Magnetic resonance imaging quantification of compliance and collateral flow in late-onset idiopathic aqueductal stenosis: venous pathophysiology revisited

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Object. Findings in animal models of noncommunicating hydrocephalus have suggested that a reduction in compliance of the superior sagittal sinus, an elevation in venous outflow pressure, and the development of venous collateral flow may be associated with this condition. Although elevated venous pressure is known to cause hydrocephalus in children, this mechanism has fallen out of favor as a theory in adults.

Methods. Twenty-one patients with late-onset idiopathic aqueductal stenosis (LIAS) underwent magnetic resonance imaging with flow quantification measuring the degree of ventricular enlargement, sulcal compression, total blood inflow, superior sagittal/straight sinus outflow, aqueduct flow, arteriovenous delay (AVD), and the extent of collateral venous flow. Data obtained in these patients were compared with those obtained in 21 age-matched control individuals.

Results. There was a reduction in compliance in the patients with LIAS in whom the AVD decreased by 50% (p = 0.01). The arterial inflow and the straight sinus outflow were normal, but the sagittal sinus outflow was reduced by 23% (p = 0.001). This indicated that significant collateral venous outflow pathways were draining blood away from the superficial but not the deep drainage system.

Conclusions. Similar to the animal models, patients with LIAS exhibit a reduced venous compliance and an elevation in venous collateral flow. Together, these findings suggest that an elevation in venous pressure may be associated with this disease process. A review of the literature has indicated that only subtle differences may exist in the pathophysiology among patients with LIAS, normal-pressure hydrocephalus, and idiopathic intracranial hypertension. (DOI: 10.3171/JNS-07/11/0951)

Key Words • aqueductal stenosis • compliance • idiopathic intracranial hypertension • normal-pressure hydrocephalus • sagittal sinus

Patients who present with a benign, nontumor-related aqueductal stenosis can be separated into two groups in terms of the age at presentation. In early life, the presentation is that of an acute, rapid increase in ventricle size, but in later life patients present with a more chronic picture, suggestive of a balanced form of hydrocephalus. The chronic form of obstructive hydrocephalus has been termed LIAS. Patients with LIAS typically present with a spectrum of features. Younger patients tend to experience headaches, but older patients tend to have NPH-like symptoms (ataxia, cognitive impairment, and incontinence). Older patients have larger ventricles than younger patients.

Late-onset idiopathic aqueductal stenosis has been modeled in dogs in which kaolin clay is injected into the ventricular system. This results in an intense inflammation of the meninges and the formation of connective and fibrous tissue, which leads to obstruction of the fourth ventricle outlet. Although it may be expected that such a maneuver would cause ventricular enlargement due to an elevation in intraventricular pressure over and above the subarachnoid pressure (that is, a pressure gradient), these pressure gradients are transient at best. No consistent pressure differences have been found between the ventricle, brain, and subarachnoid space either before kaolin injection or after. Kaolin obviously affects CSF reabsorption, but the absence of a transmantle pressure gradient, despite the obvious dilation of the ventricles, suggests that the blockage of CSF absorption must exist elsewhere along the CSF resorption pathway. The final common pathway for CSF resorption is into the venous system, and an elevation in venous pressure will reduce the CSF resorption driving pressure. With this in mind, it has been shown that the sagittal sinus pressure is elevated in these animal models of hydrocephalus. When the sagittal sinus pressure/volume response was tested by direct injection of fluid into the proximal sagittal sinus, an elevation in elastance (reduced compliance) was found. The noted elevated venous pressures were shown to be hemodynamically significant by being associated with accessory venous or collateral pathway filling.

In children it has been well established that communicating hydrocephalus can be caused by venous hypertension. Shulman and Ranoshoff found, in 15 hydrocephalic
children, that there was an elevation in CSF and SSS pressure with a loss of the driving force across the arachnoid villi. Sainte-Rose et al.\(^{10}\) reported a raised sagittal sinus pressure in hydrocephalic infants, both in patients with an interruption in venous outflow, such as in cases of achondroplasia or craniosynostosis, and even in those without an interruption of venous outflow.\(^{10}\) In adults, however, elevated venous sinus pressures cause idiopathic intracranial hypertension rather than hydrocephalus (that is, raised CSF pressure but nondilated ventricles).\(^ {22}\) It has been noted that whether a patient presents with hydrocephalus or intracranial hypertension is an age-related factor and seems to depend on whether the cranial sutures are patent or closed.\(^ {18}\) Thus, in adults the idea that there is a correlation between hydrocephalus and an elevation in venous pressure has fallen out of favor, but it has not been disproven. With this in mind, the purpose of this study was to test the hypothesis that the venous hemodynamics of clinical LIAS are similar to those demonstrated in animal models—that is, a decrease in venous compliance and an elevation in venous pressure, with both of these encouraging collateral outflow pathways.

**Clinical Material and Methods**

**Patient Population**

Twenty-one patients found to have triventricular dilation at a tertiary referral hospital between November 1998 and May 2006 were prospectively entered into the study. There were 11 men and 10 women whose mean age was 52 ± 17 years (± SD). Review of the planar MR images allowed cases to be divided on the basis of whether there was evidence of sulcal effacement over the vertex. In the noncompression group there were 14 patients—nine men and five women—whose mean age was 56 ± 13 years (± SD). In the compression group there were seven patients—two men and five women—whose mean age was 44 ± 22 years (± SD). The control group consisted of 21 individuals who were predominantly either the spouses of patients or volunteers. I confirmed that the control cases were free of chronic headache and cognitive symptoms. The control group consisted of 11 men and 10 women whose mean age was 54 ± 15 years (± SD). Informed consent was obtained from all patients, and the hospital ethics committee approved the study protocol.

**Magnetic Resonance Imaging Studies**

All patients underwent neuroimaging in a 1.5-tesla superconducting magnet MR imaging unit (Magnetom Vision, Siemens). I used standard sagittal T1-weighted, axial T2-weighted, and axial fluid attenuated inversion recovery images as well as MR imaging flow quantification sequences. A retrospectively cardiac gated phase contrast flow quantification sequence was used (TR 29 msec, TE 7 msec, flip angle 30°, slice thickness 6 mm, matrix 192 × 512, field of view 200, and a single NEX [a standard sequence available on this scanner]). The velocity-encoding values of 20 cm/second, 40 cm/second, and 75 cm/second were used. The lower velocity-encoding value was selected to maximize the measurement of the aqueduct, with the mid value used for the venous sinus measurements and the higher one used to maximize the arterial measurements.

The plane of section was selected to intersect the basilar artery and the cavernous portion of the internal carotid arteries according to the literature.\(^ {7}\) The planar imaging results, as well as the flow quantification raw data, were archived on a magneto-optical disc. Regions of interest were placed around the carotid arteries, basilar artery, the sagittal sinus, straight sinus, and the aqueduct in each patient. Care was taken to exclude aliasing by retrospectively manipulating the base lines of each resultant graph, giving an effective arterial flow limit of 150 cm/second.

The addition of the flow from the three arteries gave the total supratentorial blood inflow. The sagittal sinus and straight sinus outflow was obtained from the region of interest placed around each of these vessels. The percentage of the arterial inflow represented by the sagittal sinus and straight sinus outflow was calculated. The AVD is the time taken between the center of the arterial pulse and the center of the venous outflow pulse (measured at half height) when both are measured simultaneously, and it is inversely proportional to the pulse wave velocity between these two points. The aqueductal flow was reviewed to confirm aqueductal obstruction in all of the patients and patency in the controls. The ventricle/cerebral index (Evans index) was obtained as the ratio of maximal width of both anterior horns of the ventricles to the diameter of the inner table of skull along the line of measurement of the anterior horn transverse dimension (expressed as a percentage).

**Statistical Analysis**

Mean and SDs were obtained for each group of patients. Differences between the groups were tested using a non-paired Student t-test.

**Results**

The ventricular size, compliance, and blood flow data are summarized in Table 1.

**Clinical and Morphological Findings**

Of the 21 patients with LIAS, 12 presented with headaches as the initial symptom and nine presented with NPH-type symptoms (predominantly ataxia with some having mild memory impairment). In all patients I observed ventricular enlargement involving the lateral ventricles and the third ventricle. The mean Evans index was increased by 13% (p = 0.0001). The planar images showed occlusion of the aqueduct but no evidence of tumor or extrinsic compression in all patients (Fig. 1). Seven patients were noted to have loss of the sulci over the vertex (Fig. 2) and in 14 there was no evidence of significant compression (Fig. 3). The ventricles had enlarged 10% more individuals in the compression group than the noncompression group (p = 0.01).

**Hemodynamic Findings**

The AVD showed an overall 50% reduction in compliance between patients with LIAS and controls (p = 0.01). There was no difference in the arterial inflow volumes between patients and controls, but I did observe a 23% reduction in sagittal sinus outflow in patients with LIAS (p = 0.0001). The straight sinus outflow was not significantly different between patients and control individuals.
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TABLE 1
Summary of ventricular size, compliance, and blood flow*

<table>
<thead>
<tr>
<th>Variable</th>
<th>No. of Cases</th>
<th>Age (yrs)</th>
<th>Evans Index (%)</th>
<th>AVD (msec)</th>
<th>Cerebral Inflow (ml/min)</th>
<th>Outflow (ml/min)</th>
<th>% of Inflow</th>
</tr>
</thead>
<tbody>
<tr>
<td>control</td>
<td>21</td>
<td>54 ± 15</td>
<td>33 ± 2</td>
<td>106 ± 70</td>
<td>770 ± 140</td>
<td>350 ± 72</td>
<td>95 ± 24</td>
</tr>
<tr>
<td>all LIAS</td>
<td>21</td>
<td>52 ± 17</td>
<td>46 ± 9</td>
<td>53 ± 52</td>
<td>740 ± 150</td>
<td>270 ± 70</td>
<td>86 ± 27</td>
</tr>
<tr>
<td>p value†</td>
<td>0.64</td>
<td>0.0001</td>
<td>0.01</td>
<td>0.56</td>
<td>0.001</td>
<td>0.23</td>
<td>0.005</td>
</tr>
<tr>
<td>LIAS group</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>noncompressed</td>
<td>14</td>
<td>56 ± 13</td>
<td>43 ± 6</td>
<td>52 ± 60</td>
<td>740 ± 190</td>
<td>270 ± 80</td>
<td>82 ± 21</td>
</tr>
<tr>
<td>compressed</td>
<td>7</td>
<td>44 ± 22</td>
<td>53 ± 10</td>
<td>56 ± 30</td>
<td>740 ± 240</td>
<td>280 ± 48</td>
<td>94 ± 36</td>
</tr>
<tr>
<td>p value†</td>
<td>0.14</td>
<td>0.01</td>
<td>0.84</td>
<td>0.97</td>
<td>0.84</td>
<td>0.35</td>
<td>0.65</td>
</tr>
</tbody>
</table>

* Except for p values, data are presented as the means ± SDs. Abbreviation: ST = straight sinus.
† Determined using the Student t-test.

No significant differences were noted in any of the hemodynamic parameters between patients in whom the sulci were compressed and those in whom the sulci were not compressed.

Discussion

Compliance or the pressure–volume index is essentially a measure of the degree of stiffness of the walls delimiting the container surrounding the CSF. Compliance is the reciprocal of elastance, a measure that has been shown to be significantly correlated with clinical improvement following third ventriculostomy in patients with LIAS despite the elastance being unchanged postoperatively.45 The formal measurement of compliance is difficult to perform in vivo because it requires the invasive measurement of the pressure rise, which occurs following an increase in CSF volume instilled via an infusion. The compliance of the thoracoabdominal arterial tree is noninvasively estimated by measuring the time the pulse wave takes to travel the length of the aorta, this being inversely proportional to the compliance of the aorta.44 Similarly, the intracranial compliance has been estimated by measuring the time the pulse wave takes to travel from the arterial to the venous side of the cerebral circulation,7 which is termed the AVD. In cases of NPH, the compliance, based on the AVD and other measures, is reduced by 50%.7,28 In the present study, a reduction in compliance of 50% in LIAS was also noted (identical to NPH). The intracranial venous structures provide 80% of the vascular capacitance,10 and therefore it is likely that the reduction in vascular compliance measured in LIAS is predominantly from the venous side of the vascular tree rather than the arterial side. Thus, the findings in my study mirror those documented in the animal models of LIAS—that is, a reduction in venous compliance.33

In the animal model, previously described, investigators also noted an elevation in venous sinus pressures in aqueductal stenosis.33 Unfortunately, pressure cannot be measured in absolute terms in a noninvasive fashion. One of the outcomes of the pressure elevation noted in the sinuses of the animal models was an increase in collateral venous flow.33 In IIH venous sinus pressure gradients have been shown to be elevated approximately 10-fold that of normal gradients,24 and these elevated pressures open collateral vessels. This collateral flow has been measured as a reduced percentage of the sagittal sinus outflow (compared with inflow) of 32% in IIH compared with 47% in controls (p < 0.0001).1 Similarly, in the straight sinus a percentage return of 9% in IIH compared with 14% in controls (p < 0.0001) has indicated evidence of collateral flow in the deep system.3 In the present study, the total arterial inflow in the patients was normal, indicating that the blood flowing into the capillary bed of the brain drained by the sagittal sinus was also probably normal (or another area of the brain would have to have been hyperemic to make up the shortfall). The 38% of blood returning via the SSS in LIAS and 46% returning in the controls is almost identical to that found in IIH, underscoring a similar increase in collateral flow in LIAS and IIH. Of note, there was no evidence of collateral flow occurring in the deep venous system in LIAS (unlike in IIH), indicating that the mechanism responsible for this alteration is limited to the superficial system. Does an elevation in venous pressure, as found in IIH, also occur in the superficial veins of LIAS? An elevation in venous pressure would be a valid hypothesis to account for these findings. An elevation in venous pressure in conjunc-

![FIG. 1. Sagittal T1-weighted MR image obtained in a patient with LIAS, showing a dilated upper aqueduct and obstructed lower aqueduct.](image-url)
tion with normal aging has been suggested to be partly accountable for the senescent reduction in CSF resorption noted, but there have been no direct measurements of venous pressures in adult patients with LIAS. The data of both reduced compliance and elevated collateral flow provides strong circumstantial evidence of elevated pressures in LIAS, with other potential causes for these phenomena being less likely.

One may ask at this point whether the reduced venous compliance and elevated venous collateral flow are the causes of the ventricular dilation or an effect of compression of the veins and brain parenchyma by the dilated ventricles? First, if the ventricles were compressing the veins and elevating the pressure, then one would expect the collateral flow to come from the deep and the superficial system, but this was not the case. Second, one would expect that in patients with much larger ventricles and compressed sulci there should be a greater reduction in compliance and further elevation in collateral flow, but, as I documented (see Results), the hydrodynamic variables were identical regardless of the apparent ventricular size or extent of compression. It could be argued that enlarged ventricles may directly compress the parenchymal veins even before the subarachnoid space. Figure 4 provides a scatter plot showing no evidence of a relationship between ventricular enlargement and sagittal sinus flow in LIAS. Thus, it would appear that the findings are not merely due to the ventricular enlargement.

Pathophysiological Similarities of LIAS and NPH

Both NPH and LIAS are associated with an identical reduction in the AVD. Following shunt insertion, SSS flow in patients with NPH has been shown to increase by 28% whereas the straight sinus blood flow remained unaltered, suggesting a pathophysiology maximally affecting the superficial venous territory. In a larger cohort of 20 patients with NPH (incorporating the patients from the original study and others) I observed a reduced rate of sagittal sinus outflow compared with the inflow of 36%; it was 44% in the age-matched controls (p = 0.03). The straight sinus flow was unchanged at 13% in both groups (unpublished data). This further suggests a difference between the superficial and deep territories. Elevations in cortical venous impedance have been shown in NPH, and these are reduced after shunt insertion. Direct measurement of pressures in a model of communicating hydrocephalus has shown that cortical vein pressures were markedly elevated and sagittal sinus pressures moderately elevated in dogs in which there was a naturally occurring form of hydrocephalus. These findings, taken together, appear identical to those in LIAS and suggest that pressure and flow changes occur in the superficial venous system in NPH as well. Although the idea that the physiology associated with LIAS and NPH may be identical seems unlikely, there is evidence in the literature to suggest that this may well be the case. On theoretical grounds, Williams proposed that idiopathic aqueductal stenosis could best be understood as the result rather than the cause of hydrocephalus. Furthermore, an overlap has been noted between aqueductal stenosis and communicating hydrocephalus, with the symptoms of the former due to only partial obstruction of the aqueduct. Finally, communicating hydrocephalus has been shown to cause obstruction of the aqueduct in a number of patients, suggesting a direct causality.

Similar to the clinical and morphological findings, the physiological features of the ventricular and subarachnoid spaces in NPH and LIAS have also been shown to be similar. There are higher than normal CSF pulse pressure amplitudes in patients with and without communicating hydrocephalus, and this reflects a loss of spinal compliance in both diseases. Furthermore, there is no significant difference between the resistance to CSF resorption and elastance measured in the ventricles or the subarachnoid space in patients with aqueductal stenosis or NPH. Finally, the authors of one study reported that measurements of neuropeptide light protein, tau protein, sulfatide, vasoactive intestinal peptide, neuropeptide PYY and CSF–serum albumin ratio in ventricular CSF in patients with aqueductal stenosis and NPH showed no differences, suggesting to the authors that there were similarities in the pathophysiology and turnover rate of these proteins despite the differences in CSF dynamics.

One hypothesis to account for the aforementioned findings is that LIAS is essentially a noncommunicating form of communicating hydrocephalus. An overlap in the therapies of these conditions supports this theory. Endoscopic third ventriculostomy is the therapy of choice in LIAS, but ETV has been reported to have a success rate approaching that of shunt therapy when used in patients with NPH. Hopf and colleagues have reported a success rate of 83% after performing ETV in patients with LIAS. Although other authors have not found their results to be as promis-
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The suggestion that the same treatment works for both obstructed and unobstructed hydrocephalus indicates that these conditions must have a similar physiology. Not only the treatment successes but also the treatment failures suggest the presence of underlying communicating hydrocephalus in LIAS. Based on the failure of ETV to treat LIAS in their cases, McMillan and Williams proposed that a hydrocephalus of some other cause (a communicating hydrocephalus) was responsible for the aqueductal stenosis, and the original cause might still be incompletely resolved after treatment. Tsell and associates reported that only 50% of their patients improved permanently after ETV despite an open ventriculostomy and with most nonresponders improving after a later shunt operation. Finally, third ventriculostomy even in successfully treated cases appears to leave the patients with a balanced communicating hydrocephalus (the decrease in ventricular size that does occur is frequently minimal, usually requiring ≥ 3 weeks to occur, and, in most patients, the ventricles do not return to normal size). Thus, although ETV may be the treatment of choice for LIAS, whether the patient improves, an underlying communicating hydrocephalus may still exist and may be only partially treated.

Bulk Flow Theory of NPH and LIAS

In recent theories on the causation of NPH, proponents have stressed a hydrodynamic origin. These theories predict that a reduction in the arterial tree compliance in NPH will lead to a decrease in the conversion of the pulse pressure within the arterial tree—that is, to pulsatile flow. Essentially this reduces the ability of the arterial tree to hinder the pulse pressure and results in a greater brain parenchymal pulsation, which causes a “water hammer” effect and ventricular dilation. This theory fails to describe the cause of the abnormal CSF bulk flow in NPH. A hypothesis to explain the bulk flow changes in chronic hydrocephalus will be described with the help of the drawings in Fig. 5. Figure 5A is the hypothetical depiction of an individual with a healthy brain. Normally, jugular bulb pressure is close to atmospheric, the sagittal sinus pressure is between 4 and 10 mm Hg, and the driving pressure gradient between the subarachnoid space and the sagittal sinus is between 2 and 6 mm Hg. The pressure gradient between the CSF and the cortical veins has been shown to be 2 mm Hg in humans. No data exist regarding the pressures in the deep system, but the vein of Galen is larger than the cortical veins and has a shorter passage through the subarachnoid space. It is hypothesized that the lesser interaction between the subarachnoid space and deep veins may account for the lower deep system pressures. Note, despite the favorable pressure gradient between the ventricle and the deep capillaries depicted in Fig. 5A, bulk flow would still referentially pass to the vertex, because the pressure gradient through the arachnoid granulations is greater.

The hypothesized findings in both NPH and LIAS after third ventriculostomy are depicted in Fig. 5B. An elevated venous pressure in the sagittal sinus reduces the driving gradient below that where CSF is resorbed. It has been noted in children with hydrocephalus and in dogs that the gradient from CSF to sagittal sinus drops to 2 mm Hg without causing an apparent focal obstruction in the sinus. The lack of a focal obstruction means that the pressure gradient along the sagittal sinus (which is normally only a few mm Hg) is increased. The elevated pressure gradient along the sagittal sinus tends to maintain a lower pressure in the deep system. The small pressure gradient between the ventricle and the deep capillaries would tend to force CSF resorption to occur into the deep system (subependymal brain) and reverse the CSF flow. Subependymal CSF absorption is often observed in communicating hydrocephalus. Nuclear cisternography shows ventricular reflux but not passage of the tracer over the vertex in LIAS after ventriculostomy, also suggesting that ventricular absorption predominates postoperatively. Deep CSF resorption also suggests lower pressure. Portnoy and associates were the first to suggest that a pressure gradient between the superficial and deep systems could induce hydrocephalus. They suggested that normally the cortical venous pressure is maintained above CSF pressure by the Starling resistor effect of the lateral lacunae. A similar mechanism is absent from the deep system, meaning that the deep system pressure remains similar to the dural sinuses. If CSF and superficial venous pressures rise in hydrocephalus, then an elevation in the pressure gradient across the ependymal layer of the ventricle will ensue, encouraging transependymal CSF resorption. It has been hypothesized that without ventriculostomy in LIAS, the subarachnoid CSF would simply drain through the less favorable accessory resorption pathways available through the perineural and perivascular routes.

Idiopathic Intracranial Hypertension and LIAS

Because an elevated venous pressure in adults seems to cause idiopathic intracranial hypertension rather than hydrocephalus, how can an elevated venous pressure in LIAS also be correct? The answer may depend on the site of the pressure elevation. This will be explained with the aid of Fig. 5C. In most adults the SSS pressure remains constant regardless of an elevation in CSF pressure; in other words, the sinuses are not compressible. In the minority of patients in whom SSS pressure increases in proportion to elevations in CSF pressure, this effect appears to be due to collapse of the transverse sinuses. A collapse of the venous

Fig. 4. Scatter plot comparing the ventricular dilation with the SSS flow in LIAS showing no correlation.
The cause of the elevated sagittal sinus pressure in LIAS remains unknown. In the majority of healthy adults and in healthy dogs, the minimal CSF pressures generated by chronic hydrocephalus would not be enough to compress the sinuses or significantly increase their pressure. There is circumstantial evidence that the pressure elevation may be related to the reduced compliance of the system. First, there is the high correlation between postsurgical improvement and elastance. Second, the reduced compliance does not seem to be secondary to the ventricular enlargement. Third, patients with LIAS tend to suffer decompensation (and therefore present) at the same stage in life where there is a significant “normal” age-related decrease in compliance of the craniospinal system. Finally, reducing the CSF pressure (and as a consequence increasing compliance) reduces the sinus pressures in both children and dogs with chronic hydrocephalus. A pure elevation in sinus impedance should only increase the pulse pressure but not the mean sinus pressure. However, impedance mismatches are known to increase the amount of work required to maintain flow, and increased mismatches at the venous junctions are likely given the significantly reduced venous compliance; these would require an elevated pressure to maintain flow.

The major discrepancy between the finding of the present study and the literature is in the relation of the blood flow to the subependymal brain in hydrocephalus. It is almost universally accepted that the brain just beneath the ventricles is ischemic in chronic hydrocephalus, but this study has shown a normal blood flow passing out via the straight sinus. If the arterial inflow in LIAS to the deep portions of the brain were reduced (as suggested by studies in the literature), then for the venous outflow to be normal, part of the collateral flow from the cortex would need to return via the deep system to make up the shortfall. This could only occur if the pressures in the deep venous system were lower than those in the superficial (as hypothesized).

Study Limitations

It has been my intention to describe the physiology underlying the apparent balance that exists between the ventricular system and the subarachnoid space in LIAS during the chronic stage of the disease process. There are, however, some limitations of the current work that need clarification.

This data do not necessarily shed light on the original cause of the aqueductal occlusion. Two theories have been popularized. The first is that the aqueductal stenosis is always present but has remained asymptomatic since childhood and the second is that the aqueductal stenosis is a secondary phenomenon following an initial communicating hydrocephalus. Although the data presented may seem to support the second theory most readily, they are compatible with either.

The data do not directly address the cause of the ventricular dilation in hydrocephalus. The findings relate to the bulk flow of CSF, which is toward the vertex in healthy individuals and in those with IIH but reverses with absorption via the subependymal brain in the presence of hydrocephalus. No large transmantle or transependymal pressure gradients are needed to explain the findings as presented—only a small pressure gradient between the deep and superficial venous systems. It has been suggested that ventricular dilation in chronic hydrocephalus may relate more to dynamic brain compression and white matter damage (that is, volume loss). This effect would require only a very small pressure gradient to cause the ventricles to dilate. The dynamic components of hydrocephalus, however, are beyond the scope of the present report.

Fig. 5. Artist’s illustrations. A: Drawing depicting the normal venous and CSF pressures. Cerebrospinal fluid flows from the ventricle to the vertex (arrows). B: Drawing depicting hydrocephalus. Compared with panel A the SSS pressure is elevated, reducing the driving pressure across the arachnoid granulations (AG). The pressures in the deep system remain lower than the CSF pressure, and the flow of CSF is reversed (arrows). C: Drawing depicting IIH. Compression of the transverse sinuses abolishes the pressure gradient along the SSS, elevating the deep pressures and promoting normal CSF absorption over the vertex. J = jugular bulb; ST = straight sinus.

sinuses causing elevated venous sinus pressures has a well-documented correlation with IIH. In the 21 patients studied by King et al. there was an abrupt gradient at the level of the transverse sinus and the mean torcular pressure was almost identical to the sagittal sinus pressure. This has the effect of elevating the deep venous pressures so they are no longer conducive to CSF resorption. Janny et al. reported documenting ongoing CSF resorption over the vertex in 12 patients with IIH, and King et al. documented a normal CSF–sagittal sinus gradient despite the raised pressures.

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However, a limitation of this explanation is the apparent lack of any suitable connections occurring between the superficial and deep venous territories to allow for this flow.

Conclusions

Similar to findings in animal models, patients with LIAS experienced reduced venous compliance and an elevation in venous collateral flow. Together, these findings suggest that an elevation in venous pressure may be associated with this disease process. A literature review indicated that only subtle differences may exist in the pathophysiology between patients with LIAS, NPH, and idiopathic intracranial IHH.

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