Skull Base Reconstruction in the Pediatric Patient

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Abstract

Introduction Pediatric skull base and craniofacial reconstruction presents a unique challenge since the potential benefits of therapy must be balanced against the cumulative impact of multimodality treatment on craniofacial growth, donor-site morbidity, and the potential for serious psychosocial issues.

Objectives To suggest an algorithm for skull base reconstruction in children and adolescents after tumor resection.

Materials and Methods Comprehensive literature review and summary of our experience.

Results We advocate soft-tissue reconstruction as the primary technique, reserving bony flaps for definitive procedures in survivors who have reached skeletal maturity. Free soft-tissue transfer in microvascular technique is the mainstay for reconstruction of large, three-dimensional defects, involving more than one anatomic region of the skull base, as well as defects involving an irradiated field. However, to reduce total operative time, intraoperative blood loss, postoperative hospital stay, and donor-site morbidity, locoregional flaps are better be considered the flap of first choice for skull base reconstruction in children and adolescents, as long as the flap is large enough to cover the defect. Our “workhorse” for dural reconstruction is the double-layer fascia lata. Advances in endoscopic surgery, image guidance, alloplastic grafts, and biomaterials have increased the armamentarium for reconstruction of small and mid-sized defects.

Conclusions Skull base reconstruction using locoregional flaps or free flaps may be safely performed in pediatrics. Although the general principles of skull base reconstruction are applicable to nearly all patients, the unique demands of skull base surgery in pediatrics merit special attention. Multidisciplinary care in experienced centers is of utmost importance.

Keywords

- skull base surgery
- pediatric oncology
- microvascular
- free tissue reconstruction
- endoscopic
- skull base reconstruction

Introduction

Skull base tumors are rare and heterogeneous in the pediatric population. Malignant tumors comprise approximately half of the cases, of which about half are soft-tissue sarcomas.¹ The age distribution exhibits a bimodal pattern peaking at ages 3.5 and 17.5 years, with different pathologies afflicting each age

group. While skull base and craniofacial surgical resection and reconstruction are integral components in the management of many pediatric neoplasms, they pose a particular challenge in this age group. The potential benefits of therapy must be balanced against the cumulative impact on future craniofacial skeletal growth and soft-tissue changes, along with donor-site morbidity following harvesting tissue from a growing skeleton,

and the potentially significant psychosocial impact.² Unique anatomic considerations in children include smaller craniofacial complex, cranial fossa and paranasal sinuses, stage of tooth eruption, absence of anatomic landmarks such as the superior orbital fissure and mastoid pneumatization, and the fragility of neurovascular elements. Major concerns of skull base procedures in children are the potential impact of osteotomies on the subsequent development of the face and paranasal sinuses, and the challenges of performing microsurgery in children whose blood vessel diameters are small, anatomic landmarks are inconsistent, and growth of anastomosed vessels is anticipated. Since children are more likely to suffer from sarcomas and from tumors involving the middle cranial fossa compared with adults, surgical defects are often large and cause significant functional, aesthetic, and psychological issues. Members of a multidisciplinary team consisting of specialists in neurosurgery, head and neck surgery, plastic and reconstructive surgery, maxillofacial surgeons and prosthodontics, pediatric critical care, and pediatric oncology should work in conjunction with child life specialists. Technical developments in the fields of head and neck surgery and neurosurgery in the last few decades, allowed a growth in skull base reconstructive options. As a result of improvements in microvascular technique, endoscopic equipment, image guidance, and biomaterials, the choices in reconstructive technique have expanded,

allowing for more radical tumor extirpations, with major impact on the long-term survival of patients with skull base tumors. Although the general principles of various surgical approaches of skull base reconstruction following tumor ablation are feasible, applicable, and well established, the variety of approaches that have been developed over the years have been applied mainly to adult patients while unique issues remain to be established in the pediatric patients.³ Several retrospective studies have been conducted to shed light on the special considerations related to this age group, but the sample sizes were small, and data are limited.⁴ Because of the relative rarity and heterogeneity of skull base neoplasms, protocol-driven research has proven difficult. However, enough experience has accumulated to allow a new focus on the unique demands of skull base surgery in children. In this article, we aim to suggest an algorithm for skull base reconstruction in the pediatric population in light of the experience gained, and review of the literature (► Table 1).

Goals of Skull Base Reconstruction

The goals of skull base reconstruction in children are similar to those in adults. When tumors arising in the skull base invade both soft and bony tissues, tumor resection may create extensive skull base defects and produce a free

Table 1 Pediatric skull base reconstruction modalities

Type of defect	Reconstruction modality
Dural defects	Primary closure whenever possible
Minimal dural tear	Primary closure
Small dural defect	Temporalis fascia
Moderate to large dural defect	Double-layer fascia lata
EEA	Multi-layer fascia lata +/- NSF
Soft-tissue defects	
Orbital exenteration alone	Temporalis muscle flap, gracilis free flap (closed cavity) Skin graft or temporoparietal fascia flap (open cavity)
Bone defects	
Small or medium defects of the calvarium	Calvarial bone graft (age > 3 y)
Orbital wall resection	Coated titanium mesh, fascia lata sling, septal cartilage
Orbital wall, nasal, and frontal bones	Posterior frontal sinus wall graft, split calvarial bone graft, titanium mesh covered by pericranium
Medial maxillectomy and resection of lacrimal duct	DCR
Large defects involving more than one compartment of the skull base	
Primary reconstruction of large defects	Soft-tissue free flaps or bony free flaps
Orbitomaxillary resection	Soft tissue free flap (rectus abdominis, lateral thigh +/- obturator)
	Bony free flap (scapula, fibula)
Peri-/postoperative XRT	Pericranial wrapping of all osteotomized bony segments or titanium mesh, free flaps
Definitive procedures in survivors who have reached skeletal maturity	Vascularized bone flaps

Abbreviations: DCR, dacryorhinocystostomy; EEA, endoscopic endonasal approach; NSF, nasoseptal flap; XRT, radiotherapy; y, years.

conduit between the paranasal sinuses and the intracranial space. Following tumor extirpation, skull base cranial base defects require precise and durable reconstruction to (1) support the brain, the orbit, neural, and vascular structures; (2) form a watertight dural seal; (3) reestablish separation between the contaminated extracranial oral and sinonasal cavities and the sterile subdural compartment and intracranial contents; (4) prevent airflow into the intracranial space; (5) provide lining for the nasal cavity; (6) reestablish the nasal and oropharyngeal cavities and maintain a functional sinonasal system; (7) obliteration of dead space created by the ablative surgery; and (8) provide three-dimensional (3D) restoration for optimal function, contour, cosmetic outcome, and quality of life.⁵ Failure to create adequate reconstruction harbors significant complications, among them cerebrospinal fluid (CSF) leak, meningitis, brain herniation, and tension pneumocephalus.⁶ Children with skull base defects pose a special multidisciplinary challenge which requires adaptation of the surgical technique to minimize developmental complications and emotional toll.

Anatomic Considerations

The human skull grows rapidly during the first 4 years of life. The maximal growth rate at birth slopes down to a minimum in prepuberty and returns to a smaller second spike at puberty. The spheno-occipital synchondrosis, which is the main axis of skull base growth, fuses at the ages 12 to 15 years and ossifies only at early adulthood. Several differences exist between adults and children with regard to skull base morphology. The size of the cranial base and maxillofacial complex in children is smaller, the cranial bone is thinner, and the floor of the frontal and middle cranial fossa may be flatter.³ Neurovascular elements in children are thinner and more fragile than in adults.¹ Another difference is the inconsistency of specific anatomic landmarks in children compared with the fully developed cranium of adults. In children younger than 3 years, the mastoid tip may not be fully developed, so the facial nerve is more superficial and inferior than in adults. The mastoid air cells are not fully pneumatized in very young children, because the mastoid and middle fossa reach adult size after the age of 10 making the identification of the labyrinth more difficult.⁷ The pterion is located more anteriorly in children, and the supraorbital fissure or foramen, which serves as a landmark for orbital and orbitozygomatic approaches, may be absent before the age of 8 years.³ Another important anatomic discrepancy is the developmental stage of the paranasal sinuses: the frontal sinus starts to develop from the age of 6 years and reaches its full size at the end of puberty.⁷ In pediatric craniofacial surgery, it is essential to avoid disruption of the permanent dentition within the maxillary complex, because permanent tooth eruption does not take place before the age of 10 years. In terms of general skull base location, the pediatric skull base defects have higher proportion of pure middle area lesions (42%) and a lower rate of posterior lesions (8%) compared with adults.⁸

Preoperative Planning

The preoperative planning begins with acquiring high-resolution maxillofacial and skull base computed tomography (CT) images and dental models if necessary. The selection of the proper skull base reconstruction technique is dictated primarily by the size of the cranial defect based on radiologic and intraoperative calculations, prior radiation to the area, and the need for bone reconstruction.⁵ Additional variables which influence the reconstructive approach are the etiology of the defect, the anatomical location, presence of infection, intracranial pressure, prior surgeries, patient age and comorbidities, and surgeon experience.^{5,9} Anticipating the appropriate skull base deficit is imperative to successful reconstruction. Large defects measuring greater than 50% of the anterior skull base (ASB) often involve adjacent orbitomaxillary structures, thereby requiring significant tissue bulk. Conversely, smaller defects limited to the central region, with preservation of the orbital contents, can be reconstructed in a multitude of ways ranging from vascularized tissue to nonvascularized tissue and biomaterials.¹⁰ Bony reconstruction is recommended if greater than one-third of the orbital roof is lost; otherwise, the patient may experience pulsatile enophthalmus. In addition, loss of the supraorbital bar, frontal bone, and malar eminence necessitates bone replacement to achieve optimal aesthetic results. Patients who have undergone radiation therapy or who are likely to require postoperative radiation experience significant fibrosis and decreased vascularization. Nonvascularized grafts' reabsorption over time can be avoided by wrapping the graft in vascularized tissue.^{5,11} Alloplastic agents, such as porous high-density polyethylene (Medpor; Porex Surgical, Inc. Newnan, Georgia, United States), can be fabricated prior to surgery and can be customized to precisely match a cranial defect using thin slices or 3D CT imaging. The bony expected defects are analyzed in all planes. Volumetric and linear analysis of reduced segments is computed by virtual software to establish symmetry facial widths and heights.¹² The surface data generated can also be converted into a stereolithography model, which is computer-rendered and then 3D-printed. These processes follow well-established, computer-aided design and computer-aided design and manufacturing (CAD/CAM) protocols.¹³ The advantages of CAD/CAM include the ability to virtually manipulate skeletal segments and analyze the expected outcome to obtain the best possible result. Once the surgical plan is developed and objectively analyzed, it can then be translated into the operating room by computer-manufactured guides and models. If a vascularized pericranial flap is available, it can be used to cover the implant. However, alloplastic agents should not be used in an irradiated field owing to the high rate of extrusion.¹⁰ In cases of fibula-free flap reconstruction, CT angio of the lower extremity is used to plan accurate osteotomies on the fibula to recreate the shape and angles of the skull base or maxillofacial bony defect.¹² The development of surgical navigation systems provides real-time intraoperative guidance based on a preoperatively simulated treatment plan. The rapidly developing technologies for 3D

visualization, interactive localization, and point mapping enable the precise identification and preservation of critical structures. The incorporation of these simulation-guided navigation systems into the clinical routine improve the accuracy of craniofacial reconstructions and reproducibility of the surgical plan, and minimize the need for intraoperative improvisation and the risk of complications. The development of CAD/CAM for virtual surgical planning has enabled preoperative planning in three dimensions and manufacturing of guides and models for intraoperative use. These cutting guides can be used to make precise osteotomies based on the virtual plan. They can help avoid crucial underlying structures. Since the surgical plan is developed in a virtual environment, an unlimited number of iterations can be analyzed until the desired outcome is achieved. As the attempted repair becomes more difficult and the number of bone segments that require manipulation increases, CAD/CAM becomes increasingly beneficial for obtaining the best outcome. The combination of these tools can minimize the amount of time spent contouring bone intraoperatively to obtain the precise fit. The ability to virtually mirror one side improves symmetry, and the use of cutting and positioning guides enable maximal bony contact and minimize the need for bone grafting. The angles and distances of movement can be precisely calculated to correct vertical dystopias. Finally, multiple objective measures of accepted cephalometric points may be assessed within the virtual surgical environment for a controlled planned resection, thereby generating a precisely modeled result that can be predictably reproduced in the operating room.

Reconstruction

Nonvascularized Flaps

Dural Defects

CSF leak prevention is an essential component of the surgical success in skull base reconstruction as the risk of meningitis is substantial.¹⁴ Whenever possible, usually in limited small defects (< 1 cm), primary closure should be attempted or covered with a nonvascularized flap. Medium-sized low flow leaks can be managed by additional underlay temporalis fascia or fascia lata grafts. Local pedicled flaps or free flaps covering an osteocutaneous flap applied in a multilayered fashion are reserved for larger complex defects, with high flow leaks.^{15,16}

Autologous tissues, such as *fascia lata* and *temporalis fascia*, had been used in the past for skull base reconstruction, usually covered with a vascularized flap. In a previous publication, we described the healing process of the double-layer fascia lata graft and provided evidence that long-term viability of the fascia is achievable without an overlying vascularized flap.¹¹ A double-layer fascia lata is used in cases of extensive skull base defects. The fascia is tacked under the edges of the dura and carefully sutured in place. The dural repair is then covered with a second layer of fascia that is applied against the entire undersurface of the ethmoidal roof, the sella, and the sphenoidal area. Fibrin glue is used to provide additional protection against CSF leak. Following

dural repair, Vaseline gauze is applied below the dura and into the paranasal cavity to provide additional support against pulsation.

Nonautologous tissue, such as cadaveric fascia lata,¹¹ lyophilized dura, and cadaveric dermis (Alloderm; LifeCell Corp., Woodlands, Texas, United States),⁵ can be used for small or medium defects of the calvarium. A layered technique is often fashioned, along with fibrin sealant and supporting nasal packing materials.

Bone Defects

Small bony defects can be reconstructed with a variety of nonvascularized techniques. 3D titanium mesh can be used for small defects of the calvarium following removal of the frontal sinus outer table. *Autologous* bone, such as a *split calvarial bone graft* or a *posterior frontal sinus wall graft*, are used for reconstruction when the tumor involves the nasal bone, nasal septum, or an adjacent fronto-orbital segment, and are considered popular grafts in adults owing to ease of harvest, naturally rounded contour, accessibility within the surgical field, malleability for sizing and shaping, and regeneration of the donor site to a perfect surface.^{11,17} However, there is a minimum patient age for the performance of a split cranium graft.¹⁷ Harvesting split cranium graft can be a challenge in children younger than 3 years old because of the thinness and the underdeveloped diploic space of the skull. Another popular source of autologous bone involves split rib grafts. No minimum patient age is associated with the harvesting of rib; however, the procedure requires a second operative site and a longer operating time. The stiff angular grid formation of parallel ribs also makes rib bone unsuitable for use in most cranial defects. Bone yield from iliac crest and rib harvest is limited because of the patient's size and are difficult to contour aesthetically. If nonvascularized bone grafts are utilized in an irradiated field, vascularized tissue should be wrapped around the bone.^{5,11} Regardless of the source of autologous bone, grafts can be complicated by infection, exposure, fragmentation, growth restriction, and lifelong risks secondary to lack of incorporation, vascular ingrowth, and remodeling. Grafts may undergo partial absorption and leave an unpleasing and unsound surface that requires secondary revision. Infections with or without resorption of the native bone often complicate the repair.¹⁸ For all of these reasons, the usage of autologous bone grafts has never become standard practice despite its theoretical advantages.¹⁸ Whereas nonvascularized bone grafts successfully reconstruct small bony defects, titanium mesh is well suited for reconstructing larger areas requiring structural support.^{5,11} Desirable properties include lack of donor-site morbidity, malleability, biologic inertness, and magnetic resonance imaging (MRI) compatibility.^{5,11} The porosity of titanium mesh necessitates additional tissue such as a pericranial flap or radial forearm free flap (RFFF) to achieve a watertight seal.^{5,11} Commercially available alloplastic biomaterials and prefashioned implants for craniofacial reconstruction exist for addressing small and medium-sized calvarial defects and serve as an alternative to autologous nonvascularized bone grafts.⁵ *Porous high-*

density polyethylene (Medpor; Porex Surgical, Inc.) has been described as an ideal alloplastic material for cranioplasty. It produces no foreign body reaction, becomes well incorporated through fibrovascular bony ingrowth at its margins, and can be custom made from CT imaging to precisely fill the patient's cranial defect.¹⁸ Adequate soft tissue is a requirement before alloplastic implant cranioplasty. If soft-tissue coverage is inadequate, tissue expansion of the scalp and scalp regeneration with acellular dermal matrix are useful tools to optimize overlying soft tissue before cranioplasty, to prevent complications of implant extrusion and wound breakdown, and to improve outcomes.¹⁸ While porous polyethylene has been extensively and successfully used in adult craniofacial reconstruction, its use has been less well documented in the pediatric population.¹⁹ Lin et al¹⁸ reported that custom-made porous polyethylene implant cranioplasty can be used safely and successfully for pediatric large-scale calvarial defects, even in the setting of a previously infected, multiply operated wound bed. The potential advantages are immediate one-stage reconstruction, avoidance of donor-site complications, and excellent contour matching. However, the risks include restriction of normal calvarial growth, long-term compromise of overlying soft tissues, and possible increased incidence of infection. Furthermore, alloplastic agents should not be used in an irradiated field owing to the high rate of extrusion.¹⁰ A major concern important to bear in mind with the use of alloplastic materials in the pediatric population is the inability of the implant to grow and retain the appropriate contour with the growing skull, necessitating replacement of the implants with the skull and maxillofacial bones' complete maturation, reserving definitive reconstruction with vascularized bone until skeletal maturity is completed.

Vascularized Flaps

Local flaps, regional myocutaneous pedicled flaps, and free tissue transfer are reconstructive options with a reliable blood supply, which is not directly dependent on neovascularization.

Dural and Bone Defects

Local Flaps

The *pericranial flap* first described by Wolfe in 1978²⁰ remains the most frequently utilized local flap for small and medium central and paramedian defects.^{5,10} The pericranial flap encompasses the subaponeurotic connective tissue and underlying periosteum, and is based anteriorly and inferiorly from the supratrochlear and supraorbital arteries. It is readily available, easily harvested, and can be used to repair various dural defects, unilateral or bilateral defects of the cribriform plate, fovea ethmoidalis, planum sphenoidale, and lamina papyracea.⁵ For patients undergoing extirpation of malignant tumors and for whom adjuvant radiation therapy is planned, the nasofronto-orbital (NFO) segment should be wrapped with a pericranial flap or rotational temporoparietal fascia to prevent osteoradionecrosis.²¹ A pericranial flap is also used in cases of medial orbital wall reconstruction to cover the bone-graft segments or the titanium mesh. Alternatively, small-to-

moderate sized paramedian defects can be reconstructed with a de-epithelialized midline *forehead flap*,^{5,10} which is pedicled on the supratrochlear artery. Lateral ASB defects are often closed using the *temporoparietal fascia flap*. The temporoparietal fascia flap is supplied by the superficial temporal artery.⁵ It is often used for small lateral ASB defects, especially if orbit or malar contouring is required.⁵ Vascularized parietal bone may be concomitantly harvested with the flap.⁵ Disadvantages include the potential for alopecia, especially if electrocautery is used, and the risk to the temporal branch of the facial nerve. Alternatively, the *temporalis muscle flap* has proven useful in this setting, especially following orbital exenteration.²² The flap is supplied by the anterior and posterior deep temporal arteries that allows for the development of two separate paddles. Arc of rotations and cosmetic concerns are major disadvantages. Donor-site morbidities include damage to the temporal branch of the facial nerve and a concave deformity, resulting in not satisfactory cosmetic result.^{5,22} Local flaps are often insufficient in size for reliable restoration of extensive ASB and maxillofacial defects.¹¹

Endoscopic Reconstruction

Endoscopic expanded endonasal approaches (EEAs) to the ASB spare the morbidity of an open craniofacial resection and offer excellent visualization. Studies have shown that EEA to sellar lesions can be safely and successfully performed in young patients.²³ However, it is less clear whether the smaller facial structures of pediatric patients can accommodate reconstruction of surgical skull base defects with vascularized tissue flaps. Effective and consistent reconstruction following EEA requires complete separation of the cranial cavity from the sinonasal tract, obliteration of dead space, preservation of neurovascular and ocular function, and reconstruction of tissue barriers.²⁴ The most popular flap described for reconstruction in the setting of endoscopic ASB, trans-sellar, and transclival resection or CSF leak in adults is the neurovascular pedicled nasoseptal flap (NSF), a flap of nasal septum mucoperiosteum and mucoperichondrium based on the posterior septal branch of the sphenopalatine artery,^{24,25} avoiding separate external incision. A significant decrease in the incidence of postoperative CSF leaks following EEA surgery has been reported with the adoption of the NSF.²⁶ Although the use of NSF has been studied and proven adequate in adults,²⁷ little is known about its use in pediatric patients. Shah et al²⁴ used CT measurements to evaluate NSF size among pediatric patients. Their findings suggested that children younger than 6 to 7 had inadequate NSF length for reconstruction of sellar defects; the investigators also noted that NSF width did not appear to be a limiting factor in reconstruction. As opposed to Shah et al, Ghosh et al²⁷ reported that NSF provide sufficient and reliable coverage option in reconstruction of suprasellar defects in pediatric patients. They demonstrated that septal lengths in children less than 10 years of age were adequate to cover the defects created by the suprasellar resections. When compared with older patients, younger patients tend to have greater NSF length relative to the length of sellar defects. They concluded that preoperative radiographic assessment of NSF feasibility is critical to ensure adequate flap coverage for

suprasellar defects. Purcell et al²⁸ attempted to use a standardized measurement method to characterize the relationship between potential nasoseptal length and sellar defect length in children. This radiographic feasibility study suggested that NSF length is not a limitation in reconstruction of pediatric sellar defects. Due to pneumatization, the volume of the sphenoid sinus expands with age,²⁸ and this study found that older patients were more likely to have longer sellar defect lengths. Incomplete pneumatization remains an obstacle to performing endoscopic approaches to sellar lesions in very young children. There are case reports of successful endoscopic transsphenoidal procedures with NSF reconstruction in children under the age of 2 years.²⁸ Our experience with the NSF in a relatively large series of patients is discussed elsewhere in this supplement. With the use of image-guided navigation, it is becoming more feasible to perform EEA in the setting of incomplete pneumatization, but this remains an area for future investigation. Smaller defects involving the fovea ethmoidalis, planum sphenoidale, or sella can also be sealed endoscopically using the middle turbinate flap.²⁹ This pedicled flap is based on the middle turbinate branch of the sphenopalatine artery and is much smaller. Regardless of flap selection, endoscopic visualization of the entire defect is imperative to achieving a successful seal.

Soft Tissue and Complex Large Defects

Free Tissue Transfer

Advances in microvascular technique, enabled free tissue transfer to become the mainstay for reconstruction of complex 3D ASB defects with excellent surgical results and low complication rates.^{5,11,22} This technique provides adequate tissue bulk with a rich, reliable blood supply, allowing for improved healing, decreased hospitalization, and decreased complications including CSF leak and meningitis.⁵ Free tissue transfer is, however, a relatively complex surgical procedure that requires high technical qualifications. Another drawback of this method is the bulk of the muscular free flap,

which may mask local recurrence and make radiological follow-up more difficult.¹¹ The choice in free tissue donor site is dictated by defect size, location, need for bony restoration, and pedicle length.⁵ Frequently utilized flaps include the RFFF, rectus abdominis, and latissimus dorsi free flaps. Additional options include the fibula, anteriolateral thigh, gracilis muscle (►Figs. 1–3), scapula, lateral arm, ulna, serratus anterior, and superficial temporal fascia free flap.⁵ A temporal burr hole created during the craniotomy provides access to superficial temporal and facial vessels.⁵ When an orbital exenteration is performed, the pedicle can be tunneled through the cheek to reach the facial vessels at the mandibular border.⁵ Large ASB reconstruction often includes defects of the midface, maxilla, and orbit, thereby warranting bulky tissue to fill significant dead space.⁵ The rectus abdominis myocutaneous free flap and the latissimus dorsi free flap are ideal in this setting. Both flaps provide long vascular pedicles and large skin paddles that can be subdivided and inset in complex 3D orientations. The rectus abdominis is preferable for large ASB reconstructions⁵ since the majority of the tissue bulk provided is vascularized fat, which does not atrophy over time. Conversely, the latissimus dorsi free flap tissue bulk is muscle, which atrophies following denervation. The rectus abdominis free flap is based from the deep inferior epigastric artery that provides a large vessel diameter and wide distribution to overlying subcutaneous tissue.⁵ The anterior rectus sheath can be islanded off a perforator vessel, allowing for simultaneous dural closure and elimination of dead space with the muscular bulk.⁵ Common complications include abdominal wall hernia, wound dehiscence, and infection. A fibula free flap may continue to grow with the adjacent mandible or maxilla, but the growth of adjacent recipient tissues may be altered. Harvest of a fibula free flap before skeletal maturity may result in tibial overgrowth or a valgus deformity. The choice of local pedicled flaps versus free flaps in children is still a matter of debate. Some authors reserve pedicled flaps to very limited defects and prefer free flaps due to their versatility and reliability.⁹ Others found

Fig. 1 (A) Coronal and (B) axial gadolinium enhanced T1-weighted preoperative magnetic resonance imaging (MRI) of a 2-year-old child with right infratemporal fossa and masticatory space embryonal rhabdomyosarcoma, after chemoradiation therapy.

Fig. 2 Intraoperative images of a 2-year-old child with right infratemporal fossa and masticatory space embryonal rhabdomyosarcoma, after chemoradiation therapy. (A) A combined subcranial and transfacial approach was used for extirpation of this tumor, while preservation of the facial nerve (B) The deficit remained after tumor extirpation.

locoregional flaps preferable in children as they reduce operative time, postoperative hospitalization time and donor-site morbidity, and reserve free flaps only for large, complex defects.³⁰

Orbital Reconstruction

Reconstruction of the medial orbital wall is performed only in cases in which a total removal of this segment is necessary or if the periorbit is excised. In these situations, a split

calvarial bone graft, fascia lata sling, or 3D titanium mesh covered with pericranium can be used. Septal cartilage can be used for reconstruction of a limited defect of the inferior orbital wall. If eye globe exenteration is required, a split-thickness skin graft is used to cover the orbital socket. The orbital socket (open cavity) may be reconstructed also with the use of a small volume free flap, like the gracilis muscle (closed cavity). For a radical maxillectomy with or without orbital exenteration, a lateral thigh free flap or a rectus

Fig. 3 The reconstruction was performed with gracilis muscle free flap. (A) The specimen resected. (B) Gracilis muscle free flap harvest. (C, D) The patient at the end of the operation.

abdominis musculocutaneous free flap can be used to obliterate this large defect and to support the obturator from above. The bony component of the maxilla and orbital floor can also be reconstructed with the use of a scapula or fibula free flap.⁴ A dacryorhinocystostomy (DCR) is performed on all patients undergoing orbital wall resection or medial maxillectomy. Following tumor resection and reconstruction, a centripetal compression of both globes is performed to reduce the telecanthus.¹¹

Discussion

Successful skull base reconstruction requires a thorough understanding of the complex process of craniofacial development and of skull base anatomic considerations regarding the differences between children and adults. Although the general principles of skull base reconstruction are applicable to nearly all patients, the unique demands of skull base surgery in children and adolescents merit special attention, since the potential benefits of therapy must be balanced against the cumulative impact of multimodality treatment on craniofacial growth, donor-site morbidity, and the potential for serious psychosocial issues. Multidisciplinary care in centers experienced in the management of these complex tumors is of utmost importance. Given the psychological issues that children and adolescents with skull base malignancies face, experts in child psychology or psychiatry should be involved in these patients' care. Our experience in treating children with skull base tumors has helped us understand the considerable anatomic differences between children and adults, including the small size and fragility of neurovascular elements, the inconsistency of anatomic landmarks, and the need to avoid disruption of permanent dentition.^{1,31,32} Pediatric skull base tumors which require reconstruction after extirpation seem to arise irrespective of gender, are benign in half of cases, and are of various histological types.^{1,2,9,30,33} Because the pathological types of skull base tumors in children and adolescents vary, it can be difficult to predict the extent of a defect after extirpation and to plan a complicated reconstructive procedure, while considering the potential for subsequent growth. Although skull base surgery has not clearly demonstrated an adverse effect on the remaining craniofacial skeleton development in nonirradiated children,³³ nonoperative modalities such as radiation therapy are associated with significant negative consequences, such as retardation of facial bone and soft-tissue growth, short stature, visual or auditory dysfunction, dental abnormalities, and various endocrinopathies. With nearly half of pediatric patients with skull base tumors demonstrating some adverse effect of radiation, some groups have advocated more aggressive surgical resections to achieve a gross total resection in an attempt to avoid or delay radiation therapy.⁹ In general, pediatric skull base reconstruction follows the principles of adult skull base reconstruction, thus flap selection should be decided with the defect size, surgical stress, and donor-site morbidity. With the evolution of surgical extirpative approaches, refinements in the practice of free tissue transfer and the devel-

opment of new anesthetic and monitoring techniques, procedures once perceived to be too radical for the pediatric patient have become both safer and more reliable.³³ Free tissue transfer has been demonstrated to be the preferable technique in the reconstruction of large ASB defects in several reviews, as it promotes improved primary wound healing, restores function, and maintains esthetics, compared with regional or pedicled flaps.⁴ Reconstruction with free tissue transfer can minimize late sequelae of chemotherapy and radiation treatment for malignant tumors, such as facial hypoplasia, asymmetry, and postoperative retraction deformity.⁴ Free tissue transfer promises flexibility in flap content and design and provides the opportunity to introduce a large quantity of well-vascularized tissue to the reconstructed area in a single-stage operation. However, there are several concerns unique to pediatric patients. Pediatric microsurgery is challenging due to the small size of vascular structures, and the increased risk of donor-site long-term functional deficits. In addition to the inevitable compromise at the site of resection, esthetic defects at the donor site can have a major psychological impact on a child. Commonly used donor sites, such as the fibula, iliac, and scapula, possess epiphyseal growth centers that are central to skeletal growth and development, and soft-tissue architecture must be considered as well. Although it did not reach to the statistical difference, Yano et al³⁰ reported that using free flaps in pediatrics tended to have higher postoperative complication rate than using locoregional flaps compared with that of adult, thus advocated locoregional flaps as the flaps of first choice for skull base reconstruction if they are large enough to cover the defect. It is speculated that free flaps in pediatrics are possibly affected by preoperative chemoradiotherapy or negligible injury to the elevated flaps due to technical error during surgical procedure because a vascular pedicle and perforator of free flaps in pediatrics is rather small and narrow compared with the adult. Locoregional flaps, such as pericranial flaps and temporalis muscle flaps, have sufficient vascularity, area, and thickness to cover skull base defects, even in children and adolescents. These flaps also significantly reduce reconstructive time and postoperative hospital stay. Such flaps are also favorable from the standpoint of donor-site morbidity. Several previous reports^{9,30} have suggested that osteotomy and sacrifice of the pericranium have limited effects on postoperative craniofacial development. In a previous study, we reported that wide resection of skull base tumors decreases postoperative quality of life.³⁴ It can be speculated that decreasing donor-site morbidity in patients undergoing skull base reconstruction favorably affects postoperative quality of life, especially in growing children. Local flaps remain the mainstay for smaller defects. We use the pericranium for wrapping bony segments (i.e., NFO or split calvarial bone graft) to provide vascularization to these components in patients scheduled for postoperative radiation therapy, to decrease the risk of osteoradionecrosis²¹ since the pericranium can offer an excellent blood and nutrients supply to the osteotomized bone. Several authors believe that bony reconstruction of the skull base is necessary to support the newly

reconstructed skull base and to prevent herniation of the cranial content.¹¹ As we previously reported,¹¹ large cranial base defects and prior surgery and radiotherapy (previously considered indications for free flap reconstruction⁶) can be managed by fascia alone with excellent surgical results, and reconstruction with a double-layer fascia does not require a rigid support of bone or synthetic materials. Furthermore, this procedure does not require the high technical qualifications which are essential for free muscle transfer. The relatively low bulk of the fascia lata facilitates radiological follow-up in patients with an increased risk of local recurrence. The use of fascial flaps does not require the utilization of skin grafts or muscle flaps for covering the nasopharyngeal space. Moreover, the rapid healing process of the fascial reconstruction provides a robust physiological barrier between the nasopharynx and the intracranial space within days following surgery, an important advantage of its use. Free fascial grafts seem to survive via local proliferation of a newly formed vascular layer embedded within the fascial sheath¹¹ and are associated with low incidence and severity of perioperative complications compared with other reconstructive methods. Finally, the overall duration of the reconstructive procedure with double-layer fascia is not longer than that of a pericranial flap, and takes considerably less time than free tissue transfer procedures.¹¹ The double-layer fascia lata serves as the standard material for ASB reconstruction. The thin and low mass properties of the fascia lata enable the surgeon to cover large dural defects with a single fascial sheath. Furthermore, the flexibility of the fascia lata enables coating of extensive cranial defects, including parts of the orbit and paranasal sinuses. As we reported,¹¹ we used free flaps for reconstruction in cases which required ASB tumor resection and radical maxillectomy with or without orbital exenteration, for closed reconstruction of the orbit alone, in cases of recurrent tumors and in cases of skull base osteoradionecrosis. The rectus abdominis is the most commonly used free flap in this anatomical area, followed by the lateral thigh, radial forearm, and latissimus dorsi flaps. Bony reconstruction is our choice for orbital, nasal, and anterior frontal sinus wall reconstruction. This can be achieved with the use of biological tissue, such as split calvarial bone graft or posterior wall of frontal sinus, or with artificial materials, such as 3D titanium mesh. These materials are wrapped with a pericranial flap if postoperative radiation is planned. A host of nonautologous biomaterials are also used in the setting of small-to-moderate sized defects requiring rigid support. They should not be used in the setting of radiation owing to the high extrusion rate. Advances in endoscopic surgery, image guidance, alloplastic grafts, and biomaterials have increased the armamentarium for reconstruction of small and mid-sized defects. The advent of endoscopic tumor resection has introduced a variety of local flaps amenable to endoscopic reconstruction.

Conclusions

Comprehensive skull base reconstruction using locoregional flaps or free flaps is practical and appropriate, and may be

safely performed in children and adolescents. Because of the potentially synergistic effects of multimodality treatment for skull base malignancies on craniofacial growth and development, we advocate soft-tissue reconstruction as the primary technique, reserving bony flaps for definitive procedures in survivors who have reached skeletal maturity. Free soft-tissue transfer in microvascular technique is the mainstay for reconstruction of large, 3D defects, involving more than one anatomic region of the skull base, as well as defects involving an irradiated field. However, to reduce total operative time, intraoperative blood loss, postoperative hospital stay, and donor-site morbidity, locoregional flaps are better be considered the flap of first choice for skull base reconstruction in children and adolescents, as long as the flap is large enough to cover the defect. Our “workhorse” for dural reconstruction is the double-layer fascia lata, which provides a simple and versatile means of skull base reconstruction after resections of advanced tumors, without the need for blood supply from an overlying regional flap or free muscular flap. Future developments of biomedical materials and EEA surgery will probably bring with them continuing improvement of current techniques for skull base reconstruction with fewer complications and a better quality of life outcomes.

Note

This is an original manuscript reviewed by all authors and not under consideration for publication elsewhere.

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None.

Conflict of Interest

None.

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