Negative-pressure hydrocephalus

MICHAEL VASSILYADI, M.Sc., M.D., JEAN-PIERRE FARMER, M.D., C.M., F.R.C.S.(C), and JOSÉ L. MONTES, M.D.

Department of Neurosurgery, The Montreal Children’s Hospital, Quebec, Canada; and McGill University, Montreal, Quebec, Canada

Two cases of children with closed myelodysplasia, Arnold-Chiari malformation, and shunted hydrocephalus who underwent spinal arachnoid cystopleural shunting are presented. Postoperatively, both patients developed craniovertebral symptomatology accompanied by radiologically documented ventricular dilation in spite of negative intracranial pressure and functional ventriculoperitoneal (VP) shunts. Both patients recovered after the cystopleural shunts were revised to increase the resistance to flow within the system. The authors believe that some communication between the arachnoid cyst and the subarachnoid space existed in both cases and that the negative pleural pressure was transmitted preferentially to the spinal and cerebral convexity subarachnoid spaces with relative sparing of the ventricular system. A transmantle pressure gradient was thereby established, leading to ventricular dilation. The authors further suggest that a craniospinal gradient was possibly established as well, leading to craniovertebral symptomatology in the patients. Return of flow in the VP shunts was obtained by correcting this iatrogenic transmantle pressure gradient.

KEY WORDS • hydrocephalus • spinal arachnoid cyst • Arnold-Chiari malformation • myelomeningocele • cystoperitoneal shunt

Hydrocephalus is a condition caused by a discrepancy between the production and absorption of cerebrospinal fluid (CSF), leading to ventricular dilation. The ventricular dilation is believed to result from increased intraventricular pressure.20 In communicating hydrocephalus, this increased pressure becomes equalized throughout the central nervous system (CNS), whereas in noncommunicating hydrocephalus, pressure gradients are established between different compartments of the CNS. Shunt-dependent hydrocephalic patients, at the time of shunt dysfunction, exhibit signs and symptoms compatible with increased intracranial pressure (ICP) and are “expected” by their physicians to demonstrate enlarged ventricles. The majority of patients do, but there are some who do not6,8,21 and others who do, but only in a subtle fashion.8 In addition, at the time of shunt revision, although one would “predict” finding uniform intracranial hypertension proportional to the ventriculomegaly on manometric measurement, this often fails to be the case.19

It is clear to many researchers6,8,19,20 that chronic shunting can affect the compliance of the brain. In some patients it is increased, in others decreased. In most patients the change is of little consequence.19–21

In this paper, we describe two patients with long-standing ventriculoperitoneal (VP) shunts who, at the time of treatment of spinal arachnoid cysts with cystopleural shunts, developed acute ventriculomegaly that was not associated with shunt malfunction. Although the clinical status of each of the patients suggested intracranial hypertension, ICP measurements were subatmospheric. Iatrogenically induced negative-pressure hydrocephalus was documented and subsequently treated.

Illustrative Cases

Case 1

This 13-year-old girl presented with a history of progressive deterioration in gait. According to her history, the patient had had a low lumbar myelomeningocele that had been repaired at birth and had undergone bilateral VP shunting and a Chiari decompression at the age of 1 year. At that time, a spinal arachnoid cyst was identified. To ameliorate her present symptoms, she had undergone two attempts at subarachnoid cystopleural shunting and two attempts at cyst marsupialization, as well as release of a tethered cord.

Although initially the patient had been ambulatory, at presentation she was wheelchair-bound from progressive hamstring flexion contractures and ongoing painful spasms that were unresponsive to Lioresal or Dantrium. A spine magnetic resonance (MR) image revealed a large posterior arachnoid cyst (Fig. 1), and a computerized tomography (CT) brain scan showed a well-decompressed ventricular system (Fig. 2 left). Given the severity and relentlessness of her pain, and given her past surgical history, we inserted a right subarachnoid cyst-to-pleural shunt, which resulted in the patient’s receiving significant relief from back and right leg pain during the initial postoperative period. However, on the third postoperative
day, the girl experienced an episode of tachycardia with hypotension and associated diaphoresis, and a decreased level of consciousness. A CT scan revealed ventriculomegaly involving both of the lateral ventricles and the third ventricle (Fig. 2 right). This finding represented a significant change from the results of her preoperative examination. Her shunt was tapped for 30 cc of slightly xanthochromic fluid, which resulted in a marginal improvement in her mentation. She underwent a VP shunt revision with the placement of a medium-pressure Pudenz–Schulte valve. At the time, the distal flow was thought to be normal as determined by manometric intraoperative measures. The ventricular catheter and valve were patent and the intraventricular pressure measured 2 cm H2O. The pulsations were dampened; for this reason the proximal catheter and valve were changed. She improved for approximately 5 days at which point the headaches, nausea, vomiting, decreased level of consciousness, and hypotension associated with diaphoresis recurred. A repeat CT scan showed recurrence of ventriculomegaly to a similar degree. The shunt was once again revised and, at this point, the distal flow was marginally decreased although the proximal catheter and valve were patent; intraventricular pressure was 13 cm H2O under the existing anesthetic condition. The distal catheter was changed. The patient did not improve and we inserted a right ventricular drain set to monitor (HP Monitor/Terminal, model 78634, Hewlett Packard Corp., Waltham, MA) for ICP. The patient’s ICP varied between 0 and 5 mm Hg throughout a monitoring period of 6 hours. Chest x-ray films did not reveal any pleural effusion. The patient was intubated and placed on positive end-expiratory pressure ventilation with a slight improvement in her hypotensive status. Her cystoperitoneal shunt was percutaneously clamped and, within a few hours, her neurological status returned to baseline with, paradoxically, a progressive rise in ICP to between 2 and 5 mm Hg. The drain was kept closed and, despite a 7- to 10-mm Hg rise in ICP, the ventricular system collapsed to the preoperative state, verified by a CT scan obtained 24 hours later. The cystoperitoneal shunt was removed and replaced by a left cystoperitoneal shunt system in which a low-pressure valve was inserted. A repeat CT scan showed the ventricles to be collapsed back to the preadmission state.

Case 2

This 4-year-old girl presented with a cervicothoracic arachnoid cyst initially thought to be hydromyelia on MR imaging (Fig. 3). A brain CT scan failed to show any shunt malfunction (Fig. 4 left). The patient’s history indicated that she had had T-12 myeloschisis repaired at birth, had VP shunt-dependent hydrocephalus, and a significant yet asymptomatic Chiari II malformation. The patient was treated by insertion of a cystoperitoneal shunt. After the procedure, she was unable to assume an upright position because of the development of severe postural headaches and vomiting; she could only eat in the decubitus position. Her signs did not improve after a 3- to 4-week period of observation (Fig. 4 right). A shuntogram showed proximal patency of the VP shunt, but a slow outflow into the peritoneum; instead, the tracer was visualized within the arachnoid cyst with a quick flow into the pleural space (Fig. 5 left). A right pleural effusion was seen on chest x-ray films. Based on our previous experience with Case 1, this patient’s symptomatology was thought to be secondary to transmission of negative pleural pressure into the subarachnoid space. A flow-regulated valve (Orbis Sigma, Cordis Corporation, Valbonne, France) was placed between the arachnoid cyst and the pleural cavity, and the patient’s symptoms completely resolved. A postoperative shuntogram revealed preferential flow through the VP shunt (Fig. 5 right) and a postoperative CT scan showed

Fig. 1. Case 1. A sagittal T1-weighted magnetic resonance image of the spine demonstrating a large posterior arachnoid cyst with compression of the thoracic spinal cord.

Fig. 2. Case 1. Noninfused computerized tomography scans of the brain. Left: Preoperative study showing collapsed ventricles and two ventriculoperitoneal shunts. Right: Scan revealing enlarged ventricular system on 3rd postoperative day after placement of a cystopleural shunt.
a return of the ventricular size to baseline. Postoperative chest x-ray examination showed decreased pleural effusion.

**Discussion**

Arachnoid cysts are believed to originate either from a developmental anomaly or from a delayed complication of arachnoiditis leading to duplication of the arachnoid membranes. These cysts are classified according to whether they communicate with the subarachnoid space. In the case of communicating arachnoid cysts, a "ball-valve" mechanism allows CSF to enter the cyst, but does not allow free egress of the accumulated fluid. Transient increases in CSF pressure ensure the continued patency and enlargement of these arachnoid cysts. Hydrocephalus or other conditions that lead to increased intraspinal pressure may exacerbate the condition.

Shunting of arachnoid cysts is currently the preferred treatment. Marsupialization requires a larger anatomical exposure and a longer duration of anesthesia; it is more hazardous and is associated with a high rate of recurrence.

Shunting from the cyst to the pleural cavity establishes a pressure gradient of $-5$ to $-8$ cm H$_2$O. In addition, if communication exists between the arachnoid cyst and the subarachnoid space, negative pleural pressures are transmitted to the spinal subarachnoid space. Because of the presence of obstructive hydrocephalus in patients with Chiari II malformations, this negative pressure is transmitted preferentially to the subarachnoid space of the convexity and, only to a lesser degree, to the ventricular system (Fig. 6B). As a result, the intraventricular pressure, although low or negative, is greater than the convexity subarachnoid pressure, and a transmantle pressure gradient is established that leads to ventricular dilation (Fig. 6B). The gradient is not dissipated by the VP shunt through which CSF does not flow because the ventricular pressure is lower than the opening pressure of the valve.

Furthermore, because the craniovertebral circulation is suboptimal in these patients and because the shunt is rendered nonfunctional despite being patent, a pressure gradient is also likely to be established at the craniovertebral junction (Fig. 6B). In describing low-pressure hydrocephalic states, others have believed the symptoms of obtundation to be either due to herniation or to altered cerebral perfusion as a result of the ventriculomegaly. Although conceivably, pericallosal flow could be altered by the enlarged ventricles, cerebrovascular resistance studies have shown a correlation between reduced flow and large ventricles only in high ICP conditions and not in low or negative pressure states.

Furthermore, the level of consciousness experienced by the patient depends much more on brainstem function (theoretically unhampered in this low-pressure state) than on parasagittal perfusion.

It therefore seems difficult to reconcile the altered perfusion hypothesis with the symptomatology and pressures demonstrated by our patients. We believe that the establishment of a craniovertebral pressure gradient that leads to altered brainstem function is a more plausible explanation.

The symptoms and signs demonstrated by patients, coupled with their ventriculomegaly, can mislead one to believe in a VP shunt malfunction. At the time of shunt revision in Case 1, there was no absolute sign of obstruction shown on manometry; the dampened waves believed to reflect a partial obstruction may, in fact, simply have represented a low-pressure state. In Case 2, flow was reestablished within the shunt system without VP shunt revision. It appears, therefore, as though the clinical pre-
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Fig. 5. Case 2. Shuntogram using technetium 99. Left: Adequate ventricular filling from the right ventriculoperitoneal (VP) shunt reservoir (upper darkened circle) with slow outflow via the VP shunt (arrow), and rapid flow into the spinal subarachnoid space (arrowhead) and pleural space (lower darkened circle). Right: Shuntogram obtained 1 month after revision of the cys-topleural shunt with the addition of a flow-regulated valve showing adequate ventricular filling from the VP shunt reservoir (arrow) with preferential flow into the peritoneum (arrowhead).

sentation is due, at least in part, to herniation from negative spinal pressure and the, albeit relative (that is, less negative), ventricular hypertension. The ventriculomegaly was corrected by altering the iatrogenically introduced transmantle and craniovertebral pressure gradients (Fig. 6C). Ventriculoperitoneal shunt revisions failed twice in Case 1.

In their classic description of normal pressure hydrocephalus, Hakim and colleagues9,10 proposed that dilated ventricles were maintained, despite normal intraventricular pressure, because of Laplace’s law. In this context, the increased pressure is dissipated by the increased area of the ventricular surface.

Clinical2,3,5,6 and experimental evidence1,3 have subsequently shown the ventriculomegaly of negative-pressure hydrocephalus to be related to the establishment of a pressure gradient between the ventricles and the convexity across the brain mantle. The absolute value of the intraventricular pressure was deemed unimportant as long as a transmantle gradient existed.3,7,12 It was always assumed that the gradient was created by a relative rise in the intraventricular pressure.

In negative-pressure hydrocephalus, as described in this paper, the ventricular dilation is not created by increased pressure within the ventricular system leading to the transmantle pressure gradient. The transmantle gradient is rather created by the preferential transmission of negative pleural pressures to the subarachnoid space of the convexity. The pressure within the ventricles, although “normal” and below the opening pressure of the valve, allows the establishment of the gradient.

Fig. 6. Schematic drawing displaying a model of pathophysiology of “negative-pressure hydrocephalus.” A: Normal state of pressure equilibrium achieved with a functioning ventriculoperitoneal (VP) shunt. B: With the introduction of a cys-topleural shunt, a negative subarachnoid fluid pressure is created. The negative pressure is transmitted preferentially to the cerebral subarachnoid space and a transmantle pressure gradient is created. The dilatation dissipates the relative intraventricular pressure and the shunt valve turns off. A craniovertebral pressure gradient develops, creating posterior fossa symptomatology. C: Correction with either a cystoperitoneal shunt or by addition of resistance within the existing cystopleural system reestablishes transmantle and craniovertebral equilibrium. The VP shunt resumes function.

Why then was the pressure measured in the ventricles of the patient in Case 1 lower than atmospheric pressure? It is possible that, as postulated by Hakim and colleagues,9,10 once the ventricular system had dilated, the transmantle pressure gradient was indeed dissipated by the enlarged surface area of the ventricular system, leading to a measured intraventricular pressure that was not very different from the theoretical negative pressure of the pleural space. A second reason may also exist. The shuntogram in Case 2 would suggest that some of the “less negative” ventricular pressure may dissipate via the egress of ventricular CSF into the spinal subarachnoid space (Fig. 5 left). That is, although the hydrocephalus tends to be obstructive in nature, the obstruction is incomplete. Consequently, the intraventricular pressure may well correlate (after the initial ventricular dilation) with the negative spinal and subarachnoid pressures created by the cystopleural shunt.

In describing shunted patients with a low-pressure hydrocephalic state, Pang and Altschuler19 recently postulated that a transient shunt obstruction is followed by a change in the viscoelastic properties of the brain due to the loss of extracellular fluid. The altered pressure-volume index leads, according to them, to a new equilibrium whereby a larger ventricular volume coexists with very low pressures, preventing drainage through otherwise functional shunts. The authors describe these changes as occurring in patients with low brain turgor, for example, those experiencing postischemic, postradiotherapy states, over a very long period.19 They also describe the treatment as requiring at times prolonged ventriculostomy at sub-atmospheric levels to correct these viscoelastic changes related to the decreased parenchymal fluid compartment prior to the definitive shunt revision.19

The condition described in this paper is different in many respects. First, it would appear as if a transient
obstruction of the VP shunt and subsequent rise in ventricular pressure may not be necessary to produce negative-pressure hydrocephalus. Second, the changes occur abruptly over a course of hours to days after cystopleural shunting and, correspondingly, the response to treatment is nearly immediate as documented by clinical and radiological improvement. We do not, therefore, feel that profound changes in the values of the pressure-volume index need to be effected in our patients. Additionally, one might wonder if the low-pressure hydrocephalic state triggered by lumbar punctures in two patients with long-standing shunts, described by Pang and Altschuler, may not, in fact, have been created by a mechanism similar to the one described here: that is, by the establishment of the transmantle pressure gradient as a result of low spinal and convexity subarachnoid pressure created by leakage of CSF in the subcutaneous tissues rather than by a transient shunt malfunction.

The conditions described in this paper are not dissimilar to other conditions in which two shunts appear to “compete” for drainage of CSF; for instance, after insertion of a low-pressure shunt in a sequestered fourth ventricle of a patient with a long-standing medium-pressure shunt in the lateral ventricles. In this situation, even if the two shunt systems are completely separate, the shunt that drains the lower pressure compartment tends to “win the competition” (RA Sanford, personal communication, 1993). It may well be that the lower pressures of the new fourth ventricle shunt system are transmitted preferentially to the convexity subarachnoid space. The lateral intraventricular pressure becomes higher than the convexity pressure while remaining lower than the opening pressure of the medium-pressure valve. A transmantle gradient is created and the ventricular system dilates. At this point, there are two therapeutic options: pumping the lateral ventricular shunt or increasing the resistance of the newly placed fourth ventricular system. The latter is similar to the treatment used in our patients.

Negative-pressure hydrocephalus is therefore an iatrogenically created condition that develops as a result of secondary, negative spinal subarachnoid pressures generated by a subarachnoid cystopleural shunt inserted in patients with Chiari malformations. In our experience, this condition has not been seen to develop as a result of syringopleural shunting, despite the probable introduction of negative pressures within the syrinx. It is possible that the collapse of the syrinx, seen in these cases, prevents negative pressure from being transmitted to the cranial compartment. Also, we have not had the opportunity to treat spinal subarachnoid cysts unassociated with Chiari malformations using pleural shunts.

To prevent this potentially lethal condition from occurring, it is advisable to consider that all symptomatic spinal arachnoid cysts may communicate with the subarachnoid space and to either place a shunt from the subarachnoid cyst to the peritoneal cavity or add a flow-regulated device to the system to prevent this iatrogenically induced condition.

References