Orbital Compartment Syndrome

Orbital compartment syndrome (OCS) is an emergent condition that occurs when intraorbital pressure exceeds ophthalmic artery pressure, causing compression and ischemic compromise to the retina and optic nerve. Patients of any age or demographic can be affected. Early recognition of OCS is crucial because the condition can lead to permanent vision loss if not treated promptly. Appropriate management involves acute-care surgical maneuvers to decompress the orbit.

Causes. A common cause of OCS is retrobulbar hematoma after blunt trauma or surgery, such as sinus or orbital procedures.[1-3] Notably, the incidence of retrobulbar hemorrhage after administration of peribulbar and retrobulbar anesthesia ranges from 0.44% to 2%, but only a small fraction of these cases progress to OCS.[4] OCS can also result from an array of space-occupying processes in or posterior to the orbit, some of which are rare. When the cause is not obvious, a thorough workup is needed to identify and address the underlying process.[1]

Potentially fatal. OCS also can have systemic implications, so failure to treat it or make appropriate referrals can lead to additional morbidity or even death. It is crucial that ophthalmologists be familiar with the various causes and treatments for this vision-threatening condition.

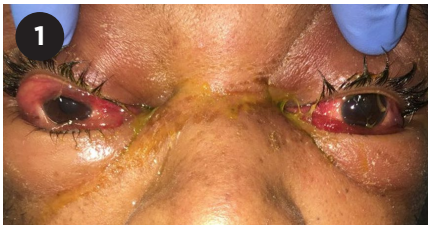
Anatomy and Pathophysiology

The orbits are pear-shaped structures, each with a volume of approximately 30 mL; they are bound by bone on all sides except anteriorly, where the border is formed by the orbital septum and eyelids. The medial and lateral canthal tendons attach the eyelids to orbital bones. The orbital apex is the entry and exit point for important neurovascular structures. Any rapid volumetric increase within the orbit may elevate intraorbital pressure, which can cause ischemia to orbital contents.

Compression can be blinding. Compression of the central retinal artery leads to retinal ischemia—a cause of irreversible vision loss. For example, in a study of rhesus monkeys, investigators observed irreparable loss of retinal function after clamping the central retinal artery for 105 minutes.[5] Another major cause of permanent vision loss in OCS is ischemic optic neuropathy, which can happen with compression of the posterior ciliary arteries. Also, stretch optic neuropathy may result from severe proptosis. Additional morbidity may result from ischemia to the extraocular muscles or to the anterior segment, where neovascular glaucoma may develop.

The Cause Is Not Always Clear

Although diagnosing OCS may be straightforward, it is important to determine the underlying cause.



BILATERAL OCS. *OCS due to septic cavernous sinus thromboses.*

Trauma or surgery. In the setting of trauma or surgery, the most likely etiology is retrobulbar hemorrhage. Other causes to consider are carotid-cavernous fistula; venous outflow obstruction; and orbital emphysema, which can result from nose blowing, when air egresses from the sinuses and enters the orbit via breaks in the orbital walls.

Hypercoagulability. If the patient is hypercoagulable, then infectious and neoplastic causes should be explored. Signs of an infectious etiology may include systemic inflammatory response syndrome (SIRS) criteria; cardinal signs of inflammation on exam; inflammatory marker elevation on labs; and evidence of sinusitis or abscess on imaging. It is helpful to note that orbital congestion may be difficult to distinguish from cellulitis on CT imaging, so it is important to take the full clinical picture into account.

"Spontaneous" cases. Seemingly spontaneous cases of OCS have occurred in the settings of malignancy, severe burn, cirrhosis, and hemophilia.[6] In severely burned patients who are resuscitated by high-volume intravenous fluid, OCS may occur from fluid extravasation due to loss of intravascu-

BY **ALEXANDER ENGELMANN, MD,** AND **KATHERINE DUNCAN, MD.** EDITED BY JEREMIAH TAO, MD, FACS.

Alexander Engelmann, MD

lar oncotic pressure.

External compression. External compression during sedation may cause OCS and should be considered in cases that occur following a prone procedure. OCS may also be seen in patients who present with an entity termed Saturday night retinopathy, which may develop after a heavily intoxicated patient has passed out in a facedown position.

Presentation and Diagnosis

Patients with OCS are commonly obtunded or unconscious, especially after major blunt-force trauma to the head.

Symptoms. However, if they are alert and able to provide history, their symptoms may include orbital pain, swelling, vision loss, double vision, or nausea.

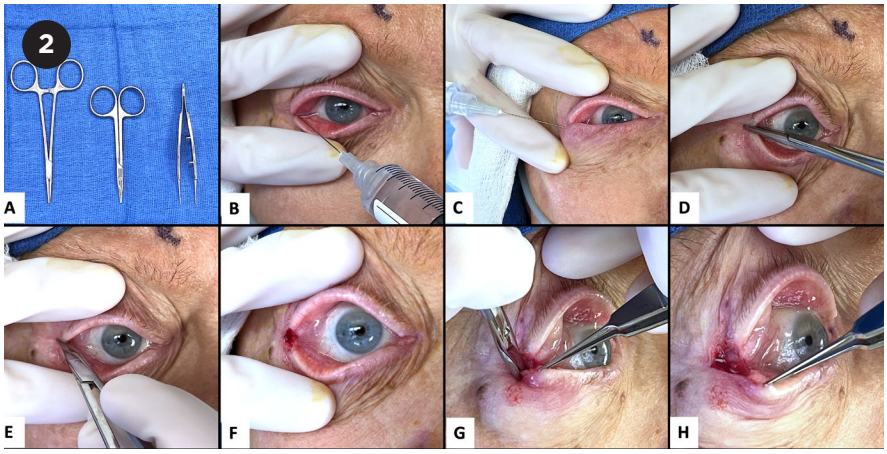
Signs. Clinical signs include a relative afferent pupillary defect, axial proptosis, tense periorbital edema, eyelid ecchymosis, resistance to retropulsion of the globe, restriction of extraocular movements, internal ophthalmoplegia, conjunctival chemosis, hemorrhage, and injection (Fig. 1), and a firm globe due to elevated intraocular pressure (IOP).[1] Hence, IOP can serve as a proxy for intraorbital pressure when orbital pressure exceeds normal IOP.

Diagnosis. The diagnosis of OCS is established clinically, based on findings indicative of optic neuropathy and intraorbital pressure elevation.

Testing

After emergency surgical decompression, ancillary testing should be performed, guided by the patient's history. The initial lab workup should include a complete blood count, metabolic panel, and coagulation testing.

Imaging. Initial neuroimaging may include CT, with contrast arteriography of the head and neck if hemorrhage or fistula is a concern. CT with venography and attention to cavernous sinus and orbital apex should be conducted if cavernous sinus or superior ophthalmic vein thrombosis is a possibility. The findings of subsequent MRI and other lab studies may improve diagnostic specificity. (See MRI images with this article at aao.org/eyenet.)



LATERAL CANTHOTOMY AND INFERIOR CANTHOLYSIS STEPS. *Demonstrated on a patient with excessive horizontal eyelid laxity. (2A) Instruments: hemostat, blunt-tipped scissors, toothed microforceps. (2B-C) Administer local anesthetic. Inject 2% lidocaine with epinephrine into the lateral canthal angle and the lower eyelid using a 27-gauge needle. (2D) Clamp with hemostat to mark and aide hemostasis. (2E-F) Use scissors to make a full-thickness cut from the lateral canthal angle to the lateral orbital rim. This is the only incision that should involve the skin. (2G) Use toothed microforceps to distract the lower lid away from the globe and identify the inferior crus of the lateral canthal tendon by "strumming" with blunt scissor-tips. Incise the lateral canthal tendon. (2H) Lower eyelid is freely mobile. If necessary, repeat step "G" and perform cantholysis of the superior crus, taking care to avoid damaging the lacrimal gland.*

Management and Prognosis

In acute OCS with compromised vision, a lateral canthotomy with cantholysis (Fig. 2) should be performed emergently, usually at bedside. If the cause of OCS is an abscess or emphysema, orbitotomy may be indicated to evacuate pus or air. In such cases, needle decompression has been used. If the OCS is recalcitrant to canthotomy and cantholysis, it may be necessary to conduct open orbitotomy as well as bony orbital decompression.

Early treatment is best. The prognosis of OCS depends on the timing of treatment; better outcomes are achieved if treatment is rendered within 90 minutes of OCS onset.[1] However, in cases when treatment was delayed by up to several days, VA improvement has occurred.[7] So, even if patients do not present immediately, they should still be offered canthotomy, cantholysis, or another form of surgical decompression. The risks associated with canthotomy and cantholysis include damage to the eyelids, lacrimal gland, and globe. Notably, the lateral canthus is very amenable to later reconstruction.

Address the underlying cause. Once the immediate threat to vision has been eliminated by orbital decompression, the underlying etiology must be addressed. If an infectious source is suspected, broad-spectrum antibiotics should be initiated.[1] If a cavernous sinus or superior ophthalmic vein thrombosis is present, anticoagulation may be indicated. If a noninfectious inflammatory process is present, corticosteroids may be considered.

Further care. IOP should be rechecked within eight hours of initial presentation. And bedside nursing is advised to monitor for signs and symptoms of increased intraorbital pressure. Other care measures include controlling pain and blood pressure; minimizing Valsalva maneuver with antiemetics, decongestants, or stool softeners; elevating the head of the bed; and deep extubation (if applicable).[3]

Medication considerations. Systemic diuretics like acetazolamide and mannitol may condense the vitreous, reducing the globe's volume within the orbit. However, the effect of this treatment is negligible and likely has no

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role in management of OCS. Similarly, IOP-lowering drops likely have no role, as ocular morbidity in OCS occurs due to a host of factors. Pursuing IOP control rather than source control may delay delivery of effective treatment.

Ongoing follow-up. After leaving the hospital, patients should follow up with their ophthalmologist to monitor for and manage development of long-term sequelae. For example, patients with significant ischemic insults should be followed monthly over the first six months for signs of neovascular glaucoma. Patients with lateral canthus deformities may be at risk for exposure keratopathy and should be treated with lubrication until reconstruction. And those with ptosis and strabismus should be referred appropriately. Monocular precautions and low vision therapy should be offered to those who fail to recover normal vision.

Key Messages

Permanent loss of vision can be prevented if OCS is recognized and treated immediately. Many patients can be managed successfully with bedside canthotomy and cantholysis, but some will require further decompression. In cases where the cause is not apparent, a medical workup is warranted to identify the cause and determine the treatment.

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7 Bailey LA et al. *Ophthalmic Plast Reconstr Surg.* 2019;35(6):586-589.

Dr. Engelmann is an ophthalmology resident at the University of Maryland in Baltimore. **Dr. Duncan** is an ophthalmologist and oculofacial plastic surgeon at the Greater Baltimore Medical Center. *Financial disclosures: None.*

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