Paparella IV: Section 2: Disciplines Closely Associated With Otolaryngology

Chapter 25: Exophthalmos for the Otolaryngologist

Lawrence W. DeSanto, James A. Garrity

Orbital and Sinus Relationships

The orbits and sinuses are related in two ways: by anatomic location and by venous drainage. Paranasal sinuses surround the orbit from the 11 o'clock position above to the 6 o'clock position below, with individual variations. The orbit is a quadrilateral and pyramidal cavity formed by several bones. The superior wall of the orbit forms some or all of the floor of the frontal sinus; the orbital floor is the roof of the maxillary antrum, and its medial wall is the lateral wall of the ethmoid labyrinth. None of these bony partitions is very thick, and the ethmoid orbital wall is aptly named the paper plate of the ethmoid bone.

The venous relationships are consequential in inflammatory disease. The venous system is devoid of valves, and this allows a two-way pathway for infection to and from orbits from the sinuses, face, nasal cavity, and pterygoid fossa. At the superior medial angle of the orbit, the superior ophthalmic vein is continuous with the nasofrontal vein, which is an extension of the angular vein of the face. The superior ophthalmic vein courses posteriorly and laterally, passing between the two halves of the lateral rectus muscle, through the superior orbital fissure, and into the cavernous sinus. Infection can follow the vein from sinus to cavernous sinus to orbit or from sinus to orbit to cavernous sinus. The inferior ophthalmic vein begins along the floor and medial wall of the orbit, with contributions from veins about the lacrimal sac, eyelids, and orbital muscles. This vein courses posteriorly in the lower orbit and then usually divides, one branch going through the inferior orbital fissure to the pterygoid venous plexus, and the other branch exiting the orbit through the superior orbital fissure and entering the cavernous sinus.

The volume of the orbital socket is approximately 30 mL. Because of the rigid confines of the orbital walls, any disease process within or adjacent to the orbit displaces volume, with a resultant proptosis - globe displacement. Proptosis is a forward displacement of the globe, which is a nonspecific response. Other symptoms of orbital disease include diplopia or visual loss related to compression or infiltration of vital structures.

Clinical Evaluation

The evaluation and treatment of orbital disease falls within the domain of several specialties, including neurosurgery, ophthalmology, otorhinolaryngology, and plastic surgery. Many of the disease processes affecting the orbit are best managed by a team approach, which is advocated for any complex orbital problem.

A patient with proptosis must be evaluated to determine whether there is true proptosis or pseudoproptosis.

An accurate history can assist the examination by historically documenting previous ocular trauma or inflammation. Either trauma or inflammation can ultimately lead to an

atrophic or phthisical globe, resulting in enophthalmos and in either apparent proptosis or, more correctly, pseudoproptosis in the contralateral eye. Unexpectedly, patients may be unable to recall previous trauma that has "blown out" the floor or medial wall of the orbit. Another example of apparent proptosis caused by contralateral enophthalmos is that arising from metastatic scirrhous adenocarcinoma (of breast, lung, or stomach) to the orbit. The restrictive orbitopathy is secondary to a generalized contraction that the tumor promotes. Again, historical information can lead one to suspect this diagnosis.

An easily overlooked cause of pseudoproptosis is a large globe from unilateral high myopia. Lid retraction can also give the appearance of ipsilateral proptosis. An ophthalmologic examination can document the large refractive error, along with the attendant amblyopia or lid retraction as a source of pseudoproptosis.

When the clinical evaluation has determined that proptosis does exist, we have found the mnemonic VEIN (vascular, endocrine, inflammation-infection, neoplasm) helpful for clinical classification. Clinically significant proptosis is regarded as a difference of 2 mm or more, as measured by either the Krahn or Hertel exophthalmometer.

Radiologic Evaluation

Radiologic evaluation of the orbit can be done with computed tomography (CT), magnetic resonance imaging (MRI), or ultrasonography (B- or A-scan modes). Modern CT scanners can produce 1.5-mm axial cuts through the orbit. The data collected from these thin-cut axial images can be used to reconstruct coronal images. Alternatively, direct scanning of the orbit in both the axial and the coronal planes can be performed.

We recommend cuts no thicker than 3 mm in both the axial and coronal planes (it is possible to obtain reconstructed coronal scans from 3-mm axial cuts that are of good quality but of less resolution than 1.5-mm cuts). Extensive metallic dental work or a patient's inability to extend the neck make reformatted coronal images a more viable option. We stress the importance of coronal imaging in the evaluation of orbital disease, particularly when lesions are along the roof or floor. Orbital CT scanning is the standard of current imaging, in part because the orbital fat serves as an intrinsic contrast agent.

Orbital MRI holds much promise, particularly as experience is accumulated. One notable advantage of MRI over CT is that the former gives an idea of the histologic appearance. A noticeable disadvantage of MRI is the long scanning time required, although this has been reduced on the newer units.

Orbital ultrasonography is a valuable ancillary test for diagnosis. High-frequency sound waves are propagated through soft tissue, and the differential reflection of these waves by tissues in the path of the beam is recorded. A-scan sonograms provide a one-dimensional tracing that allows specific comparisons of tissue reflectance as well as accurate measurements of tissue dimensions. B-scan sonograms give a two-dimensional cross section of the eye-orbit. The transducer can be used with either a water bath or a contact method. The contact B-scan technique is most likely to be used in a general office setting: its advantages are low cost, non-invasiveness, and ready availability; its disadvantages is the high level of technical and interpretive skills needed by the operator. The posterior portion of the orbit is

not seen reliably, and lesions located along the orbital roof or floor may be missed, particularly when the contact B-scan method is used. A water bath B-scan is not as readily available and is more cumbersome to use, but it does offer better resolution than the contact method. On a practical note, CT and MRI have virtually replaced ultrasonography, but the latter still is helpful in certain situations.

Clinical Classification of Proptosis

Vascular Exophthalmos

Vascular causes of proptosis are seen from either the venous (varix) circulation or the arterial circulation (dural fistula or carotid cavernous fistula). A venous varix has a characteristic history of either positional proptosis induced by Valsalva maneuvers. A long-standing varix, because of eventual orbital fat atrophy, may produce enophthalmos in the primary position, until a Valsalva maneuver reveals the true identity. Suspicion of a varix should alert the radiologist to perform CT, either with jugular vein compression or during a Valsalva maneuver. Operative intervention can be fraught with difficulty, and observation may be the most prudent therapy.

A dural sinus fistula or low-flow shunt may masquerade as unilateral glaucoma. Because of the insidious onset of proptosis, a high index of suspicion is necessary, particularly if the globe is not injected or the proptosis is minimal. Headache or tinnitus need not be present. Orbital CT or B-scan ultrasonography may reveal a slightly enlarged superior ophthalmic vein with either an enlarged or a normal-sized muscle. No treatment other than observation may suffice, or antiglaucoma therapy alone may be required. In addition to being diagnostic, a selective carotid angiogram is often therapeutic. Closure of a fistula is a well-recognized entity after angiography, although the fistula often closes spontaneously. Embolization with polyvinyl alcohol or small detachable balloons is another therapeutic option.

Carotid cavernous fistulas (high-flow shunts), which arise either spontaneously or after trauma, are symptomatic in terms of subjective bruits, proptosis, or visual loss. Objectively, the proptosis, chemosis, and ophthalmoplegia are related to the congested orbit. The arteriolized conjunctival vessels that approach the limbus in a corkscrew fashion are characteristic of this condition. CT and B-scan ultrasonography reveal both symmetrically enlarged extraocular muscles and a dilated superior ophthalmic vein. The fistula of spontaneous onset has a better chance of spontaneous resolution, although the shunt often must be closed with either a detachable balloon or carotid artery ligation.

Inflammatory-Infectious Exophthalmos

In inflammatory proptosis, the lesion either is an idiopathic inflammatory orbital pseudotumor (nonspecific orbital inflammation) or has an infectious cause. Both types have a fairly typical sudden onset with associated discomfort. The inflammatory pseudotumors generally have an explosive onset with erythema, swelling, pain, and loss of function. The inflammation may be diffuse or located in either the anterior or the posterior portion of the orbit. The inflammation also can be localized to certain areas or tissues within the orbit, such as in myositis, dacryoadenitis, and perioptic neuritis. Clinically, these patients are not "toxic"

or febrile, as are patients with infectious proptosis. Myositis may involve one or more extraocular muscles and the patient may present with pain, proptosis, diplopia, lid erythema, swelling, chemosis, and injection over the involved muscle(s). The involved muscle(s) will show radiographic evidence of enlargement, often with involvement of the tendinous insertion, a differential point in favor of diagnosing myositis rather than Graves' disease.

Acute dacryoadenitis presents with pain, erythema, and swelling of the upper lid. Proptosis and globe displacement are minimal. The upper outer lid is tender to palpation, and swelling in this area produces a characteristic S-shaped lid. CT shows an enhancing mass in the lacrimal fossa, with edema extending into Tenon's space. The importance of coronal views in the evaluation of lacrimal fossa masses cannot be overemphasized.

Another form of localized orbital inflammation is perioptic neuritis. This simulates optic neuritis, but proptosis and pain with retropulsion of the globe are present. The optic nerve is enlarged on CT.

All three entities (myositis, dacryoadenitis, and perioptic neuritis) share a common feature: an exquisite response to oral prednisone. Most episodes resolve after a short course of prednisone, but for patients in whom the condition is chronic, biopsy and radiation therapy should be considered, particularly when inflammatory pseudotumors and dacryoadenitis are present. If the biopsy reveals vasculitis, cytotoxic therapy may be required. The use of radiation is not advised in this setting.

Endocrine Exophthalmos

Endocrine exophthalmos is one of the most frequent causes of exophthalmos seen in routine clinical practice. The diagnosis is seldom difficult, except in unusual instances of a unilateral presentation. Lid signs provide valuable clinical evidence of an underlying thyroid disorder. This entity is considered in greater detail later in this chapter.

Neoplastic Exophthalmos

Orbital neoplasm also may be a source of proptosis. Most orbital tumors are associated with proptosis, some more than others. For example, a meningioma of the optic nerve sheath takes up little space in the orbit and thus displays minimal proptosis. The same is true of some compressible vascular lesions. However, tumors with firm tissue consistency, such as orbital optic nerve gliomas or neurinomas, are associated with greater proptosis. Tumors within the muscle cone produce axial proptosis, while extraconal tumors displace the eye out and in a direction opposite that of the lesion. Another important clinical feature of orbital neoplasm is the lack of pain, with one notable exception: adenoid cystic carcinoma of the lacrimal gland, because of perineural invasion, characteristically produces a deep-seated orbital pain early in its course.

Operative Considerations

CT features of orbital lesions help to ascertain whether a lesion is benign and resectable or malignant and infiltrative. Rather than list individual tumors, it is more instructive to consider the location of the lesion and plan the operative approach on the basis

of suspected histology and location, as determined by imaging techniques. Lesions in the orbital apex are best dealt with by a combined neurosurgical-ophthalmologic approach. A frontal craniotomy with unroofing of the orbit provides access to this region. The craniotomy may either incorporate the superior orbital rim or spare it. Incorporating the rim involves less need for retraction of the frontal lobes; also the roof fractures approximately halfway back, so there is no need for alloplast material to reconstruct a roof. There remains enough of a roof to prevent a pulsating globe. A disadvantage of this approach is that the frontal sinus is invariably violated. A rim-sparing craniotomy is more likely to be associated with a pulsating globe after operation, although generally the patient is asymptomatic. A technical point regarding the operation is that brain retraction can be minimized by drainage of spinal fluid.

Both approaches are facilitated by microsurgical techniques for work in the orbital apex. This is particularly true in attempts to save the trochlear nerve.

The craniotomy with removal of the orbital roof also can be used to expose the superior portion of the orbit. If the lesion is as large as or larger than the globe, a transcranial approach may facilitate excision of the mass, particularly if it is located extraconally. Intraconal lesions are probably best managed by another approach. A transcranial approach to an intraconal lesion would increase the risk of sensory disturbance, ptosis, and motility imbalance, because of the necessity to retract the frontal nerve and the levator palpebrae superioris and rectus superior muscles. A lateral orbitotomy would be useful for either intraconal or extraconal lesions, especially if the lesion were not located medial to the plane of the optic nerve. Two types of medial orbitotomies are useful in approaching lesions that are medial to the plane of the optic nerve: an anteromedial orbitotomy (conjunctival) with disinsertion of the medial rectus muscle, and a Lynch or lateral rhinotomy-type approach. The anteromedial orbitotomy is useful when the lesion is intraconal; the Lynch-type approach is helpful when the lesion is extraconal. A superior transseptal or subbrow incision also may give adequate exposure.

Medial orbital lesions are approached as previously described. The anteromedial approach may be "augmented" by temporary removal of the lateral wall of the orbit; additional deep exposure can be achieved in this fashion. Caution must be taken because of the possibility of damaging the central retinal artery as it enters the posteromedial optic nerve. A more standard lateral rhinotomy may be performed for inferomedial lesions.

Inferior orbital lesions are also amenable to many approaches. A lateral orbitotomy may be used for lesions below or lateral to the optic nerve, located either intraconally or extraconally. An intraconal lesion sited inferonasally to the optic nerve may best be approached by an anteromedial orbitotomy. Inferior orbital lesions located in the extraconal space also may be exposed by a transseptal approach, provided the lesion is anterior in the orbit. The transseptal approach is performed by marking a subcilial incision under the lashes of the lower lid. As this incision nears the lateral canthus, it is directed posteriorly, as is a standard blepharoplasty incision of the lower lid. This will enhance exposure without sacrificing cosmesis. The skin muscle flap is reflected downward, and the orbital septum can be incised directly over the lesion. The septum may be loosely approximated if the incision is large, but generally a tight septal closure is not done, so that any postoperative orbital bleeding can decompress itself through the incision. The skin muscle flap may be closed in one layer.

The most versatile approach probably is the lateral orbitotomy, performed as described by Maroon and Kennerdell. Lesions in the lacrimal fossa, in the lateral orbit both intraconally and extraconally, and above or below the optic nerve may be approached in this fashion. The incision may be shaped like a "lazy-S" or in the shape of a hockeystick along the lateral orbital rim. This incision does not violate the lateral canthus, and the posterior extension lies in an area that would normally be covered by the bow of a pair of eyeglasses. Hemostasis is aided by the use of 1:200.000 epinephrine subcutaneously before the incision is made. The incision is taken down to the level of the temporalis fascia and undermined. The exposed periosteum of the lateral orbital rim is cut in a T-shaped fashion and reflected off the bone and orbital wall with an elevator. Temporalis fascia (but not muscle) is cut posteriorly. Temporalis muscle is cut from its attachment to the orbital wall. With malleable brain retractors to protect the orbital contents and the temporalis muscle, a reciprocating saw is used to incise the lateral wall of the orbit above the zygomaticofrontal suture. The second incision is made approximately 25 mm below this. A rongeur is used to fracture the bone posteriorly at its attachment to the greater wing of the sphenoid. Additional exposure may be obtained by using a diamond bur to remove bone from the greater wing of the sphenoid. Although this bone can be very vascular, the diamond bur, along with supplemental bone wax, can adequately control the bleeding. A traction suture previously placed around the insertion of the lateral rectus will allow its identification through the periorbital fascia. Entry into the muscle cone above or below the lateral rectus muscle may then be made with confidence. Moist Cottonoids are most helpful when orbital fat is being retracted. A self-retaining orbital retractor and an operating microscope are useful in many orbital procedures. Solid tumors may be "grasped" with an ophthalmic cryoprobe. This method permits gentle retraction without breaking the capsules.

Before skin closure, the bone flap is reinserted and periosteum is closed over it. This provides a splint sufficient to allow adequate healing; we do not routinely wire these bone flaps in place. The skin is then closed in two layers. We also do not routinely use drains. It is our policy to have a dry field before closure. A mild pressure patch is left in place overnight. The use of steroids during and after surgery is optional.

Fine-Needle Aspiration Biopsy

The use of fine-needle aspiration biopsy is not advocated for lesions that are judged resectable by CT. Its greatest value if for infiltrative lesions, particularly in suspected metastatic disease. The procedure requires special training, but most ophthalmologists are familiar with retrobulbar injection techniques for sample collection. The other required member of the team is competent cytologist, who can interpret the tissue sample.

Fine-needle aspiration biopsy has been useful in dealing with suspected orbital abscesses. In addition to being helpful in making the diagnosis, biopsy can provide an offending pathogen that may be cultured before antibiotic therapy. As mentioned previously, we do not advocate that this technique be performed without previous instructions.

Representative Tumors

The most common benign primary orbital tumor is a cavernous hemangioma. This benign encapsulated tumor is found predominantly in females. The average age at onset of

symptoms is 42 years. Because the tumor grows slowly, its presence is tolerated well until the inevitable effects of its mass become apparent. The ophthalmic cryoprobe was first used to assist in the removal of this type of tumor.

The lacrimal gland, which is a type of minor salivary gland, may give rise to an epithelial tumor termed "a benign mixed tumor". This lesion preferentially affects men, whose mean age at presentation is 39 years. Clinically, symptoms may be present for at least a year; proptosis and globe displacement are the cardinal features. Ophthalmoscopy may show the tumor indenting the globe. CT shows a mass in the lacrimal fossa, which may indent the globe (a lacrimal fossa mass conforming to the shape of the globe is a feature of inflammatory or lymphomatous infiltration of the lacrimal gland). There may be thinning of the lateral orbital wall due to pressure. The tumor is best removed by a lateral orbitotomy; it should be delivered with its "capsule" intact. The capsule is a condensation of surrounding tissue that is produced as the enlarging tumor incorporates it. Incomplete removal or rupture of the capsule leads to recurrence in one-third of patients. Recurrent lesions may be more aggressive than the primary tumor.

The lacrimal gland is also host to a malignant epithelial tumor, the adenoid cystic tumor. Generally, symptoms are present for a shorter period than when the lesion is a benign mixed tumor, but in contradistinction, pain may be present early. Adenoid cystic tumors display perineural invasion and bone destruction to account for this. There is no consensus as to what constitutes the proper management of these patients. Patients with features of basiloid tissue were reported to have a median survival of 3 years. Whether exenteration, en bloc excision, or radiation is the best treatment awaits the test of time.

The lacrimal gland is also subject to lymphomatous infiltration. The findings on examination may be indistinguishable from those of a benign mixed tumor, although symptoms are generally present for a much shorter time. CT shows the lacrimal gland molding to the globe. Lymphomas also can infiltrate any structure within the orbit, including orbital fat, extraocular muscles, and even the optic nerve. Once the diagnosis is established, systemic evaluation for lymphoma is recommended. Lymphoma isolated to the orbit responds to radiation in doses of 2500 to 3000 rad (25 to 30 Gy).

Rhabdomyosarcoma is the most frequent primary orbital malignancy of childhood, and its presence should be suspected in any child who has acute proptosis. The average age of presentation is 7 years, and boys are affected slightly more often than girls. A suspicion of this diagnosis should lead to a biopsy without delay. Biopsy specimens may reveal a very poorly differentiated tumor that awaits immunocytochemical or electron microscopic evaluation for final diagnosis. Combination radiotherapy and chemotherapy has led to a survival rate of 80 per cent, although the rate is worse if the tumor extends into the surrounding sinuses.

Metastatic disease to the orbit is usually seen in patients in their sixth or seventh decade. Orbital involvement can occur long after the primary tumor has been treated. As mentioned previously, a scirrhous adenocarcinoma may paradoxically present with enophthalmos on the affected side. Metastatic disease from tumors such as oat cell carcinoma, renal cell carcinoma, and prostate carcinoma may also cause focal enlargement of an extraocular muscle. These lesions may also give rise to metastatic orbital disease. Treatment

is generally nonsurgical, although biopsy should be performed.

Sinus Diseases That Can Cause Proptosis

The association of orbital inflammation and proptosis with purulent bacterial sinusitis is well known and usually easily recognized. At one time, bacterial sinusitis was the most common interrelated orbital-sinus problem. In the developed world the ready use of antibiotics has changed the relative frequency, but in the lesser developed parts of the world the relationship still holds.

Infection from the sinus is spread by two routes: by the venous connections through venous periphlebitis or thrombophlebitis, and by direct spread, usually through the paper plate of the ethmoid bone.

The usual organisms are *Streptococcus pneumoniae, Haemophilus influenzae,* beta-haemolytic streptococci, and *Staphylococcus aureus.* The usual sites are the ethmoid sinus in children and all sinuses in adults.

Proptosis alone is rare in acute bacterial sinusitis. Other manifestations include inflammatory edema of the eyelids, chemosis, and decreased extraocular muscle motion. The final stage, which is not inevitable, is a subperiosteal abscess between the orbital contents and the affected sinus. The globe protrusion occurs anteriorly and away from the abscess and sinus. Abscess within the orbit may occur after the subperiosteal collection progresses into orbital fat or through localization of the orbital cellulitis. Visual impairment and ophthalmoplegia occur. Visual impairment is a result of increased orbital pressure from the inflammatory process.

The symptoms of sinusitis vary according to the sinuses involved and the intensity of the infection. Headache is not the hallmark of sinusitis. Pain and tenderness over the affected sinus and nasal obstruction are usual. Purulent drainage may be observed in the nose or nasopharynx or in both. Purulent drainage in the nasopharynx without nasal obstruction or discharge is seen in isolated sphenoid sinusitis. Tenderness over the affected sinus on palpation is found in antral and frontal infections but not in sphenoid disease. Most acute sinusitis occurs without orbital signs or symptoms.

The clinical diagnosis of sinusitis with orbital involvement is usually obvious. Roentgenograms and CT scans are more often used for treatment planning to specify the target of surgical intervention. The plain roentgenograms and the Caldwell, Waters, and submental vertex views show clouding of the affected sinus, with or without an air-fluid level.

Treatment of sinusitis complicated by orbital inflammation varies with the degree of involvement. Most patients with mild orbital cellulitis are treated as outpatients and given antibiotics and nasal decongestants. Cultures are taken, but the initial antibiotic usually is either some type of penicillin or cephalosporin for patients allergic to penicillin. Only when the abscess is within the orbit and conservative management fails is sinus surgery with drainage of the orbital collection needed. Visual loss and proptosis are indications for drainage when there is an orbital abscess along with cellulitis. The development of chemosis, edema, proptosis, and limitation of muscle motion in the opposite eye requires anticoagulant therapy

as well as an increase in antibiotic therapy for cavernous sinus thrombophlebitis.

Patients who experience a second episode of sinusitis with orbital symptoms should be surgically treated after the episode subsides.

The external ethmoidectomy will provide access to ethmoid cells and the sphenoid sinus when needed, and can be used as an approach to the frontal sinus for drainage as well as an approach to the orbit. If a subperiosteal abscess is encountered, it can be drained into the nose by way of the ethmoidectomy. If the frontal sinus is involved, an inferior opening can be made in the sinus floor and drainage can be accomplished into the nose. In the absence of bone infection, drainage of the frontal sinus is all that is needed. Our group uses a thin rolled sheet of soft Silastic to maintain drainage from the frontal sinus to the nasal cavity for several weeks. The key to the surgical treatment is a careful and complete ethmoidectomy.

Mucormycosis

Rhinomucormycosis is caused by fungi of the Phycomycetes class, which have nonseptate mycelia. The vast majority of active infections are caused by *Rhizopus* genera. Others are caused by *Mucor* and *Absidia* organisms. More than half of the patients have poorly controlled diabetes mellitus. Mucormycosis is also seen rarely in patients with malnutrition, immunodeficiency syndromes, chronic renal failure, cancer, and cirrhosis or patients on long-term antibiotic, steroid, or cytotoxic therapy.

The fungi are normal respiratory flora that proliferate in the sinuses and nose. The sinus proliferation can extend to the orbit. In the sinuses and the orbit, the fungi invade arterioles and arteries, producing thrombosis and ischemic infarctions.

The clinical examination shows black, crusted, and necrotic areas over the turbinates and nasal septum. The eye may show chemosis with decreased ocular motility. Roentgenograms of the sinuses reveal nodular mucosal thickening in the antrum and ethmoids. The frontal sinus is generally spared. Fluid levels are unusual. Bone destruction can be seen in the sinuses.

Biopsy material usually shows the fungus, necrosis, and acute and chronic infection.

The key to treatment is to modify the underlying cause, as well as provide specific antifungal therapy. Diabetes ketoacidosis is treated. If the patient is taking steroids or cytotoxic agents, these regimens need to be modified. Surgical excision of the necrotic areas and drainage are appropriate. At times, aggressive resection or even orbital exenteration is required, because extensive orbital disease can extend to the cranial space and cause death.

Aspergillus

Aspergillus is another fungus found in soil and decaying fruits and plants. Aspergillosis can occur in otherwise healthy people. Polyp disease can recurrent sinusitis in a tropical setting predisposes to the infection. The usual infected area is the antrum and to a lesser degree the ethmoids. The orbit becomes involved slowly over months as a progressive granulomatous process with fibrosis. In the orbits the fibrosis (a granulomatous process)

produces rigid proptosis and optic nerve damage. A roentgenogram will show involvement of a sinus, along with orbital extension. The diagnosis is made on the basis of biopsy, which reveals fungal septa within the matrix of dense fibrous tissue, epithelioid and foreign body-type giant cells, plasma cells, histiocytes, and lymphocytes.

Although amphotericin B is of value in the treatment of mucormycosis, it is not effective for aspergillosis. Wide surgical excision is needed, along with surgical exteriorization and packing.

Wegener's Disease

Wegener's disease, a poorly understood vasculitis with granulomatous features, is most often a pulmonary disorder having associated renal involvement and nasal manifestations. Patients of all ages and both sexes can be affected, but most are adults.

The nasal finding is a characteristic crusting of the septum and turbinates with underlying mucosal necrosis secondary to vasculitis. The histologic verification of small-vessel perivasculitis is the key to the diagnosis. The lesion is seen in the sinuses, trachea, or subglottic larynx together, or initially at only one site. Pulmonary granulomas with cavitation occur. If untreated, this disease can involve almost any organ, including the skin, and death is likely.

Pansinusitis is common. All sinuses may be involved. The antrum and ethmoid sinuses are the regions most often diseased. Secondary infection within the sinuses may add confusion to the diagnosis. When the underlying necrosis extends beyond the sinus lining, bone necrosis may occur. With loss of bone the process may extend to the orbit and present as a pseudotumor having a mass effect on the orbital contents and causing proptosis with limitation of ocular motility, congestion, central retinal vein congestion, and papilledema. The clinical appearance of an orbital inflammation with lid edema and chemosis also occurs. There may also be retinal cellulitis, posterior uveitis, conjunctival inflammation, scleritis, and even corneal involvement.

The vasculitis, the key to the diagnosis, can be seen around arterioles and arteries in any of the sites. The granuloma formation alone is not diagnostic. Granulomas are sometimes found in walls of vessels near the vasculitis.

Diagnosis is made on the basis of appropriate biopsy. Nasal biopsy specimens usually can be taken, but the pathologist needs to be told what to look for, and care must be taken to obtain a specimen of an active lesion and not an associated crust or area of necrosis. Systemic manifestations such as anemia, elevated sedimentation rate, microscopic hematuria, and roentgenographic evidence of pulmonary involvement help in the diagnosis.

The orbital signs of the disease are rarely the first and only manifestation of the process. The clinical findings are those of euthyroid Graves' disease. Proptosis, extraocular muscle mobility problems, and chemosis may lead to the mistaken diagnosis of Graves' disease. Orbital decompression does not improve the situation, and the diagnosis becomes more obvious when there is progression of the other ocular or sinus-nasal symptoms.

The choice of treatment is uncertain. Various immunosuppressive regimens have been used, including cyclophosphamide, methotrexate, chlorambucil, and azathioprine. Recently, long-term antibiotic therapy such as with trimethoprim-sulfamethoxazole has been used empirically on the premise that the disease is an infectious disease and that the organism is unknown.

Giant-Cell Reparative Granuloma

This rare entity usually involves the mandible but may occur in the maxilla or ethmoid, and may extend secondarily into the orbit and cause proptosis. The granuloma occurs in young persons and usually is associated with trauma. The lesion is a bony reaction to uncontrolled repair. The pathologic findings are those of multinucleated giant cells intermixed with a stroma of spindle-like cells. There is variable vascularity, hemorrhage, and inflammation. Conservative surgery is sometimes indicated because of proptosis.

Benign Tumors and Tumor-like Growths of the Sinuses Affecting the Orbit

Inverting Papilloma

The inverting papilloma is the most common nasal benign tumor that involves the orbit. Orbital extension is not common, but a few properly situated, extensive, or very aggressive papillomas have been noted to cause proptosis and other orbital signs.

Most authorities consider the inverting papilloma a true neoplasm. Other names used for this growth are inverted papilloma, schneiderian papilloma, papillary sinusitis, polyp with inverting metaplasia, benign transitional growth, epithelial papilloma, inverted schneiderian papilloma, soft papilloma, transitional cell papilloma, squamous papillary epithelioma, papillomatosis, papillary fibroma, and cylindrical cell carcinoma. These names reflect the various histologic interpretations, which range from an inflammatory disease to a malignant neoplasm.

The usual site of the papilloma is the lateral wall of the nose, with secondary involvement of the ethmoid cells. The growth arises from the epithelium of the mucous membrane. Grossly, the papilloma is a fleshy gray to gray-red mass that can be very extensive. The symptoms are those common to all nasal tumors: unilateral obstruction and occasional bleeding. Pain is unusual. Sinusitis from obstruction may occur secondarily. The orbit is involved by pressure on the orbital ethmoid wall, and the eye is displaced laterally and forward. Papillomas extend to the frontal sinus or frontoethmoid cells. In this rare occurrence, the protrusion of the eye is forward and down.

The epithelial surface is usually composed of respiratory cells or a mixture of respiratory and squamous cells. The growth within a confined space causes the projections of the lining to invaginate upon themselves, giving the appearance of internal papillae; hence, the common synonym of inverted or inverting papilloma. The basement membrane of the papillae usually is intact, but studies of cross sections of tissue may give a false impression of invasion because islands of epithelial cells can be seen within the stroma; degrees of dyskeratosis also may be noted.

There is some doubt whether papillomas undergo malignant changes. Malignant tumors have been documented in areas previously involved with papilloma and have caused death. Although this phenomenon is rare, all papilloma specimens should be examined carefully under the microscope. In the study by Suh and colleagues of 57 patients, four (7 per cent) had invasive carcinoma associated with papilloma; one of the four had received irradiation.

Papilloma is often clinically treated as nasal polyps, and the specimens are not examined by the pathologist. Suh and colleagues noted that 63 per cent of patients who had definitive surgery for papilloma had had previous nasal surgery for obstruction. In 36 patients who had undergone previous surgery, 76 nasal operations had been performed before the papilloma was diagnosed.

Treatment of papilloma usually consists of wide excision through an external approach. The lateral rhinotomy is the approach generally recommended for the usual large growth. Because of the superior localization that is now available with CT, more localized treatment of some papillomas may be possible. A small growth localized to an anterior site may be adequately treated through the nostril if disease is localized to that site only. Lateral rhinotomy removes the lateral wall of the nose, including the turbinates, ethmoid bulla, and uncinate process, and converts the nasal cavity, ethmoid, and antrum on one side into a single cavity. This approach makes follow-up easier, but the loss of the inferior turbinate may have the functional consequences of dryness and crusting. In some patients, when localization is precise, it may be possible to perform a modified lateral rhinotomy safely and to preserve the inferior turbinates. Total removal is needed to prevent recurrence.

Radical procedures such as maxillectomy are seldom required unless histologic evidence of malignant disease is convincing. In that situation, the disease should be treated as a cancer and not as a papilloma.

Orbital involvement with proptosis is managed in a relatively conservative manner as for strictly nasal papilloma.

Mucocele

Mucoceles of the sinuses are benign, encapsulated, mucus-filled swellings lined with respiratory, columnar, or squamous epithelium. The mucocele evolves from isolated and encapsulated mucus-secreting epithelium. There are several theories of how this happens. One theory considers the mucocele a retention cyst that develops from obstruction of the ducts of one or more mucous glands. Another theory speculates on obstruction of the sinus ostium, with retention of the normal sinus mucus within the sinus cavity. The obstruction may arise from any cause, such as trauma, mucosal thickening from chronic sinusitis, or congenital narrowing or obstruction of the ostium.

Mucoceles are most common in the frontal and ethmoid sinuses. Mucoceles are seen in the maxillary sinus, are usually small and asymptomatic, and are generally seen serendipitously on a roentgenogram or at sinus exploration for some other reason. In the maxillary sinus, mucoceles are referred to as cysts. A mucocele is rarely seen in the sphenoid sinus, and diagnosis can be difficult. An infected mucocele in the sphenoid sinus may cause pain, visual loss, or pituitary suppression or insufficiency. As they expand, sphenoid

mucoceles can extend forward into the posterior ethmoids and involve the orbit or may extend upward, causing a superior orbital apex syndrome with diplopia, ophthalmoplegia, and visual loss.

The most common mucocele is the frontal or frontoethmoid lesion. Clinical symptoms and orbital signs vary with the size of the lesion. Some lesions may be asymptomatic, the only sign being when the position of the affected eye is noted to be different from that of the opposite eye. Such a patient may notice discomfort around the eye or may experience intermittent frontal headaches. The expanding lesion usually enters the orbit through the floor, which has the thinnest bone. A cystic soft tissue swelling may be palpated in the mediosuperior angle of the orbit. The mucocele also may erode through an intersinus septum, involve the opposite frontal sinus, and push down into that orbit. Rarely, the mucocele displaces or erodes the anterior wall of the frontal bone and presents as a soft cystic swelling in the forehead. Likewise, the mucocele may penetrate posteriorly into the anterior cranial cavity.

The eye displacement from the frontal mucocele is downward, forward, and lateral. Diplopia occurs after eye displacement.

The ethmoid mucocele is the second most common mucocele. This lesion may erode the paper plate of the ethmoid bone and displace the eye laterally and forward, after which diplopia occurs. The lacrimal system may be obstructed and there is tearing. An infected mucocele is called a mucopyocele. The symptoms are those of acute sinusitis with orbital symptoms.

The roentgenographic findings of mucocele are clouding of all or part of the affected sinus and thinning of the bone in the path of least resistance. For the frontal mucocele, the roentgenographic pattern is that of smooth oval erosion of the floor of the sinus. Plain roentgenograms are usually sufficient for the diagnosis and treatment planning of mucocele. Tomography and CT may better demonstrate loss of bone in the paper plate of the ethmoid bone and sphenoid sinus lesion. The scans and tomograms are also useful in evaluating the posterior wall of the frontal sinus and intracranial extensions. Most mucoceles have a homogeneous character on CT.

The treatment of mucocele is surgical. The frontal sinus is opened either by expanding the ethmoidectomy from below, or by way of an osteoplastic frontal flap using a coronal incision. It is unsettled whether these lesions should be merely exteriorized by ethmoidal nasal drainage and nasofrontal duct reconstruction, or whether the sinus should be obliterated. The uninfected frontal mucocele is usually seen with relatively normal or normal ethmoids and nasofrontal area. In that case, a complete ethmoidectomy with extension of the bony removal to enter the frontal sinus exposes the lesion. If the mucosa that lines the nasofrontal duct area is normal, excision of the cyst with good drainage into the nose is all that is usually needed. If the nasofrontal duct area is devoid of mucosa, the duct can be relined with a flap folded up from either the nasal septum medially or the superior turbinate laterally. The key to the drainage of the frontal sinus is complete ethmoidectomy plus a mucosa-lined frontal duct area. A coronal incision and osteoplastic flap are often excessive for the simply uninfected mucocele. In this situation, sinus obliteration likewise is not usually needed.

Complete ethmoidectomy by an external or intranasal approach, depending on the surgeon's experience, is usually enough for excision of the ethmoid lesion. Sphenoid sinus mucocele may be approached and drained either by way of the external ethmoidectomy or by the transseptal approach to the sphenoid. Maxillary sinus mucoceles do not always need treatment. They are often present as asymptomatic sinus opacification on a sinus roentgenogram taken for some other reason. The Caldwell-Luc antrostomy is the traditional approach. Mucoceles that require treatment may also be approached transnasally or externally, using the newer micronasal sinus surgery techniques. Some sort of nasal antral drainage augmentation - either an inferior meatus nasoantral window or middle meatus enhancement - is believed to be needed after a maxillary mucocele is evacuated.

In mucopyocele, antibiotics should be used both preoperatively and postoperatively. Cultures provide guidance in the selection of the proper antibiotic.

Osteoma

The frontal sinus is the most common site of this benign, slow-growing bony tumor. The tumor often is asymptomatic and is seen on skull or sinus roentgenograms taken for other reasons.

When symptomatic, the osteoma may cause proptosis, facial asymmetry, and displacement of the eye. Only when the lesion obstructs a sinus ostium is there pain or sinusitis. The otherwise asymptomatic frontal or ethmoid osteoma may subtly or obviously displace the eye. The ethmoid osteoma may displace the eye laterally and present as a firm palpable mass in the upper inner corner of the orbit. Sphenoid sinus osteomas are rare. As with the ethmoid osteoma, when large enough, these benign growths may lead to papilledema and optic atrophy with obstruction of the blood supply of the optic nerve. Amaurosis fugax with change in eye rotation has been reported.

Diagnosis requires roentgenography. The lesion is radiodense, well outlined, homogeneous, and calcified. Microscopically, it contains a cortex of compact, laminated bone with a central core of trabeculated bone, blood vessels, fibrous tissue, and fat.

Treatment is needed only when the osteoma is symptomatic. When the lesion is in the frontal sinus, either the frontal osteoplastic flap or a frontoethmoidectomy approach is used, depending on the size of the osteoma, its location in the sinus, and the size of the sinus. Osteomas without symptoms or progression that are located away from the frontal sinus ostia do not require treatment. The ethmoid lesion is approached by means of external ethmoidectomy. The sphenoid sinus lesion is also approached favorably by an external ethmoidectomy. These growths in a tight space such as the ethmoid or sphenoid sinus may be difficult to dislodge and remove. A drill with suction irrigation may be used to core out the lesions and permit their infracturing and piecemeal removal, to avoid what could be dangerous pulling and tearing of important surrounding structures.

Fibrous Dysplasia

Fibrous dysplasia is a benign growth consisting of bone-forming fibrous tissue in one of the bones of the face and sinuses. When the tumor is polyostotic and occurring with

precocious puberty and cutaneous pigmentation in females, the condition is called Albright's syndrome. In the polyostotic form, deformities of the long bones may occur, along with the skull lesions. The disease does not progress rapidly. When the dysplasia is monostotic, eye symptoms are prominent. Proptosis, deformity of the orbit, and even optic pressure atrophy may occur.

In most patients with fibrous dysplasia of the sinuses, the orbital occurrence is a part of Albright's syndrome. The bones and sinuses involved are the frontal, sphenoid, and ethmoid. Also, disease in the zygoma and maxilla may encroach on the orbit. Moore's review of the ophthalmic literature describes the rapid onset of proptosis over weeks to months. There is downward displacement of the globe, diplopia, nasal obstruction, and epiphora. With sphenoid sinus involvement, optic atrophy occurs.

The roentgenogram shows a homogeneous lesion having a ground-glass appearance, with a gradual blending into normal bone; this appearance comes from the mixture of tiny spicules of bone within the dysplasia. There is no capsule in fibrous dysplasia. Microscopic examination reveals a moderate to highly cellular fibrous stroma composed of uniform, benign-appearing spindle cells. Irregular trabeculae of bone without lamellae and osteoblastic rims are mixed in the fibrous stroma. The condition is considered to be a bony hamartoma.

Treatment is needed only when there are symptomatic growths; when the process threatens important structures, such as the eye or optic nerve; or when there is consequential cosmetic deformity or serious pain that is clearly related to the process. Decompression of the optic nerve has been performed for the treatment of sphenoid sinus lesions, but the results are unpredictable. Radiotherapy should not be used because osteosarcoma may be induced in lesions so treated.

Juvenile Nasopharyngeal Angiofibroma

Juvenile nasopharyngeal angiofibroma is a peculiar benign tumor typically found in young boys who have nosebleeds. Symptoms may be present for years.

The precise origin of this tumor is subject to some speculation. At onset the tumor is a lateral nasal growth that seems to originate in the posterior nasal cavity rather than the nasopharynx itself. The specific site is the posterolateral and superior nasal cavity at the point where the sphenoid process of the palatine bone meets the horizontal ala of the vomer and the root of the pterygoid process of the sphenoid bone. This site is near the upper margin of the sphenopalatine foramen.

The site of origin permits extension beyond the nasal cavity at the time of usual diagnosis. The juvenile nasopharyngeal angiofibroma grows forward behind the posterior wall of the maxillary sinus, pushing the thin bony wall forward. The tumor may grow laterally into the pterygoid fossa; from here the lesion may grow into the inferior and the superior orbital fissure, pushing the eye forward and sometimes forward and down. The tumor may erode the greater wing of the sphenoid by pressure, and tumor may be left extradurally in the middle fossa near or adjacent to the cavernous sinus. Posterior extension into the sphenoid sinus through its floor or ostium fills the sinus and pushes upward and back to displace the pituitary gland and fill the sella turcica. Tumor in the orbit or sella turcica may cause loss of vision.

The rate of growth is not known but is usually slow.

Because the tumor is rarely seen in adults, spontaneous regression is believed to occur. Clinical regression after incomplete removal or after radiation therapy is recognized. Since regression cannot be assumed and most of the lesions are symptomatic, the tumors should be removed.

Radiographic Studies

The plain roentgenographic findings are characteristic. Holman and Miller emphasized the posterior bowing of the posterior wall of the maxillary sinus. CT permits precise localization because the tumor is seen as a defined homogeneous mass. Some studies have advocated angiography as being useful in the diagnosis and treatment planning, but knowledge of the vascular supply provides no extra options, and the usual major feeding vessel is the internal maxillary artery.

Other vessels that may contribute to the blood supply of the tumor are the dural, sphenoidal, and ophthalmic branches from the internal carotid system and vessels from the thyrocervical trunk. Because of this diverse blood supply, preliminary ligation of the external carotid artery is of little help in limiting bleeding at excision. Ligation of the artery before definitive surgery may increase the problem of bleeding by encouraging the blood supply from the inaccessible vessels. At times the internal maxillary artery is pushed forward by the tumor and may be ligated early in the exposure, which may decrease the bleeding. Because the histologic appearance of the tumor may vary from predominantly fibrous to more angiomatous, it is difficult to assess the value of a preliminary procedure to limit loss of blood.

Many techniques have been advocated to decrease blood loss during surgery for angiofibroma. These include arterial ligation, embolization of some of the arterial supply from the external carotid system, electrocoagulation, interstitial irradiation, hormones, cryotherapy, and external irradiation. Anesthesia adjuncts, such as hypotensive techniques and hypothermia, also have been recommended.

The value of all of these is conjectural because some tumors bleed more than others. Experience with the management of these tumors has resulted in the discarding of the less practical adjuvants. The internal maxillary artery is ligated if it is anterior, and angiography is not necessary to determine this. Hypotensive anesthesia techniques are introduced if the preliminary bone work for exposure is particularly bloody. Surgery for large juvenile nasopharyngeal angiofibromas requires rapid and efficient procedures, alert anesthesia personnel, continued communication between the surgeon and the anesthesia stuff, and multiple routes for the infusion of blood. Surgery for angiofibroma may be dangerous, but there are few reported deaths associated with the initial treatment.

Large juvenile nasopharyngeal angiofibromas that extend to the middle fossa and cavernous sinus are considered by some to be inoperable or incurable, and external-beam radiation has been recommended for their treatment. In fact, these large tumors can be removed with some degree of safety by an external extracranial approach. They are seldom intradural or intracavernous. The dura and the cavernous sinus may be displaced but usually

are not invaded. An extended lateral rhinotomy always access to the tumor, which may be displaced from the hole created by its progression by means of progressive packing away from the floor of the middle fossa. Cavernous sinus bleeding is low-pressure bleeding that is controllable by packing. The internal carotid vessels are near the tumor, but if vigorous evulsion is avoided they can be left undamaged. Intracranial staged or simultaneous exposure offers no extra safety, either in decreasing the surgical risk of frequency or in frequency of incomplete removal.

Extension of the tumor through either the inferior or the superior orbital fissure may be managed in the same manner as an extradural intracranial tumor. The tumor pushes into these fissures and, in the process, widens them. With the rhinotomy approach, the cheek flap is lifted as far laterally as the zygoma, and the tumor projection into the orbit can be levered out, much like extracting a cork from a wine bottle. Care must be taken to avoid leaving a projection of tumor in the orbit, but this can be checked by inspecting the specimen while it is being extracted from the orbital fissure. The surgery for angiofibroma is not the same as that for invasive cancer. The goal is to remove all projections of this benign tumor but not to create an oncologic block.

These large tumors recur in some patients. All recurrences do not have to be retreated. The purpose of treatment is to eliminate symptoms, which consist of epistaxis and nasal obstructions; unless the patient is symptomatic, expectant watching is judicious. The exception is the lesion that recurs within the sella or with suprasellar extension; in this situation, radiation may be appropriate. There is no evidence in the literature that asymptomatic recurrent juvenile nasopharyngeal angiofibromas have caused death, although there is evidence that retreatment itself has.

Taking a biopsy specimen may be bloody, and the tissue diagnosis is usually apparent from the clinical presentation and the roentgenographic studies. Preliminary biopsy may be more trouble than it is worth, and excisional biopsy is preferred at the time of definitive treatment, when the patient is asleep and prepared for tumor removal.

There are many surgical approaches to the juvenile angiofibroma. Neel and colleagues cited Hellat's 1911 survey in which he documented 65 different techniques used to remove the tumors. There are about eight logical approaches: through the nose, through the palate, through the mandible, through the zygoma, through the bed of the hyoid bone, through the antrum, by combined craniotomy and rhinotomy, and by lateral rhinotomy.

Of all the surgical approaches, the lateral rhinotomy is the most logical and versatile. It is relatively unrestricted and may be modified to provide access to all potential sites of extension, including the extradural, anterior, and middle cranial fossae. The rhinotomy provides lateral, inferior, and medial access rather than the single frontal, inferolateral attack permitted by the first six approaches mentioned. The rhinotomy is an old operation that allows, in one procedure, the removal of the extensions and point of origin of this tumor, without disturbing facial growth or causing deformity.

Radiation Therapy

Some authors have advocated that radiation therapy be used for the juvenile nasopharyngeal angiofibroma, but there is concern about this. Biopsy is still needed, and this has risks. The purpose of treatment is to control nosebleed and nasal obstruction, but irradiation is a slow and ineffective way to treat either of these. The regression of tumor in patients so treated is slow.

Even if all tumors regressed and all symptoms disappeared, the concern about using radiation in young patients remains, because of the long-term carcinogenic effect. Angiofibroma is a benign tumor, and short-term expediency should not be permitted to create later problems. The proponents of radiation base their recommendations on the mortality rate associated with tumor removal and on the fact that the amount of radiation given is only half that used for malignant tumors. With an experienced surgical team, there should be little risk of death from excision, even with extradural intracranial extension. The amount of radiation used is irrelevant.

Ameloblastoma

Ameloblastoma is a benign odontogenic tumor that arises from oral ectoderm. The tumor can arise in the enamel organ, either within the dental lamina or in the labyrinth crest cells. The tumor also arises from the epithelium of odontogenic cysts, surface epithelium of the jaw, and other bones of the body, particularly the tibia.

Clinically, ameloblastoma is a slowly enlarging mass that expands within the involved maxilla. It may displace teeth, and if the orbit is involved the eye may be displaced, with resultant diplopia.

Radiographically, the tumor may be cystic or solid. Neighboring bone is displaced rather than invaded. Dental radiographs show that the tumor may enclose adjacent teeth rather than displace them. Reabsorption and exposure of dental roots is observed.

The microscopic appearance is that of interlacing strands of odontogenic epithelium within a fibrous connective tissue stroma. Cystic degeneration may be seen and squamous cell metaplasia is common. These tumors do not undergo malignant degeneration.

Complete removal is required; incomplete removal is followed by local recurrence. Total maxillectomy may be needed when the tumor is large but orbital exenteration usually is not necessary. Because orbital periosteum is almost always intact, the orbital contents can be preserved. Either orbital floor support or reconstruction may be needed, but the intact periorbita is usually sufficient, along with the remaining medial orbital support, to sustain eye position.

Malignant Sinus Tumors That May Involve the Orbit

Malignant tumors of the nose and paranasal sinus are rare, and the opportunity for one surgeon or center to have an extensive experience with nose and sinus tumors is limited. The tumors tend to be aggressive, and in some series as many as 75 per cent of the growths

extend beyond the site of origin. The orbit is a common site of secondary involvement. Only thin bone separates the orbit from the four surrounding sinuses. Also, the orbital fissures and foramina provide a preexisting route for extension of the tumor. This proximity allows various eye signs and symptoms to occur early in the course of the tumors. Eye symptoms include epiphora from nasolacrimal drainage obstruction, globe displacement by mass effect, limitation of eye motion, diplopia, conjunctival chemosis, bony orbital erosion, palpable orbital mass, and fundus findings due to pressure. Orbital infection as a secondary effect is also possible.

Frequency of Site of Origin

Hospital studies characteristically report that about 55 per cent of nose and sinus tumors originate from the maxillary sinus, 35 per cent from the nasal cavity, 9 per cent from the ethmoid sinus, and the rest from the frontal and sphenoid sinuses. The reality is that these sites merge together, and at diagnosis the site of tumor origin may be surmised on the basis of the place of dominant involvement. In treatment planning and execution, the nasal cavity and paranasal sinuses must be considered as a single oncologic unit. Symptoms depend on the site of the tumor. Maxillary tumors that arise from the floor are less likely to involve the orbit than are superior maxillary tumors or those originating from the ethmoid or sphenoid. Frontal sinus tumors are extremely rare.

Patients with sinus neoplasms show few or no early symptoms, and the symptoms that they do have are not unique for serious disease. Thus, most sinus tumors that present with orbital signs or symptoms are advanced at diagnosis. Pain is not a prominent finding of any sinus disease, except acute infection. Therefore, facial pain, facial neuralgia, or facial sensory changes require particular diagnostic attention. Pain with roentgenographic evidence of bony destruction requires the taking of tissue for diagnosis. If there is cloudy sinus without bony destruction, the tumor is not likely to be malignant. A cloudy sinus associated with pain, eye signs, or sensory changes also is suggestive of a tumor.

Roentgenographic Diagnosis and Treatment Planning

Plain sinus films are seldom adequate to evaluate the suspicious sinus properly. CT has been very valuable in providing evidence of bony involvement. CT has supplanted conventional polytomography in the accurate diagnosis of malignant sinus disease. The physician who orders these studies must be assured that the scan includes all the sinuses and the base of the skull. Both coronal and frontal projections may be needed for treatment planning.

Even with the most sophisticated radiographic assessment, the data usually are inadequate for diagnosis and especially for planning treatment. CT is much superior to what has been available in the past. Differences in tissue density allow conjecture about where tumor stops and inflammatory tissue begins, but it is not possible to distinguish tumor accurately on a microscopic basis. The actual situation may still be better or worse than that seen on the scans. Robin and Powell emphasized the likely sites of radiographic error: the ethmoid complex, pterygopalatine fossa, infratemporal fossa, and orbit. They noted that there is still an error of about 33 per cent when the usual methods of tumor assessment are used, although all patients in their study did not undergo CT. Surgery remains the ultimate staging

procedure.

There are some guidelines regarding when a sinus tumor is not likely to be cured by operation. Tumor into the nasopharynx is unlikely to be cured; some aggressive surgeons using so-called skull base surgery have attacked this site, but results have been questionable. Extensive bilateral involvement high in the nasal cavity, distant metastasis, and insurmountable regional disease all contraindicate a curative surgical effort.

Complete tumor removal is unlikely if there is bony destruction of the base of the skull, with the exception of the cribriform region, where an anterior cranial approach may make a block possible. The farther back that a tumor extends into the ethmoid, the less likely complete resection becomes. Likewise, posterior orbital extension decreases the probability of cure. Cure is unlikely if there is bony destruction of the orbital plate of the frontal bone, the posterior wall of the frontal sinus, the wings of the sphenoid bone above the infratemporal fossa, or the pterygoid process of the sphenoid bone - although surgery is not contraindicated. The decision not to operate is a difficult one. When these sites are involved, the probability of a meaningful operation is decreased if a goal of cure is declared.

Histologic Types

Squamous cell carcinoma is the most common malignant neoplasm of the sinuses. Some of these lesions originate in the sinuses and others in the lateral wall of the nose. Differentiating them by site is difficult and unnecessary, because the lateral wall of the nose is the medial wall of the maxillary sinus and part of the ethmoid complex. Squamous cell carcinoma accounts for 75 to 90 per cent of the paranasal sinus tumors. There is a male predominance of almost 2:1 and the tumor is more common in the elderly. Whereas the sinuses are lined with respiratory epithelium, the squamous cells of the neoplasm are presumed to develop after metaplasia secondary to chronic infection, irradiation, or carcinogen (such as nickel dust) exposure.

The appropriate choice of treatment of these tumors is unresolved. Current therapy is not much different from what was done 100 years ago. In 1848 Fergusson noted that "for centuries tumors have been removed from this area"; the earliest reference he found in the English literature was in 1671. The primitive operations consisted of cutting, tearing, and scraping away far-advanced tumors and applying hot irons to the base of the tumor. Fergusson contrasted that approach to the "modern" operation of the 19th century, which consisted of exposing the area by a skin incision, sawing and chiseling the bony attachments, and wrenching the part out with a large forceps. Currently, the same is done but the tools are newer. Instead of inserting hot irons, we use such adjuncts as ionizing particles, pulsed laser beams, profound cold, hyperthermia, and toxic systemic drugs to improve the results of treatment, which were unsatisfactory in Fergusson's time and are still not very good. The introduction of each new modality has provided the promise of more sophisticated and successful treatment at additional expense. Electrosurgery was followed by radium and radium by conventional x-ray, cobalt irradiation, and now particulate emissions. After the initial enthusiasm for all these waned, surgeons tended to return to surgical procedures with knives, hammers, saws, and chisels.

The current preferred treatment for large sinus cancers is a combination of radiation, chemotherapy, and surgery. The rationale of combined therapy is that each of these modalities when used alone has some effect on the tumor and some likelihood of eliminating it. If these tools can be used together, the net effect in terms of cure should be improved over that of a single modality used alone. However, the rationale breaks down in practice. Many combinations of radiation, surgery, and chemotherapy have been tried, but the net effect in terms of survival is not obvious. Patients die as a result of distant metastasis rather than local failure, or they may die of the treatment. However, the number of survivors at 5 years is not much different according to stage of lesion than the results of surgery alone. It is not even certain whether radiation should be used, and if so, whether before or after surgery. Irradiation before surgery is appealing from a radiobiologic point of view because the oxygenated parts of the tumor will be reduced in volume. It is not known whether volume reduction makes any difference, and the prognosis does not seem to be better if viable tumor is not found at histologic study of the surgical specimen.

So many of these tumors are partially necrotic that a hypoxic tumor is almost always radiated. It may be that whatever survival benefit is derived from the preliminary radiation is canceled out by the tumor being left longer in the lymphatic field, which has been altered by the radiation, or by the effect of radiation on the resistance of the host tumors.

Currently, postoperative radiation is advocated by some. This is an old idea that has been criticized in the past. The theoretical concept of postoperative radiation is one of "microscopic disease", which is based on an animal experiment using a mouse adenocarcinoma model. Cohen recognized that about 90 per cent of microscopic clusters of cancer cells can be killed by radiation given within the range of normal tissue tolerance. In human cancer, it is conjectured that if all gross cancer is removed, the radiation will kill the cells that the human eye cannot see and the patient will benefit. This concept has not been verified in the treatment of human squamous cell cancers, and whatever support there is for the use of postoperative radiation on human cancer has more to do with site of failure than increased survival at any stage of sinus cancers. If this concept of "microscopic disease" were accepted as an oncologic concept, the implications are that bits of cancers can be left behind as long as they are microscopic. It may be true that surgery cannot remove every single cancer cell, but to accept the premise of radiation for "microscopic cancer" invites taking the concept one step further and accepting the idea that less than radical surgery may be tolerated if radiation is to be added later. This is a tempting concept for the treatment of sinus cancer in which radical surgery has functional consequences in that eyes and palates are at risk. This concept was actually proposed by Robin and Powell, who questioned whether "radical" excision is ever indicated, because they believed it succeeds so rarely; they proposed and use subradical excision. "Subradical" means subtotal tumor removal, which others might call debulking, whereas "radical" implies an effort to remove the entire tumor and a margin of normal tissue. The argument is that, if trying to perform a complete operation it not satisfactory, a less radical one followed by radiation may be just as good. This thinking is a major deviation from contemporary oncologic thought and is difficult to accept on the basis of current evidence.

For these tumors, radiation alone is not satisfactory. The anatomy of the nose and sinuses and the vagaries of the tumors in and near bone make the delivery of homogeneous radiation difficult. Tumor hypoxia or necrosis, overlapping fields, and undertreatment of

portions of the tumor make the actual cure of a sinus tumor by radiation alone unpredictable and unlikely.

The use of radiation in combination with surgery for early lesions curable by surgery alone must be carefully considered in light of the long-term problems associated with radiation. Morita and Kawabe observed that, in all their patients who received 5800 rad (58 Gy) to the eye, severe panophthalmitis with corneal ulceration developed within 2 years of the radiation. Of 21 eyes exposed to between 2800 and 5400 rad (28 and 54 Gy), 18 experienced visual disturbances and radiation-induced cataracts.

Surgical excision, when practicable, is still the mainstay of the treatment of squamous cell carcinoma of the sinuses. With complete removal, patients do better than if tumor is left behind. The concept of subtotal removal (debulking) is attractive but dangerous. Treatment of small tumors is more successful than that of large ones. Early diagnosis is the best way to improve survival. Combined therapy is acceptable and may help to save an occasional palate or eye, but has done nothing to improve survival. Currently, nobody knows the most effective way to combine radiation and surgery.

Adenocarcinoma

Tumors of the minor salivary glands and glandular epithelium within the sinuses are a diverse group, all of which may involve the orbit in the same manner as squamous cell carcinoma. There are two types of adenocarcinomas: adenoid cystic carcinoma and "other" glandular tumors. each may be high grade or low grade.

Adenoid Cystic Carcinoma

Low-grade adenoid cystic carcinoma is a form in which the histologic pattern is a mixture of cellular and cribriform formations of epithelial cells, with a few solid cellular areas. The cribriform pattern is the classic "Swiss-cheese" pattern of cells arranged in elongated tubular structures. The high-grade tumor has the pattern of the classic tumor, but solid areas of malignant cells make up most of the mass. This distinction between high and low grade is probably important because vascular invasion and distant metastasis are more frequent and death is earlier with the high-grade tumors. Perineural invasion is a common feature of both types of tumor. This route of spread makes the surgical removal more complex and less predictable and is a reason for frequent local recurrence, regardless of the extent of surgery.

Surgical excision is the mainstay of treatment. The surgery must be extensive, when practical, because the more thorough the excision, the longer a patient may be expected to be free of tumor. The concept of cure with this tumor is relative. Patients may live a long time after treatment, with or without local recurrence. There is considerable doubt whether cure can be achieved in any patient if cure is defined as freedom from any trouble throughout the individual's life, but there is ample justification for aggressive treatment, nevertheless. When an oncologic block can be obtained, long trouble-free intervals may be anticipated. Five-year survival is not adequate to define cure. The 10-year recurrence-free rate for patients with nasal tumors is approximately 60 per cent, a figure that encourages the surgeon to treat. Some patients live their normal life span and die of other causes, with or without recurrence and

even with pulmonary metastasis.

The role for radiation therapy is not well defined. There is no evidence that preoperative or postoperative radiation will decrease the frequency of local, regional, or distant metastasis. With low-grade tumors, which are the most common, the radiation should be reserved for patients who have recurrence and pain. These carcinomas regress with radiation, and symptoms may be lessened or eliminated, but the relief is seldom permanent.

Some tumors are so extensive that they may fill the roof of the nose and both ethmoid spaces and extend into both orbits. Surgery requires the removal of the entire midface, the floor of the anterior cavity, and both eyes. This is not practical and is not likely to cure the patient. In a situation such as this, local "debulking" with a goal of preserving vision can be justified. There is often very little pain, and in very elderly patients the decision not to treat may be proper. Support and local treatment without surgery can be provided to care for some elderly patients who have extensive midface tumors. These patients can live out their lives and usually die of something other than the tumor.

Other Adenocarcinomas

Other adenocarcinomas are classified in several ways. One method uses the same terminology that describes tumors of the minor salivary glands. The terms used are mucoepidermoid and acinic cell cancer. Batsakis and associated described a practical classification based on growth form: papillary, sessile, and alveolar-mucoid. In the first two forms, mucin production is variable and the level of cellular differentiation may range from modest to poorly differentiated.

Papillary adenocarcinomas are usually the most localized of the three forms. A special type of mucus-producing papillary adenocarcinoma is described as "colonic type" because the tumor has the microscopic appearance of a colon cancer. It is important to be aware of this variant, because one should avoid extensive investigation of the digestive tract for a primary tumor on the assumption that the nasal-sinus tumor is a metastatic lesion.

A sessile adenocarcinoma has a broad base and surface and has cells that are not typical for a sinus origin. These tumors are invasive and are associated with a poorer prognosis than is a similarly staged papillary tumor.

The alveolar-mucoid tumor is the most aggressive and is associated with a poor prognosis. The tumor is characterized by abundant mucin, in which there are nests of individual malignant cells. All these "other" adenocarcinomas are more aggressive than the usual adenoid cystic carcinoma. An Armed Forces Institute of Pathology (AFIP) study of 50 cases suggests that low-grade tumors can be differentiated from high-grade tumors and may be treated with more optimism. For high-grade tumors combined therapy is used, but the results are disappointing.

Sarcoma

Sarcomas of the sinuses are rare. Osteosarcoma of the maxilla tends to occur in the older age group. The prognosis is poor, the overall survival at 5 years being 19 to 30 per cent.

Some of these sarcomas are radiation induced. The only chance for cure is complete excision. The exception to this grim generalization is the chondrosarcoma that arises in the nasal cavity. Sarcomas of the sinuses are slow growing and usually quite large when diagnosed. Coates and colleagues reported 13 nasal chondrosarcomas seen during a 25-year period at the Mayo Clinic. The typical chondrosarcoma was histologically low grade and was seen as a pale, glistening obstructing mass that sometimes extended into the medial orbit. Local excision was used in seven patients, with recurrence in five. Definitive en bloc resection cured four of six patients. The others had a long protracted course, with local recurrences and finally death from intracranial extension. Radiation therapy did not influence tumor growth after recurrence. One of us (LWD) performed bilateral maxillectomy in a 32-year-old female who had a unilateral recurrent nasal chondroma. She had had a wide local excision by rhinotomy at the age of 19 years. She was free of disease at 8-year follow-up.

Olfactory Esthesioneuroblastoma

A rare neuroepithelial malignant lesion that originates in the olfactory area high in the nose is the olfactory esthesioneuroblastoma. Other terms used for this tumor are olfactory esthesioneuroma, neuroesthesioma, and olfactory neurocytoma. The growth was described by Berger and Luc in 1924. The olfactory neuroectoderm is the presumed site of origin. Cantrell and associates noted that 160 of these tumors had been reported since 1924, and 125 during the previous 15 years. This finding reflects either a greater awareness or an increased incidence. It is more likely that these tumors were called something else in the past.

Orbital extension is not uncommon and the first sign may be eye displacement, although nasal obstruction is more usual.

On gross examination the tumor is seen as a red polypod mass high in the nose. Symptoms may be present for months or years. One patient had progressive nasal obstruction for 10 years and complete obstruction for 6 years. The tumor may be multicentric, with separate tumors above and below the cribriform plate. Olsen and DeSanto described a patient who had two separate tumors (one above and the other below the cribriform plate) with no gross or microscopic connection between the two. Djalilian and colleagues found that in one of four patients the tumor either was in the anterior cranial cavity at diagnosis or developed later. They noted metastasis in eight of 19 patients studied. Metastatic lesions in cervical lymph nodes were present in two children at diagnosis and in two adults 6 and 8 years after the nasal tumors were removed. One of us (LWD) has treated a patient in whom bilateral parotid metastasis, bilateral cervical metastasis, thyroid gland metastasis, a frontal lobe esthesioneuroblastoma, and then a local recurrence developed 6, 9, 11, 13, and 17 years, respectively, after the nasal tumor was removed. The esthesioneuroblastoma is obviously an unpredictable or diffusely multicentric tumor having a variable clinical behavior.

Surgical removal is recommended. Kadish and associates described a patient whose tumor responded to radiation. Frontal cranial resection has been advocated, but there is no long-term follow-up to document that this will result in fewer local recurrences. The combined cranial facial resection is a sound concept, but because so few patients have been treated and staging has been inadequate, it is difficult to know the ideal role for this approach. Postoperative radiation likewise is worth considering in the treatment of clinically aggressive tumors. Lateral rhinotomy may be adequate for small tumors that originate in the lateral wall

of the nose, away from the cribriform area. Persistence in the treatment of recurrent disease, intracranial tumor, and manageable metastasis may be worthwhile in keeping patients from complications or death. The concept of a 5-year period after which a cure may be declared is not appropriate for this tumor.

Graves' Ophthalmopathy

Graves' ophthalmopathy is usually related temporally to hyperthyroidism but can be seen in euthyroid or hypothyroid persons. The diagnosis seldom is a challenge because of the classic orbital changes or overt hyperthyroidism, although the diagnosis may be elusive in clinically euthyroid patients with asymmetric proptosis. In these patients the most helpful signs are lid retraction and lid lag. A temporal flare of the upper lid, which seems characteristic of thyroid eye disease, also has been noted. Other features of ophthalmopathy include orbital congestion (injection and chemosis of the conjunctiva and lid edema), myopathy, and optic neuropathy, with or without proptosis. None of the eye signs are pathognomonic of thyroid eye disease. At times, the diagnosis in subclinical hyperthyroidism is made or confirmed with the suppression test of thyrotropin-releasing hormone. Another newer laboratory test is the serum analysis of thyroid-stimulating hormone, which is easier to perform and provides the same information.

CT of the orbit usually shows enlarged extraocular muscles, which is the principal cause of proptosis. The inferior rectus is usually the first muscle involved, followed in order by the medial rectus, superior rectus, and lateral rectus muscles. Because of congestion at the orbital apex, a dilated superior ophthalmic vein may be seen in Graves' ophthalmopathy. The muscle enlargement is fusiform, with sparing of the tendinous insertions. This finding, in combination with the characteristic muscle enlargement, differentiates Graves' ophthalmopathy from other entities such as orbital myositis. A bulging orbital septum due to protruding fat is another useful CT finding.

The eye findings in Graves' ophthalmopathy vary from mild protrusion, which may be attractive, a stare, to blindness. The anterior protrusion of the eye is the only natural way that the globe can accommodate to the increased tissue volume in the orbit. Treatment is needed only when the anterior protrusion reaches a point at which there are other problems. Because muscle myopathy is the basic pathologic change, muscle dysfunction may be a problem if the muscle disease is asymmetric. The result is diplopia. The optic neuropathy ranges from visual loss to visual field defects. Corneal exposure has been listed as a common problem in Graves' ophthalmopathy, although this seems to be rare. Of more than 450 patients treated by orbital decompression, only a few had corneal exposure and ulcers.

The visual loss and the degree of proptosis appear to be inversely related: ie, patients with severe proptosis usually have little or no visual loss. Thus, exophthalmos probably is a natural anterior orbital decompression. Some patients have extreme proptosis with no congestion, diplopia, or visual loss; to these the problem is cosmetic and practical; at times, their eyes touch their glasses.

There is no consensus concerning (1) whether the orbital space should be enlarged, (2) the indications for enlarging the space, (3) the sequence of therapy that is most successful in restoring the eyes to near normal, (4) the value or mechanical enlargement of the space,

and (5) the nonsurgical alternatives that are useful in seriously affected patients, and who (what specialty) should enlarge the space.

Because of these variable clinical possibilities, there can be no single treatment. The ideal treatment would be to identify the process that causes the muscle enlargement and reverse it, but currently this is not possible. Present therapy includes reassurance, lubricating eye drops, oral steroids, orbital radiation, and orbital decompression.

Oral steroids have been used for years, with variable effects. A common observation is that the congestive signs and even the visual loss improve during oral steroid therapy, but that the symptoms return when the therapy is decreased or stopped. An observation of 103 patients given 40 to 120 mg of prednisone before orbital decompression is illustrative. In this group, 26 improved while on drug therapy; an improvement was recorded if the congestive signs regressed and pain was relieved. A positive effect was also recorded if vision improved in a meaningful way or if proptosis regressed by 3 mm or more. In terms of these broad and mixed criteria, one of four patients improved. All 26 patients who improved underwent orbital decompression later because of the side effects of steroids (facial swelling, emotional changes, diabetes, and osteoporosis). Some of the patients requested decompression in order to discontinue the use of steroids. These results should not be taken to represent the global value of steroids. The group was selected because they ultimately underwent decompression. Nevertheless, steroid intolerance or failure was a major secondary indication for orbital decompression.

Radiation likewise has been used, with mixed results. Improvement in congestive changes has been reported. Visual loss was stabilized in some patients, but the results were not predictable.

Bilaterality

Graves' ophthalmopathy is a bilateral disease, although it may be asymmetric and may seem to be unilateral. Both eyes are not affected equally at the same time. Careful measurements of the exophthalmos almost always show that the measurement in the normal-appearing eye exceeds the extreme of the range of normal in unaffected persons. The normal-appearing eye is unlikely to remain that way. This observation is important when a patient with asymmetric Graves' ophthalmopathy is considered for orbital decompression. It makes little practical sense to decompress only the most involved eye in what is a bilateral process. Of the 450 patients previously mentioned who had decompression, only two underwent unilateral decompression, and in both cases the decision was a poor one.

Orbital Decompression

The concept of orbital decompression is simple. There is too much tissue in a bony space, and either the tissue must be removed or the space made larger. Removal of tissue is not practical, so the space must be enlarged. The space can be enlarged in as many ways as there are bony walls to the orbit. The superior wall is the orbital plate of the frontal bone, and this wall can be approached from the anterior cranial fossa, which is the basis of the transfrontal decompression. The lesser and greater wings of the sphenoid bone and zygomatic bone form the lateral wall. The maxilla is inferior, and the paper plate of the ethmoids and

lacrimal bone is medial.

The lateral approach was the first one used for orbital decompression. In 1910 Dollinger removed the lateral wall, leaving the lateral rim intact. This is the same procedure that Krönlein used to remove orbital tumors, and the approach has his name. The lateral approach has been favored by ophthalmologists. Not much has been published regarding this approach, reflecting its infrequent use. Long and Ellis reported 67 cases in 1966 collected during a 16-year period, and Kennerdell and Maroon used this approach to gain access to all four walls. Starting in 1969, our group tested this approach on patients with measurable field defects and visual loss, but we quickly abandoned it because of its lack or predictability.

The superior approach is through the anterior cranial cavity and carries the name of Naffziger, who described and popularized the operation. Another superior approach is the pterion, described by Welti and Offret, as cited by Hamby. At first glance a craniotomy to decompress the orbit may appear to be excessive until one appreciates the natural progression of serious ophthalmopathy and the limited options. Several series of transfrontal orbital decompression have had significant number of cases. In 1954 Naffziger reviewed his overall experience in 40 cases, Poppen reported on 66 patients in 1950, and MacCarty and colleagues collected a series of 46 patients in 1970.

The results of this operation are generally good. Visual loss is stabilized or vision is improved, and appearance is satisfactory. There is usually pulsation of the eye from transmitted dural pulsations. Published reports have commented about the morbidity, but postoperative death has not occurred. Because of the perceived magnitude of a frontal craniotomy, the use of this operation has been restricted to the most extreme manifestations of Graves' ophthalmopathy. It is not offered to patients with lesser signs and symptoms nor to those who need only cosmetic decompression.

The pterion is the point where the sphenoid, frontal, and parietal bones meet in the infratemporal fossa. This bone can be exposed through an incision anterior to the ear in a hair-bearing area, and the fibers of the temporalis muscle can be split to approach the anterior and middle cranial fossa to remove the superior and lateral walls of the orbit. Both sides can be treated at the same session, although the procedure is usually carried out on one side only. In 1972 Moran and colleagues reported their experience with more than 100 cases.

Currently, the inferior approach is the one most often used and may be made either transantrally or transorbitally. Removing bone inferiorly and medially is logical in that access to the greatest area of bone can be achieved through a single incision, both sides can be done at one session, and the morbidity is that of sinus surgery. The transantral approach enlarges the orbital space to use the actual space of the antrum and ethmoid sinuses, whereas the superior, lateral, and pterional approaches decompress into potential spaces only.

The first report of the use of this concept was by Hirsch and Urbanek in the German literature in 1930. They removed the roof of the antrum on one side, noted a 3-mm recession of the eye, and considered the procedure successful. In 1936 Sewall described orbital decompression through an external ethmoidectomy, but he based his report on laboratory work and never actually performed an orbital decompression. Kistner was probably the first to carry out a transantral-ethmoidal decompression using Sewall's method. Hirsch and Urbanek

carefully reviewed the orbital decompression techniques and recommended the transantral approach, but Walsh and Ogura published the first of what was to be a series of reports in which the medial and inferior walls of the orbit were removed. From 1946 to 1980, they operated on 252 patients. Other series have been reported by Stell (six cases), Golding-Wood (ten cases), Sessions and colleagues (nine cases), Calcaterra and Thompson (104 cases), Baylis and colleagues (24 cases), and DeSanto (200 cases). Our personal experience now involves 440 cases, and from this experience the concept of total rehabilitation of Graves' ophthalmopathy has evolved and addresses in what seems to be a logical order, when required, all the manifestations of Graves' ophthalmopathy.

Ocular protrusion is only one manifestation of Graves' ophthalmopathy. Enlarging orbit corrects and may eliminate the proptosis. In the process, most patients have improved vision and a reduction or elimination of visual field defects. The degree of myopathy is unknown before decompression, and remains so afterward. Some, all, or none of the affected muscles may return to their predisease state. What happens to the muscles depends on the duration and degree of the myopathy, and all muscles do not recover, but those that do recover do so to different degrees. Thus, muscle imbalance and diplopia may remain after the eye returns to a more normal position, and muscle surgery may be needed to restore balanced vision. Occasionally, in patients with severe visual loss, diplopia occurs when it did not exist before. As vision returns, it is double; this is considered a positive benefit compared with no vision before operation, but for total rehabilitation, muscle balance should be restored.

Retraction of the upper or lower lid or of both lids is almost always a problem in Graves' ophthalmopathy. After decompression, retraction remains and the cosmetic appearance is not satisfactory in all patients. Lid retraction may be corrected by appropriate lowering of the upper lid and raising of the lower lid. After the muscles and lids are restored, unsightly lid skin may remain. Upper and lower lid blepharoplasty is rarely needed.

The logical sequence for total rehabilitation thus seems to be (1) decompression, (2) extraocular muscle surgery, (3) lid surgery, and (4) lid skin surgery. In our total experience involving more than 400 patients, about 50 orbital decompressions required muscle surgery, 25 needed orbital decompression lid surgery, and fewer than five required orbital decompression lid skin surgery.

Results

On Vision. A detailed analysis has been made of the results of transantral orbital decompression in the first 200 patients. In 87 (143 eyes with visual loss), visual loss was the main indication. In each patient, the vision was threatened either by optic neuropathy and the associated visual field defect or by serious corneal ulceration (corneal ulceration alone is very rare; most patients had neuropathy). A few patients had chronic papilledema. Follow-up measurements of visual fields and acuity were obtained in 84 of the 87 patients between 6 months and 3 years afterward. Of the 84 patients, 79 had improved visual acuity and elimination of the field defect. One patient who showed no improvement had a deep corneal ulcer that healed, leaving an opaque scar. Another patient had chronic papilledema and finger-counting vision in both eyes, with improvement in one. Three eyes with dense field defects did not improve. Four of the five patients in whom orbital decompression failed showed some initial improvement in both acuity and field defects but later had regression and a return of

a dense field; steroid therapy was resumed. All patients but the five with previously cited failures noted improvement of vision at the first follow-up. Improvement usually occurs within the week, although some patients had no visual improvement during the first days. In these patients with field defects, improvement occurs slowly over months. Six patients had continued improvement over 1 year. The fact that vision returns so quickly suggests that the visual loss is a vascular problem, either arterial ischemia or impaired venous return. Vision return per se is not proof that orbital decompression caused the improvement; it is well known that Graves' ophthalmopathy often stabilizes and improves spontaneously. Nevertheless, the improvement in 79 of 87 patients, almost all of those who had been treated with steroids before decompression and 12 of those who had radiation therapy, was impressive. Several patients had previous lateral decompression that failed.

On Ocular Protrusion. The changes in ocular protrusion, as measured by Krahn or Hertel exophthalmometers, were determined by subtracting the last measurement from the immediate preoperative measurement. On 334 eyes, at least two measurements were available. A mean recession of 5.5 mm was observed. The range was 0 to 13 mm. Between the first and second follow-up, 28 patients had an increase in protrusion in one eye. In other words, the process continues, and eyes that retract do not always stay retracted. There was slight recession in 273 eyes when more than one postoperative measurement was made, but this recession was seldom more than the test-retest error of the measuring instruments.

Generally, most of the changes were noted during the first month after decompression. The exophthalmometer readings at 4 to 6 months are usually maintained.

Complications. Because of the different indications for and goals of the operation, it is difficult to establish consistent criteria for evaluating complications of surgery such as orbital decompression. For example, all patients have a temporary sensory disturbance of the upper lid from the sublabial incision and retraction, and this may be considered a complication if the operation is performed for cosmetic reasons. The same residual in a patient who has severe visual loss, painful proptosis, congested orbits, and a field defect or chronic papilledema may be judged differently. Likewise, diplopia that evolves in a legally blind person may be considered a positive benefit rather than a complication in that diplopia is preferable to no vision. Another "complication" is an oroantral fistula after Caldwell-Luc antrostomy. This complication in a person with steroid-induced diabetes who presents for decompression with failing vision in spite of 120 mg of prednisone daily for months or years is probably inevitable.

Complications in our first 200 patients studied are listed in Table 1. Since that study was completed, there have been two deaths. One elderly patient died of meningitis within 6 weeks of surgery. She had been taking 160 mg of prednisone for 9 months and had osteoporosis and diabetes; her surgical wounds did not heal. Another patient died during the immediate postoperative period. The surgical morbidity, as measured by hospital time ranged from 1 day to 13 days (mean, 3.5 days). The day of surgery was counted as the first day.

Table 1. Complications in 200 Patients Undergoing Orbital Decompression

Complication	No. of Patients
Cerebrospinal rhinorrhea	4
Nasolacrimal duct obstruction	9
Nasolacrimal duct obstruction requiring surgery	5
Oroantral fistula requiring surgery	7
Blindness (one eye)	2
Steroid use postoperatively	3
Diplopia	159
Numb lip (permanent)	8
Numb lip (temporary)	200.

Treatment Planning

The technical aspects of orbital transantral decompression are straightforward and well within the capability of the otolaryngologist who is comfortable with antral and ethmoid surgery. Some technical details seem to make a difference between a successful operation and a disappointing one. If the operation was all that was required of the otolaryngologist, the role would be simple. The surgeon operates, supervises the convalescent period, and steps aside. Patients and professional satisfaction require more than this.

Patients with Graves' ophthalmopathy who are candidates for orbital decompression need considerable counseling. They must be provided with the insight into the rationale, risks, and limitations of the procedure in order to obtain a realistic feeling for the probability of benefit from decompression for a specific problem. Patients also need to know that the operation does not solve the basic problem, that simply enlarging the orbital space does not provide total rehabilitation, and that other surgery for muscle imbalance and lid retraction or redundant skin may be needed. If patients' expectations for improvement exceed the potential of the operation, that fact should be established preoperatively.

The surgeon's goals and those of the patients must be compatible. For example, a young woman with disfiguring proptosis, as well as vision-threatening effects of ophthalmopathy, must be aware that the purpose of the operation is vision preservation and not cosmetic, and that after a successful decompression her appearance may be less than perfect and may never be what it was before the disease began. When the patient has different ideas of what constitutes success, both the surgeon and the patient will be disappointed.

When otolaryngologists operate for an eye disease, the patient needs to know that they are only serving a technical role for which he or she is qualified. Otolaryngologists must realize that they are functioning as consultants to the ophthalmologist and endocrinologist. Otolaryngologists who become involved with the many problems of Graves' ophthalmopathy will find themselves with problems that they are not prepared to manage: specifically, extraocular muscle problems. It is far better that a team approach be used: the ophthalmologic experts will manage the patient after the orbit is enlarged and prescribe whatever further rehabilitative efforts are needed. The idea of the team concept needs to be reinforced in the preliminary discussion.

The patient needs to know that other options are available for the problem, to understand why the decompression option is recommended, and to be aware that decompression is not always successful. The more that has been done to a patient surgically or with radiation before decompression, the less predictable the procedure becomes.

Preoperative Evaluation

The preoperative evaluation involves endocrine and ophthalmologic studies. If time permits (ie, if the eye problem is not an emergency), patients with hyperthyroidism should be rendered euthyroid. If urgent decompression is required, hyperthyroidism can be controlled by propranolol.

Eye studies should include both corrected and uncorrected acuity measurements, exophthalmometer readings, assessment of extraocular and levator muscle functions, palpebral fissure measurements, measurement of ocular tension, and a complete fundoscopic examination. Patients with visual loss that is not correctable with lenses require visual field assessment. The field measurements document the seriousness of the eye problem and provide an objective baseline for determining the effectiveness of the operation.

Roentgenograms of the sinuses are not needed if there is no history of sinus disease. Sinus roentgenograms are not helpful, even when there is a history of sinus disease, because the actual state of the cavities is determined at surgery. Membrane thickening and polypoid disease are not contraindication for decompression, because these can be treated at the time of decompression. CT is useful only when the diagnosis is not certain; then the typical muscle changes are observed, and the diagnosis is confirmed. Another exception when CT is helpful is the patient with asymmetric exophthalmos who has no history of hyperthyroidism and in whom the proptosis measurement of the least involved eye is within the normal range.

Patients who are taking corticosteroids systemically or who have taken these drugs during the 9 months before surgery require special precautions during and after the operation. Our policy is to prepare these patients by either giving an intramuscular injection of 200 mg of cortisone acetate the evening before surgery and the morning of surgery, or giving at least 40 mg of prednisolone during surgery and tapering the oral corticosteroid doses that are related to the patients' previous intake during the postoperative period.

Complete decompression is possible only if a complete ethmoidectomy is accomplished. This point cannot be overemphasized. The inability to do a complete ethmoidectomy by the conjunctival route is a concern that we have regarding an attempt to do a transantral-ethmoidal orbital decompression through approaches that are more familiar to the ophthalmologic surgeon. There is a belief that the same removal of bone can be accomplished this way as through the sinuses. This is not true. In patients who have undergone redecompression after anterior orbital approaches, it is obvious that there is a timid attitude toward or an inability to perform a proper posterior ethmoidectomy. Perhaps this area cannot be reached or the surgeon's inexperience in this area leads to a timid approach. Unfortunately, a few operations were repeated for nothing but little blow-out fractures of the medial orbital floor. This is not an orbital decompression. It is our opinion that the incidence of inadequate decompression is higher with the anterior approach than with the transantral approach. This difference has tempered our enthusiasm for the anterior approach, and we

select the transantral operation for patients who need orbital decompression.

Surgical Technique

The patient is placed supine on the operating table in about 15 degrees of reverse Trendelenburg position (ie, sitting slightly up). This position allows more comfort for the surgeon and assistant in relation to the operating microscope, which is used in all cases (300-mm lens).

The microscope provides two benefits: (1) it permits the assistant to observe directly through a side piece or television monitor and (2) the brilliant light and magnification make the operation easier and safer.

The eyelids are stitched together and the eye is left undraped. A standard sublabial incision for anterior antrostomy exposes the maxillary antrum. Bone is removed anteriorly up to the infraorbital rim and neurovascular bundle. The microscope is introduced and the ethmoids are entered with a punch instrument. The precise site of entry is half the distance from front to back at the junction of the superior and medial antral wall. The angle of entry is about 45 degrees upward, directed to the medial canthus of the opposite eye. The ethmoid cells are then gently crushed and removed up to the cribriform plate and back to the sphenoid sinus, using the ethmoid exenteration instrument. The angle between the cribriform area and the roof of the ethmoid capsule from front to back can be estimated, because if the head is slightly up, the posterior cribriform is lower than the anterior cribriform. The bone of the cribriform plate is easily visualized and can be protected by working away from it with the instruments, rather than toward it. The decompression is not adequate unless the level bone of this structure is visualized. The most anterior ethmoid cells are not directly visualized, and these cells must be removed to complete the ethmoidectomy. This is the most often neglected site of the ethmoidectomy.

An adequate complete ethmoidectomy can be performed when the ethmoid capsule will easily accommodate three 0.75-inch transantral Cottonoid strips soaked in epinephrine solution. If the three strips do not fit the ethmoid capsule easily, the ethmoidectomy is not complete. This is an important clue to technique.

If there is bleeding from the anterior or posterior ethmoid arteries, the arteries should be packed temporarily with a Cottonoid pledget soaked in epinephrine solution, and the surgeon should proceed with the operation. The bleeding usually stops; if not, gentle cautery may be applied.

Occasionally, cerebrospinal fluid is seen. This is no cause of alarm. A patch of antral mucosa can be laid over the site of the fluid and held in place with oxidized cellulose (Oxycel) to stop the leak. To our knowledge, only one in more than 400 patients required repair of a CSF leak months after decompression.

After the ethmoidectomy is completed, the bone of the orbital floor is fractured away with a small curette or chisel. The bone medial to the course of the infraorbital nerve is usually very thin and can be pulled away without difficulty. The bone around the nerve and lateral to it should also be removed. The bone lateral to the nerve is much thicker, and a

sphenoid punch type of instrument is useful. Likewise, the thicker bone just behind the infraorbital rim is easier to remove with the sphenoid punch (the rim is not removed). After the bony floor is removed, the paper plate of the ethmoid can be fractured into the ethmoid capsule, after the pledgets are removed. After the pieces of bone are rinsed out, the orbital fascia is split. Splitting is done by placing a long-handled knife as far forward and as far up as possible in the ethmoid, and incising the periorbita as far back as possible. Succeeding cuts are made more laterally. By working from ethmoid medially to antrum, one can maintain direct vision; going in the other direction puts orbital fat in the way. Usually, the fascia is excised in two places medial to the infraorbital nerve and with one slit lateral to the nerve. At times, the nerve is far lateral and all the slits are made medial to it. If extreme proptosis is an issue, the orbital fat can be teased out of the orbit to increase the decompression. This can be done by looking at the patient's eye position while teasing out the fat. If a lesser amount of exophthalmos is present, this is not done. In other words, the operation is tailored to the patient's needs.

One of the major technical difficulties involves attaining the proper exposure of the orbital floor and ethmoid by retraction of the sublabial incision. We have tried a number of the available self-retaining retractors and have designed a few ourselves. None works well for us, and the best that we can recommend is a hand-held Army-Navy retractor and a steady-handed assistant.

The same procedure is carried out at the same session on the patient's other eye. Unilateral decompression should not be performed.

If decompression is not immediate, something is wrong. In most instances, orbital fat fills the ethmoid completely and the antrum in the upper half. Occasionally, very large muscles and less fat are seen; in these patients, decompression is not easily accomplished.

Conclusions

Until the biochemical process that produces the orbital changes in Graves' ophthalmopathy is better understood and can be prevented or reversed, orbital decompression will be needed in some patients. Enlargement of the orbital space does not in itself cure the orbital process but rather allows more space for the process to work itself out. The operations to enlarge the space are only palliative or rehabilitative. Selection of the appropriate patients is essential.

The predictability of recession of the eyes after decompression depends on the patient's problem. The result of the operation is most predictable in patients with congestive signs and visual loss of recent onset. The more treatment that has preceded an unsuccessful decompression, the less predictable the reoperation becomes. Muscle surgery before decompression and orbital radiation are the two forms of preliminary treatment that limit, or at least create uncertainty regarding, the predictable value of decompression.

Because of the short hospitalization period, few complications, and high percentage of satisfied patients when selection is careful, the transantral orbital decompression appears to be a conservative form of treatment and not a treatment of last resort. For these specific problems, decompression seems more conservative than long-term, high-dose corticosteroid

therapy. Currently, we are less reluctant to decompress orbits early during the course of the disease process than when our experience began in 1969.

Assessment of the effectiveness of transantral decompression becomes more subjective as indications expand to include cosmetic goals, and when the decompressions are done as a preliminary to muscle surgery. In these circumstances, the decompression is only the first part of a more complex rehabilitative sequence that includes extraocular muscle surgery, lid surgery, and occasionally facial skin surgery.

The anterior approach to the orbit, which is more familiar to the ophthalmologic surgeon, can remove some of the bone through the transantral route, but there is obvious hesitation with respect to the ethmoid cells when they are approached through eyelid incisions. A complete ethmoidectomy is the key to successful orbital decompression.

Conditions That Mimic Graves' Disease

In our experience, four patients were treated who, in retrospect, did not have Graves' ophthalmopathy. These individuals were not helped by the operation. Each patient underwent decompression during an earlier period, when CT was not available.

Case 1 (Radiation Retinopathy)

A 55-year-old man was seen in 1975. He had been treated for classic clinical hyperthyroidism. In 1971, his eyes became prominent and his vision decreased. Steroid therapy of unknown dosage failed to alter progressive visual deterioration. Cobalt irradiation was administered in 16 fractions to a dose of 2800 rad (28 Gy), and vision returned. Because the proptosis progressed, steroid therapy was restarted 1 month after the radiation. Diplopia followed. In March 1974 visual loss again began. Fundoscopic examination showed atrophic choroidal retinal areas and papilledema. A second course of cobalt was given to a dose of 1000 rad (10 Gy), and there was some improvement in vision. In February 1975 a third course of cobalt in an unknown dosage was given, without benefit. When the patient was first seen at the Mayo Clinic, his visual acuity was 20/200 right and 20/30 left. A visual field defect was plotted in both eyes. Both optic discs were elevated, and retinal hemorrhage, neovascularization, and exudative retinopathy were observed. Atheromatous material was seen in the arterioles. Transantral decompression was performed in August 1975. There was little change in vision. The eyes were recessed by 3 mm. By May 1977 vision was 20/200 in the left eye and finger-counting only in the right eye.

In this patient the process started as Graves' ophthalmopathy, but the treatment with radiation led to the retinopathy. The orbital fat had the same appearance as the orbital contents of other nonradiated patients. This patient was one of 16 patients treated with orbital decompression after radiation, and was the only patient with retinal signs characteristic not of Graves' ophthalmopathy but of radiation retinopathy. The indications for orbital decompression in the other patients varied, and the results were generally no different from those that may be anticipated in nonradiated patients. Our attitude toward orbital radiation has been consistent over the years. We believe that (1) radiation is excessive treatment and unpredictable and (2) just as with long-term corticosteroid therapy, orbital decompression is more predictable than orbital radiation and is a more conservative approach to Graves'

ophthalmopathy in properly selected patients.

Case 2 (Wegener's Granulomatosis)

This patient, a 52-year-old man, had clinical findings consistent with Graves' disease, but the process evolved into classic Wegener's granulomatosis, the first manifestation being in the orbit. If CT had been available, decompression might have been avoided.

In June 1974 he experienced progressive loss of vision and diplopia. Visual examination showed 2 diopters of papilledema on the left, a relative enlargement of the blind spot on visual field study, and a central scotoma on the left. Results of examination of the right retina were normal. There was no history of hyperthyroidism or thyroid disease. Prednisone (60 to 80 mg per day) did not help. Decompression in June 1974 revealed massively enlarged extraocular muscles and little orbital fat. The eyes did not recede. Vision improved, and the field defect cleared. By August 1974 severe periorbital edema, chemosis, and ocular congestion returned and double vision was troublesome. Orbital radiation of 2000 rad (20 Gy) in 20 fractions and steroids were given.

In November 1974 the congestive symptoms were worse, although vision was maintained and the visual fields were normal. Because of the possibility that another orbital process existed, the orbital fat was biopsied. Biopsy revealed acute and chronic inflammation with acute necrotizing vasculitis and focal deposits of fibrinoid material. The diagnosis of orbital pseudotumor was considered. Shortly after this, crusting and bleeding from the nose began and fluid was observed in the ears. Nasal examination showed extensive crusting and a septal perforation. The anterior ends of the turbinates were gone. Multiple biopsies of the nasal mucosa showed subacute and chronic changes, ulceration, plasmacytosis, and necrotizing vasculitis consistent with Wegener's granulomatosis. The sedimentation rate was 98 mm in 1 hour. Prednisone therapy, 60 mg every other day, was started.

By January 1975 proteinuria was present and the creatinine level was elevated. Prednisone therapy was continued and the symptoms gradually improved. In October 1976 papilledema of 2 diopters was noted in the right eye. Cyclophosphamide, 100 mg per day, was added to the program. Within 3 months all systems were stable. Prednisone was stopped in October 1977 and cyclophosphamide in August 1978. At all subsequent visits the patient was well. His last visual field examination was normal and vision was corrected to 20/20 in both eyes.

Case 3 (Chronic Sinusitis)

The third patient had clinical symptoms of chronic sinusitis and presented with proptosis and orbital congestion suggestive of Graves' disease. This was the only patient seen with this presentation. Although roentgen examination is not a routine part of the evaluation, appropriate roentgenograms are needed if the situation is atypical or symptoms are suggestive of sinusitis.

Case 4 (Orbital Lymphoma)

One patient had orbital lymphoma diagnosed at the time of decompression for what was considered to be Graves' ophthalmopathy. This patient had proptosis, decreased vision, and bilateral visual field defects, consistent with optic neuropathy. At decompression a bilateral orbital process consistent with orbital lymphoma was diagnosed, and the decompression was terminated.