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Authors

Yoon, Michael K
Parsa, Andrew T
Horton, Jonathan C

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Skull thickening, paranasal sinus expansion, and sella turcica shrinkage from chronic intracranial hypotension

Case report

MICHAEL K. YOON, M.D.,^{1,2} ANDREW T. PARSA, M.D., PH.D.,³
AND JONATHAN C. HORTON, M.D., PH.D.^{4–6}

¹Department of Ophthalmology, Harvard Medical School; ²Massachusetts Eye and Ear Infirmary, Boston, Massachusetts; and Departments of ³Neurosurgery, ⁴Ophthalmology, ⁵Neurology, and ⁶Physiology, University of California, San Francisco, California

In children or young adults, the morphology of the skull can be altered by excessive drainage of CSF following placement of a ventriculoperitoneal (VP) shunt. In Sunken Eyes, Sagging Brain Syndrome, gradual enlargement of the orbital cavity occurs from low or negative intracranial pressure (ICP), leading to progressive bilateral enophthalmos. The authors report several heretofore unrecognized manifestations of this syndrome, which developed in a 29-year-old man with a history of VP shunt placement following a traumatic brain injury at the age of 9 years. Magnetic resonance imaging showed typical features of chronic intracranial hypotension, and lumbar puncture yielded an unrecordable subarachnoid opening pressure. The calvaria was twice its normal thickness, owing to contraction of the inner table. The paranasal sinuses were expanded, with aeration of the anterior clinoid processes, greater sphenoid wings, and temporal bones. The sella turcica showed a 50% reduction in cross-sectional area as compared with that in control subjects, resulting in partial extrusion of the pituitary gland. These new features broaden the spectrum of clinical findings associated with low ICP. Secondary installation of a valve to restore normal ICP is recommended to halt progression of these rare complications of VP shunt placement.
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KEY WORDS • enophthalmos • ventriculoperitoneal shunt •
cerebrospinal fluid • hydrocephalus • pneumocephalus •
Sunken Eyes, Sagging Brain Syndrome

IN 1996 Meyer and colleagues²³ described a new syndrome characterized by progressive bilateral enophthalmos in 3 adults who had undergone VP shunting as children. The enophthalmos was so severe that poor eyelid-globe apposition developed, leading to the breakdown of the corneal epithelium. Orbital CT evaluation revealed no abnormalities of the bony orbit or surrounding sinuses, although no formal volumetric analysis of the bony orbit was performed. These authors were intrigued by the history of VP shunting in each patient but warned that “a causal relationship between hydrocephalus and enophthalmos is not proved by our cases.” Similar cases were reported sporadically in subsequent years, each associated with a history of CSF shunting in childhood.^{4,7,14}

In 2011 Hwang and colleagues¹⁸ described 4 patients

with progressive bilateral enophthalmos after VP shunting. No control valve was installed in the patients at the time of surgery to prevent excessive shunting of CSF. All 4 patients showed typical neuroimaging signs of low ICP. In 2 patients, the ICP was measured and found to be extremely low or negative. These 2 patients were treated by incorporating a valve into their shunt, resulting in slight improvement in the enophthalmos. The phenomenon of enophthalmos from chronic intracranial hypotension was named “Sunken Eyes, Sagging Brain Syndrome.”

Analysis of orbital CT scans revealed a 35%–40% increase in orbital volume in patients with enophthalmos from chronic intracranial hypotension.¹⁸ This increase in volume was principally caused by remodeling of the orbital roof, which assumed an arched configuration that

Abbreviations used in this paper: ICP = intracranial pressure; VP = ventriculoperitoneal.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.

expanded it into the anterior cranial fossa. It was hypothesized that low or negative ICP from overshunting caused upward bowing of the orbital roof, slowly leading to progressive enophthalmos.

We describe here another patient with progressive enophthalmos from overshunting. In addition to orbital expansion, novel features of chronic intracranial hypotension were present. Neuroimaging revealed marked thickening of the skull from contraction of the inner table, expansion of the paranasal sinuses, and shrinkage of the sella turcica. The current report expands the spectrum of clinical findings associated with Sunken Eyes, Sagging Brain Syndrome, which should facilitate the recognition of this rare but preventable condition.

Case Report

History. This 29-year-old man was struck by a motor vehicle in 1991 when he was 9 years old, resulting in a severe brain injury and a prolonged coma. An emergency bifrontal craniotomy was performed to remove a large intracerebral hematoma. Subsequently, ventricular enlargement developed, requiring placement of a left parietal VP shunt. The shunt did not incorporate a valve to regulate ICP. Eventually, the patient regained consciousness, but he had permanent neurological deficits, including quadriplegia, seizures, and impaired mentation.

In 2003 the patient, in a stupor after a 10-minute tonic-clonic seizure, was taken to a hospital emergency room. Computed tomography showed left frontal pneumocephalus (Fig. 1 left). Enlargement of the paranasal sinuses and thickening of the calvaria were also present (Fig. 1 right). He underwent repair of a leak in the posterior wall of the left frontal sinus and skull reconstruction with a titanium mesh. Over the next 3 years he had several episodes of frontal sinusitis, and a sinocutaneous fistula developed, requiring surgical closure.

In 2009 he was hospitalized with lethargy and fever. To address the possibility of a VP shunt infection, a spinal puncture was performed with fluoroscopy while the patient was prone. The radiologist reported no spontane-

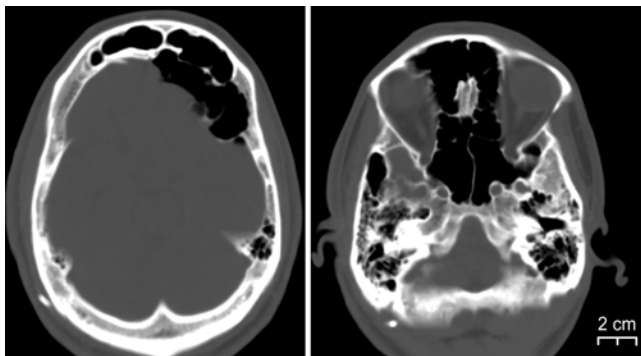


FIG. 1. Computed tomography scans obtained when the patient was 21 years old, 12 years after placement of a VP shunt with no valve. **Left:** Axial section showing an extraaxial air collection adjacent to the left frontal sinus. The calvaria is abnormally thick. **Right:** Lower axial section demonstrating enlargement of the ethmoid and sphenoid sinuses, along with abnormal proliferation of the mastoid air cells.

ous egress of CSF when the needle tip was placed in the lumbar subarachnoid space, suggesting a low or negative ICP. The patient was treated with intravenous antibiotics and recovered to his baseline level of function.

Examination. In 2011 the patient was referred for evaluation because of ocular misalignment and progressive enophthalmos. Visual acuity was 20/40 in each eye with refractive correction. Pupils were normal. Visual fields were intact to confrontation testing. Extraocular eye movements were full. There was an alternating exotropia measuring 25°. The globes were enophthalmic, with a deeply recessed superior eyelid sulcus. A Hertel exophthalmometer yielded a reading of 10 mm in each eye (normal mean: 16.6 mm, 95% range: 10.9–22.4 mm⁶). Despite the enophthalmos, the eyelids were apposed to the globe. Slit-lamp examination showed no corneal exposure. Examination of the fundi through dilated pupils was normal.

Neuroimaging and Skull Measurements. A CT scan showed thickening of nearly the entire skull, principally due to excessive separation of the inner and outer bony tables (Fig. 2). As a result, the diploic space was abnormally wide. The frontal, sphenoid, and ethmoidal sinuses were enlarged. There was aeration of the greater wings of the sphenoid bone and the anterior clinoid processes. The mastoid air cells were more extensive than normal and extended into the squamous portion of the temporal bone.

The thickness of the patient's calvaria was measured on the CT scans using open source imaging software (OsiriX, version 3.9.4, OsiriX Foundation). This software allows one to adjust the imaging plane for each measurement point to obtain an image oriented perpendicular to the skull surface. To image the skull in perfect cross-section was important, because an oblique cut would give a falsely thick reading. A total of 15 sites were measured on each side, located along parasagittal lines 1.5, 3.5, 5.5, and 7.5 cm from the midline (Fig. 3). These loci were chosen because control data were available from normal adults.¹⁷ In our patient, the mean (\pm standard deviation)

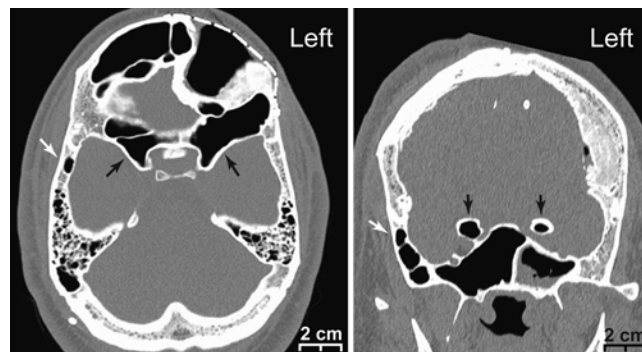


FIG. 2. Computed tomography scans revealing thickening of the calvaria with expansion of the diploë. **Left:** Axial view showing enlarged frontal sinuses and a titanium mesh closing a defect in the left frontal bone. The sphenoid sinus has expanded into the greater wings of the sphenoid bone (*black arrows*). Mastoid air cells are present in the squamous portion of the temporal bone (*white arrow*). **Right:** Coronal view showing aeration of the anterior clinoid processes (*black arrows*), large sphenoid sinuses, and air pockets in the temporal bone (*white arrow*).

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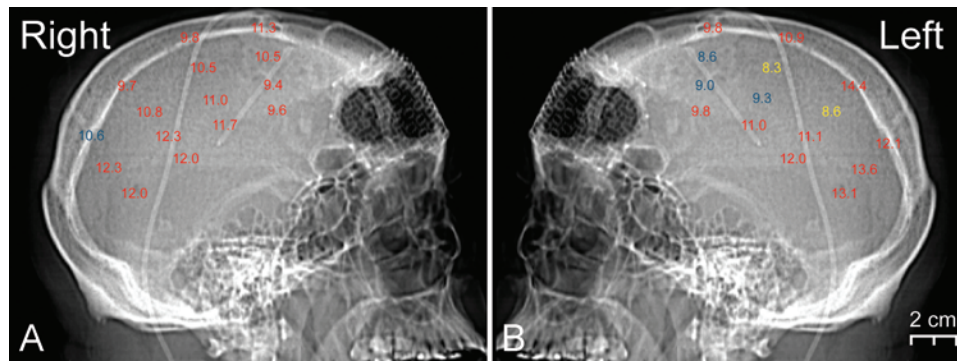


Fig. 3. Lateral scout film of the skull showing the locations where measurements of skull thickness were made from CT scans. The image was flipped horizontally to depict sites on the right (A) and left (B) calvariae. Values are expressed in millimeters. Color denotes the number of standard deviations greater than the published mean normal values (*red* = 3, *blue* = 2, *yellow* = 1). Note the shunt tubing, with no valve to regulate flow of CSF.

value for skull thickness was 10.8 ± 1.52 mm, compared with a value of 5.6 ± 0.56 mm in normal subjects. At 24 of 30 sites, skull thickness was 3 SDs greater than the mean normal adult skull thickness. At every site, skull thickness was at least 1 SD greater than the mean normal value.

To learn how the patient's skull thickness had changed over 2 decades, we compared the CT scan with a study from 1991 that had been obtained 10 days after his accident. The 1991 CT scan was preserved only on film. Without an electronic data file, the image plane could not be adjusted to make accurate measurements of skull thickness. However, it was evident that the skull had approximately doubled in thickness (Fig. 4). To determine the internal length of the skull in the sagittal plane, the distance from the internal occipital protuberance to the frontal crest was measured. The value was 15.2 cm at the age of 9 years and 14.2 cm at the age of 29 years. The accuracy of this comparison was limited by changes in overall skull and brain morphology as well as differences in the imaging technique, but the measurement suggests that the internal volume of the skull had actually diminished from age 9 years to adulthood, owing to contraction of the inner table of the skull.

An MR image confirmed that the skull was thick-

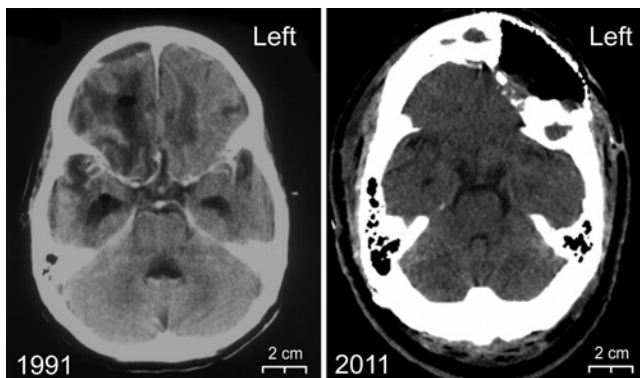


Fig. 4. Axial CT scans from 1991 (left) just after the traumatic injury and from 2011 (right) showing an increase in skull thickness over 20 years. The imaging planes are not identical because of changes in head shape over 2 decades, but a marked increase in skull thickness is apparent.

ened, with widening of the diploë (Fig. 5). There was marked Gd enhancement of the dura mater, a characteristic feature of low ICP. The sphenoid sinus was enlarged and the clivus was partially aerated. The sella turcica was unusually small. As a result, the pituitary gland was extruded superiorly into the suprasellar cistern, deforming the optic chiasm.

The size of the sella turcica was measured by drawing a straight line on the sagittal MR image from the dor-

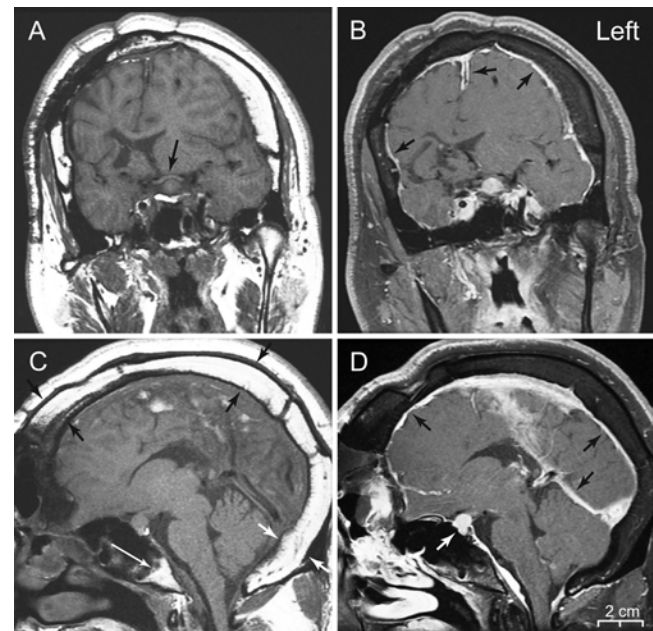


Fig. 5. Magnetic resonance images with features of chronic intracranial hypotension. Coronal T1-weighted MR image (A) showing thickening of the skull from widening of the diploë. The optic chiasm (arrow) is flattened by the pituitary gland. The brain morphology is distorted with midline shift to the right from the old traumatic injury. Coronal image (B) obtained after Gd administration and fat suppression, showing excessive enhancement of the dura (arrows). Sagittal T1-weighted image (C) showing marked expansion of the diploë (opposing arrow pairs), which is 1-cm thick in some areas. The sphenoid sinus has enlarged into the clivus (single white arrow). Sagittal image (D) obtained after Gd and fat suppression, showing dural enhancement (black arrows). The sella turcica (white arrow) is small, extruding the pituitary gland upward into the optic chiasm.

sum sella to the tuberculum sella to delineate the sellar opening. This dimension measured 8.9 mm. The line was continued along the border between the sphenoid sinus and the pituitary fossa to encircle the sella turcica. The cross-sectional profile had an area of 33 mm². The same measurement was made in 10 normal adults with a mean age of 28 years (range 25–32 years). The mean sagittal cross-sectional area of the sella turcica in these normal subjects was 62 ± 14 mm². The sella turcica in our patient was smaller than the mean control value by 2 SDs.

An operation was recommended to add a pressure-control valve to the VP shunt. The patient's guardian declined but agreed to allow the procedure in the future if pneumocephalus recurred.

Discussion

Shunting of CSF is performed frequently to treat elevated ICP, especially in cases of obstructive hydrocephalus. The incorporation of a valve to regulate the flow of CSF is intended to prevent overshunting. Even when a valve is installed, however, overshunting can be a problem following VP shunt placement. The frequency of this complication ranges widely in different neurosurgical series, from 1.5%–34%.^{2,9,10,21} Symptoms include postural headache, nausea, vomiting, lethargy, and coma.^{20,25} Neuroimaging may demonstrate subdural hematoma, flattening of the pons, herniation, Gd enhancement of the dura, and slit ventricles.⁵ These changes can also occur in spontaneous cases of intracranial hypotension from dural leakage.^{13,16,28}

More than a half century ago, Nulsen and Spitz²⁶ were the first to incorporate a valve in a shunt for the treatment of hydrocephalus.³ Most neurosurgeons now install some type of valve in the shunt apparatus. Only a single report in the modern literature specifies how often a valve is omitted. Robinson and colleagues³⁰ reported that no valve was placed in 17 children (11%) among a cohort of 157 patients treated with shunting from 1990 to 1998. After 5 years, there was a significant difference in the rate of complications from overshunting. Problems occurred among 43% of children with no valve or a low-pressure valve, as compared with 17% of children with a medium- or high-pressure valve. These data underscore the importance of limiting the flow of CSF but confirm that excessive drainage can occur even if a valve is used.

Although the short-term complications of overshunting are familiar to neurosurgeons, the long-term manifestations have received less attention until recently. Eleven cases of progressive enophthalmos have been reported, each occurring many years after placement of a VP shunt in a child or young adult.^{4,7,14,18,23} This condition was called Sunken Eyes, Sagging Brain Syndrome after it was recognized that intracranial hypotension produces the enophthalmos.¹⁸ The mechanism responsible for the sunken eyes has not been established conclusively, but a pressure gradient across the orbital roof may play a role. In normal subjects, the ICP measured at the vertex occasionally drops below atmospheric pressure while sitting or standing.^{12,22,29} In a patient with a VP shunt and no valve, ICP is likely to become negative even more frequently when the

patient is upright. In our patient there was strong evidence for negative ICP: a history of spontaneous pneumocephalus, excessive dural Gd enhancement, and a “dry” spinal tap by a radiologist using fluoroscopy. If atmospheric pressure is greater than ICP, the gradient will exert an upward force on the orbital roof. This force will cause the orbital roof, a structure composed of much thinner bone than the external skull, to arch into the anterior cranial fossa. This remodeling of the orbital roof will enlarge the orbit, resulting in progressive enophthalmos.

A precedent for this proposed mechanism is provided by the silent sinus syndrome, another rare cause of progressive enophthalmos.^{8,11,27} In this disease, negative pressure within the maxillary sinus occurs because of blockage of the sinus ostium. As a result, the orbital floor is pulled inferiorly, giving rise to hypoglobus. Reventilation of the maxillary sinus by antrostomy alone can lead to partial restoration of the normal orbital floor position, eliminating the need for secondary surgical reconstruction in some patients.³¹

Our previous report of progressive enophthalmos from VP shunt placement focused on the change in the anatomy of the orbital roof induced by chronic intracranial hypotension.¹⁸ However, we noted that skull remodeling was not limited to the orbit. Patients also exhibited enlargement of the frontal, ethmoidal, maxillary, and sphenoid sinuses, as well as aeration of the anterior clinoid processes. In the present report, the patient had striking enlargement of the paranasal sinuses, as well as aeration of the clivus, anterior clinoid processes, greater sphenoid wings, and squamous portions of the temporal bones (Figs. 1 and 2). These structures all have thin bony walls that border the intracranial compartment. Just as for the orbit, the volume of these structures appears to have expanded after many years in response to the force exerted by atmospheric pressure against an intracranial compartment below atmospheric pressure.

The most remarkable finding in our patient was the diffuse thickening of the skull that occurred from centripetal migration of the inner table (Fig. 2). Comparison with published control data revealed that his skull was about twice the normal thickness (Fig. 3). Magnetic resonance imaging showed that the expansion was due mainly to increased diploë (Fig. 5). We theorize that chronically negative ICP pulled the inner table away from the outer table, reducing the volume of the intracranial compartment. This was verified by measuring a change in the internal sagittal length of the skull from 15.2 to 14.2 cm over 20 years.

In 1966 Moseley and colleagues²⁴ described thickening of the skull in children after ventricular shunting, a process they called “hyperostosis cranii ex vacuo.” They attributed the skull thickening to premature closure of the cranial sutures. In 1970, 2 groups independently confirmed the rare occurrence of skull thickening after shunting for hydrocephalus. Anderson et al.¹ found thickening of the calvaria in 7 of 230 patients who had undergone shunt surgery in the period from 1938 to 1967. All 7 patients were younger than 12 years of age at the time of surgery. These authors stated, “The reason why these few patients (out of a much larger post-shunt group

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not so affected) show the changes described is unknown.” Griscom and Oh¹⁵ identified 5 of 140 children who demonstrated pronounced skull thickening after ventricular shunting. These authors concluded that inward growth of the inner table was “an uncommonly seen but physiologically reasonable accompaniment of relief of childhood hydrocephalus.” None of these reports linked skull thickening after shunting to excessively low ICP.

Another unusual feature of the patient described in the present report was the small size of the sella turcica, which measured only 33 mm² in the sagittal cross-section. Comparison with a control group of patients showed a nearly 50% reduction in area, with partial extrusion of the pituitary gland. A previous report featured a patient with intracranial hypotension and an abnormally small sella turcica, although the finding was not appreciated by the authors.¹⁸ We measured the size of the sella turcica in that published case (their Fig. 3A), obtaining a value of 31 mm². These 2 cases demonstrate that the sella turcica shrinks in response to chronic intracranial hypotension in some patients who have undergone VP shunting. This observation was first reported by Kaufman and colleagues¹⁹ in a series of shunted patients, but the authors attributed it to thickening of the bone from loss of CSF pulsations.

Strabismus has occurred in 6 of 12 patients with Sunken Eyes, Sagging Brain Syndrome, including the patient in the present case report.^{4,7,14,18,23} Given the complex neurological history of the patients, there are many potential explanations for their ocular misalignment, but in some cases altered muscle dynamics from posterior migration of the globe into an enlarged orbit may be a contributing factor.³³

In slit ventricle syndrome, excessive drainage after VP shunt placement produces small ventricles and cranial vault collapse with secondary craniosynostosis.³² This can lead to scaphocephaly or microcephaly, scalloping of the inner calvarial table, and high ICP. It is unknown why overshunting leads to slit ventricle syndrome in some patients and Sunken Eyes, Sagging Brain Syndrome in others. It is also uncertain why Sunken Eyes, Sagging Brain Syndrome is so rare, even among patients without a shunt valve.^{1,15} Clearly, additional factors remain to be elucidated in the pathogenesis of this syndrome.

Sunken Eyes, Sagging Brain Syndrome should become even rarer in the future, because few VP shunts are installed today without a valve. Nonetheless, there is a small cohort of patients who underwent shunt placement as children or young adults in an era when valves were less ubiquitous and who are now manifesting signs of this condition as they enter middle age. Placement of a valve and restoration of normal ICP can arrest progression.¹⁸

Disclosure

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Author contributions to the study and manuscript preparation include the following. Conception and design: Horton, Yoon. Acquisition of data: all authors. Analysis and interpretation of data: Horton, Yoon. Drafting the article: Horton, Yoon. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf

of all authors: Horton. Statistical analysis: Horton. Administrative/technical/material support: Horton. Study supervision: Horton.

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Address correspondence to: Jonathan C. Horton, M.D., Ph.D., University of California, San Francisco, 10 Koret Way, San Francisco, California 94143. email: hortonj@vision.ucsf.edu.