# Sinking Into His Sockets

Ora Cornelius* is a 15-year-old boy who over the last five years had experienced increasing foreign body sensation, blurry vision, and diplopia, as well as constant tearing that failed to improve with lubricating eyedrops and ointment.

After noticing that Ora had bilateral asymmetric enophthalmos, his local ophthalmologist referred him to our oculoplastics clinic.

## We Get a Look

**History.** Ora's medical history is significant for a left middle cerebral artery (MCA) stroke in utero with resultant intractable epilepsy, which necessitated a left functional hemispherectomy. Postoperatively, he developed hydrocephalus requiring a ventriculoperitoneal (VP) shunt. A few weeks prior to his oculoplastic consultation, Ora had visited his neurosurgeon who had found no indication of shunt failure or over-drainage and therefore did not recommend shunt revision or removal.

When we spoke to Ora, he said that he felt self-conscious about the appearance of his eyes, especially since starting high school this past year. He also reported a history of sinus congestion, balance and gait disturbances, focal weakness, memory difficulties, and muscle weakness.

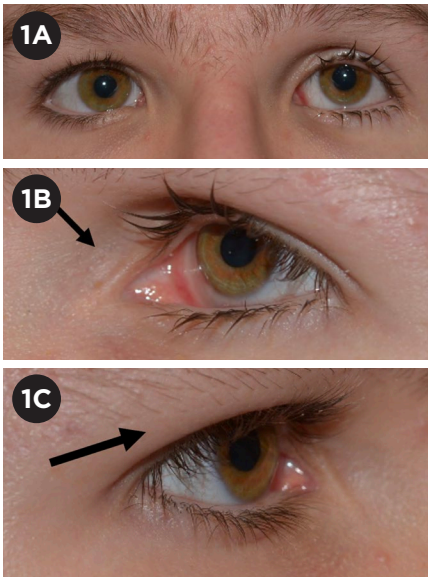
**Exam.** Ora's BCVA was 20/20 in both eyes. His pupils, IOP, and extraocular movements (EOMs) were all normal. The confrontation visual fields test revealed bilaterally constricted fields. Hertel exophthalmometry was 8 mm and 11 mm for the right and left eye, respectively. The external exam demonstrated a prominent glabella, poor eyelid-to-globe apposition bilaterally, epicanthus, a deep superior sulcus (with the left eye greater than the right), and bilateral, asymmetric ptosis (Figs. 1A-1C). The anterior segment exam was notable for meibomian gland dysfunction, scattered superficial punctate keratitis, and trace injection. The posterior segment exam was normal.

**Imaging and HVF testing.** We ordered a computed tomography (CT) maxillofacial scan, which revealed marked bilateral enophthalmos, upward bowing of the orbital roofs, air entrapment under the upper eyelids, and pseudoptosis of the upper eyelids (Fig. 2). Humphrey Visual Field (HVF) testing revealed congruous right homonymous hemianopia with macular sparing in each eye (Fig. 3; see "More Online"), consistent with his history of a left hemispherectomy. OCT of the retinal nerve fiber layer (RNFL) demonstrated bilateral optic nerve thinning consistent with his VF defects (Fig. 4; see "More Online").

## The Differential Diagnosis

When you examine a patient presenting with bilateral enophthalmos with ocular irritation, epiphora, diplopia, VF constriction, and poor eyelid-to-globe apposition, it is important to categorize the differential diagnoses into causes of pseudo-enophthalmos, orbital structural abnormalities, orbital soft tissue loss, or orbital retraction.

Ora's history of childhood VP shunting along with his external presentation and largely benign ocular exam indicated an orbital structural abnormality.




***WE GET A LOOK.*** *(1A) We noted a prominent glabella, epicanthus, deep superior sulcus more appreciable on the left eye, bilateral ptosis (with margin to reflex distance greater on the left eye), and more visible caruncle on the left side. In looking at his right (1B) and left (1C) eyes, we also noted poor eyelid-globe lid apposition (predominant feature), left more than right. Fig. 1C shows a deep sulcus with a heavy brow ridge.*

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His CT maxillofacial scan confirmed normal-sized globes with posterior displacement from the eyelids and excluded the presence of microphthalmos or any lesions on the globe, extraocular muscles, or orbital apex. His EOMs were full on exam, which allowed us to exclude conditions associated with extraocular muscle retraction that can cause enophthalmos, such as Duane retraction syndrome. We excluded silent sinus syndrome based on our patient's bilateral enophthalmos with a lack of sinus opacification, convex shape of the orbital floor, or relative shrinkage of the maxillary sinus on CT maxillofacial imaging.[1]

## Our Diagnosis

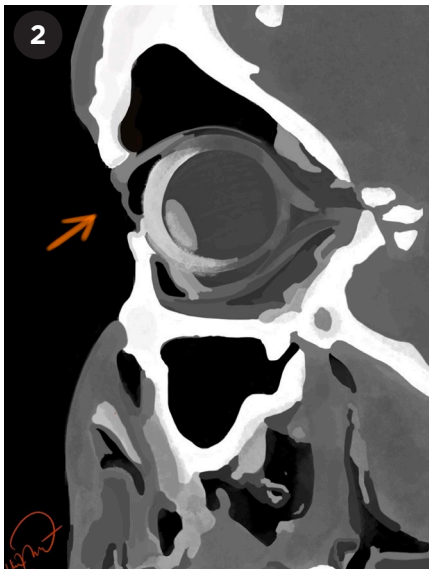
Based on the constellation of clinical findings in the setting of a childhood VP shunt, Ora was diagnosed with silent brain syndrome (SBS). This was supported by the bony remodeling with upward bowing of the orbital roof and clear sinus cavities on CT maxillofacial imaging. The diagnosis of SBS can be challenging due to its rarity; despite the prevalence of childhood VP shunts, the condition has only appeared in case reports.

## Discussion

**Epidemiology.** SBS is classically reported in patients who have a history of VP shunt placement for hydrocephalus at a young developmental age. Patients typically present in their 20s as sequelae typically develop at this age. No gender or racial predilection has been identified.[1]

**Pathophysiology.** Although the pathophysiology of SBS is not well defined, the leading theory is that the malleable nature of bone formation in children and young adults (up until about 22 years old) along with chronic decreased intracranial pressure (ICP), which is found in young patients with VP shunts, leads to the development of bilateral enophthalmos.

If a patient has excess outflow from their VP shunt prior to permanent bone development, the thin cranial bones of the orbital roof can remodel toward the area of least resistance, causing an upward bowing deformity.[2-4] The subsequent enlargement of the conical orbit allows the globe to sink backward, creating a progressive bilateral enophthalmos that may be asymmetric.




***IMAGING AND VHF TESTING.*** *CT maxillofacial illustrative interpretation from patient's original CT read demonstrates superior orbital roof bowing allowing large orbital space and posterior displacement of the eye.*

**Complications.** The most common complications from progressive enophthalmos can be subdivided into those associated with the anterior segment and those associated with the asymmetric globe position, though the negative impact on visual acuity is associated with both.

**Anterior segment complications** include epiphora and ocular surface disease due to poor eyelid-to-globe apposition. These patients are at an increased risk of corneal abrasion and ulceration.

**Complications due to the globe's sunken position** can include superior VF deficit from upper eyelid obstruction, nasal field deficit due to visual axis obstruction from the nose, and decreased depth perception. Cosmetic concerns about the altered appearance can cause a psychological burden, especially since the sequelae of SBS most commonly develop in teenagers and young adults.

**Treatment.** Nonsurgical treatment of SBS is directed at managing the corneal sequelae with aggressive lubrication. Surgical management is targeted at improving eyelid-to-globe apposition, globe symmetry, and cosmesis. The most common surgical approach involves orbital volume augmentation using an orbital floor or roof implant. Another treatment option is eyelid malposition repair, which involves repositioning the medial or medial and lateral canthal tendons and is considered after orbital augmentation.[1,5] If the patient still has strabismus after these surgeries, strabismus surgery may be necessary to further align globe position and movement.

## Follow-Up

To accommodate Ora's academic schedule, we plan to update orbital imaging in the next few months and then perform an orbital buildup of the right eye when school closes in the summer. If Ora is symptomatic after completing postoperative recovery, we will consider referring him to an adult strabismus specialist for further visual and cosmetic improvement.

*Patient name is fictitious.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**MORE ONLINE.** See this article at aao.org/eyenet for the initial Humphrey 24-2 VF testing (Fig. 3), which demonstrates congruous right homonymous hemianopia with macular sparing, and OCT (Fig. 4), which reveals diffuse thinning of the RNFL in both eyes.