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

Section 2: Disciplines Closely Associated With Otolaryngology

Chapter 24: Ophthalmology Relevant to the Practice of Otolaryngology

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In the crush of information constantly being developed about each medical and surgical subspecialty, practitioners within a given subspecialty sometimes forget that the object of their devotion does not exist as an island unto itself. The fields of otolaryngology and ophthalmology are two such that richly interrelate, owing to the physical proximity of the structures involved (for example, the paranasal sinuses are also paraorbital). There is also a similarity in philosophical approach, in that both fields deliver medical and surgical care to their patients. This changes the intimacy of involvement with the patient.

Nonetheless, it is all too common that circumstances that should stimulate interactive concern between the two fields in a particular patient are resolved without benefit of such interaction. This can occur from the constant pressure of time as well as incomplete knowledge regarding when interaction should be sought. This chapter offers an opportunity to enhance the relationship between the two fields for the common good of patients.

Our goals, therefore, are to provide necessary information on topics in which there is a substantial degree of interrelationship between ophthalmology and otolaryngology, in order to foster a climate in which practitioners from both fields will know when to involve the other. The future of medicine must include such a spirit of cooperation if it is to survive less tarnished in a world that increasingly wishes to treat it as just another business.

Anatomy

Many outstanding texts describe the clinical aspects of the anatomy of this region. Our purpose here is limited to describing the features we feel are of most importance to the otolaryngologist: those that further diagnostic and surgical acumen. Most of this chapter deals with the orbit and lids, with only a brief description of the globe itself.

Orbital Anatomy

The first rudiments of the bones that make up the orbit are assembled within the first 2 months of embryogenesis. Ossification of these is not completed until the sixth to seventh months, allowing for substantial plasticity in the face of globe or other malformation. There are seven orbital bones: the sphenoid, lacrimal, palatal, maxillary, ethmoid, frontal, and zygomatic bones.

The configuration of the orbital cavity has been likened to that of a pear, with the floor, roof, and lateral and medial walls tapering from the wide opening to the orbital apex. It should be noted, however, that the orbital floor does not extend to attain the apex. The medial walls come to lie almost parallel to one another, while the lateral walls diverge from

one another by about 90 degrees.

The volume of the orbit is approximately 30 mL, within which lie the globe (approximately 7 mL); the optic nerve; cranial nerves III, IV, V1, and VI; the extraocular muscles (EOMs); the autonomic nerves and the ciliary ganglion; the blood vessels; the lacrimal gland and sac; and the orbital fat. The arterial system originates from the internal carotid's ophthalmic branch. The valveless orbital veins condense to two pathways: the superior ophthalmic vein (the major effluent) and the inferior ophthalmic vein. The former drains to the cavernous sinus, the latter to the pterygoid plexus. The lack of valves allows for the retrograde passage of bacteria from an even initially trivial infection, which can culminate in the catastrophe of cavernous sinus thrombosis if unimpeded by therapeutic measures.

The orbital fat is separated into lobules by fibrous septa; some of these interdigitate into the epimysium of the EOMs and explain the degree of limitation of ocular motility sometimes observed after herniation of fat in blow-out fractures, even if muscles themselves are not entrapped. The fat is richly invested with blood vessels with a marked propensity to bleed if the unwary surgeon proceeds without caution. Such bleeding would cause extensive scarring and limitation of motility, and should thus be avoided.

The deep orbit itself is possessed of neither a standing population of lymphocytes nor lymphatic channels. Thus, lymphoid inflammations or neoplasms must invade the orbital fat via extravasation from the blood vessels. There is a resident lymphoid tissue within both the substantia propria of the conjunctiva and the lacrimal gland, which drain into preauricular, postauricular, submandibular, and cervical nodes. These should all be routinely palpated as part of an orbital assessment.

Let us consider some specifics of the osteology of the orbit. The orbital rim extends approximately 40 mm horizontally and 35 mm vertically. The superior aspect of the rim is formed by the frontal bone, with the supraorbital notch to be found in its nasal third. The notch is completely enclosed by bone in 25 per cent of people, and is then termed a foramen. It transmits the supraorbital nerve (V1 branch of frontal nerve) and vessels, which supply the medial aspects of the eyebrow and upper lid. Posterior to this area lie the frontal sinuses.

The medial aspect of the rim is made up of the maxillary process of the frontal bone, the lacrimal bone, and the frontal process of the maxilla. There is a discontinuity of the inferonasal aspect of the medial rim, which allows for the formation of the fossa for the lacrimal sac. The lacrimal sac fossa is bounded anteriorly by the anterior lacrimal crest, continuous with the inferior rim, and posteriorly by the posterior lacrimal crest, continuous with the medial rim. Parallel to the anterior lacrimal crest is a groove containing a branch of the infraorbital artery, important surgically. The cribriform plate lies just 12 mm above the lacrimal sac fossa and can be inadvertently fractured during conjunctivo-dacryocystorhinostomy, with the risk of meningitis. It should be noted that the lacrimal bone is incompletely ossified even in adults, and hence is readily traversed for nasolacrimal surgery. The nasolacrimal canal transmits the nasolacrimal duct down through the maxillary bone to exit beneath the inferior nasal turbinate. Rarely, it may terminate in the maxillary sinus.

The inferior rim is formed from the maxilla and zygoma. The suture between these two lies about halfway along the length of the rim. Ten mm below this suture lies the infraorbital foramen, transmitting the infraorbital nerve (V2) and artery (branch of the internal maxillary artery). The presence of the infraorbital nerve in the floor of the orbit explains the hypesthesia frequently noted consequent to fracture therein.

Finally, the lateral rim is composed of the zygomatic process of the frontal bone and the frontal process of the zygomatic bone. The zygomaticofrontal suture occurs at the superolateral rim. The floor of the anterior cranial fossa lies 5 to 10 mm above and posteromedial to the zygomaticofacial suture; 10 mm below this suture lies the lateral orbital tubercle, to which is attached the lateral canthal tendon, the lateral horn of the levator aponeurosis, and other suspensory structures of the lid. Failure to attend to these attachments during surgery in the region will have disastrous consequences for both cosmesis and function.

Trauma to the rim most frequently occurs at either the inferior rim or the zygoma. In the former case, the force of the blow is usually transmitted to the weak floor or lamina papyracea, with a resultant orbital blow-out. The zygoma is the strongest bone of the orbit, and hence fracture usually occurs only at its sutures. Such fractures are considered extensively later in this chapter.

The orbital walls are important not only for their structural guidelines but also for the vessels and nerves transmitted through them.

The orbital floor is formed by the orbital plate of the frontal bone and the lesser wing of the sphenoid posteromedially. The frontal sinus lies above the anterior orbital roof. In the superonasal area of the roof, 5 mm posterior from the rim, lies the trochlea for the tendon of the superior oblique muscle. This area is another to respect during surgery, in order to avoid vertical diplopia. Interestingly, if it should be detached, the trochlea may reanneal to its former site of attachment without other manipulation. The lacrimal gland fossa occurs in the superolateral roof anteriorly. The gland has both a deep orbital and a superficial palpebral lobe, separated by the lateral horn of the levator aponeurosis.

The medial orbital wall consists of the frontal process of the maxilla, the lacrimal bone, the lamina papyracea of the ethmoid, and the body of the sphenoid bone. Most of the extent of the wall is the extremely delicate and friable lamina papyracea. Within it lie the anterior and posterior ethmoidal foramina and their constituent vessels, as well as the ethmoid sinuses. The anterior ethmoidal foramen is located about 25 mm posterior to the rim, transmitting the anterior ethmoidal nerve and artery. This foramen must be respected during medial wall decompressions, otherwise severe hemorrhage may result. The posterior ethmoidal foramen is 12 mm further posterior, transmitting the posterior ethmoidal artery and the sphenoethmoidal nerve of Luschka. It usually is not interfered with during wall decompression procedures, being so posterior. However, if decompression of the posterior region is required, a direct approach via a skin incision may be preferable to an indirect approach via the maxillary sinus. This entire region constitutes that at greatest risk for orbital cellulitis, from spread of paranasal sinus infection or mucocele.

One person's floor is another person's roof; the orbital floor is the roof of the maxillary sinus. The major constituent of the orbital floor is the orbital plate of the maxilla, with additional contribution from the zygomatic and palatine bones. We have already noted the presence of the infraorbital canal in the floor, and it serves as the thinnest and hence weakest constituent. Surgery in this area should be conducted with regard for the origin of the inferior oblique muscle, which arises anteromedially just behind the orbital rim. The muscle then passes laterally and posteriorly, to insert upon the posterior pole of the globe. Disruption of this muscle will cause vertical diplopia, and it is the muscle at highest risk for entrapment in the case of blow-out fracture.

The lateral orbital wall is composed of the zygoma and the greater wing of the sphenoid. It is considered to be the strongest of the orbital walls owing to the thickness of the zygoma, but at its articulations the zygoma is susceptible to trauma. The posterior boundaries of the lateral wall may be thought of as the superior and inferior orbital fissures. It is surprising to many students of anatomy that the former is approximately 10 mm long while the latter is twice that length.

The gap between the lesser and greater wings of the sphenoid bone forms the superior orbital fissure (SOF). It transmits, proceeding medially to laterally, the inferior division of III (supplies the medial rectus, inferior rectus, and inferior oblique), nasociliary nerve (V1), VI (lateral rectus), superior division of III (superior rectus and levator palpebrae superioris), IV (superior oblique), frontal nerve, and lacrimal nerve (both V1)). The most apical portion of the SOF is enclosed by the annulus of Zinn, a condensation of tissue from whence the rectus muscles arise. This opening, called the oculomotor foramen, transmits nerves II, III, and VI and the nasociliary nerve. Note that these structures are thus ensconced within the muscle cone, and that cranial nerve IV lies outside the cone. This explains the frequent exclusion of the superior oblique from anesthesia using a standard retrobulbar block, observable as preserved intorsion in the face of otherwise complete akinesia of the globe. The SOF extends posteriorly to the cavernous sinus, and thus the otolaryngologist must recognize the signs of an inflammatory orbital apex syndrome that may lead to cavernous sinus thrombosis. Classically, infection occurring anteriorly, ie, at the junction of the SOF and the anterior cavernous sinus, will involve only V1. Once the cavernous sinus is totally involved, V1-3 will be dysfunctional. Ophthalmologists may use the term "orbital apex syndrome" to imply that the optic nerve is involved in addition to the contents of the superior orbital fissure, but in practice these distinctions are less pure.

The superior greater sphenoidal wing supports the middle cranial fossa and contains the foramen rotundum (V2) and the foramen ovale (V3, accessory meningeal artery, lesser petrosal nerve). The posterior greater sphenoidal wing contains the foramen lacerum (carotid artery) and the pterygoid canal (Vidian nerve, secretory innervation to the lacrimal gland).

Between the superomedial SOF and the optic foramen lies the optic strut, part of the lesser wing of the sphenoid. This thin section of bone is an important radiologic landmark, because erosion of the optic canal (as by optic nerve glioma) will affect it. The optic canal itself transmits the optic nerve, the ophthalmic artery, and some ocular sympathetic nerves. The upper limit of normal for its width in the adult is 6.5 mm; radiologically, asymmetry of the two canals is the most reliable sign of enlargement, especially in infants and children. The optic canal may not be a complete bony canal, so that in a few patients the medial dura may

be in direct continuity with the basal layer of the ethmoid sinus mucosa.

The inferior orbital fissure (IOF) is located at the junction between the lateral wall and the floor of the orbit. It transmits the infraorbital nerve (V2, supplying sensation to the lower lid and adjacent cheek), including zygomatic and alveolar branches; the infraorbital artery; the anterior superior alveolar nerve (supplying incisors and canines); the posterior superior alveolar nerve (cuspids); and the inferior ophthalmic vein. One may think of the infraorbital canal as an anterior extension of IOF, conveying the structures noted above.

Lids and Attendant Structures

We shall try to emphasize that which is necessary for successful cosmetic and functional repair of the lids. We also encourage a team approach in cases of substantial repair of the head and face, whether due to trauma, spread of disease, or correction of senile changes. This ensures maximal patient satisfaction, the ultimate goal in all cases. It is most effective to consider the lids as multilaminar structures, with attention due to each layer.

Skin

It is well known that the skin overlying the eyelids is among the thinnest in the body. This is quite helpful to proper functioning, but renders the area susceptible to the ravages of time and use. It is very well vascularized, which is extremely important in lid surgery, since it allows for ease in grafting, relative resistance to infection, and rapidity of healing.

Vessels and Lymphatics

The arterial supply to the lids comes essentially from two sources: the internal carotid artery, via branches of the ophthalmic artery; and the external carotid artery, via the anastomosing arteries of the face.

An important surgical landmark is the angular artery and its attendant vein. This derives from the external carotid through the facial artery, lying superficially beneath the orbicularis muscle 6 to 8 mm medial to the medial canthus, and 5 mm anterior to the lacrimal sac. It is essential to identify and avoid these vessels during nasolacrimal or ethmoidal sinus procedures. Other lid branches from the external carotid to the lids derive from the superficial temporal artery: the frontal, zygomaticofacial, and transverse facial branches.

The cutaneous terminations of the ophthalmic artery make up the lacrimal, frontal, supratrochlear, and nasal branches. Important in the upper lids is the anastomosis between the marginal and deep arterial arcades. The marginal arcade lies on the anterior tarsal surface, about 2 to 3 mm from the lid margin. This bleeds profusely if injured, spreading unwanted ecchymosis through the lids. This is also of concern in reconstructive procedures that incorporate tarsal-sharing procedures. The arcades arise nasally from terminal nasal branches of the ophthalmic artery, laterally from the lacrimal artery. There are also anastomoses with the angular vessels, as well as with deep orbital contributaries. In the lower lid, there is a similar marginal arcade, connecting with the nasal artery medially and lacrimal and zygomaticofacial arteries laterally. Veins are roughly parallel structures.

Again, let us stress the importance of control of superficial facial sepsis. Perhaps the correct way to think of the face is that there can be no truly "superficial" infection because of the anastomotic connections to deep vessels. The cavernous sinus can be attained through the angular, supraorbital, and superior ophthalmic veins, and the facial vein drains to the pterygoid plexus (which drains through the IOF to the cavernous sinus).

A brief restatement of lymphatic drainage: usually, the medial lids drain to the submandibular nodes, the lateral lids to the preauricular nodes. A wise clinician also palpates the cervical nodes, especially in cases of suspected neoplasia.

Lid Protractors

The orbicularis oculi muscle, innervated by the zygomatic branches of the facial nerve, subserves as protractor for the upper and lower lids. It has three regions based on underlying structures as well as anatomic considerations: pretarsal, preseptal, and orbital. The pretarsal orbicularis is fused to the tarsus; nasally, it arises from the posterior lacrimal crest and the medial canthal tendon; laterally it contributes to the lateral canthal tendon. The preseptal orbicularis arises nasally from around the lacrimal sac, the posterior lacrimal crest, and the medial canthal tendon; laterally, from the lateral palpebral raphe. Normal eyelid closure recruits the pretarsal and preseptal components; only vigorous forced closure involves the orbital portion. It arises from the medial canthal tendon, and laterally from the periosteum.

Orbital Septum

The septum represents the delineation between lids and orbit, and arises from the periosteum of the superior and inferior orbital rims. In the upper lid, it descends to fuse to the levator aponeurosis 3 mm above the superior tarsal border. In the lower lid, it fuses with the capsulopalpebral fascia 3 to 5 mm inferior to the tarsal border. This fused tissue then inserts onto the anterior and posterior aspects, as well as the inferior border, of the tarsus. It can be identified during surgery by tugging on it to feel the firm attachment to the periosteum; it will not move as the patient attempts to raise or lower the lid. Immediately behind the upper lid orbital septum lies another hallowed anatomic landmark: the pre-(levator) aponeurotic fat pad.

Lid Retractors

Upper Lid. Retractors consist of the levator palpebrae superioris, innervated by the oculomotor nerve, and the tarsal muscle of Müller; the latter arises from the undersurface of the former and is sympathetically innervated. It is usually estimated that Müller's muscle adds 2 mm of elevation to the upper lid, leaving most of that effort to the levator. Twelve mm above the superior border of the tarsus, Müller's muscle arises from the undersurface of the aponeurosis. It is strongly adherent to the underlying conjunctiva, but loosely so to the aponeurosis. The sympathetic denervation of Horner's syndrome affects this muscle, usually producing 2 mm of ptosis. It is now recognized that most cases of ptosis are not the result of neuropathic or myopathic disorders, but of a mechanical problem: disinsertion of the levator aponeurosis. Other mechanical problems that cause ptosis are frontal sinus mucoceles with orbital extension and allergic thickening and edema of the conjunctiva near the upper tarsus, as in giant papillary conjunctivitis.

The levator arises at the orbital apex, from the periorbita of the lesser wing of the sphenoid, immediately above the annulus of Zinn. It rests on the superior rectus, with which it shares innervation (see above). Its muscular portion is perhaps 40 mm long, and its aponeurosis 15 to 20 mm. At the level of the superior orbital rim, and at the point of conversion from muscle to aponeurosis, there is a condensation of tissue known as the transverse ligament of Whitnall. It is the suspensory fulcrum of the lid and is crucial to its proper functioning. At Whitnall's ligament, the levator muscle is reoriented from an anteroposterior to a superoinferior course. Its suspensory function for the superior orbit as well as the lacrimal gland was not initially recognized. It inserts medially in the trochlear region, and laterally passes through the lacrimal gland and onto the lateral orbital wall. In the past these insertions were cut during upper lid procedures, especially ptosis repairs, but also in reconstructive efforts. Doing so changes the appearance and function of the upper lid, and also allows for lacrimal gland prolapse temporally; this should usually be avoided.

As the levator aponeurosis descends, it sends out fibers laterally, which contributes to the lateral canthal tendon, and medially, to the posterior medial canthal tendon. Proper repositioning of the anterior and posterior crus of the medial canthus in, for example, trauma cases requires both medial and posterior fixation; otherwise, the canthus is not properly reformed and upper lid function is suboptimal.

At the level where the septum fuses with the orbicularis, about 3 to 4 mm above the superior tarsal border, the aponeurosis splits into anterior and posterior portions. The former finely inserts into the pretarsal orbicularis, causing the upper lid crease. Disinsertion of the aponeurosis destroys the normal position of the lid crease. The aponeurosis then recedes and the superior orbital sulcus has a sunken appearance. This condition is repaired by reinserting the levator aponeurosis on the upper border of the tarsus. In Oriental people, the insertion occurs much lower, allowing orbital fat to descend farther inferiorly, with the resultant typical Oriental appearance; this should be kept in mind during repair of lids in Asian patients. The posterior portion of the levator aponeurosis inserts upon the anterior face of the tarsus.

Lower Lid. In the lower lid, fibrous attachments condense from around the inferior rectus muscle to form the capsulopalpebral head, the analog to the levator and its aponeurosis. This divides to encircle the inferior oblique, and fuses just anterior to it to form Lockwood's suspensory ligament. Five mm below the inferior tarsal border, it fuses with the orbital septum and goes on to insert upon the tarsus. Modern surgical treatment of most cases of entropion and some types of ectropion relies on repositioning the lower lid retractors with regard to the lower lid tarsus.

Tarsus

The tarsi are dense plates of fibrous tissue that provide a kind of skeleton for the lids. In the upper lid, the maximal height is 10 mm centrally, tapering medially and laterally. In the lower lid, the maximal height is 3 to 5 mm centrally, also tapering to either side. The tarsus is approximately 1 mm thick but can attenuate remarkably with advancing age. Ophthalmologic colleagues may ask you to harvest ear or nasal cartilages as a replacement for weak or missing tarsus; periosteum and dura are also suitable but less stiff replacements. Within the substance of the tarsi lie the Meibomian glands, 25 in the upper lid and 20 in the lower lid; these elaborate the oily component of the tear film.

Conjunctiva

The conjunctiva provides the most posterior layer of the lid. On the back surface of the lids, it is termed palpebral; when reflected upon the globe, it is termed bulbar. Between the bulbar and palpebral conjunctiva lie the fornices, which can be regarded as little pouches of potential space, sometimes filling with small air bubbles detected on computed tomographic (CT) scans.

The conjunctiva is a nonkeratinizing, stratified squamous epithelium, essential for elaboration of crucial tear constituents: aqueous from accessory lacrimal glands, and mucus from goblet cells. The third tear layer, lipid, is contributed by the Meibomian glands found within the tarsi.

Superficial Lid Innervation

It is important to know how to anesthetize the lids adequately. Sensation at the medial canthus is provided by the supratrochlear nerve above, and the infratrochlear nerve below. The supraorbital nerve supplies the central upper lid, the infraorbital nerve the central lower lid. The lateral canthus is subserved by the lacrimal nerve above, and the zygomaticofacial nerve below. One should use this information to maximize patient comfort during local procedures; for example, injecting local anesthetics along the upper lid does *not* anesthetize the medial canthal region. Injections 1.5 and 2.5 cm posterior to the medial canthal tendons anesthetize the anterior and posterior ethmoidal nerves. Anesthesia of the anterior ethmoidal nerve permits probing of the lacrimal drainage system. Akinesia of the eyelids can be achieved by local infiltration of the orbicularis oculi muscles, or by injecting the facial nerve at its exit at the facial canal or more peripherally.

Lacrimal Secretory System

The tear film in the normal eye is a trilaminar fluid: a mucin layer coating the corneal epithelium, central aqueous layer, and outermost lipid layer. These components are produced by conjunctival goblet cells, lacrimal glands, and Meibomian glands, respectively. The importance of the tear layer-corneal interface cannot be overstressed; it provides approximately 45 out of the total refracting power of 50 diopters attainable by the eye. For our purposes here, we need to devote some time to the source and control of the production of the aqueous layer of the tear film, for the interfaces with otolaryngology as seen below.

Most experts agree that there are essentially two kinds of tearing: reflex and basal. The latter is mediated by the accessory lacrimal glands of Krause (found in the fornices), and Wolfring (found at the extremes of the upper and lower lid tarsus). Reflex tearing is thought to issue from the lacrimal gland itself, located as previously described in the superolateral corner of the orbit. There are two lobes to the lacrimal gland, divided by the insertion of the levator aponeurosis. The larger, superior orbital lobe rests on the aponeurosis and channels all its output through tubules that must run through the smaller, inferior palpebral lobe. Thus, destruction of the palpebral lobe by inflammation, neoplasia, or surgical carelessness will ablate the secretory function of the gland.

The palpebral lobe lies between the levator and the conjunctiva, with firm attachments to the latter. Nonetheless, with senile involution, the lacrimal gland may prolapse from its normal perch and may be mistaken for prolapsed orbital fat during a blepharoplasty. Attention to the color of the material being excised during such a procedure can save the patient from the disastrous consequences of this action; look for the pinkish-gray color of the lacrimal gland to distinguish it from the yellow or yellow-white of orbital fat. The major ducts of the lacrimal gland empty into the superior fornix 5 mm above the lateral tarsal border. What causes the onset of reflex tearing? The reflex afferent stimulation is mediated through the lacrimal branch of V. However, reflex tearing can occur as a result of stimulation of almost any branch of the trigeminal nerve. The efferent innervation begins in the pons in the lacrimal nucleus, and courses forward to leave the pons as the nervus intermedius at the cerebellopontine angle. Here it lies between the motor root of VII and the acoustic nerve. It then travels through the internal auditory canal and geniculate ganglion, joins the maxillary nerve, and finally joins the lacrimal nerve posterior to the lacrimal gland. Many lesions of otolaryngologic interest can interrupt the efferent innervation, and thus assessment of reflex tearing is a not unimportant part of the examination in an appropriate situation.

A brief mention should be made at this point of tumors of the lacrimal gland. Approximately half of these are lymphoid or inflammatory in nature, and half epithelial; half again of the epithelial tumors are malignant. The most common benign tumor is the benign mixed adenoma; the most common malignant tumor is the adenoid cystic carcinoma. The differences in presentation and pathology are detailed elsewhere.

Lacrimal Excretory System

Once tears have been secreted and have circulated over the globe and fornices, the lacrimal excretory system is called upon to make room for fresh tears. Current concepts initiate this process by postulating an active tear-pump mechanism caused by lid movement.

Both the upper and lower lids possess a puncta, an opening to the excretory system that lies slightly elevated (the lacrimal papillae) and inverted so as to lie against the globe. These are at the medial ends of the lid margins, parallel with the openings of the Meibomian glands. Each is the aperture of the vertical 2-mm canalicular ampulla, which then takes a right-angled bend to become the lacrimal canaliculus. The canaliculus runs medially for about 8 to 10 mm, whereupon, in 90 per cent of individuals, the upper and lower canaliculi join to form the common canaliculus. This enters the lateral wall of the lacrimal sac, approximately 2 to 3 mm below the dome of the sac. The nasolacrimal sac lies in the lacrimal fossa of the medial wall of the orbit, as described above. At this point the valve of Rosenmüller guards against tear reflux into the canalicular system.

The sac lies ensconced within the anterior and posterior crus of the medial canthal tendon, periosteum separating it from the lacrimal bone and frontal process of the maxilla. Thus, the middle meatus of the nasal cavity lies immediately medial to the sac. The nasolacrimal sac has a length of 10 mm and its dome rises above the medial canthal tendon. Interestingly, inflammatory dacryocystitis tends to cause swelling below the tendon, while tumors of the sac usually cause swelling above the tendon, in the dome of the sac.

Dacryocystitis is most common in middle-aged women, usually beginning as infection within the sac, but it occasionally takes the form of spread of infection from contiguous paranasal sinuses. If not adequately controlled, persistent infection can lead to permanent obstruction of the drainage apparatus, or even mucocele. The most common primary tumor of the sac is the squamous papilloma, which, owing too its frequently inverted growth predisposition, tends to recur and may even undergo malignant transformation. Squamous cell carcinoma may occur, as may invasion of the sac by contiguous skin tumors or paranasal sinus squamous carcinoma.

The sac is continuous inferiorly with the nasolacrimal duct, with its initial interosseous section traversing 12 mm laterally and posteriorly, and its membranous portion extending 10 mm to end at the valve of Hasner, beneath and lateral to the inferior turbinate of the nasal cavity. The valve of Hasner is a mucosal fold partly covering the end of the nasolacrimal duct, and it is here that most congenital nasolacrimal obstruction occurs. Two to 4 per cent of newborns have this obstruction, and one-third of these are bilateral. Most perforate spontaneously by 2 months after birth; if not, nasolacrimal probing by age 8 months is successful in opening the obstruction in over 90 per cent of patients. Such probing usually is not performed before the age of 6 months in view of the increased risk of serious complications with anesthesia.

Acquired obstruction of the nasolacrimal duct usually occurs in middle age, is more common in women, and is most often caused by involutional stenosis. Granulomatous entities such as sarcoidosis, Wegener's granulomatosis, or idiopathic midline destructive disease can also wreak havoc upon the excretory apparatus. The surgical procedures for overcoming obstruction of the nasolacrimal system, such as dacryocystorhinostomy, are beyond the scope of this chapter.

The Globe

Although it is difficult to restrain an ophthalmologist when the beauty and grandeur of the eye is considered, we shall try to relate only those points pertinent to the otolaryngologist's practice.

The adult eyeball has an anteroposterior expanse of 23 mm. Its weight approaches 7.5 gm, its volume 6.5 mL, and its circumference 75 mm. The area of widest transverse diameter is referred to as the equator of the globe; the posterior pole is the most posterior portion of the globe, which contains the optic nerve and macula. The sclera comprises the outer fibrous coat of the globe itself, covering 80 per cent of the total surface area. It has two apertures: anteriorly, for the cornea, and posteriorly, to transmit the optic nerve and associated structures. It is thickest (about 1 mm) back by the optic nerve and thinnest just posterior to the insertions of the rectus muscles. This explains the propensity of the globe to rupture at these sites. The rectus muscles extend anteriorly from the annulus of Zinn, to insert 5.5 to 9 mm from the corneoscleral limbus in a stereotypical pattern (the "spiral of Tillaux"). The superior oblique passes inferior to the superior rectus, the inferior oblique inferior to the inferior rectus, to insert upon the globe.

Tenon's capsule is a fibrous membrane that envelopes the globe and is connected to the sclera via fine collagenous trabeculae. On its outer surface, it is attached to the sheaths

of the EOMs, and it sends fibers to the interlobular fat septa and conjunctiva. It keeps the orbital fat away from the mechanics of motility; violation of this structure may cause troublesome adhesions.

The cornea is the most transparent tissue of the eye, with horizontal and vertical dimensions of 12 mm and 11 mm, respectively. It and the precorneal tear film provide the major refracting power of the eye (45 out of 50 diopters). It is covered with a stratifying squamous epithelium and has multiple subepithelial layers. The area where it joins the sclera is called the limbus; this region overlies the iridocorneal angle of the eye. This is made up of the trabecular meshwork, among other structures, which is the major site of drainage of the aqueous humor. Pathologic conditions of iridocorneal angle structures are responsible, in most cases, for glaucoma.

Between the posterior surface of the cornea and the anterior surface of the iris is a space called the anterior chamber, the depth of which is maximally 3 mm at its center. Abnormalities in the depth and clarity of the anterior chamber may accompany trauma. For example, posterior rupture of the globe or dislocation of the lens may cause deepening of the anterior chamber. Normally, it is filled with aqueous humor, and it may be the site for numerous inflammatory processes.

The iris is the most anterior division of the three-membered uvea; the other two structures are the ciliary body and the choroid. The iris has both a dilator muscle, sympathetically innervated, and a constrictor muscle, parasympathetically innervated. It thus can respond to changing levels of illumination and other stimuli.

Behind the iris is the ciliary body, responsible for the elaboration of the aqueous humor and the suspensory ligament of the lens, the zonules. The lens is thereby suspended behind the pupillary aperture, having an equatorial diameter of 9 to 10 mm and an anteroposterior thickness of 4 to 5 mm. In comparison with the cornea, it is the second most transparent ocular structure and the second strongest refracting power (about 12 diopters). The space between the ciliary body and the lens is called the posterior chamber; it is also aqueous filled.

Posterior to the lens is the vitreous cavity, which makes up about two-thirds of the volume of the eye. It is composed mainly of water and collagenous tissue, and exists in the young individual as a gel. As people age, the vitreous undergoes syneresis, a process of liquefaction. Since the collagenous skeleton of the vitreous is firmly attached to the optic disc, fovea, and vitreous base, the collapse of the collagenous skeleton due to liquefaction may allow tractional tearing of the retina and retinal detachment.

One can view the retina as the ocular component around which all the others are designed, for it is the anterior neural tissue of vision. It is a multilaminar structure with a total area of 1250 mm^2. Its thickness varies; it is 100 microm in the periphery and at the macula, 180 microm at the equator, and thickest around the optic nerve, 560 microm. The macula subserves central vision, possessing the greatest density of photoreceptors over a 1.5-mm area about 2.5 disc diameters temporal to the disc. The optic nervehead (disc) is slightly taller than it is wide (1.86 x 1.75 mm); since there are no neural components at the disc, it is totally impercipient, and it thus produces the well-known physiologic blind spot on visual field

testing. A total of 1.3 million axons course through the disc, the retina having already accomplished a considerable amount of processing of visual data. The depression in the center of the optic disc is called the physiologic cup, a remnant of the withering of the embryologic hyaloid vascular system. The normal ratio of the cup:disc diameters is 0.3; this may differ on the basis of congenital development or postnatal pathologic processes. In some patients, deep in the cup an area of yellowish cross striations is barely visible; this represents the lamina cribrosa, which consists of the scleral trabeculations crossing at the posterior scleral foramen. With substantial atrophy of the nervehead, such as seen in glaucoma, the lamina becomes more and more apparent.

The central retinal artery, a branch of the ophthalmic artery, enters the optic nerve about 10 to 15 mm posterior to the globe, and branches extensively to supply the nerve. Then, at the level of the cup, it branches into the nasal and temporal vascular arcades. Veins follow the arterial pattern and join together at or below the cup to form the central retinal vein. The optic nerve is covered with the meninges, and its cerebrospinal fluid (CSF) is continuous with that of the brain.

Beneath the retina lies the retinal pigment epithelium, providing support, a nourishment, and recycling of photoreceptor components, among many other functions. It is a monolayer of cells, with pseudocilia interdigitating with photoreceptor outer segments. Between the two is a mucopolysaccharide matrix and no cell-to-cell bonds; thus, between the retinal pigment epithelium and the retina lies a potential space, achieved in the case of retinal detachment. Supporting the retinal pigment epithelium is the choroid, a fenestrated vascular plexus that nourishes up to and including the outer one-third of the retina; the retinal blood vessels nourish the inner two-thirds. The extreme outer coat of the globe is the sclera, which is continuous anteriorly with the transparent cornea.

Conditions of Otolaryngologic-Ophthalmic Interface

Congenital Conditions

A myriad of congenital conditions, by virtue of the fact that they involve the orbital walls, fall into this interface category. Most are covered in great detail elsewhere in this volume.

Hypertelorism Versus Telecanthus

These are terms frequently confused by otolaryngologists and ophthalmologists alike, despite the fact that the significance of and surgical approaches to the two are quite disparate. It is essential that members of a craniofacial surgical team be able to communicate information to one another precisely and accurately.

Orbital hypertelorism is defined on the basis of bony landmarks, and means that there is an increased distance between the medial orbital walls. Telecanthus, on the other hand, is a measurement based on soft tissue landmarks, and refers to an increased distance between the apices of the medial canthi. At birth the average interorbital distance measured at the orbital equator is 16 mm, increasing gradually with age and finally ceasing such growth in early adulthood. Average measurements for interorbital distance are 25 mm for adult females

and 28 mm for adult males. In making the distinction between hypertelorism and telecanthus, it is helpful to recall that the intercanthal distance is normally approximately one-half of the interpupillary distance (PD, the distance between lines drawn vertically through the centers of the pupils).

The examiner must beware of conditions that convey the impression of hypertelorism, but are not: flattening of the bridge of the nose (very common in newborns and infants), epicanthus, and exotropia. True hypertelorism occurs in a plethora of conditions; some of the more important include severe facial trauma, blepharophimosis, Turner's syndrome, Edwards' syndrome (trisomy 18), Patau's syndrome (trisomy 13), craniostenoses (Apert's, Crouzon's syndromes), Rubenstein-Taybi syndrome, Hurler's syndrome, median facial cleft syndrome, oculopalatodigital syndrome, Waardenburg's syndrome, Treacher Collins syndrome (mandibulofacial dysostosis), cretinism, Marfan's syndrome, Hallermann-Streiff syndrome (oculomandibulodyscephaly), Ehlers-Danlos syndrome. These are covered elsewhere in these volumes.

Craniostenoses

The most common of these disorders are Apert's (acrocephalosyndactyly type I) and Crouzon's (craniofacial dysostosis) syndromes, actually now thought to represent extremes in a spectrum of related abnormalities. Apert's is felt to be less severe expression of the pattern, and Crouzon's, inherited in an autosomal dominant pattern with high penetrance, more severe. In all these entities, there exists a symmetric premature closure (synostosis) of all or some of the sutures in the upper skull accompanied by synostosis or shortening of the skull base. Usually, these skull malformations are accompanied by midfacial bony anomalies such as hypoplastic maxilla, narrow rhinopharynx, cleft palate, dental anomalies, and shallow orbits. Shallow orbits are caused by an anterior displacement (frontalization) of the greater wing of the sphenoid bone and vertical malalignment of the frontal lobe. Interestingly, the primary cause of these syndromes is now believed to be failure in development of the blastemic mesoderm, from which the skull bones develop, which can occur as early as 4 weeks of gestation.

Ophthalmic consultation is mandatory at an early stage in all cases of craniostenosis because of the exceedingly high rate of optic atrophy in these individuals (30 to 80 per cent). This is due to a combination of factors: elevated intracranial pressure, intracranial or intraorbital stretching of the optic nerves, and transverse narrowing of the optic canals or foramina. There may also be genetically determined optic atrophy that does not depend on simple mechanical problems. Exophthalmos is usually present owing to the decrease in anteroposterior diameter of the various orbital walls. Decisions on the best time to operate on these individuals must therefore take into account the status of the eye, lest surgical cosmetic perfection be accompanied only by blindness. If there is substantial compression of the optic nerve, surgery is best performed in concert with an orbitally trained ophthalmic surgeon to safeguard vision. Visual evoked potentials and other techniques may be required to assess the vision of these children, particularly when language skills are impaired by deafness. The problem of evaluating vision in the absence of hearing is similarly complicated in children with osteopetrosis.

Facial Dysplasias

Craniofacial clefting is a very common congenital problem. These entities are very commonly accompanied by severe ophthalmic abnormalities, some obvious, but many not so. Many clefting syndromes are inherited and there is considerable overlap in their features. Using the nomenclature of Tessier, clefts 0 to 4 commonly have anomalies of the nasolacrimal apparatus, 5 to 8 have lower lid deformities, 6 is associated with Treacher Collins syndrome, and 7 is associated with otomandibular dysostosis and Treacher Collins syndrome.

Goldenhar's syndrome, oculoauriculovertebral dysplasia, is noted with cleft 8. This entity may manifest epibulbar dermoids and be associated with the oculomotor abnormalities of Duane's syndrome.

Cleft 10 is usually caused by a frontal encephalocele, which is crucial to recognize so that CT scanning can be used to guide the establishment of surgical planes without endangering the brain. Clefts 10 to 14 are those most commonly associated with hypertelorism and ocular colobomas.

Children with the Pierre Robin syndrome have a 50 per cent chance of also having Stickler's syndrome, hereditary arthro-ophthalmopathy. It is vital to establish this diagnosis, since there is an extremely high rate of high myopia and retinal detachment in this condition. Stickler's syndrome is also noted to cause cataracts and strabismus. It is thought to be a disease of type II collagen.

Selected Soft Tissue Deformities

Colobomas. Colobomas of the upper lid more commonly exist in isolation, but may rarely be associated with extensive facial deformity. It is of paramount importance to ensure adequate coverage of the cornea, otherwise severe corneal exposure may cause abscess, perforation, and ultimately endophthalmitis. This may be accomplished presurgically by the frequent and generous use of ocular lubricants. Ointment provides more resistant coverage, but should not be relied on for long-term use, because occlusion amblyopia is possible in infants and children.

Lower lid colobomas are usually seen as the most superior extension of a lower facial cleft.

Ectopic Brain. Encephaloceles and meningoencephaloceles appear within the orbit but are also noted at the base of the nose, along the cranial sutures, and in the oropharynx. In the orbit, those anteriorly located occur at the junction of the frontal and lacrimal bones, at the cribriform plate, or at the superior maxilla. These may cause pulsating exophthalmos. Posterior encephaloceles herniate through posterior foramina such as the optic canal, superior and inferior orbital fissures, and ethmoidal foramina. Because their repair is usually intracranial, it is imperative to identify these before surgery of the facial area is begun. It is possible to confuse them with dermoid or epidermoid cysts, or with cholesterol granulomas, since these entities can occur in some of the same sites. A direct coronal CT scan allows optimal evaluation of the interface between the cranial cavity, the orbit, and the paranasal

sinuses; axial views can easily miss these relationships. Alternatively, parasagittal magnetic resonance imaging (MRI) may reveal the same features.

Trauma

Orbital Wall Fractures

Orbital floor, and medial wall, fractures are common traumatic entities that clearly interface the line between the otolaryngologist and the ophthalmic surgeon. This is not only because of the region of the possible surgical repair, but because an estimated 25 to 50 per cent of patients with orbital fractures also sustain ocular or intraocular injury. These injuries may include scleral or corneal laceration, dislocation or tear of the lens, retinal detachment, vitreous hemorrhage, hyphema, extraocular muscle laceration, cranial nerve damage, or optic nerve transection, and so many will not be readily apparent on gross examination. Despite other features of the injury, some of these are absolute contraindications to immediate surgical repair of a floor fracture, lest the globe be irretrievably lost. Thus, it is common sense that an assessment by the otolaryngologist of severe damage accompanying floor fracture mandates an accompanying careful ophthalmic examination. A sensible combined surgical approach can then be formulated, with minimal risk to the eye.

Blow-out fractures may be classified as either direct, involving fracture of the inferior orbital rim, or indirect, in the absence of such; the latter is the more common. The classical concept of the genesis of blow-out fractures postulated that a foreign body, of a diameter larger than the orbital rim, struck the orbital contents, greatly raising the intraorbital pressure. This would act to compress the orbital contents toward the orbital apex, with the bones giving away at their weakest points, which are the medial wall lamina papyracea and the posteromedial aspect of the floor. Thus, fractures of the floor may be accompanied by medial wall fractures, which may endanger the medial canthal support elements. Clues to the existence of a medial wall fracture include medial subcutaneous emphysema and epistaxis. More recent hypotheses of blow-out genesis suggest that the foreign object causes a compressive force at the inferior rim itself, which directly leads to buckling of the floor or contiguous medial wall.

Classical clues to the diagnosis of a floor fracture are lid ecchymosis, diplopia and/or EOM limitation, enophthalmos, and decreased sensation in the distribution of the infraorbital nerve (branch of V2). How can one assess the status of the EOMs in the face of such an injury? Radiologic examination is, of course, mandatory. Waters views have been superseded by CT as the best means of assessment, especially if direct coronal views are obtained (barring that, coronal reformatting may be acceptable). One can quite readily observe the inferior herniation of orbital contents into the maxillary antrum, if present. However, this does not distinguish herniated fat and connective tissue from EOM.

If the fracture is laterally displaced from the infraorbital groove and canal, direct entrapment of the inferior oblique, and/or inferior rectus muscles becomes much more likely. Next, one can resort to forced duction testing. If employed, this should be performed correctly: the area between the corneal periphery (limbus) and the insertion of the inferior rectus muscle (6.9 mm from the limbus) should be anesthetized by placement of a cotton pledget, soaked in 4 per cent cocaine, for a few minutes. Next, a toothed forceps should be

applied to the aforementioned area, and the patient instructed to look down. Gentle traction will then establish whether there is restriction or not. However, the mere presence of blood and soft tissue swelling may be enough to produce limitation. This technique also does not establish whether there is diplopia due to cranial nerve trauma *unless* tissue swelling has been minimal. Then, manipulation reveals no restriction in the face of motor dysfunction. In all, one should not rely on forced duction tests as the final word on the question of entrapment, but use them in combination with other testing.

The timing or necessity of surgical intervention in the event of floor fractures remains one of the more controversial topics in both otolaryngology and ophthalmology. Some authors advocate immediate surgery in almost all cases, citing the fact that 40 to 63 per cent of patients with isolated floor fracture present with a restrictive strabismus. Others advocate waiting up to 6 months before performing surgery, maintaining that most patients do well without it. The authors feel that a compromise position is most sensible. Certainly, some factors press for immediate surgery: eg, detection of retrobulbar hemorrhage with impending or advancing optic nerve compromise. Another factor is the need to attend to intraocular conditions, particularly in exploration for scleral rupture. Patients with a fracture destroying more than half of the floor or demonstrating soft tissue prolapse into the antrum are at the highest risk for late vertical diplopia and enophthalmos, and can be most easily operated on within 2 weeks of injury should disabling diplopia or enophthalmos over 2 mm persist. This allows for reduction in swelling, and hence more accurate assessment of motility derangement and greater ease of surgical correction. Residual diplopia only in upgaze occurs in about 20 per cent of patients who have no surgery, but this is problematic to a very few specialized occupations (eg, librarians shelving books) and need not indicate a need for surgery. There is a small (less than 1 per cent) risk of optic nerve damage during the course of surgical repair of floor fractures, and thus surgeons would be well counseled to merely observe small fractures that are causing only minor soft tissue problems.

Although individual surgeons differ in their opinions, we consider that one is well advised to give a course of antibiotics to patients with maxillary antrum involvement. It is theoretically quite possible for infection to ascend to the orbital apex, and hence the cavernous sinus, by this route. These patients should also be cautioned to avoid blowing their noses for several weeks after the injury, to avoid subcutaneous emphysema.

The currently favored approach for many ophthalmologists is the inferior forniceal conjunctival incision, which is thought to be associated with the lowest incidence of postoperative complications. Ectropion following subciliary incision, and impaired lower lid movement after some inferior orbital rim incisions, do occur.

Tripod Fractures

These fractures are frequent consequences of falls, fistfights, or motor vehicle accidents. They are termed "tripod" because they involve fractures of the zygoma near three of its articulations: along the lateral orbital rim (hinged at the zygomaticofrontal suture), the inferior orbital rim (hinged at the zygomaticomaxillary suture), and the zygomatic arch. However, tripod fractures may often be accompanied by other fractures, such as those of the orbital floor, and the clinician must be assiduous in scanning roentgenograms for further damage.

Even without a concomitant floor fracture, but especially with one, the clinical signs of tripod fractures may include all those previously listed for floor fractures. In addition, if there is impingement of the coronoid process of the mandible, the patient will have difficulty and pain with attempted mastication. The appropriate facial landmarks will be flattened, depending on the type of fracture. An extremely important point to recall is that the same types of serious intraocular damage listed under orbital wall fractures above occur with tripod fractures, and the same precautions should be observed.

As described elsewhere in the text, there is a classification scheme for tripod fractures, grouping them on the basis of the site of fracture, the presence of residual attachments ("hinging"), and the degree and direction of displacement. As with orbital wall fractures, most clinicians would agree on initial antibiotic coverage. Surgery should be delayed until adequate assessment of globe status and proper precautionary measures have been accomplished. It then is usually advisable to allow 10 to 14 days to elapse before surgery, to allow for partial resolution of swelling, unless fracture severity and functional compromise demand earlier attention.

Fractures hinged at the zygomaticofrontal suture often result in inferior displacement of the lower lid, which may be obscured by soft tissue swelling. Repair is mandatory to prevent ultimate corneal exposure. Hinging at the zygomaticomaxillary suture will disrupt the lateral canthal angle. It is crucial to ensure that surgery incorporates reestablishment of this structure, to allow proper function of lower and (especially) upper lids.

Complete tripod fractures leave no residual articulations and often cause substantial diplopia. In all tripod fractures, but especially those hinged at the zygomaticofrontal suture, retrobulbar hemorrhage may cause optic nerve compression and resultant loss of vision. Proptosis is a clue to this condition, but soft tissue swelling may obscure this.

Furthermore, a fracture along the optic canal may cause compression in the absence of proptosis, so it is essential to obtain, record, and follow visual acuity. The patient's glasses are frequently lost at the time of injury, but every attempt should be made to obtain a visual acuity reading. A pocket visual acuity card will suffice, as long as it is held at the same distance with each reading, and the patient does not have advanced presbyopia! It should be obvious that occlusive eye bandage would be counteractive to these measurements and thus should be avoided. It is important to record the initial absence of an afferent pupillary defect, in case one develops later owing to optic nerve compression.

The early and late complications of tripod fractures overlap those of floor fractures: intraocular damage, optic nerve compression, damage to the lacrimal gland or nasolacrimal apparatus, lid malalignment, keratitis sicca (dry eye from corneal exposure), diplopia, enophthalmos, and lid ptosis.

Other Midfacial Fractures

A LeFort fracture type I, a low transverse maxillary fracture, does not involve any of the orbital walls. Type II involves the medial wall, and type III the medial and lateral walls and the floor.

Orbital apex fractures may damage the optic canal or superior orbital fissure. Thus, any structure traversing these canals can sustain severe injury. Radiographic visualization of blood in the *ethmoid* sinuses is frequently a sign of optic canal fractures, and should be checked. CSF leaks may complicate the clinical picture, as may early or late cavernous-carotid fistula. One should look for arterialization of the conjunctival blood vessels, proptosis, and a bruit over the globe in the latter. These bruits are best heard if the patient is instructed to close both eyes, the bell of the stethoscope is gently applied to the globe in question, and the patient is then told to gently open the eyes. Only the nonauscultated eye will open. This technique avoids the muscular movement artifacts that may be mistaken for bruits.

Finally, orbital roof fractures may seriously involve the brain. A team approach with a neurosurgeon is well advised.

Inflammatory Entities of Special Interest

Orbital Cellulitis, Abscess, and Cavernous Sinus Thrombosis

Orbital cellulitis remains the most common cause of proptosis in the pediatric years, but there is sometimes confusion over terms. As discussed in the section on anatomy, the orbital septum is a continuation of the frontal periosteum and serves as a natural boundary for less virulent advanced infections. Preseptal cellulitis, then, refers to inflammatory signs limited to eyelid tissues anterior to the septum.

The term postseptal or orbital cellulitis implies that the infection began posterior to or has traversed the natural boundary, and is a much more severe threat to vision and, indeed, life. Cellulitides are most frequently the result of bacterial spread from contiguous paranasal sinuses: up to 84 per cent of orbital cellulitides have been estimated to derive from this route. In children, the maxillary and especially the ethmoid sinuses are the predominant source; in adults, there is a roughly equal contribution from the frontal, maxillary, and ethmoidal sinuses. Not surprisingly, the most common infecting organisms are *Staphylococcus aureus, S. epidermidis,* streptococci, and *Haemophilus influenzae.* Other organisms include *Escherichia coli,* diphtheroids, and anaerobes.

Infection spreads either via the valveless orbital veins or directly, since the orbit itself has no naturally occurring lymphatic tissue. The extensive venous plexus connects nose, nasopharynx, pharynx, orbit, sinuses, and cavernous sinus. If a pocket of bacteria and purulence forms within orbital fat or other soft tissue, it is termed an orbital abscess. If it occurs beneath the periosteum, it is a subperiosteal abscess. Most of these entities arise in patients under the age of 16 years. Although the overall frequency of orbital complications from paranasal sinus disease is rather small (0.5 to 3.9 per cent), these complications may be severe even with high-dosage systemic antibiotics and surgery.

Osteomyelitis may develop, usually in the frontal bone. Spread of infection from any source to the superior orbital fissure produces the so-called orbital apex syndrome: lid swelling, proptosis, optic nerve dysfunction, decreased sensation in the distribution of V1, and internal and external ophthalmoplegia. Further superior progression will involve the cavernous sinus. Sometimes a violaceous hue seeps into the lids, and the patient becomes systemically septic and obtunded; meningeal signs may be noted. It is a short step thereafter to frank

meningitis (2 per cent of all patients with orbital cellulitis in one study despite treatment), with or without brain abscess.

It is the job of the otolaryngologist and ophthalmologist to ensure that these sequelae occur as infrequently as possible. Over clinical signs are not as helpful as one might like, because there is considerable overlap. Moreover, the situation may change rapidly when the orbital venous drainage or volume becomes critically compromised. In the evaluation, one should always check the visual acuity, pupillary response (to rule out afferent pupillary defect), motility, and appearance of the optic nervehead. Should any of these become abnormal while the patient is on systemic therapy, a CT scan should be performed. This does not differentiate preseptal from orbital cellulitis, but the former should not have any of the aforementioned signs positive. CT does identify subperiosteal abscess, especially with the use of contrast material. This necessitates surgical drainage as well as antibiotics. Furthermore, in the instance of absence of both localizing abscess and improvement on antibiotics over 36 to 48 hours, surgical exploration and drainage may not be required. Ultrasonographic evaluation can identify subperiosteal abscess, but is not as reliable as CT in most hands. In summary, a high index of suspicion should be coupled with prompt and aggressive therapy in response to clinical signs.

Phycomycosis

Also termed mucormycosis by some, this represents a special and particularly lethal variant of the above processes. Nonseptate, branching fungi of the class Phycomycetes and order Mucorales comprise the pathogens, with *Mucor* and *Rhizopus* the most common genera. Interestingly, these organisms are part of the normal flora of the human upper respiratory tract, and seem to cause trouble only in certain populations. Systemic acidosis (such as seen in diabetic ketoacidosis or renal failure) or immunosuppression (as in carcinomatosis, therapy with antimetabolites, or large-surface-area body burns) predispose to phycomycosis.

The fungi cause a vaso-obliterative thrombosis and resultant tissue necrosis that progresses at an amazingly rapid rate. By the time many cases are correctly diagnosed, proptosis and an orbital apex syndrome have occurred, the condition is advanced, and the prognosis is grave (50 per cent mortality). Although the ophthalmologist is frequently asked to see such patients to test for orbital swelling and suspected orbital vein thrombosis, it is the arterial component of the vaso-obliterative condition that can contribute to the diagnostic process. Cutaneous infarction, central retinal artery occlusion, or intracranial carotid occlusion may each be presenting features. It is imperative to recognize such cases very early; a mild and early case presents with naso- or oropharyngitis with or without sinusitis (most commonly ethmoid and sphenoid involved), facial pain, and mild obtundation. If this is noted in any patient who is immunosuppressed as above, lesions in the naso- or oropharynx and face should be sought and biopsied immediately. It is easy to demonstrate the fungi, which stain avidly with hematoxylin and eosin, in distinction to most other fungi. Alternatively, the Gomori methenamine silver (GMS) stain may be used.

Treatment should be swiftly instituted, to secure control of the systemic ailment as much as is possible; there should be aggressive surgical debridement of involved areas, including orbital exenteration if necessary, combined with systemic courses of amphotericin B, flucytosine, and/or rifampin. The goal is to save the patient's life, and cosmesis must take

a back seat to this expediency. The key to success is a high degree of suspicion in the population at risk.

Wegener's Granulomatosis

This presumed collagen vascular disorder has received greater attention in recent years, and some excellent reviews are available. The full-blown syndrome includes granulomatous lesions of the sinus mucosa, with contiguous bony erosion; tracheobronchial necrotic lesions; cavitary lung lesions, predominantly at lung bases; and glomerulonephritis.

Although this condition is thought of as primarily an otolaryngologic disorder, orbital involvement has been observed in up to 47 per cent of patients. This may be not only the presenting symptom but the sole clinically significant manifestation. Ocular manifestations may be quite diverse and destructive, and can include inflammation and obstruction of the distal lacrimal system, conjunctiva, episclera, and sclera; corneal ulceration; uveitis; retinal vasculitis; optic neuritis; proptosis; and eyelid edema and retraction. Biopsy of the more accessible of these orbital lesions may demonstrate the anticipated vasculitis with granulomatous infiltration and fibrinous necrosis. The hallmark of the disease is the multinucleated Langhans' giant cell. Thus, the clinician can use the frequently more accessible lesions of the ocular system as a means of corroborating the diagnosis when necessary. It is of particular interest that at times, albeit less commonly than in cases of coexistent ocular disease, Wegener's granulomatosis may initially present ophthalmologically. Otolaryngologists should not hesitate to consult ophthalmic colleagues to establish the diagnosis of this frequently misdiagnosed or belatedly diagnosed clinical entity.

Idiopathic Midline Destructive Disease

Idiopathic midline destructive disease (IMDD) is the new nomenclature for lethal midline granuloma, clinical entities well known to otolaryngologists as a group of relentlessly progressive obliterative diseases of the naso- or oropharynx and paranasal sinuses. It is mentioned here because, in its manifestation as a focal granulomatous vasculitis, it can masquerade as Wegener's granulomatosis. The distinction is a critical one to make, because the prognosis and course of the two disorders are quite different: Wegener's is multisystemic, whereas IMDD rarely involves the lungs and never the kidneys. Furthermore, in its other expression, as central facial lymphoma, its treatment, consisting of irradiation, contrasts with the prednisone and sometimes cyclophosphamide therapy useful for Wegener's. Once again, biopsy of involved ocular and orbital regions may assist the diagnosis.

Sjögren's Syndrome

This fascinating clinical entity, which has both otolaryngologic and ophthalmic manifestations, may be a troublesome diagnostic entity. Basically, there are two groups of patients with Sjögren's syndrome. The first includes those with lacrimal and salivary gland dysfunction in the setting of a systemic collagen vascular disease, most characteristically rheumatoid arthritis. The second group consists of those who manifest only xerostomia and xerophthalmia. Interestingly, however, a certain proportion in this latter group progress after a number of years to frank autoimmune disease. Patients are characteristically women in middle age.

Thirty to 50 per cent of patients with Sjögren's syndrome probably present to the otolaryngologist with complaints of dry mouth, dry nose, tracheobronchial abnormalities, external otitis sicca, or enlarged salivary or lacrimal glands. The enlarged glands may cause confusion with Mikulicz's syndrome. Classically, this nomenclature was used to represent involvement of the lacrimal and salivary glands by masses of lymphocytes, and no doubt incorporated within it many cases of Sjögren's syndrome. Later, this term was expanded to include any enlargement of these glands, including etiologies of lymphoma, leukemia, sarcoidosis, and syphilis. The current practitioner should be aware that this term is now looked disdainfully upon, and should refer to more precise etiologic factors rather than coalescing many diseases into this one "lump".

Other effects of Sjögren's syndrome may be wrought upon the cranial nerves, which again present symptoms bringing a patient to the otolaryngologist: trigeminal neuropathy, dysgeusia, and anosmia. The reason for including Sjögren's syndrome in this chapter is that there are many coexistent ophthalmic manifestations that the otolaryngologist clinician should have some knowledge of to assist the diagnosis, especially in troublesome cases. One should look for symptoms of dry eye, caused mostly by the involvement of the eyelid's accessory lacrimal glands, and conjunctival hyperemia. Patients initially may describe a gritty sensation such as sand in the eyes, reflex epiphora that ceases as the disease progresses and the lacrimal gland's parenchyma is effaced. This involvement should always be bilateral. The quest for diagnostic certainty will be abetted by the presence of characteristic decreased tear lysozyme or, especially, lacrimal gland biopsy revealing classical myoepithelial islands. These are formed by the proliferation of residual terminal ductules, and are surrounded by sheets of lymphocytes and plasma cells. Since most cases do not involve enlargement of the lacrimal glands, it may be desirable for an ophthalmic surgeon to perform the biopsy, hoping to preserve upper lid function so as not to worsen the preexistent xerophthalmia by corneal exposure.

Finally, slit lamp examination may reveal an early tear breakup time (associated with abnormal tears), and fluorescein or rose bengal staining may show a compromised cornea and conjunctiva. Fluorescein will stain the collagen of epithelium-denuded Bowman's membrane of the cornea; rose bengal will stain devitalized or dying epithelial cells of the cornea or conjunctiva. Schirmer's test will demonstrate the decreased basic as well as reflex tearing present. One should not hesitate to use these additional data; referral to an internist or oncologist to rule out coexistent occult foci of systemic lymphoma is also an important part of the treatment.

Mucoceles

Jakobiec and Font perfectly described these entities as "orbital space-occupying lesions of sinus provenance". Their etiology is purely otolaryngologic: chronic sinusitis is thought to cause occluded ostia and accumulation of inflammatory tissue and mucus, with eventual enlargement of the sinus that encroaches into the space of the orbit. It is most frequently associated with the frontal sinus, closely followed by the ethmoid; sphenoid mucoceles are the most rare. Maxillary mucoceles are not seen; apparently, chronic sinusitis here tends toward shrinkage of the sinus, not enlargement.

The lesions are most common in adults. If they are found in children, the ethmoids are preferentially involved, since they aerate first. The presence of a mucocele in a child should alert the otolaryngologist to the possibility of cystic fibrosis. Recurrent mucoceles of the frontal and ethmoid sinuses can cause significant depression of the globe and be very difficult to eradicate. Unlike the anterior displacement of the globe that occurs in Graves' ophthalmopathy and orbital pseudotumor, inferior displacement of the globe and ptosis, rather than proptosis, predominate in patients with frontal sinus mucoceles. This is a helpful diagnostic point. Both ocular motility and visual acuity are much less affected by mucoceles than by Graves' ophthalmopathy and orbital pseudotumor. However, if the sphenoid sinus is the encroacher, the more posterior location provides prime access to the optic nerve.

Ophthalmic consultation should be sought for all cases that intrude upon the globe to ensure optimal evaluation and results. Roentgenographic evaluation usually provides the diagnosis, but confusion with ossifying fibroma may occur, so caution is advised. As a final rare avis, beware the prolapse of ethmoid sinus polyps into the orbit. Fortunately uncommon, it yields quite a different radiologic picture and may be confusing.

Orbital Pseudotumor and Graves' Ophthalmopathy

These two entities are the most common causes of proptosis in adults. Even for experts the differential diagnosis can be confusing, so we hope to point out salient differential features.

Orbital pseudotumor is the term given to inflammations of the orbit that have no known cause (such as a retained foreign body), produce swelling and proptosis, and are not neoplastic processes. It is often a multicentric process, and may involve myositis, dacryoadenitis, sclerotenonitis, trochleitis, optic nerve perineuritis, and orbital fat and connective tissue. A special case, the Tolosa-Hunt syndrome, is discussed later among the facial pain syndromes.

Presenting features may be almost identical to those of Graves' ophthalmopathy: pain, diplopia, proptosis, periorbital swelling, inflamed and edematous conjunctiva, and decrement in visual acuity. However, orbital pseudotumor is more classically noted for its acute evolution, as opposed to the more gradual onset in Graves' ophthalmopathy.

Radiologic evaluation, by axial and direct coronal CT or MR scans of the orbit, is crucial. In Graves' ophthalmopathy there is thickening of multiple muscles, while the tendinous insertions of those upon the globe is spared. The most commonly involved muscle is the inferior rectus, and thickening of muscles appear to "hug" the medial wall to involve the medial rectus next, and then the superior rectus. Involvement of only one muscle is very rare, especially if it is the lateral rectus in isolation. Direct coronal views complement axial CT of pseudotumor, disclosing thickening of the rectus muscles *involving* the tendinous insertions. It is quite common to observe a muscle enlarged in isolation, as well as other foci of involvement as noted above. If the aforementioned clinical features are noted in a child, a diagnosis of orbital cellulitis must be entertained, since that is the most common cause of proptosis in the pediatric age group. Graves' disease is uncommon, but not impossible, in this age group.

One might expect that evidence of other systemic symptoms and chemical studies would reveal the diagnosis of Graves' ophthalmopathy, but the otolaryngologist should be reminded that many individuals with true Graves' ophthalmopathy are both systemically and chemically "normal". This is especially true since most "thyroid profiles" performed with modern techniques do not include a thyrotropin-releasing hormone test. One cannot exclude the diagnosis of Graves' disease until this has been performed, even in the presence of normal T3, T4, and TSH levels.

If the otolaryngologist ascertains that one of these entities is involved, there should be swift referral to an ophthalmologist specializing in orbital conditions, since vision may be severely threatened, and any diagnostic work and therapeutic surgery is best done by someone with considerable experience in the matter. We usually prefer a team otolaryngology-ophthalmology approach to the orbital decompressions sometimes required by the Graves' patient. When the purpose of the surgery is to relieve the posterior compression of the optic nerve by the origin of the medial rectus muscle, we feel that extension of the medial wall decompression should extend as far as prudence allows. Sometimes this may best be accomplished by an external approach to the ethmoid via a Lynch incision. Conversely, a decompression for cosmesis may be adequately performed by means of a lateral orbital and inferior fornix incision. We have been concerned that pulling the orbital fat into the maxillary sinus also contributes to diplopia by virtue of connective tissue strands connecting the fat with the muscles. The surgical details of orbital decompression and the post-decompression surgical rehabilitation of patients with lid lag, diplopia, and corneal exposure are beyond the scope of this chapter.

Neoplasia

This section describes tumors that commonly involve both otolaryngologic and ophthalmic provenance, and therefore should not be viewed as an exhaustive review of either orbital or nasopharyngeal neoplasia. For the sake of simplicity of approach for the clinician, we have divided it into two sections: common tumors arising in situ and those arising elsewhere.

Neoplasia Arising in the Orbit and Contiguous Areas

Rhabdomyosarcoma. This is the most common primary orbital malignancy of childhood, with an average onset at age 7 years. It presents acutely, with proptosis that usually advances rapidly. The diagnosis should be kept in mind, because patients and their parents often cite a history of trauma to the area, which may delay correct diagnosis and treatment. Lethal if not treated, rhabdomyosarcoma now carries a very good prognosis when a combination of chemotherapy and radiation are used.

Neurofibromatosis. This disease is a phakomatosis, inherited dominantly with incomplete penetrance, which produces tumors of proliferating Schwann cells. Neurofibromatosis is classified according to three types: NF1, NF2, and NF3. NF1 (von Recklinghausen's disease) involves many regions of the body; NF2 (NF central type) has bilateral acoustic neuromas; and NF3 (intestinal neurofibromatosis) is very rare. Each of these dominantly inherited disorders may involve different abnormalities in the production of nerve growth factor. Recently, abnormalities on two chromosomes have been identified that may

further aid in the diagnosis.

In NF1, neurofibromas can arise essentially anywhere, and one may be called upon to make a diagnosis of a tumor presenting in sites of primary otolaryngologic interest. In that circumstance, ophthalmic evaluation can enable a diagnosis to be made noninvasively, given the wealth of ocular signs in this disease. To wit: plexiform neurofibromas are pathognomonic for neurofibromatosis; 92 per cent of patients over the age of 6 years have iris Lisch nodules; there is an increased incidence of both gliomas and meningiomas of the optic nerve; pulsatile exophthalmos from sphenoid dysgenesis may be present; and unilateral or bilateral glaucoma may be found owing to the locations of the neurofibromas.

Fibro-osseous Tumors. These can arise from either orbital bones or paranasal sinuses. The most common simulator here is the ivory osteoma, predominantly from the frontal synostoses adjacent to the ethmoid sinuses. Fibrous dysplasia not infrequently causes displacement of the globe and, less often, narrowing of the optic canal. Fortunately, consequent optic atrophy is unusual. Ossifying fibroma, frequently confused with fibrous dysplasia, is much more aggressive.

Osteogenic sarcoma is massively destructive, and is of special interest because in patients treated with irradiation for retinoblastoma in early childhood it has a much increased incidence, both in the portal of treatment and elsewhere. This predisposition has been linked to oncogenes.

Other Sinus Tumors. Of carcinomas found in the orbit and sinuses simultaneously, most originate in the sinuses. Maxillary origins are much more common than others; frontal or sphenoid sources are decidedly rare. Symptoms depend on the vector of tumor expansion: eg, upward displacement of the globe with a maxillary source.

Most of these carcinomas are squamous. The presence of adenocarcinoma is so rare that one should consider metastatic disease as the most likely source. Second most common is the inverted squamous papilloma, which can undergo malignant transformation. Sinus tumors that occasionally invade the orbit include adenoid cystic carcinoma, malignant mixed tumor, and muco-epidermoid carcinoma. Adenoid cystic carcinoma can also present primarily in the lacrimal gland and invade and metastasize to other structures.

Metastatic Tumors

There are no surprises in this category. The most common metastatic tumor in adult females is breast carcinoma; in males, testicular and prostate carcinoma; in both sexes, lung and kidney carcinoma. Children manifest metastatic neuroblastoma, usually late in the course of the disease as a preterminal finding, and metastatic Ewing's sarcoma of the bone.

Oculomotor-Vestibular Interactions

In no other realm do the fields of otolaryngology and ophthalmology interrelate so fundamentally as in the vestibular system. Nonetheless, the vestibular apparatus and its plenitude of interactions with the extraocular motility system, among others, remains a little-understood realm by practitioners in both fields. A great deal of information has been revealed

about its internal structure and physiology, but this sometimes seems dwarfed by what remains to be unraveled.

Many patients approach the otolaryngologist with vague complaints consistent with vestibular dysfunction: dizziness, vertigo, or disequilibrium. A proper evaluation of these patients requires thorough understanding of some basics of oculomotor-vestibular interactions.

Pertinent Vestibular Anatomy

The peripheral vestibular apparatus contains two main sensory structures: the semicircular canals and the otolith crystals. The former are three in number: the horizontal (lateral), anterior (superior), and posterior (vertical). Within the bony canals lie membranous canals, filled with endolymph. These are dilated on one end to form ampullae, and on the other to form the utricle. An additional membranous structure, the saccule, attaches to the utricle.

Within the ampullae, utricle, and saccule are the ciliary sensory epithelia. The cupula, consisting of gelatinous material overlying the epithelium, also contains the calcium carbonate-comprised otoliths in the utricle and saccule. The otoliths are sensitive to linear acceleration of the head, while the semicircular canals are receptive to angular acceleration.

Nerve fibers condense around the three ampullae, the utricle, and the saccule to form the vestibular nerve. This traverses the cerebellopontine angle, lying posterior to the cochlear nerve and below the facial nerve. The vestibular nerve enters the brain stem to terminate in the vestibular nuclei, which lie in the medulla beneath the floor of the fourth ventricle. Alternatively and additionally, connections are made to the vestibulocerebellum. This consists of the flocculus, nodulus, ventral uvula, and ventral paraflocculus. The vestibular nuclei number four main regions with other less well characterized ones.

Eye Movements: Vestibular Reflexes

To understand the vestibulo-ocular reflex and its various neuronal connections, it is helpful to understand the type of eye movements involved and their generation.

Saccades. Saccades are rapid eye movements that serve to place the image of the object of regard upon the fovea. They can be voluntary or involuntary in nature. The stimulus is usually an object perceived in the visual periphery. Latency approximates 250 msec, but it can vary with stimulus characteristics. The larger the scope of the eye movement, the higher the peak velocity will be; it usually varies from 30 to 700 degrees per second. Velocity is not under conscious control, but can be influenced by the overall level of arousal of the subject.

Voluntary saccades are initiated by the frontal eyefields (FEFs) and the superior colliculi (SCs). These two are thought to be parallel pathways, because damage to both, but not just one, is required for permanent serious disruption of saccadic generation. Each FEF or SC activates the contralateral paramedian pontine reticular formation (PPRF), which in turn activates the abducens nucleus ipsilateral to itself. Abducens interneurons transmit activation via the MLF to the medial rectus subnucleus on the side of the original FEF-SC. Thus, activation of the right FEF causes gaze to the left via a saccade. The electrophysiologic

production of vertical saccades necessitates the simultaneous stimulation of corresponding points in both FEFs or SCs.

Involuntary saccades, also known as "quick phases", are generated through the vestibular and oculomotor systems, as described below.

Smooth Pursuit. Pursuit eye movements allow one to follow a moving target, whether one's head is moving or not. Retinal slip is the usual stimulus for its initiation, but other generators include other sensory systems monitoring target motion or body motor functions, or high level perceptual representation of target movement in space. Pursuit latency is about 130 msec, and a target traveling less than 50 degrees per second is easily followed.

Although the pathways for voluntary generation of smooth pursuit movements are not as well known as those for saccades, it is known that damage to a hemisphere's parieto-occipitotemporal junction disrupts ipsilateral smooth pursuit. These regions project to ipsilateral pons and cerebellum; the flocculus of the latter is especially important. Again, the abducens nucleus is stimulated, with activation of the contralateral medial longitudinal fasciculus, and hence the contralateral medial rectus muscle.

Central Pathways for Vestibule-Induced Eye Movements. Generation of these is based on sensory input from the semicircular canals and the otoliths. There are two basic pathways, one for horizontal and one for vertical movements. For the former, the pathway is the same as for voluntary horizontal gaze as above, the PPRF contralateral to the stimulated labyrinth being the premotor stimulus. For vertical and torsional movements, the MLF, the brachium conjunctivum, and other brain stem pathways project to the oculomotor and trochlear nuclei. This is expanded below.

Ocular Reflexes. Since we as living beings are constantly moving within our environment, some mechanisms must exist for coordinating the movement of our eyes with that of our heads. There are indeed several of these, and the vestibulo-ocular reflex is the most important. Two more minor systems are the cervico-ocular and vestibulo-ocular reflexes.

The cervico-ocular reflex relays information from the muscles and joints of the neck to the vestibular nuclei and cerebellum. Its importance becomes apparent only if a labyrinth is destroyed, producing "head nystagmus" during whole body rotation when the head is free to move.

The vestibulo-ocular reflex (VOR) is a phylogenetically ancient and powerful aligner of head and eye movements. It ensures that eye movement is equal and opposite to head movement. It is mediated mostly through the semicircular canals and responds best to the effects of brief, high-frequency head movements. The importance of a normal VOR is underscored by the fact that even minimal movements such as the bounce of a moving automobile or those associated with pulse transmissions can be enough to blur vision in its absence.

The concept of gain of the VOR is important in detecting vestibular disease. It is defined as the ratio of the amplitude of the eye movements to the amplitude of the head movements, calculated from the peak velocities attained. The ideal normal gain is equal to

1.

Semicircular Canals. Upon the onset of head motion, the semicircular canals function reciprocally so that if one excites, the other inhibits. These impinge upon a preexisting resting tonus to modulate responses. Each canal predominantly excites two ocular muscles, one in each orbit, which will serve to rotate the globe in approximately the same plane as that in which the semicircular canal is located. Thus:

Horizontal canal: ipsilateral medial rectus and contralateral lateral rectus.
Anterior canal: ipsilateral superior rectus and contralateral inferior oblique.
Posterior canal: ipsilateral superior oblique and contralateral inferior rectus.

The pathway between the posterior semicircular canal and the superior oblique muscle is via the ipsilateral medial vestibular nucleus and the ipsilateral MLF to excite the trochlear nucleus.

Ocular Tilt Reflex. The ocular tilt reflex that follows stimulation of the utricle is similar, in that the ipsilateral superior oblique muscle is activated by a pathway that involves relay in the lateral vestibular nucleus. Thus, the eye on the stimulated side intorts while the contralateral eye extorts. The ocular tilt reaction also includes movement away from the stimulated utricle, and a vertical skew deviation in which the eye on the stimulated side is elevated while the contralateral eye is depressed. An identical effect can be produced by stimulation of the contralateral interstitial nucleus of Cajal.

Control of Eye-Head Movements

Interaction of visual and vestibular system occurs when a target is perceived in the peripheral visual field. After a 250-msec latency a saccade begins to achieve foveation. Then, 20 msec later, head movement begins in the same direction. This excites the labyrinth, which results in an oppositely directed compensatory slow vestibular eye movement, the VOR.

Rotation of the head in the light activates both the VOR and the optokinetic nystagmus system (OKN). The latter is a retinally directed system that causes slow phase movements in the direction of the movement of an external visual pattern. There is an accompanying refixational quick phase, described as "nystagmus". Interestingly, OKN can never be active without the accompaniment of activation of the smooth pursuit system. Sustained rotation in the dark initially activates both the VOR and OKN, but the VOR extinguishes after about 30 sec. The OKN (pursuit) system then continues alone. If the rotation were to cease, the change in angular acceleration would activate the VOR, which would in this circumstance produce undesired post-rotational nystagmus and vertigo. This may be neutralized by the presence of optokinetic after-nystagmus (OKAN), which represents the continuation for a few seconds of the preexistent OKN.

OKAN does not have input from the pursuit system, so it is the only example of a centrally generated OKN, important for those who wish to study the phenomenon.

The synergy between the VOR and OKN is extremely important for control of head-eye movements. If one is tracking an object moving in space the head movement excites the

vestibular system, but this is negated by a simultaneous pursuit eye movement command.

Finally, it has been demonstrated by many investigators that the VOR may be subject to adaptation, both short and long term. Lesions of the vestibulocerebellum may modify or abolish the ability to adapt.

Clinical Assessment of Vestibular System

There are several techniques by which one can assess the integrity of the vestibular and oculomotor systems.

General Bedside Techniques. First, observe patients at rest, with their eyes fixating on a distant target. Is there nystagmus? Small-amplitude movements may be hard to see; a +20 diopter (Fresnell) convex lens can be used to magnify the image of the patient's eyes, or the direct ophthalmoscope employed to observe movement of the optic disc. While improving the physician's view, the 20-diopter lens also impairs the patient's ability to employ fixation to prevent nystagmus (darkness also releases vestibular nystagmus, but it then cannot be observed). The patient should then close the eyes to determine whether loss of fixation provides onset of nystagmus.

A reliable but nonlocalizing sign of vestibular disturbance is obtained by having the patient shake the head in the horizontal plane at 10 times per second, then repeat the same in the vertical plane. Check thereafter for transient jerk nystagmus.

Paroxysmal vertigo can be addressed by postural testing, wherein the tilted head and trunk are swiftly moved to 30 degrees below the horizontal and nystagmus sought in primary, right, and left gaze.

The status of the VOR gain may be deduced by two techniques. First, a near-card is used to determine the patient's visual acuity. Next, the patient should shake the head vertically and then horizontally at a frequency of 2 times per second while reading the card. If the gain is abnormal, the patient's vision will blur several lines above the acuity previously ascertained. Telltale corrective saccades should be sought at the same time.

The second technique is more sensitive. Using a direct ophthalmoscope, observe the patient's optic nervehead as the patient moves the head at 2 times per second. If the VOR gain was normal, there should be no perceived movement of the optic nervehead. If the optic nervehead moves opposite to the head movement, the gain is <1 and the reflex hypoactive; if it moves in the same direction as the head, the gain is >1 and the reflex is hyperactive. If the eye drifts in the same direction irrespective of head movement, this indicates general vestibular dysfunction. This technique works even in the comatose patient, for the head can be moved with a lid speculum in place.

Oculocephalic Maneuvers. The oculocephalic maneuver is also known as the doll's-eyes phenomenon. If the patient under study is awake, he is instructed to fixate on a target directly in front of him. Simultaneously, the examiner turns the patient's head horizontally or vertically. If the patient is comatose and there is no reason to suspect a cervical fracture, the eyelids are held open by the examiner, and the same maneuver is performed. The normal

response is now predictable from the above discussions: the eyes will move conjugately in the direction opposite of that of head movement. This ensures the integrity of nuclear and infranuclear oculomotor pathways.

Caloric Testing. These tests are used predominantly to test the horizontal semicircular canals, in order to demonstrate the integrity of the brain stem. An important first step, often omitted, is to check both external auditory canals for the presence of intact tympanic membranes. Once this is done, the supine patient should have the neck flexed approximately 30 degrees, so as to place the horizontal semicircular canal in the vertical plane. The warm water should be at 44°C, the cold water at 30°C. Gentle irrigation of the external auditory canals is then performed.

In an awake patient, there will be a conjugate nystagmus; with warm water the *slow,* vestibular movement phase will be to the opposite side, and with cold water to the same side. Recall that the mnemonics "COWS" refers to the *quick* refixational movement phase, and that the vestibular response is the slow "pursuit" movement. This is underscored by the fact that in comatose patients, all that is observed is a tonic conjugate deviation as above, with no significant refixation movement. This is because the PPRF is not functioning in a comatose patient.

For vertical caloric tests one must stimulate both ears simultaneously with either cold or warm water. In the awake patient, warm water produces a conjugate nystagmus with upward *slow* phase (mnemonic: "warms things up"), and cold water a downward slow phase. In comatose patients the movements are tonic, without refixation movements. Because vertical caloric tests involve questions of the function of the entire system, they are thought not to be as reliable as horizontal caloric tests, which are often used to evaluate a part of the system. Oculocephalic tests may be preferable to vertical caloric testing. However, it cannot be denied that calorics remains the best way to evaluate unilateral peripheral vestibular function.

The otolaryngologist should be alerted to the existence of pseudocaloric nystagmus. This occurs in the setting of labyrinthine dysfunction; the quick phase of nystagmus here always beats away from the injured ear and does not respond to warm or cold stimulation. It has been suggested that this represents the unmasking of a preexistent vestibular nystagmus through tactile stimulation.

Clinical Interpretation of Vestibular Nystagmus. Table 1 is intended to bring all the foregoing information together and aid in the making of clinical decisions concerning vestibular dysfunction. First, a few specific considerations.

Vestibular imbalance causes spontaneous nystagmus. This can be horizontal or vertical jerk nystagmus, which follows Alexander's law: there is an increase in intensity when the eyes are moved in the direction of the fast phase. Lesions of the central vestibular mechanisms always give a mixed-type nystagmus, unlike peripheral nystagmus. Problematically, some brain stem lesions may create a nystagmus, mimicking the loss of one semicircular canal.

Peripheral vestibular disorders cause nystagmus that decreases upon fixation because of the intact ocular stabilization provided by the smooth pursuit system. Flourens' law dictates that disease affecting one semicircular canal will produce abnormal eye movements in a plane

parallel to the plane of the disease canal. The most common manifestation of unilateral peripheral vestibular disease is a mixed horizontal-torsional nystagmus. Central vestibular derangement is usually accompanied by concomitant derangement of smooth pursuit, so visual fixation does not stabilize the eyes.

Table 1. Aid to Clinical Diagnosis of Peripheral Nystagmus

Symptom	**Peripheral**	**Central**
Severity of vertigo	Marked	Mild
Direction of nystagmus	Usually unilateral; fast opposite lesion	Bi- or unidirectional
Duration	Finite, recurrent	May be chronic
Tinnitus/deafness	Common	Rare
Effect of fixation	Inhibits nystagmus	Not inhibited
Directional environment spins	Toward fast	Variable
Direction of pastpointing	Toward slow	Variable
Direction of Romberg fall	Slow	Variable
Effect of head turning	Changes Romberg fall	No effect
Torsional nystagmus	Never pure	Possible
Common causes	Infection / inflammation, Ménière's, vesicular, trauma, toxic	Acoustic neuroma, demyelination.

The term lateropulsion describes the sensation of the body being pulled toward one side - usually toward that of the lesion. The eyes tonically deviate toward the lesion with the corrective quick phases in the opposite direction. Lateropulsion may appear in brain stem ischemia (the Wallenberg syndrome) or in more peripheral disease. Destructive lesions of the labyrinth behave analogously to cold water caloric testing. The slow phase of the nystagmus is to the side of the lesion. Peripheral vestibular disease never produces purely rotational eye movements.

Vestibulocerebellar disease can often disrupt the ability of the VOR to adapt. This occurs in the face of little change in the VOR or OKN. Most commonly, however, there are abnormalities in the smooth pursuit gain. Usually, the gain is decreased so that "catch-up" saccades are required.

Lesions of the anterior visual pathways usually impair OKN. With acute vestibular disease, a directional preponderance of OKN will be observed with increased slow phase toward the side of the lesion.

Selected Aspects of Cranial Syndromes

Bell's Palsy and Melkersson-Rosenthal Syndrome

It is not unusual for a patient to present to the otolaryngologist with the hallmarks of Bell's palsy: a lower motor neuron, facial nerve paresis. In adults it usually has a sudden onset. Interestingly, it may be preceded or followed by trigeminal-type pain. Its etiologies are thought to be either ischemic or viral, but it may also be associated with diabetes mellitus, hypertension, and pregnancy. We are most concerned when the problem results from neoplasm, surgery, or both. Such patients may be at high risk for corneal complications because of coexistent anesthesia (trigeminal nerve), absent tearing (greater superficial petrosal nerve), and impaired upward eye movement (of supranuclear, nuclear, or intranuclear origin). These are the patients most likely to need frequent eye care or tarsorrhaphy.

Seventy-five per cent of patients with idiopathic Bell's palsy recover spontaneously by the third month after onset; 25 per cent do not recover well, and aberrant regeneration is common in this group. Curiously, the prognosis for correct somatotopic reconnection of regenerating axons is much higher in immature animals and humans than in adults. This implies that the regenerating axons of young animals is guided by non-neural cues! For humans, poor prognostic signs include advanced age at the onset of disease, the presence of dysacusis, or impaired lacrimation.

The controversy over whether or not to employ corticosteroids in the treatment of this condition is discussed elsewhere in these volumes. We feel that a short (1-week) course of oral steroids in patients without risk factors for steroids may avoid continued damage to a presumably swollen facial nerve in a rigid fallopian canal.

Interface with the ophthalmologist should be rapid, since these patients may develop a corneal exposure syndrome that can progress rapidly to become a corneal ulcer. Usually, a combination of an intense lubrication regimen with or without patching at night suffices; occasionally, lateral tarsorrhaphy is necessary. We suggest placing the patient on an artificial tear ointment on the first visit, with swift referral. We recommend the sterile lubrication ointments that contain no preservatives, since chronic use of ointments with preservatives can cause epithelial damage.

An interesting related clinical entity is the Melkersson-Rosenthal syndrome, which combines uni- or bilateral facial palsy with chronic facial swelling. Some patients also have a markedly furrowed tongue. This generally occurs in childhood, and is distinct from Bell's palsy.

Blepharospasm and Hemifacial Spasm

The otolaryngologist may see these patients because of their "abnormal facial movement". Ophthalmologists usually see them either because of the mechanical disruption of vision, which can be totally disabling if severe, or on referral for botulinum A toxin injections - a temporary treatment. So-called "benign essential blepharospasm" is manifested by intermittent, occasionally constant closing of the palpebral fissures due to abnormal excitation of the orbicularis oculi muscle. It can have a multitude of ophthalmic causes, which

should be eliminated carefully before a diagnosis of "benign essential blepharospasm" is made.

Meige's orofacial dystonia is a more complex syndrome in which there is spasmatic lid closure associated with contracture of the middle and lower facial musculature. The first sign of this disease is usually blepharospasm, in isolation. The cause is unknown, and attempts to control it pharmacologically with Baclofen (a-GABA-ergic drug) or other central nervous system active agents are variable and unpredictably successful.

Both Parkinson's disease and Huntington's chorea feature blepharospasm as prominent signs. Other clinical signs steer toward neurological referral.

The most common external cause of blepharospasm is inflammation of the anterior segment of the globe, resulting in reflex trigeminal spasm. Clues include injection of the conjunctiva, corneal staining with fluorescein, dry eye syndromes, or pronounced photophobia. If these are present, an ophthalmologist should be consulted to confirm suspicions. The blepharospasm ceases with treatment and resolution of the ocular condition.

It is not unusual to see mild contracture of the facial muscles after the resolution of a Bell's palsy. However, vermiform (wormlike) movements of the facial musculature raise the possibility of pathologic facial myokymia, and further studies should be obtained to rule out pontine compromise by either intrinsic or extra-axial tumors.

In the past, proximal sectioning of the facial nerve, eg, at the stylomastoid foramen, was advocated, but there is a high rate of recurrence and a risk of total facial paralysis. More peripheral sectioning leads to even higher rates of recurrence. Baclofen, benztropine mesylate (Cogentin), clonazepam, trihexyphenidyl hydrochloride (Artane), and other drugs have been tried and are successful for some patients. Orbicularis myectomy is the surgery of choice for this condition, owing to better postoperative facial appearance and a lower recurrence rate. Injection with botulinum A toxin is another alternative, very successful for many patients, but the injections need to be repeated about every 3 months or more frequently.

Hemifacial spasm should not be confused with blepharospasm; it usually is unilateral tonic or clonic activity seen in the muscles of facial expression. It continues during sleep, unlike blepharospasm. It also should be differentiated from seizure activity; if Jacksonian progression is absent, electroencephalography may resolve the diagnosis. A small number of these cases occur as postparalytic facial nerve disease, but most are believed to arise as a result of compression by vascular loops on the facial nerve as it leaves the brain stem, which causes irritation of the nerve and spontaneous recurrent firing. Emphatic transmission between fibers also occurs. Some patients complain of simultaneous noise in the ear caused by stimulation of the stapedius nerve. The vessels most commonly implicated are the anterior or posterior cerebellar arteries. Carbamazepine and baclofen taken orally and botulinum injections have been very helpful; intracranial surgical decompression is advocated for young, healthy patients who are noting progressive facial weakness and who have good hearing in the opposite ear.

Gradenigo's Syndrome

The classic case of Gradenigo's syndrome involves an abducens nerve palsy, pain due to involvement of the gasserian ganglion, and ipsilateral facial nerve palsy due to suppurative otitis media. In the case of severe mastoiditis, the petrous bone can be involved to its apex and produce a focal meningitis. The segment of the abducens nerve involved is that which passes beneath the petroclinoid (Gruber's) ligament adjacent to the mastoid air cells.

Pain is usually the first finding, usually preceding the palsies by a few days. The pain may be quite severe, may localize in or around the eye or contiguous face, and may be worse at night. In rare cases the patient may experience occipital pain from a recurrent branch of the trigeminal nerve.

The prompt recognition of otitis media coupled with efficacious antibiotic regimens has decreased the overall frequency of this syndrome. However, the clinician must be aware of other possible causes; tumors as well as aneurysms of the intrapetrosal (carotid canal) internal carotid artery have been reported. In the absence of clinically demonstrable otitis media, it is advisable to refer to the neuro-ophthalmologist and obtain proper neuroradiologic evaluation.

Ramsay Hunt Syndrome

Ramsay Hunt described this clinical entity in the early years of this century as characterized by severe pain and ipsilateral facial palsy due to herpes zoster otitis externa. The neuritic pain usually precedes the vesicular eruption by several days, and the involved skin may experience hyperalgesia and paresthesias. Resolution is slow.

Hunt himself thought that the pain came from involvement of the geniculate ganglion itself, but data have accrued in recent years making that theory unlikely. Lymphocytic infiltration of the facial nerve has been observed throughout its length. Other evidence suggests that the syndrome represents a "cranial polyneuropathy".

It is important to arrange rapid ophthalmic consultation for a patient with this syndrome, since keratouveitis (requiring corticosteroid therapy) may be present at the same time, masked by the overall pain status. Frequent examinations may improve the opportunity for an unblemished recovery. Topical or systemic acyclovir may be helpful in ameliorating some acute cases. The management of postherpetic neuralgia is beyond the scope of this chapter.

Tolosa-Hunt Syndrome: Painful External Ophthalmoplegia

This clinical entity was briefly mentioned above and warrants further discussion here. Description of the syndrome was first made by Edvardo Tolosa in 1954, followed by Hunt's compilation of six cases. The otolaryngologist should be aware of the ramifications of this disease.

Typically, there is the acute onset of pain of a particular nature: retro-bulbar and forehead pain, steady, not throbbing, and boring in quality. This is to be distinguished from

the sharp, lancing pain of tic douloureux. The pain usually precedes the ocular motor dysfunction by several days; thereafter, there is involvement of cranial nerves III, IV, and VI with predictable results. V1 is usually involved, but V2 may be also. Severe disease may involve orbital sympathetic nerves, with consequent pupillary constriction and/or lid droop, and the optic nerve, with impairment of vision. The cause is inflammation, usually within the cavernous sinus, but this may extend to the superior orbital fissure (SOF) or even the optic foramen. The inflammation causes an inflammatory perineuritis and vasculitis. Sometimes narrowing of the intracavernous segment of the carotid artery is demonstrated angiographically. Patients most usually are in their middle to advanced years.

True Tolosa-Hunt syndrome usually responds exquisitely to relatively moderate doses of prednisone. It has been recommended that a trial of 60 to 80 mg of prednisone be administered for 24 to 48 hours; success in controlling the pain with this regimen often confirms the diagnosis. However, this cannot be asserted so boldly, because the syndrome of painful ophthalmoplegia can be caused by other entities. Neoplastic entities familiar to the otolaryngologist, such as nasopharyngeal carcinoma, perinasal sinus adenoid cystic carcinoma, and squamous carcinoma, may be the culprits and may be steroid responsive, at least initially. The same is true of parasellar neoplasms such as pituitary adenomas, craniopharyngiomas, and meningiomas. Arterial aneurysms may mimic the syndrome, including the steroid responsiveness, as may syphilis, temporal arteritis, diabetes mellitus, rheumatoid arthritis, and systemic lupus erythematosus. Trauma to the head may produce Tolosa-Hunt syndrome by damaging the contents of the SOF.

Thus, the otolaryngologist may quite easily be involved in a case presenting as a Tolosa-Hunt syndrome. It is imperative that such patients be subjected to rigorous examination of the oropharynx, blood studies (VDRL, ESR, fasting serum glucose, rheumatoid factor, and antinuclear antibodies), and neuroradiologic evaluation, including attention to the base of the skull (this last should not be omitted even though symptoms focus on the cavernous sinus - SOF area). Steroid trial may be begun as soon as neuroradiologic study is completed, but the clinician should be advised that some patients require higher doses than those quoted above for analgesia. Furthermore, the ophthalmoplegia may not recover for days or months after cessation of pain, and indeed may never resolve completely. It is advised that the patient be assessed by an ophthalmologist, so that motility dysfunction can be determined precisely and recovery charted, and to ensure that the optic nerve is not compromised. Recurrence may be noted and should not be taken to represent an initial misdiagnosis. Gradual tapering of steroids may be necessary to prevent inflammatory rebound.

Temporal Arteritis

Temporal (giant cell) arteritis is only one cause of ischemic optic neuropathy, but it is the one that may present most commonly to the otolaryngologist. This disease is very much one of the elderly, its incidence increasing with age. It is caused by granulomatous vasculitis affecting the arteries feeding the globe and optic nerve, but it must be viewed for what it is: a whole-body vasculitis. This explains its usual acute presentation as an acute loss of vision in one eye, as well as its other attendant symptomatology.

These nonocular symptoms may be the ones that motivate patients to seek medical attention other than with an ophthalmologist. They may be having chronic headaches, with

exquisite scalp tenderness over the superficial temporal artery. There may be moderate to severe jaw pain upon mastication; this should be rather diffuse and not related to the temporomandibular joint. This should also be contrasted with the pain in trigeminal neuralgia (see below). Symptoms of polymyalgia rheumatica may occur: malaise, anorexia, and joint pain, especially in the cervical and shoulder region.

The presence of nonophthalmic symptoms may well precede the devastating acute loss of vision. With patients who have jaw pain as described above, not ascribed to temporomandibular joint syndrome or tic douloureux, it is important to ask whether there have been any transient visual obscurations; these occur in up to 10 per cent of patients. One should also ascertain whether there is or has been diplopia, noted in up to 10 per cent of patients with temporal arteritis.

Obtaining a Westergren erythrocyte sedimentation rate (ESR) is crucial in patients with "atypical" jaw pain, for this may make the diagnosis. Appropriate upper limits of normal ESRs adjusted for age follow the formulas age/2 for men, age + 10/2 for women. Strong suspicion warrants a temporal artery biopsy. A patient may be begun on steroids, the "cure" for the disease, immediately; histopathologically positive temporal arteries will safely remain so for at least 2 weeks after initiation of steroid therapy, perhaps longer. Criteria for positivity include not only the presence of giant cells but also destruction of the elastica.

Prompt referral of suspected or clear cases of temporal arteritis to an eye specialist is imperative, because the severe visual loss is irreversible (often the final acuity is less than 20/200), and there is a high incidence of involvement of the second eye. A useful guide to such occurrence is that in untreated patients one-third of second eyes will be involved 24 hours after the first, one-third within 1 week, and one-third within 1 month. A high index of suspicion should be coupled with immediate eye examination for elderly patients with jaw pain.

Raeder's Paratrigeminal Neuralgia

This constitutes another instance of facial pain. The classic description includes severe unilateral headache, ipsilateral facial pain or dysesthesias, usually in the distribution of cranial nerve V1; and ipsilateral Horner's syndrome. It is not uncommon to find nasal congestion and rhinorrhea as well. Middle-aged and elderly individuals are predominantly affected. The facial pain is easily confused with the pain of sinusitis.

The literature provides descriptions of subtypes of this disorder. The most ominous variant demonstrates multiple parasellar cranial neuropathy, is indicative of serious middle cranial fossa disease (trauma, neoplasia, internal carotid anomalies, inflammation), and must be distinguished from Tolosa-Hunt syndrome. A second variant is thought to represent cluster headache. A third manifests with headache and V1 signs; this last may be an early form of the first subtype, and should be carefully followed for the later insidious development of ocular cranial nerve palsies. From the foregoing, it should be apparent that a multispecialty approach to this syndrome is essential for thorough patient care.

Trigeminal Neuralgia

The purpose of this section is not to review the vast literature on trigeminal neuralgia, but to attempt to distinguish the pain described as inherent to the syndrome from that of others, such as Tolosa-Hunt syndrome or temporal arteritis.

The pain of an attack is paroxysmal in nature, consisting of extreme, sharp, lancing pain in the distribution of one or more branches of cranial nerve V. This distribution alone is an important differential: the mandibular division is involved in perhaps up to 70 per cent of cases; the ophthalmic is least involved. Isolated V1 involvement is as low as 1 to 2 per cent.

Each paroxysm is very brief, lasting from a few seconds to 1 minute. However, there may be repeated paroxysms, and these may result in lasting pain for several hours. Another important differential is the existence of trigger zones; attacks may be precipitated by touching a certain part of the face, or by a specific facial movement, such as chewing. As a result, patients with this disease frequently immobilize their faces in an attempt to avoid further torture, sometimes holding their faces with their hands. Between attacks, they report complete relief.

The age group predominantly affected is identical with that of most facial pain syndromes, but instances have been reported in children. This is most certainly not the case in temporal arteritis.

We prefer the term atypical facial pain for all cases not securely categorized; this ensures that the examiner remains alert to the diagnostic possibilities even while performing therapeutic trials. Relief of pain does not confirm a diagnosis.