

Key Points

1. Pseudotumor cerebri, or idiopathic intracranial hypertension (IIH), is a disorder involving elevated intracranial pressure with no visible cause
2. There appears to be a link between IIH and Chiari; either a subset of Chiari patients also have IIH or one causes the other or surgery can lead to IIH
3. Most common symptom of IIH is pressure headache, made worse with straining
4. Visual problems are also common and permanent visual damage is a concern
5. Treated with drugs; sometimes the optic nerve is surgically decompressed to preserve visual function
6. More common in women than men; especially common among obese women
7. About 70% get better with treatment in 3 months or less, but about 25% experience protracted symptoms

Definitions

benign intracranial hypertension - another name used to refer to idiopathic intracranial hypertension

fenestrate - create perforations or openings

idiopathic - due to an unknown cause

idiopathic intracranial hypertension (IIH) - condition where a person suffers from elevated intracranial pressure with no visible cause

intracranial pressure (ICP) - the pressure of the cerebrospinal fluid in the skull

lumbar puncture (LP) - procedure where a needle is inserted into the CSF space of the lower back region; can be used to remove CSF or insert drugs

Idiopathic Intracranial Hypertension (Pseudotumor Cerebri)

April 20, 2006 - Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri (PTC) and benign intracranial hypertension, is a condition characterized by an increase in intracranial pressure (the pressure of spinal fluid in the head) with no apparent cause. Like Chiari, its main symptom is a pressure headache, made worse by straining, coughing, etc. Since both IIH and Chiari are fundamentally tied to the CSF system, perhaps it should not be surprising that there appears to be some type of connection between the two.

Recently, there have been a couple of studies published which indicate that a percentage of Chiari surgeries fail due to the presence of IIH in addition to Chiari. However, this research is early-stage, and the exact link between Chiari and IIH is not understood. It could be that the sustained increase in pressure associated with IIH eventually leads the cerebellum to herniate out of the skull and creates a Chiari malformation. If this were the case, then decompression surgery would help symptoms associated with direct compression of the malformation, but would not relieve the symptoms associated with the elevated pressure of IIH.

It may also be the case that the blockage caused by a Chiari malformation, which we know can elevate the intracranial pressure, may lead to a fundamental change in the CSF system and eventually IIH. In this case, even though the region around the Chiari malformation is decompressed surgically, for unknown reasons, the intracranial pressure remains high.

A third possibility is that both Chiari and IIH are manifestations of a more fundamental problem, such as a too small posterior fossa (the skull region where the cerebellum sits). Perhaps some people with a small or abnormally shaped skull develop IIH, while others develop Chiari, and still others develop both.

Finally, it may be that Chiari surgery itself plays a role in the development of IIH. There appear to be some cases where symptoms associated with IIH don't appear until after Chiari surgery. How, and even if, decompression surgery could lead to IIH is not at all clear.

While the nature of the connection is a mystery, and likely will turn out to be a combination of the possibilities listed above, the importance of IIH in the context of Chiari is clear. Although they didn't mention IIH's apparent relationship to Chiari, a group from Denmark (Skau, Brennum, Gjerris, Jensen) recently published a very informative review of IIH in the April, 2006 edition of the journal *Cephalalgia*. The following description of IIH is based upon that review.

As mentioned previously idiopathic intracranial hypertension is a condition involving elevated pressure of the CSF in the brain region due to an unknown cause. Idiopathic is a term used to mean of unknown origin; intracranial means within the skull; and hypertension means raised, or elevated, pressure. IIH was first described in 1937 and has remained largely a mystery ever since.

Being idiopathic, by its very nature, IIH is difficult to define and diagnose. The name IIH really only applies when no cause can be found, and IIH likely encompasses a number of related conditions. Over the years, as different, specific conditions which lead to elevated ICP have been identified, they have been split off from IIH and labeled separately (perhaps one day, Chiari related intracranial hypertension will be thought of differently than IIH).

Because of this, IIH is largely diagnosed by excluding other possibilities. If a patient (especially an obese woman as will be explained below) has symptoms of elevated ICP, the suspected pressure can be confirmed through an eye exam and or a lumbar puncture. Once confirmed, the treating physician will look for any of a myriad of potential causes of elevated ICP, including: tumors, vascular disease, infections, drug reactions, and circulatory problems to name a few. Once all other causes are ruled out, the condition is considered to be idiopathic in nature, or IIH.

Given its somewhat obscure definition, the number of people affected by IIH is not known. However, like Chiari, recently there has been a rapid rise in diagnoses. It appears to effect women more than men, perhaps as much as 15 to 1, and IIH is especially common among obese women. In fact, some studies of IIH have found that as many as 70% of IIH patients were obese women. Interestingly, among children there appears to be no difference between the number of boys and girls affected, and a link to obesity in adult men has not been identified.

Although the most common symptom is a pressure headache, symptoms also include double vision, visual blurring, nausea, vomiting, dizziness, and ringing in the ears (see Table 1). The most serious symptom associated with IIH is vision loss. The sustained pressure associated with IIH can eventually damage the optic nerve - the bundle of fibers which connect the eye to the brain - and if not treated can lead serious vision problems.

optic nerve - large bundle of nerve fibers which connect the eye to the visual region of the brain, critical for visual function

pseudotumor cerebri (PTC) - another name for idiopathic intracranial hypertension; not used as much anymore

shunt - a small tube which is inserted into the body to divert spinal fluid

Source

Source: Skau M, Brennum J, Gjerris F, Jensen R. What is new about idiopathic intracranial hypertension? An updated review of mechanism and treatment. Cephalalgia. 2006 Apr;26(4):384-99

Table 1
Common Symptoms Of Idiopathic Intracranial Hypertension

	Study 1	Study 2	Study 3
# of Patients	62	63	57
Headache (%)	95	75	81
Double Vision (%)	31	35	33
Visual Blurring	65	68	72
Nausea, Vomiting	24	21	—

Note: Study 1: Johnston et al., Brain 1974; Study 2: Rush et al., Mayo Clin Proc 1980; Study 3: Corbett et al., Arch Neurol 1982

Since the cause of IIH is unknown, treatments tend to focus on the symptoms and can involve drugs or surgery. Unfortunately, to date there have not been any rigorous studies comparing the effectiveness of drug treatments versus surgery.

On the medicine side, acetazolamide is commonly one of the first drugs tried. It is believed to act by reducing the production of cerebrospinal fluid and thus leads to a decrease in ICP. If the acetazolamide does not work, it is sometimes supplemented with a second medicine, furosemide. Recently, an anti-convulsant, topiramate, has been reported to be used to treat IIH, but more research is required to determine its effectiveness. In addition to treating the symptoms with medicine, if the patient is obese, weight loss has been shown to be an effective approach to alleviating symptoms.

On the surgical side, a shunt can be inserted to divert CSF and lower the pressure in the head. While shunting is very effective in relieving symptoms, a patient then has to deal with potential problems related to having a shunt inside of them; namely mechanical malfunction and infection. Such problems are not uncommon can lead to the need for additional surgeries to revise or replace the shunt.

For a patient whose vision is at risk, a surgeon may elect to decompress the optic nerve by perforating the sheath over the nerve bundle. This reduces pressure on the nerve and according to published reports is usually successful in stabilizing or improving visual function. However, this type of surgery does not address the problem of elevated pressure directly and relief from other IIH symptoms is not as great.

One approach to treating IIH which may be falling out of favor is to repeatedly drain CSF through lumbar punctures. Draining a large quantity of CSF in this way usually provides temporary relief from symptoms, but again does not really address the underlying problem.

Overall, treatments for IIH are effective for a majority of people. Research has found that 70% or more of patients experience symptom relief or resolution within a couple months of starting treatment. However, there does appear to be a subset of patients, perhaps as many as 25%, for whom IIH becomes a long, difficult battle. In addition, much like Chiari, recurrence of symptoms, even years down the road, has been noted.

Although the exact mechanism which leads to the increased pressure in IIH is not known, from an abstract point of view there are several possibilities: increased production of CSF, abnormal absorption of CSF, increased brain mass, and obstruction of blood outflow from the brain. However, and somewhat surprisingly, research has failed to consistently find any of the above to be the problem in IIH patients. Interestingly, studies have shown an increase in resistance to CSF flow (much like exists with Chiari) in 75% or more of IIH patients.

For now, IIH remains largely a poorly understood condition, but with its connection to the CSF system, similar symptoms, and presence in a subset of Chiari patients, it appears IIH is a Chiari cousin worth staying in touch with.

-- Rick Labuda

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