Hyperemic hydrocephalus

HAROLD L. REKATE, M.D.

Barrow Neurological Institute, St. Joseph’s Hospital and Medical Center, Phoenix, Arizona

Dr. Bateman’s article, “Hyperemic hydrocephalus: a new form of childhood hydrocephalus analogous to hyperemic intracranial hypertension in adults,” in this issue of the Journal of Neurosurgery: Pediatrics represents another advance in the use of MR imaging techniques applied to the study of the pathophysiology of hydrocephalus and other abnormalities of CSF dynamics. Previous work in this series of studies showed that vascular and CSF dynamics were similar in certain cases of hydrocephalus and idiopathic intracranial hypertension (IIH). This new study takes that concept a step further and suggests that the underlying cause of IIH when a venous sinus stenosis cannot be demonstrated is hyperemia. Hyperemia is present in a number of patients with hydrocephalus when the underlying cause is not obvious. Such cases have been termed idiopathic.

Dr. Bateman accepts the premise that all IIH and some forms of hydrocephalus derive from significantly increased pressure in the venous sinuses. He divides IIH into venostenotic and hyperemic forms. In the hyperemic form of IIH, there is no pressure gradient across different levels of the dural venous sinuses. Magnetic resonance imaging measurements of cerebral blood flow (CBF) show significant increases in the pressure in those vessels. Dr. Bateman does not speculate on the cause underlying the increase in CBF but states that the hyperemia causes the venous hypertension that leads to the increase in intracranial pressure (ICP). In these same patients he has found that measurements of venous blood flow out of the intracranial compartment through the dural venous sinuses are significantly lower than would be predicted by the arterial inflow of blood and also significantly less than in normal controls. His new study involved 9 children with hydrocephalus without an obvious cause and therefore considered idiopathic. Their arterial flow (markedly increased) and venous outflow (decreased) were identical to those of the IIH patients that Dr. Bateman had previously reported on. These findings support the conclusion that an increase in dural sinus pressure in children with open sutures and fontanels leads to hydrocephalus, while an increase after the sutures have closed leads to IIH.

In our initial work on the pathophysiology of IIH, we measured right atrial and dural sinus pressure in 10 patients with documented pseudotumor cerebri. Five thin patients had significant pressure gradients within the dural venous sinuses. In obese patients the pseudotumor cerebri was related to increased right atrial pressure. We subsequently demonstrated that IIH in the obese could be treated successfully with bariatric surgery. This success suggests that the elevated venous pressure resulted from the obesity itself. None of the patients reported by Dr. Bateman were reported to be obese, and I am not aware of any measurements of this kind being performed on obese patients with pseudotumor cerebri.

Measurements of CBF and venous outflow rates are derivative: they are not validated by actual physical measurements. These derivations require the acceptance of a number of assumptions that would be unnecessary with direct physical measurements. I am not competent to comment on the subtleties of the techniques employed in this study, but I would like to suggest an alternative explanation of the findings and ask for help in understanding the process.

In our continuing study of ICP in general and IIH and complex hydrocephalus in particular, we have had the opportunity to perform retrograde venograms in a series of patients with severe slit-ventricle syndrome. In these patients, the ventricles did not expand at the time of shunt failure despite severe increases in ICP. All of these patients, whose condition has been referred to as normal volume hydrocephalus, had high superior sagittal sinus (SSS) pressure. The cause of the venous hypertension in 3 of these patients was elevated right atrial pressure. None of these 3 patients were obese, and none had a clear underlying cause of their hydrocephalus. Increased right atrial pressure can cause obesity-related IIH. It now appears that this process can also be operating in nonobese patients and can begin early in life.

Dr. Bateman uses a hydraulic analog of Ohm’s law (V = IR) to explain how increased arterial inflow can lead to increases in venous pressure in patients with venostenotic states. He then states that in the absence of signs of congestive heart failure there is no reason to assume that there is a primary venous cause of the venous hy-
pertension and that the increased venous sinus pressure is due to the increased arterial inflow. Using the same equation as a concept, we can document the same interdependence of arterial blood flow and dural venous sinus pressure in patients in whom the venous sinus pressure is due to increased right atrial pressure.

Another way of expressing the hydraulic equivalent of the Ohm’s law equation would be as follows: CPP = CBF × resistance, where CPP (cerebral perfusion pressure) equals mean arterial pressure minus SSS pressure; CBF has already been defined, and resistance is a function of the diameter of the precapillary arterioles.

Assuming that cerebral autoregulation is intact—and in this situation there is no reason to suspect that it is not—increasing the pressure in the SSS by increasing right atrial pressure would lower CPP. For CBF to remain the same, the luminal diameter of the precapillary arterioles would necessarily have to increase to lower the resistance. In turn, cerebral blood volume (CBV) and brain turgor would increase. What would be the effect of this vasodilation on arterial CBF as measured by these techniques? Hyperemia is defined as increased volume of blood in tissues or blood vessels, and right atrial hypertension causes hyperemia.

A traditional treatment of IIH has been the use of the carbonic anhydrase inhibitor acetazolamide. The putative method of decreasing ICP in IIH has been to decrease the production of CSF. We have found that acetazolamide works quite well when IIH is related to obesity but is not helpful in the management of venostenotic IIH. We have also shown that using acetazolamide to treat obesity-related IIH decreases right atrial pressure. This medication leads to a mild metabolic acidosis and therefore tends to decrease CBF by constriction of the precapillary arterioles, which would tend to decrease CBV. This process again emphasizes the interdependence of CBF, CBV, and pressure in the dural venous sinuses.

The techniques described by Dr. Bateman have the potential to help diagnose and treat complex forms of both IIH and hydrocephalus. I suspect what Dr. Bateman is describing is the substrate for a variety of enigmatic conditions such as normal volume hydrocephalus and chronic compensated hydrocephalus found incidentally in adults and possibly even in patients with normal pressure hydrocephalus.

In a landmark paper, Dr. Rekate and associates studied 10 patients with IIH using manometry and found 2 subtypes. The first group contained patients who were predominantly thin and had venous outflow stenoses. A large pressure gradient was found across these stenoses suggesting hemodynamic significance. The second group were described as morbidly obese and were found to have elevated right atrial pressures but no evidence of an outflow stenosis. I have also investigated IIH. Excluding patients with thromboses, I too have found 2 subgroups. Seventy-five percent of my patients had outflow stenoses on MR venography similar to Dr. Rekate’s findings. My flow quantification technique also indicated that these stenoses were hemodynamically significant. The remaining 25% of patients in my cohort had no evidence of outflow stenosis and were mostly obese, again similar to Dr. Rekate’s second group. I found cerebral hyperemia in this subgroup. Clearly, we are describing different findings in similar groups of patients. Dr. Rekate favors right atrial pressure elevation secondary to obesity as the cause of the IIH in those patients without stenoses, and I favor hyperemia. The 2 suggestions are not necessarily mutually exclusive and indeed may be synergistic in some instances, but which one is more important?

I believe that raised central venous pressure (CVP) may be associated with IIH but by itself cannot cause IIH. I have several reasons for believing this.

First, elevated CVP is a common accompaniment of obesity. De Divitiis et al. noted that elevated right atrial pressure was strongly correlated with obesity even in the absence of IIH. The elevation in CVP in obesity is caused by a combination of increased abdominal and thoracic pressure compounded by an increase in the circulating blood volume and cardiac output. Given these findings, it is obvious that if obesity and elevated CVP could induce IIH on their own, then the latter condition should be much more common than it is, particularly in males.

Secondly, Dr. Rekate’s group measured CVP in the 5 patients with morbid obesity in the supine position and found a mean pressure of 11.8 mm Hg. In the supine position the SSS is only marginally above the level of the

### References


### Response

**Grant Bateman, M.B.B.S., F.R.A.N.Z.C.R.
John Hunter Hospital, Newcastle, Australia**

I would like to thank Dr. Rekate for his thoughtful review and comments regarding the impact and relevance of this manuscript. He raises several important points and I would like to respond to his comments regarding the relative importance of central venous pressure and hyperemia in the physiology underlying IIH and childhood hydrocephalus.

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