Malignant tumors of the skull base

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Malignant tumors of the skull base have traditionally been difficult to control because of adjacent crucial anatomical structures and because of the inherent difficulty in accessing/resecting tumors and reconstructing surgical defects. Techniques in craniofacial surgery for malignant tumors of the skull base have advanced significantly since their first description. Advances in neuroimaging, surgical technique, perioperative care, and adjuvant treatment have contributed to improved results. Because the majority of malignant tumors requiring craniofacial resection involve the anterior skull base, this review focuses mainly on state-of-the-art surgical techniques as well as pertinent variations, complications and results.

Key Words • skull base tumor • paranasal sinus • reconstructive surgery • outcome

The skull base is the anatomical region that forms the interface between the cranial cavity and the structures of the rest of the head. Pathological conditions, especially malignant tumors, have traditionally been difficult to control because of the crucial anatomical structures located in this region and the inherent difficulty in gaining access, resecting the tumor, and reconstructing surgical defects. With the advent of modern neuroimaging, innovative methods of surgical access, and more reliable reconstructive options, the indications and scope of skull base surgery for malignant tumors have constantly evolved in recent years. Because the majority of malignant tumors that require CFR involve the anterior skull base, we will focus mainly on the current state-of-the-art operative techniques as well as pertinent variations, complications, and results of anterior CFR.

HISTORY OF SKULL BASE PROCEDURES

The first descriptions of a transcranial transfacial approach for orbital tumors were reported by Cushing in 1938 and Dandy in 1941. Over the ensuing decade, other authors also reported on the technique, but it was Ketcham’s systematic description of the combined transcranial transfacial approach to tumors of the paranasal sinuses in 1963 that had the most significant impact on the treatment of these tumors. Since then, numerous other factors have contributed to the progress in enhancing and sustaining the field of skull base surgery. Because of reconstructive techniques involving microvascular free tissue transfer, physical constraints pertaining to the amount of tissue that can be resected have essentially been eliminated. Newer radiological techniques such as CT scanning and neuroimaging modalities such as MR imaging have vastly improved the surgeon’s ability to map the tumor accurately and to conceptualize and devise innovative surgical and reconstructive approaches. Advances such as intraoperative neuroimaging and navigation devices continue to enhance the accuracy and safety of resection. Modern interactive frameless stereotactic devices allow the surgeon to orient the position and motion of instruments in real time to previously imaged anatomical structures. Preoperative CT or MR images are used along with external fiducial markers placed in the patient to establish a relationship between the patient’s reference...
frame and the images. Although this modality provides three-dimensional information on the position of any pointer or instrument relative to the reference images obtained preoperatively, it is limited by potential changes in anatomy that can occur intraoperatively. With the availability of high-speed CT and MR imaging combined with modern high-performance computing technology, it is now possible to integrate high contrast images with frameless stereotactic, interactive localization in near-real time in the operating room. Coregistration of CT, MR, and MR imaging angiography images may be invaluable in surgery for neurovascular skull base tumors of the middle and posterior fossae. The role of these anatomical modalities combined with functional imaging such as single-photon emission CT and other technological advances will doubtless contribute to the armamentarium at the disposal of the multidisciplinary team treating skull base tumors in the future. One of the major limitations to aggressive skull base resections has been the ability of preoperative tests to predict the risk of stroke and death following occlusion or division of the CA. Anatomical neuroimaging modalities such as four-vessel arteriography are not reliable when used alone. Provocative testing with temporary balloon occlusion of the CA (with or without induced hypotension) to determine the physiological effects and/or electroencephalographic changes involves risk, and is not entirely reliable. Functional modalities such as Xe-CT CBF scanning or 99mTc-hexamethylpropyleneamine oxine single-photon emission CT are noninvasive and capable of providing accurate assessment of cerebral perfusion. It is currently believed that combining preoperative CA temporary balloon occlusion with one of these functional neuroimaging modalities, most commonly Xe-CT CBF scanning, provides the most reliable measure of the collateral CA circulation. In addition, advances in perioperative monitoring and care have contributed to decreased complication and mortality rates. Multidisciplinary collaboration allows the head and neck surgeon, neuroradiologist, neurosurgeon, neuroanesthesiologist, microvascular plastic surgeon, critical care specialist, maxillofacial prosthodontist, neurologist, and rehabilitation specialist to plan and complete successful therapy for patients with tumors of the skull base.

SURGERY-RELATED ANATOMY

The skull base is the interface between the cranial cavity and the rest of the body. It is composed of several flat bones that transmit vital nerves and vessels through a number of foramina and fissures. Tumors in the vicinity can either enter the cranial cavity or exit it through these foramina, which offer very little resistance. Successful skull base surgery without undue complications requires a thorough knowledge of regional anatomy, which is clearly beyond the scope of this review. For descriptive purposes, however, the skull base is divided into anterior, middle, and posterior regions based on the cranial fossae.

The floor of the anterior cranial fossa is formed by the crista galli in the midline, cribriform plates, orbital roof, and the planum sphenoidale (Fig. 1). Because the vast majority of malignant tumors of the paranasal sinuses requiring combined CFR necessitate an anterior approach, this will be the major focus of this review.

The middle cranial fossa floor is formed by the squamous temporal bone, the greater wing of the sphenoid, and the planum sphenoidale (Fig. 1). Tumors of the temporal region, auditory canal, and ear may require temporal bone resection (Fig. 3).

The posterior aspect of the temporal bone around the cerebellopontine angle, the occipital bone, and the inferior clivus comprise the posterior cranial fossa (Fig. 4). The head and neck surgeon is only very rarely involved in the

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**Fig. 1.** Computer-generated images. The anatomical extent of the anterior fossa skull base and examples of some tumors requiring anterior CFR.

**Fig. 2.** Computer-generated images. The middle fossa skull base and examples of some tumors requiring middle fossa skull base surgery.
Malignant tumors of the skull base

INDICATIONS AND CONTRAINDICATIONS OF CRANIOFACIAL RESECTION

Any tumor involving or abutting the interface between the cranial cavity and rest of the head and/or neck merits consideration of CFR. The primary surgery-related goals are to provide exposure for total en bloc resection of the tumor and adequate surgical margins while avoiding injury to vital adjacent structures.

Initial skepticism associated with the efficacy and relevance of the procedure in the 1960s was soon replaced by enthusiasm. During the 1980s and 1990s, however, many surgeons realized that although CFR may be technically feasible, it does not benefit all patients. Although it is true that the resectability of a tumor often is a reflection of the expertise and philosophy of the surgeon and support staff, there are cases in which the risks to the patient far outweigh the benefits and CFR is clearly contraindicated. Such contraindications include gross massive invasion of the brain; invasion of the optic chiasm; invasion of both orbits; gross invasion of the internal carotid artery (relative); and elderly patients physiologically and psychologically unsuitable for a major operation. Patients with locally advanced unresectable tumors should be offered alternative therapeutic options including chemotherapy and/or radiotherapy, as will be discussed.

PREOPERATIVE WORKUP

Preoperative workup should include a detailed clinical history, physical examination, and laboratory tests. If feasible, a biopsy sample of the lesion should be obtained to establish a tissue diagnosis. Additional consultations with appropriate specialists are sought, as appropriate, for assessment of cardiopulmonary status. A multidisciplinary team, including the head and neck surgeon, neurosurgeon, reconstructive surgeon, prosthodontist, and radiation oncologist, is essential for delivering optimal care, and all members need to be involved in treatment planning. Clinical nurse specialists and the rehabilitation team provide psychosocial counseling to patients and their family.

Because the skull base is an anatomical area relatively unamenable to clinical examination, neuroimaging is relied on for the accurate mapping of the extent of the tumor. Advances in surgical technique and improved results can be attributed largely to the surgical team’s preoperative ability to plan and define the operation accurately. Axial and coronal contrast-enhanced CT scanning performed with bone and soft-tissue windows provides optimal infor-
mation on tumor-induced bone erosion or destruction. In the absence of gross bone destruction, indirect findings such as widening of skull base foramina should raise suspicion that malignancy is involved. On the other hand, MR imaging demonstrates superior soft-tissue resolution and can visualize anatomy in the sagittal plane. Neuroimaging assessment of structures such as the orbit, dura mater, and brain can provide data detailed and reliable enough to guide the surgeon intraoperatively (Fig. 5). It is crucial to realize that CT and MR imaging are complementary rather than mutually exclusive techniques, and the majority of cases benefit when information is culled from both examinations. In cases of tumors in proximity to or actually involving major blood vessels such as the internal carotid artery, resection carries the risk of cerebral vascular compromise. Neuroimaging evaluation in such cases may include MR imaging or invasive angiography, temporary balloon occlusion or Xe-CT CBF scanning studies as indicated. Selected vascular tumors of the skull base may be embolized to decrease intraoperative blood loss and facilitate excision. As previously discussed, neuroimaging studies are also necessary if intraoperative neuronavigation is part of the operative plan.

HISTOLOGICAL TUMOR TYPE

Craniofacial resection is most commonly undertaken for malignant tumors, and the majority are squamous cell carcinomas. Benign tumors such as nasopharyngeal angiofibromas, chondomas, and osteomas of the ethmoid or frontal sinuses may require CFR for complete excision. Other histological types that may be encountered are shown in Table 1. Most major reports on CFR include a heterogeneous population of tumors, but a particular histological subtype may predominate depending on the referral patterns within that practice; for example, the authors of most European studies report larger numbers of adenocarcinomas whereas skin cancers predominate in Brazil. The pie chart in Fig. 6 illustrates the histological distribution of malignant tumors in a large international database of patients who underwent CFR.

SURGICAL TECHNIQUE: VARIATIONS AND ADVANCES

Although techniques of anterior CFR have been evolving, key aspects of the procedure remain established. The goal of the operation is to obtain adequate exposure of the tumor and important structures in the vicinity, which can therefore be preserved to ensure good surgical margins around the tumor. The CFR-related principles for malignant tumors include: adequate exposure of the tumor and adjacent vital structures; minimal or no brain retraction; (intraoperative spinal drainage ± mannitol diuresis to maintain brain slack); watertight dural repair by using dural patch (pericranium, fascia lata, bovine pericardium, cadaveric dura, or allograft) for large defects and direct suture for smaller ones; and reconstruction of the skull base defect by using galeal pericranial flap or free tissue transfer (rectus abdominis flap for large defects). The cranial aspect of the operation is most often performed via a standard bifrontal craniotomy incision whereas inferior access is obtained via one of several transfacial approaches. The basic technique for anterior CFR has been well described14–19 and is summarized in Table 2. Certain key aspects of the procedure, however, should be highlighted. Preoperative intravenous antibiotic agents are administered during induction of general anesthesia to provide broad-spectrum coverage during the procedure. An antibiotic regimen at our institution combines ceftazidime, vancomycin, and Flagyl to provide prophylaxis against Gram-positive and -negative bacteria as well as Bacteroides. Intravenous antibiotic administration is continued through the postoperative period as long as the nasal pack is in place.

With the patient in the lateral decubitus position, a lumbar puncture is performed and an intrathecal catheter placed for intra- and postoperative CSF drainage. Intra-

<p>| TABLE 1 |</p>
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<th>Common histological subtypes of skull base tumors*</th>
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<tr>
<td>malignant</td>
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<tr>
<td>adenocarcinoma</td>
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<tr>
<td>undifferentiated carcinoma</td>
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<tr>
<td>neuroendocrine carcinoma</td>
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<tr>
<td>melanoma</td>
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<tr>
<td>SCC &amp; BCC of skin</td>
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<tr>
<td>salivary gland carcinoma</td>
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<tr>
<td>sarcomas (soft tissue &amp; bone)</td>
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<tr>
<td>chordomas</td>
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<tr>
<td>benign</td>
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<tr>
<td>angiofibroma</td>
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<tr>
<td>fibro-osseous lesion</td>
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<td>neurovascular tumor</td>
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* BCC = basal cell carcinoma; SCC = squamous cell carcinoma.
operatively, the anesthesiologist drains 10-ml aliquots of CSF until adequate brain relaxation is achieved to minimize handling and retraction of the brain. Generally, 30 to 60 ml of CSF are drained for a standard anterior CFR.

The standard bicoronal frontal incision extends from the tragus of one ear to that of the other, just within the hairline (Fig. 7 upper). The scalp is incised down to the plane between the epicranial aponeurosis and the pericranium. The posterior scalp flap is elevated in the loose areolar tissue plane between the galea and pericranium several centimeters posteriorly to expose the pericranium. Hemostatic Raney clips are applied to the cut edges of the scalp. The pedicled galeal pericranial flap (Fig. 7 lower left and right) is considerably more robust than a pericranial flap and is the flap of choice when reconstructing the skull base defect. It is important to harvest a sufficient length of the flap posteriorly so that it can extend to the skull base defect without undue tension. To provide additional length to the graft, the skin incision is made well posterior on the scalp and through the pericranium. The posterior scalp flap is elevated in the plane between the pericranium and the skull all the way up to the supraorbital ridges. During reconstruction of the skull base, the galeal pericranial flap is separated from the anterior scalp flap, remaining superficial to the galea. Sufficient length of the flap is separated to obtain adequate tissue for repair, without compromising its blood supply from the supraorbital and -trochlear vessels. The pericranium is released posteriorly and laterally along the temporalis insertions. The anterior flap is then elevated in a subperiosteal fashion to expose the supraorbital ridge, glabella, and upper half of the nasal bones. Care should be taken to preserve the supraorbital and -trochlear vessels that supply the pericranial–galeal graft. Once the anterior flap is turned, the galeal–pericranial flap is dissected using a No. 15 blade and wrapped in a bacitracin-soaked gauze throughout the procedure. A 1-cm cuff of galea should be preserved on the anterior flap for subsequent scalp closure. The incision should be fashioned to provide approximately 8 cm of pericranial–galeal graft to cover the skull base deficit. The remaining 10 cm of pericranial graft can be folded over to create third layer of closure. On occasion, the scalp incision may be limited to one side if only unilateral exposure is desired as, for example, in cases of orbital tumors that do not approach or cross the midline.

With the anterior scalp flap elevated to expose the glabellar region and the upper half of the nasal bones, the proposed craniotomy bone cuts are out marked with methylene blue on the anterior wall of the frontal sinus and the bone. Cosmetic saddle-nose deformity can be avoided by placing this bone cut so that the nasal bones are retained in situ and excluded from the bone plate. A single midline burr hole is made in the frontal bone approximately 5 cm above the glabella, and the dura is separated from the under side of the bone. Arachnoid granulation bleeding is readily controlled using thrombin-soaked Gelfoam. The B1 bit on the Midas Rex drill is used to make bone cuts to approximately the level of the midorbits bilaterally without entering the frontal sinuses. The frontal sinus cuts are made using the 2-mm cutting burr through the anterior and posterior walls of the sinus. This procedural strategy has decreased the incidence of dural tears adjacent to the frontal sinus. A small frontal craniotomy is favored (Fig. 8) to increase the surface area of the frontal bones for the surgeon’s hands to rest and consequently decrease the possibility of pressure inadvertently transmitted to the frontal lobes. Dissecting the sagittal sinus and dura from the bone elevates the flap. The remainder of the posterior wall of the frontal sinus is removed using a rongeur and the mu-

### TABLE 2

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<th>Standard steps involved in craniotomy</th>
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<tr>
<td>1) preop placement of lumbar catheter</td>
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<td>2) supine position on op table w/ head in neutral position</td>
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<tr>
<td>3) ceramic corneal shields, or eyelids sutured to prevent corneal exposure &amp; injury</td>
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<tr>
<td>4) bicoronal skin incision w/ in hairline</td>
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<tr>
<td>5) adequate elevation of posterior scalp flap superficial to galeal plane to allow generous-length galeal–pericranial flap for skull base repair</td>
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<tr>
<td>6) anterior scalp flap galeal–pericranial flap elevated up to supraorbital ridges to maintain blood supply of flap</td>
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<tr>
<td>7) anteriorly based U-shaped galeal–pericranial flap elevated</td>
</tr>
<tr>
<td>8) midline frontal burr hole &amp; small frontal osteotomy</td>
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<tr>
<td>9) frontal bone plate preserved in sterile saline for replacement at end of op</td>
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cosa is exenterated. A bone rongeur is use to remove the septae in the frontal sinus and all sharp spicules of bone are smoothed using a burr. It is important to curette the entire mucosa of the sinus and to cranialize the frontal sinus by excising its posterior wall. The nasofrontal duct openings are plugged with Gelfoam. Technical variations of craniotomy include multiple frontal and parasagittal burr holes; single frontal burr hole and small osteotomy; anterior subcranial osteotomy; and facial disassembly. The practice of using multiple burr holes has largely been abandoned because of the associated cosmetic deformity. A small frontal osteotomy in which a single frontal burr hole is made provides adequate access for exposure of the anterior skull base without the morbidity associated with multiple burr holes. The anterior subcranial approach has been reported to provide adequate exposure of the anterior skull base and the advantage of precluding frontal lobe retraction and lateral rhinotomy.13 The size of the osteotomy can be varied depending on the exposure desired. Although effective in providing exposure for correction of craniofacial deformities and fractures, this approach does not allow retraction of the frontal lobe nor inspection or resection of the dura at the anterior skull base, which are crucial steps for total en bloc tumor excision in most cases requiring anterior CFR. The technique of facial disassembly allows the nasal bone complex to be removed en bloc to facilitate transfacial access to the nasal cavity (Fig. 9 left and center). The bone complex is then replaced in its original position at the end of the procedure and fixated using miniplates (Fig. 9 right).

Once the craniotomy is completed, the dura of the floor of the anterior cranial fossa is elevated, sharply dividing its attachments to the crista galli. The dural sleeves along the olfactory nerves need to be individually divided and ligated. Immediate ligation of the dural sleeves ensures that contamination of the brain during subsequent phases of the operation is avoided. This segment of the dura along with the olfactory sleeves obviously needs to be resected in cases of esthesioneuroblastomas or other tumors that perforate the cribiform plate. Once the dura is elevated, the exposed crista galli can be excised using a rongeur. Small dural defects can be repaired using a free graft of pericranium (Fig. 10 left) whereas larger defects require bovine pericardium (Fig. 10 center and right) or reconstituted cadaveric dura. Use of fibrin glue can be effective not only in supplementing repairs of dural tears and over the ligated olfactory dural sleeves but also as a sealant to prevent postoperative pneumocephalus.

Dissection is then continued in a posterior direction to expose the posterior part of the cribiform plate and the planum sphenoidale. It must be reemphasized that retraction and handling of the brain should be minimized to avoid postoperative complications. Slackening of the brain by draining 30 to 60 ml of CSF via the intrathecal catheter is usually sufficient. A wide malleable retractor is used gently to protect the frontal lobes in the midline posteriorly to expose the planum sphenoidale and the cribiform plate. The bone cuts in the floor of the anterior cranial fossa are completed using a fine burr on the high-speed drill. In cases of tumors restricted to the midline of the anterior cranial fossa, a circumferential cut around the cribiform plate is adequate. More extensive tumors, such as those involving the lamina papyracea or the orbit, require the removal of appropriate amounts of the orbital roof to allow en bloc resection of the tumor (Fig. 11). This concludes the first phase of the operation.
The tumor can be approached from the inferior aspect via one of many transfacial approaches (Table 3). The traditional Weber–Ferguson incision and its modifications are no longer acceptable because the cosmetic results are generally suboptimal (Fig. 12 upper left and right). Our current preference is to use a modified Weber–Ferguson incision placed in the nasal subunits along the nasolabial fold (Fig. 12 lower left and right). This basic incision is modified depending on the anatomical access required. The upper lip is split in the midline if exposure of the lower half of the nasal cavity, hard palate, or maxilla is required. The cosmetic outcome after using this modified incision is considerably better because the scars situated at the borders adjoining the subunits of the nose are less conspicuous. Although the sublabial approach avoids making facial scars, its exposure is generally adequate only for inferiorly located tumors. Facial incision may be avoided in cases of selected tumors of the superior nasal vault or ethmoid sinuses if adequate exposure and resection can be accomplished transcranially.

The skin incision is taken through the entire thickness of the soft tissue and musculature of the nasolabial region. If a facial disassembly of the nasal bones is planned, the incision can be extended across the glabella up to the contralateral eyebrow. Alternatively, subciliary extension up to the lateral canthus can provide exposure for a total maxillectomy (Fig. 13). After the skin incision is completed, the osseous rim of the piriform aperture and the inferior orbital rim are delineated. The attachment of the medial canthal ligament is sharply divided and tagged with a No. 4-0 Neurolon suture for subsequent reattachment to the nasal bone during closure. Fine periosteal elevators are used to dissect the nasolacrimal duct from its fossa, and the duct is divided flush with the rim of the orbit by using a scalpel. Before the nasal pack is introduced, nasolacrimal drainage needs to be reestablished using silastic stents.

It is vital to plan and make soft-tissue and bone cuts around the tumor so that total en bloc resection of the tumor is achieved (Fig. 14). Tumors that are soft and friable may fracture during removal, but piecemeal resection should never be the aim of the procedure. Once the tumor
has been surgically invaded into, blood obscures normal tissue planes and surrounding margins. Although there are no data to support the en bloc resection, there can be little doubt that if piecemeal resection is the intent, no patient should be subjected to the morbidity associated with CFR.

Reconstruction of the surgical defect entails repositioning the divided medial canthus to its normal position, providing watertight support across the anterior skull base, reestablishing nasolacrimal drainage, and accurate reapproximation of the facial incisions. It is important to match the position of the divided medial canthal ligament accurately to the contralateral side to avoid telecanthus. The previously tagged end of the medial canthal ligament is sutured via an appropriately positioned drill hole in the nasal bone.

As described, the primary goal of reconstruction of the skull base defect after anterior CFR is to isolate the cranial cavity from the upper airway to prevent contamination and infection. Various options have included split thickness skin graft and pericranial flap. None is as robust and reliable as the galeal pericranial flap. The flap is anchored in place by suturing it through drill holes along the rim of the osseous defect so that it provides support to the brain while isolating the cranial cavity from contaminants of the nose and upper aerodigestive tract. Because the nasal cavity/nasopharyngeal surface of the flap epithelializes over time, skin grafting is unnecessary. Similarly, osseous support for the skull base defect is not necessary because brain herniation is not a problem. If a functioning eye has been preserved, the floor of the orbit must be reconstructed using a split calvarial graft or rib graft to avoid postoperative diplopia.

If extirpation of the tumor requires craniectomy, reconstruction-related options include no osseous support, alloplastic material, split calvarial graft, or free flap reconstruction. Small craniectomy defects may be left open, without bone reconstruction, if the patient is able and willing to take measures to protect the underlying brain from injury. Alloplastic material such as acrylic plates, titanium plates, or synthetic mesh and methylmethacrylate cement may interfere with healing and delay initiation of radiotherapy. The approach-related risks and benefits need to be carefully considered to avoid compromising the quality of treatment.

Nasolacrimal drainage needs to be reestablished to avoid postoperative epiphora. A silastic stent is used to cannulate the puncta of the nasolacrimal duct, and the stent is kept in place by knotting its two ends within the nasal cavity. The entire nasal cavity is then packed snugly with a Xeroform roller gauze packing to provide support to the galeal pericranial flap superiorly and the medial orbital periosteum laterally. Towards the end of the 1st postoperative week, the pack is removed in stages over a period of 2 to 3 days via the nostril.

For locally advanced tumors requiring maxillectomy, the traditional approach has been to surface the cavity with a split thickness skin graft and rehabilitate the patient with a palatal obturator. The advent of microvascular surgery allows these defects to be reconstructed using vascularized flaps that provide lining, soft-tissue bulk, and osseous support. The case depicted in Fig. 15 left is one of locally advanced tumor of the maxillary antrum; a radical maxillectomy, including orbital exenteration via a craniofacial approach, was required. After such an extensive resection, the surgical defect is necessarily complex, requiring tissue to support the skull base defect and provide internal as well as external lining (Fig. 15 right). The free rectus abdominis flap is ideally suited to provide these components and, although cosmetically not pleasing, results in prompt healing that allows for timely institution of adjuvant radiotherapy (Fig. 16). Alternatively, if the soft-tissue defect is not extensive, other flaps such as the radial forearm, scapula, or latissimus dorsi may be considered. A bulky intraoral component of the flap can interfere with dental prosthetic rehabilitation, but patients who retain healthy dentition after resection are able to function without being inconvenienced by the necessity for care of the prosthesis. In such patients the external appearance can be greatly improved by a facial prosthesis. The services of a skillful maxillofacial prosthetist are invaluable in rehabilitating patients with facial deformity following extensive CFS (Fig. 17).

Middle Cranial Fossa Resection

In tumors of the parapharyngeal space or pterygoid fossa extending into the infratemporal fossa, CFR can be achieved via a middle fossa approach. More commonly,
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Fig. 12. **Upper Left and Right:** The traditional Weber–Ferguson incision and its variants generally provide suboptimal cosmesis. **Lower Left and Right:** A modified Weber–Ferguson incision respecting the nasal subunits yields improved postoperative cosmesis.

tumors of the ear or auditory canal require temporal bone resection for total en bloc excision. The patient shown in Fig. 18 **upper left** harbored a locally advanced squamous cell carcinoma of the external auditory canal; the neoplasm invaded the parotid gland and overlying skin. Neuroimaging revealed extensive invasion of the parotid space and temporal bone. The patient underwent a subtotal temporal bone and pinna resection, radical total parotidectomy with segmental mandibulectomy, and neck dissection in an en bloc fashion (Fig. 18 **upper right**). A surgical defect of this magnitude (Fig. 18 **lower left** and **right**) can be promptly reconstructed using a free rectus abdominis flap so that radiotherapy can be instituted early in the postoperative course. The technical details of this and other middle cranial fossa procedures are clearly beyond the scope of this review, but it is important to realize that every procedure requires specific tailoring to the individual case.

**COMPLICATIONS AND AVOIDANCE**

In approximately one third of patients undergoing CFR postoperative complications develop, and the inhospital mortality rate in most major series is less than 5%. The most common complications following anterior CFR relate to wound sepsis and the central nervous system. Postoperative wound sepsis can lead to meningitis, sub- or intradural abscess, or even osteomyelitis and ultimately loss of the bone flap. Table 4 provides a summary of complications following CFR for malignant tumors. These complications may be minimized by appropriate perioperative antibiotic therapy and meticulous surgical technique, including the use of the galeal pericranial flap to isolate the cranial cavity from the upper airway. Cerebrospinal
fluid leakage can also be avoided by ensuring watertight dural closure and reconstruction. Larger composite defects with soft-tissue loss require free flap reconstruction. The lumbar spinal catheter is kept in situ while the patient is recovering in the neurosurgical unit postoperatively, and spinal fluid can be drained intermittently to manage CSF leakage conservatively. Depending on the confidence in the dural repair, 10-ml aliquots are drained by the intensive care unit nurse every hour for 48 to 72 hours postoperatively to ensure a watertight dural seal. Draining 10 ml/hour prevents overdrainage, which may occur if one attempts to maintain drainage to a prescribed pressure. Increasing the hourly drainage to 15 to 20 ml may treat persistent leaks. Leaks that do not respond to adequate conservative treatment can be localized using fluorescein and need to be repaired either by the transnasal endoscopic approach or, more rarely, transcranially.

Postoperative obtundation and confusion (“acute brain syndrome”) is not unusual. It can be minimized by avoiding unnecessary intraoperative retraction of the frontal lobes of the brain. It is crucial to keep the brain parenchyma slack by obtaining adequate drainage of CSF through the spinal catheter so that the frontal lobes can be handled minimally. Altered neurological status can also be caused by pneumocephalus, but an intracranial hemorrhage must be ruled out. Pneumocephalus can be avoided by ensuring good separation of the cranial cavity from the nasal cavity and sinuses by using the aforementioned techniques. The patient should be instructed to avoid straining or trying to blow his/her nose in the immediate postoperative period. Some authors have advocated routine of tracheostomy to avoid this complication, but we have generally found this to be unnecessary. An epidural drain on gravity drainage is routinely placed to prevent pneumocephalus and is maintained until lumbar CSF drainage is discontinued or until there is cessation of the air leak. This drain prevents brain herniation, which can potentially result from pneumocephalus (that is, intracranial mass) and lumbar CSF drainage. As noted, thrombin glue is used to augment the skull base repair and has been found to decrease pneumocephalus and air leakage by acting as a sealant. Dural tacking sutures are not used because the epidural drain on suction creates severe headaches when the dura is tacked to the bone.

**IMPACT OF MULTIMODALITY THERAPY**

There are no published randomized data to support the observation that postoperative radiotherapy has improved surgery-related outcome in patients with skull base tumors. The authors of retrospective reviews have generally failed to identify any statistically significant improvement in results after postoperative radiotherapy likely because of the selection bias associated with treatment choice; thus, patients undergoing postoperative radiotherapy gen-
eraly harbor higher-risk tumors compared with those in whom adjuvant treatment is deemed unnecessary. Nonetheless, most clinicians would currently consider postoperative radiotherapy in the majority of patients undergoing CFR because of the inherent difficulty in obtaining wide surgical clearance of the tumor.

In cases of locoregionally advanced tumors that are unresectable or resectable only with unacceptable functional results, the patients must be considered for alternative management. Concurrent chemotherapy and accelerated hyperfractionated radiotherapy in patients with unresectable nasopharyngeal/paranasal sinus lesions has yielded a 2-year local progression-free survival rate of 94% and an overall survival rate of 80%. Obviously, the approach-related data requires further evaluation and long-term follow-up examination, but initial findings are encouraging for patients for whom there are otherwise no viable treatment options.

RESULTS OF THERAPY AND PREDICTORS OF OUTCOME

Anterior Craniofacial Resection

In a recent review of major studies in the literature the authors found that 5-year survival rates up to 80% have been reported after CFR.² Because of the heterogeneity of the patient populations reported and the relatively small numbers in each series, an accurate assessment of the efficacy of the procedure has been difficult. Local disease control is achieved in approximately two thirds of patients, and approximately 60% survive free of disease 5 years after treatment.¹⁸ The authors of a large series from Italy reported comparable results.³ Regional nodal failure is not common, whereas distant metastases have been reported in up to 25% of patients.⁸,¹⁸ In a recent review of a large international collaborative cohort, the 5-year overall survival and disease-specific survival rates (calculated using the Kaplan–Meier method) were 53 and 57%, respectively, in patients followed up for a median of 25
months (range 1–940 months) (unpublished data, International Collaborative Study on Craniofacial Surgery).

Table 5 provides a summary of factors predictive of prognosis in patients undergoing anterior CFR for malignant tumors. In cases of esthesioneuroblastoma and low-grade sarcoma, overall outcome is the best, whereas in those of melanoma and anaplastic carcinoma outcome is the worst (Fig. 19 upper). Intracranial extension (dural and/or brain invasion) is a significant adverse factor (Fig. 19 center), as is the ability to achieve tumor-free surgical margins (Fig. 19 lower). Although CFR may be technically possible for locally extensive tumors invading the dura or brain, local recurrence is a frequent problem (Fig. 20).

TEMPORAL BONE RESECTION

Treatment-related outcomes following temporal bone resection have been reviewed in a recent report.1 An improvement in results was noted over the past half century, with survival doubling to 50% compared with rates in initial series. The results of temporal bone resection for cutaneous carcinoma of the ear are slightly better, with an approximate 80% survival rate for basal cell and 60% for squamous carcinomas.6,11 Prognostic factors predictive of adverse outcome include the extent of disease, facial nerve paralysis, tumor-positive surgical margins, involvement of dura, and regional lymph node metastases.

CONCLUSIONS

Craniofacial surgery for malignant tumors of the skull base has advanced significantly since its earliest description. Advances in neuroimaging technology, operative technique, perioperative care, and adjuvant treatment have contributed to improved results. Although complication rates are high and surgery-related mortality is low, further research is required to assess the impact on quality of life in patients with these lesions.

References

5. Dandy WE: Orbital Tumors: Results Following the Transcranial Operative Attack. New York: O Piest, 1941