Hypertensive slit ventricle syndrome: pseudotumor cerebri with a malfunctioning shunt?

Report of 3 cases

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Symptomatic shunt malfunction without ventricular enlargement is known as slit ventricle syndrome (SVS). Patients presenting with this syndrome are not a homogeneous group. Of the 5 different types classified by Rekate, Type 1 is caused by CSF overdrainage and is associated with low pressures; Types 2 and 3 are associated with shunt blockage and elevated CSF pressures; Type 4 is cephalocranial disproportion that increases brain parenchymal pressure but not CSF pressure; and Type 5 is headache unrelated to shunt function. The low and normal CSF pressure types are relatively well understood, but the high-pressure forms are more problematic. In the high-pressure forms of SVS it is said that the lack of ventricular dilation is related to a reduction in brain compliance analogous to idiopathic intracranial hypertension or pseudotumor cerebri. Despite this, there is little evidence in the literature to support this conjecture. With this in mind, 3 cases of SVS associated with elevated CSF pressure are presented. The MR venogram findings and hemodynamics of these 3 cases are shown to be identical to those of pseudotumor cerebri. A literature review indicates that an underlying venous impairment may be functioning in the patients who re-present with small ventricles following shunt malfunction.

(keywords: slit ventricle syndrome, hydrocephalus, venous sinus stenosis, cryptococcal meningitis)

SLIT ventricle syndrome (SVS) can be defined as a severe headache disorder in patients with previously inserted intraventricular shunts who are found to have small ventricles on cross-sectional imaging. Rekate has subclassified this syndrome into 5 types: Type 1, severe intracranial hypotension secondary to shunt over-drainage; Type 2, intermittent obstruction of the ventricular catheter with episodic pressure spikes; Type 3, normal-volume hydrocephalus (intracranial hypertension with small ventricles and a failed shunt); Type 4, cephalocranial disproportion often with Chiari I malformation (parenchymal compression due to premature suture closure with a working shunt tube); and Type 5, headache unrelated to shunt function. It can be seen that this is not a homogeneous group; patients with the various subtypes experience low, normal, or elevated CSF pressure. Type 1 is analogous to the low-pressure spinal headaches that occur following LP, and Types 4 and 5 do not involve alterations in CSF pressure, and so these types are not difficult to understand from a fluid-dynamics point of view. Why patients with Types 2 and 3 should re-present with elevated CSF pressures and small ventricles, especially when they may have been previously documented to have enlarged ventricles, is more difficult to explain.

The ependyma lining the ventricles represents an interface between the CSF and brain. As the CSF pressure rises, the ability of the brain to deform and allow ventricular enlargement is dependent on the brain’s compliance, a measure of the rate at which a volume changes following a pressure change. Simplistically, given an elevation in CSF pressure, those in whom the ventricles dilate must have higher brain compliance than those in whom the ventricles do not dilate. Rekate has suggested that the reduced brain compliance may be due to an elevation in venous outflow pressure analogous to idiopathic intracranial hypertension (IIH) or pseudotumor cerebri. He mentions 5 children with “normal volume hydrocephalus” or Type 3 SVS who underwent retrograde manometry and venography and showed elevated dural sinus pressure, but these cases were not further documented. A single patient with Type 3 SVS was shown to have venous outflow impairment on an MRV. The purpose of this paper is to document the MRI cross-sectional findings as well as the MR venography and hemodynamic data in 3 cases of SVS associated

Abbreviations used in this paper: CTV = CT venogram; ICP = intracranial pressure; IIH = idiopathic intracranial hypertension; LP = lumbar puncture; MRV = MR venogram; PVI = pressure volume index; \( R_{out} \) = CSF outflow resistance; SSS = superior sagittal sinus; SVS = slit ventricle syndrome.
with elevated CSF pressure to show that they are identical to the published findings in pseudotumor cerebri. A review of the literature will be undertaken in light of this.

Methods

Study Participants

My colleagues and I have previously noted an association between hydrocephalus and venous outflow abnormalities in children and young adults. Patients undergoing MRI examinations at a tertiary referral center for the initial diagnosis or follow-up of treated hydrocephalus are screened with additional MR venography and MR flow quantification sequences. The 3 patients included in this review were found to have symptomatic shunt malfunction and elevated intracranial pressures (ICPs), despite no evidence of ventriculomegaly, between August 2011 and October 2012.

Protocol for MR Sequences and Analysis

All patients underwent imaging on a 3-T superconducting magnet (Vario; Siemens). Standard 3D T1-weighted sagittal, and T2-weighted and inversion-recovery axial MRI sequences were performed. A time-of-flight MRV sequence was performed. A retrospectively cardiac-gated phase-contrast flow quantification sequence was used, with a TR of 26.5 msec, TE 6.9 msec, flip angle 15°, slice thickness 5 mm, matrix 192 × 512, FOV 150, and a single excitation. The velocity-encoding values of 40 cm/sec and 75 cm/sec were used. The lower velocity-encoding value was selected to maximize the measurement of the venous structures, with the higher one used to maximize the arterial measurements. The plane of section was selected to intersect the SSS approximately 2 cm above the confluence of sinuses and to pass through the basilar artery and the cavernous portion of the internal carotid arteries as per the literature. Regions of interest were placed around the SSS, the carotid arteries, and basilar artery in each patient. Care was taken to exclude aliasing by retrospectively manipulating the baselines of each resultant graph, giving an effective venous upper flow limit of 80 cm/sec and an arterial flow limit of 150 cm/sec. Background correction was used.

The addition of the flow in both the carotid and basilar arteries gave the total arterial inflow for each patient. The SSS outflow was calculated from the respective region of interest. The percentage of the arterial inflow returning via the sagittal sinus was calculated.

Case Reports

Case 1

This 46-year-old man presented with headache, nausea, vomiting, and fever. A CTV performed to rule out thrombosis showed a hypoplastic left transverse sinus with a dominant right transverse sinus. There was a prominent arachnoid granulation in the right sinus, giving a 70% stenosis by area (Fig. 1A). An MRI study performed the next day showed no parenchymal abnormality and no abnormal enhancement. The MRV was similar to the CTV except that in addition there were prominent scalp and facial veins noted (Fig. 1B). The ventricles were not dilated (Fig. 1C). The arterial inflow was 720 ml/min, with a sagittal sinus returning 38% of the arterial inflow. An LP showed an elevated CSF pressure of 34 cm/H₂O, with cryptococcal meningitis being confirmed.

Over the next 3 weeks intravenous antibiotics were given, but severe headaches continued. Repeat LP showed no evidence of infection, but the pressures remained high. A shunt tube was inserted but was malpositioned, passing through the right basal ganglia. This had no effect on the patient’s symptoms and was repositioned 1 week later. His headache improved and a repeat LP showed a pressure of 6 cm H₂O. Three weeks later he again developed headache and vomiting. A CT scan at this stage showed a small right ventricle. Ten days after the CT he developed worsening of the headache with confusion. Repeat LP showed a CSF pressure of 27 cm H₂O but no evidence of infection. An MRI study showed a small right ventricle around the tube and displacement of the septum pellucidum to the right; the left ventricle was normal (Fig. 1D). Results of the MRV were unchanged from the previous study. The arterial inflow was 910 ml/min with a 34% venous return. The patient was placed on a course of steroids and became asymptomatic. He remained well and was slowly weaned off the steroids over the course of several months.

Case 2

This 24-year-old man underwent shunt insertion at 6 months of age for congenital hydrocephalus. He re-
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presented at the age of 8 years with a blocked shunt, and this was revised. He suffered chronic headaches for many years with an exacerbation of the headaches at 21 years of age. An MRI study obtained at this time showed small ventricles (Fig. 2A). His arterial inflow was 1080 ml/min, with a sagittal sinus returning 35% of the arterial inflow. He was not treated and his symptoms waxed and waned over the next year.

He returned at the age of 22 years for a follow-up MRI sequence while asymptomatic, and the ventricles appeared slightly larger than before (Fig. 2B). At this point the arterial inflow was normal and the sagittal sinus was returning 47% of the inflow. He returned 1 year after this at the age of 23 with severe headache, nausea, and vomiting. He complained of photophobia, blurred vision, and visual obscurations. He was noted to have papilledema. A nuclear medicine shunt study showed no blockage. A repeat MRI study showed small ventricles (Fig. 2C), with the MRV showing a hypoplastic left transverse sinus and a high-grade stenosis of the right transverse sinus (Fig. 2D). At this point the arterial inflow was 1030 ml/min with a venous return of 34%. At LP the pressure was greater than 35 cm H2O with normal biochemistry. The shunt was revised, a higher-pressure valve was inserted, and he became asymptomatic.

Case 3

This 29-year-old man underwent shunt insertion at 18 weeks of age for idiopathic congenital hydrocephalus. He was well until 18 years of age when he required a revision for shunt blockage. He suffered intermittent severe headaches but remained relatively well in the meantime, until the age of 28 years when he presented with headache, seizure, blurred vision, and papilledema. A CT scan showed small ventricles and results of a nuclear medicine shunt study were normal. Overnight monitoring of a Rickham reservoir showed pressures between 20 and 60 cm H2O. An MRI sequence confirmed a slitlike right ventricle compressed around the shunt tube (Fig. 3A). The MRV showed a tight stenosis of the SSS just above the torcular and prominent collateral veins in the scalp and face (Fig. 3B). His arterial inflow was 740 ml/min, with his sagittal sinus returning only 16% of the arterial inflow. His vision began to deteriorate and the shunt tube was revised.

His initial response was favorable but he re-presented 8 months later with headaches, loss of vision, and papilledema. His CT scan again showed small ventricles. Findings on a repeat nuclear medicine shunt study were again normal. His MRI study again showed the sagittal sinus stenosis and very poor venous return. Due to the anticipated difficulty in shunt replacement, it was decided to perform a retrograde venogram with manometry. The venogram confirmed the tight stenosis of the distal sagittal sinus (Fig. 3C), with the pressure gradient across the stenosis measuring 18 mm Hg. The stenosis was treated with stent insertion, with the abolition of the pressure gradient (Fig. 3D). The patient became asymptomatic, with resolution of his papilledema at follow-up 2 months later.

Discussion

Shunt malfunction is not uncommon in patients treated for either hydrocephalus or pseudotumor cerebri. In those treated for hydrocephalus, when a shunt malfunction two groups of shunt-dependent patients emerge. In the first group the symptoms evolve rapidly, with little enlargement of the ventricles despite significantly elevated CSF pressures (SVS); in the second there is a more subtle deterioration, with lower CSF pressures but enlarged ventricles (similar to “standard” hydrocephalus). The difference between these two outcomes depends on the compliance of the brain. Compliance is a measure of the degree of change in volume that occurs secondary to a change in pressure. Obviously, if the ventricles enlarge in some patients but not in others when the CSF pressure rises, then there must be a difference in the compliance of the brain between these two groups. Two theories have been suggested to account for this—in the first, subependymal gliosis is thought to reduce ventricular enlargement; in the second a change in brain turgor is thought to be responsible.

It has been suggested that the long-standing presence of a venricular catheter may promote subependymal gliosis, which may lead to decreased ventricular compliance. The stiff subependyma is hypothesized to restrict the ability of the ventricles to enlarge. However, Del Bigio showed at autopsy that the degree of subependymal gliosis is no different in those with dilated or nondilated ventricles at shunt malfunction. In an elegant study, direct
measurement of the compliance of the ventricular wall was measured in 15 children who presented with slit ventricles following shunt malfunction. By simultaneously measuring the ventricular and brain parenchyma pressure adjacent to the ventricle wall and by altering the intraventricular pressure, it was shown that there was no change in wall compliance.25

Rekate16 defined brain turgor as the ability of the parenchyma to resist volumetric changes. It depends on the density of the brain parenchyma (fixed in the short term), intra- and extracellular water, and the intraparenchymal venous blood volume. Sood et al.24 reviewed this hypothesis and noted that although the brain parenchyma is deformable, it is not compressible and will not contribute to changes in compliance. Similarly, CSF is incompressible and will not affect compliance over the short term. Compliance is rather determined by the displaceable intracranial blood volume, which is predominantly within the veins. Therefore, brain compliance in the short term is the result of expulsion of blood from the veins and depends on the venous stiffness and the volume of the veins themselves. Stiffer veins have lower compliance, larger veins have higher compliance.12

**Total Craniospinal Compliance in SVS**

One would imagine that if the compliance of the brain were lower in patients with the high-pressure form of SVS, then this should show up in a measure of the total craniospinal compliance, such as the pressure volume index (PVI). The PVI is measured by infusing mock CSF into the lumbar canal to gauge the volume of fluid required to increase the CSF pressure by a factor of 10. Fried and Shapiro,12,22 in a series of studies, measured the PVI in 20 children who presented with normal or small ventricles following shunt failure and 18 who represented with moderately to severely enlarged ventricles following shunt failure. The compliance of those whose ventricles enlarged following shunt malfunction was twice that of those whose ventricles did not enlarge. The confusing factor was that although the compliance in the patients with SVS was lower than in the patients with dilated ventricles, it was still within the normal range for age-matched controls.

However, a total craniospinal compliance measure, such as the PVI, may be insensitive to changes in brain turgor for two reasons. First, approximately 70% of the total craniospinal compliance is provided by the spinal canal and only 30% by the cranial cavity,26 so we may be looking for a change in only a small component of the PVI. The spinal canal may also compensate in hydrocephalus. Second, if the underlying pathology is a limitation in venous outflow over the vertex, then the elevation in venous pressure may tend to decrease the compliance of the brain parenchyma but also simultaneously increase the vein volume in the subarachnoid space.12 This will tend to increase compliance around the brain despite increasing turgor within it, so the net effect globally may be balanced.

**Cause of the Shunt Blockage**

In the 3 cases reviewed, a shunt was inserted for control of CSF outflow pressure but later failed. In the patient in Case 1, the reason for shunt insertion was continuing elevated CSF pressure following treatment of cryptococcal meningitis. The patient appeared to have a pseudotumor-like syndrome with normal-sized ventricles despite the elevated pressure. Tan27 found that 11 of 34 patients with cryptococcal meningitis presented with papilledema and visual failure, with 9 of these having small or slitlike ventricles despite intracranial hypertension. The symptoms rapidly improved on LP, but 7 of these patients eventually required shunt insertion. Cremer et al.3 similarly noted a syndrome of intracranial hypertension without ventricular enlargement in 30%–70% of cryptococcal meningitis cases without dilation of the ventricles despite intracranial hypertension.

Thus, the presentation of Case 1 from this series is not unusual. Before shunt insertion the ventricles in this patient were of normal size and symmetrical (Fig. 1C). After the previously successful shunt had failed, the follow-up MRI (Fig. 1D) showed that the ventricle had collapsed around the shunt and the septum pellucidum had displaced to the right. The left ventricle was now larger than previously but still within the normal range. It has been suggested that if the septum pellucidum is intact, drainage of one lateral ventricle can cause it to collapse against the head of the caudate nucleus, closing the foramen of Monro and leading to a pressure change between the two sides.20

**Fig. 3.** Case 3.  **A:** A T2-weighted axial image showing the ventricle collapsed around the shunt (arrow).  **B:** An MRV showing very poor flow in the distal sagittal sinus (arrow) and collateral flow in the scalp and face.  **C:** An image of the sagittal sinus obtained during a retrograde venogram showing the high-grade stenosis (arrow); the pressure gradient across this stenosis was 18 mm Hg.  **D:** The sagittal sinus following a stent procedure in which the pressure gradient was abolished.
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The undrained left ventricle in this case compressed the septum and acted as a flap valve occluding the shunt.

Cases 2 and 3 appear similar. In Case 2, during a time of normal shunt function, there was fluid around the shunt (Fig. 2B). When the shunt failed it was compressed adjacent to the ventricular walls (Fig. 2A and C). Thus, the 3 cases would be classified as Type 2 in the Rekate classification. Nuclear medicine shunt studies in both Cases 2 and 3 showed no evidence of malfunction. This is probably because the obstructions were intermittent and functional. Injecting an amount of fluid into the shunt increases the volume of the ventricle into which it resides and reverses the obstruction temporarily. Thus, shunt studies may be misleading in this syndrome. Cases 2 and 3 occurred in patients who had previously undergone shunt insertion as children for congenital hydrocephalus, which is a common antecedent to SVS.11,17 It appears that the effect of the CSF absorption abnormality in children changes from one of ventricular dilation to slit ventricles sometime during successful shunt drainage.3

Evidence of Venous Outflow Impairment—MRV

All 3 cases show evidence of intermittent shunt blockage. The lack of ventricular enlargement despite the elevated CSF pressure indicates that there is probably increased brain parenchymal turgor underlying this. The previous discussion implicates venous outflow impairment as the most likely cause of the elevated brain turgor. The CTV in Case 1 shows a hypoplastic left transverse sinus and a very prominent arachnoid granulation in the right transverse sinus (Fig. 1A). Perhaps the inflammation and blockage of the arachnoid granulations that occur in cryptococcal meningitis increase their size and induce venous outflow impairment in some predisposed individuals. The MRV taken concurrently with the CTV shows prominent collateral veins in the scalp and face (Fig. 1B). In Case 2 a very high-grade stenosis is seen in the dominant transverse sinus (Fig. 2D), and in Case 3 there is a high-grade distal sagittal sinus stenosis (Fig. 3B) that is later confirmed by direct manometry (Fig. 3C). Similar to Case 1, Case 3 shows prominent collateral circulation in the face and scalp.

High-grade stenoses have been noted in up to 80% of cases of pseudotumor in adults.13 In a review of 145 children presenting with IIH, Dwyer et al.9 found that 50% had a dominant venous outflow stenosis, with most of these having MRV evidence of substantial nonphysiological collateral circulation. This compares to the controls in whom dominant outflow stenoses were noted in only 4% and collateral veins were even less common. Thus, the MRV data in the current 3 cases appears to be identical to the findings in cases of pseudotumor from the literature: that is, dominant outflow stenoses and prominent collateral flow.

Of perhaps more interest, Dwyer’s group went on to review the MRVs in a similar cohort of children who presented with hydrocephalus, and found significant venous outflow impairment in more than 75% of these children (Dwyer C et al.: Venous sinus obstruction in pediatric hydrocephalus and pseudotumor cerebri. Paper presented at the meeting of the International Society for Hydrocephalus and CSF Disorders in Copenhagen, Denmark, on September 7, 2011). This suggests that venous stenoses can present as either hydrocephalus or pseudotumor in children, possibly depending on the response of the brain parenchyma.1 The 3 cases we have provided suggest that venous hypertension is involved in Rekate Type 2 SVS as well as in Type 3, with only the level of the obstruction of the tube being different.

Evidence of Venous Outflow Impairment—Hemodynamics

In adults with IIH, as the venous outflow becomes more limited, the venous pressure increases. As the venous pressure rises, collateral veins open (already discussed in the MRVs of these patients). Therefore, as the amount of blood passing via collateral vessels increases, the amount of blood returning via the main pathway is reduced. Thus, the percentage of the arterial inflow coming back via the sagittal sinus is a surrogate marker of the degree of the significance of the collateral flow and the venous pressure. In adults with symptomatic IIH, the sagittal sinus returned 35% ± 5% of the inflow compared with the control value of 48% ± 6% (that is, 13% of the inflow returning as collateral flow). Similarly, in symptomatic children with hydrocephalus the sagittal sinus returned 38% ± 11% compared with 51% ± 8% (that is, 13% collateral flow). In the current cohort the sagittal sinuses returned an average of 31% from 5 measurements performed while the patients were symptomatic. In Case 2 the venous return was in the normal range, while the patient was asymptomatic on one occasion, indicating a large change in collateral flow between symptomatic and asymptomatic in this patient. Thus, the hemodynamic findings in this cohort are found to be identical to the pseudotumor literature.

Venous Outflow Impairment: Hydrocephalus or Pseudotumor? The data presented suggest that the 3 cases have small ventricles because they have a syndrome identical to pseudotumor cerebri with an intermittently blocked shunt tube. The question remains, was the venous stenosis acquired due to the inflammation of the enlarged arachnoid granulation. In Cases 2 and 3, however, the stenoses may have been preexisting.

As my colleagues and I have previously noted,3 a large percentage of children with hydrocephalus have venous stenoses. The stenoses appear to behave hemodynamically in an identical manner to those in adults with pseudotumor cerebri. Therefore, it seems likely that the stenoses were always present. Figure 4 is an example of such a case. This child was found to have hydrocephalus on an 18-week antenatal ultrasound. An in utero MRI confirmed communicating hydrocephalus, but there was no evidence of spinal dysraphism (Fig. 4A). An MRV obtained 7 days after birth showed a high-grade stenosis of the distal sagittal sinus (Fig. 4B). An axial T2 slice taken just above the stenosis showed a normal 6-mm equilateral triangle representing the sinus lumen (Fig. 4C), but just below the arrow in Fig. 4B the sinus had collapsed to less than 1 mm (Fig. 4D): that is, a 97% stenosis.
Venous Stenosis or Blocked Arachnoid Granulations?  Thus we return to the question posed by Sainte–Rose et al. some 30 years ago. Intracranial venous sinus hypertension: cause or consequence of hydrocephalus in infants? Are we really looking at the cause of hydrocephalus or some secondary event leading to venous hypertension? The CSF infusion studies done in children with hydrocephalus tend to show an increase in CSF outflow resistance ($R_{\text{out}}$), and this is thought to signify blockage of the arachnoid granulations. If this is true, then the sinus stenosis would be a secondary event. However, the interpretation of infusion studies may be problematic in patients with venous sinuses that collapse. In a recent study, 9 patients with IHH underwent a lumbar infusion study with simultaneous SSS manometry. Mock CSF infusion provoked rises in both CSF pressure and sagittal sinus pressure, with the venous pressure being closely coupled to the ICP, suggesting there was venous collapse providing a functional obstruction.

![Fig. 4.](image)

**Fig. 4.** A: A T2-weighted sagittal image of a fetus with hydrocephalus in utero. B: An MRV showing very poor flow in the distal sagittal sinus (arrow). C: A T2-weighted image of the sagittal sinus taken just above the level of the arrow in panel B, showing the sagittal sinus to be a 6-mm equilateral triangle (arrows). D: A T2-weighted image of the sagittal sinus taken just below the level of the arrow in panel B, showing the sagittal sinus to be a 1-mm equilateral triangle (arrows) giving a 97% stenosis.

Rekate et al. noted, “infants have open fontanelles and sutures that are capable of expanding. Decreased venous return in infants leads to hydrocephalus rather than pseudotumor. After a shunt is inserted, the ventricles return to normal (or even reduced size), the fontanelles close and the skull is no longer distensible. What remains is a drained ventricular system with a brain with increased stiffness or turgor.” So we can see that a previously shunted hydrocephalus can re-present as pseudotumor much later once the shunt has become blocked, because the skull is no longer expandable. This then begs the question “Why do some children re-present with dilated ventricles if the venous sinuses are so prevalent?”

The same group went on to calculate the apparent $R_{\text{out}}$ by assuming that the sagittal sinus pressure was constant during the infusion study (as is the standard technique), and then later corrected the $R_{\text{out}}$ for the sinus pressure. The apparent $R_{\text{out}}$ was overestimated 5-fold compared with the corrected value, which was actually shown to be within the normal range (Pickard JD et al.: CSF dynamics in idiopathic intracranial hypertension: how to estimate the resistance to CSF outflow? Paper presented at the meeting of the International Society for Hydrocephalus and CSF Disorders in Kyoto, Japan, on October 21, 2012). Thus, if you ignore the venous collapse you can overestimate the $R_{\text{out}}$ by an enormous amount in both pseudotumor and possibly also in childhood hydrocephalus, provided the venous sinus pressure varies with the CSF pressure. The latter has been shown to be the case in childhood hydrocephalus. In studies simultaneously measuring the ICP and sagittal sinus pressure in infants with hydrocephalus, the CSF pressure was found to translate directly through the sinus wall so that the sinus pressure directly followed the CSF pressure. It was concluded this was “brought about by a reversible collapse of the sinus wall due to the intracranial hypertension.” The mechanism behind this is further discussed in the Appendix.

Thus, by ignoring the venous collapse in infantile hydrocephalus the pathophysiology could be ascribed to blocked arachnoid granulations rather than the venous pressure itself. In the infusion studies performed by Fried and Shapiro (previously discussed) the children who presented with SVS following shunt malfunction showed a close coupling of the $R_{\text{out}}$ with the ICP over a wide range of pressures; that is, the calculated $R_{\text{out}}$ was dependent on the level the CSF pressure was raised to for each child. The slope of the correlation was 1, indicating that a 1–mm Hg increase in ICP corresponded to an increase in $R_{\text{out}}$ of 1 mm Hg/ml/min. The children who presented with dilated ventricles similarly showed a calculated $R_{\text{out}}$ that increased linearly with ICP, but the slope of the line was less (0.64), indicating that the coupling of ICP and $R_{\text{out}}$ was lower.

This is an unexpected finding in patients with blocked granulations, because in healthy children (with open granulations) the $R_{\text{out}}$ is found to be a constant over a range of ICPs. Blocked granulations also should not vary in resistance due to changes in pressure because they are blocked. As discussed in the Appendix, the answer is that estimating $R_{\text{out}}$ in children with collapsible sinuses by ignoring the venous pressure (similar to that found in pseudotumor, discussed above) overestimates the $R_{\text{out}}$. The degree of sinus collapse is probably a function of the ICP, so the apparent overestimation in $R_{\text{out}}$ will increase as the ICP is increased (there is progressive vein collapse) during the infusion study in these children, giving the results Shapiro and Fried found. The difference between whether a child with venous collapse re-presents with slit ventricles or dilated ventricles appears to depend on whether the sinuses collapse quickly as pressure rises, giving a higher venous pressure and higher brain turgor (SVS), or whether the sinuses collapse slowly; the sinus pressure thus lags behind the ICP to a greater degree, and the brain turgor is less (dilated ventricles).
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Fig. 5. A graph of ICP versus infusion rate plotted with feedback fractions of 0%, 70%, 80%, and 90%.

Appendix

Lumbar infusion studies are based on the Davson equation regarding ICP; that is

\[ ICP = R_{\text{out}} \times FR_{\text{CSF}} + P_{\text{SSS}} \]  

where \( R_{\text{out}} \) is the resistance to CSF outflow, \( FR_{\text{CSF}} \) is the rate of CSF formation, and \( P_{\text{SSS}} \) is the SSS pressure. If we insert normal values from the literature into Eq. 1, using an \( R_{\text{out}} \) of 7.5 mm Hg/ml/min, \( FR_{\text{CSF}} \) of 0.4 ml/min, and \( P_{\text{SSS}} \) of 7 mm Hg we can see the ICP would be 10 mm Hg. In practice \( R_{\text{out}} \) is measured by artificially elevating \( FR_{\text{CSF}} \) with a constant lumbar infusion performed at several known rates. It can be shown by simple arithmetic that the slope of the resultant line obtained when ICP is graphed against the infusion rate is the \( R_{\text{out}} \), provided the PSSS does not change during the test; that is, it is a constant. The Davson equation must be slightly modified to account for the infusion rate (I):

\[ ICP = R_{\text{out}} \times (FR_{\text{CSF}} + I) + P_{\text{SSS}} \]  

If the assumption that the PSSS is a constant is incorrect, then we need to insert a variable to account for this. To simplify the mathematics we will initially assume that the PSSS follows the ICP as a constant percentage of the ICP once the resultant PSSS rises above the nominal baseline of 7 mm Hg (the feedback fraction \( F_f \)). Thus PSSS would become ICP \( \times F_f \), and Eq. 2 becomes

\[ ICP = R_{\text{out}} \times (FR_{\text{CSF}} + I) + ICP \times F_f \]  

Solving for the ICP we get

\[ ICP = R_{\text{out}} \times (FR_{\text{CSF}} + I)/(1 - F_f) \]  

We can insert the normal values for \( FR_{\text{CSF}} \) (0.4) and \( R_{\text{out}} \) (7.5) and plot the ICP versus I, with a variable \( F_f \), and the resultant graph is Fig. 5. The PSSS cannot be less than 7 mm Hg, so if ICP \( \times F_f \) \( \geq \) 7 then Eq. 4 applies, and if ICP \( \times F_f \) \( \leq \) 7 then Eq. 2 applies. Thus with an \( F_f \) of zero, Eq. 2 applies and the slope of \( R_{\text{out}} \) is 7.5. If the PSSS is made to be 70% of the ICP, then \( F_f \) is 0.7 and the line is much steeper. The estimated \( R_{\text{out}} \) is 3 times the actual \( R_{\text{out}} \) if the PSSS is thought erroneously to be a constant. At higher feedback fractions the slope of the line becomes 5 and 10 times normal (80% and 90% feedback, respectively). In addition, the resting pressure rises above 10 mm Hg as the \( F_f \) goes above 70%, because the pressure gradient from the ICP to PSSS must reach a minimum of 3 mm Hg for CSF to drain and the pressures to stabilize. In actually, the feedback fraction is unlikely to be a constant over all ICPs. This is because the veins are fully open at low ICP and progressively collapse at higher pressures. Thus, the feedback fraction would be zero at low pressures and increase from there to a maximum at higher pressures. The speed of this change in the feedback fraction depends on the compliance of the walls of the veins themselves. Thus, in the case of the children with SVS investigated by Shapiro and Fried,2 the rapid change in calculated \( R_{\text{out}} \) would indicate a rapid collapse in the vein wall with a change in ICP, and thus a higher overall venous pressure at any ICP and a greater brain turgor with less venous expansion. In the children with dilated ventricles after shunt failure the \( R_{\text{out}} \) did not change as quickly with a change in ICP. Thus, the venous pressure lagged behind the ICP to a greater degree, the brain turgor was less, and so the ventricles dilated.

Disclosure

The author reports no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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