Pearls & Oy-sters:

Low-pressure hydrocephalus and inadequate shunting

Alex P. Michael, MD
Ahmad Elkouzi, MD
Rodger J. Elble, MD,
PhD

Correspondence to
Dr. Michael:
amichael@siumed.edu

PEARLS

- Disabling acute or subacute hydrocephalus can occur at low intraventricular pressures (<5 – 8 cm H₂O).
- A major risk factor for low pressure hydrocephalus (LPH) is chronically shunted hydrocephalus with a medium pressure valve (valve opening pressure ≈ 7 – 10 cm H₂O).
- The treatment of LPH typically requires ventricular drainage at negative pressures.

OY-STERS

- Adults with normal pressure hydrocephalus (NPH) frequently improve following ventriculoperitoneal shunting, but many regress to baseline in less than 3 years.
- The regression following initial improvement is largely unexplained.
- It is possible that persistent ventriculomegaly, resulting from inadequate drainage, promotes the occurrence of LPH in some patients with NPH.

We report a case of LPH and discuss how this phenomenon could play a role in the poor response of NPH to ventriculoperitoneal shunting.

CASE REPORT A 30-year-old woman was in good health except for a history of frequent headaches since childhood. These headaches occurred every 1–2 weeks and were often associated with photophobia, phonophobia, and nausea. There was a history of depression but no history of traumatic brain injury, prior CNS infections, or developmental delay. The patient's hydrocephalus was discovered fortuitously with a CT head scan (figure 1A) when she was evaluated in an emergency department for a scalp laceration sustained during a domestic altercation. An MRI revealed partial aqueductal stenosis (figure 1, B and C) and no acute intracranial abnormality. A neurologist and neurosurgeon found no abnormalities on physical examination except for mild optic disk blurring in the right eye. An ophthalmologist

confirmed the presence of mild papilledema. Visual acuity and visual fields were normal. At the time she was 66 inches tall and weighed 130 pounds. She had 2 sons and 1 daughter. One son received a ventriculoperitoneal (VP) shunt at 3 months of age for congenital aqueductal stenosis. Neither she nor her son was tested for a genetic cause of aqueductal stenosis. She had no other family history of hydrocephalus.

Twenty-eight days after the altercation, a VP shunt was inserted with a PS Delta valve (Medtronic; Minneapolis, MN) set at performance level 1.5 (opening pressure 7 cm H₂O supine and 8.5 cm H₂O standing). The patient's opening pressure was 25 cm H₂O by manometry at the time of surgery. CSF was slightly traumatic (10 leukocytes and 700/mm³ erythrocytes), but protein, glucose, gram stain, and cultures were normal. A postoperative CT revealed no change in ventricle size.

The patient's headaches did not change with the initial shunt. Four months later, a 50-lb box fell on her head, prompting her to seek medical attention in a local emergency department. There was no loss of consciousness or other apparent injury at this time. A CT scan of the head revealed ventricular enlargement that was slightly less than that which preceded her shunt (figure 1D). A shunt series was normal.

The patient's headaches began increasing a few days after the head injury. Fifteen days after the head injury, she began vomiting and became stuporous and mute within 24 hours. A CT head scan revealed increased ventricular size and effacement of the cortical sulci (figure 1E). An emergency ventriculostomy was placed, and the initial pressure was only 5 cm CSF. CSF was slightly traumatic (8 leukocytes and 9,850/mm³ erythrocytes), but glucose, protein, gram stain, and cultures were normal. Her clinical condition did not improve unless CSF was siphoned at negative pressures. Her VP shunt was revised with an adjustable PS Strata II valve (Medtronic) placed on the lowest possible resistance (performance level 0.5: opening pressure 1.5 cm H₂O supine, 3 cm H₂O standing). This continued to result in depressed

From the Department of Surgery, Division of Neurosurgery (A.P.M.), and Department of Neurology (A.E., R.J.E.), Southern Illinois University School of Medicine, Springfield.

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Top row (A–C): Axial CT on the day of altercation and axial T2 fluid-attenuated inversion recovery MRI and sagittal T1 MRI on day of initial shunt surgery. Bottom row (D–F): Axial CT scans on the day a box fell on the patient's head, 15 days later when she was stuporous and vomiting, and after 7 months of drainage with a valveless shunt.

consciousness unless the external ventricular drain press was <5 cm H_2O . After several days, the external drain was removed, and a valveless VP shunt was placed to allow continuous drainage. With this treatment, she recovered consciousness and was discharged home with no neurologic deficit. However, her chronic headaches continued as before, even though CT scans 7 months, 2 years, 3 years, and 5 years later revealed no hydrocephalus (figure 1F). Five years after placement of the valveless shunt, a fiber optic intraparenchymal monitor revealed normal pressures. Her chronic headaches persisted and were treated as migraine.

DISCUSSION Four characteristics of patients with LPH have been described: (1) successful initial treatment of hydrocephalus with a medium pressure shunt or external drain (valve opening pressure ≈ 7 – 10 cm H_2O), (2) subsequent development of acute or subacute neurologic deterioration and ventriculomegaly at intraventricular pressures below the opening pressure of the shunt or drain, (3) persistence of ventriculomegaly and neurologic deterioration in the presence of ventricular drainage at mid-normal pressures, and (4) clinical and radiologic improvement with drainage at

negative pressures.^{1,2} Our case illustrated all of these characteristics. Her obstructive hydrocephalus with mild papilledema was discovered fortuitously during an evaluation for head trauma that produced a scalp laceration without loss of consciousness. Her initial VP shunt produced little change in ventricular volume, but her papilledema resolved. Four months later, she experienced a dramatic episode of LPH following seemingly insignificant head trauma, which required ventricular drainage at negative pressures to relieve symptoms and normalize ventricular volume. Her stupor, mutism, and vomiting repeatedly occurred at intraventricular pressures of 5–8 cm H_2O , suggesting that mechanical distortion of the brain, not high pressure, was the cause of her symptoms. Her surgeon ultimately resorted to a valveless shunt that resulted in complete resolution of her acute symptoms and ventriculomegaly.

Several authors have described a hysteretic pressure–volume relationship for the ventricular system in hydrocephalus, similar to that shown in figure 2.^{1,3} In acute experimental hydrocephalus, there is a sharp initial rise in intraventricular pressure from the initial condition (point 0, figure 2), with little change in ventricular size.⁴ Within about 48 hours, intraventricular pressure begins to fall due to

Figure 2 An artist's conception of the hysteretic pressure-volume relationship of low and normal pressure hydrocephalus

In acute hydrocephalus, starting at point 0, there is a rapid rise in intraventricular pressure with little change in ventricular size during the first 24–48 hours. Plastic change in the surrounding brain tissue ensues, resulting in an increase in ventricular volume and a decrease in pressure. A new chronic hydrocephalic state may be achieved with normal intraventricular pressure (point 1). A subsequent insult may result in a second expansion of ventricular volume, producing low pressure hydrocephalus (point 2). Shunting at negative pressures is typically necessary to produce additional plastic change that restores the ventricles to normal (point 0).

increasing ventricular compliance and ventriculomegaly. This ventricular distension is associated with plastic change (loss of elasticity) in the brain parenchyma, producing sustained ventriculomegaly at normal intracranial pressure (ICP) (point 1, figure 2). Adequate drainage of CSF at point 1 (figure 2) may reduce ventricular volume and ICP, resulting in further plastic change in which the ventricles return to normal size and elasticity (point 0). This was not achieved when our patient was initially shunted, and we speculate that this may have predisposed her to LPH.

LPH is an uncommon entity that has been described in several case series.^{1,2} LPH usually begins months or years after ventricular shunt placement, but it can also occur in the setting of acute ventricular drainage.² Insults such as trauma, infection, and subarachnoid hemorrhage may perturb the ventricular system from point 1 in figure 2 into another hysteretic cycle of ventricular enlargement, resulting in increased ventricular size at persistently low pressure (LPH: point 2, figure 2).² Drainage at negative pressures is usually needed to reduce ventricular size and restore neurologic function.

Several investigators have speculated that a phenomenon similar to LPH might underlie shunt failure and clinical relapse in patients with NPH.^{1,5,6} Shunt resistance and clinical relapse are common in patients

treated for NPH.⁷ The initial clinical improvement in NPH following CSF drainage is often associated with little or no change in ventriculomegaly, and this has led to the notion that a reduction in ventricular size is not necessary for successful treatment of NPH.^{8,9} However, some investigators found a beneficial response to shunting primarily in patients who experienced reduced ventricular volume in response to lowered shunt valve pressure settings.¹⁰ Persistent ventriculomegaly in elderly patients with NPH is often attributed, perhaps falsely, to cerebral atrophy or permanent ventricular deformity. The initial medium pressure shunt did not correct the ventriculomegaly in our young patient, and the reversibility of her ventriculomegaly was only discovered when she was treated aggressively for disabling acute LPH. Ventricular drainage at negative pressures is needed to reduce ventricular size and restore parenchymal elasticity in LPH, and this may also be true for some cases of NPH.⁶ The clinician's urge to aggressively drain these patients is tempered by the risks of overdrainage.⁸ Protocols for aggressive drainage at negative pressures have been described,^{1,5} and a siphoning shunt, similar to the one used in our patient, was used to treat 3 patients with NPH.⁶ Controlled studies are needed.

AUTHOR CONTRIBUTIONS

Alex P. Michael: intellectual content and original manuscript draft. Ahmad Elkouzi: critical revision of the manuscript. Rodger J. Elble: intellectual content and original manuscript draft.

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