

# Idiopathic Intracranial Hypertension (IIH)

Thomas J. Walsh

## Abstract

The first report of Idiopathic Intracranial Hypertension (IIH) was by Quinche in 1893. Throughout history it has had many names. When I was a child growing up, before we had antibiotic therapy, middle ear infections occasionally would trigger intracranial inflammation and the disease was called Otitic Hydrocephalus. Then it was called Benign Intracranial Hypertension which we now know is far from benign. In 1931 Symonds was the first to suggest that it was caused by dural sinus obstruction. Loss of vision or field is not uncommon in untreated cases. We now use the term Idiopathic Intracranial Hypertension to express our ignorance about the cause and mechanism of the disease. Some

cases are related to causes that we can identify and correct. These include Vitamin A, Tetracycline, Lithium and Corticosteroids. The mechanism of these are not fully understood but their causal relationship is historically known. Many cases still fall into the unknown cause category.

Submitted: 27 February 2008

Accepted: 19 March 2008

From the Department of Ophthalmology, Yale Eye Center, Yale University, USA.

Address correspondence and reprint request to: Dr. J Walsh Professor of Ophthalmology and Neurology, Yale University, 330 Cedar Street, P.O. Box 208061, New Haven, Connecticut, P. C. 06520-8061, USA

E-mail: twalsh13@optonline.net

## History

There are three ways to increase intracranial pressure without dilating the ventricles:

1. You can obstruct venous drainage, which we can see after radical neck surgery, with a loss of drainage after resection of one side of the Jugular system.
2. An alternative way is dilation of the Arterial system and microcirculation.
3. The last way is edema of cerebral tissue.

## Treatment

The unwarranted clinical effects of the disease are centered around the loss of vision and/or field. Headaches are a symptom but no permanent defect in and of itself. The pressure over a period of time causes atrophy of the optic nerve. The treatment has been to decrease the Intracranial Pressure by reducing CSF pressure. One of the techniques to reduce the pressure has been repeated spinal fluid taps. This has not been a practical technique because of the frequency required and how quickly the pressure builds up again. A tap may be useful for an acute effect but not chronic use. The usual choice is the oral medication Diamox. This is a Carbonic Anhydrase inhibitor and reduces cerebral spinal fluid production. The use of other diuretics do not have this property because they are not Carbonic Anhydrase inhibitors and therefore are of no use. If the patient has drug sensitivity Topamax has some of the Carbonic Anhydrase qualities and can be substituted but is not as effective. If they do not work, or loss of vision is a concern, then surgical intervention to decrease pressure around the nerve like optic nerve sheath fenestration or a shunt of the CSF can be performed.

Unfortunately, both of these options have their problems as well as a significant fail rate.

Walter Dandy in 1937,<sup>1</sup> outlined the rules for aggressive surgical treatment which are applicable to management today. His surgical approach was a subtemporal decompression. The rules for surgical intervention are gliosis of the disc, increasing obscuration, decreasing field or vision and double vision. These are all signs of an impending decompensation of the visual system.

The two surgical treatments optic nerve fenestration and CSF shunts do not treat the problem but only reduce the complication until such time as the process resolves spontaneously. It is hoped that the surgical treatments will last long enough so they do not have to be repeated.

## Mechanism

The problem that confounds us is as follows: intracranial pressure is the first culprit and then develops a decrease in spinal fluid or was a decrease in fluid flow through the sigmoid sinus the cause of the IIP? Several authors have come up with some new theories and possible modes of therapy.

## New Ideas

Loss of weight has always been part of the therapy since most patients are at least moderately obese. However, most patients are not what we would call morbidly obese. Sugarman performed gastric bypass surgery on 6 patients.<sup>2</sup> Prior to surgery he found increase abdominal pressure and increase cerebral venous pressure. This impeded venous return from the brain and also increased CO<sup>2</sup> levels. These

two problems and the IIP improved with gastric bypass surgery. I do not feel this is appropriate therapy for the average patient who is mildly obese. In those cases a diet regimen is sufficient.

The surgical technique mentioned above appears to confirm the work of King who concludes that decreasing intracranial pressure would decrease venous pressure. He did this experiment by removing CSF from between a tap at C1-2. Simultaneously he performed manometry of the Superior Sagittal Sinus which revealed a rapid decrease in pressure. He concluded that obstruction of venous outflow was due to ICP and caused by collapse of the wall of the Superior Sagittal Sinus and Transverse Sinus. Partial obstruction of the Transverse Sinus suggests that this may be an asymptomatic phase of the illness possibly occurring for several months with raised intracranial pressure. Once the Transverse Sinus begins to collapse, the Sigmoid Sinus pressure will rise rapidly and further impair passive CSF absorption. This mechanism could quickly produce an escalation of IIP and the onset of symptoms.

Farb used a new MR technique.<sup>3</sup> His studies demonstrated congenital narrowing of the Transverse Sinus and a decrease in CSF absorption. This resulted in IIP which in turn exacerbated the underlying venous sinus abnormality and created a flow limiting stenosis. The pressure was transferred to the Sigmoid Sinus like King has demonstrated,<sup>4</sup> but now we see a reason for this happening in the Transverse Sinus.

Owler took this principle and applied it to a surgical approach.<sup>5</sup>

He operated nine patients with IIP who demonstrated dural sinus obstruction. He used a well known surgical procedure called stenting. He inserted a stent into the Transverse Sinus. He did these cases under general rather than local anesthesia because dural sinuses are very sensitive to pain. The result of opening the Transverse Sinus led to normal IIP and a curing of the Idiopathic Intracranial Pressure. These results were followed for one year with no failure.

### **Conclusion**

These clinical conclusions and newer surgical approach seem promising but need to be confirmed. This approach may not be useful to all cases but may reduce the number of idiopathic cases for which we have only a watch and wait approach until we see optic nerve damage.

### **References**

1. Dandy WE. Intracranial Pressure without brain tumor, Diagnosis and treatment. *Ann. Surg.* 1937; 106:492-513.
2. Sugerman HJ, DeMaria EJ, et al. Increased intradominal pressure and cardiac filling pressures in obesity-associated pseudotumor. *Neurol.* 1997; 49:507-511.
3. Farb RI, Vanek I. Idiopathic intracranial hypertension. *Neurol.* 2003; 60:1418-1424.
4. King JO, Mitchell PJ, et al. Manometry combined with cervical puncture in idiopathic intracranial hypertension. *Neurol.* 2002; 58:26-30.
5. Owler B, et al. Pseudotumor cerebri syndrome: venous sinus obstruction and its treatment with tent placement. *J. Neurosurgery.* 2003; 98: 1045-1055.