Paparella: Volume IV: Plastic and Reconstructive Surgery and Interrelated Disciplines

Section 2: Disciplines Closely Associated With Otolaryngology

Chapter 23: Neurosurgery of the Head and Neck

John M. Tew, Raymond Sawaya, Myles L. Pensak

Many responsibilities and problems are shared by the otolaryngologist and the neurosurgeon. Disorders that begin in the ear and its environs or in the paranasal sinuses commonly invade the intracranial cavity. Occasionally the reverse is true. Also, many lesions of the central nervous system either refer or produce symptoms that lead patients and referring physicians to consult the otolaryngologist.

From time to time the development of new medications, diagnostic methods, and improved instrumentation modifies treatment and indications for operation. Computed tomography (CT) of the head, microsurgical techniques, and new chemotherapeutic agents are cases in point.

Space precludes a comprehensive presentation of the specialty of neurosurgery here. This chapter is devoted to those areas in which the responsibilities of the specialists overlap and those areas in which the authors' opinion differs from that of our otolaryngologic colleagues. Included in this chapter are certain important examples of disorders that are rare or sufficiently unusual to escape the reader's attention if not recorded here. Finally, because of the effect that CT has had on neuro-otologic diagnosis, pertinent examples of the use of this valuable tool will be presented.

Advances in Neurosurgery

Over the last decade, tremendous advances have been made in the management of complicated neurosurgical disorders involving the base of the skull. Although the basic principles of intracranial surgery were established decades ago, including proper flap positioning, meticulous handling of tissues, and careful hemostasis, modern neurosurgery has benefited from numerous technical advances that have become essential components of a technically successful operation.

The bipolar coagulation offers the advantage of efficacious hemostasis without the danger of spreading current encountered with the older monopolar coagulator. The introduction of the microscope along with sophisticated microsurgical instruments represents the single most important technical breakthrough, improving technical capabilities and, therefore, results. The microscope provides a wide range of magnification and enhanced illumination. With the addition of long, bayonet-shaped microsurgical instruments, it is now possible to approach lesions in any intracranial location and to maintain good visualization of the vascular supply and of the critical areas of the central nervous system.

Self-retaining brain retractors have been designed to aid in improving the surgical exposure while at the same time maintaining a constant pressure force on the brain and

freeing the surgeon's hands. The prototype retractor we have been using can accommodate several retractors placed circumferentially. Additional arms can be added to hold smoke suction devices necessary during laser surgery.

More recently, we have seen the application of laser science to the extirpation of intracranial neoplasms (Tew and Tobler, 1984). The infrared carbon dioxide laser is most commonly used for surgical applications, although other laser sources such as the argon and the neodymium:YAG are being used with increasing frequency. Laser light is amplified in a resonator that is an essential part of the device; the energy is then directed by reflective mirrors into the operative field, using either a handpiece or a micromanipulator attached to the microscope. The beam can be manipulated by adjusting the power intensity of the laser source and the focal point of the beam. The functions served by the laser are cutting, evaporation, and coagulation. When used to cut or evaporate tissue, the laser is actually vaporizing cells by overheating them; however, because of the precise localization of the laser beam, surrounding tissue suffers minimal injury. Lower power density is used for coagulating blood vessels. The advantages offered by the laser are numerous and include minimal retraction of nervous tissue, a bloodless surgical field, precise non-touch vaporization of tissue, reduced blood loss, and elimination of interference with monitoring equipment. The application of laser technology to surgery for intracranial neoplasms is constantly expanding: it is now possible to completely vaporize an acoustic neuroma without damaging the facial nerve, or to remove a previously inaccessible meningioma at the base of the skull with minimal or no morbidity (Gongbai and Xu, 1983; Tew and Tobler, 1984).

A new instrument using ultrasonic energy that fragments and aspirates tissue has been introduced (Flamm and colleagues, 1978). Large blood vessels are not affected by the ultrasonic vibrations, which allow removal of tumor directly from the outer surface of the adjacent vessels.

Other factors that have greatly enhanced the safety of intracranial surgery include modern neuroanesthesia, improved methods of managing increased intracranial pressure, and improved control of fluids and electrolyte balance.

Intraoperative monitoring of neural tissue function is gaining wide acceptance and applicability, especially in surgery of the cerebellopontine angle region. Intraoperative monitoring of evoked potentials may be useful when four conditions are met: (1) a part of the nervous system is at risk or needs to be identified, (2) sites are accessible for stimulation and recording, (3) personnel and equipment are available, and (4) there is a possibility of intervention by the surgeon or anesthesiologist if damages in the sensory evoked potentials are detected (Grundy, 1982; Grundy and colleagues, 1982).

Sensory evoked potentials can, for instance, guide the surgeon during excessive manipulation of vascular and neural structures, and allow the surgeon to be more aggressive in the resection of lesions intimately related to cranial nerves.

Interventional neuroradiology is a new field aimed at aiding in the management of intracranial vascular lesions. Large vascular tumors, such as meningioma and chemodectoma, can be devascularized by selective embolization of feeding tumor arteries, thereby markedly reducing the intraoperative blood loss. This procedure is most indicated when most of the

blood supply of the tumor originates from the external carotid artery (Bernstein and Kircheff, 1981; Teasdale and colleagues, 1984).

Materials used for embolization includes Gelfoam, silicone or acrylic polymer, and metallic pellets and polyvinyl alcohol sponge. Tumor necrosis and obliteration of the capillary bed occur after embolization treatment.

The most recent technical advance in the surgery of deep-seated brain lesions is the CT-guided stereotaxic apparatus (Apuzzo and Sabshin, 1983; Heilbrun and colleagues, 1983). With the use of a special head frame, specific sites deep in the brain can be identified on a CT scan, and coordinates can be calculated from the scanner and transferred to a stereotaxic apparatus by a portable computer in the operating room. This technique offers the advantage of precision and safety, and is successful in the biopsy of obscure, deep-seated lesions and in the drainage of intracranial cysts and abscesses. This technique is preferred for determining proper therapy for the high-risk patient, and is also being used for the implantation of radioactive sources and chemotherapeutic agents within brain tumors. More recently, the laser has been introduced through stereotaxic attachments to facilitate the rapid and precise removal of neoplastic tissue (Kelley and colleagues, 1982).

Surgical Approaches to the Skull Base

Within the past decade, interdisciplinary approaches to lesions involving the skull base have allowed physicians to deal optimally with previously unresectable lesions (Glasscock and colleagues, 1985; Fisch and Kumar, 1985). The safe extirpation of tumor masses or the reconstruction of vascular abnormalities has increasingly drawn attention to this anatomic region. This section addresses the complex and irregular osteology of the skull base, and examines current therapeutic modalities and surgical approaches to pathologic conditions of this area that are of common interest to the otolaryngologist and neurosurgeon.

Anatomic Definition

The cranial vault is divided into anterior, middle, and posterior fossae. The presence of numerous foramina, fissures, suture lines, and bony appositions are characteristic features of the irregular surface of the skull base. Moreover, variability of bone form and density is appreciated (ie, the thin lamina cribrosa of the ethmoid plate and the dense otic capsule housed within the temporal bone), thus predisposing certain areas to an increased degree of vulnerability or surgical access.

The anterior cranial fossa is bounded by the frontal bone anteriorly, the orbital roofs, inferiorly, and the sphenoid posteriorly. In the midline the cribriform plate of the ethmoids is an area of defined weakness. Attendant to the cribriform plate is the crista galli, which fixes (along with the frontal crest) the falx cerebri. The anterior and posterior ethmoid arteries pass through named foramina in this bone, while the olfactory bulbs lie just above the plate.

The lesser wings of the sphenoid, forming the posterior wall of the anterior fossa, form the paired anterior clinoid processes just posterior to the jugum sphenoidale. The optic chiasm, and more posteriorly the tuberculum sellae, are intimately related to the midline positioning of the lesser wings of the sphenoid. The posterior clinoid processes form the back

of the dorsum sellae. Although pneumatization of the paranasal sinuses may be variable (Ritter, 1978), access to anterior fossa floor may be obtained through the transnasal-transsinus route in addition to the standard frontal craniotomy.

More posteriorly, the petrous portion of the temporal bone defines the borders of the middle and posterior fossae. The petrous bone is trifaced, with its narrow leading edge directed in an anteromedial direction. The steep anterior wall forms the posterior margin of the middle fossae. The posterior surface is bounded by the superior and inferior petrosal sinuses running along the longitudinal axis of the bone. The internal auditory canal lies midway across the posterior surface, transmitting cranial nerves VII and VIII as they exit the cerebello-pontine angle.

Within the internal auditory canal, at the level of the fundus, a bony partition, the crista falciformis, separates the canal into upper and lower passages. Within the upper bony canal lies the facial nerve anteriorly, separated from the superior vestibular nerve by a bone strut commonly called Bill's bar. The lower canal is occupied by the anteriorly located cochlear nerve and the posteriorly positioned inferior vestibular nerve. Laterally, the inferior vestibular nerve gives off a branch to the ampullae of the posterior semicircular canal (singular nerve), which travels within its own canal. The course of the facial nerve is of utmost importance to the temporal bone surgeon, and is discussed in detail in other sections of this book.

The otic capsule housing the end organs of hearing and balance lies within the temporal bone. The phylogenetically ancient vestibular labyrinth is directed posteriorly; the cochlea is located more anteriorly.

When viewed from below, the inferior surface of the skull base is highly irregular. Laterally, the digastric muscle attaches to the mastoid portion of the temporal bone. At the stylomastoid foramen the facial nerve exits the temporal bone. A constant landmark for this locus is the tympanomastoid suture line. The jugular foramen lies medial and inferior to the stylomastoid foramen transmitting cranial nerves IX, X, and XI as well as the jugular vein. Separated by a wedge of bone, the internal carotid artery lies anterior to the jugular foramen. Upon entering the temporal bone, the artery passes along the anterior margin of the promontory (the basal turn of the cochlea) before entering its intracranial portion by passing through the foramen lacerum. As the carotid exits the temporal bone it is in juxtaposition to the eustachian tube, just below the semicanal of the tensor tympani muscle.

Inferiorly, and central in position, is the foramen magnum, bounded laterally by the occipital condyles and anteromedially by the basioccipital synostosis. Inferior to the jugular fossa, the hypoglossal canal and foramina transmit cranial nerve XII.

Tumors that invade the skull base may extend from the clivus anteromedially to the jugular fossa, extensively involving the infratemporal fossa. The margins of limitation within this area are generally recognized to be the nasopharynx medially, the scaphoid fossa of the sphenoid anterolaterally, and the foramen spinosum superiorly. Extension into the middle and posterior cranial fossae is not a contraindication from a surgical anatomic vantage.

Surgical Techniques

The techniques described herein pertain to lesions that commonly interface the two disciplines. Tumors that are primarily sinal in nature or extensive lesions requiring a craniofacial resection are addressed in other sections of this text.

Access to the Pituitary

Sublabial Transseptal-Transsphenoidal Approach. Approaches to the sella through the sphenoid sinus have been recorded extensively (Hardy, 1985; Sasaki, 1984; Lee, 1978), since Koenic (1898) first approached the sella by this route (Calcaterra and colleagues, 1985). Transfrontal procedures may be employed for large asymmetric lesions, but the transsphenoidal technique is most commonly used.

Technique. The patient is placed in the supine position with the back, shoulders, and head elevated after oroendotracheal anesthesia. The nasal mucosa and sublabial regions are injected with lidocaine (Xylocaine) and epinephrine in a 1:200.000 dilution. The mucosa is incised above the buccogingival sulcus, while pledgets impregnated with cocaine solution are topically applied to the nasal mucosa. The periosteum is elevated from canine fossa to canine fossa across the piriform crests. The maxillary spine is identified as well as the caudal septum. The mucoperichondrium is elevated from the septal cartilage and the anterior maxillary spine is separated from the maxilla. With inferior and superior tunnels created, the quadrangular cartilage is disarticulated from the vomer and delivered to the left side. Mobilization separates it from the ethmoid posteriorly and maintains a superior attachment. A hypophysectomy speculum is placed and the perpendicular plate of the ethmoid removed with a rongeur. Using the vomer as a guide to the sphenoidal rostrum, the anterior wall of the sinus is noted and the sinus opened.

The mucosa is removed from the sinus, as well as the septum, once these are carefully identified. The bone separating the sella is removed, exposing the pituitary tumor. After tumor extirpation, fascia lata or muscle is employed to obliterate the sella, and harvested abdominal fat is used to fill the sphenoid defect. Nasal packing is placed as in a septoplasty and the sublabial incision is closed with absorbable suture.

Anterior Approaches to the Clivus and Craniocervical Regions. Transpalatal and transoral approaches give excellent visualization for a variety of conditions ranging from choanal atresia to juvenile angiofibromas, clival chordomas, and craniocervical abnormalities (Pásztor and colleagues, 1984; Hayakawa and colleagues, 1981; O'Laoire and Thomas, 1982; Jenkins and Canalis, 1984).

Technique. The patient is placed in the supine position and an oroendotracheal tube is placed. In cases requiring an extended field of resection or in which postoperative airway compromise is anticipated, a tracheostomy is performed. An oral cavity retractor (Dingman or Whitehead)0 is placed and the palate (soft and hard) is injected with lidocaine and epinephrine 1:100.000. By injecting the greater palatine foramen directly, bleeding attendant on flap elevation will be minimized.

A U-shaped incision is made circumferentially from the third molar around the alveolar ridge at the level of the gingivobuccal sulcus from one side to the opposite. Care should be taken to avoid the greater palatine arteries. A posteriorly based mucoperiosteal flap is elevated, exposing the bony hard palate. As needed the palate (hard) may be drilled or rongeured off, exposing the nasopharynx and floor of the nose posteriorly.

For further exposure on the pharyngeal wall, the soft palate may be split lateral to the uvula. A midline incision is then made in the posterior pharyngeal wall, exposing the soft tissue just anterior to the anterior longitudinal ligament. Nonparallel incisions allow for a tighter closure through the suture lines of the retropharynx after removal of tumor or work on the cervical spine.

Transbasal Approach. This procedure, adapted from work done by Tessier and colleagues (1973), was described by Derome (1977) and should be employed to remove extensive anterior skull base lesions.

Technique. A bitemporal coronal skin flap is incised and a large bifrontal craniotomy performed with the anterior margin reaching the supraorbital ridge. The frontal periosteum is carefully preserved and the subfrontal dura gradually separated from the floor of the anterior fossae. The dissection must reach the posterior limits of the anterior fossa, the posterior edge of the lesser sphenoidal wings, the tuberculum sellae, and the base of the anterior clinoid processes. Dural tears must be closed before the base of the skull and the tumor are resected (Derome, 1977). Through this approach it is now possible to resect lesions involving the roof of the orbit, the ethmoid plate and tuberculum sellae, the sphenoid sinus area, and the clivus down to the anterior arch of C1 and the precervical space.

Reconstruction of the base is necessary before closure to avoid enophthalmos and pulsatility, to reduce dead space and potential encephaloceles, and for cosmetic reasons (Derome, 1977).

Middle Fossa Approach. This operative approach was described by House (1961) for the removal of small acoustic neuromas. In patients with bilateral tumors or with serviceable hearing and intracanalicular tumors, this operation affords the opportunity of preserving hearing (Glasscock, 1978). The authors feel that once tumors extend into the cerebellopontine angle beyond the porus, hearing preservation is best attempted through a suboccipital approach.

Technique. The patient is placed in the supine position with the involved ear upward. The surgeon positions himself at the head of the table. After an injection of lidocaine and epinephrine 1:100.000, an incision is made 1 cm in front of the tragus, and extended from the root of the zygoma to the squamoparietal suture line. The temporalis is divided and the pericranium incised. A temporal craniectomy is performed and the bone plug placed on the back table until the end of the procedure.

The temporal lobe is elevated off the petrous bone and held in place with a House-Urban retractor. Using continuous suction irrigation and a diamond burr, the greater superficial petrosal nerve is followed from the facial hiatus to the geniculate ganglion. The epitympanum may be opened to orient the surgeon, especially in well-pneumatized bone. The

superior semicircular canal is "blue lined" and the facial nerve is followed into the internal auditory canal. Bill's bar is identified with the facial nerve lying anterior and the superior vestibular nerve lying posterior. The dura is incised and the surgical defect widened.

Care must be maintained to avoid injury to the basal turn of the cochlea, which may sit as close as 1 mm to the facial nerve.

After removal of the tumor, muscle is placed over the defect to the internal auditory canal and the epitympanum, and the wound is closed in layers after replacement of the bone plug.

Translabyrinthine Approach. Initially employed by Panse (1904), this procedure was not used extensively until House and Luetje (1979) began to use it routinely for moderate-sized cerebellopontine angle tumors in the 1960s. Since that time it has become an accepted standard procedure for small to moderate-sized tumors in patients who do not have serviceable hearing.

Technique. The patient is placed in the supine position with the involved ear upward. An extended postauricular incision is made and soft tissue is elevated off the mastoid region. A cortical mastoidectomy is performed. The dura overlying the middle fossa is exposed, as is the posterior fossa dura, the sigmoid sinus being decompressed with a bipolar cautery.

The facial nerve is identified in the vertical segment of the mastoid and the facial recess is opened. The incudostapedial joint is separated and the incus removed. The malleus head is removed and the anterior epitympanum exposed.

An organized labyrinthectomy is performed. Neural tissue is removed from the vestibule and the region of the lamina cribrosa superioris (Mike's dot) is noted. Inferiorly, the jugular bulb is exposed, and just superior to this the cochlear aqueduct is opened, further decompressing the cranial vault by draining spinal fluid.

The internal auditory canal is opened medially and the dura incised. At the lateral end of the canal, the superior vestibular nerve is separated from the facial nerve by a wedge of bone (Bill's bar). With a small right-angled hook the superior vestibular nerve with tumor is mobilized, demonstrating the anterior position of the facial nerve.

The tumor is then mobilized from the cerebellopontine angle and removed.

Fat harvested from the abdomen is used to fill the defect, and the wound is closed in layers.

Suboccipital Approach. This technique has been the standard by which neurosurgeons during the 20th century have measured the merits of other procedures for the treatment of cerebellopontine angle lesions (Cushing, 1917; Rand, 1985). Currently, this technique is advocated for all procedures in which tumor extends into the cerebellopontine angle and hearing is sufficient to attempt preservation.

Large tumors can be satisfactorily exposed only by this method, often used in conjunction with a translabyrinthine exposure to optimize nerve VII preservation (see combined approach below).

Technique. Historically the patient was placed in a sitting or semisitting position, but we employ a modified lateral oblique position for the suboccipital approach.

A paramedian incision is made and a suboccipital craniectomy performed. Because of the lateral positioning, generally only mild retraction need be placed on the cerebellum to expose the tumor mass.

The tumor mass is reduced in size and gutted with the CO_2 laser to preserve its capsule until the attendant cranial nerves are identified. Dura is elevated off the petrous ridge and the posterior wall of the internal auditory canal is opened to expose the nerves and tumor within the canal. Generally, nerve VII has been displaced anteriorly, but this is "never for certain". Mobilization of the nerves and tumor laterally toward the fundus optimizes identification of both the facial and cochlear nerves.

Once mobilized, the dissection continues from the brain stem where the root entry zone of the facial nerve is appreciated. After total removal the wound is closed in layers, with muscle placed into the internal auditory canal and bone wax applied to the petrous ridge that was drilled.

Combined Approach. This procedure is used in cases of large tumors in patients with serviceable hearing. The major advantage is that it allows identification of nerve VII out in the most lateral aspect of the internal auditory canal and, if necessary, nerve VII may be appreciated in the labyrinthine segment. The technique does imply that the surgical team work anterior and posterior to the bridge created by the patent sigmoid sinus.

Retrolabyrinthine Approach. Originally described to give access to the nerve V in patients with trigeminal neuralgia (Hitselberger and Pulec, 1972), its most current application is to give access to the posterior fossa for vestibular nerve sections, microvascular decompression, or exploration of the posterior fossa.

Technique. The patient is placed in the supine position with the involved ear up. An extended postauricular incision is made and a cortical mastoidectomy performed. The facial nerve is identified and the labyrinth skeletonized. The bone over the posterior fossa and sigmoid sinus is removed.

With care taken not to incise the endolymphatic sac, the posterior fossa dura is opened anterior to the sigmoid and superiorly at the level of the superior petrosal sinus.

Once exposed the sigmoid sinus may be retracted (with a Silverstein or Jannetta retractor) and the procedure completed.

The dura is closed and the wound ablated with harvested abdominal fat.

Transcochlear Approach. This procedure, described by House and Hitselberger (1976), was designed to approach lesions of the posterior fossa, of the petrous bone anterior to the internal auditory canal, and as far forward as the clivus.

Technique. The patient is placed supine with the involved side upward. An extended postauricular incision is made. A wide cortical mastoidectomy is performed from the zygomatic root anteriorly to the subocciput. Bone over the sigmoid sinus is skeletonized. The facial nerve is identified, and the facial recess is opened and extended after division of the chorda tympani nerve.

Subsequently, with the incus and the malleus head removed, an organized labyrinthectomy is performed. The facial nerve is followed from the stylomastoid foramen to the internal auditory canal.

The greater superficial petrosal nerve is divided from its anterior takeoff at the geniculate ganglion. The facial nerve is then mobilized out of its canal and delivered posteriorly.

The promontory of the basal turn of the cochlea is drilled away to expose the internal carotid artery anteriorly. Superiorly the superior petrosal sinus up to Meckel's cave is exposed, and inferiorly the jugular bulb is exposed.

The dura is opened posterior to the internal auditory canal, and the tumor visualized and extirpated. Closure is performed in similar fashion to that employed for a translabyrinthine closure.

Infratemporal Fossa Approach. Access to the infratemporal fossa was pioneered by Glasscock and colleagues (1974, 1978, 1985) and Fisch and colleagues (1979, 1982, 1985). Current classification schemes for glomus tumors are shown under the section on brain tumors.

Technique. The patient is placed in the supine position with the involved side upward. A large, C-shaped incision is carried from the temporal region, 4 to 5 cm behind the auricle, and gently swept anteriorly into the neck, forming a wide, broadly based anterior pedicle that will elevate, giving exposure to the parotid as well as the vital neurovascular structures in the neck.

The external auditory canal is closed over after being cut at the bony cartilaginous junction. Exploration of the neck identifies cranial nerves IX to XII, the common carotid artery, the internal and external carotid arteries, and the jugular vein. A superficial parotidectomy is performed with clear identification of the facial nerve.

The mastoid is exposed and the posterior canal wall removed. The mastoid tip is removed, as is the styloid process. The facial nerve is mobilized anteriorly from the stylomastoid foramen to the geniculate ganglion. To gain additional access anteriorly, the mandible may need to be mobilized or partially resected.

Tumor dissection follows to the posterior or middle cranial fossae as indicated. The sigmoid sinus is packed off with Surgical and the ligated jugular vein is mobilized with the tumor mass. Intracranial invasion often requires complete labyrinthectomy to gain exposure. At the level of the jugular bulb, bleeding is encountered as the inferior and superior petrosal sinuses are opened and packed off.

Anteriorly, the dissection often follows the carotid artery to the level of the cavernous sinus.

Care should be exercised to protect nerves IX to XI at the jugular foramen if possible. Inferiorly, resection may extend to the level of the foramen magnum; anteriorly, access to the clivus is made possible.

After complete tumor removal the wound is closed with a rotated temporalis muscle fascia flap.

Tumors

Encephalomeningoceles

Encephalomeningoceles are developmental abnormalities that represent limited closure defects of the encephalic neural groove. A cephalic hernia occurs, which may consist of meninges, cerebrospinal fluid, and portions of brain tissue.

Encephalomeningoceles may be divided into the following types: occipital, sincipital, and basal (Blumenfeld and Skolnik, 1965). The majority occur in the occipital region (75 per cent). Of the 15 per cent presenting in the sincipital region, the following variations have been documented: (1) nasofrontal, in which the mass presents in the midline at the bridge of the nose; (2) naso-orbital, in which the mass presents in the anteromedian aspect of the orbit and causes proptosis; and (3) nasoethmoidal (Suwanwela and Horgsaprabhas, 1966), in which the mass distorts the side of the nose at the junction of the nasal and ethmoid bones.

The basal region accounts for only 10 per cent of encephalomeningoceles. However, this group is of greatest interest and importance to this discussion, since they commonly present in the nasal or pharyngeal region, where they may be mistaken for less complex lesions indigenous to the area (Anderson, 1947; Finerman and Pick, 1953). Accurate preoperative definition is necessary in order to prevent recurrence and complications such as spinal fluid rhinorrhoea, which frequently follows attempts at endonasal excision. The following types of basal encephalomeningoceles have been recognized: (1) spheno-orbital, in which the mass presents in the superior orbit via the superior orbital fissure and causes exophthalmos; (2) intranasal, in which the lesion passes through the lamina cribrosa and presents between the middle turbinate and the nasal septum; (3) sphenopharyngeal, in which the mass escapes through the sphenoid bone and presents in the epipharynx; and (4) sphenomaxillary, in which the lesion passes through the infraorbital fissure into the pterygomaxillary fossa and presents as a mass on the medial aspect of the maxilla.

Preoperative differentiation of these masses from other lesions indigenous to the area is sometimes difficult but is necessary for proper surgical treatment. CT scanning has

provided a noninvasive means of delineating the contents and limits of these lesions. The occipital lesions do not usually present a diagnostic problem because of obvious external deformities. The prognosis for these patients is guarded in view of the high incidence of contained brain tissue in the hernia. Most sincipital and basal lesions are isolated lesions and do not preclude normal mental and physical development. However, occasionally they may be associated with other severe developmental anomalies such as microcephaly and hydrocephalus (Mealey and colleagues, 1970). If there is no gross deformity, an anterior lesion should arouse suspicion in the presence of a nasal or paranasal mass in early life, widening of the nasal bridge, hypertelorism, and (in the case of an intranasal mass) attachment between the middle turbinate and the nasal septum (an unlikely site for polyp formation).

Encephalomeningoceles demand intracranial surgical treatment with correction of the dural defect. Treatment in early infancy may be indicated for rapid enlargement of the lesion; encroachment on eyes, nose, or mouth; danger of rupture or infection; and associated hydrocephalus (Whatmore, 1973). Otherwise, definitive surgical treatment may be deferred in order to facilitate any requisite reconstructive surgery.

Nasal Gliomas

Nasal gliomas that have no direct connection with the central nervous system may originate from embryonic nests that may have been isolated from the frontal lobe by closure of the cranial sutures during embryonic development, or may arise from the primitive olfactory membrane (Mood, 1938). In most instances, there are neoplasms that are of low growth potential and can be eradicated by local excision (Black and Smith, 1950). However, the olfactory nerve neuroblastoma, a highly malignant neoplasm that arises from the olfactory apparatus (Holland and colleagues, 1959), erodes the floor of the frontal fossa. This lesion may present as a mass in the nose or cranial cavity (Robinson and Solitare, 1966). It must be treated by radical excision and high-voltage radiotherapy, to which it is highly sensitive. Despite this regimen, distant metastases and limited survival are the common results (Hutter and colleagues, 1963).

Craniopharyngiomas

The diagnostic and therapeutic problems associated with craniopharyngiomas remain controversial. These tumors, which can be solid or cystic, arise as a proliferation of the squamous cell nests in the superior region of the anterior lobe of the pituitary gland (Matson, 1969). Despite the characteristic location of this tumor in the parasellar region and its predilection for children, the diagnosis of craniopharyngioma is not always easy because of the frequent occurrence of other tumors in the same region that may have similar presentations, ie, optic gliomas and diencephalic tumors. The clinical presentation is usually a result of compression of adjacent neural structures. Therefore, headache, visual disturbances, and endocrine dysfunction can all be presenting symptoms. The sella turcica is eroded or enlarged in 60 per cent of all patients (Kahn and colleagues, 1973). Parasellar calcifications are present in 80 per cent of children and approximately 60 per cent of adults with this tumor.

The diagnosis of a craniopharyngioma is made by the use of skull films, cerebral angiography, and CT. However, the diagnosis may sometimes be made at the operating table

because of the similar radiographic features that may be seen in other tumors of this region. CT has virtually eliminated the use of the pneumoencephalogram in studying these tumors. The latter test is now reserved for patients in whom there is difficulty in determining the amount of suprasellar extension of the tumor.

Treatment of this tumor is one of the burning controversies in neurosurgery. Matson and Crigler (1960, 1969), Sweet (1968a), and Shillito (1976) advocated radical surgical extirpation with an attempt at total removal. In the event of symptomatic recurrence as documented by radiographic studies, the patient is referred for radiation therapy (Shillito, 1976). If there is symptomatic recurrence, reoperation for total removal is performed, followed by radiation treatment. Kramer (1976) recommended total removal only when the tumor is small and easily encompassed. Otherwise, he believed that surgery should be used for biopsy and decompression, to be followed by irradiation at doses of 5500 to 6500 rads, depending on the age of the patient. Most surgical approaches consist of transfrontal craniotomy. The use of the operating microscope has decreased the morbidity and increased the chances of total tumor removal. The transsphenoidal approach has been proposed for excision of lesions confined to the sella (Rathke's cleft cyst) and for drainage of recurrent cystic lesions. With the advent of stereotaxic methods, intracystic implantation of phosphorous 32 has been successfully used to treat recurrent cystic craniopharyngioma. The efficacy of these various modes of treatment is under study at the present time.

Pituitary Tumors

Tumors of the pituitary gland, along with acoustic nerve tumors, are responsible for most of the collaboration between neurosurgeon and otolaryngologist in recent years. The resumption of the transsphenoidal surgical approach to these lesions has, in our opinion, been one of the most significant contributions to contemporary neurosurgery.

Pituitary adenomas have classically been divided into three types: (1) chromophobic, (acidophilic), and (3) basophilic (Russell and Rubenstein, 1963; Martins and colleagues, 1965). Chromophobic adenomas are the most common and have been considered to be nonsecretory. However, electron microscopic studies of these tumors show secretory granules in the chromophobe adenoma. At the present time the classification of these tumors has been reorganized (Landolt, 1978; Wilson, 1984).

Clinically, pituitary tumors may present because of abnormal endocrinologic function (panhypopituitarism, acromegaly, amenorrhea, galactorrhea) or because of direct extension to contiguous structures (optic pathways, third ventricle, hypothalamus, cavernous sinus).

The surgical approach to pituitary tumors is usually determined by the presence and degree of extrasellar extension of the tumor (Hardy and colleagues, 1976). This is determined by a complete diagnostic workup, which includes a combination of (1) plain roentgenographic films of the sella, (2) tomograms of the sella (especially for microadenomas), (3) CT scan, and (4) angiography (to rule out a vascular anomaly).

In the early years of neurologic surgery, a large ballooned sella turcica was regarded as the classic sign of a pituitary tumor. Oscar Hirsch, pioneering Viennese otolaryngologist (1910), was quick to recognize that these lesions could be treated by transnasal intracapsular

removal. Indeed, because of the initial high mortality associated with intracranial removal of pituitary tumors, Harvey Cushing (1912) employed a modification of this approach for many years until he accumulated experience that decreased the complications associated with the intracranial approach. He became convinced that greater return in visual function could be achieved by transfrontal extirpation of these tumors.

Analysis of Cushing's large and expertly documented series (Henderson, 1939) showed little difference between the mortality involved in the two approaches; however, the incidence of recurrence even with radiotherapy was considerably greater after transsphenoidal extirpation. Despite this disadvantage, others continued to employ the transsphenoidal route (Guiot and Thibaut, 1959; Hamberger and colleagues, 1961; Jamlin, 1962; Bateman, 1962; Montgomery, 1963; Svien and Litzow, 1965). In this regard, one must cite the tremendous contribution of Hirsch (1959), which has extended over a half-century. With the advent of sophisticated contrast studies, improved radiotherapy techniques, and the unique contribution of the microsurgical techniques (Hardy and Wigser, 1965; Guiot and colleagues, 1962) employing radiographic control, the disadvantages of the transsphenoidal technique have been minimized. Indeed, the transsphenoidal approach might be considered in (1) the aged or debilitated individual who tolerates intracranial surgery poorly; (2) those on the verge of blindness in whom manipulation of the optic nerve from intracranial approach might increase the deficit; (3) patients with cystic or acutely hemorrhagic tumors; (4) individuals with lesions confined to the sella turcica or sphenoid sinus; or (5) those with recurrent cystic or solid lesions causing compression of the optic nerves or chiasm, in whom decompression by the sphenoidal route can be safely achieved (Ray and Patterson, 1962).

The major benefits of the transsphenoidal approach include the low morbidity and the ability to treat endocrinologically active tumors successfully while they are confined to the sella turcica. Preservation of normal pituitary function can be expected in most patients, and reversal of sterility has been documented in many case reports. The excitement created by the advances in the early recognition and treatment of secretory pituitary tumors is an experience shared by many fortunate patients. Advocates of this technique reported large series with excellent results (Laws, 1977; Wilson and Dempsey, 1978; Tindall and colleagues, 1978).

It should be noted that the transsphenoidal surgical approach has been effective and safe in the treatment of other pathologic states. Metastatic carcinoma, diabetic retinopathy, craniopharyngiomas, clival chordomas, and cerebrospinal fluid (CSF) fistulas have all been approached by this technique (Laws, 1977; Collins, 1971; Ciric and Tarkington, 1974; Hardy and Ciric, 1968).

Intracranial Aneurysms

The initial intracranial course of the internal carotid artery through the cavernous sinus places it adjacent to the sphenoid sinus and the anterior clinoid of the sella turcica. Therefore, aneurysms of the internal carotid artery, particularly those within the cavernous sinus where they are unlikely to produce subarachnoid hemorrhage, may present as a mass lesion that is indistinguishable from tumors of the parasellar region (Cushing, 1912). Because of the disastrous events that may follow failure to recognize aneurysms, bilateral carotid angiography or digital subtraction angiography should be completed prior to definitive therapy for all parasellar lesions in which the diagnosis is unclear. Advances in computerized brain scanning

have helped clarify most cases that previously proved to be diagnostic problems. This concept is particularly relevant in patients undergoing radiotherapy and in those requiring exploration of the sella turcica via the transsphenoidal approach. It may be impossible to differentiate the progressively enlarging intrasellar aneurysms from tumors that produce a characteristic expansion and erosion of the sella turcica. This is well documented by the case of Ellen O'B, a 61-year-old woman whose radiographs were reproduced by Holmes and Robbins (1955) as showing a typical example of erosion and expansion of the sella turcica by a pituitary adenoma. Because of progressive deterioration in pituitary function and visual fields, the patient underwent a transsphenoidal approach to the sella turcica by Drs. Hirsch and Hamlin. However, when the floor of the eroded sella turcica was entered, a gush of arterial blood was controlled with difficulty, and subsequent angiography demonstrated a 3x2.5 cm aneurysm filling the sella turcica.

In their outstanding study of the subject, White and Ballantine (1961) reported three personal cases and 22 cases communicated by their colleagues. They emphasized the importance of careful preoperative studies, particularly angiography, if the transsphenoidal approach is to be employed.

Aneurysm may be suspected by circular calcification in the aneurysmal wall (Zollinger and Cutler, 1933) and erosion of the outer wall of the optic foramen (Jefferson, 1955b). However, even with angiography, aneurysms may be overlooked because of thrombus in the lumen of the aneurysm (Rhonheimer, 1959). Clinical features of supraorbital pain, unilateral blindness, diminished sensation of the forehead, and ophthalmoplegia (cavernous sinus syndrome) suggest an infraclinoidal aneurysm.

It is important for the surgeon to recognize these aneurysms, since early treatment may reverse the neurologic deficit. Proper therapy for aneurysms in this location may require common carotid ligation in the neck or balloon occlusion rather than direct surgical treatment (Jefferson, 1937; Gallagher and colleagues, 1956).

Malignant Tumors of the Nasopharynx and Paranasal Sinuses

Carcinoma of the nasopharynx and paranasal sinuses frequently presents with neurologic symptoms, particularly if early localized symptoms such as obstruction of the eustachian tube and sinus ostia are overlooked. Focal invasion of the orbit, the superior orbital fissure, and the base of the skull may produce atypical facial pain (see later section on pain), proptosis, ocular palsies, and cranial nerve palsies. A study showed that 25 per cent of patients had one or more cranial nerve palsies; 12 per cent presented with persistent facial pain (Lederman, 1966). Attention should be directed toward early signs, since the prognosis is extremely poor when the tumor has invaded the nervous system (Shedd and colleagues, 1967). In addition, one must recognize the more benign nature of the nasopharyngeal fibroma, which is locally invasive but may be totally removed even if the orbit and base of the skull have been invaded (Hall and Wilkins, 1968).

Malignant tumors of the nose and paranasal sinuses have a more favorable prognosis than those of the nasopharynx. Perhaps this is related to earlier recognition and more radical surgical treatment (Oliver, 1967). Smith and co-workers (1954), Ketcham and associates (1966), and Leffall and White (1966) advocate radical resection of tumors of the sinuses; they

propose that recurrence frequently is the result of invasion of the base of the frontal fossa in the region of the cribriform plate. Early attempts to achieve radical resections through a transfacial approach resulted in an extremely high incidence of complications and led to a combined craniofacial approach in which the entire cribriform plate, sphenoid and ethmoid sinuses, and orbit, if necessary, could be resected (Ketcham and associates, 1966; Van Buren and associates, 1968). To avoid injury to the basal dura, the orbital surface of the frontal lobe, and the optic nerves, a neurosurgical team reflects the dura from the frontal fossa and cribriform plate, where it is extremely adherent. Through a small frontal craniectomy, the frontal sinus is exenterated, and osteotomy of the planum sphenoidale is extended until adequate resection is achieved. The procedure is then continued by the maxillofacial team, who complete the radical maxillectomy and orbitectomy. Following this procedure, it is essential that any rents in the basal dura be closed with care to avoid cerebrospinal fluid fistula. Dural grafts are poorly tolerated in this situation, in which vascularity is marginal and contamination is certain.

The extensive experiences of Van Buren and co-workers (1968), Ketcham and associates (1966), and Leffall and White (1966) indicate that this radical procedure has distinct value and may be performed with acceptable morbidity.

Chordoma

The chordoma is a rare, invasive tumor that arises in the basal portion of the sphenoid and occipital bones. It originates from the notochordal remnants in these bones or at the spheno-occipital synchondroses (Friedman and colleagues, 1962; Falconer and colleagues, 1968), and erodes the clivus and adjacent bone to present in the sphenoid sinus (Harrison, 1961) or nasopharynx (Ormerod, 1960; Batsakis and Kittleson, 1963; Wright, 1967). Simultaneous intracranial spread usually occurs, so that invasion of the dura and distortion of strategic structures at the base of the brain interfere greatly with radical surgical removal.

Clinical presentation depends on the direction of local extension. Danzinger and co-workers (1974) pointed out that the position of the tumor on the clivus correlated well with the direction of extension and with signs and symptoms. Lesions arising high on the clivus show forward extension into the sphenoid sinus and may result in visual impairment. Lower clivus lesions disrupt posterior fossa dura and result in multiple cranial nerve palsies and brain stem and cervical cord signs. The invasion of bone and dura makes headache a common complaint.

For lesions presenting in the sphenoid and nasopharynx, the transsphenoidal approach offers certain advantages: biopsy and partial resection may be performed with ease (Wright, 1967; Guiot, 1967; Hardy, 1969). Similarly, the transcervical-transclival approach to the ventral surface of the brain is of value in the radical attempt to extirpate the tumor (Stevenson and colleagues, 1966). Guthkelch and Williams (1972) described a transpalatal approach to clival chordomas designed to improve exposure of the posterior wall of the nasopharynx.

Although the ultimate outlook for patients with intracranial chordoma is poor, the tumors are slow growing and they create a bulky mass that is placed ventral to the brain stem and cranial nerves. Thus, radical intracapsular removal may result in great symptomatic improvement and significant prolongation of the patient's life (Poppen and King, 1952;

Falconer and colleagues, 1968). High-voltage radiotherapy and intracavitary radiation have been advised but are of unproved value (Zoltan and Fenyes, 1960; Wright, 1967; Falconer and colleagues, 1968). A few instances of metastatic chordoma were documented, but the histologic appearance of these lesions was benign (Russell and Rubinstein, 1963).

Fibrous Dysplasia

Fibrous dysplasia, an osseous hyperplasia of unknown etiology, may involve the craniofacial bones (Schlumberger, 1946). The disorder is characteristically monostotic in form, and either the frontal or the sphenoid bone is involved in most cases (Ramsey and colleagues, 1968). The feminine sex preponderance, sexual precocity, and skin lesions associated with the polyostotic form (Turner's syndrome) are seldom encountered in association with the monostotic form. The disorder occurs during childhood and becomes inactive when skeletal growth is completed. The presenting findings are ptosis, exophthalmos, diminution in visual acuity, diminished hearing, and cosmetic deformity. The radiographic picture is of marked thickening and increased density of the involved bone. In 50 cases reported by Sassin and Rosenberg (1968), the frontal and sphenoid bones were involved in 50 per cent of patients; the optic canal was involved in 20 per cent of patients, most of whom had severe visual deficits secondary to optic nerve compression. Hearing loss and tinnitus secondary to temporal bone dysplasia have been reported (Basek, 1967). Occasionally, dysplasia of the sphenoid and frontal bone produces an intracranial lesion as well as an orbital mass. Increase in the serum alkaline phosphatase level and increased radioactive uptake by the bony lesion (isotopic brain scan) are confirmatory laboratory findings. CT has also become a valuable tool in delineating the bony abnormalities.

Conservative treatment is indicated, ie, curettage and partial resection, unless complete resection can be achieved without creating a significant cosmetic and functional deficit. In the case of optic foramina encroachment, careful serial evaluation of visual acuity and visual fields is mandatory. If progressive deterioration occurs, surgical treatment consists of removal of the orbital roof and the medial wing of the sphenoid until the optic nerve is free in its canal (Matson, 1969). Similar but less extensive decompression may be required in cases of progressive exophthalmos. Surgical treatment for cosmetic reasons alone has seldom been necessary.

Aneurysmal Bone Cysts

Aneurysmal bone cysts, commonly found in long bones, are rare lesions that occasionally involve the bones of the cranium and base of the skull. The facial bones, mandible, and maxilla are more frequent hosts for this benign, although highly vascular, lesion, which is characterized by destruction and expansion of the parent bone (Lichtenstein, 1957).

The radiographic picture consists of central rarefaction, expansion of the bone, and preservation of a cortical shell of bone outlining the mass lesion.

The lesion commonly presents as a cosmetic deformity of the face. However, we saw one patient in whom a large mass expanded into the temporal fossa and produced a severe neurologic deficit. A similar case was reported by Constantini and co-workers (1966).

Therefore, in lesions of the cranial bones, careful preoperative study is indicated to exclude intracranial spread.

Surgical treatment consists of radical curettage and packing of the cavity with cancellous bone chips. The highly vascular nature of this tumor may make it a formidable surgical challenge (MacCarty and colleagues, 1961).

Tumors of the Posterior Cranial Fossa

The posterior fossa is a relatively small and closed compartment of the intracranial vault. The anatomic structures contained therein are of primary interest to both neurosurgeons and otolaryngologists. The intimate relationship of the auditory, vestibular, and labyrinthine apparatus to other neural structures brings the two specialists together as a working team more often than does any other anatomic region.

Acoustic Nerve Tumors

Cranial nerve VIII is the site of tumor origin more frequently than any other cranial or spinal nerve. The tumor arises from the sheath of the nerve and has been variously classified as a neuroma, neurinoma, or neurilemoma. In addition, however, it has become synonymous with a term that is incorrect, ie, tumor of the cerebellopontine angle.

The acoustic nerve tumor arises from the Schwann sheath, cells of which cover the axons from a point at which the latter penetrate the pia mater to their termination. The point of penetration of the leptomeninges varies greatly with different nerve roots and, in the case of the auditory and vestibular nerves, the distance may be as great as 1 cm (Skinner, 1963). This fact may well account for the frequency with which acoustic tumors arise within the internal auditory canal (Henschen, 1963), where the nerve root usually acquires its reticulin and Schwann cell components. In this location the expanding tumor causes erosion of the internal auditory meatus prior to escape of the mass into the cranial cavity. Indeed, we have designated these lesions as tumors of the ear or of the brain.

Grossly, these tumors are well encapsulated. There may be cystic enlargements, with necrotic or hemorrhagic centers. The vascularity is variable, as is the fatty density. Not uncommonly, calcium specules are noted. In addition, some acoustic neuromas are mobilized quite readily, while others, irrespective of size, may be firmly adherent to adjoining neural and vascular structures.

Histologically, two common variants are described, Type A Antoni cells are characterized by a dense, fibrillar cytoarchitecture; type B Antoni cells are characterized by a less dense cell population and a loose reticular pattern.

Several histologic observations raise questions regarding the best method of treatment. In studying the neuronal population and relationship to tumor infiltration, Neely (previously cited by Nager, 1964) found that the tumor generally arises from the vestibular division of nerve VIII, in the neurilemmal segment lateral to the glial-neurilemmal junction. He noted that in 21 of 22 patients tumor filled the internal auditory canal to the crista falciformis or beyond. Histologic observations confirm that in some instances (3 of 15 cochlear nerves, 5

of 13 superior vestibular nerves, and 6 of 6 nerve trunks) in which there appeared clinically to be no tumor, there was in fact tumor on the histologic appearance.

Tinnitus, unilateral sensorineural hearing impairment, and mild unsteadiness are the early symptoms of the small tumors that are still confined to the internal auditory canal. We personally have not had a patient with an acoustic tumor complain of episodes of true vertigo, although such cases have appeared in the literature (Hitselberger, 1967; Rand and Kurze, 1968; Sheehy, 1968). It should be noted that the vestibular responses may well be normal in these small acoustic tumors. This presents a problem in differentiating these tumors from Ménière's or other cochlear diseases. As the tumor grows and expands into the cranial cavity, neurologic symptoms and signs of a cerebellopontine angle mass appear. Diminished sensation of the cornea and face indicates compression of the trigeminal nerve. Remarkably, a facial nerve paresis is seldom seen, even when the tumor becomes very large (Mack, 1968). However, sophisticated clinical tests may show disturbances in taste, hearing, and sensation of the posterior wall of the external auditory canal (House, 1964). Other cranial nerves may become involved as the tumor expands, and extraocular movement disorders may occur if the tumor ascends into the incisural notch. The tumor may expand to compress the cerebellum and brain stem, resulting in nystagmus, ataxia, spasticity of gait, and pyramidal tract signs. Finally, obstruction of the aqueduct produces hydrocephalus and papilledema as a late finding. Tew and co-workers (1974) staged these tumors according to their size and the amount of extension into the cranial cavity. They found that this correlated with the radiographic findings but not necessarily with the clinical presentations. That is, it is possible for even a very large tumor to cause minimal signs and symptoms.

The diagnosis of acoustic nerve tumors has undergone much improvement because of the widespread use of CT and the contributions of modern otology. Otolaryngologists now have the ability to diagnose the presence of a lesion while it is still contained inside the canal and is causing only auditory and vestibular symptoms and signs, and before the characteristic radiographic findings appear. The usual battery of diagnostic tests includes tomography of the petrous bones and internal auditory canal, posterior fossa angiography, and CT of the head. Occasionally, posterior fossa air myelography is used to prevent negative exploration or to help plan a surgical approach (Baker, 1963; Scanlan, 1964; Hitselberger and House, 1968).

Consideration of the surgical treatment of acoustic tumors should begin with a review of the history of the tumor. This falls into three definite periods of development: (1) Cushing, (2) Dandy, and (3) House.

Cushing, whose experience (dating from 1890 to 1933) and skill with brain tumors were both extensive and consummate, was convinced that the large tumors he was called on the deal with could not be removed totally, even though they were known to be benign. This view was based on overwhelming mortality and morbidity rates when total removal was undertaken. Accordingly, he advised intracapsular enucleation of the lesion as a palliative procedure.

Dandy followed this principle for a time. However, early recurrence and an overwhelming mortality rate after secondary operation (55 per cent of patients dead within 5 years) led him to undertake total removal in all cases, and he achieved a surgical mortality rate of 10 per cent.

After Dandy's (1925) epoch-making publications in this field, all neurosurgeons began to undertake total removal as a primary procedure for tumors of this type, with a surgical mortality rate ranging from 5 to 15 per cent. Preservation of the facial nerve in this type of procedure was rare, but subsequent operations in which the facial nerve was replaced by a graft with the hypoglossal nerve and the spinal accessory nerve usually provided an acceptable, if not totally satisfactory, result.

House (1968), already a master with the surgical microscope in procedures involving the temporal bone, encountered and removed a small acoustic neuroma in the internal auditory canal of a patient who was being treated for Ménière's disease. Recognizing the probability that tumors of this type might be the cause of unilateral deafness (sensory neural deficit) and tinnitus, and proceeding with characteristic vigor and skill, House confirmed that the lesion is indeed common. He also found that the tumor can be recognized in many instances while it is still small and can be removed through the ear totally and safely. The observations of House and associates (1979) stimulated many otolaryngologists and neurosurgeons to look anew at this problem; in a sense, a new specialty - neuro-otology - has developed. The selection of the most favorable method of approaching acoustic neuromas (transaural or transcranial) is one of the many factors that they are attempting to solve.

At the present time there are essentially three operative approaches to acoustic tumors: (1) translabyrinthine, (2) subtemporal, and (3) suboccipital. The translabyrinthine approach was first described by Panse in 1904 (Buchheit and Johnson, 1977). Those acquainted with the approach recommend it for tumors of moderate size. Unfortunately, it precludes preservation of any auditory function (Hitselberger and House, 1966), since the otic capsule is destroyed. More recently, Ojemann and co-workers (1972) and Yasargil and Fox (1974) published their series using the combined approach (translabyrinthine and suboccipital).

The subtemporal, or middle fossa, approach is reserved for tumors confined to the auditory canal. It is felt that this approach affords the best chance to preserve, and in some instances to improve, neurologic function when only auditory and vestibular symptoms are present (Hitselberger and House, 1966). It is our opinion that this approach is rarely indicated. However, with the advent of more sophisticated audiologic testing, these tumors will be diagnosed at this stage more frequently.

The suboccipital approach as conventionally performed remains (in the authors' opinion) the procedure of choice for any acoustic tumor that has appreciable intracranial extension. This is because the opportunity for total removal of a tumor with an intracranial component is greater with the suboccipital approach owing to better exposure. It is our opinion that because of microsurgical techniques and new lateral oblique positioning of the patient, the morbidity and mortality rates have been minimized. It is felt that with the microsurgical technique the anatomy of the cerebellopontine angle (Rand, 1985) can be adequately exposed and visualized to allow for preservation of neural function including hearing. Therefore, for all but very small intracanalicular tumors, we attempted complete removal by the suboccipital approach, employing microsurgical technique (Dandy, 1925; McKissock, 1965; Drake, 1967; Olivecrona, 1967; Rand and Kurze, 1968).

It is obvious that the treatment of this tumor and others in this location brings us to a neuro-otologic cross-road that in future years may provide data for the best treatment of this

benign, but devastating, disease (DiTullio and colleagues, 1978).

Other Posterior Cranial Fossa Tumors

Trigeminal Neuroma. This relatively rare tumor characteristically presents as a trigeminal nerve pain syndrome. The mass may be of variable size, which does not correlate with the severity of symptoms. Symptoms may include head and face pain associated with sensory and motor dysfunction of the trigeminal nerve. The authors treated one patient who had intractable facial pain for 2 years and paralysis of cranial nerve V for 6 months. During these 2 years, cerebral angiography, posterior fossa myelography, tomograms of the base of the skull, and repeated CT scans were normal. Exploratory subtemporal craniotomy demonstrated a 2.5-cm tumor infiltrating the gasserian ganglion. This diagnosis should be considered in all patients with atypical facial pain.

Meningioma. Although the acoustic nerve tumors account for 85 to 90 per cent of neoplastic lesions arising in the area of the cerebellopontine angle, tumors of diverse origins occur in this location. In his original monograph on meningiomas, Cushing and Eizenhardt (1938) recognized that meningiomas arose in the cerebellopontine angle; in several instances they were unable to distinguish the clinical picture from that of the acoustic nerve tumor. Indeed, in their monograph in which 200 acoustic nerve tumors were reported, House and colleagues (1968) found 13 meningiomas. It is difficult to distinguish meningiomas clinically from tumors of cranial nerve VIII. It is important to note, however, that plain radiographs of the skull may show an osteoblastic reaction in the petrous portion of the temporal bone, a feature characteristic of intratemporal meningioma. Occasionally, marked bone destruction may also occur (Nager, 1964).

The CT appearance of a meningioma may be identical to that of an acoustic nerve tumor if it is present in the cerebellopontine angle. In such cases the angiographic studies may be diagnostic if a vascular stain can be demonstrated. In addition, if there is no erosion of the internal auditory meatus, meningioma is more likely. Treatment of these tumors consists of total removal whenever possible through a suboccipital approach. All other approaches are inadequate; therefore, preoperative planning for this lesion is valuable.

Cholesteatomas. These benign tumors may be of two types. Congenital cholesteatomas arise from embryonic rests of epidermal tissue in the petrous bones. Secondary cholesteatomas result from reaction to chronic mastoid infection. These lesions usually present with progressive sensorineural deafness and facial paralysis (Olivecrona, 1949; Cawthorne and Griffin, 1961). The CT scan shows low density, which should eliminate acoustic tumor or meningioma as a possibility. Tomograms demonstrate marked erosion of the petrous bone. Cholesteatomas should be removed surgically in case life-threatening neural compression results.

Glomus Tumors. These uncommon tumors, referred to as paragangliomas, are derived embryologically from neural crest elements. First described by Guild (Glasscock, 1982), glomus bodies may be found closely attendant to the carotid artery and jugular vein; those associated with the latter are in intimate juxtaposition to the path of Jacobson's nerve.

Because they lack the positive response to chromaffin staining associated with neural crest tumors of the adrenal, glomus tumors have been called nonchromaffin paragangliomas. Vasoactive substances have been identified (Farrior and colleagues, 1980; Jackson, 1981) as a chemical concomitant with the tumor syndrome. Jackson and colleagues (1982) reported that a multicentric origin may be present in upward of 10 per cent of cases. Furthermore, both pheochromocytoma and thyroid carcinoma have been associated with glomus tumor development.

Symptoms are referable to both local and regional extension (Alford and Guilford, 1962; Glasscock and colleagues, 1974; Brammer and colleagues, 1984). Involvement of the middle ear space produces a conductive hearing loss and pulsatile tinnitus. Progressive growth may eventually erode the otic capsule, producing cochlear and labyrinthine dysfunction. Cranial neuropathies may be evidenced as nerves VII and VIII become involved in the internal auditory canal or nerves IX, X, and XI become disrupted at the jugular foramen. Rarely, extension to the hypoglossal canal or extension to the petrous apex involves nerve XII, and V or VI, respectively. In addition, growth of tumor into the posterior cranial fossa is appreciated more commonly than involvement of the middle cranial fossa, although both may occur.

Clinically, a blue-red mass behind or protruding through the tympanic membrane may be seen on otoscopic examination. The blanching of the tumor mass (Brown's sign) with pneumatic otoscopy is often seen with this type of tumor. When observed, the mesotympanic and hypotympanic elements may be just a small demonstration of the eventually detected tumor mass.

The diagnosis of glomus tumors can be made definitively on biopsy only. However, when there is a high degree of suspicion, most surgeons prepare the patient for total extirpation before obtaining a tissue diagnosis. As with other lesions of the skull base involving the temporal bone, audiometric testing, including Brain Stem Evoked Response (BSER), vestibulometric studies with electronystagmography, and CT scanning are the diagnostic tools most commonly employed. It is important to request bone density-enhanced windows when reviewing skull base bony pathology. Furthermore, because of the vascular nature of these tumors, carotid angiography with crosscompression is mandatory before surgery (Glasscock and colleagues, 1985). The use of nuclear magnetic resonance imaging (MRI) is currently being evaluated.

To obviate the possibility of an intraoperative or postoperative hemodynamic crisis due to vasoactive substance release, all patients are preoperatively screened with a 24-hour urine collection for vanillylmandelic acid (VMA), metanephrine, and normetanephrine. Some work suggests that there may be other neuropeptides secreted by these tumors, such as VIP vasoactive intestinal peptide (VIP) (Pensak and colleagues, 1984).

Tables 1 and 2 list the two most commonly employed classifications tumor location and relative size (Fisch, 1982; Jackson, 1981).

Table 1. Fisch Classification of Glomus Tumors

Type A. Tumors confined to middle ear space.

Type B. Tumors confined to mastoid and middle ear; no infralabyrinthine involvement.

Type C. Tumors extending to infralabyrinthine region of temporal bone and petrous apex.

Type D1. Tumors with less than 2 cm diameter intracranial extension.

Type D2. Tumors with greater than 2 cm diameter intracranial extension.

Table 2. Glasscock-Jacobson Classification of Glomus Tumors

Glomus Tympanicum

I. Small mass limited to promontory.

II. Tumor completely filling middle ear space.

III. Tumor filling middle ear and extending into mastoid.

IV. Tumor filling middle ear, extending into mastoid or through tympanic membrane to fill EAC; may also extend anterior to internal carotid artery.

Glomus Jugulare

I. Small tumor involving jugular bulb, middle ear, and mastoid.

II. Tumor extending under IAC; may have ICE.

III. Tumor extending into petrous apex; may have ICE.

IV. Tumor extending beyond petrous apex into clivus or intra-temporal fossa; may have ICE.

IAC, Internal auditory canal; ICE, intracranial extension; EAC, external auditory canal.

The current treatment of glomus tumors is aimed at complete removal. For intratympanic glomus tumors a transtympanic or postauricular approach is sufficient. For larger lesions, especially with intracranial and skull base extension, varying forms of the infratemporal fossa procedure earlier described in this chapter are employed. The indication for embolization of the tumor before surgery is based on a reduction of the blood supply to

both the tumor mass and the neighboring surgical field. The concomitant employment of this modality varies from institution to institution (Glasscock and colleagues, 1985). Currently at the University of Cincinnati we do not use radiation therapy to treat glomus tumors.

Miscellaneous Tumors. A number of other space-occupying lesions in the posterior fossa may come to the attention of the otolaryngologist. Exophytic brain stem gliomas (Matson, 1969), aneurysms of the anterior cerebellar artery (Drake, 1968), arachnoid cysts (Tew, 1967), and metastatic tumors may all present in the cerebellopontine angle and other portions of the posterior fossa. Therefore, one must be aware of these possibilities and be prepared to deal with them appropriately.

Cranial Trauma

Craniocerebral trauma is a major cause of death and disability in the USA. A brief neurosurgical overview of this area is therefore warranted. Attention will be given to problems of otologic significance.

Skull Fractures

There are basically three types of skull fracture.

Linear Skull Fractures. These are usually of no clinical significance unless they traverse a vascular groove, ie, the middle meningeal artery. Vascular injury enhances the possibility of subsequent development of an intracranial hematoma. If loss of consciousness occurs, serious considerations should be given to hospitalization of the patient for skilled observation.

Depressed Skull Fractures. These are more significant because when the bone fragments project inward there is likelihood of dural and cortical laceration. This complication increases the risk of hematoma formation and post-traumatic seizures. It is generally accepted that a fracture with 1 cm of bony depression should be elevated, particularly if there is a focal neurologic deficit that correlates with the fracture site. *A compound fracture,* whether linear or depressed, demands immediate debridement and antibiotic therapy, especially in the presence of contaminated wounds.

Basal Skull Fractures. These represent a final type of bone disruption that is not readily seen on x-ray studies, including basal views.

The vault of the adult skull is composed of dense, strong, laminated, and resilient membranous bone. The base of the skull is composed of relatively inelastic cartilaginous bone. The base is not subject to direct force and hence can be fractured only when the vault undergoes considerable deformation. Since the base of the skull is fragile and houses important structures such as the carotid arteries, cranial nerves, and brain stem, fractures in this region may assume particular importance. Fractures of the base are frequently difficult to visualize radiographically, particularly by routine techniques. There are certain clinical signs that identify a fracture of the base of the skull that otherwise might not be apparent. Bleeding from the auditory canal or from the nose or mouth without evidence of direct injury to the part usually is an expression of basal skull fracture.

At times ecchymosis may appear over the mastoid (Battle's sign) several days after head injury. This presumably occurs as a result of bleeding into the tissue about the base of the skull adjacent to a fracture site. The same is true of ecchymosis about the orbits, which appears as a result of extravasation of blood into the facial tissues after fractures that traverse the anterior fossa and has been referred to as "panda eyes". It is of critical importance to recognize the leakage of cerebrospinal fluid from the nose, eustachian tube, or external auditory canal is indicative of a basal fracture in either the frontal or posterior cranial fossae.

The basal fractures are often associated with cranial nerve injuries, including cranial nerves II, VII, and VIII. These fractures are therefore of more otologic interest than the others.

The emergency evaluation of head trauma is one of the areas in which the CT scan has contributed most. Being a fast, noninvasive diagnostic tool, it allows the physician to diagnose life-threatening intracranial lesions earlier than previously. Epidural, subdural, and intracerebral hematomas are discovered and treated in shorter time periods, thus increasing the chance of survival. In addition, severe head injuries not requiring surgery, such as cerebral contusion and hemorrhagic contusions, can be followed with serial CT scans. CT scanning, enhanced by the addition of intracranial pressure monitoring, permits judicious but aggressive treatment with mannitol, high-dose steroids, and barbiturates, all of which are being applied in the hope that the prognosis of severe head injuries can be improved (Becker and colleagues, 1977; Bruce and colleagues, 1978a, b; Smith and colleagues, 1974).

Cranial Nerve Injuries Associated with Skull Fractures

The cranial nerves are often injured in fractures of the base of the skull that involve their foramina. Anosmia occurs secondary to disruption of the olfactory nerve filaments as they traverse the cribriform plate. Indeed, the olfactory nerve is the most commonly injured cranial nerve in craniocerebral trauma (Turner, 1943). The optic nerve is rarely injured in basilar fractures, but any injury is seldom recognized early because of the difficulty of testing vision in the severely injured patient with depressed consciousness and facial swelling (Hughes, 1962). Oculomotor palsy is uncommon in the absence of orbital fracture; however, abducens palsy is frequent but has little or no localizing value in the head-injured patient.

Facial Nerve Injuries. Peripheral facial paralysis is a common occurrence following fracture of the temporal bone. The site of injury to the nerve appears to be related more to the structure of the temporal bone than to the nature of the head injury producing the fracture (McHugh, 1963). The most frequent site of injury to the nerve is within the middle ear, where it is crushed against the underlying labyrinthine capsule or contused and compressed by the shattered fragments of the tegmen tympani and the thin bone overlying the facial canal (McHugh, 1959). In transverse fractures of the temporal bone, the facial nerve is frequently lacerated or severed, and facial paralysis is immediate in onset. In longitudinal fractures, by far the more common type, the nerve is more likely to be contused, compressed, or stretched, and accordingly the paralysis of the facial muscles may be delayed in onset. In the case of transverse fractures in which the nerve is severely injured and paralysis is immediate in onset, recovery is unlikely in the absence of operative decompression and intrapetrous anastomosis, as advocated by Ballance and Duel (1932). However, in the case of longitudinal fracture, spontaneous recovery from paralysis occurs in most patients (McHugh, 1959). Signs of

recovery should appear within 21 days. Facial electromyography and other electrodiagnostic steps provide a controlled basis on which to determine the need for surgical exploration (Collier, 1963).

Auditory Nerve Injuries. Transverse fractures of the temporal bone are associated with a high incidence of permanent hearing loss due to contusion or transection of the auditory nerve (McHugh, 1963). Such fractures are frequently accompanied by irreversible facial paralysis, as previously noted. However, longitudinal fractures are more likely to involve the middle ear, spare the labyrinth, and cause a conduction-type hearing loss. Thus, if there is a unilateral conductive hearing loss with a marked air-bone gap and an intact tympanic membrane, disruption in the ossicular chain has occurred (Kossner, 1961; Jackson and Magi, 1966)l. Successful techniques have been developed for the repair of this disturbance (Kossner, 1961; Perri, 1962; Gunderson, 1964). Recognition of this condition has been greatly facilitated by techniques for temporal bone laminography, which precisely identifies the fracture sites and the position of the ossicles.

Vascular Injuries Associated with Skull Fractures

Carotid-Cavernous Sinus Fistula. Fractures and penetrating wounds of the base of the skull may result in injury to the cranial vessels (Cairns, 1942). Traumatic occlusion of carotid or vertebral arteries is, however, a rare circumstance. More commonly, the lumen of the carotid artery is lacerated by the sharp fragments of bone lining the carotid canal, and because of the unique location of the artery in the cavernous sinus, the blood remains within the vascular system. Accordingly, signs of intracranial hemorrhage do not occur. Moreover, the development of ocular and neurologic symptoms secondary to elevation of pressure within the veins draining the cavernous sinus may be delayed several days, a feature attributable to secondary enlargement of the fistula and dilatation of orbital venous channels. Pulsating exophthalmos, failing visual acuity, diplopia, retro-ocular pain, and a continuous bruit constitutes the characteristic findings of this disorder (Hamby, 1966; Mayfield and Wilson, 1967).

Surgical treatment is indicated as early as the patient's general condition allows (Dandy, 1937). Failure to lower the arterial pressure in channels leading to the fistula results in progressive enlargement of veins draining from the sinus and diminishes the likelihood of success with subsequent simple surgical maneuvers. Ligation of the common and external carotid arteries in the neck occasionally succeeds in reducing the bruit and orbital pathologic findings, although additional maneuvers, such as intracranial ligation of the carotid and ophthalmic arteries and embolization of the cavernous carotid artery, are necessary for complete control of the fistula. Parkinson (1965) made an extensive study of this condition and demonstrated several intracavernous branches of the normal carotid artery that account for persistence of the fistula, even though all apparent inflow has been obliterated. The senior author and others having neurovascular interests developed techniques for balloon catheterization of the carotid artery and definitive occlusion of the fistulous opening (DeBrun and colleagues, 1978; Serbienko, 1974).

Epistaxis. Massive epistaxis may result from blunt or penetrating injuries of the internal carotid artery (Hamilton, 1953; Petty, 1969). We had two such cases, one of which was secondary to a missile injury to the carotid artery 2 months before the epistaxis. In

treating epistaxis of such magnitude, ordinary maneuvers may not be satisfactory. In this regard, we devised a technique in which a 30-mL Foley catheter balloon is placed in each nasopharynx and posterior choana. With each bag inflated and the nostrils manually occluded, it is possible to control the hemorrhage until surgical treatment can be instituted (Moore, 1979).

Cerebrospinal Fluid Fistula

Leakage of CSF occurs in approximately 2 per cent of all head injuries (Ommaya, 1968). The onset is usually within 48 hours of the trauma but may be as late as 3 months after injury. Recognition of the leak by the physician is important because the patient must be observed closely for signs of infection and meningitis. This is especially true in comatose patients who cannot report the leakage themselves. Patients with clinical signs of basilar skull fractures should be observed with a high index of suspicion. Fifty per cent of traumatic leaks stop within 1 week; 80 per cent cease within 1 month (Ommaya, 1968). Because of the self-limiting nature of these leaks, most neurosurgeons advocate surgical intervention only in cases of persistent leak (Coleman, 1937; Adson, 1941; Gurdjian and Webster, 1944; Morley and Hetherington, 1957; Ray and Bergland, 1969). The use of prophylactic antibiotics is a controversial matter that has an equal number of proponents and opponents. The incidence of meningitis after traumatic CSF leaks has varied from 3 to 50 per cent (Davis, 1976). Scientific data to support the efficacy of antibiotics in preventing meningitis are scarce. MacGee and co-workers (1970) reviewed 402 cases of traumatic CSF fistula: 325 of these patients received antibiotics, and 46 cases of meningitis occurred (14 per cent); 77 patients did not receive antibiotics, and four cases of meningitis were found (5 per cent). The authors felt that no statistical conclusion could be made. Ignelzi (1974) made a prospective study of 100 patients, alternated the treatment, and reached the same conclusion (Davis, 1976).

In our experience the following regimen has been successful: (1) broad-spectrum antibiotics are used initially; (2) the patient is kept quiet, and the head of the bed is elevated 30 degrees; (3) the CSF is allowed to flow freely from either the nose or the ear, and no attempt is made to block its escape; (4) is any signs of infection appear (fever, meningismus, and so forth), the spinal fluid and nose and throat secretions are cultured again, and antibiotic treatment is adjusted when so indicated; (5) if the leak persists for longer than 5 to 7 days, daily lumbar punctures or spinal drainage may be used; and (6) if the leak persists beyond 5 to 7 days, laminograms of the base of the skull are obtained in anticipation of surgical closure.

Localization of persistent leaks is a requisite for surgical repair. The following methods have been advocated in the more difficult cases:

1. Injection of radioiodinated serum albumin (RISA) into the subarachnoid spaces was advocated by DiChiro and co-workers (1964) but has not proved valuable in our experience unless the leakage of fluid is brisk. Montgomery (1968) used a modification of this technique, collecting the accumulated radioactive material in pledgets placed in the cavities of the sinuses overnight after injection of intrathecal RISA. However, results of this technique have been difficult to interpret because of the unbound RISA that is freely secreted in tears and mucus.

2. Pantopaque cisternography was advocated and found particularly helpful in spinal fluid leak through the internal auditory meatus (Teng and Edalatpour, 1963; Rockett and colleagues, 1964).

3. Metrizimide CT is the current method of choice.

The surgical technique for closure of spinal fluid fistula developed by Dandy (1926, 1944) consists of a transcranial intradural exposure of the dural defect and its closure with a patch of autogenous fascia. Few refinements have been made since this procedure was devised. The extradural approach proposed by Adson (1941) has gained few advocates. However, all have recognized that at times it may be extremely difficult to localize the fistula during surgery once the cranial cavity has been entered. In this regard it is important to have excluded a fistula through the internal auditory canal or mastoid prior to craniotomy. Morley and Wortzman (1965) reported the significance of lateral extension of the sphenoid sinus into the greater wings of the sphenoid in 25 per cent of normal skulls. This condition creates a situation in which the fistula may originate in the middle rather than the anterior cranial fossa. Similarly, Ray and Bergland (1969) proposed that the sphenoid be opened and packed with muscle in patients in whom the fistula cannot be identified at operation. Hirsch (1952) successfully closed fistulas extending through the sphenoid sinus by the endonasal approach, and Montgomery (1963) popularized the transethmoidal approach, in which the sphenoid is packed with fat and a septal mucosal flap is rotated to cover the defect in the floor of the anterior fossa. Observed success with this simple technique justifies its further use, particularly in the following situations: recurrence of fistulas following transcranial procedures, fistulas that can be defined as traversing the sphenoid sinus, and fistulas that occur after transsphenoidal operative procedures.

There are numerous other lesions that lead to CSF fistulas. These fistulas do not have as great a tendency to cease spontaneously and should therefore be approached more vigorously. They include tumors of the pituitary fossa (Norsa, 1953); tumors of the nose, nasopharynx, and paranasal sinuses; infections of the paranasal sinuses; congenital defects in the dura (Rockett and colleagues, 1964); nasal encephalomeningoceles (Suwanwela and Horgsaprabhas, 1966); arachnoid cysts of the sella turcica (Brisman and colleagues, 1969); spontaneous rhinorrhea resulting from lesions obstructing CSF circulation and causing increased intracranial pressure (hydrocephalus) (Ommaya and colleagues, 1968; Rovit and colleagues, 1969); and surgical procedures in the region of the paranasal sinuses and mastoid. The high incidence of cerebrospinal fistula and meningitis secondary to stereotaxic implantation of yttrium in the pituitary fossa led to the virtual abandonment of this procedure (Sweet, 1969). With other stereotaxic and open procedures, special precautions must be exercised to prevent persistent spinal fluid fistula. This precaution is particularly important in view of the continued popularity of transnasal and translabyrinthine surgical approaches to the cranial cavity (House and Hitselberger, 1964).

Infections

Neurologic Complications of Sinus and Ear Infections

The neurologic complications of infections of the ear and paranasal sinuses are extracranial and intracranial in location. The sinuses are mucosa-lined cavities within bone, the apertures of which are readily obstructed by inflammatory changes within the lumina. As the intraluminal pressure increases, pain occurs, which may be localized over the sinus as well as referred to other sites about the face. The intracranial complications of mastoid and paranasal sinus infections occur as a result of extension through a fistula or the oval or round windows. However, facial nerve paralysis and labyrinthitis are more commonly complications of osteomyelitis of the temporal bone. Exophthalmos, oculomotor nerve palsy, diplopia due to abducent nerve palsy, and diminished sensation of the forehead are features of the superior orbital fissure syndrome that in partial or complete form may result from paranasal sinusitis. Venous thrombosis of the retro-orbital veins is an early manifestation of orbital cellulitis, which may lead to chemosis, exophthalmos, diplopia, and immobility of the globe. Necrotizing orbital cellulitis and sinusitis may be seen in debilitated individuals as a result of mucormycosis (Green and colleagues, 1967) and Wegener's granulomatosis. Extensive bone destruction and cranial nerve palsies are common.

In each of these conditions early recognition, prompt antibiotic therapy, and drainage when indicated are the therapeutic methods of proven value, and undoubtedly are responsible for the continuing low incidence of neurologic complications of ear and sinus infections.

Pyoceles may develop in the sinuses. Similar to chronic cholesteatoma found in the mastoid, the lesions require drainage and radical resection in case persistent destruction of bone leads to compression of the cranial nerves confined in the bone adjacent to the cavities.

Mucocele

Mucocele is defined as the accumulation and retention of mucoid material within a sinus as a result of continuous or periodic obstruction of the ostium of the sinus (Hayes and Creston, 1964). The secretion is usually clear, thick, and tenacious unless invasion by bacteria has occurred and has created a pyocele, in which case the color and consistency of the accumulated material vary according to the infecting organisms (Krueger and colleagues, 1965). Secondary erosion and distention of the sinus walls occurs as the intraluminal pressure increases.

Mucoceles are most frequently found in the frontal sinus (Mortada, 1968). Chronic in nature and a source of recurrent frontal headache, they may erode through either the anterior or the posterior wall of the frontal sinus. In the former instance, a tender, fluctuant mass presents beneath the periosteum of the frontal bone, commonly known as Pott's puffy tumor, and requires local drainage and exenteration of the sinus. If the posterior sinus wall is destroyed, epidural abscess, subdural empyema, meningitis, or brain abscess may develop. This complication is covered in a separate section of this chapter.

Mucoceles of the ethmoid sinuses commonly destroy the adjacent thin lamina papyracea and displace the orbital contents laterally or downward, resulting in exophthalmos

and diplopia. Compression of the superior orbital fissure produces ocular palsies and diminished sensation in the forehead.

Mucoceles of the maxillary sinus rarely lead to neurologic complications (Bloom, 1965; De, 1966); only a few well-documented cases are recorded. Proptosis due to upward displacement of the orbital contents (Parker, 1961), exophthalmos caused by destruction of the floor of the orbit (Montgomery, 1964), diplopia due to displacement of orbital contents, and paresis of the oculomotor as well as the optic nerve, as in the orbital apex syndrome (Pooley and Wilkinson, 1913; Lundgren and Olin, 1961) are documented features.

Mucoceles of the sphenoid sinuses have been infrequently reported. However, this condition must always be suspected when one is confronted with an expanding mass in the sphenoid sinus and sella turcica. Recurrent headaches and visual disturbances (diplopia and visual deficits) are characteristic symptoms (Reinecke and Montgomery, 1964). Indeed, this condition may be confused with ophthalmoplegic migraine (Pincus and Daroff, 1964), a plausible error considering the chronic nature of both diseases. Signs of chiasmal compression, ocular nerve palsies, exophthalmos, and intermittent CSF rhinorrhea have been recorded. However, spontaneous intracranial infection is most unusual (Hayes and Creston, 1964).

The radiographic findings consist of (1) opacification of the sphenoid sinus, (2) ballooning and rarefaction of the bony wall of the sphenoid sinus, (3) destruction of the interseptum of the sinus, (4) erosion of the floor and walls of the sella turcica, and (5) lateral and upward displacement of the carotid arteries. Tomography and carotid angiography are essential in the investigation of this lesion (Norman and Yanagisawa, 1964; Bloom, 1965; Nevins and Leaver, 1967; Diaz, 1978).

It is imperative that the correct diagnosis is made before treatment (radiotherapy or surgical) of all masses in the sphenoid sinus and sella turcica. Although sphenoid mucoceles account for only a small portion of these lesions, they may always be cured by adequate endonasal drainage and exenteration of the sinus.

It should be emphasized that intracranial exploration of these lesions must be avoided, since spinal fluid rhinorrhea and meningitis are almost certain to follow.

Osteomyelitis and Epidural Abscess

Osteomyelitis of the bone of the calvarium occurs by direct extension of infection from the mastoid and nasal sinuses (French and Chou, 1969) and spreads through the haversian canals by thrombosis of intraosseous vessels. The spread of the process is facilitated on reaching the diploë because of the absence of valves in diploic veins. The frontal, parietal, and temporal bones are most commonly involved. Rupture of the suppurative lesion into the subperiosteal space causes a localized collection beneath the skin, Pott's puffy tumor, facilitating recognition and external drainage. However, if the lesion ruptures into the epidural space and remains unrecognized, serious neurologic complications may develop. Osteomyelitis and epidural abscess are to be suspected if there are local and systemic signs of sepsis - pain, fluctuance, tenderness, erythema, fever, and malaise. Rarely is there nuchal rigidity or any other sign of meningeal irritation. Radiographic evidence of osteomyelitis may develop slowly

and appear as only small areas of rarefaction of the bone. However, confirmatory findings of opacification of the adjacent sinus and clouding of the mastoids are early radiographic signs.

Obviously, early recognition requires direct exploration; there is no role for delay in this disorder. Drainage of the sinus and extensive removal of the infected bone are desirable. The bone usually regenerates quickly and readily because of the retention of the periosteum. If the suppuration has extended to the epidural space, this material must be debrided in a careful manner that will allow removal of granulation tissue and pus but will maintain the integrity of the dura mater.

Subdural Empyema

Subdural infection (empyema) in the absence of direct implantation of a foreign body is a result of extension from adjacent areas of infection, commonly in the sinuses and ears. The infection probably reaches the subdural space by extension through the dura mater via small arterial and venous channels, or by direct extension in thrombi propagated in the dural sinuses or emissary veins. The latter has been documented in a number of patients in whom osteomyelitis and epidural abscess have been absent.

The clinical features should distinguish subdural empyema from other forms of intracranial infection. The illness usually begins with localized headache and focal signs of infection in the diseased mastoid or sinus. Development of systemic signs of infection - fever and malaise - is followed shortly by alterations in consciousness that may progress to stupor or coma. As the process of spread through the subdural space continues, seizures are a frequent occurrence and progressive neurologic deficits may develop. Nuchal rigidity, hemiparesis, aphasia, diminished sensation, and visual field deficits have all been recorded in a majority of patients. Skull radiography demonstrates the focus of mastoiditis or sinusitis and perhaps an area of osteomyelitis. Carotid angiography may show displacement of the intracranial vessels from the inner table of the skull, indicating a peripheral extracerebral mass. However, CT scanning provides the best evidence of the subdural collection. Spinal fluid examination usually shows elevated pressure, mild pleocytosis (less than 1000 cells), normal sugar and chloride concentrations, and absence of any organisms on Gram stain or culture.

Treatment of subdural empyema consists of effective drainage of the infected sinus. There is no reason to delay this maneuver pending treatment of the intracranial sepsis, since further delay serves only to propagate infection into the subdural space. Botterell and Drake (1952) stressed the mode of spread of infection in the subdural space, as follows: "From the paranasal sinuses it spreads along the interhemispheric fissure, over the frontal convexity, and under the orbital surfaces of the frontal lobes; from the mastoid purulent material spreads underneath the temporal and occipital lobes, over the cerebellum, over the convexity, and along the posterior interhemispheric fissure. Loculation of pus may occur in any of the areas; however, it is more likely to be concealed between the hemispheres, underneath the cerebral lobes, and over the cerebellum. Appropriate placement of cranial burr holes is facilitated by this knowledge". Rather than placing multiple bur holes as advocated by Botterell and Drake (1952), McLaurin (1969), and others, we prefer to use a 2-inch trephine craniotomy in the frontal and occipital regions. This technique provides ample room for inspection and exploration of the subdural space. Catheters are placed in situ for repeated irrigation and

instillation of antibiotics. In addition, it is important to change the patient's position frequently to diminish loculation in the occipital region.

Despite these vigorous surgical maneuvers and administration of massive intravenous and local antibiotics, the mortality rate for this disease remains exceedingly high, 50 to 60 per cent.

Meningitis

Meningitis, the most common intracranial infection, is a pyogenic infection of the pia-arachnoid of the brain and spinal cord. In the preantibiotic era, meningitis was a common and deadly complication of middle ear and sinus suppuration. Fortunately, recognition of the mechanism of spread to the subarachnoid space (Shambaugh, 1967) and institution of proper antibiotic therapy have greatly decreased the incidence of mortality from this condition. Meningitis accounts for about 95 per cent of all intracranial infections associated with sinus and mastoid disease (Beekhuis and Taylor, 1969). Septic thrombophlebitis, direct extension through the subdural space, and hematogenous spread are the modes of seeding the subarachnoid space from sinus and mastoid infection (Alpers, 1958). The mortality rate remains high in meningitis that is associated with spread from both these foci, particularly in cases of sinus suppuration, being 31 per cent compared with 7 per cent for otitic meningitis (Beekhuis and Taylor, 1969). This unfortunate figure is perhaps related to late recognition of meningeal spread from sinus disease because of masking of the signs by the local process, inadequate treatment of sinus suppuration, and a generally higher index of suspicion of meningitis when ear infection is concerned.

Accordingly, it is important to watch carefully for the symptoms and signs of meningitis. Headache, lethargy, and irritability are the early symptoms of meningeal infection and should lead the physician to elicit signs of meningeal irritation (nuchal rigidity, limited flexion of the legs, fever, and alterations in mental status). Ultimately the diagnosis rests with spinal fluid analysis. Certainly a spinal puncture must be performed as soon as meningitis is suspected, except of course when there are elevated intracranial pressure and focal signs indicating a mass lesion (brain abscess), which can be ruled out with the help of CT. Bacterial meningitis usually is accompanied by elevated spinal fluid pressure, pleocytosis (greater than 1000 cells, predominantly polymorphonuclear leukocytes), elevated protein levels, decreased sugar concentration (less than 50 per cent of blood sugar value; simultaneous blood sugar determination should always be obtain), and decreased chloride content (Alpers, 1958). The cloudy appearance of the fluid is related to the degree of pleocytosis and protein content. The value of meticulous cytologic, chemical, and bacteriological analysis of the spinal fluid cannot be overemphasized. Broad-spectrum antibiotic coverage should be initiated as soon as spinal puncture has been completed. Specific antimicrobic therapy may then be instituted in accordance with the results of culture and antibiotic sensitivity testing. In some severe cases, systemic corticosteroids may be of benefit (Toole and colleagues, 1969; Eisen and colleagues, 1969). Of course, prompt attention to the local infection in the sinus or ear is necessary to prevent continued seeding of the subarachnoid space.

Brain Abscess

Brain abscess is a localized collection of pus in a cavity created by the necrosis of cerebral tissue, and encapsulated by deposition of inflammatory cells from the blood and glial reaction of the adjacent cerebral tissue. Despite the reduced incidence of brain abscess attributable to antibiotics and early recognition of extracranial suppuration, mastoid and paranasal sinus infection remains the leading cause of brain abscess, except in medical centers with a large population of patients with cardiac defects (Matson and Salom, 1961; Beekhuis and Taylor, 1969). The mechanisms of propagation from sinus and mastoid suppuration include (1) direct extension, (2) vascular extension via septic thrombophlebitis and perivasculitis, and (3) extension through preformed pathways, as in congenital defects, tumors, or traumatic fistulas.

The brain abscess begins as an inflammatory process (cerebritis), which consists of leukocytic infiltration and microscopic necrosis. At this stage clinical and diagnostic efforts usually fail to localize the focus of infection. However, in 7 to 10 days a capsule is initiated about the necrotic core by the deposition of a granulomatous layer and the proliferation of glial and fibrous tissue elements from the adjacent blood vessels. Enlargement of the mass and provocation of edema in the adjacent cerebral tissue usually produce focal neurologic abnormalities unless the mass is located in the frontal lobe, where it may produce only signs of increased intracranial pressure.

The diagnostic workup for a suspected abscess has changed with the availability of CT scanning. The combination of isotopic and CT scans ensures 90 to 95 per cent accuracy in diagnosing abscesses (Wang and Rosen, 1965; Tefft and colleagues, 1966). Rosenblum and coworkers (1978) pointed out that since the advent of the CT scan there has been more accurate diagnosis and localization of abscesses and more rapid detection of postoperative complications, which previously accounted for most of the deaths. Hoff (1978) further maintained that observation based on serial CT scanning might well permit medical treatment of some abscesses previously treated with intracranial surgery. Cerebral angiography remains one of the most accurate means of identifying and localizing abscesses. In some patients, only certain angiographic findings may differentiate an abscess from some other intracerebral mass lesion (Chou and colleagues, 1966).

Otogenic sources probably account for 40 to 50 per cent of all brain abscesses, whereas sinus infection causes 10 to 15 per cent. This feature accounts for the greater incidence of abscesses in temporal and cerebellar locations. The symptoms of brain abscess are general and focal in nature. General symptoms consist of mental dullness, lethargy, headache, and occasionally fever. The symptoms may be masked or confused with those related to the condition in the sinus or mastoid. Fever and meningeal signs usually are not present in the absence of coincidental meningitis. Focal signs (hemiparesis, hemisensory deficit, and hemianopia) frequently accompany temporal and parietal abscesses, a most common site for otitic propagation of infection. Ataxia, vertigo, cranial nerve palsies, and increased intracranial pressure (due to obstruction of the aqueduct of Sylvius or fourth ventricle outlet) are signs of cerebellar hemisphere abscess, also a favorite site for spread from the infected middle ear. Frontal lobe abscesses are almost always the result of sinus infection and seldom have consistent localizing neurologic features. Increased intracranial pressure, papilledema, and stupor are features that precede the onset of fatal brain stem

herniation or rupture of the abscess into the ventricular system.

Lumbar puncture and air encephalography must be performed with great care in suspected brain abscess and carried out only when brain scan and angiography have failed to localize the lesion sufficiently. Such a circumstance seldom occurs except in cerebellar abscess. In this situation, positive contrast ventriculography may be the preferable procedure (Siquera and colleagues, 1968; Wilkinson, 1969). CT scanning has made these considerations obsolete and greatly reduced the risk of misdiagnosis.

The treatment of brain abscess consists of broad-spectrum antibiotic coverage, systemic corticosteroids to reduce brain swelling, and surgical evacuation of the abscess. It is our opinion that delay of definitive surgical treatment to obliterate meningitis or local infection in the sinuses is ill advised. In this regard it must be emphasized that the cause of death in brain abscess is brain stem compression secondary to herniation of the medial temporal lobe or cerebellar tonsils, or rupture of the abscess into the ventricular system (Ballantine, 1969; Krayenbühl, 1966).

The surgical treatment of brain abscess is a matter of dispute, since some surgeons feel strongly that the mass should be excised completely via craniotomy and direct vision as soon as the diagnosis is made (Ballantine, 1969; Matson, 1969), whereas others prefer to puncture the abscess through a bur hole, aspirate the purulent material, and instill a radiopaque material (Kahn and Arbo, 1939; Maxwell and DeLong, 1968), either Thorotrast or micropaque barium. If this method is chosen, repeated taps or catheter drainage may be performed on the basis of serial studies of the size of the cavity. Obviously, the ideal treatment is complete excision of the abscess, but this may be a dangerous and unwise step in individuals who are extremely ill because of systemic toxicity of local infection and meningitis. In patients with lesions located beneath vital neurological structures, such as the left motor and speech areas, total excision may produce a profound neurologic deficit. Accordingly, in such circumstances we are prepared to tap the abscess, repeatedly if necessary. Furthermore, we rely on systemic antibiotics and hyperosmolar agents to control the cerebral edema. Prompt treatment of the local infection is imperative (Morgan and colleagues, 1973; Carey and colleagues, 1972).

Painful Disorders of the Head and Neck

Pain in the head and neck is probably one of the most complex and difficult disorders encountered by the otolaryngologist and neurosurgeon. How does one distinguish the various neuralgias of the cranial and upper cervical nerves (a condition in which severe paroxysms of pain radiate into the distribution of a nerve that has nos structural evidence of disease) from the symptomatic head and neck pain in which distinct pathologic lesions involve the peripheral nerve, ganglion, posterior rootlet or brain stem, and upper spinal cord?

Fortunately, there are many successful techniques for relieving pain in the region of the head and neck. Accordingly, it is important to obtain the correct diagnosis before alleviating pain, which may be the only alerting feature of an underlying pathologic disorder. In order to achieve this goal, precise definition of the clinical features of facial and cervical neuralgias is important. We shall therefore review the classic features that have been so carefully documented by numerous observers.

Neuralgias

Trigeminal Neuralgia

Trigeminal neuralgia, or tic douloureux, is a common disorder in which the patient is so miserable that it has been recognized since the earliest days of medical history (Crawford and Walker, 1951). This form of neuralgia is characterized by (1) paroxysms of intense pain lasting seconds to minutes and separated by intervals of freedom from pain; (2) provocation of the paroxysms by peripheral stimuli to the face and mouth, as in chewing, swallowing, shaving, touching, or, at times, even a whiff of air striking the face; (3) radiation of pain into the distribution of the trigeminal nerve, usually second or third division; (4) restriction of pain to a single side of the face; and (5) no neurologic deficit in the region of the trigeminal nerve, ie, no diminution of sensation or weakness of the masticatory muscles.

Departure from these features may occur but must be considered with skepticism. Slight sensory deficit of the face or cornea; persistence of a dull, aching pain between the paroxysms of pain; and flight of pain beyond the trigeminal distribution or across the midline of the face are features that should suggest the possibility of multiple sclerosis, brain stem vascular anomaly, syringomyelia, or posterior fossa tumors. It has been noted that facial pain is more likely to occur in association with lesions involving the gasserian ganglion than in those affecting the posterior root (White and Sweet, 1969). Acoustic neuromas, neuromas of the trigeminal nerve, and meningiomas are rarely associated with typical trigeminal neuralgia. However, congenital cholesteatoma causes ticlike pain in a high percentage of patients (Revilla, 1947; Taarnhoj, 1952), particularly when the tumor compresses the ganglion in the middle fossa (Krayenbühl, 1936).

Tumors invading the divisions of the trigeminal nerve may produce facial pain. Malignant tumors of the sinus and nasopharynx frequently invade the cranial nerve, but generally the associated pain is accompanied by neurologic deficit. Cranial nerve palsies are common, as in the superior orbital fissure and jugular foramen syndromes.

If symptomatic causes of trigeminal neuralgia can be excluded by careful history taking and neurologic examination, few diagnostic procedures are necessary. However, in doubtful cases the previously noted lesions may be excluded by intracranial angiography, pneumoencephalography, posterior fossa myelography, and CT of the head.

The initial treatment of trigeminal neuralgia is medical. Carbamazepine (Tegretol) has proved extremely beneficial in relieving severe bouts of tic pain. Loss of effectiveness appears in some persons, and others have relapses shortly after cessation of the drug. Toxicity poses another hazard; indeed, some patients find the nausea, ataxia, and vertigo too troublesome. Severe complications, such as agranulocytosis and renal and hepatic toxicity, have been infrequent but require careful monitoring of appropriate laboratory values. Tegretol is of particular value for the patient with severe pain in need of urgent relief, for treatment of mild protracted pain, and for recurrent pain following operative procedures.

The various operative procedures that have been developed to alleviate the pain of trigeminal neuralgia are numerous, and all have some unique value. In particular, differential section of the posterior root in the middle fossa has found great favor with neurologic

surgeons (Frazier, 1925; Stookey, 1955). However, in an attempt to avoid loss of touch perception over the face, Dandy (1929) sectioned the posterior root in the posterior fossa. Because of his great ability, he was apparently able to identify a small group of intermediate fibers in the posterior fossa that relayed proprioceptive sensation from the face. With the aid of the operating microscope, Jannetta (1967) confirmed Dandy's impression that preservation of these fibers preserves light touch intact while eliminating pain in the same area. However, a number of less complicated and hazardous procedures have been developed. Injection of the various nerve trunks with neurotoxic agents has been, and remains, a highly effective and simple procedure for the temporary relief of pain. Injection of alcohol or phenol into the gasserian ganglion, designed to produce destruction of the ganglion and thus permanent relief of tic (Harris, 1940), has been associated with a high incidence of complications (nonparalytic keratitis and anesthesia dolorosa). Seeking to avoid such an occurrence, Jaeger (1959) injected 0.5-mL increments of boiling water into the gasserian ganglion. This maneuver achieved a significant lowering of the complications but has not gained favor with other surgeons.

In 1931 Kirschner introduced a technique of coagulation of the gasserian ganglion. Large series of patients have been treated by this procedure over the past 5 decades in Europe, where it has enjoyed considerable approval (Kirschner, 1942). However, despite continued improvement in technique and results, it was not introduced in the USA until the late 1960s. Sweet (1968b) and Wepsic (1969) were responsible for bringing this technique to our attention. Their results in more than 500 cases have been extremely favorable. We have evaluated the features of this technique and found it extremely desirable because it is capable of relieving trigeminal neuralgia while sparing proprioceptive sensation over the face and cornea and reducing injury to the other cranial nerves, including the motor root of the trigeminal nerve. Furthermore, it is simple in principle and execution and carries no significant morbidity. Therefore, it can be performed on people who are not candidates for more major surgical procedures.

The selection of a surgical procedure should be considered only after medical treatment has been thoroughly explored. Sensory preservation is important; therefore, considerable interest has been directed toward techniques that induce minimal sensory deprivation. The hypothesis by Jannetta that vascular compression of the nerve root at the pons is the major etiologic factor in tic douloureux led to a new microsurgical procedure (Jannetta, 1976) that is currently being used. Further study is required to determine the ultimate place of this procedure in the total surgical treatment available for trigeminal neuralgia.

Application of stereotaxic principles to the treatment of trigeminal neuralgia (Tew and colleagues, 1978) led to a remarkable degree of safety and reliability in results. This procedure compares favorably with any other technique, whether destruction or decompression is the intent of the surgeon (Tew and Keller, 1976).

Nervus Intermedius Neuralgia

There is substantial anatomic evidence that pain fibers from the geniculate ganglion innervate a portion of the ear, the deep structures of the head and neck, and the facial muscles. Ramsay Hunt (1915) determined the cutaneous and mucosal distribution of the facial nerve, primarily on the basis of the location of vesicular eruptions in herpes zoster of the

geniculate ganglion. Additional evidence comes in the form of paresthesias, which have been noted to radiate into the medial surface of the auricle and anterior two-thirds of the tongue in patients with Bell's palsy. Other pain fibers pass in the greater superficial petrosal nerve, in branches of the facial nerve to the expressive muscles, and perhaps together with the taste fibers of the chorda tympanic nerve. Anatomic dissections in humans have shown great variation in the anastomosis between facial, vagus, and trigeminal nerves, a finding that undoubtedly accounts for the confusion surrounding the clinical features of painful disorders affecting these nerves.

The typical pain referred over the nervus intermedius is usually continuous rather than paroxysmal. It is dull to aching in character, although severe episodes of sharp, burning pain radiating deep into the ear and mouth have been recorded. The pain usually radiates into the depth of the ear and external canal, which may be sensitive to manipulation. The conclusive diagnosis depends on reproduction of the pain by stimulation of the nervus intermedius at the pons; long-term relief of pain is obtained by section of the fibers of the nerve. Such a delicate maneuver requires an awake, cooperative patient and the visualization provided by the operating microscope. The anatomy and operative approach to the nervus intermedius were described by Rhoton and co-workers (1968) and Sachs (1968).

Vagoglossopharyngeal Neuralgia

Although cranial nerves IX and X individually may conduct paroxysms of pain in the throat, tongue, ear, and jaw, or any combination of these areas, it frequently is impossible to divide the characteristics of this disorder into two separate clinical categories. Therefore, White and Sweet (1969) suggested the term vagoglossopharyngeal neuralgia as a more realistic title. Recognition depends on the following features: (1) unilateral paroxysms of pain occur in the region of the tongue, throat, ear, and larynx; (2) the pain is seldom confined to the area from which it initially arose and frequently involves all areas just referred to; (3) the pain is usually brief in duration and sharp in character, although prolonged bouts of constant dull or burning pain may occur; (4) paroxysms of pain occur that may be induced by swallowing or by stroking the skin of the ear or mucous membranes of the tonsillar and pharyngeal regions; and (5) attacks of pain are sometimes accompanied by anatomic and cardiovascular responses, ie, salivation, bradycardia, hypotension, and syncope. If these features are prominent, one must consider hypersensitivity of the carotid sinus (Ray and Steward, 1948). However, bradycardia, syncope, and hypotension may be associated with severe episodes of pain in the throat and ear (Riley and colleagues, 1942).

In view of the considerable variation in the character of neuralgia of the vagoglossopharyngeal nerve, astute otolaryngologic and neurologic evaluation is essential to exclude symptomatic lesions such as tumors of the nasopharynx, paranasal sinuses, or larynx; local infection; and tumors compressing the ganglia or posterior rootlets of these nerves in the posterior fossa.

Tegretol has been effective in the therapy for this disorder, as for trigeminal neuralgia. Removal of the styloid process has been noted to relieve a painful condition of the throat attributed to elongation of the styloid process or to ossified extensions that may irritate the lateral pharyngeal wall as the stylopharyngeal muscles pull it across the anomaly during the act of deglutition (O'Brien, 1962). Superior laryngeal neurectomy may be successful in

relieving pain that radiates outside the confines of the larynx (Echols and Maxwell, 1934; Ballantine, 1966). If the simple methods fail, rhizotomy of cranial nerve rootlets IX and X in the posterior fossa is advisable (Dandy, 1927; White and Sweet, 1969). Care should be taken to avoid cutting the lower rootlets of the vagus, lest undue hoarseness occur (Walker, 1966).

Periodic Migrainous Neuralgia

Periodic migrainous neuralgia is a conspicuous syndrome distinguished by a bevy of eponyms and descriptive titles: Horton's syndrome, histamine cephalalgia, autonomic faciocephalalgia, petrosal neuralgia, cluster headaches, and erythromelalgia.

Clinical features of the syndrome resemble both migraine cephalalgia and trigeminal neuralgia. Unilateral paroxysms of constant, severe, burning pain, beginning in the eye or forehead and radiating into the face, temple, and neck, distinguish this condition. The episodes of pain occur in clusters during the day or, more characteristically, awake the patient from sleep. An episode of pain usually builds to a peak intensity in a crescendo fashion in a period of 30 to 45 minutes, then promptly ends. Although rarely preceded by a visual or sensory aura, the paroxysms of pain are commonly accompanied by autonomic disturbance in the form of tearing, rhinorrhea, salivation, ptosis, myosis, injection of the conjunctiva, and dilatation of facial and scalp vessels. All these features are unilateral in occurrence and are not provoked by external stimuli; however, some of our patients have noted that ingestion of even modest amounts of alcohol precipitate an attack. Although sinusitis and other nasal disorders have been implicated in the pathogenesis of this condition, Schiller (1960) and others were singularly unimpressed by this association.

The medical treatment is similar to that effective for migraine cephalalgia, ie, ergotamine tartrate (Schiller, 1960) and methysergide (Gragam, 1964). However, the undesirable and occasionally disabling side effects of methysergide necessitate careful evaluation and surveillance of patients receiving this drug. Corticosteroids have been beneficial in the control of severe and protracted bouts of pain (Graham, 1964).

Surgical procedures vary from simple to complex and are generally designed to attack anatomic components of the disorders at various sites. Sphenopalatine ganglionectomy, Vidian neurectomy, and stellate ganglionectomy are procedures that have been largely abandoned because of lack of value. Resection of the superficial temporal artery has provided striking relief in occasional patients; however, there is no method for selecting those who might benefit from this simple procedure (White and Sweet, 1969). Greater superficial neurectomy was of distinct value in 50 to 75 per cent of patients in the experience of Gardner and co-workers (1947). The recurrence of pain after petrosal neurectomy led to section of the nervus intermedius in the posterior fossa (Sachs, 1968).

Atypical Facial Pain

This final group of neuralgias contains the painful disorders that are poorly characterized and therefore deviate from the commonly occurring features of other neuralgias. Features that may indicate atypical facial pain include radiation of pain into deep structures beyond individual cranial nerve zones; lack of restriction to hemicranial location; pain that

is constant, dull, and aching in character; and a tendency of patients toward drug addiction and personality disorders. Obviously, this is not a very gratifying diagnosis. Thus, one must search carefully for obscure conditions that may be the basis of these symptoms. One must also avoid the temptation to perform destructive procedures, because these patients are frequently more miserable after each new procedure.

Symptomatic Pain of the Head and Neck

Costen's Syndrome

Temporomandibular neuralgia was described by Costen (1934) and is said to consist of medical movement of the condyle, leading to pressure against the auriculotemporal and chorda tympanic nerves. Headache and burning in the throat, tongue, and nose are characteristic complaints. Diminished salivation has been noted. Relief is said to be obtained by repositioning the jaw with dental prostheses (Smolik and Hempstead, 1952). We have not seen any patients treated successfully in this fashion, although most patients with facial pain have been treated by dentists or orthodontists prior to neurosurgical consultation.

Dental Disease

Dental infection, pulpitis, apical abscess, and impacted teeth must be considered in the evaluation of atypical facial pain that may even be diffuse in nature.

Tumors, Infections, and Trauma

Deep-seated neoplasms, infection, and traumatic lesions frequently give rise to atypical head and neck pain. The features of each condition have been covered in the appropriate section.