A lumbar drain is inserted through a split mattress into

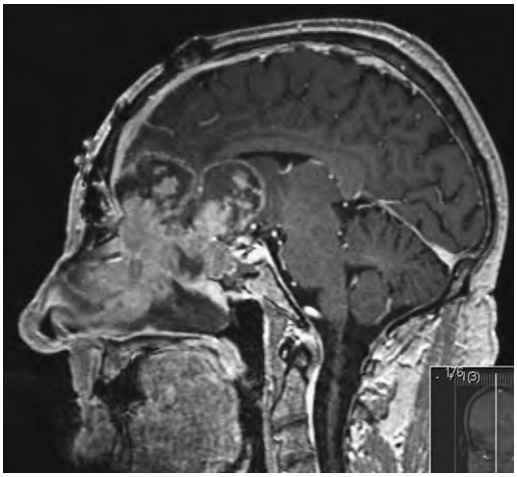

Fig. 3. Sagittal MRI of Malignant Meningioma extending intracranially and extracranially.

the lumbar thecal sac and connected to a spinal drainage set (Fig. 5). The head, face, and neck are prepared and draped in the usual manner. The right thigh of the patient is also prepped and draped for possible harvesting of fascia lata and split-thickness skin grafts. The 2 operating disciplines (neurosurgery and otolaryngology) maintain separate instruments and fields to minimize cranial contamination by nasopharyngeal flora [2].

Cranial component

A bicoronal incision is most commonly used for standard craniofacial approach. A skin flap is elevated and retracted anteriorly. The frontalis fascia underlying the galea and temporal fascia are initially preserved. The frontalis fascia is incised bicoronally as posterior as possible along the superior temporal lines, elevated anteriorly without temporalis muscles, and kept attached at the supraorbital margin supplied by the supraorbital vessels. This provides a superbly vascularized pericranial flap that not only offers a barrier at the craniofacial defect but also provides strong support to the brain (Fig. 6).

A rectangular craniotomy based at the midline is performed using the high-speed drill. Two posterior holes on each side of the sagittal sinus are connected using a rongeur. The midline burr hole and the keyhole are connected using a Gigli saw. The remaining holes are connected using the

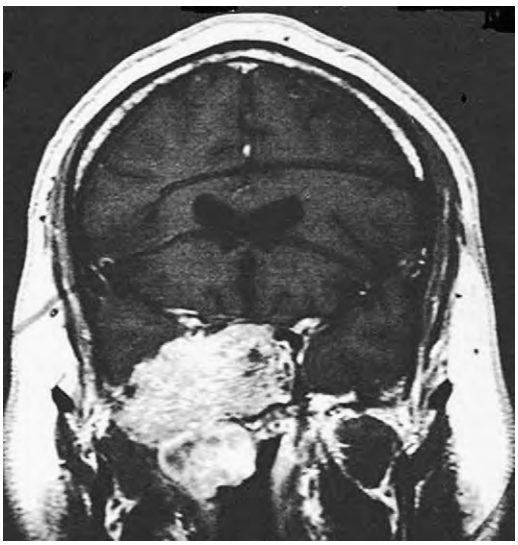

Fig. 4. Coronal MRI of invasive Adenocarcinoma extending intracranially into middle cranial fossa.

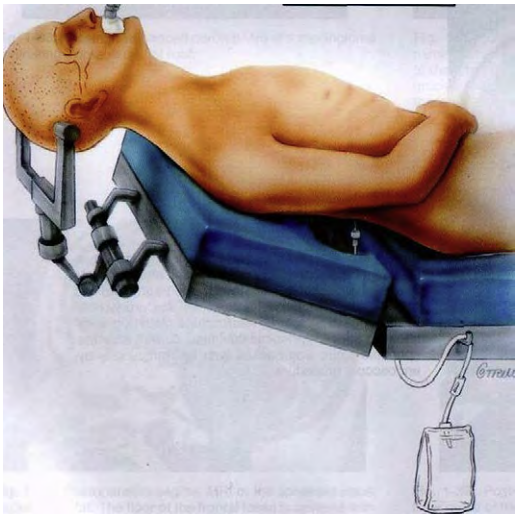

Fig. 5. Patient is positioned in supine position with the head in a Mayfield head holder. A lumbar drain is inserted through a split mattress into the lumbar thecal sac and connected to a spinal drainage set. (*From* Al-Mefty O. Meningiomas of the anterior cranial base. Operative Atlas of Meningiomas. Philadelphia, PA: Lippincott-Raven; 1998; with permission.)

craniotome, and the bone flap is removed. The frontal sinus mucosa is exenterated and the posterior wall of the frontal sinus is removed from the cranial flap (Fig. 7).

The dura is then carefully elevated from the floor of the anterior cranial fossa. The olfactory fibers are sectioned and the frontal lobes, covered by the dura, are elevated to the planum sphenoidale. A high-speed drill or small rongeur is used to remove crista galli and expose the anterior skull base reaching the planum sphenoidale.

Facial component

Extracranial exposure is more frequently gained by a Weber-Fergusson incision on the side of greater tumor involvement. Depending on the tumor extension and need, the upper lip can be divided to gain good exposure to the maxilla following definition of a cheek flap (Fig. 8). This step is required especially if a maxillectomy is indicated for complete tumor removal. The periosteum is incised and the medial canthal ligament detached. The surgeon should make an effort to preserve the lacrimal sac. The periosteal incision is carried into the medial and superior orbital walls. The periosteal attachment of the nasal bones should be preserved to ensure the blood supply. A lateral osteotomy in the nasal maxillary area is then performed. Some bone removal from the pyriform aperture can facilitate intranasal exposure. The periosteum from the medial and superior walls of the opposite orbit can be raised

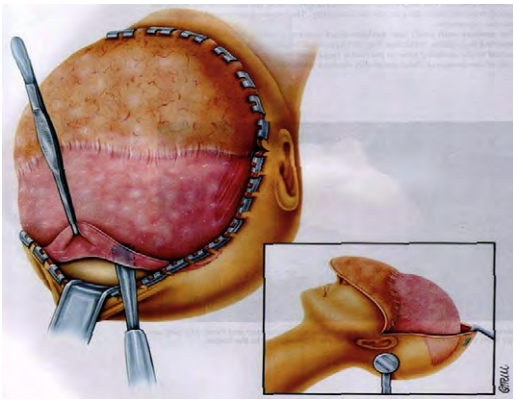

Fig. 6. A large pericranial flap, based on the supraorbital and frontal vessels, then is incised as far posteriorly as possible, dissected forward, and reflected over the scalp. (*From* Al-Mefty O. Meningiomas of the anterior cranial base. Operative Atlas of Meningiomas. Philadelphia, PA: Lippincott-Raven; 1998; with permission.)

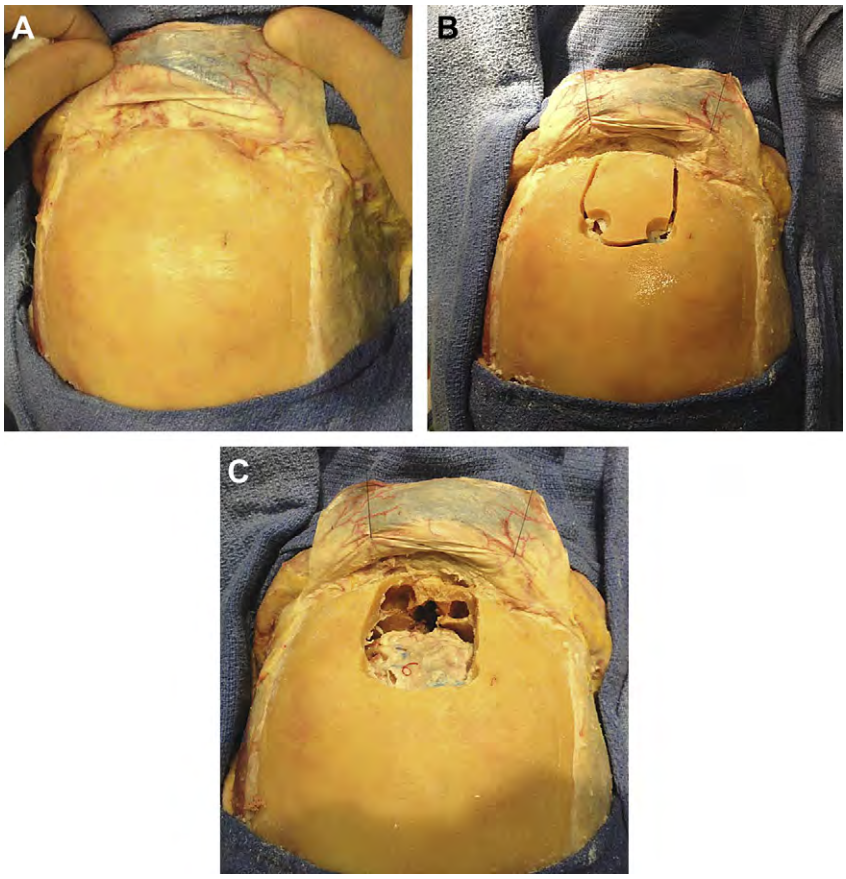

Fig. 7. Cadaveric illustrations outlining the harvest of central pericranial flap, a midline bifrontal craniotomy flap before and after elevation of bone flap. The frontal sinus mucosa is exenterated and the posterior wall of the frontal sinus is removed from the cranial flap. Anterior ethmoidectomy was performed for access into the nasal cavity. This flap is used when the orbit is not included with the resection (*A–C*). (Surgical dissections performed by Samer Elbabaa, MD; courtesy of Samer Elbabaa, MD, Little Rock, AR.)

in a similar manner. The anterior and posterior ethmoidal bundles can be cauterized with a bipolar cautery and divided.

A transverse osteotomy at the root of the nose can then be performed to allow adequate rotation of the nose. With proper retraction of the periorbita, appropriate osteotomies are performed at the inferomedial orbital wall junction, or the zygomatic arch and lateral orbital wall if a total maxillectomy is performed in conjunction. In the event of a total maxillectomy, hard palate cuts can be made with a Gigli saw. The surgeon must exercise caution to preserve as much of the soft palate as possible to maintain velopharyngeal functions.

Recently, some literature reported that the transnasal endoscopic approach combined with a frontal craniotomy represents an alternative to the traditional transfacial approaches and allows adequate resection of extensive malignant sinonasal tumors with comparable oncologic outcomes [13].

Craniofacial approach modifications

The extent of craniofacial resection is tailored to adequately expose each individual lesion. This craniofacial approach can be modified in the presence of an intracranial tumor extension or when the orbit, the maxilla, or both are to be included in the specimen.

Various incisions have been used in craniofacial resection, including a bicoronal incision, which is the most commonly used in the craniofacial approach; the Weber-Fergusson incision, which may be extended into the philtrum and upper lip inferiorly and around the subciliary area; and a butterfly incision, which is helpful if the tumor is localized to the anterior cranial floor with minimal inferior extension (see Fig. 8).

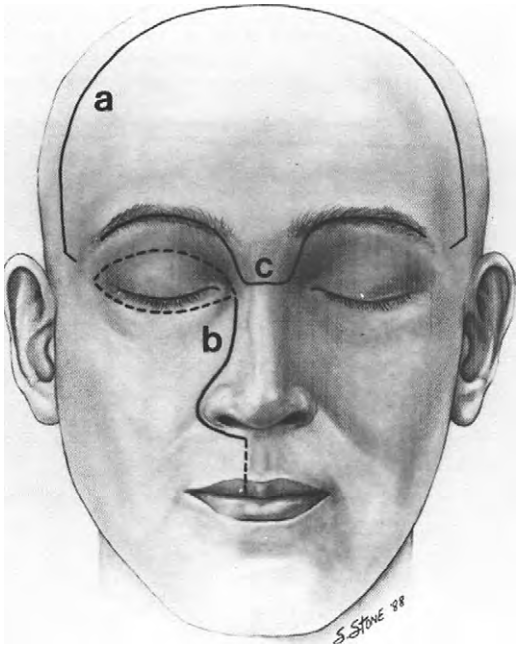

Fig. 8. The commonly used incisions for craniofacial approaches include: (a) a bicoronal incision; (b) the Weber-Fergusson incision. This incision may be extended into the philtrum and upper lip inferiorly, and around the subciliary area. If the orbital contents can be preserved, the incision is extended into the upper eyelid as shown; (c) A butterfly incision will be helpful if the tumor is localized to the anterior cranial floor with minimal inferior extension. (*From* Al-Mefty O. Surgery of the cranial base. Boston: Kluwer Academic; 1989; with permission.)

A supraorbital unilateral or bifrontal bone flap is recommended when the orbit is to be included with the specimen. Figs. 9 and 10 demonstrate the position of the holes and bony cuts performed in the mobilization of supraorbital unilateral or bifrontal flap.

If the tumor extends intradurally, the dura is opened with a transverse incision (Fig. 11). The superior sagittal sinus is ligated and sectioned at its insertion. The frontal lobes are elevated and, with an

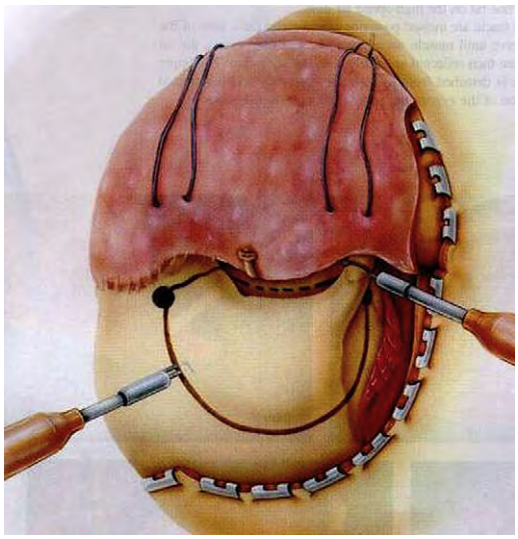

Fig. 9. Illustration demonstrate the supraorbital unilateral bone flap. Notice the position of the holes and bony cuts. (*From* Al-Mefty O. Meningiomas of the anterior cranial base. Operative Atlas of Meningiomas. Philadelphia, PA: Lippincott-Raven; 1998; with permission.)

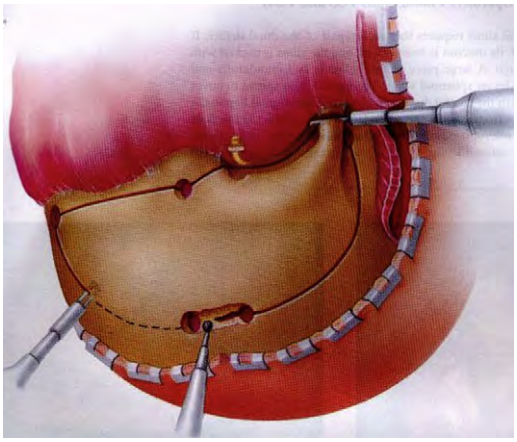

Fig. 10. Illustration demonstrate the supraorbital bifrontal bone flap. The flap is used for large tumors that require the inclusion of the orbit with the specimen. Notice the position of the holes and bony cuts. (*From* Al-Mefty O. Meningiomas of the anterior cranial base. Operative Atlas of Meningiomas. Philadelphia, PA: Lippincott-Raven; 1998; with permission.)

adequate margin, the dura is excised around the tumor. Preference in this case is to repair the dura with a fascia lata graft at this stage to protect the intracranial contents from contamination [2].

When resection is limited to the ethmoid, the bony incision is extended longitudinally on both sides of the cribriform plate to the planum sphenoidale. Two connecting transverse incisions are made posteriorly and anteriorly, and the specimen is freed intracranially for further resection using the transfacial approach. To avoid inadvertent injury to the optic nerve or entry into the middle cranial fossa, the surgeon must take extreme care when making posteromedial orbital osteotomies. Finally, to include the sphenoid in the specimen when required, the sphenoid bone between the optic nerves is drilled. When the orbit is to be included, a single longitudinal osteotomy is made along the cribriform plate on the opposite side, extending transversely across the planum sphenoidale and the posterior orbital roof down to the lateral orbital wall (Fig. 12).

The zygomatic arch is then sectioned and the orbital foramen is unroofed. The optic nerve and the ophthalmic artery are exposed, coagulated, and transected. Once the cranial osteotomy is completed,

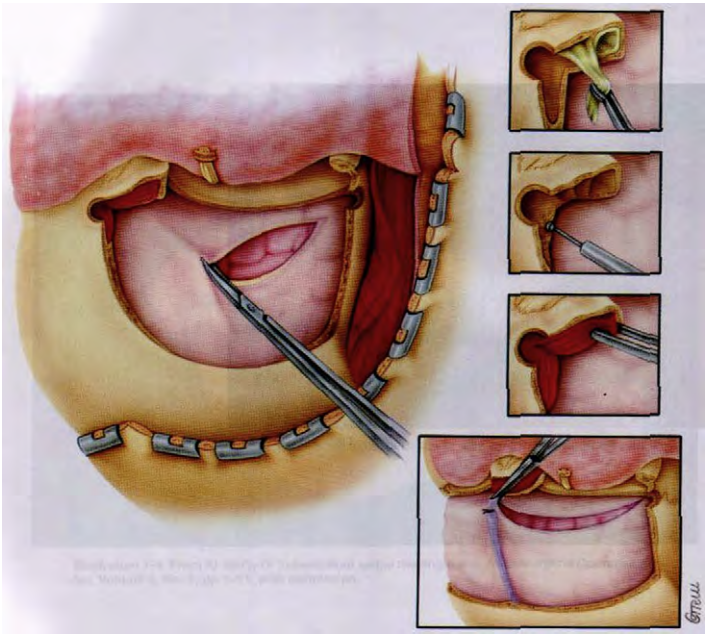

Fig. 11. If the tumor extends intradurally, the dura is opened with a transverse incision. (*From* Al-Mefty O. Meningiomas of the anterior cranial base. Operative Atlas of Meningiomas. Philadelphia, PA: Lippincott-Raven; 1998; with permission.)

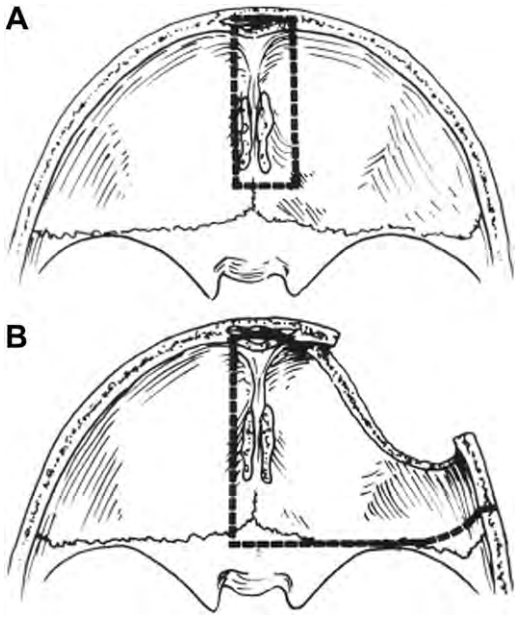

Fig. 12. Illustration outlining the intracranial osteotomy: (*A*) in case of ethomoidectomy; (*B*) when the orbit is included in the specimen. (*From* Al-Mefty O. Surgery of the cranial base. Boston: Kluwer Academic; 1989; with permission.)

the frontal lobes are protected with surgical patties, and the head-and-neck surgeon embarks on the facial exposure.

Complication Avoidance

Neurosurgical monitoring is required to rule out pneumocephalus, CSF leak, postoperative hemorrhage, or infection. Despite the extensive nature of the operative procedure, it is surprising how few complications are encountered when the surgery is performed by experienced surgeons.

To avoid CSF leakage and septic complications, closure should be meticulous. Any dural defect is grafted with a fascia-lata or synthetic graft using a water-tight closure technique. The pericranial temporalis flap is then brought into the cranial-base defect and secured with a suture to the deepest part of the dura on the anterior fossa floor, or to small holes made in the planum sphenoidale. In addition to preventing a CSF leak and being a well-vascularized tissue, this flap serves as a sling, supporting the frontal lobes. It also covers the opening of the frontal sinuses. The bone flap is then secured in place with heavy sutures. The galea and skin are closed in 2 layers.

Acknowledgments

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Evolution of Endoscopic Endonasal Surgery of the Skull Base and Paranasal Sinuses

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For more than 1 hundred years, lesions and disorders of the skull base have provided significant challenges to neurosurgeons, otolaryngologists, and craniofacial surgeons for many reasons. Lesions of the skull base are seated deep in the skull, are often midline, and usually have multiple delicate critical neurovascular structures intermingled between the pathology and the route required to access the lesion. Injuries to any of these structures could result in significant permanent morbidity and even mortality to patients requiring treatment of a skull base lesion. If patients were to survive these risks, postoperative recovery is also fraught with potential problems including cerebrospinal fluid (CSF) leaks, meningitis, swallowing dysfunction requiring tracheostomy or gastrostomy tube placement, and corneal anesthesia resulting in infection or potential visual loss. In the past 20 years, significant advancements in technology and techniques in neurosurgery, otolaryngology, and craniofacial surgery have allowed for a revolution in skull base surgery with the introduction of minimal access endoscopic approaches.

Endoscopic skull base surgery is an extension of endoscopic sinus surgery pioneered by the work of Messerklinger, Stammberger, and Kennedy during the 1970s and 1980s. Their advent and refinement of rod lens endoscopy overcame the limitations of visualization and illumination encountered previously with microscopic endonasal approaches, allowing for less invasive approaches via an endoscopic endonasal route to the paranasal sinuses. As collaborative efforts between neurosurgery and otolaryngology improved outcomes with open skull base approaches, neurosurgeons began to work with sinus otolaryngologists to address pathology of the parasellar regions via a totally endoscopic transnasal approach.

When it became evident that endoscopes allowed better illumination and visualization than microscopic approaches, neurosurgical otolaryngologic teams began to approach pathologies outside the sella. These early attempts of endoscopic skull base surgery, however, were very challenging and resulted in unacceptably high complication rates for many reasons. Pioneers of endoscopic skull base surgery including Sethi, Jho and Carrau, Cappabianca and de Devitiis, Frank and Pasquini, Kassam and Snyderman, and Schwartz and Anand methodically addressed each of these obstacles:

Infection
Imaging
Neurophysiologic monitoring
Dural reconstruction
Radiation therapies/radiosurgery
Operative visualization/magnification
Instrumentation

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Neuronavigation
Hormonal management
Hemostatic management
Neuroanesthesia
Operative illumination.

The development of safe surgical corridors based on anatomic cadaveric endoscopic work in combination with the integration of improved imaging and image-guided stereotactic systems gave the skull base endoscopic surgeon confidence when manipulating pertinent anatomy relative to the location along the approach. The development of endoscopic endonasal coagulation devices and hemostatic materials to manage bleeding as safely as open approaches allowed for more aggressive sharp dissection in the skull base, and allowed the surgeon to extend beyond structures of the skull base that were previously forbidden to operate around because of bleeding concerns, such as the intercavernous sinuses, the medial cavernous sinus, the petrous carotid artery, the basilar plexus, and the anterior cranial fossa. The development of endoscopic vascularized autologous flaps, reconstructive allografts such as DuraGen (Integra Lifescience Corporation, Plainsboro, NJ, USA) and AlloDerm (Lifecell Corporation, Branchburg, NJ, USA), dural sealants such as DuraSeal (Confluent Surgical, Inc, Waltham, MA, USA) and fibrin glue, and techniques to keep reconstructions in place have significantly decreased the CSF leak rates and infection rates to acceptable levels allowing for the expansion of endoscopic endonasal skull base surgery far beyond the sella.

Although advancements in techniques and technologies allowed for expansion in where the endoscopic skull base surgeon could operate, Kassam and colleagues developed what is now called the expanded endonasal approach to access the entire ventral skull base. They divided the approach into units or modules based on its anatomic orientation in the skull base (Box 1). This approach has now become the workhorse for endoscopic skull base surgery from which multiple centers have made slight modifications based on their individual experiences, comfort levels, training, and pathologies treated. Fig. 1 shows a diagrammatic comparison of transsphenoidal microscopic surgery of the sellar region to the expanded endonasal approach to the ventral skull base.

As endoscopic skull base surgery has continued to develop, it is important for the neurosurgeon, otolaryngologist, or craniofacial surgeon who is contemplating the integration of these approaches into their practice to understand that this discipline is a team approach. As opposed to earlier techniques in sellar and skull base surgery, neurosurgeons and otolaryngologists do not work

Box 1. Kassam-Snyderman classification of endoscopic approaches

Sagittal or midline plane
Transfrontal
Transcribiform
Transplanum-suprasellar/infrachiasmatic
Transphenoidal-sellar/transcavernous
Transclival
Posterior clinoidal/transdorsal
Mid-clival
Cervicomedullary
Foramen magnum
Transodontoid
Coronal or paramedian plane
Transorbital
Medial transpetrous-petrous apex
Transcavernous
Transpterygoid
Transpetrous
Suprapetrous
Infrapetrous
Paraphayrngeal space

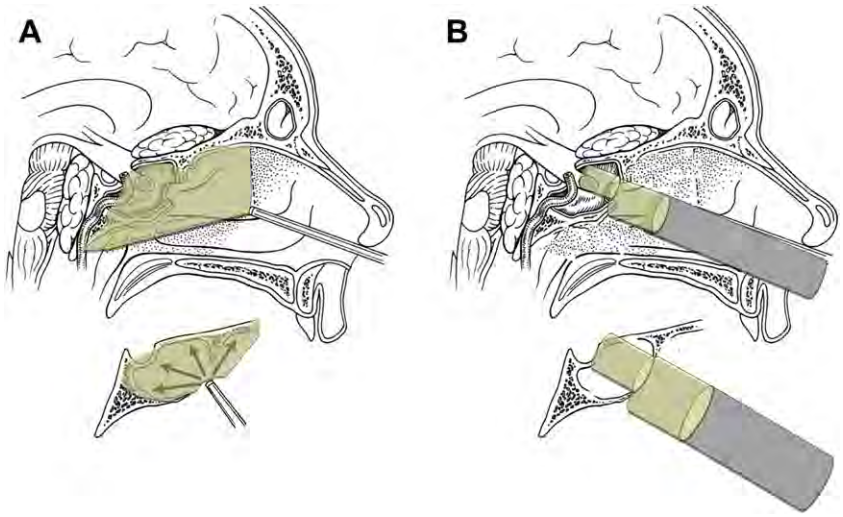

Fig. 1. A comparison of the visualization obtained via the nose for the skull base using an endoscope or a microscope. (*A*) With the endoscope, a cone of light is created extending from its tip to visualize more of the skull base laterally, rostrally, and caudally. (*B*) With a microscopic view, the light and image are directed down a tube forming a cylinder of light that provides excellent visualization of anatomy in direct line of sight of the tube, but is limited laterally, rostrally, and caudally. The endoscope can also be advanced through small corridors that would limit light when using a microscope.

sequentially but rather simultaneously as a team. One person functions as the eyes of the team by manipulating and controlling the endoscope to help the other surgeon best visualize and dissect the pertinent tissues. The second surgeon uses both hands via a binaral technique to meticulously dissect the lesion and expose the skull base anatomy to allow for removal or repair of the skull base pathology.

Patient evaluation for potential endoscopic skull base surgery

Image-guided endoscopic approaches to the skull base can be used to treat a variety of different pathologies. Table 1 shows a list of potential pathologies now amenable to an endoscopic skull base approach. However, in some instances preoperative imaging, patient history, and patient symptoms cannot reliably determine the pathology. In these cases an image-guided endoscopic endonasal biopsy is performed. With little morbidity, a precise diagnosis is made and the best treatment is determined. In some cases this may include an open approach if the lesion cannot be adequately removed via an endoscopic endonasal route. For lesions such as infections, plasmacytoma or lymphoma, a biopsy can prevents more extensive and often unnecessary surgery as these are best treated with nonsurgical therapies.

Lesions involving the sella or pituitary gland can significantly alter a patient's endocrine status and all patients with such lesions should have a basic pituitary hormonal evaluation before any form of

Table 1
Pathologies treatable via an endoscopic endonasal approach

Tumors Derived from Adenohypophyseal Cells	Pituitary Adenoma (Secretory and Nonsecretory) Pituitary Carcinoma
Other primary tumors of the sellar region	Angiofibroma, angiosarcoma, chordomas, chondrosarcoma, choristoma, craniopharyngiomas, fibroma, fibrosarcoma, ganglioglioma, ganglioneuroma, germinoma (ectopic pinealoma), hamartoma (hypothalamic), melanoma, meningioma, paraganglioma, sarcoma, teratoma, glioma (optic nerve, infundibulum. posterior lobe. hypothalamic), granular cell tumor (posterior lobe, pituitary stalk)
Malignant sinus tumors	Sinonasal malignancies, esthesioneuroblastoma
Metastatic tumors	Carcinoma, sarcoma, leukemia, lymphoma
Tumorlike conditions	Inflammatory pseudotumor (fibroblastic tumor), infectious, mucocoele, allergic fungal sinusitis
Other lesions involving skull base	CSF fistula, meningoceles, encephaloceles, Rathke cleft cysts, pituitary apoplexy hemorrhage, chondroma, fibrous dysplasia, medial orbital lesions, basilar invagination, optic nerve decompression for trauma, dermoid cyst, epidermoid, osteoma, inverted papillomas

surgery. Table 2 lists our basic preoperative laboratory tests used in the evaluation of all sellar lesions. This information can be invaluable when creating a plan of care for these patients and in some cases can result in the prevention of surgery. For example, prolactinomas can often be treated medically with bromocriptine or cabergoline. However, other hypersecretory states, such as with acromegaly caused by excess tumor production of growth hormone or Cushing syndrome caused by excess tumor adrenocorticotropic hormone (ACTH) production, have no reliable medicinal therapeutic options and are often only curative with surgery accomplishing a gross total resection. Direct pituitary gland and stalk compression can result in hyposecretory states that do not improve with surgical removal of the tumor, but can put the patient at perioperative risk if not identified preoperatively.

When deciding whether a lesion can be adequately reached or removed via an endoscopic endonasal approach, a full understanding of the associated anatomy of the region is absolutely necessary. The relationship of neurovascular structures and whether the pathology violates pial planes can have a significant effect on the resectability of the lesion or the risk of injury to the patient via an endoscopic endonasal approach. As a result, we prefer to use magnetic resonance imaging (MRI) with and without gadolinium with high-resolution thin cuts through the region of the pathology of interest and through the path that would be taken to reach the lesion. We also prefer fine-cut computed tomography (CT) of the sinuses and skull base with sagittal and coronal reconstructions to understand the bony anatomy of the region and whether any particular sinus barriers exist limiting an endoscopic endonasal approach. In some cases a high-resolution CT angiogram of the skull base is also performed to determine the course of the carotid arteries relative to the pathology of concern as well as determine whether a dehiscent carotid may be present, which is important to know before performing bony decompression to reach the lesion. As a general rule, if we find on imaging that the pathology of interest is distal or beyond the critical neurovascular structures of the skull base, an endoscopic endonasal approach is unlikely to result in an adequate exposure to remove the lesion or repair the defect without significant injury to the patient and thus is not recommended (Fig. 2A). An open approach is then performed. Most lesions of the ventral skull base, however, tend to displace the neurovascular structures laterally, rostrally, or posteriorly, and thus are on the tumor periphery allowing for an endoscopic endonasal approach to be performed without having to go through these structures (see Fig. 2B).

The comfort and the experience of the endoscopic skull base team are also important in determining the resectability of a skull base lesion. Lesions requiring vascular sharp dissection and manipulation as well as intradural lesions require a more experienced skill set than lesions purely in the paranasal sinuses or sella. The Pittsburgh group has divided pathology complexity into 5 levels and has recommended mastery of all lower levels before moving on to the next to minimize the risk of complications (Table 3).

Operative principles of endoscopic endonasal skull base surgery

Patient Preparation and Room Setup

All patients undergoing endoscopic skull base surgery undergo a preoperative image-guided stereotactic imaging study the night before or on the morning of surgery in addition to the

Table 2
Basic serum preoperative pituitary hormone laboratory tests in the evaluation of sellar and pituitary lesions

ACTH[a]	Adrenocorticotrophic hormone (used to evaluate for Cushing disease)
am cortisol[a]	Hyper- or hypocortisolism
Growth hormone	Acromegaly
IGF-1	Insulin growth factor-1 (used to evaluate acromegaly)
LH	Luteinizing hormone
FSH	Follicle-stimulating hormone
Testosterone	Often low in pituitary lesions
Prolactin	Markedly increased in prolactinomas and mildly to moderately increased in tumors compressing the gland or infundibulum, called stalk effect
TSH	Thyroid-stimulating hormone
Free thyroxine (T_4)	Helps to distinguish pituitary versus nonpituitary sources of thyroid dysfunction

[a] If abnormal may proceed to dexamethasone suppression test and 24-hour urinary cortisol testing.

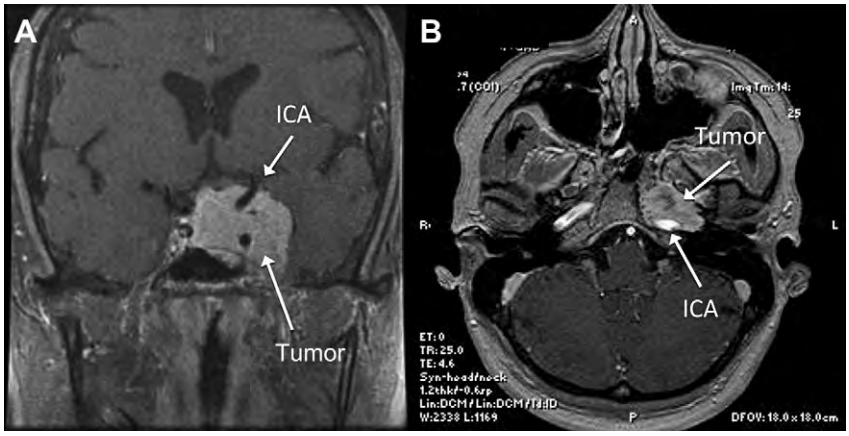

Fig. 2. Location of critical skull base neurovascular structures relative to abnormal pathology in determining resectability via an endoscopic endonasal approach. (*A*) A coronal T1 gadolinium-enhanced MRI scan of the brain showing a left petroclival meningioma encasing the left cavernous and supraclinoidal internal carotid artery. The presence of significant tumor lateral and above the supraclinoidal internal carotid artery limits a gross total resection via an endoscopic approach. (*B*) An axial T1 gadolinium-enhanced MRI of the brain showing a left vidian nerve schwannoma that has displaced the petrous internal carotid artery posteriorly without encasement. By displacing the artery posteriorly, the tumor can easily be approached via an endoscopic endonasal route with a high likelihood of a gross total resection and low risk of vascular injury. ICA, internal carotid artery.

preoperative imaging studies mentioned earlier as part of the initial evaluation of patients with skull base lesions. Depending on previous imaging and suspected neural or vascular structures involved in the approach, this study will be a gadolinium-enhanced MRI of the brain, skull base, and paranasal sinuses or a CT angiogram of the same anatomy. When both bony, vascular, and soft tissues windows are desired, image-fusion studies are used. We find image-guided neuronavigation invaluable for

Table 3
Endoscopic endonasal skull base surgery approach complexity levels

Level	Approach/Procedure
I	Endoscopic sinonasal surgery including sphenoethmoidectomy
	Sphenopalatine artery ligation
	Endoscopic frontal sinusotomy
II	Sellar and ethmoidal CSF leaks
	Lesions of the lateral recess of the sphenoid
	Pituitary surgery
III	Medial orbital decompression
Extradural	Optic nerve decompression
	Petrous apex
	Transclival extradural approaches
	Transodontoid extradural approaches
IV	A. Presence of cortical cuff between vasculature and lesion
Intradural	Transplanum approach
	Transcribiform approach
	Pre-infundibular craniopharyngiomas
	B. Absence of cortical cuff with direct vascular contact
	Foramen magnum approach
	Transplanum approach
	Transcribiform approach
	Infundibular and retroinfundibular craniopharyngiomas
	Transclival approach
V	A. Coronal paramedian plane
Cerebrovascular	Suprapetrous and infrapetrous carotid approaches
	Transpterygoid approach
	Infratemporal fossa approach
	Jugular foramen approach
	Hypoglossal canal approach
	B. Vascular disease
	Aneurysms and vascular malformations

complex lesions of the skull base. It assists us in identifying distorted anatomy and allows individually tailored exposures to be created based on that individual's lesion minimizing extent of exposure while still allowing us to safely remove a lesion (Fig. 3). Although we prefer a fiducial-based system that requires frame fixation to the skull, other nonfiducial, non−frame-based systems have been successfully used for this application and are perfectly acceptable.

Once our patients enter the operative suite, prophylactic antibiotics are administered. If no contraindications exist, we generally give our patients 1 g of ceftriaxone at the time of induction. If the patient's lesion involves the sellar region, potentially requires pituitary gland manipulation for removal, or has already resulted in hypocortisolism, then 100 mg of hydrocortisone is also administered at the time of induction as stress prophylaxis. Traditional neuroanesthesia is then administered and the patient undergoes endotracheal intubation. Once asleep, radial arterial access is obtained and a central venous catheter is placed. A Foley urinary catheter is inserted, lower extremity thromboembolic deterrent (TED)/sequential compression device (SCD) hose are applied, and neurophysiologic monitoring electrodes are placed. A Mayfield 3-pin head holder fixation frame (Integra Lifescience Corporation, Branchburg, NJ, USA) is then applied and the patient is positioned on the operating table for surgery.

The standard operating room setup is shown in Fig. 4A. Anesthesia equipment, intravenous lines, and monitoring leads/devices are positioned to come off to the left of the patient; the surgeons and scrub nurse are on the patient's right side. The endotracheal tube is positioned down and to the left, away from the patient's nose and the surgeons' hands (see Fig. 4B). Electric instrumentation cords (bipolar, monopolar, drill, ultrasonic aspirator) and suction tubing coming off the surgical field are

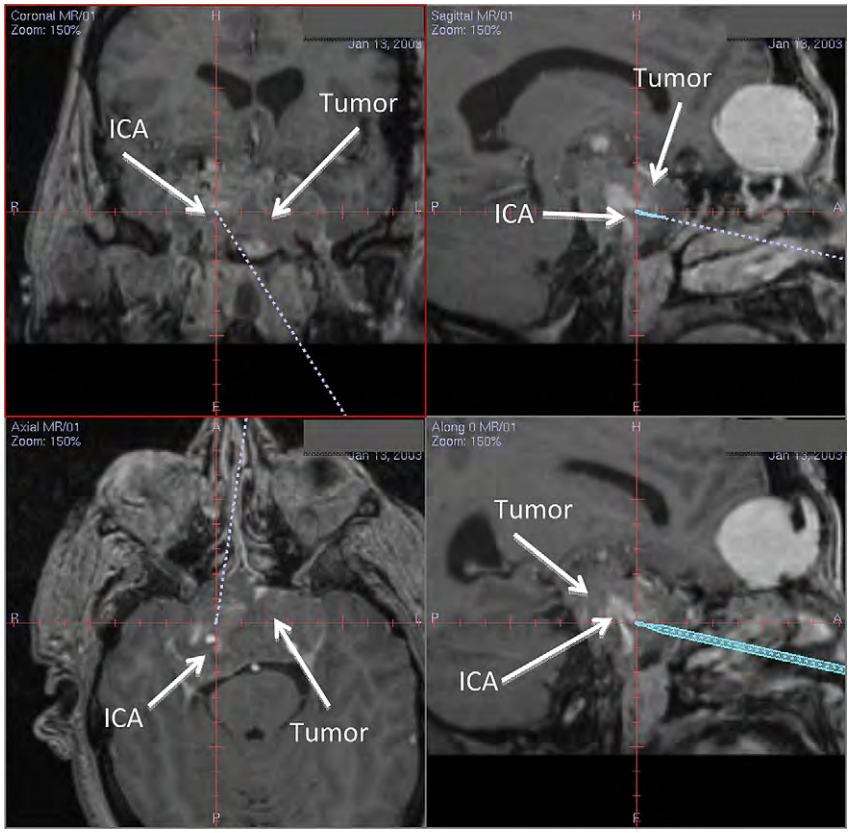

Fig. 3. An example of the benefits of image-guided stereotactic neuronavigation. In this case of a large prolactinoma presenting with acute and rapidly progressive visual loss and diplopia, the patient underwent an endoscopic endonasal removal of the tumor. The goal of this approach was cytoreduction, cavernous sinus and optic nerve decompression to preserve vision with a plan for medical or radiation therapy postoperatively. The tumor completely encased the internal carotid artery. Image guidance was invaluable when removing this fibrous vascular tumor to confirm localization of the artery and prevent injury. The tip of the neuronavigation probe was on what we believed was the internal carotid artery by endoscopic visualization. Image-guided neuronavigation confirmed this and helped to prevent injury to the vessel or the nerves lateral to it. ICA, internal carotid artery.

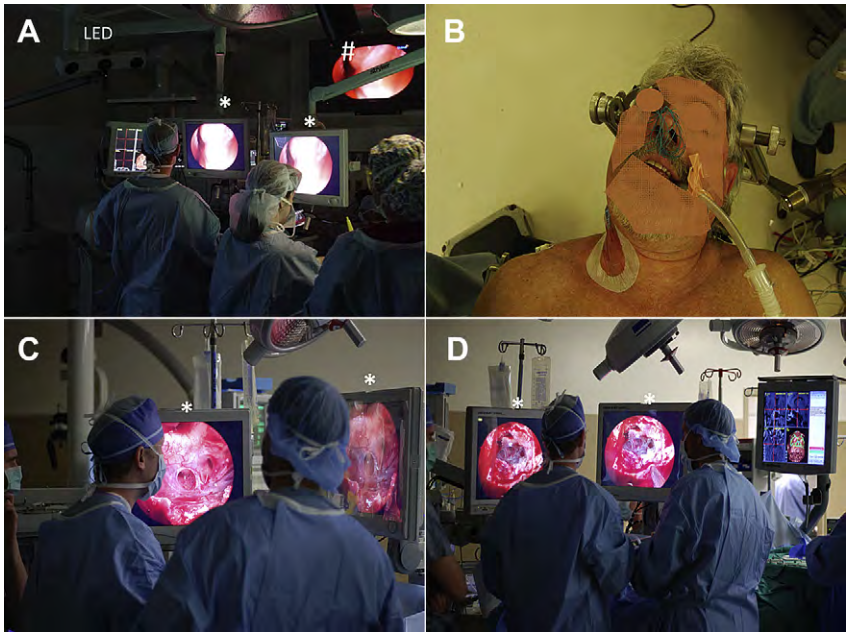

Fig. 4. Operating room positioning for endoscopic endonasal skull base surgery. (*A*) Basic monitor setup with 4 monitors. Two monitors are for surgeon endoscopic visualization, 1 monitor is for neuronavigation (at the head of bed), and the fourth monitor is a room monitor for nursing staff, anesthetists, and students to visualize the endoscope image. Monitors are placed on the patient's left, between the patient and anesthetist. The neuronavigation camera is set up to prevent obstruction by other monitors, instruments, or personnel between its real-time light-emitting diodes (LEDs) and the patient. (*B*) Basic head positioning with endotracheal tube positioned down and to the left away from the nose and the path for passing instrumentation through the nose. The head is tilted to the left and rotated slightly to the right. (*C*) Each surgeon sets up their own high-definition monitor for endoscope image visualization, minimizing strain on the back and neck. (*D*) Variation of neuronavigation monitor placement at the patient's hip toward the foot of the bed based on surgeon preference. *, surgeon endoscope monitor; #, OR staff monitor, ■, neuronavigation monitor; LED, neuronavigation LED camera.

positioned on the patient's left, away from the head and the surgeons' working area. The patient's torso is placed as far to the right of the operating room (OR) table as possible to minimize the amount of reach necessary to operate through the nose. The head is generally tilted slightly (10−15°) to the left and rotated slightly (10−15°) to the right giving the surgeon's as close to a straight trajectory to the paranasal sinuses as possible (see Fig. 4B). Depending on the location of the pathology, the head may be slightly extended for lesions involving the anterior skull base and slightly flexed for lesions lower in the skull base such as the lower clivus and cervicomedullary junction (Fig. 5). Such slight modifications to head positioning significantly improve the angle for visualization and dissection of

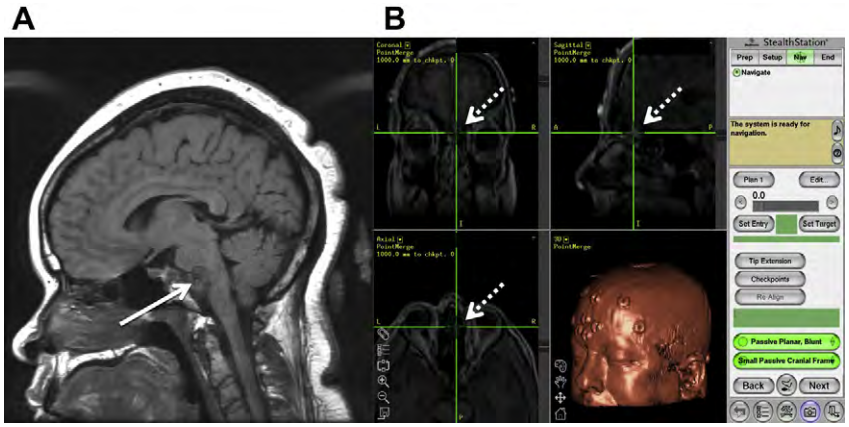

Fig. 5. Lesion location and head positioning. (*A*, *solid arrow*) Clival tumor requiring slight head flexion to maximize exposure via an endoscopic endonasal approach. (*B*, *dashed arrow*) Cribiform plate osteoma requiring slight head extension to best visualize via an endoscopic endonasal route.

pathologies along the extreme margins of the skull base. Neuronavigation optical tracking systems can be placed to the left of the patient either above the head of the bed or by the patient's feet, away from the surgeons' hands and body. We generally use 3 monitors to visualize the operative field of the paranasal sinuses and skull base. Each surgeon has their own viewing monitor positioned at eye level and on the patient's left side to minimize obstruction by the other surgeon's and scrub nurse's body and to minimize neck, back, and arm fatigue that can occur with longer procedures (see Fig. 4C). The third monitor is positioned for anesthetists, circulating nursing staff, students, and other OR staff. A fourth monitor is used for image-guided neuronavigation. It can be placed at the head of the bed or on the left side of the patient at the level of the patient's hip (see Fig. 4A, D).

Neurophysiologic monitoring is routinely used for endoscopic endonasal approaches to the skull base when dural exposure, cranial nerve dissection, or carotid artery dissection is required. Somatosensory evoked potentials and electroencephalography are used to identify changes in blood flow caused by vascular injury, increased intracranial pressure, hypotension, significant blood loss, or direct neural injury. Brainstem evoked response monitoring is used for clival and posterior fossa disease and cranial nerve electromyography is used when specific cranial nerves may be at risk for injury.

Operative approach

Endoscopic endonasal surgery of the skull base involves 3 main components: exposure of lesion and critical associated anatomy, removal of lesion, and reconstruction of the skull base defect created.

Exposure

Once the patient is properly positioned and all equipment is setup, 1% lidocaine with 1:100,000 epinephrine is infiltrated intranasally into the mucosa along the base of the right middle turbinate, posteriorly at the level of the sphenopalatine foramen, the sphenoethmoidal recess, and posterior nasal septum. In some cases, a greater palatine block is also performed transorally. Following direct injection, 0.5% oxymetazoline−soaked cottonoid pledgets are placed between the middle turbinates and the nasal septum bilaterally and are allowed to soak the mucosa of these structures for 5 minutes at which point they are removed. This results in significantly improved visualization during the initial dissection by causing vasoconstriction of the anterior paranasal mucosal vasculature. No antiseptic preparation is used, as we have not found it effective in decreasing the incidence of postoperative infection.

During the initial phase of exposing the skull base a 0° Hopkins 4-mm endoscope (Karl Storz GmbH & Company KG, Tuttlingen, Germany) is used. For straightforward sellar disease, such as adenomas less than 2.5 cm in size, we make every attempt to minimize disruption of the normal paranasal anatomy. For such cases, most of the paranasal dissection is performed on the right side where the endoscope is usually positioned and dissecting or suctioning instruments are placed. The middle turbinate is lateralized to the right, the superior turbinate is visualized, and any septal spurs are removed (Fig. 6A). If the corridor created is adequate then the turbinate does not need to be removed and the posterior portion of the nasal cavity can be well visualized. For more complex lesions for which a broader more lateralizing or rostral-caudal exposure is necessary, the inferior turbinates are fractured out laterally using a Cottle dissector and the right middle turbinate is partially resected. Once the initial corridor is created, the choana, sphenoethmoid recess, and sphenoid ostium are identified (see Fig. 6B). The sphenoid may be entered via the natural ostia or centrally, through the rostrum. We prefer to identify the natural ostium of each sphenoid sinus with a blunt instrument such as a Cottle or freer. The same instrument may be used to dilate the ostium inferiorly and medially. A rotating 2-mm Kerrison rongeur is then used to enlarge the natural ostium medially and inferiorly (see Fig. 6C). The mucosa and bone of the anterior sphenoid is then removed to expose the sinus and reveal the sellar and parasellar landmarks. As the exposure continues laterally toward the lamina papyracea, it may be necessary to trim the superior turbinate and enter the posterior ethmoid cleft. The patient is cautioned preoperatively about the risk of temporary or permanent anosmia. Inferolaterally, the posterior nasal branch of the sphenopalatine artery may be encountered and typically responds to suction Bovey electrocautery.

Although the degree of exposure may vary, it is often useful to remove anterior sphenoid bone superiorly to the skull base and inferiorly to the sphenoid floor. In some cases the natural ostium can

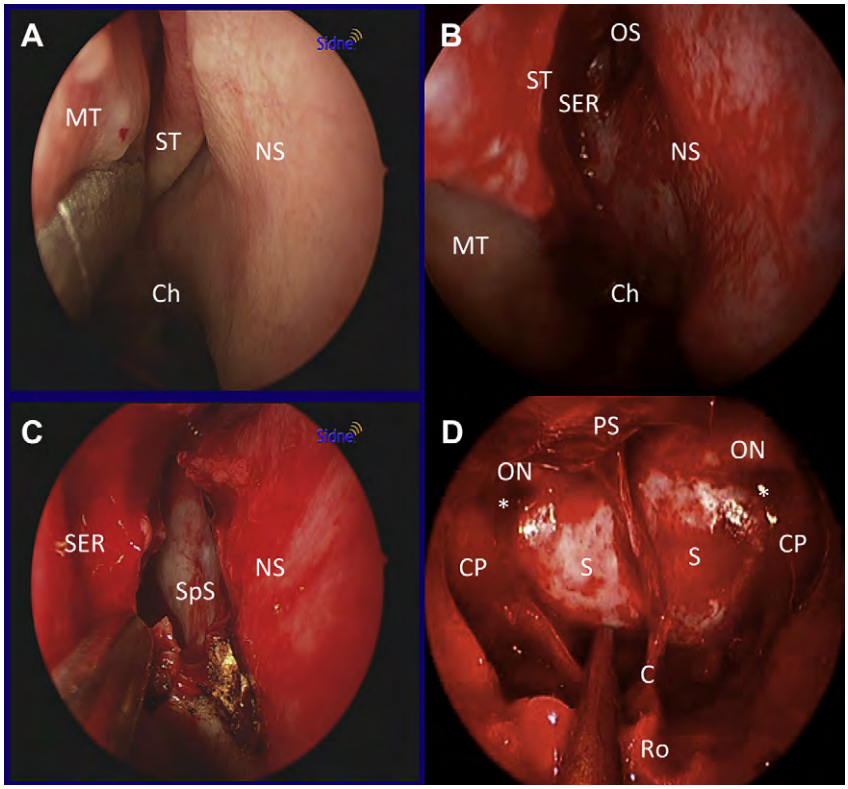

Fig. 6. Basic exposure of the sellar and parasellar region. (*A*) Middle turbinate (MT) is lateralized or removed and the superior turbinate (ST) is visualized. NS, nasal septum; Ch, choana. (*B*) Superior turbinate is lateralized exposing the sphenoid ostium (OS). (*C*) The sphenoid ostium is opened inferiorly and medially exposing the sphenoid sinus (SpS). (*D*) Once the anterior wall of the sphenoid sinus is completely removed and the posterior nasal septum is opened, the entire posterior sphenoid and parasellar bony anatomy is visualized. C, clivus; CP, carotid protuberance; ON, optic nerve protuberance; PS, planum sphenoidale; Ro, rostrum of sphenoid; S, sella floor; SER, sphenoethmoid recess.

be distorted by the pathology or not easily visualized by the surgeon. When this occurs we then enter the sphenoid at the junction of the nasal septum and rostrum of the sphenoid using a Cottle dissector. Once the anterior wall is opened on the right, a Cottle dissector is used to separate the posterior septal nasal bone from the sphenoid rostrum and a back-cutting antrum punch is used to open the posterior nasal septum up to 2 cm from its attachment from the sphenoid rostrum. After this, the left anterior sphenoid wall is opened via the left nare using the same technique described for the right. The bony rostrum is removed using Kerrison rongeurs or irrigating microdrills and wide sphenoidotomies are completed bilaterally that extend laterally to the medial pterygoid plates and lateral sphenoid sinus wall. Midline and paramedian sphenoid septa are then removed until flush with the posterior sphenoid wall using either an irrigating microdrill or punch. However, a keen understanding of the bony anatomy relative to the underlying neurovascular anatomy is crucial before removing any sphenoid septum as they commonly may lateralize to the carotid artery at its vertical segment or even the optic nerve. Neuronavigation is helpful at this point to determine this. When the septum base is away from the carotid artery or optic nerve, it can safely be removed with a punch. When the base extends to the carotid or optic nerve, an irrigating drill with diamond bit should be used carefully to remove the bone until flush with the sellar floor. Once completed, the endoscope is advanced into the sphenoid and the entire sellar and parasellar region can now be well visualized. With the endoscope, this exposure allows for visualization of the planum sphenoidale, clivus, carotid protuberances, optic protuberances, and the opticocarotid recesses (see Fig. 6D). This wide exposure allows for better visualization and for binarial technique to be used by both surgeons to access the skull base. The wide exposure prevents crowding of instrumentation when 1 endoscope, 1 suction, and 2 dissecting instruments are being inserted via both nares simultaneously. By using 4 hands, 1 surgeon can advance or adjust the endoscope and use endoscopic microsuction as needed to maximize

visualization for the second surgeon who is using bimanual sharp endoscopic microdissection technique to manipulate delicate neurovascular structures and resect the pertinent pathology. By placing the endoscope superiorly and retracting the nasal vestibule rostrally, a second instrument can easily be placed underneath the endoscope to reach the skull base without compromising visualization or instrument mobility (Fig. 7).

To keep the camera image sharp and clear during surgery, a suction-irrigation sheath is used over the endoscope to allow cleaning of the lens tip during surgery without having to remove the endoscope from the nose. This minimizes the trauma that can occur with multiple passes in and out of the nasal passageway, shortens surgery time, and permits continuous visualization of pertinent anatomy at times when active bleeding is occurring or when loss of visualization significantly compromises safety to the patient during critical stages of dissection. Once within the sphenoid, monopolar coagulation is not used because of potential risk of injury to adjacent structures, and either bipolar coagulation, warm saline irrigation, mild pressure, or other forms of topical hemostatic agents are used to obtain hemostasis. Arterial bleeding generally can only be adequately controlled with bipolar electrocautery; low flow capillary and venous bleeding is easily controlled with any of the other methods.

Transellar Approach

With this approach, the compartmentalized sphenoid sinus is opened to create a single large window. The exposure extends laterally to the sphenoid recesses bilaterally, superiorly to the posterior ethmoid sinus air cells, and inferiorly to the level of the clivus. The removal of the sphenoid floor allows for direct access to the suprasellar space through the sella by creating more room to look up and move instruments in a caudal-rostral direction. This is particularly helpful for large adenomas and craniopharyngiomas that extend up into the hypothalamus, third ventricle and suprasellar cistern. The mucosa of the sphenoid is removed and the bone of the sellar floor is opened using a combination of Kerrison rongeurs and an irrigating endoscopic microdrill. The basal dura is preserved. The bony opening extends from the superior intercavernous sinus to the inferior intercavernous sinus in the rostral-caudal plane and to the bilateral medial margins of the cavernous sinus in the lateral plane. The basal dura is then opened sharply in a cruciate fashion and the leaflets are reflected laterally and rostrally/caudally exposing the pituitary gland and the sellar lesion for removal. The transellar opening is well demonstrated in Fig. 8.

Transplanum/Transtuberculum Approach

For lesions with a large suprasellar component a transtubercular/transplanum approach is added to the sellar approach allowing for exposure of the optic apparatus, anterior cerebral vasculature, gyrus rectus, and pituitary stalk. By adding this module to the transellar approach, the endoscope can be advanced through the floor of the third ventricle to visualize is structures (Fig. 9). To expose this region, bilateral posterior ethmoidectomies are performed until flush with the floor of the anterior

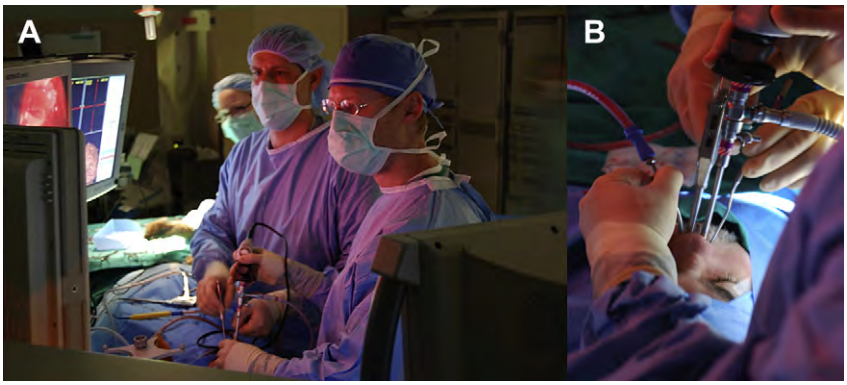

Fig. 7. Binaral technique. (*A*) Otolaryngologist stands closer to head of bed with neurosurgeon to his right allowing for up to 4 instruments to be introduced into the nose at once (*B*).

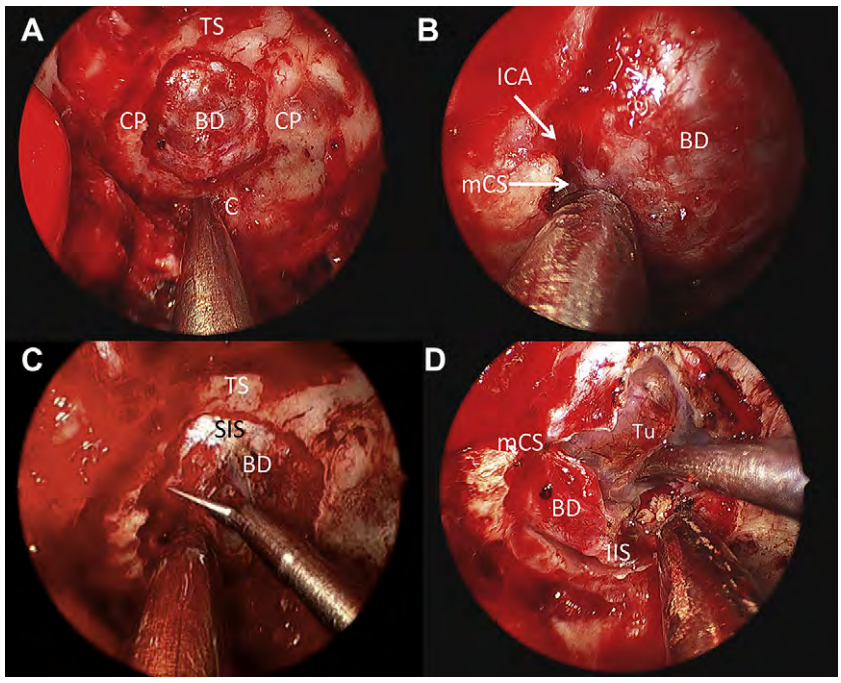

Fig. 8. The transellar approach. (*A*) The mucosa of the sphenoid and sphenoid septum is removed. The sellar bony floor is opened to the medial cavernous sinus bilaterally and intercavernous sinuses rostrocaudally. (*B*) Careful examination of the lateral bony opening to the right cavernous sinus identifies the medial bend of the cavernous internal carotid artery (ICA). (*C*, *D*) Basal dural (BD) opening up to medial cavernous sinus (mCS) bilaterally and intercavernous sinuses rostrocaudally using endoscopic scissors. C, clivus; CP, carotid protuberance; IIS, inferior intercavernous sinus (cut); SIS, superior intercavernous sinus; TS, tuberculum sellae; Tu, tumor.

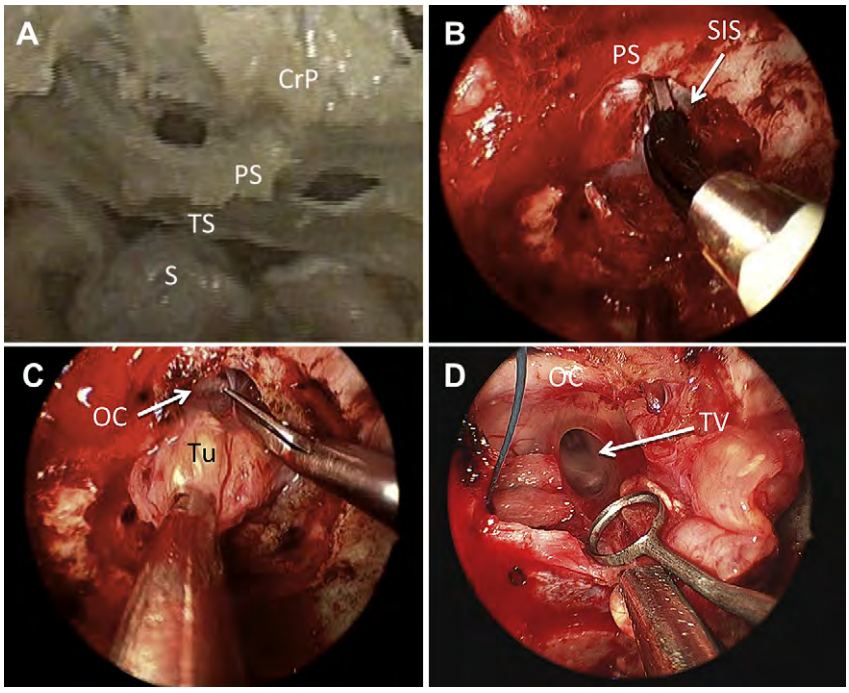

Fig. 9. Transplanum approach. (*A*) Cadaveric dissection showing relationship of planum sphenoidale (PS) to cribiform plate (CrP) and sella (S). TS, tuberculum sellae. (*B*) Superior intercavernous sinus (SIS) coagulation once bony decompression is complete. (*C*) Transplanum extracapsular dissection of tumor (Tu) off optic chiasm (OC). (*D*) Visualization of optic chiasm and third ventricle (TV).

cranial fossa. The exposure can be made laterally as needed up to the lamina papyracea on either side and anteriorly up to the posterior ethmoidal arteries. Extending more anterior than the posterior ethmoidal arteries will result in altered or lost olfaction. The bone overlying the superior intercavernous sinus (SIS) is removed by using the irrigating endoscopic microdrill to remove the tuberculum sellae and planum sphenoidale. The dura is then opened anterior and posterior to the SIS. The SIS is coagulated with angled 45° bipolar electrocautery and sharply incised with angled endoscopic microscissors to complete the dural opening and expose the pathology of interest (see Fig. 9B).

Transcribiform Approach

For lesions extending more anteriorly than the planum sphenoidale, a transcribiform approach can be used. This approach allows for removal of lesions involving the frontal sinus and crista galli (Fig. 10). To gain access to this region, the middle turbinates are removed and the complete ethmoidectomies are performed to lamina papyracea bilaterally. Using a 30° or 45° endoscope, the nasofrontal recess is identified and the superior nasal septum is opened between the 2 recesses. The bone is then drilled to connect the frontal sinuses across midline. The soft tissue in the midline, medial to

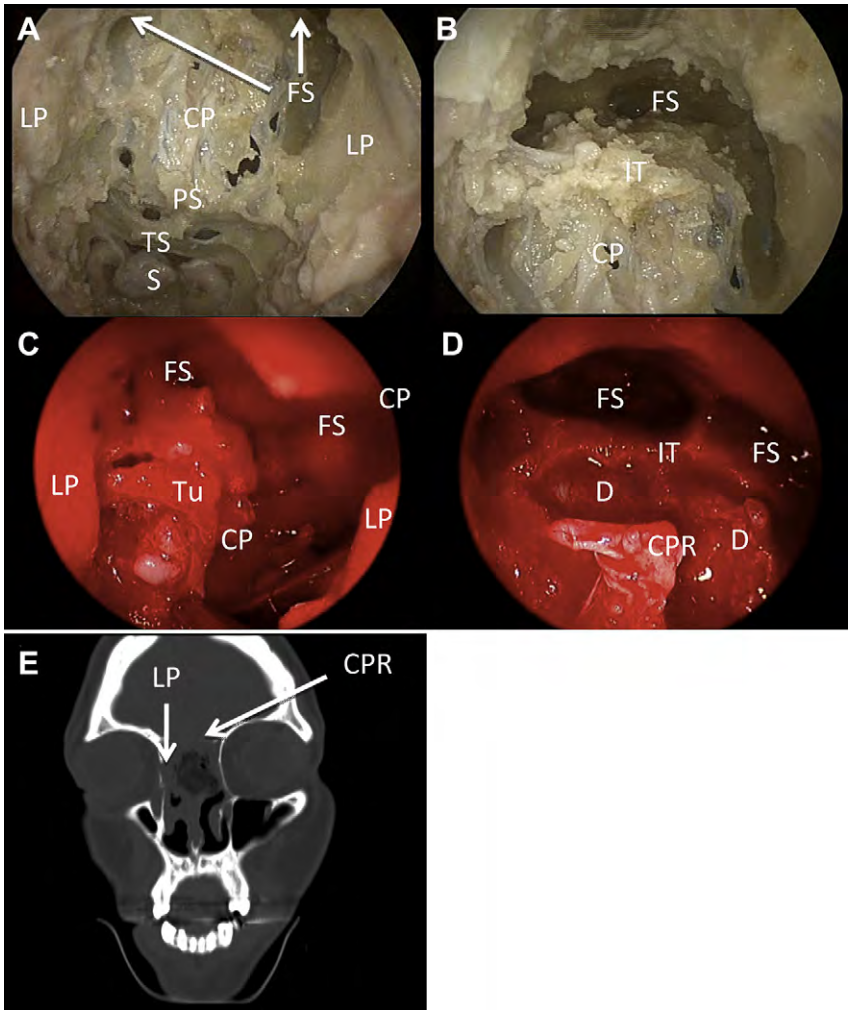

Fig. 10. Transcribiform approach. (*A*) Cadaveric dissection showing relationship of cribiform plate (CP) to frontal sinus (FS), planum sphenoidale (PS), and sella (S). (*B*) Cadaveric dissection showing frontal sinuses opened using a 70° endoscope. (*C*) Image showing a patient with recurrent fibroblastic tumor (Tu) of the anterior ethmoids, cribiform plate, and right frontal sinus causing headache, sinus obstruction with recurrent infections, and anosmia. (*D*) Using a transcribiform approach, the tumor is removed and the frontal sinuses are opened. (*E*) Postoperative sagittal CT of the sinuses showing the bony defect from removal of the cribiform plate. CPR, resected cribiform plate; D, dura of anterior cranial fossa; IT, inner table of frontal sinus; LP, lamina papyracea; TS, tuberculum sellae.

where the middle turbinates are removed and posterior to the frontal sinuses, overlies the cribiform plate and its bleeding is controlled with bipolar electrocautery. To open the cribiform plate, first the bone lateral to it is opened using an irrigating drill and Kerrison rongeurs from the frontal sinuses to the planum sphenoidale. Once the lateral dura is exposed, the bony removal proceeds medially as the midline anterior cranial fossa dura tends to descend inferiorly into the operative field and can be violated inadvertently when the bone-dural plane is not initially established laterally. The crista galli is then identified and drilled to eggshell thickness for removal if necessary. In some cases, the anterior and posterior ethmoidal arteries need to be ligated for tumor removal and can be identified at the junction of the sphenoid sinus and the posterior ethmoid sinuses.

Transclival Approach

The transclival approach is used for lesions involving or requiring removal of the superior one-third of the clivus. Like the transtubercular/transplanum approach, it is an extension of the sellar approach (Fig. 11). Opening is bound laterally by the petrous carotid arteries anteriorly and the sixth cranial nerve superolaterally at the dural opening. The approach allows access to the interpeduncular cistern, third cranial nerve, upper basilar artery, and the posterior cerebral and superior cerebellar arteries (see Fig. 11B). It allows access to lesions such as retroinfundibular retrochiasmatic craniopharyngiomas but requires removal of the posterior clinoids and opening of the inferior intercavernous sinus (IIS). This requires using a 1-mm diamond irrigating endoscopic drill to remove the posterior clinoid bone that is bordered by the carotid arteries laterally and the sixth nerve posteriorly. Once the sellar dorsum opening and the clival bone with the posterior clinoids are removed, the dura can be opened giving a direct view of the retrosellar interpeduncular space. Dural bleeding during this exposure is controlled by opening the sellar basal dura and by using angled bipolar electrocautery in the midline (away from the sixth nerve). The IIS and clival dural are coagulated in a caudal direction before opening with angled endoscopic scissors. Bony bleeding during the extradural component of the exposure is controlled with venous bleeding hemostatic techniques mentioned earlier.

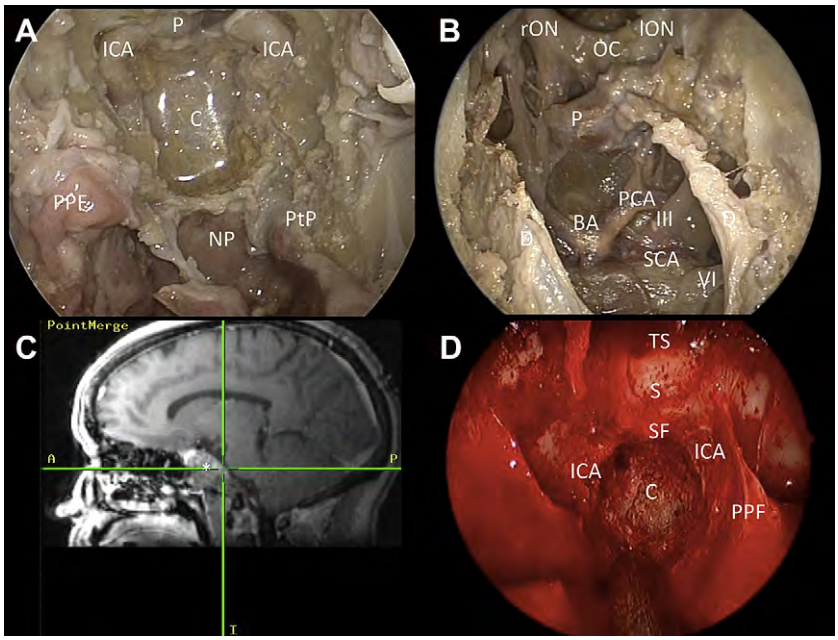

Fig. 11. Transclival approach. (*A*) Cadaveric dissection from planum to foramen magnum. (*B*) Dural opening from tuberculum sellae to base of clivus revealing ventral posterior fossa structures and parasellar structures. (*C*) Neuronavigation image guidance to back of clivus during removal of clival chordoma seen in (*D*). (*D*) Drilled out clivus just below sellar floor with instrument tip corresponding to cross-hatch seen with neuronavigation in (*C*) confirming the posterior wall of the tumor cavity with ventral pontine clival dura seen at base of cavity. BA, basilar artery; C, clivus; CD, clival dura; ICA, petrous internal carotid artery; lON, left optic nerve; NP, nasopharynx; OC, optic chiasm; P, pituitary; PCA, posterior cerebral artery; PPF, pterygopalatine fossa; PtP, pterygoid process; rON, right optic nerve; SCA, superior cerebral artery; III, cranial nerve III; VI, sixth cranial nerve.

For lesions involving the mid or lower clivus, a complete exposure of the clivus is necessary and the medial margin of the petrous carotid arteries needs to be identified. To do this, the vomer and floor of the sphenoid sinus have to be completely removed. The medial pterygoid plates and pterygoid wedges are identified bilaterally just lateral to the vomer-sphenoid junction. Then the sphenopalatine artery is identified and dissected posteriorly to its foramen. The foramen is opened using the irrigating drill exposing the pterygoid canal (Fig. 12A, B). The vidian nerve and artery are identified coming out of the canal. The vidian artery and nerve travel back to the junction of the horizontal and vertical internal carotid artery (ICA) segments at the anterior genus and the medial extent of the petrous carotid at the petroclival junction can also be identified. Drilling medial and inferior to the vidian artery and nerve along their canal allows the surgeon to safely expose the petrous carotid artery and define the lateral bony opening of the clival exposure. Inferiorly, the bony and soft tissue dissection is limited by the eustachian tubes bilaterally and the hypoglossal canals at the craniovertebral junction. Once the critical lateral landmarks are identified, the clival bone is removed using an irrigating endoscopic drill until the clival dura and basilar venous plexus are exposed. Clival bone bleeding is easily controlled with venous hemostatic agents. Once the dura is exposed, the dura is opened in the midline and dural bleeding and dural plexus bleeding, which can be profuse, is controlled with bipolar electrocautery and venous hemostatic agents. Dural opening laterally under the horizontal ICA extends to the Rosenmuller fossa on either side and opening laterally above

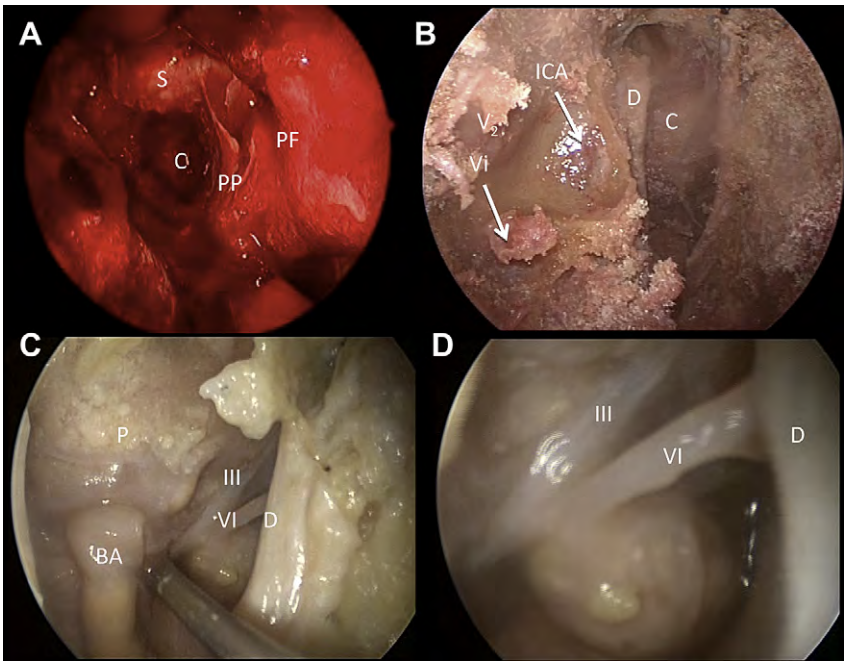

Fig. 12. Limitations to lateral exposure with the transclival approach: the petrous carotid artery and abducens nerve. (*A*) Intraoperative view for resection of a clival chondrosarcoma with bony resection of the clivus (C). The sellar floor (S) is visualized as the rostral extent of bony decompression. Laterally the pterygopalatine fossa (PF) and pterygoid process (PP) are visualized allowing for identification of the vidian nerve and artery. Once identified, the vidian artery is followed to the petrous carotid artery and allows for a corridor to be created exposing the lateral sphenoid sinus wall. (*B*) Cadaveric dissection with removal of the right middle turbinate and medial maxillary sinus wall. The posterior wall of the maxillary sinus is also the anterior wall to the pterygopalatine fossa where the vidian artery, vidian nerve, pterygopalatine ganglion with the infraorbital and maxillary nerves, and the maxillary artery can be found. It is removed to expose the posterior and lateral walls of the pterygopalatine fossa. In this approach, laterally the maxillary nerve (V_2) is visualized and can be skeletonized posteriorly to foramen rotundum. Opening the foramen rotundum then allows access to the middle cranial fossa. Posterior to the sphenopalatine artery (excised) the vidian artery (Vi) has been skeletonized from the pterygoid canal. Further removal of bone from 4 to 8 o'clock under the vessel leads to the anterior genu of the petrous internal carotid artery (ICA). Once identified, the ICA can be skeletonized and then mobilized laterally exposing the petrous apex. The clivus and sella are seen deep to the pterygopalatine exposure. (*C*, *D*) Opening of the clival dura (D) to expose the ventral posterior fossa brainstem is limited laterally by the abducens nerve (VI) where it runs by the superolateral margin of the dura next to the dorsal meningeal artery to enter the ventral dura en route to the Dorello canal and the cavernous sinus. BA, basilar artery; III, oculomotor nerve.

the anteromedial ICA genu can extend to where the sixth nerve enters the dura at the Dorello canal bilaterally (see Fig. 12C, D).

Transcavernous Approach

The transcavernous approach is an extension of the sellar approach and allows medial and lateral cavernous sinus exploration. Once the sellar approach bone removal is complete, the bone over the carotid protuberances is removed by proceeding laterally using a diamond burr, irrigating endoscopic drill and 1- to 2-mm Kerrison rongeur in an extradural manner. Venous bleeding, which is common during this bony decompression, is easily managed with warm irrigation and thrombin-soaked gel foam powder. Medial access to the cavernous sinus is easily performed via a trans-sellar approach with opening of the medial cavernous sinus wall. Many tumors create this opening for you, but in the event no medial cavernous sinus defect exists, a safe corridor between the anteriorly positioned carotid artery and posteriorly positioned posterior clinoid can be accessed (Fig. 13). This is best visualized with a 30° or 45° endoscope. Lateral cavernous sinus dissection has a higher risk for diplopia or facial hypesthesia and thus is avoided unless the pathology cannot be effectively treated via other modalities such as radiosurgery.

Tumor Resection

Endoscopic endonasal removal of skull base lesions follows the same general neurosurgical principles applied for removal of these lesions via an open approach. After an adequate exposure is accomplished to visualize and manipulate adjacent neurovascular structures, tumor capsule is coagulated and sharply opened. A frozen section is then obtained if the tissue diagnosis is in question. If the result comes back consistent with a pathology whose outcome is not helped with maximal resection or is better managed via other therapies with lower risk, then we proceed to close. If the pathology's long-term outcome is positively affected by maximal resection, then tumor resection proceeds. Using endoscopic malleable suction, endoscopic scissors, endoscopic ultrasonic aspirators, pituitary rongeurs, and other endoscopic instruments, the tumors are internally debulked. This helps create more room for dissection with less need for neurovascular retraction. As the space becomes

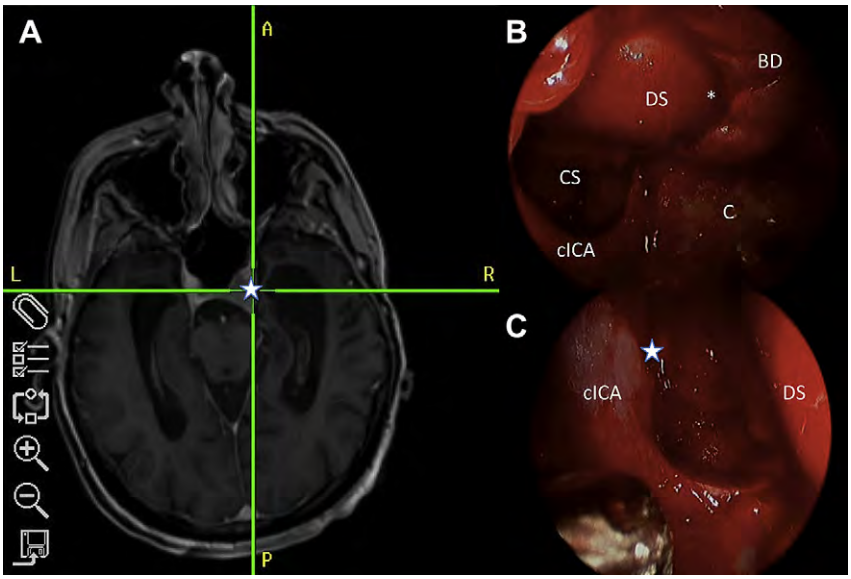

Fig. 13. Medial cavernous sinus approach. (*A*) Intraoperative neuronavigation image showing a right cavernous sinus mass approach via a transellar medial cavernous approach. (*B*) Sphenoidal view using a 0° endoscope showing a right medial cavernous sinus approach to the mass. BD, basal sellar dura; DS, diaphragma sella; cICA, cavernous internal carotid artery; C, clivus; CS, medial cavernous sinus wall defect for entry into cavernous sinus. *, midline. (*C*) Sellar-medial cavernous view using a 30° endoscope showing the medial cavernous sinus defect, cavernous internal carotid artery and looking back toward the top of the petrous apex. ★, location of intraoperative neuronavigation probe tip.

decompressed, the capsule often softens and allows for easier extracapsular dissection. The tumor capsule is then mobilized and is coagulated with bipolar electrocautery. Using endoscopic bipolar and sharp dissection, the tumor is devascularized and freed from adjacent normal tissues (Fig. 14A–C). When the plane between tumor and normal tissue is defined, micropatties are placed against the normal structures to prevent injury to them with further dissection and tumor manipulation (see Fig. 14B, D). Once the lesion is completely removed, the patties are meticulously removed with warm irrigation and the cavity is inspected for bleeding. Absolute hemostasis is obtained and defect reconstruction is then performed.

Skull Base Reconstruction

One of the greatest limitations of endoscopic skull base surgery in the past has been the inability to adequately repair osteodural defects created from these approaches resulting in persistent CSF fistula and/or infection. Advancements in the past decade, however, allow these procedures to be performed with a low risk of postoperative CSF leak or intracranial infection. Depending on the size of the defect and the vascularity of the surrounding tissues, there are several approaches for repair. Obese patients, patients with previous surgery or radiation to the resection bed, and patients with large dural defects are at the highest risk of postoperative CSF leak. In such cases, we often use a vascularized nasomucosal flap obtained during initial paranasal sinus exposure.

In cases where no dural violation occurs and no CSF drainage is seen, resorbable thrombin-soaked gel foam and fibrin glue are used to create a barrier between the dura and paranasal sinus flora. No splinting or other packing is used. For cases in which a small dural arachnoid opening or diaphragmatic opening is seen with spinal fluid drainage, a small onlay graft with DuraGen is used

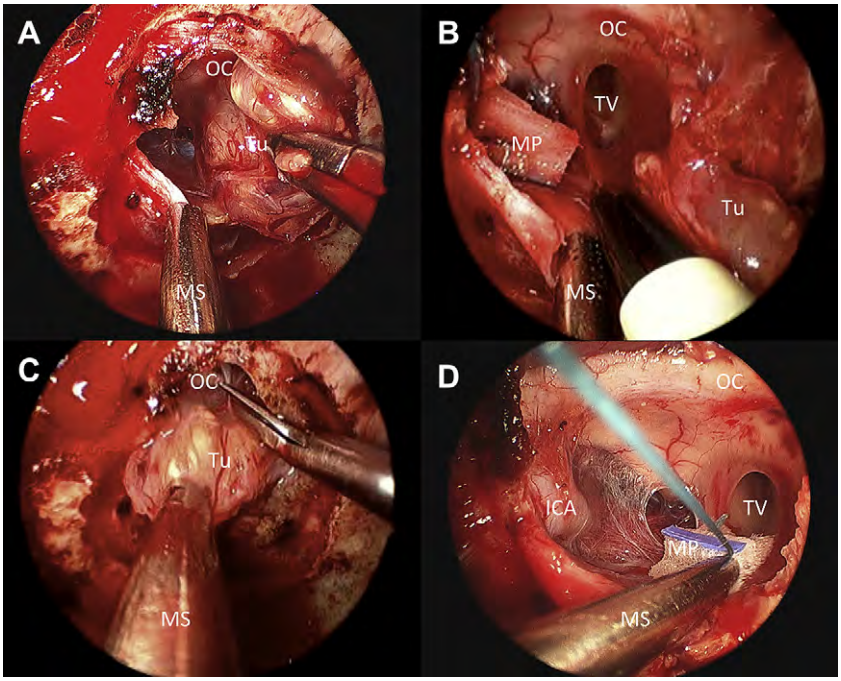

Fig. 14. Techniques of tumor dissection. (*A*) Using the same principles of open intracranial microsurgery, tumors are removed using extracapsular dissection whenever possible. The tumor (Tu) is being dissected away from the optic chiasm (OC) using bimanual technique. Just like with open microsurgery, endoscopic dissection is performed in a manner to remove the mass without having to retract, pull, compress, or coagulate the adjacent and often-adherent critical neurovascular structures. The endoscopic malleable suction (MS) is a great instrument for soft tissue traction and dissection. (*B*) During endoscopic bipolar coagulation, delicate adjacent structures are protected with micropatties, the coagulation settings are kept low, and the bipolar site is irrigated to minimize thermal damage to adjacent structures and sticking of the bipolar to the coagulated tissue. (*C*) Extracapsular dissection requires sharp dissection with endoscopic microscissors and bipolar electrocautery to minimize neurovascular tissue avulsion that can result in hemorrhage, nerve injury, or stroke. (*D*) Suction is applied to the field to improve visualization, but delicate neurovascular structures are protected by micropatties to prevent trauma by the suction tip. ICA, internal carotid artery; MP, micropatty; TV, third ventricle.

extending across the defect with good contact over intact dura or bone (Fig. 15B). This is then covered with a thin layer of fibrin glue and the associated sinus is packed with gel foam and an additional thin layer of fibrin glue. For larger dural openings, as can be seen with suprasellar adenomas with diaphragmatic invasion, that are still less than 5 mm in greatest dimension, we still generally do not use allograft or autograft materials. An inlay graft with DuraGen is placed through the defect (see Fig. 15A) followed by an onlay DuraGen graft covering and contacting the intact dural margins and/or bone. DuraSeal is then placed to cover the onlay graft and keep it in place (see Fig. 15E). DuraSeal is a synthetic absorbable sealant composed of 2 solutions, a polyethylene glycol ester solution and a trilysine amine solution. When mixed together, the precursors cross link to form the hydrogel sealant, which is absorbed in approximately 4 to 8 weeks, allowing sufficient time for healing. The sinus is then packed with gelfoam and a thin layer of fibrin glue is placed superficially to help seal off the space (Fig. 15F, G). No splints or other packing is used. In cases in which a larger dural defect is anticipated, a vascularized nasomucosal flap is then harvested at the beginning of the procedure (see Fig. 15C). This mucosal flap is harvested from the nasal septum and its pedicle is supplied from the posterior nasal artery. In patients who have had previous transphenoidal surgeries or patients in whom a large wide sphenoidotomy is performed before considering a flap, this pedicle is often destroyed and the vascularized flap cannot be harvested. When anticipated and harvested, the flap is placed in the nasopharynx or into the maxillary sinus via a medial antrostromy to prevent flap injury and maximize access to the pertinent anatomy and pathology behind the pedicle's source. If a nasomucosal flap cannot be harvested, we often use AlloDerm as a substitute, but with less success (see Fig. 15D). With large dural defects (>5 mm), the reconstruction begins with an inlay DuraGen

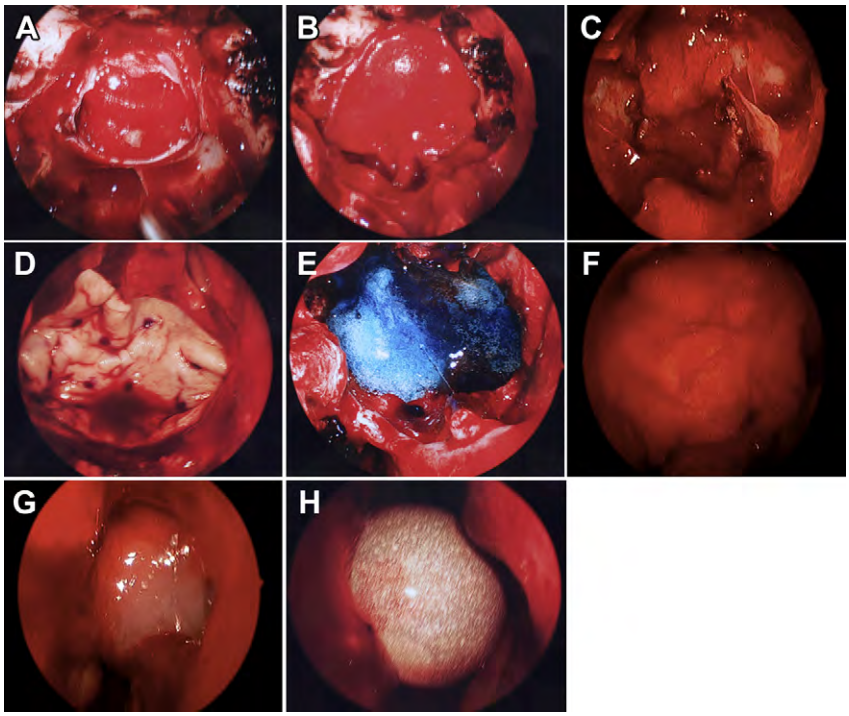

Fig. 15. Methods to reconstruct dural skull base defects created during endoscopic endonasal skull base approaches. (*A*) Dural inlay with DuraGen collagen matrix graft. (*B*) Dural onlay with DuraGen collagen matrix graft for small dural defects or cases with an intact diaphragma sella. (*C*) Dural onlay with a vascularized nasomucosal autograft flap for large dural defects. (*D*) Dural onlay with AlloDerm regenerative acellular human tissue matrix allograft for large dural defects in which a vascularized nasomucosal flap cannot be harvested. (*E*) DuraSeal hydrogel dural sealant is applied on top of the dural onlay graft to hold the graft against the patient's dura and bone adjacent to the skull base defect. (*F*) Obliterated sinus with multiple small pieces of thrombin-soaked compressed gel foam. The sinus is stripped of its mucosa before application to prevent delayed mucocele formation. (*G*) Application of Tisseel fibrin glue sealant (Baxter AG, Vienna, Austria) over the packed sinus to help prevent dislodging of gel foam and seal area from CSF leakage. (*H*) Positioning of a 12-French 5-mL nasal Foley catheter with the balloon filled with 5 mL of saline. The balloon is inflated once positioned more than the graft construct. This is used in cases where a large dural defect is created or when there is a high clinical suspicion that the graft will migrate or not easily adhere to the adjacent tissues. An example would be a patient with previous radiation to the area or previous CSF leak after an endonasal approach.

graft. If a large space then exists between the inlay and where an onlay vascularized graft would sit, then an abdominal fat graft is placed over the inlay graft and is then covered with the nasomucosal flap (or AlloDerm when no flap can be harvested) so that the onlay flap contacts all bony margins. This is then sealed in place with DuraSeal and then the sinus space is obliterated with gelfoam and further sealed with fibrin glue. A 12-French nasal Foley catheter is then placed over the reconstruction and the balloon is filled with 5 to 7 mL of saline so that the balloon conforms to the bony defect and holds the graft over the vascular bony margins to enhance adherence (see Fig. 15H). The catheter is then removed in 4 to 7 days.

Postoperative care

Postoperative imaging is obtained in all patients within 24 hours of surgery to confirm the degree of resection, evaluate reconstruction, and to look for evidence of complications such as hemorrhage, stroke, or tension pneumocephalus. We encourage patients to keep the nasal mucosa moist by using saline nasal spray 4 to 5 times per day. Patients are told not to blow their nose, use straws or engage in activities that induce a Valsalva-type response. Patients are followed in the office every 2 to 4 weeks until the wound appears well healed for routine sinus care including removal of crusting and disruption of any synechiae that form via office nasal endoscopy. When patients describe symptoms suggestive of a CSF leak, direct inspection for graft migration is performed in the office and any drainage is sent for a β_2 transferrin. If obvious copious CSF drainage is occurring and graft migration can be visualized, then the laboratory β_2 transferrin testing is deferred and the patient is brought back to the OR for re-exploration and revision. Once the reconstruction appears healed, the patient is allowed to resume regular activities. A 3-month imaging study is also obtained to serve as a new postsurgical baseline for future comparison. At this point most postoperative inflammatory changes have resolved, graft incorporation should have occurred, and postoperative bed debris should have absorbed.

Summary

In the past decade, tremendous strides in endoscopic instrumentation, visualization, and technique have made endoscopic endonasal surgery much safer and technically feasible for many pathologies of the skull base and paranasal sinuses. Using a graded step-wise progression in modular approaches to the skull base, surgeons can gain confidence and proficiency with the endoscopic endonasal approach. Progressive levels build on the previous levels in a logical manner that many surgeons can understand and add to their armamentarium of ways to treat skull base pathologies as their comfort levels with previous levels increase. Using current image-guided endoscopic endonasal techniques, surgeons can now access the skull base from the frontal sinuses all the way down to base of the dens and laterally to the internal auditory canal, jugular foramen, eustachian tubes, and mid-occipital condyles. With this approach cosmesis is improved, no brain retraction is required, lesions can often be reached faster than with open approaches, and less neurovascular manipulation occurs compared with traditional open approaches. As a result, more areas of the skull base can be reached and more pathologies can be successfully treated.

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Frontobasilar Fractures: Contemporary Management

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Approximately 5% of patients presenting to a level 1 trauma center in the United States will have sustained basilar skull injuries. The etiology of frontobasilar trauma is variable but is often associated with severe multisystem trauma caused by the significant force necessary to disrupt the thick frontal bone. Nahum demonstrated in an experimental model that forces of 800 to 2200 pounds are required to fracture the frontal bone, a force more than 2 to 3 times that required to fracture any other facial bone. Neurologic injuries are therefore common in this population of patients. Cerebral contusion is generally the most common radiographic finding; however, subdural and epidural hematomas requiring operative intervention are present in as many as 10% of the patients. Open brain trauma, globe injury, cerebrospinal fluid (CSF) leak, frontal sinus disruption, frontonasal fractures, and frontotemporal-orbital fractures are common manifestations of frontobasilar trauma, and may require multidisciplinary management. Thus, contemporary management of frontobasilar trauma in the United States should occur at designated level I or II trauma centers, whenever possible, due to the availability in these centers of maxillofacial surgeons, neurosurgeons, and ophthalmologists.

Classification

Several classification schemes have been advocated by various investigators in an attempt to describe injury patterns for frontobasilar fractures and to provide a clinically relevant decision-making tool. While a few are fairly descriptive of the injury components based on intraoperative and radiographic examination, most are too complicated to be clinically useful. The authors' preferred classification scheme is that described by Raveh in 1992, which is based on the treatment of 355 patients with severe skull base and frontal bone involvement (Fig. 1). In this classification the frontal, maxillary, and ethmoidal sinuses, the orbital cavity, and the nasal buttress may be regarded as shock absorbers, injury to which results in 2 broad injury categories: Type 1, which consists of frontonaso-ethmoidal and medial orbital frame fractures without skull base involvement; and Type 2, which consists of combined skull base, frontonaso-ethmoidal, and medial orbital frame fractures with frequent optic nerve compression. In Type 1 fractures, the external facial frame and buttresses give way and absorb and neutralize the impact, thereby preserving the posterior wall of the frontal sinuses, the frontal skull base, and the optic nerve canal. These injuries are not characterized by significant brain injury or dural laceration, and are generally managed by reduction of the external frame fractures and nasoorbital-ethmoidal component, if present, generally without the need for neurosurgical involvement. Type 2 fractures, on the other hand, are caused by high-energy forces that are not absorbed and neutralized by the external frame, and result in severe disruption of not only the external frame but also the internal palisades and buttresses. Type 2 fractures are characterized by intracranial dislocation of the posterior frontal sinus wall, inward telescoping of the nasal

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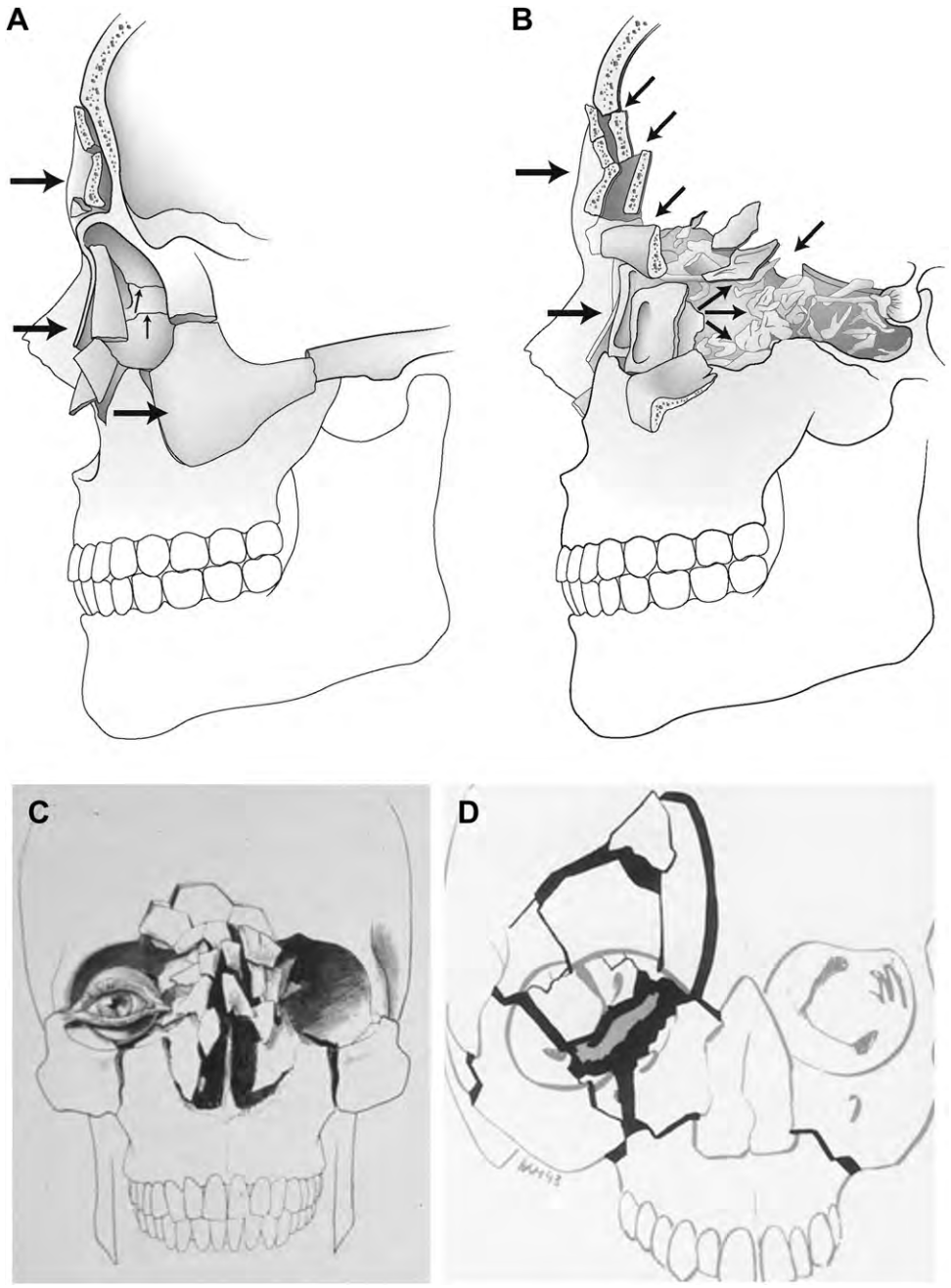

Fig. 1. Classification of frontobasilar injuries. (*A*) Type I, frontonaso-ethmoidal and medial orbital frame fractures without skull base involvement. (*B*) Type II, combined skull base, frontonaso-ethmoidal, and medial orbital frame fractures with frequent optic nerve compression. (*From* Bell RB. Management of frontal sinus fractures. Oral Maxillofacial Surg Clin N Am 2009;21:227–42; with permission). (*C*) Type IIa (central injuries), with or without anophthalmia, typically involve the frontonasal region and nasoorbital-ethmoidal complex, and virtually always involve the frontal sinus, with or without obstruction of the nasofrontal duct. (*From* Raveh J, Laedrach K, Vuillemin R, et al. Management of combined frontal-nasoorbital/skull base fractures and telecanthus in 355 cases. Arch Otolaryngol Head Neck Surg 1992;118:605; with permission). (*D*) Type IIb (lateral), or temporal-frontoorbital injuries, are not necessarily associated with nasofrontal duct obstruction or nasoorbital-ethmoid fractures, but may involve portions of the anterior or posterior tables of the frontal sinus, the orbital roof, and severely comminuted and displaced orbital "blow-in fractures" that result in optic neuropathy or other cranial neuropathies. (Used by permission *from* Orbital Fractures by Hammer, ISBN 0-88937-139-3 and ISBN 3-8017-0785-2, p. 68 © 1995, Hogrefe & Huber Publishers.)

pyramid, and intracranial dislocations along the anterior skull base planes including the orbital roof and sphenoidal-parasellar areas. Significant neurologic injury is common, including dural tears, CSF leak, brain tissue herniation, intracranial hematoma, and optic nerve compression. Treatment often includes a subcranial or transcranial approach to remove (cranialize) the frontal sinus, repair underlying dural lacerations and, if indicated, decompress the optic nerve. Most Type 2 injuries require interdisciplinary cooperation between a maxillofacial surgeon and a neurosurgeon, and are in fact the subject of this article.

Both Type 1 and Type 2 fractures can also be classified into subgroups based on the patterns of injury and the vector of force applied: either central or lateral. Central injuries typically involve the frontonasal region and nasoorbital-ethmoidal complex, and virtually always involve the frontal sinus, with or without obstruction of the nasofrontal duct (see Fig. 1C). Severe orbital or globe injuries are less common. Lateral, or "temporal-frontoorbital" fractures, on the other hand, are not necessarily associated with nasofrontal duct obstruction or nasoorbital-ethmoid fractures, but may involve portions of the anterior or posterior tables of the frontal sinus, the orbital roof, and severely comminuted and displaced orbital "blow-in fractures" that result in optic neuropathy or other cranial neuropathies (see Fig. 1D, E). Either subgroup may involve in its constellation of injuries an unsalvageable globe and anophthalmic socket that requires reconstruction prior to prosthetic globe replacement.

Epidemiology

Review of the trauma registry at the authors' trauma center uncovered 735 patients with basilar skull fractures during a 10-year period (Box 1). Most of these patients were male (598:169) with a mean age of 33.8 (age range, 1−91 years). All patients sustained significant, often multisystem injuries, primarily from motor vehicle collisions, and had a mean Injury Severity Score of 22.2 (range, 14−64). Concomitant midfacial fractures were present in 84% of the patients, cranial neuropathies in 24%, and CSF leaks in 4.6%.

Diagnostic studies

High-speed helical computed tomography (CT) with 1- to 3-mm cuts is the imaging study of choice for diagnosing and planning treatment for basilar skull fractures. Axial cuts alone are inadequate for thoroughly interrogating frontobasilar fractures, particularly when evaluating for bony disruption at the orbital apex. It is essential to obtain and evaluate coronal and sagittal views as well. Sagittal reconstructions from axial data require less than 3-mm slice thickness to avoid stepladder

Box 1. Trauma registry at the authors' trauma center

Patients: n = 735
Male: n = 598
Female: n = 169
Mean age: 33.8 years (range, 1−91 years)
Mean Injury Severity Score: 22.2 (range, 14−64)
Group 1
  No cerebrospinal fluid (CSF) leak: n = 701 (95.4)
Group 2
  CSF leak: n = 34 (4.6%)
    Rhinorrhea: n = 9 (26.5%)
    Otorrhea: n = 25 (73.5%)
  Persistent CSF leak: n = 6 (0.8%)

Data from Bell RB, Dierks EJ, Homer L, Potter BE. Management of cerebrospinal fluid leak associated with craniomaxillofacial trauma. J Oral Maxillofac Surg 2004;62:676−84.

artifact. The routine use of plain films is no longer advocated. Three-dimensional (3D) reconstructions are routinely performed to assess the injury from all angles.

Goals of treatment and reconstruction

There are a wide variety of philosophies, protocols, and procedures used in the treatment of frontobasilar injuries, each with the goal to provide an esthetic outcome, restore function, and prevent complications. These goals may be accomplished by adherence to the following sequential principles: (1) approach skull and facial skeleton via coronal incision; (2) repair of intracranial injuries, including evacuation of hematoma, debridement of nonviable brain, and neurologic decompression when necessary; (3) optic nerve decompression and/or globe repair when necessary; (4) creation of a "safe frontal sinus" through cranialization of the sinus and removal of all mucosa extending into the frontal recess; (5) repair of the frontal bandeaux and/or orbital roof; (6) dural repair and prevention of CSF leak by separating of the neurocranium from the nasopharynx with vascularized tissue (eg, pericranial flap, microvascular free flap) into the skull base and nasofrontal recess; (7) reestablishment of the facial projection and symmetry through reduction, stabilization, and fixation of the frontal bone, craniofacial buttresses, external orbit, and nasoorbital-ethmoidal complex (if involved); and (8) restoration of internal orbital volume. Various approaches, techniques, and materials are used to achieve these goals and there is no universal acceptance as to which approach, technique, or material is best in all instances.

Approach

The neurocranium and facial skeleton is approached via a coronal incision designed in sinusoidal fashion to enhance esthetics (Fig. 2A). Care must be taken to place the incision well into the hairline for individuals who are experiencing alopecia or male-pattern baldness. A preauricular extension is only necessary if the zygomatic arch is to be exposed. Otherwise the temporal extent should be completed just above or slightly behind the root of the helix.

Dissection of the coronal incision is performed in a subgaleal plane and care is taken to develop a robust, anteriorly based pericranial flap whenever possible, to be used for lining the anterior skull base or plugging the nasofrontal duct (Fig. 2B). Based on the supraorbital and supratrochlear arteries located in the galea-frontalis muscle layer, the pericranial flap can be developed as a single flap, or 2 laterally based flaps may be divided in the middle no less than 1 cm from the supraorbital rim.

The nasofrontal region and lateral orbits are approached with a subperiosteal dissection beginning 2 to 3 cm superior to the supraorbital rim. If necessary, the supraorbital foramina are osteotomized to release the supraorbital nerves (Fig. 2C). The orbits can be skeletonized almost circumferentially, but care should be taken to avoid stripping the medial canthal tendons (Fig. 2D).

If necessary, the zygomatic arch may be approached through a plane of dissection just superficial to the temporalis fascia (Fig. 2E). A 45° incision is made through the temporalis fascia 2 to 4 cm superior to the zygomatic arch, and sharp dissection proceeds between the superficial and deep layer of the temporalis fascia to the zygoma, which is then skeletonized (Fig. 2F). Exposure of the arch and any fractured segments begins first by exposing the anterior arch through the dense, frontotemporal fibrous connective tissue, then the root of the zygoma posteriorly, followed by the mid portion, which is approached inferomedially, to avoid injury to the facial nerve. The temporal branch of the facial nerve courses from 0.5 cm below the tragus to 1.5 cm above the lateral eyebrow and is located superficial to the zygomatic arch 0.8 to 3.5 cm anterior to the external auditory canal (average, 2 cm).

Neurologic decompression/repair of intracranial injuries

The treatment of severe head injuries associated with blunt or penetrating trauma has evolved over the last century from Cushing's recommendation of aggressive brain debridement and watertight dural closure in World War I to minimal brain debridement instituted by Israeli physicians during the Lebanon War of 1982, resulting in improved long-term seizure outcomes. Today, operative

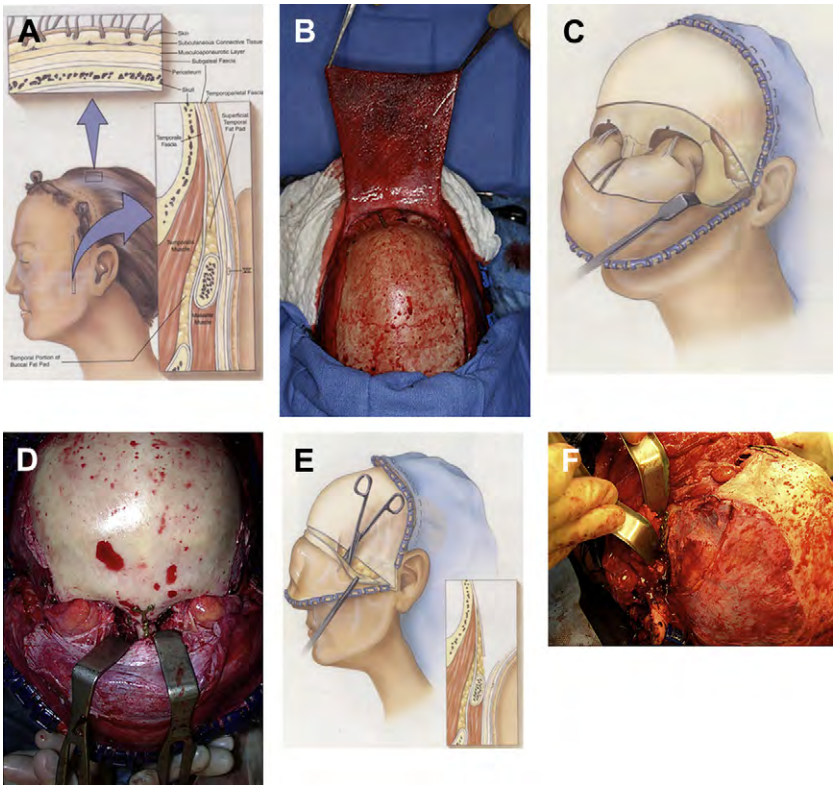

Fig. 2. Coronal incision. (*A*) Sinusoidal incision design and layers of the scalp above and below the superior temporal line. (*B*) Clinical case demonstrating subgaleal dissection and pericranial flap. (*C*) The nasofrontal region and lateral orbits are approached with a subperiosteal dissection beginning 2 to 3 cm superior to the supraorbital rim. If necessary, the supraorbital foramina are osteotomized to release the supraorbital nerves. (*D*) Clinical case demonstrating how the orbits can be skeletonized almost circumferentially, but care should be taken to avoid stripping the medial canthal tendons (note wide medial exposure). (*E*) The zygomatic arch may be approached through a plane of dissection just superficial to the temporalis fascia. A 45° incision is made through the temporalis fascia 2 to 4 cm superior to the zygomatic arch, and sharp dissection proceeds between the superficial and deep layer of the temporalis fascia to the zygoma, which is then skeletonized. (*F*) Clinical case demonstrating dissection of the zygomatic arch and body. The superficial layer of the deep temporal fascia and periosteum is retracted inferiorly and anteriorly. (*A*), (*C*), (*E*) (*From* Ellis E, Zide MF. Surgical approaches to the facial skeleton. Philadelphia: Lippincott Williams & Wilkins; 2005. p. 66, 80, 86; with permission.)

decision-making in a civilian setting is influenced by several clinical and logistical factors, but is often guided by invasive intracranial pressure (ICP) monitoring.

Multimodal brain monitoring has come to the fore as a means to optimize brain ICP, chemistry, and oxygenation in the face of the injury. One of the primary modes of monitoring and treating ICP is with the use of the ventriculostomy. In patients who undergo the repair of frontobasal injuries, the authors routinely place a ventriculostomy, which may be inserted by the neurosurgeon just anterior to the coronal suture in the midpupillary line on the right side. This may be tunneled out of a burr hole in the posterior aspect of the frontal bone flap and out of the skin posteriorly. The ventriculostomy may be used to monitor ICP via a transducer. Furthermore, the ventriculostomy is used to divert CSF to aid in the healing of the pericranial flap and to decrease the chance of a CSF leak. This catheter is connected to a drainage bag that is left open to drain about 5 to 10 cm above the level of the external auditory canal. Typically, this catheter may be removed after 3 to 5 days of drainage.

Frontobasal fractures are frequently associated with concomitant underlying injuries to the dura and the brain tissue. It is not uncommon in severe injuries to visualize brain tissue and CSF emanating from the site of the frontal bone implosion. The wide bicoronal incision discussed earlier allows ready access and exposure to areas of depression. If the zygomas do not need to be skeletonized to facilitate facial fracture repair, then the coronal incision can be developed as a myocutaneous flap by incorporating the temporalis muscle into the flap. When elevating the frontal bone, the authors perform a wide craniotomy to encompass the area of depression, allowing them to strip the normal

dura away from the bone and approach the area of depression, thus minimizing the possibility of pushing bone fragments or debris further into the brain. Particular care is taken if the injury is bilateral and if the frontal bone is removed in a bifrontal manner. The sagittal sinus is in the midline and may be torn. Even though this is far anterior, there may be profuse bleeding from the torn sinus, which can be ligated with surgical clips or sutures if necessary to control bleeding so long as the surgeon is in the anterior one-third of the sinus. This procedure is done frequently when bifrontal craniectomies are performed for intractable increased ICP.

The bifrontal craniotomy is performed by placing burr holes into the pterion, root of the zygoma just below the superior temporal line, and straddling the superior sagittal sinus (Fig. 3). Bilateral frontal and subtemporal osteotomies are performed, exposing the frontal and anterior temporal lobes. Additional removal of the squamous portion of the temporal bone and greater wing of the sphenoid bone is accomplished with a rongeur if access to the middle cranial fossa is required.

Contusions in the frontal and subfrontal regions are not uncommon with a frontobasal injury/fracture (Fig. 4). The natural history of these contusions is coalescence, and increase in size and surrounding edema, thus leading to increases in ICP. The authors debride these contusions, as they are associated with bone fragments and contaminants from the open wound. These contusions are frequently large and contribute significantly to ICP, and are removed even in the absence of an open depressed frontal fracture (see Fig. 4C). Careful hemostasis is achieved with bipolar coagulation

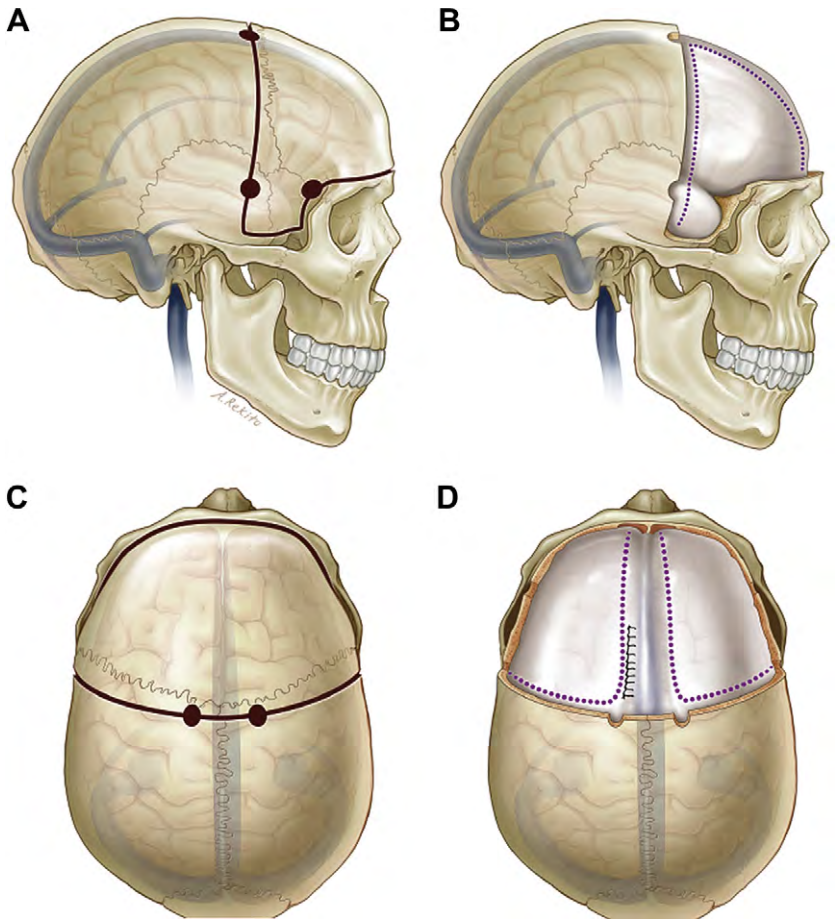

Fig. 3. Bifrontal craniotomy for access to anterior skull base and neurologic repair/decompression. The bifrontal craniotomy is performed by placing burr holes into the pterion, root of the zygoma just below the superior temporal line (*A*), and straddling the superior sagittal sinus (*B*). Bilateral frontal and subtemporal osteotomies are performed, exposing the frontal and anterior temporal lobes (*C, D*). Additional removal of the squamous portion of the temporal bone and greater wing of the sphenoid bone is accomplished with a rongeur if access to the middle cranial fossa is required to access, repair, or decompress neurologic injury. (*From* Ragel BT, Klimo P, Martin JE, et al. Wartime decompressive craniectomy: technique and lessons learned. Neurosurg Focus 2010;28(5):E2; with permission.)

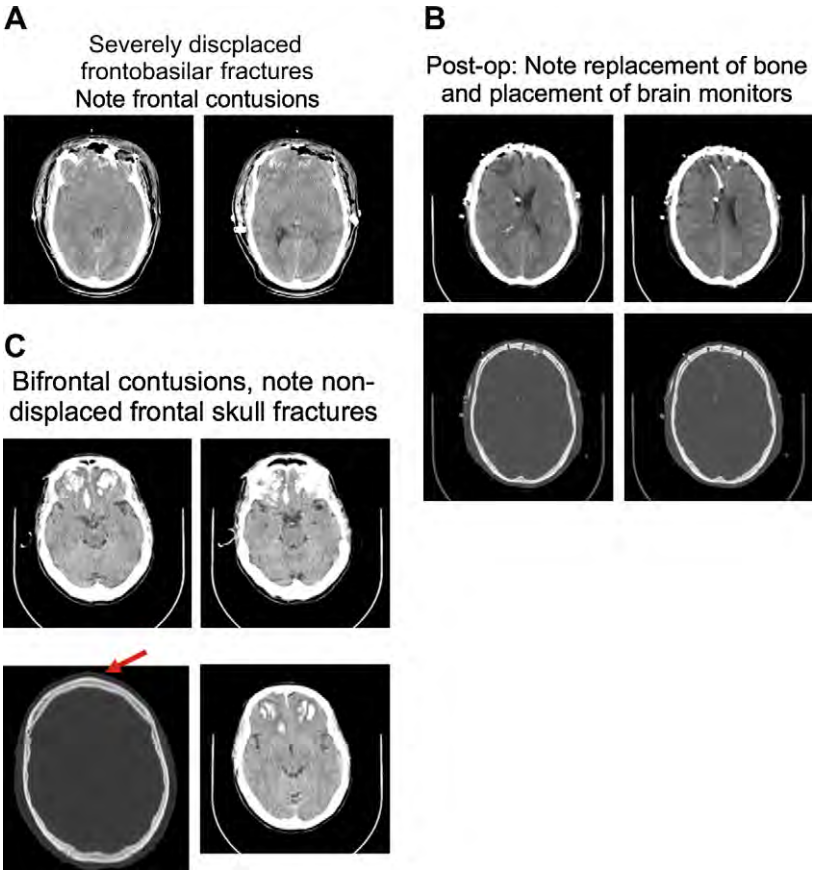

Fig. 4. Brain contusions associated with frontobasilar skull fractures. (*A*) Bifrontal contusions associated with severely displaced fractures. (*B*) Patient shown in A following decompressive craniectomy, ventriculostomy, and subsequent replacement of bone flap. Note placement of brain monitors. (*C*) Bifrontal contusions associated with nondisplaced fractures.

and careful monitoring of the intraoperative prothrombin times, activated partial thromboplastin times, and platelet counts; this decreases the risk of rebleeding and the subsequent increase in ICP.

Surgery for frontobasal injuries is directed not only at the reconstruction of the bone and dural defects as already described but also at control of ICP. If the injury is severe and there is significant cerebral edema that is not reduced by debridement of the frontal lobes and associated blood clots (ie, intraparenchymal contusions, subdural hematoma, epidural hematoma), the bone flaps may not be replaced immediately. The so-called decompressive craniectomy has evolved to become the procedure of choice for penetrating head injury and recalcitrant ICP elevation in most military centers and in many civilian centers throughout the United States. Frontotemporo-parietal or bifrontal decompressive craniectomies may be necessary to preserve brain function or facilitate transfer to a higher level of care (Fig. 5). Decompressive craniectomies allow for control of the cerebral swelling and ICP; however, one must be cognizant about potential early and late complications associated with these craniectomies. When this is necessary, delayed cranioplasty will be indicated following primary repair of the concomitant facial fractures. A detailed description of cranioplasty is beyond the scope of this article.

If there is a concern for brain swelling or cerebral edema, patients are monitored in the intensive care unit with multimodal monitoring, including cerebral microdialysis, intraparenchymal brain oxygenation monitors, and cerebral blood flow monitors. These relatively new technologies provide additional information that may guide the neurosurgeon to reexplore/redebride the areas of concern or to administer mannitol or hypertonic saline to help with ICP control. These new monitoring tools have not been demonstrated to affect the outcome. The authors use these tools judiciously and primarily in patients in whom a neurologic examination is not possible because they remain intubated and sedated, and where there is concern about cerebral edema and elevated ICP. As a result of the more reliable

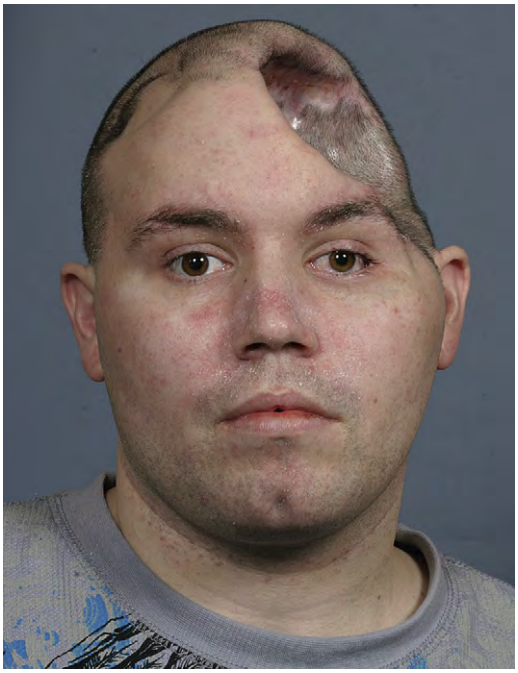

Fig. 5. A patient demonstrating craniofacial deformity following decompressive craniectomy before cranioplasty.

neurologic examination of the patient or if the monitors indicate normal ICP and brain chemistry (cerebral microdialysis), the monitors may be removed by pulling the catheters out at the bedside.

Repair of Globe Injury/Optic Nerve Decompression

Data from the authors' level I trauma center suggest that the orbit is involved in 47% of the facial fractures admitted by the trauma service as activated level I or II injuries. The majority of eye injuries consist of corneal abrasion, hyphema, and globe rupture. Compressive neuropathy, superior orbital fissure syndrome, and orbital apex syndrome are rare but functionally and esthetically devastating injuries.

The preponderance of scientific evidence suggests that progressive visual loss, optic canal fracture, retrobulbar hematoma with increased intraocular pressure, or perineural edema warrants urgent surgical exploration and/or decompression. In one study by Rajiniganth and colleagues, 70% of patients with visual deterioration and CT evidence of compressive neuropathy responded favorably to surgical intervention, if the operation was performed within the first 7 days after injury. The success rate decreased to 24% when decompression was performed more than 7 days after the injury.

If there is radiographic evidence of compression at the orbital apex/optic canal, then this area is surgically disimpacted and reduced in an attempt to maximize the potential for vision recovery (Fig. 6). Decompression of the optic nerve canal following traumatic optic neuropathy can be achieved subcranially either by an intranasal transethmoidal transphenoidal route via a Lynch approach or an endoscopic transethmoidal approach, or transcranially via a bifrontal approach. Transnasal and/or transorbital endoscopic surgery is the preferred option for patients with displacement of the optic canal and delayed visual deterioration. In the authors' experience, however, open surgery is often necessary and indeed facilitated by transcranial neurosurgical repair of concomitant intracranial injuries. Surgical techniques for optic nerve decompression are beyond the scope of this article.

It is important to coordinate all disciplines in a trauma center so that all treatment options, both medical and surgical, are available. In general, the authors' approach at Legacy Emanuel Medical Center is to employ high-dose steroids immediately for patients with failing vision and to consider surgical decompression for the following indications: (1) decreasing vision despite steroids; (2) visual loss with fracture compressing nerve; (3) increasing edema of nerve or hematoma with decreasing vision; (4) no light perception with fracture of optic canal; (5) CT evidence of optic nerve compression in the comatose patient (Box 2 and Box 3).

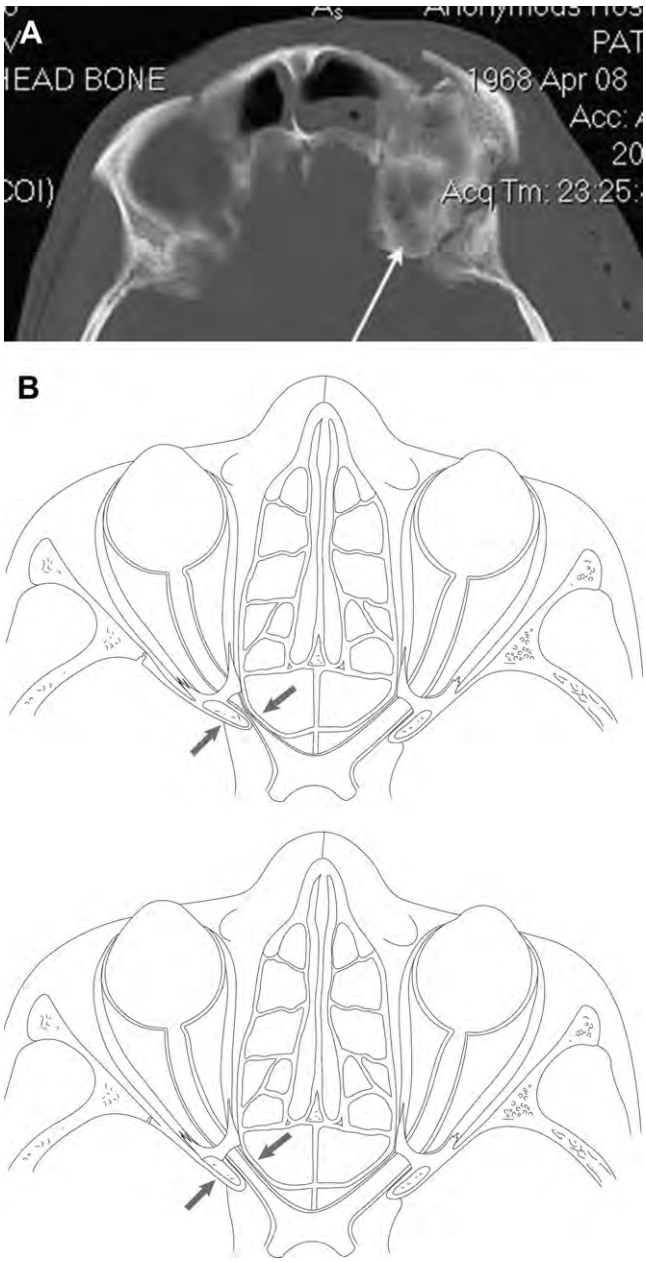

Fig. 6. Orbital blow-in fracture associated with type IIb injury. (*A*) Axial CT image of patient with a type IIb frontobasilar injury associated with orbital blow-in fracture and compression of the optic nerve (*arrow*). Such injuries should disimpacted, reduced, and stabilized. Associated globe rupture should be managed by the ophthalmology team as soon as possible. (*B*) Major relationships of the optic canal to the sphenoid sinus and sphenoid bone.

Repair of globe rupture is performed immediately. Enucleation is reserved for patients with a nonsalvageable globe rupture and is usually performed within 24 hours of the injury to prevent sympathetic ophthalmia.

Repair of the Frontal Bandeaux/Orbital Roof

Lateral temporal-frontoorbital fractures are reduced first, so as to provide a stable base on which to reestablish facial projection and orbital symmetry, and decompress the orbital contents. Minimally displaced fractures that will not result in a cosmetic deficit are not treated. Linearly displaced temporal-frontoorbital fractures may occasionally be reduced with a simple “pop up”; however, ORIF

Box 2. Sequential goals of treatment

1. Coronal approach
2. Bifrontal craniotomy (when necessary) for neurologic decompression, hematoma evacuation, repair of intracranial injuries
3. Repair globe injury/optic nerve decompression
4. Repair of the frontal bandeaux/orbital roof
5. Cranialization of frontal sinus
6. Prevention of CSF leak
7. Open reduction and internal fixation (ORIF) craniofacial buttresses, external orbital frame
8. Restore internal orbital volume

with titanium or biodegradable plates and screws is necessary for significantly displaced and comminuted fractures.

Orbital blow-in fractures are a distinct entity, occasionally associated with temporal-frontoorbital fractures (see Fig. 6). The distinguishing feature is the displacement of fractured segments into the orbit, which results in decreased orbital volume, elevated intraorbital pressure and, occasionally, compression or impingement of the orbital apex. Immediate posttraumatic exophthalmos is the commonest clinical manifestation, and the incidence of associated trauma to the optic globe, nerves, and extraocular muscles is high. These injuries are potentially reversible by prompt and early orbital decompression. Fractures are disimpacted and reduced under direct inspection and the orbit is decompressed. The orbital rim and walls are then immediately reconstructed in one stage and skeletal defects, when present, are primarily bone grafted.

Once the frontolateral components are repositioned and the orbital apex decompressed, the orbital roof is repaired, if necessary. The later can be done either with autogenous bone harvested from the parietal skull, or by using a commercially available titanium plate. The author prefers calvarial bone if the basilar skull defect is large and extends into the medial orbit (Fig. 7). Consideration can also be given to performing bone graft reconstruction of the cribriform plate/fovea ethmoidalis, as necessary. However, if the orbital roof is comminuted and displaced, but there is no large missing component, titanium mesh is effective in reestablishing the orbital volume and preventing a pulsating globe (Fig. 8).

Once the frontotemporal elements are reduced, attention is then turned to the nasofrontal region or the central component, and the frontal sinus is cranialized.

Cranialization of frontal sinus

Frontal sinus cranialization is considered a workhorse operation for patients with severe head injuries that involve the frontal sinus or anterior skull base. The most important principles for successful cranialization include complete removal of the posterior wall of the frontal sinus, meticulous removal of all visible mucosa, removal of the inner cortex of the anterior sinus wall, and the permanent occlusion of the frontonasal duct. The frontal sinus is completely eliminated and the nasofrontal recess is separated from the neurocranium, precluding any potential mucosal regrowth from the nasal epithelium.

Box 3. Relative indications for surgical decompression of the optic nerve

1. Vision decreasing despite steroids
2. Visual loss with CT evidence of fracture compressing optic nerve
3. Increasing edema of nerve or hematoma with decreasing vision
4. No light perception with fracture of optic canal
5. CT evidence of optic nerve compression in comatose patient

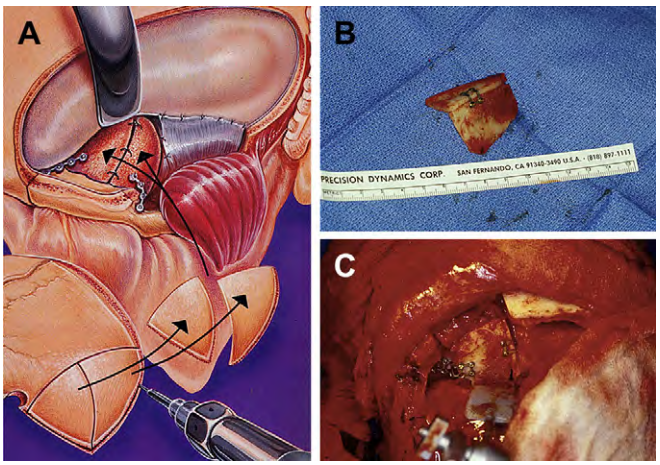

Fig. 7. Reconstruction of the orbital roof. (*A*) After reduction of the superior and lateral orbital margins, the roof and lateral wall of the orbit are reconstructed with autologous calvarial bone grafts harvested from the inner table of the frontal bone, or more posteriorly in the parietal region.*From* Rootman J. Orbital surgery: a conceptual approach. Philadelphia: Lippincott-Raven; 1995. p. 316; with permission. (*B*) Two pieces of bone are fixated to replace either the superior-lateral orbit (sphenoid component) or the superior-medial orbit (ethmoid component), or both if necessary. (*C*) Inset of the bone graft is facilitated by superior orbitotomy and retraction of the frontal lobes.

The cranialization procedure may be done with the assistance of a neurosurgeon, depending on the extent of underlying brain injury, the presence of dural lacerations, or the need for an extensive craniotomy. The procedure can also be combined with immediate reconstruction of the orbital roof, medial orbit, or nasoorbital-ethmoid fractures using autogenous bone grafts or alloplastic materials. Once the frontal bone is exposed and the anterior table removed, either by removing the fractured segments or by formal frontal craniotomy, the posterior table is completely removed and the adherent underlying dura or brain is exposed (see Fig. 8E). Care should be taken to maintain the integrity of the cribriform plate, if possible, and to avoid the sagittal sinus where the bone invaginates on either side. Occasionally, it is possible to preserve half of the frontal sinus if the intrasinus septum remains intact and the contralateral sinus remains functionally and anatomically inviolate. Once all the sinus mucosa and the posterior bony table have been removed, the nonviable bone, soft tissue, or damaged brain debrided, and dural lacerations repaired, the frontal bandeaux is restored by repositioning the anterior fragments and preparing a foundation for the restoration of nasal projection. Before replacing the bone flap or comminuted anterior skull segments, attention is turned toward prevention of CSF leak.

Prevention of CSF Leak

The dura is again inspected for CSF leak, repaired if possible, and patched if necessary. An anteriorly or laterally based pericranial flap is then rotated into the anterior cranial fossa defect and the nasofrontal recess to isolate the splachnocranium from the nasopharynx prior to wound closure (Fig. 9). The obstructed duct is then sealed with fibrin glue. The brain is allowed to expand into the extradural dead space, and the anterior table is reconstructed and stabilized with plates and screws.

ORIF Craniofacial Buttresses/Orbital Frame

Once the anterolateral skull component is reduced and the central skull base safely reconstructed, attention is turned to repairing the concomitant facial and orbital fractures. Gruss and colleagues recognized the importance of the zygomatic arch in complex midfacial fractures repair and correction of posttraumatic orbitozygomatic deformities. There is a reciprocal relationship between anteroposterior projection and facial width. As projection of the zygoma decreases, facial width increases. The zygoma, therefore, is key to restoring facial/orbital projection in severely displaced and comminuted fractures, and should be returned to its natural “flat” contour. The most common error in

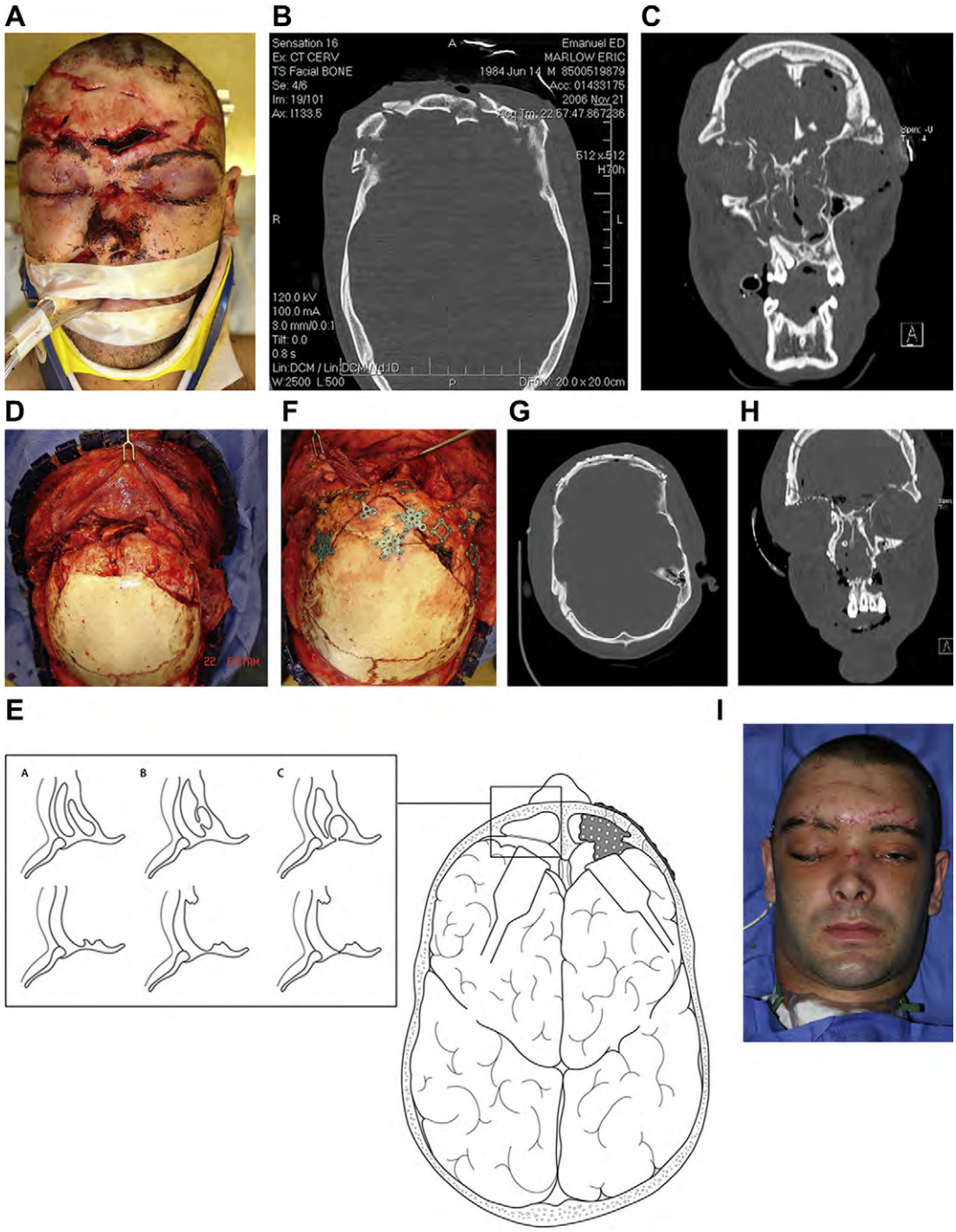

Fig. 8. Repair of the frontal bandeaux and cranialization of the frontal sinus in a 26-year-old man involved in motor vehicle collision. (*A*) Preoperative appearance. (*B*) Preoperative axial CT scan. (*C*) Preoperative coronal CT scan; note disruption of the orbital roof. (*D*) Bifrontal exposure. (*E*) The frontal lobes of the brain retracted, exposing the frontal sinus, which is being cranialized (the posterior wall removed), the frontal bandeaux repaired, and the orbital roof reconstructed with titanium mesh; also includes an inset/magnified view of the frontal sinus from the lateral view that demonstrates frontal sinus variation and removal of the posterior wall. (*F*) Frontal bone, lateral orbit, and nasoorbital-ethmoidal fractures reconstructed. (*G*) Postoperative axial CT image demonstrating favorable reduction, stabilization, and fixation of the frontal bandeaux and cranialization of the frontal sinus. (*H*) Postoperative coronal CT image demonstrating favorable restoration of orbital volume. (*I*) Postoperative appearance of patient 4 weeks after surgery; note superior orbital fissure syndrome.

reestablishing zygomatic projection is failure to reduce the segments out of a displaced "arch" into a flat zygoma, which will result in lack of malar projection and facial widening.

The lateral external orbital component is repositioned and stabilized with a mini plate at the zygomaticofrontal suture. Following this, proper projection of the zygoma is achieved by reducing

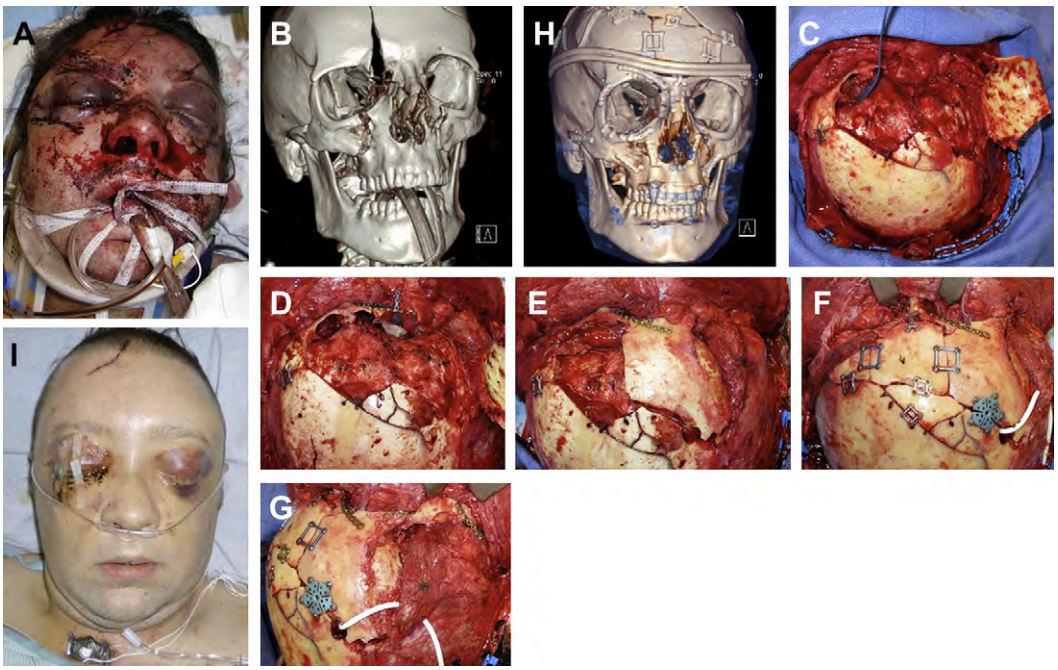

Fig. 9. A 24-year-old man with severe frontobasilar trauma associated with midfacial fractures at the Le Fort III level. (*A*) Preoperative appearance. (*B*) Preoperative 3D CT scan. (*C*) Frontotemporal craniotomy exposing the frontal and temporal lobes of the brain, frontal sinus, and right orbital roof. (*D*) Frontal bandeaux and lateral orbit are reduced, the frontal sinus cranialized, the superomedial orbital roof reconstructed with titanium orbital plate, and the dura repaired. (*E*) The skull base is lined with an anteriorly based pericranial flap and the frontotemporal bone flap is replaced. (*F*) The remainder of the central skull fragments are reconstructed along with the nasoorbital-ethmoidal component of the skeleton. (*G*) Facial projection and width are reestablished by reconstructing the zygoma and lateral-inferior orbit, and the remaining facial buttresses. (*H*) Postoperative 3D CT image. (*I*) Postoperative appearance of patient 1 week following repair.

and stabilizing the zygomatic arch. The maxillary buttress is then stabilized via a transoral approach and finally the infraorbital rim connects the lateral skeletal components to the medial nasoorbital-ethmoidal complex. Once the external orbital frame is reconstructed, attention is directed toward repairing the central skeletal components, if disrupted.

Severe telescoping or comminution of the nasal bones must be reduced, stabilized, and fixated to reestablish normal nasal projection. Although strut calvarial bone grafts have been advocated for this purpose and are occasionally necessary, the authors find that adequate anatomic reduction of the nasal bones with stable fixation is often all that is necessary as long as the frontal bandeau is adequately restored. Once the internal orbital frame has been reduced and stabilized with mini plates and nasal projection reestablished, attention must be directed to management of the central fragment. Medial canthopexy is facilitated using commercially available titanium barbed wire and titanium miniscrews placed in a posterior-superior position within the medial orbit behind the lacrimal crest.

Restoring the Internal Orbital Volume

Attention is then directed toward restoring normal orbital volume and globe position. This portion of the reconstruction may be delayed 7 to 10 days to allow for the resolution of edema, or alternatively may be performed primarily, depending on the clinical situation and surgeon's preference. The internal orbit is typically approached via a transconjunctival incision to access portions of the inferomedial orbit that cannot be reached via the coronal incision. The lower lid incision is made at the conjunctival fornix with a fine-needle–tipped Bovie and is carried through conjunctiva and orbital fat to the orbital floor as a postseptal approach. Herniated soft tissue is teased back into the orbit from the maxillary sinus or ethmoid cavities and stabilized in place with a malleable retractor. Attention is then directed toward reestablishing normal orbital anatomy with particular attention paid to restoring the contours of the critical ethmoidal and antral bulges. The authors' preference is to use

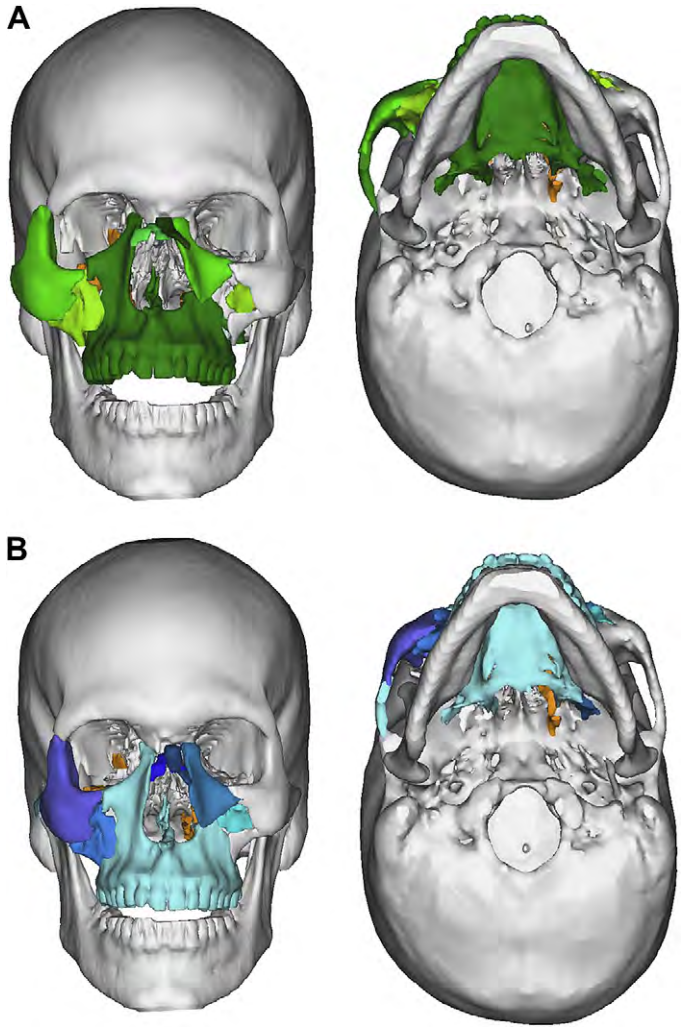

Fig. 10. Virtual reconstruction based on CT data. (*A*) 3D reconstruction of patient with midfacial fractures at the Le Fort III level. (*B*) Virtual reconstruction to achieve ideal facial projection, symmetry, and orbital volume. (*C*) Surgery is confirmed with intraoperative navigation and intraoperative CT scanning. (*D*) Portable CT scanner.

titanium orbital plates for most routine fractures. Alternatively, porous polyethylene (Medpor) with impregnated titanium may be used. Porous polyethylene without impregnated titanium is not advised because it is not radiographically apparent. Every effort is made to identify a stable posterior ledge within the orbit, and the critical ethmoidal and antral bulges are restored.

In an effort to improve predictability in orbital reconstruction, Metzger and colleagues described a semiautomatic procedure for individual placement of titanium meshes for orbital fractures. By using CT scan data, the topography of the orbital floor and wall structures is recalculated. After mirroring the unaffected side onto the affected side, the defect can be reconstructed virtually. Data for the individual virtual model of the orbital cavity are then sent to a template machine that reproduces the surface of the orbital floor and medial walls automatically. The titanium mesh can then be adjusted preoperatively for exact 3D reconstruction and an individually milled titanium implant is used with navigation to guarantee intraoperative placement of the preformed mesh. Bell and Markiewicz recently reported favorable restoration of orbital volume and external orbital contours using a combination of intraoperative navigation and stereolithographic models in complex orbital reconstruction. In the future, it is the authors' opinion that custom plates such as these placed under intraoperative guidance with intraoperative confirmation by CT scan will be the method of choice for repair of complex orbital injuries (Fig. 10).

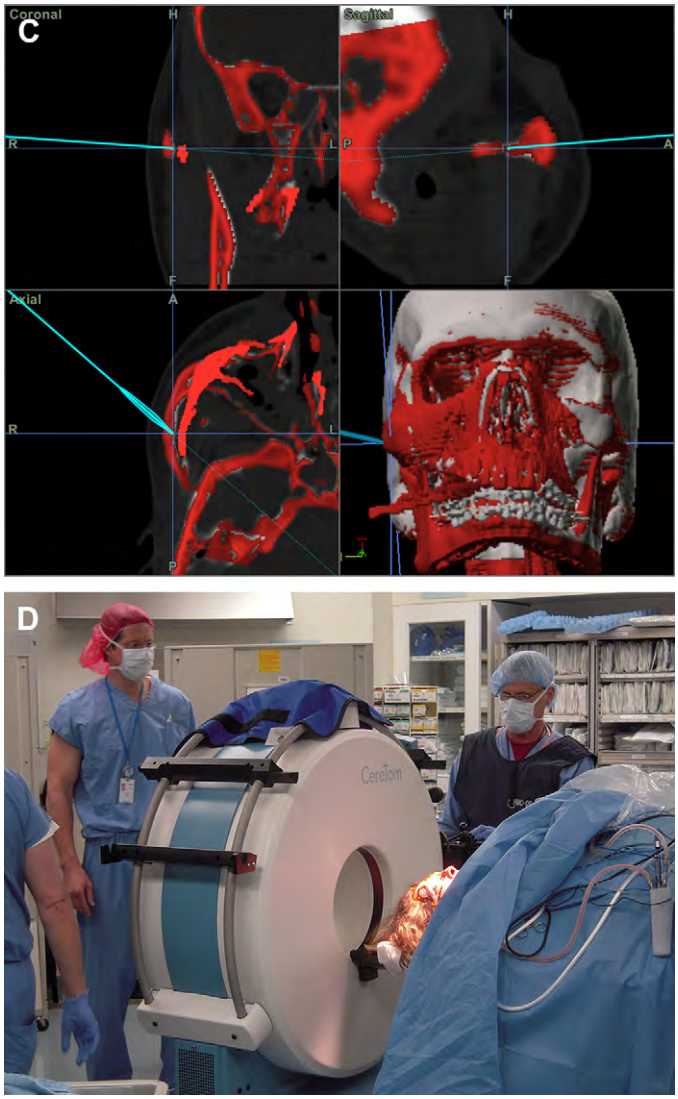

Fig. 10 (*continued*)

Postoperative care

The coronal incision is closed over a suction drain. The patient is admitted to the intensive care unit postoperatively and monitored closely for changes in vital signs or neurologic status. Prophylactic antibiotics are prescribed for 1 week postoperatively and strict sinus precautions (avoidance of Valsalva maneuvers, nose blowing, heavy lifting) are recommended. The patient is observed for signs and symptoms of infection, CSF leak, or intracranial bleeding.

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