Editorial

Hydrocephalus and idiopathic intracranial hypertension

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Hydrocephalus and idiopathic intracranial hypertension (IIH) have in common an increase in intracranial pressure (ICP) and responsiveness to treatment by means of shunt placement. Is that all they have in common? Dr. Bateman and his colleagues have analyzed information obtained from assessment of dynamic MR imaging to study these relationships in children and adults and present their findings in this issue of the Journal of Neurosurgery: Pediatrics. Some of the data on which their conclusions are based, such as flow velocities in venous sinuses and larger cortical veins, are actually measurable on dynamic imaging, and some are calculated using information gained from other measurements, such as collateral venous flow indicating increased resistance to outflow through the expected venous channels. From these data they postulate that there is increased pressure in the dural venous sinuses in both IIH and childhood hydrocephalus. They also note that in some cases the increase in dural venous pressures is the inciting event in the production of both of these conditions, and sometimes it is a result of pressure on the dural venous sinuses from a primary brain abnormality causing increased ICP.

Dr. Bateman and colleagues have built on the important work of Sainte-Rose et al. who investigated whether increased intracranial venous pressure was a cause or an effect of hydrocephalus. In one patient with achondroplasia Sainte-Rose was able to actually treat the hydrocephalus with a transverse sinus to jugular vein bypass. Increased venous pressure was suggested as a cause of hydrocephalus at least as early as 1961, and the association between venous hypertension and hydrocephalus in children has received a good deal of interest. It has been studied prospectively in patients with known elevations of right atrial pressure from congenital heart disease, and in those patients with modest increases in this parameter there was no increase in the incidence of hydrocephalus.

We have studied the relationship of ICP and superior sagittal sinus (SSS) pressure in normal and hydrocephalic animals in some detail. As ICP rises there occurs a distortion of the dural venous sinuses, turning them from equilateral to isosceles triangles and increasing the resistance to flow out of the SSS, which we have suggested as an increase in SSS elastance (ΔP/ΔV).

What significance is found in these observations? What happens differently in hydrocephalus that begins in adulthood from what happens in children, why and what clinical relevance does it have? There are two major differences between hydrocephalus that begins in infancy and that which begins in adulthood. Adult-onset hydrocephalus results from a limited number of pathological processes, and the obstruction that causes it occurs at a relatively predictable site. Tumors usually cause the hydrocephalus by blocking the flow of cerebrospinal fluid (CSF) among the ventricles (intraventricular obstructive hydrocephalus as opposed to obstructive hydrocephalus), and hemorrhage and infection cause a block at the outlet foramina of the fourth ventricle or block the cortical subarachnoid space (extraventricular obstructive or “communicating” hydrocephalus). Infantile hydrocephalus results from a wide range of pathological conditions, and the actual site of the obstruction may not be obvious at the time of presentation. The second difference is a biomechanical one, and it involves the distensibility of the head itself and the fact that the fontanel is open in infants. Assuming that there is a need for ICP to exceed SSS pressure by a certain level, in adult hydrocephalus the ICP will rise until the CSF can be absorbed. In the infant, however, the distensibility of the head prevents the increase in ICP and CSF cannot be absorbed. This results in macrocephaly and ventricular dilation. This problem led Epstein and colleagues to treat posthemorrhagic hydrocephalus of the premature newborn by wrapping of the head with an elastic bandage, a strategy with which they achieved significant success.

Based on the work presented by Bateman and colleagues and the discussion above, it is clear that venous obstruction can cause hydrocephalus in babies. That leaves two important questions. What happens when an adult experiences a markedly increased dural sinus venous pressure, and what happens when a patient who was treated for hydrocephalus from venous hypertension in infancy has a shunt malfunction as an adult?

Restriction of venous outflow from the dural venous sinuses causes increased ICP without ventriculomegaly and is probably a universal mechanism in patients with pseudotumor cerebri, described by Bateman and colleagues as IIH.
I have an objection to the use of the term idiopathic which means “arising spontaneously from an obscure or unknown cause.” Recent work has shown that the etiology of this condition is definable in all or most patients by measuring the pressure in the dural venous sinuses. When related to extreme obesity it is due to elevations of right atrial pressures, and in the nonobese patients it is related to measurable pressure gradients within the dural venous sinuses and therefore increased pressures in the SSS.

It has also been shown that, at least in some patients, lowering ICP results in a decrease in SSS pressure leading to the question of which comes first, which is cause and which is effect. It has also been demonstrated that these pressure gradients may persist after ICP is lowered. To put this all together, it is clear that all patients with pseudotumor cerebri will have increased SSS pressure. In most of these patients—including patients with anatomical venous constriction, SSS thrombosis, or obesity-related right atrial hypertension—the increase in venous sinus pressure causes the pseudotumor cerebri. It seems likely that there are a number of patients in whom there is an increase in ICP that causes the increased venous pressure by changing the geometry of the dural venous sinuses. The system therefore reflects a positive feedback loop in which geometric changes in the dural venous sinuses lead to increased venous sinus pressure. This increased venous sinus pressure in turn leads to increased ICP, which leads to further distortion of the dural venous sinuses and so on. It does not matter at which point the perturbation begins—the effect is the same, so an increase in ICP can be the inciting event. What is not known is whether placement of a venous stent could break the positive feedback loop. This question has no answer at this time.

Finally and probably most critically is the issue of shunt failure in the older child or adult whose hydrocephalus presented early in life and was due to venous hypertension. What happens to these patients when their shunts fail? In 1979 Engel and colleagues recognized an entity that they called “normal volume hydrocephalus.” They described a situation in which patients who had previously been treated with shunt placement presented with signs and symptoms of shunt failure without an increase in ventricle size. Measurement of ICP or shunt revision led to the diagnosis of shunt failure, and the symptoms remitted with repair of the shunt. Since that work, there has been much attention paid to this entity, its cause, and its effect. As many as one in four older children or adults with a history of infantile hydrocephalus will have nonresponding ventricles at the time of shunt failure. We have had the opportunity to study some of these children and young adults and have found demonstrable venous pathological conditions in all of them.

These patients now suffer from a condition identical to pseudotumor cerebri as postulated in the work reviewed here. They are subject to severe problems with intermittent or sequential failures of the ventricular catheters at times when access to the cerebral ventricles is problematic. In such cases management strategies should be similar to management strategies for pseudotumor cerebri and shunting should include the management of the CSF in the cortical subarachnoid space.

In conclusion, Bateman and colleagues have shown us a noninvasive method of studying venous outflow in various disorders of CSF flow and intracranial dynamics. Using this technique they have shown a clear interrelationship between hydrocephalus in infants and pseudotumor in adults. The authors’ findings as well as their technique support both experimental evidence and clinical observations made on these two groups of patients and may serve to help select patients for novel treatments such as venous stent placement in the future.

References

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