Subtle deterioration in shunted childhood hydrocephalus

A biomechanical and clinical profile

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Eighteen hydrocephalic children who presented with subtle deterioration when their shunts malfunctioned were studied during shunt revision by means of the pressure-volume index (PVI) technique. Bolus manipulation of cerebrospinal fluid (CSF) was used to determine the PVI and the resistance to the absorption of CSF (R0). Ventricular size was moderately to severely enlarged in all the children. Steady-state intracranial pressure (ICP) at the time of shunt revision was 17.5 ± 7.3 mm Hg (range 8 to 35 mm Hg). Pressure waves could not be induced by bolus injections in the 8- to 35-mm Hg range of ICP tested. The mean ± standard deviation (SD) of the predicted normal PVI for this group was 18.5 ± 2.7 ml. The mean ± standard error of the mean of the measured PVI was 35.5 ± 2.1 ml, which represented a 187% ± 33% (± SD) increase in volume-buffering capacity (p < 0.001). The ICP did not fall after bolus injections in three children, so that the R0 could not be measured. In the remaining 15 patients, R0 increased linearly as a function of ICP (r = 0.74, p < 0.001). At ICP's below 20 mm Hg, R0 ranged from 2.0 to 5.0 mm Hg/ml/min, but increased to as high as 21 mm Hg/ml/min when ICP was above 20 mm Hg.

This study documents that subtle deterioration in shunted hydrocephalic children is accompanied by abnormally compliant pressure-volume curves. These children develop ventricular enlargement and neurological deterioration without acute episodic pressure waves. The biomechanical profile of this group differs from other children with CSF shunts.

KEY WORDS • hydrocephalus • shunt dependency • pressure-volume index • cerebrospinal fluid absorption

In an earlier report, we described neural axis biomechanics in hydrocephalic children who had rapidly deteriorated after malfunctions of their shunts. Occlusion of the shunts was accompanied by signs of raised intracranial pressure (ICP), namely emesis, irritability, and rapid deterioration of consciousness. While many of these children had premonitory symptoms of headache which abated spontaneously and probably indicated intermittent shunt malfunction, relatively abrupt clinical deterioration occurred after the shunts occluded totally. The symptoms were so overt in this group, labeled "acute deteriorators," as to make clinical diagnosis obvious to the parents and their physicians. Shunt revisions reversed all symptoms of raised ICP within 48 hours. The biomechanical studies in these children demonstrated waves of intracranial hypertension which were precipitated by small changes of intracranial volume at relatively low baseline ICP. We believed that these findings accounted for the abrupt clinical deterioration characteristic of this group.

This earlier report was derived from a consecutive series of all children admitted to the hospital for evaluation of cerebrospinal fluid (CSF) shunt malfunction. All were sequentially entered into a study to characterize neural axis biomechanics performed during shunt revisions. As this study progressed, we encountered children whose clinical course and biomechanical profiles differed from the group described above. In contrast to the majority of shunt-dependent children who deteriorated rapidly after shunt occlusion, 20% of children with malfunctioning shunts exhibited no overt signs of raised ICP. In some children, the identification of inadequate shunt function often went unrecognized until deterioration in schoolwork or behavioral problems prompted neurosurgical evaluation. Other cases were recognized only when routine computerized to-
mography (CT) scans showed a change in ventricular size or failure of the ventricles to diminish in size after a shunt procedure. In a few children, accelerated head growth was documented on a routine examination and led to the diagnostic CT scan. Some children had been followed jointly by other physicians who had assured parents that the persistent ventricular enlargement was a sign of shunt independence. These opinions led some parents to miss scheduled follow-up appointments with neurosurgeons. Because malfunction of the shunt had not been obvious to the parents, it was impossible to document when occlusion had occurred. Since all these children were toddlers or older, routine follow-up review consisted of either semiannual or annual neurosurgical evaluations so that the malfunctions could have occurred anywhere within the 6- to 12-month period between evaluations. Interestingly, this group of "subtle deteriorators" showed dramatic anatomical and functional changes after the shunt revision. Most showed reduction in ventricular size, and many reverted toward normal or slit-like ventricles. Although all showed some behavioral improvement, the degree of neuropsychological improvement after shunt revision varied.

As we recognized this group of children, they were prospectively separated from "acute deteriorators" on the basis of their clinical presentation to determine whether biomechanical characterization could differentiate these two groups. We used the PVI technique of bolus manipulation of CSF to characterize the neural axis volume buffering capacity (PVI) and the resistance to the absorption of CSF (Ro) in shunted children with subtle deterioration after shunt malfunction.

Clinical Material and Methods

Shunt malfunction was suspected in 18 shunted children (aged 2½ to 15 years) based on loss of developmental milestones, deteriorating neuropsychological function, chronic papilledema, CT scans showing large ventricles, or increasing head circumference. Deterioration of intellectual function was not apparent to the parents of 12 of the 18 children, but was suspected by their teachers at school. None of the children had acute symptoms of intracranial hypertension such as lethargy, vomiting, or sudden severe headache. Shunt taps were performed to confirm that the shunt was occluded. The proximal end was occluded in eight children and the distal catheter in 10 children.

The size of the ventricles was graded using the ventricular/biparietal ratio, which was obtained by dividing the ventricular span through the body of the lateral ventricles by the width of the brain at this level. Before the shunt malfunction, CT scans had shown that four children had normal-sized ventricles, nine had moderately enlarged ventricles, and five had markedly enlarged ventricles. Most of the children comprising this group, therefore, had had enlarged ventricles long before shunt malfunction was present or suspected.

Pressure-volume studies were performed with the children under general endotracheal anesthesia for revision of the shunt. No premedication was given and isoflurane was used as the anesthetic agent because of its minimal effect on cerebral hemodynamics and ICP. The blood pressure and pulse were recorded throughout the study period and did not change in any child. If needed, the ventricular portion of the shunt was revised and connected via rigid saline-filled tubing to a strain gauge transducer in order to measure ICP. The ventricular fluid pressure was referenced to the right atrium and recorded on conventional strip charts at 1 mm/sec. All pressure measurements were extracted from the strip chart recordings using the diastolic component of the cardiac and respiratory cycles.

After establishing an initial steady-state intracranial pressure (P), withdrawal of a 1- to 3-ml bolus of CSF (ΔV) was performed, and the immediate pressure response after withdrawal (P) was recorded. The PVI was calculated using the formula PVI = ΔV/log (P/P0) Further withdrawals of CSF were performed as needed to reduce ICP to 8 mm Hg. Using this measured PVI, a safe volume for subsequent bolus injections was calculated which would ensure that peak ICP (Pp) did not exceed 25 to 35 mm Hg. Multiple bolus injections of 1 to 7 ml mock CSF were performed in each child at a rate of 1 ml/sec to measure PVI and Ro, through a range of ICP's from 8 to 35 mm Hg. After each injection, the following parameters were extracted: 1) Pp, the ICP prior to bolus injection; 2) Pp, the peak ICP immediately following the injection; and 3) Pp, the ICP at 1 minute after the injection. The PVI was calculated using the formula PVI = ΔV/log (P/P0) Resistance to the absorption of CSF (Ro) was calculated using the formula: Ro = P/PVI × log [(P/P0) × (P0 - P)/P0].

Predicted normal PVI was compared to the measured PVI in each child. Predicted normal PVI was determined by measuring head circumference and length of the spinal axis and extracting predicted normal PVI from a previously established nomogram. The measured Ro was compared with values previously determined in normal children using the bolus injection technique.

Results

The ventricles were enlarged in all 18 children. At the time of shunt revision, the ventricles were severely enlarged in 15 children (83%) and moderately enlarged in three (17%). Compared to the ventricular size prior to the present shunt malfunction, the ventricles enlarged further in 11 children (61%) and were unchanged in seven (39%). After the shunts were revised, ventricular size decreased in 16 children (89%). In 12 (67%), the follow-up CT scans showed normal or subnormal ventricular size. Although formal neuropsychological testing was not performed on all children, intellectual performance improved after the revision in 10 (56%) as judged by teachers and parents.
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FIG. 2. Graph showing the predicted versus the measured pressure-volume index (PVI) in the 18 "subtle deteriorators" studied. The shaded area represents the 95% confidence band for predictability of PVI in normal children. The measured PVI in each hydrocephalic child exceeded the predicted normal for that child.

FIG. 1. Strip chart recordings showing the response of intraventricular pressure to successive bolus fluid injections (ΔV) of cerebrospinal fluid (CSF) in a hydrocephalic child with a shunt who showed subtle deterioration. The steady-state intracranial pressure (ICP) at shunt revision was 35 mm Hg; ICP was initially reduced by bolus withdrawal of CSF. The measured pressure-volume index (PVI) was increased to 187% + 33% (+ SD) in measured PVI compared to predicted normal values for these children. There was no correlation between the measured ventricular size (ventricular/biparietal ratio) and the measured PVI among this study group (p > 0.1). In most of the children studied, the PVI was relatively constant from one perturbation to the next. However, in some children the values varied considerably. In order to quantitate this variation from one bolus manipulation to the next, PVI variation was calculated in each child by dividing the mean PVI determined for each child into the SD (SD/mean PVI) to determine the variance. In seven children, the variation in PVI was 13.0% ± 5.6%. In the other 11 children, the variation in PVI was 29% ± 7.0%. In these children the measured PVI appeared to increase as bolus injections elevated ICP through the range of 8 to 35 mm Hg. However, when the pressure response to multiple bolus injections was plotted, the pressure-volume relationship described an exponential curve in all of these children.

In three of the 18 children studied, ICP did not decrease after each bolus injection so that the Ro could not be calculated. In the remaining 15 children, Ro was not constant through the ICP range of 8 to 35 mm Hg. With successive bolus fluid injections, Ro increased linearly as a function of ICP (r = 0.74, p < 0.001). At ICP's below 20 mm Hg, Ro ranged from 2.0 to 5.0 mm Hg/ml/min, but increased to as high as 21 mm Hg/ml/min when ICP exceeded 20 mm Hg (Fig. 3).

At the time of shunt revision, the mean steady-state ICP (+ standard deviation (SD)) was 17.5 ± 7.3 mm Hg (range 8 to 35 mm Hg). Intracranial pressure in eight children was 15 mm Hg or less. There was no correlation between ventricular size and the steady-state ICP among these children. Episodic increases of ICP were not observed, nor could they be induced by injections of fluid (Fig. 1).

The mean predicted normal PVI calculated for the entire group was 18.5 ± 2.7 ml (+ SD). The mean ± standard error of the mean of the measured PVI for all children studied was 35.5 ± 2.1 ml. Using the paired t-test, the difference between predicted normal and measured PVI was statistically significant (p < 0.001). As seen in Fig. 2, the measured PVI for each child was beyond the 95% confidence band for predictability of PVI in normal children. The elevated PVI for this group represented an increase of 187% ± 33% (+ SD) in measured PVI compared to predicted normal values for these children. There was no correlation between the measured ventricular size (ventricular/biparietal ratio) and the measured PVI among this study group (p > 0.1). In most of the children studied, the PVI was relatively constant from one perturbation to the next. However, in some children the values varied considerably. In order to quantitate this variation from one bolus manipulation to the next, PVI variation was calculated in each child by dividing the mean PVI determined for each child into the SD (SD/mean PVI) to determine the variance. In seven children, the variation in PVI was 13.0% ± 5.6%. In the other 11 children, the variation in PVI was 29% ± 7.0%. In these children the measured PVI appeared to increase as bolus injections elevated ICP through the range of 8 to 35 mm Hg. However, when the pressure response to multiple bolus injections was plotted, the pressure-volume relationship described an exponential curve in all of these children.

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In contrast to acutely deteriorating shunt-dependent children whose symptoms are overt and most readily detected by parents, the identification of shunt mal-function in the children reported here was often undetected by their parents and recognized only by school-teachers or routine neurosurgical evaluation.

Intuitively, the easiest way of identifying inadequate shunt function is to measure steady-state ICP by tapping the shunt. However, the steady-state ICP measured at the time of shunt revision ranged from 8.0 to 35.0 mm Hg among this group. Eight of the 18 children had ICP below 15 mm Hg at shunt revision. In the other 10 children, initial steady-state ICP was elevated (15 to 35 mm Hg). This latter group of children presented (as did those with lower ICP) with subtle insidious neuropsychological deterioration, but without acute symptoms or the changes in mental status characteristic of the acutely deteriorating shunt-dependent child.

The children with relatively normal ICP resemble those included by Di Rocco, et al.4 They described 18 children with CSF shunts who fulfilled the clinical and radiological criteria of shunt-independent arrest of hydrocephalus. Although the ICP measured in their children was below 14 mm Hg, eight of them had abnormal constant CSF infusion tests indicating impaired absorption of CSF, or had episodes of intracranial hypertension documented by continuous ICP monitoring. These findings were interpreted to show that hydrocephalus was still active despite the normal steady-state ICP measured initially. Further evidence that the hydrocephalus was not arrested was the demonstration of improved neuropsychological performance after shunt revision in the patients described by Di Rocco, et al.,4 as well as in most of our children with subtle deterioration.

The most striking difference between this group of "subtle deteriorators" and a group of "acute deteriorators" reported earlier24 was the absence of waves of intracranial hypertension in these patients.3 In the children who deteriorated acutely after occlusion of their shunts, we found that waves of elevated ICP occurred spontaneously or could be induced by bolus manipulation of CSF. These pressure waves occurred only after the patient's ICP rose to the beginning of the steep portion of their pressure-volume curve. The pressure-volume curve in each of these "acute deteriorators" was normal for body size.24 We concluded that the normal steepness of the pressure-volume curves in these small children made them vulnerable to ICP instability when compliance diminished solely as a function of the exponential nature of the pressure-volume curve.

In the children described in this study of subtle deterioration after shunt malfunction, the measured PVI was 187% of predicted value. This marked increase in PVI allows ICP to rise slowly along an abnormally compliant pressure-volume curve. These curves are so compliant that, despite the relatively high ICP documented in some children, they still operated on the relatively horizontal portion of their pressure-volume curves. In order to reach the steeper vulnerable portion of their pressure-volume curves at which ICP instability might occur, ICP would have to exceed 50 mm Hg.

The various factors that can alter the neural axis volume-buffering capacity or measured PVI include the collapsible venous pool and the biomechanical properties of the brain parenchyma.18 Since an expansion of the venous pool is unlikely in these shunted children, we postulate that the increased PVI is caused by changes in the biomechanical properties of the brain parenchyma that accompany the hydrocephalic process. Many of the well-documented morphological changes in the hydrocephalic brain, which include axonal disruption and loss of myelin, may correlate with the increased volume-buffering capacity documented in this study.21,22,33 Similar anatomical and biomechanical alterations (elevated PVI) are found in untreated hydrocephalic infants.25,28,32 However, the increased ventriculostomy...
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ular volume alone does not explain the finding of elevated PVI. In this group as well as in untreated hydrocephalic infants, there was no correlation between PVI and ventricular size. It is unclear to us whether these biomechanical changes reverse after treatment and reconstitution of the cortical mantle, or whether the children reported herein have always had enhanced volume-buffering capacity.

The resistance to the absorption of CSF measured in these children is similar to that documented in the group of children with acute deterioration after shunt malfunctions. Rather than augmenting the rate of CSF absorption at higher ICP, 

the resistance to the absorption of CSF (R_e) increases linearly as a function of ICP. Similar findings have been reported in other clinical studies using the bolus technique, and are consistent with profiles obtained by constant-infusion techniques in other settings. All of these studies show the pattern of CSF absorption described by Lorenzo, et al., who used perfusion techniques.

This linkage between outflow resistance and ICP may lead to persistent shunt dependency in either the acute or subtle form of clinical presentation. In both groups of children with CSF shunts, the CSF absorptive reserve that is normally present and is found in untreated hydrocephalic patients is lost; thus, ICP will rise along a pressure-volume curve unique to each group of children and determine the clinical presentation accompanying suboptimal shunt function. The ICP, however, will not reach a new steady state after shunt malfunction because the R_e increases in a stepwise fashion as CSF accumulates (Fig. 3).

The biomechanical profile of the hydrocephalic children with CSF shunts described in this study serves to explain why these children are shunt-dependent despite the absence of acute and overt symptoms. When compared to normal children and to infants with active hydrocephalus, the enhanced volume-buffering capacity (elevated PVI) and the R_e, which increases with ICP, were observed in untreated hydrocephalic patients and in children with normal ICP; thus, ICP will rise along a pressure-volume curve unique to each group of children and determine the clinical presentation accompanying suboptimal shunt function. The ICP, however, will not reach a new steady state after shunt malfunction because the R_e increases in a stepwise fashion as CSF accumulates (Fig. 3).

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While this can identify those shunt-dependent children with transient or sustained elevations of ICP, the method will fail to identify many of the children in this group who are protected from elevated ICP by their abnormally high neural axis volume-buffering capacity. Since the present study did not include any children with arrested hydrocephalus, we cannot offer a biomechanical profile which identifies this group of patients. However, we would be reluctant to consider hydrocephalus in a child as arrested if biomechanical testing revealed the pattern described in this report. In our group of almost 500 hydrocephalic children, we have encountered only two children who have maintained adequate function after the shunt was removed. Both children had a PVI appropriate for estimated size of neural axis and outflow resistance in the normal range.

Since we cannot describe the biomechanical profile of shunt-independent arrest of hydrocephalus, we suggest that a biomechanical profile including increased PVI, resistance to the absorption of CSF (R_e) linked to ICP, and a normal to slightly elevated steady-state ICP without pressure waves signifies a child who continues to rely on shunting to maintain optimal function. While these studies offer a profile of the shunt-dependent child with subtle deterioration, they also indicate that careful follow-up monitoring of all hydrocephalic children with CSF shunts is necessary to identify children whose shunts malfunction without overt signs.

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References

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