Gross Enophthalmos After Cerebrospinal Fluid Shunting for Childhood Hydrocephalus: The "Silent Brain Syndrome"

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Purpose: To describe the clinical characteristics and ophthalmic management of 2 patients who developed gross enophthalmos after ventriculo-peritoneal shunting performed in their teenage years. A key radiologic feature is presented, and a conjectural mechanism is proposed for this disfiguring condition.

Methods: Retrospective case note review for 2 patients requiring ophthalmic care for gross enophthalmos after prior ventriculo-peritoneal shunting.

Results: Two patients, aged 24 and 25 years, presented with severe bilateral enophthalmos, bridging of all the tarsal plates off the ocular surface with secondary upper eyelid entropion, and significant lagophthalmos, associated with diffuse keratopathy. Both patients were of normal body weight, and neither had a history of anorexia nervosa. CT of the orbit revealed gross enophthalmos, with air entrapment between the globe and upper eyelids, together with a marked upward bowing of the orbital roof in the anterior cranial fossa, a newly recorded sign in this condition. One patient underwent bilateral orbital roof implants, and the other had bilateral upper eyelid entropion repair.

Conclusions: Progressive, severe, bilateral, symmetrical enophthalmos with bridging of the eyelids across the ocular surface due to upward bowing of the orbital roof many years after venticulo-peritoneal shunt in the absence of symptomatic intracranial disease are pathognomonic features of the "silent brain syndrome." A common feature was shunting in the early teenage years; although the enophthalmos had been noted for several years before presentation, the corneal symptoms had only become troublesome enough to seek ophthalmic care in their third decade, and the speed of development for this condition remains unclear. The authors suggest that a sudden reduction of raised intracranial pressure causes an "implosion" of the only available thin cranial bone—namely, the frontal plate of the orbit. Such remodeling might be greater if the bone was still relatively unmineralized, because of youth or preceding hydrocephalus. The expansion of orbital volume is responsible for the characteristic clinical features and symptoms and can be treated with placement of appropriately sized orbital roof implants or, if this is not desired, by upper eyelid entropion repair.

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Nontraumatic enophthalmos is generally due to 1 of 3 mechanisms: 1) orbital fat atrophy, as in cases of hemifacial atrophy or HIV-lipodystrophy; 2) severe fibrosis of orbital fat, as with metastatic scirrhous breast carcinoma; or 3) expansion of the orbit, as in "imploding antrum" or "silent sinus" syndrome; only rarely, however, are these conditions bilateral.[1–6] Both of our patients were free of any prior or concurrent systemic disease. Severe bilateral enophthalmos after ventriculo-peritoneal (VP) shunting was first reported in 3 patients without an explanation for this condition.[7] Together with description of a surgical correction for the condition, Cruz et al.[8] were the first to report the association of upward bowing of the orbital roof and prior shunting in one patient. We now describe the clinical features in 2 further cases and propose a conjectural mechanism for this disfiguring condition that we call "silent brain syndrome."

METHODS

The clinical notes and imaging were retrospectively reviewed for 2 patients with gross enophthalmos and persistent keratopathy after VP shunting.

Case 1. A 25-year-old white woman was referred to Moorfields Eye Hospital with a 3-year history of ocular irritation and watering, having undergone VP shunting for idiopathic congenital hydrocephalus at age 14 years. Her presenting visual acuity was 20/20 OU, with normal optic discs and intraocular pressure. Ocular motility was normal despite gross enophthalmos (hertel exophthalmometry 6 mm OU), and there was marked bridging of all 4 eyelids away from the ocular surface, lagophthalmos with 3 mm upper eyelid retraction, and trichiasis secondary to upper eyelid entropion (Fig. 1A); both corneas had severe upper-half erosive keratopathy, and there was inferior vascular pannus due to right exposure keratopathy. The lacrimal drainage system was normal. Orbital CT showed gross enophthalmos due to marked upward bowing of the orbital roof, with widespread air entrapment under the upper eyelid (Fig. 1B, C). The patient declined orbital implantation to address the severe enophthalmos but underwent upper eyelid entropion repair with moderate symptomatic improvement.

Case 2. A 24-year-old white man was referred with a 3-year history of ocular irritation and persistent eyelid rubbing, having been followed at the Gaslini Children's Hospital after VP shunting at the age of 12 years; shunting was required for intracranial hypertension after removal of a third ventricular tumor. Visual acuity, fundus examination, and ocular motility were normal, but gross bilateral enophthalmos with Hertel readings of approximately 0 mm was noticed. There was complete bridging of the upper tarsi away from the ocular surfaces, with secondary upper eyelid entropion, 4 mm of lagophthalmos, and severe bilateral erosive kerathopathy due to trichiasis (Fig. 2A). Severe en-

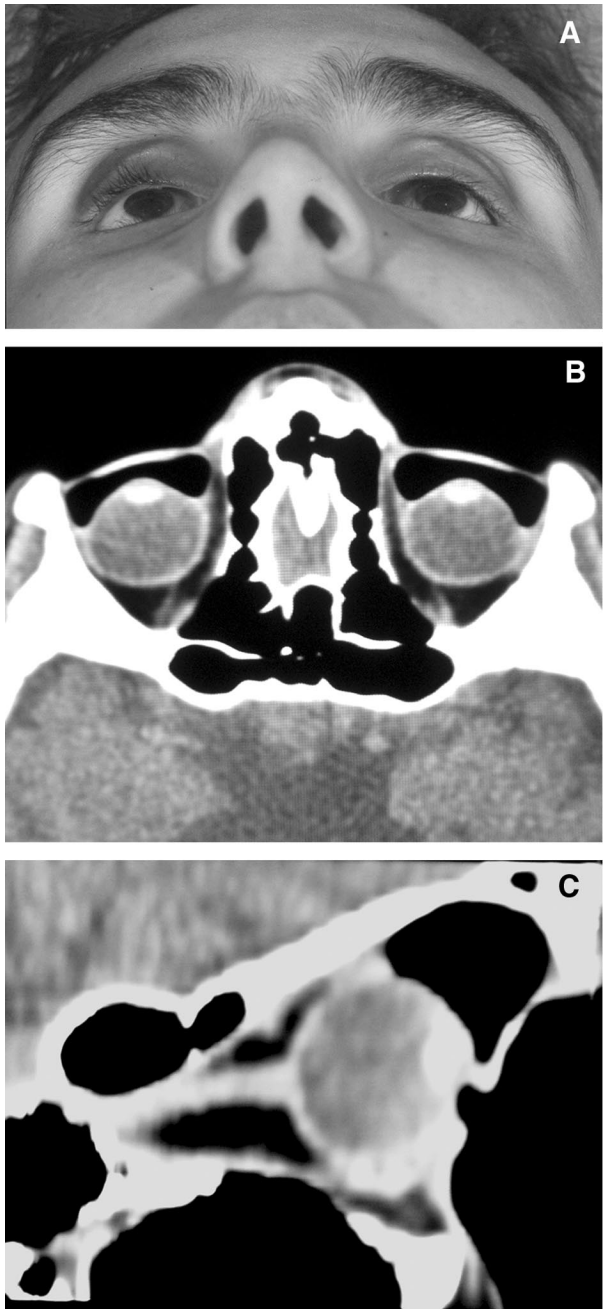

FIG. 1. Case 1. **A,** Severe, bilateral enophthalmos with bridging of all 4 eyelids away from the ocular surface. **B** and **C,** Orbital CT confirms gross enophthalmos due to a marked upward bowing of the orbital roof, rather than a loss of orbital fat; there is widespread air entrapment under the upper eyelids.

ophthalmos, with gross air entrapment under the upper eyelids was evident on orbital CT (Fig. 2B, C), and there was marked and symmetrical upward bowing of both orbital roofs. Under general anesthesia and avoiding damage to the supraorbital neurovascular bundles, the extraperiosteal plane was dissected widely through upper eyelid skincrease incisions, and multiple pieces of 0.4- or 0.8-mm–thick porous polythene sheet (Medpor; Porex, Inc., Fairburn, GA, U.S.A.) were shaped and stacked in the cavities to correct the contour defect of each orbital roof. The periosteum was closed along the orbital rims using interrupted 5-0 soluble sutures. There was an excellent reduction of

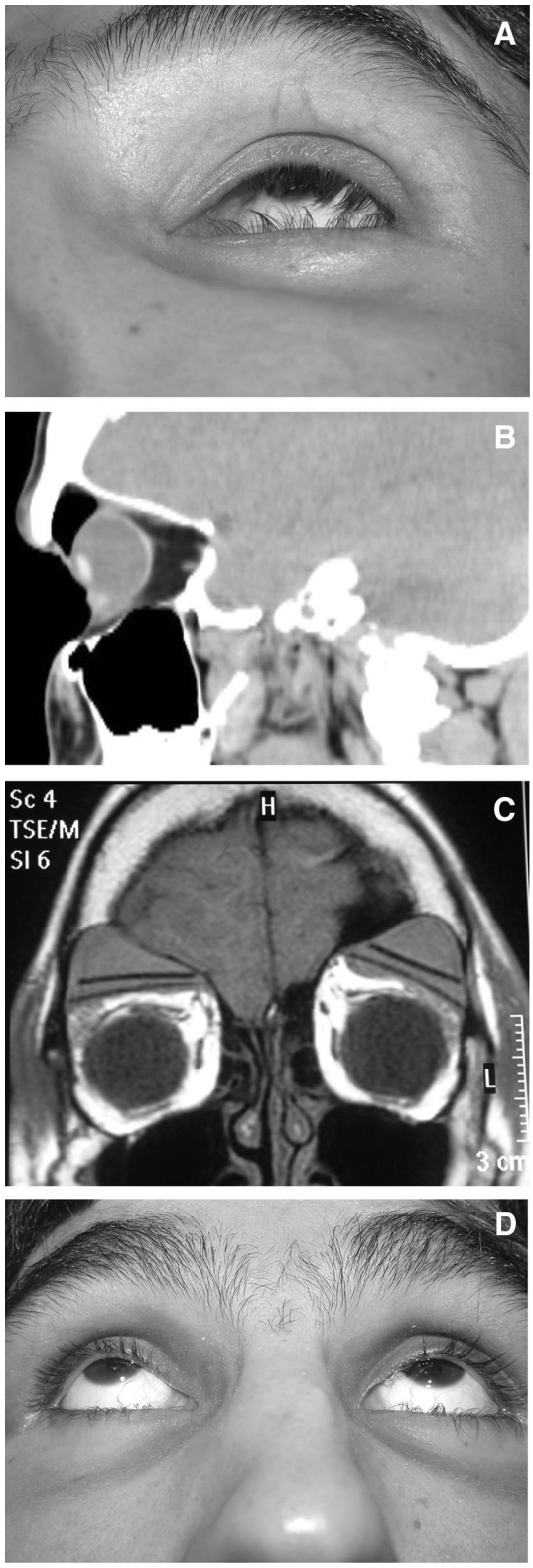

FIG. 2. Case 2. **A,** Severe, bilateral enophthalmos with bridging of all 4 eyelids away from the ocular surface. **B,** Gross enophthalmos on CT with upward bowing of the orbital roof and air entrapment under the upper eyelids. **C,** Orbital MRI showing the stacked Medpor plates on the orbital roofs bilaterally. **D,** Postoperative restoration of eyelid globe apposition and correction of enophthalmos.

Age characteristics for 6 patients developing gross bilateral enophthalmos after ventirculo-peritoneal shunting for childhood hydrocephalus

Patient	Age at CSF shunting (years)	Age at onset of ophthalmic symptoms (years)	Age at ophthalmic presentation (years)	Reference
1	14	22	25	Current series
2	12	21	24	Current series
3	8	32	33	8
4	0.5	Unreported	24	8
5	3	14	37	8
6	13	13	22	9
Series characteristics				
Mean	8.3	20.4	27.5	
Median	10	21	24.5	
Range	0.5–14	13–32	22–37	

Data derived from the current series and the English ophthalmic literature.

enophthalmos (to Hertel 12 mm; Fig. 2D), restoration of contact between the palpebral conjunctiva and globes, and a resolution of his symptoms.

DISCUSSION

There appear to be only 4 other reported cases of severe enophthalmos after VP shunting, these patients presenting with identical symptoms to our 2 new cases—namely, severe keratopathy due to lagophthalmos, upper entropion, and trichiasis due to a loss of apposition between the upper tarsi and the ocular surface.[7,8] Cranial bone growth in childhood is dependent on cerebrospinal fluid pressure, and its reduction with VP shunting can lead to various skull anomalies.[9–11] Cruz et al.[8] were the first to note that postshunting enophthalmos was associated with upward expansion of the orbital roof, rather than loss or fibrosis of orbital fat. There are remarkable similarities between our 2 patients and the other 4 reported cases: all were shunted in the early teenage years (mean 8.3 years; range 3 months to 14 years) after a period of childhood hydrocephalus, and all became symptomatic a few years after shunting (mean 19.3 years; range 11–34 years) (Table).[7,8] We propose that the sudden drop of intracranial pressure in the early teenage years leads to an "implosion" of the calvarium; at this age, the only thin (and possibly poorly calcified) part of which would appear to be the very thin floor of the frontal cranial fossa (the orbital roof). As the only area flexible enough for remodeling is the orbital roof, this gets progressively bowed upward in response to the low intracranial pressures over the ensuing years. This upward bowing of the roof leads to the characteristic clinical and radiologic changes, in the absence of symptomatic intracranial disease, of this newly recognized postshunting enophthalmos syndrome that we call "silent brain syndrome." Most cases reported to date, including our series, went undiagnosed for a long time despite frequent ophthalmic and neurosurgical follow-up examinations. This fact led us to believe that silent brain syndrome may in fact be underrecognized and that an increased awareness of this condition and its surgical management will help identify and treat many more patients in the future. The treatment that we recommend consists of orbital roof implantation via an upper eyelid crease approach or, less satisfactorily, by addressing the upper eyelid retraction and severe entropion.

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