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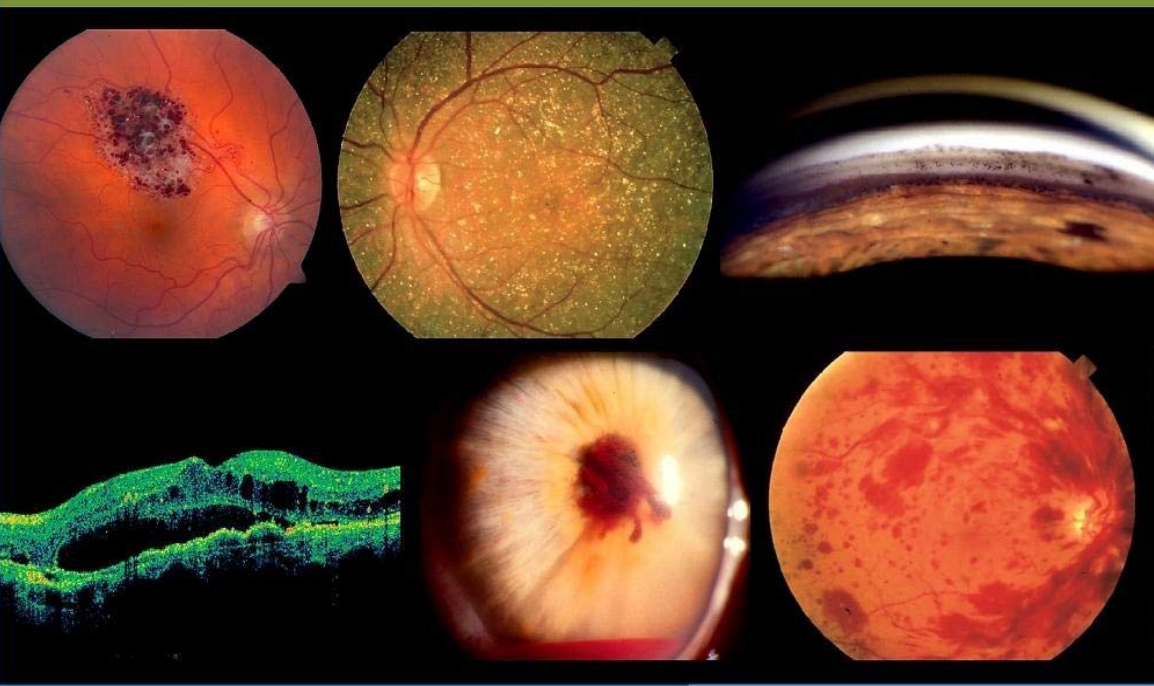
THE MASSACHUSETTS EYE AND EAR INFIRMARY
ILLUSTRATED MANUAL OF

OPHTHALMOLOGY

NEIL J. FRIEDMAN | PETER K. KAISER
ROBERTO PINEDA II



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Videos are courtesy of Y. Joyce Liao, MD, PhD, with special thanks to Dr. Thomas Hwang, Angela Oh, and Ali Shariati for their assistance.

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Preface

We started this process more than two decades ago when the first edition was published, and now we are excited to share the fifth edition of this book with you. Our goal remains the same: to produce a concise manual that covers a broad variety of ophthalmic disorders and present it in a user-friendly diagnostic atlas. With each update, we strive to improve on the previous version. We believe that this edition continues to set the bar higher.

We have expanded many chapters by adding new sections and figures, we have completely revised various sections, and we have updated numerous evaluation and management algorithms to incorporate the most up-to-date diagnostic and treatment options. Current residents, fellows, and attending physicians have reviewed and contributed to the book to ensure that the text is relevant to all ophthalmologists. The new figures include more clinical photos and images of various tests (i.e., fluorescein angiography, spectral domain optical coherence tomography, fundus autofluorescence, and visual fields) as well as updating some of the older images.

In addition, the fifth edition contains newer classification systems for various entities and updated epidemiology information. In the companion online material, we have added more videos of motility disturbances.

We believe that the new and improved fifth edition retains its previous attributes and incorporates important updates to keep pace with all the recent changes in our specialty. We hope you enjoy it.

Neil J. Friedman, MD
Peter K. Kaiser, MD
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We are most appreciative of the contribution of the following colleagues who helped review and edit various chapters of this text:

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We especially acknowledge our editorial and publishing staff at Elsevier—Russell Gabbedy, Kayla Wolfe, Nani Clansey, Radjan Selvanadin, and the members of their department—for their expertise and assistance in once again producing a work that we are excited to share with you.

In addition, we owe a debt of gratitude to Tami Fecko, Nicole Brugnoni, Anne Pinter, Shawn Perry, Louise Carr-Holden, Ditte Hesse, Kit Johnson, Bob Masini, Audrey Melacan, Jim Shigley, Huynh Van, and their photography departments for the wonderful images that truly set this book apart from other texts. We also appreciate the contributions of the numerous physicians whose photographs complete the vast array of ophthalmic disorders represented herein.

Finally, we acknowledge our families, including Mae, Jake, Alan, Diane, Peter (PJ), Stephanie, Dawn, Peter, Anafu, Christine, Roberto, Anne, Gabriela, and Nicole, for all their love and support.

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Introduction

Given the visual nature of ophthalmology, an illustrated manual is of utmost importance to our specialty. Continuing in the tradition of excellence forged by Drs. Friedman, Kaiser, and Pineda in the first four editions, the fifth edition of the *MEEI Illustrated Manual of Ophthalmology* continues to provide us with an accessible and portable yet comprehensive compendium that optimizes its availability for use by the practitioner.

Reflecting recent advances in ophthalmic care and understanding, the fifth edition contains a number of new diagnoses, new photos, updated treatment regimens, and major updates to many of the sections within the manual. New sections in this edition cover infectious uveitis, limbal stem cell deficiency, graft-versus-host disease, neurotrophic keratitis, hereditary color blindness, and refractive surgery procedures. The fifth edition continues to highlight the importance of our most sophisticated imaging technologies with optical coherence tomography angiography, wide-field angiography, and wide-field fundus photography. It serves as both a valuable teaching tool and a standard reference for practicing ophthalmologists.

The authors—all of whom trained at Harvard Medical School as students, residents, or fellows—continue to embody the best of the clinician–teacher paradigm. While advances in research (basic, translational, and clinical) drive our practice forward, it is a critical to share this knowledge to successive generations of ophthalmologists. We are especially grateful for their commitment, wisdom, and diligence in updating this classic text.

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1 Orbit

Trauma

Blunt Trauma

Orbital Contusion

Periocular bruising caused by blunt trauma; often with injury to the globe, paranasal sinuses, and bony socket; traumatic optic neuropathy or orbital hemorrhage may be present. Patients report pain and may have decreased vision. Signs include eyelid edema and ecchymosis, as well as ptosis. Isolated contusion is a preseptal (eyelid) injury and typically resolves without sequelae. Traumatic ptosis secondary to levator muscle contusion may take up to 3 months to resolve; most oculoplastic surgeons observe for 6 months before surgical repair.

Fig 1.1 • Orbital contusion demonstrating severe eyelid ecchymosis and edema, subconjunctival hemorrhage, and conjunctival chemosis.

- In the absence of orbital signs (afferent pupillary defect, visual field defect, limited extraocular motility, and proptosis), imaging studies are not necessarily required but should be considered with more serious mechanisms of injury (e.g., motor vehicle accident [MVA], massive trauma, or loss of consciousness) even in the absence of orbital signs. When indicated, orbital computed tomography (CT) scan is the imaging study of choice.

- When the globe is intact and vision is unaffected, ice compresses can be used every hour for 20 minutes during the first 48 hours to decrease swelling.
- Concomitant injuries should be treated accordingly.

Orbital Hemorrhage and Orbital Compartment Syndrome

Accumulation of blood throughout the intraorbital tissues caused by surgery or trauma (retrobulbar hemorrhage) may cause proptosis, distortion of the globe, and optic nerve stretching and compression (orbital compartment syndrome). Patients may report pain and decreased vision. Signs include bullous, subconjunctival hemorrhage, tense orbit, proptosis, resistance to retropulsion of globe, limitation of ocular movements, lid ecchymosis, and increased intraocular pressure (IOP). Immediate recognition and treatment are critical in determining outcome. Urgent treatment measures may include canthotomy and cantholysis. Evacuation of focal hematomas or bony decompression is reserved for the most severe cases with an associated optic neuropathy.

Fig 1.2 • Retrobulbar hemorrhage of the left eye demonstrating proptosis, lid swelling, chemosis, and restricted extraocular motility on upgaze.

OPHTHALMIC EMERGENCY

- If orbital compartment syndrome is suspected, lateral canthotomy and cantholysis should be performed emergently.
- **Lateral canthotomy:** This procedure is performed by compressing the lateral canthus with a hemostat, and Stevens scissors are then used to make a full-thickness incision from the lateral commissure (lateral angle of the eyelids) posterolaterally to the lateral orbital rim. Some advocate compression of the lateral canthal tendon before incision. The inferior crus of the lateral canthal tendon is then transected by elevating the lateral lower lid margin away from the face, placing the scissors between the cut edges of lower lid conjunctiva and lower lid skin, palpating the tendon with the tips of the scissors, and transecting it. If the inferior eyelid is not extremely mobile, the inferior crus has not been transected adequately, and the procedure should be repeated. If the IOP remains elevated and the orbit remains tense, the superior

OPHTHALMIC EMERGENCY—cont'd

crus of the lateral canthal tendon may be cut. Septolysis, blunt dissection through the orbital septum at the base of the cantholysis incision, may be performed when pressure is not adequately relieved with lysis of the canthal tendon.

- Emergent inferior orbital floor fracture, although advocated by some, is fraught with complications and is not advised for surgeons with little experience in orbital surgery; however, it should be considered in emergent situations with risk of blindness.
- Canthoplasty can be scheduled electively ~1 week after the hemorrhage.
- Orbital CT scan (without contrast, direct coronal and axial views, 3 mm slices) after visual status has been determined and emergent treatment (if necessary) administered (i.e., after canthotomy and cantholysis). Magnetic resonance imaging (MRI) is contraindicated in acute trauma.
- If vision is stable and IOP is elevated (>25 mm Hg), topical hypotensive agents may be administered (brimonidine 0.15% [Alphagan P] 1 gtt tid, timolol 0.5% 1 gtt bid, and/or dorzolamide 2% [Trusopt] 1 gtt tid).

Orbital Fractures

Fracture of the orbital walls may occur in isolation (e.g., blow-out fracture) or with displaced or nondisplaced orbital-rim fractures. There may be concomitant ocular, optic nerve, maxillary, mandibular, or intracranial injuries.

Orbital floor (blow-out) fracture

This is the most common orbital fracture requiring repair and usually involves the maxillary bone in the posterior medial floor (weakest point) and may extend laterally to the infraorbital canal. Orbital contents may prolapse or become entrapped in maxillary sinus. Signs and symptoms include diplopia on upgaze (anterior fracture) or downgaze (posterior fracture), enophthalmos, globe ptosis, and infraorbital nerve hypesthesia. Orbital and lid emphysema is often present and may become extensive with nose blowing.

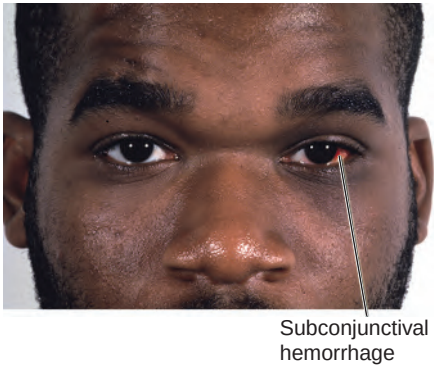

Fig 1.3 • Orbital floor blow-out fracture with enophthalmos and globe dystopia and ptosis of the left eye.

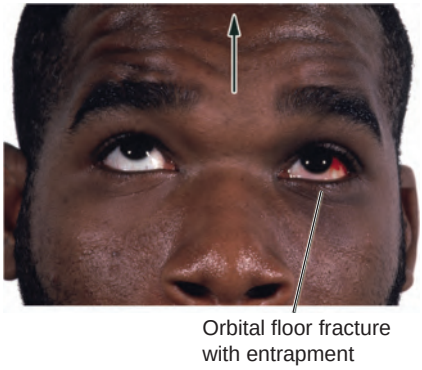

Fig 1.4 • Same patient as in Fig. 1.3 demonstrating entrapment of the left inferior rectus and inability to look up.

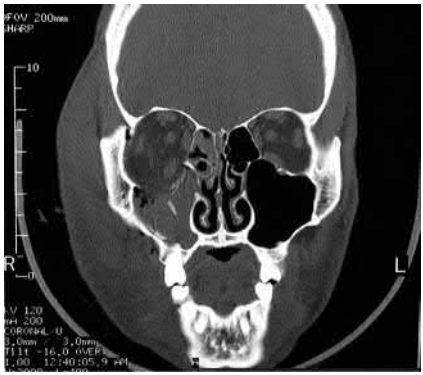

Fig 1.5 • Orbital computed tomography scan demonstrating large right orbital floor fracture.

- For mild trauma, orbital CT scan need not be obtained in the absence of orbital signs.
- Orbital surgery consultation should be considered, especially in the setting of diplopia, large floor fractures (>50% of orbital floor surface area), trismus, facial asymmetry, inferior rectus entrapment, and enophthalmos. Consider surgical repair after 1 week to allow for reduction of swelling except in cases of pediatric trapdoor-type fractures with extraocular muscle entrapment, in which emergent repair is advocated.

Pediatric floor fracture

This differs significantly from an adult fracture because the bones are pliable rather than brittle. A "trapdoor" phenomenon is created in which the inferior rectus muscle or perimuscular tissue can be entrapped in the fracture site. In this case, enophthalmos is unlikely, but ocular motility is limited dramatically. The globe halts abruptly with ductions opposite the entrapped muscle (most often on upgaze with inferior rectus involvement) as if it is "tethered." Forced ductions are positive; nausea and bradycardia (oculocardiac reflex) are common. Despite the severity of the underlying injury, the eye is typically quiet, hence the nickname "white eyed blowout fracture."

- Urgent surgery (<24 hours) is indicated in pediatric cases with entrapment.

Medial wall (nasoethmoidal) fracture

This involves the lacrimal and ethmoid (lamina papyracea) bones. It is occasionally associated with depressed nasal fracture, traumatic telecanthus (in severe cases), and orbital floor fracture. Complications include nasolacrimal duct injury, severe epistaxis caused by anterior ethmoidal artery damage, and orbit and lid emphysema. Medial rectus entrapment is rare, and enophthalmos caused by isolated medial wall fractures is extremely uncommon.

- Fractures extending through the nasolacrimal duct should be reduced with stenting of the drainage system. If not repaired primarily, persistent obstruction requiring dacryocystorhinostomy may result.
- Otolaryngology consultation is indicated in the presence of nasal fractures.

Orbital roof fracture

This is an uncommon fracture usually secondary to blunt or projectile injuries. It may involve the frontal sinus, cribriform plate, and brain. It may be associated with cerebrospinal fluid (CSF) rhinorrhea or pneumocephalus.

- Neurosurgery and otolaryngology consultations are advised, especially in the presence of CSF rhinorrhea or pneumocephalus.

Orbital apex fracture

This may be associated with other facial fractures and involve optic canal and superior orbital fissure. Direct traumatic optic neuropathy is likely. Complications include carotid–cavernous fistula and fragments impinging on optic nerve. These are difficult to manage because of proximity of multiple cranial nerves and vessels.

- Obvious impingement by a displaced fracture on the optic nerve may require immediate surgical intervention by an oculoplastic surgeon or neurosurgeon. High-dose systemic steroids may be given for traumatic optic neuropathy (see Chapter 11).

Tripod fracture

This involves three fracture sites: the inferior orbital rim (maxilla), lateral orbital rim (often at the zygomaticofrontal suture), and zygomatic arch. The fracture invariably extends through the orbit floor. Patients may report pain, tenderness, binocular diplopia, and trismus (pain on opening mouth or chewing). Signs include orbital rim discontinuity or palpable "step off," malar flattening, enophthalmos, infraorbital nerve hypesthesia, emphysema (orbital, conjunctival, or lid), limitation of ocular movements, epistaxis, rhinorrhea, ecchymosis, and ptosis. Enophthalmos may not be appreciated on exophthalmometry caused by a retrodisplaced lateral orbital rim.

Le Fort fractures

These are severe maxillary fractures with the common feature of extension through the pterygoid plates:

Le Fort I: low transverse maxillary bone; no orbital involvement

Le Fort II: nasal, lacrimal, and maxillary bones (medial orbital wall), as well as bones of the orbital floor and rim; may involve the nasolacrimal duct

Le Fort III: extends through the medial wall; traverses the orbital floor and through the lateral wall (craniofacial dysjunction); may involve the optic canal

- Orbital CT scan (without contrast, direct axial and coronal views, 3 mm slices) is indicated in the presence of orbital signs (afferent papillary defect, diplopia, limited extraocular motility, proptosis, and enophthalmos) or ominous mechanism of injury (e.g., MVA, massive facial trauma). MRI is of limited usefulness in the evaluation of fractures because bones appear dark.
- Otolaryngology consultation is indicated in the presence of mandibular fracture.
- Orbital surgery consultation is indicated in the presence of isolated orbital and trimalar fractures.
- Instruct the patient to avoid blowing the nose. A "suck-and-spit" technique should be used to clear nasal secretions.
- Nasal decongestant (oxymetazoline hydrochloride [Afrin nasal spray] bid as needed for 3 days. *Note:* This may cause urinary retention in men with prostatic hypertrophy).

- Ice compresses for the first 48 hours
- Systemic oral antibiotics (amoxicillin–clavulanate [Augmentin] 250–500 mg po tid for 10 days) are advocated by some.
- Nondisplaced zygomatic fractures may become displaced after initial evaluation because of masseter and temporalis contraction. Orbital or otolaryngology consultation is indicated for evaluation of such patients.

Penetrating Trauma

These may result from either a projectile (e.g., pellet gun) or stab (e.g., knife, tree branch) injury. A foreign body should be suspected even in the absence of significant external wounds.

Intraorbital Foreign Body

Retained orbital foreign body with or without associated ocular and optic nerve involvement. Inert foreign body (e.g., glass, lead, BB, plastics) may be well tolerated and should be evaluated by an oculoplastic surgeon in a controlled setting. Organic matter carries significant risk of infection and should be removed surgically. A long-standing iron foreign body can produce iron toxicity (siderosis), including retinopathy.

Patients may be asymptomatic or may report pain or decreased vision. Critical signs include eyelid or conjunctival laceration. Other signs may include ecchymosis, lid edema and erythema, conjunctival hemorrhage or chemosis, proptosis, limitation of ocular movements, and chorioretinitis sclopetaria (see Fig. 10.9). A relative afferent pupillary defect (RAPD) may be present. The prognosis is generally good if the globe and optic nerve are not affected.

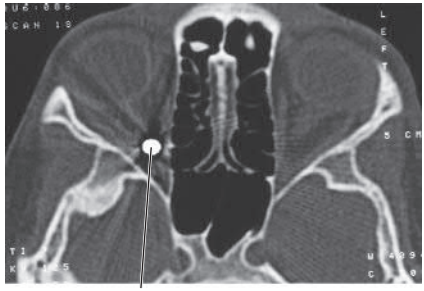

Fig 1.6 • Orbital computed tomography scan demonstrating a foreign body at the orbital apex.

- Precise history (may be necessary to isolate a minor child from the parents while obtaining history) is critical in determining the nature of any potential foreign body.
- **Lab tests:** Culture entry wound for bacteria and fungus. Serum lead levels should be monitored in patients with a retained lead foreign body.
- Orbital CT scan (without contrast, direct coronal and axial views). The best protocol is to obtain thin-section axial CT scans (0.625–1.25 mm, depending on the capabilities of the scanner) and then to perform multiplanar reformation to determine character and position of foreign body. MRI is *contraindicated* if the foreign body is metallic.
- If there is no ocular or optic nerve injury, small inert foreign bodies posterior to the equator of the globe usually are not removed but observed.
- Patients are placed on systemic oral antibiotic (amoxicillin–clavulanate [Augmentin] 500 mg po tid for 10 days) and are followed up the next day.

- Tetanus booster (tetanus toxoid 0.5 mL IM) if necessary for prophylaxis (>7 years since last tetanus shot or if status is unknown).
- Indications for surgical removal include fistula formation, infection, optic nerve compression, large foreign body, or easily removable foreign body (usually anterior to the equator of the globe). Surgery should be performed by an oculoplastic surgeon. Organic material should be removed more urgently.

Globe Subluxation

Definition Spontaneous forward displacement of the eye so that the equator of the globe protrudes in front of the eyelids, which retract behind the eye.

Etiology Most often spontaneous in patients with proptosis (e.g., Graves' disease) but may be voluntary or traumatic.

Mechanism Pressure against the globe, typically from spreading the eyelids, causes the eye to move forward, and then when a blink occurs, the eyelids contract behind the eye, locking the globe in a subluxed position.

Epidemiology Occurs in individuals of any age (range, 11 months to 73 years) and has no sex or race predilection. Risk factors include eyelid manipulation, exophthalmos, severe eyelid retraction, floppy eyelid syndrome, thyroid eye disease (TED), and shallow orbits (i.e., Crouzon's or Apert's syndrome).

Symptoms Asymptomatic; may have pain, blurred vision, and anxiety.

Signs Dramatic proptosis of the eye beyond the eyelids. Depending on the length of time the globe has been subluxed, may have exposure keratopathy, corneal abrasions, blepharospasm, and optic neuropathy.

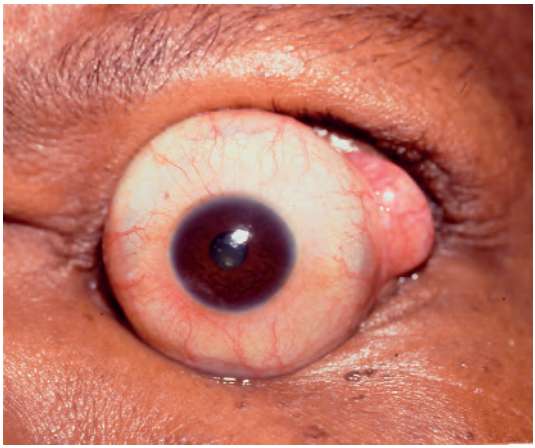

Fig 1.7 • Globe subluxation with equator of globe and lacrimal gland in front of the eyelids.

Evaluation

- Complete ophthalmic history with attention to previous episodes and potential triggers.
- Complete eye exam (after the eye has been repositioned) with attention to visual acuity, pupils, motility, exophthalmometry, lids, cornea, and ophthalmoscopy.

Management

- Immediately reposition the globe. Relax the patient, instill topical anesthetic, and digitally reduce the subluxation by one of the following methods:
  1. While the patient looks down, pull the upper eyelid up and depress the globe.
  2. Place a retractor under the center of the upper eyelid, push the globe downward, and advance the eyelid forward. When the eyelid is past the equator of the globe, have the patient look up to pull the eyelid over the eye.
- May require a facial nerve block, sedation, or general anesthesia.
- Instruct the patient to avoid triggers and how to reduce a subluxation.
- Treat any underlying condition.
- Surgical options include partial tarsorrhaphy and orbital decompression.

Prognosis Good unless complications develop.

Carotid–Cavernous and Dural Sinus Fistulas

Definition
Arterial venous connection between the carotid artery and cavernous sinus; there are two types.

High-Flow Fistula

Between the cavernous sinus and internal carotid artery (carotid–cavernous fistula).

Low-Flow Fistula

Between small meningeal arterial branches and the dural walls of the cavernous sinus (dural sinus fistula).

Etiology

High-Flow Fistula

Spontaneous; occurs in patients with atherosclerosis and hypertension with carotid aneurysms that rupture within the sinus or secondary to closed-head trauma (basal skull fracture).

Low-Flow Fistula

Slower onset compared with the carotid–cavernous variant; dural sinus fistula is more likely to present spontaneously.

Symptoms May hear a "swishing" noise (venous souffle); may have a red "bulging" eye.

Signs

High-Flow Fistula

May have orbital bruit, pulsating proptosis, chemosis, epibulbar injection and vascular tortuosity (conjunctival corkscrew vessels), congested retinal vessels, and increased IOP.

Low-Flow Fistula

Mild proptosis and orbital congestion. However, in more severe cases, findings are similar to those described for carotid–cavernous fistula may occur.

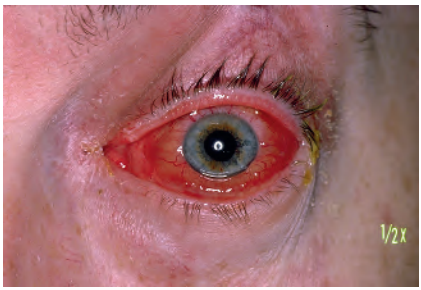

Fig 1.8 • Carotid–cavernous fistula with conjunctival injection and chemosis.

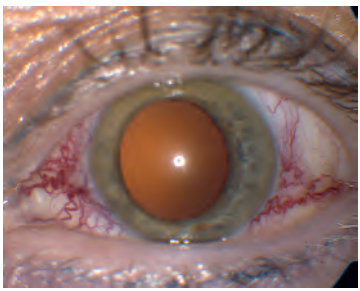

Fig 1.9 • Carotid–cavernous fistula with dilated, corkscrew, episcleral, and conjunctival vessels.

Differential Diagnosis Orbital varices that expand in a dependent position or during Valsalva maneuvers and may produce hemorrhage with minimal trauma. Carotid–cavernous fistula may also be mistaken for orbital inflammatory syndrome and occasionally uveitis.

Evaluation

- Complete history with attention to onset and duration of symptoms and history of trauma and systemic diseases (atherosclerosis, hypertension).
- Complete eye exam with attention to orbital auscultation, exophthalmometry, conjunctiva, tonometry, and ophthalmoscopy.
- **Orbital CT scan or MRI:** Enlargement of the superior ophthalmic vein.
- Arteriography usually is required to identify the fistula; CT angiography and magnetic resonance angiography have largely replaced conventional angiography.

Management

- Consider treatment with selective embolization or ligation for severely symptomatic patients (uncontrolled increase in IOP, severe proptosis, retinal ischemia, optic neuropathy, severe bruit, involvement of the cortical veins).
- Treatment for all cases of carotid–cavernous fistula has been advocated but is controversial.

Prognosis Up to 70% of dural sinus fistulas may resolve spontaneously.

Infections

Preseptal Cellulitis

Definition Infection of the eyelids not extending posterior to the orbital septum. The globe and orbit are not involved.

Etiology Usually follows periorbital trauma or dermal infection. Suspect *Staphylococcus aureus* in traumatic cases and *Haemophilus influenzae* (nontypeable) in children <5 years old.

Symptoms Eyelid swelling, redness, ptosis, and pain; low-grade fever.

Signs Eyelid erythema, edema, ptosis, and warmth (may be quite dramatic); visual acuity is normal; full ocular motility without pain; no proptosis; the conjunctiva and sclera appear uninflamed; an inconspicuous lid wound may be visible; an abscess may be present.

Differential Diagnosis Orbital cellulitis, idiopathic orbital inflammation (IOI), dacryoadenitis, dacryocystitis, conjunctivitis, and trauma.

Fig 1.10 • Mild preseptal cellulitis with right eyelid erythema in a young child.

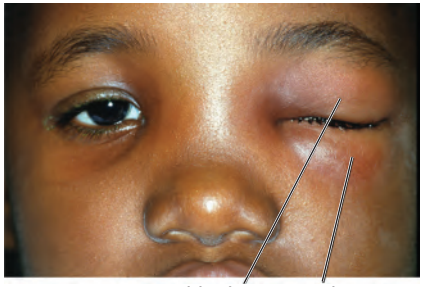

Fig 1.11 • Moderate preseptal cellulitis with left eyelid edema and erythema.

Evaluation

- Complete ophthalmic history with attention to trauma, sinus disease, recent dental work or infections, history of diabetes or immunosuppression.
- Complete eye exam with attention to visual acuity, color vision, pupils, motility, exophthalmometry, lids, conjunctiva, and sclera.
- Check vital signs, head and neck lymph nodes, meningeal signs (nuchal rigidity), and sensorium.
- **Lab tests:** Complete blood count (CBC) with differential, blood cultures; wound culture if present.
- Orbital and sinus CT scan in the absence of trauma or in the presence of orbital signs to look for orbital extension and paranasal sinus opacification.