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Diagnosing idiopathic intracranial hypertension

What patients should know

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WHAT IS IDIOPATHIC INTRACRANIAL HYPERTENSION? Idiopathic intracranial hypertension (IIH) is a condition that is more common in women of childbearing age who are overweight. Headaches are a frequent complaint. A lumbar puncture (“spinal tap”) can show increased pressure in the spinal fluid around the brain. When the eyes are examined, swelling in the main nerve that connects the eye to the brain—the optic nerve—can often be seen. Permanent vision loss is the most feared complication of this disorder, so prompt evaluation and treatment are important.

WHY IS THIS STUDY IMPORTANT? This study¹ is important because patients who do not have IIH may be incorrectly diagnosed. Patients may be examined by their primary care provider, emergency department staff, eye care provider, or neurologist and be told that they have swollen optic nerves. This may lead the provider to make a diagnosis of IIH when it does not exist. There are other causes for swollen optic nerves, and not everyone with this finding has IIH. Those with IIH are often treated with medicine and sometimes with surgery. People who don’t have IIH should not have needless treatments that might produce side effects. This study examined the reasons for misdiagnosis of IIH.

WHAT DID THE AUTHORS STUDY? The authors reviewed charts over an 8-month period looking for patients diagnosed with IIH or possible IIH by various providers. They separated these patients into those who had been treated and those who had not received treatment. They collected information about the patients, recorded the appearance of the optic nerves, and recorded spinal fluid pressure. They documented any medical or surgical treatment for IIH.

WHAT DID THE AUTHORS FIND? A total of 1,249 new patients were seen in the eye clinic where this study was done. Eighty-six were referred with a preexisting diagnosis of IIH. On closer examination, 34 (39.5%) of the 86 patients did not have IIH. Most of the misdiagnosed patients were women (94%) with an average age of 38 years. Of these

misdiagnosed patients, 27 had undergone at least 1 lumbar puncture and 10 had undergone 2–4 lumbar punctures. Twenty-six (76.5%) had been treated with medications for IIH, 1 had undergone a procedure to drain spinal fluid from the lower back to relieve pressure, and 4 had been referred for surgery. Twenty-two of these 34 patients had slightly elevated spinal fluid pressures, but this alone is not enough to make a diagnosis. Some findings on brain imaging can support the diagnosis, but only 3 of the 34 patients had imaging findings that would indicate increased intracranial pressure. The main reason patients were misdiagnosed was misinterpretation of the results from a single examination: the eye examination. This misinterpretation prompted unnecessary testing in about one-third of patients and led to invasive procedures in about one-third. The authors’ main conclusion was that the combination of common, harmless headaches and obesity in young women often leads to costly tests for presumed IIH.

WHAT ARE THE FUTURE DIRECTIONS? A swollen optic nerve signals a problem and suggests the diagnosis of IIH in an overweight headache patient. This study shows that many practitioners do not correctly assess the appearance of the optic nerve, which leads to unnecessary testing and treatments. Many patients had borderline elevated spinal fluid pressures, but it is unclear how the lumbar punctures were performed. It has been shown that spinal fluid pressures in normal patients can appear falsely elevated if pressure is applied to the abdomen during pressure measurement.² This “erroneously elevated” pressure can alarm practitioners into believing that the patient has IIH.

As the authors note, physicians often get very little education in viewing the optic nerve. The training of physicians in medical school and residency needs to be improved. It would also be helpful if doctors who first see these patients in the emergency department or primary care clinic had the technology to be able to “see” the back of the eye with photography. These images could be given to experts who could make better decisions about the appearance of the optic nerve. Some specialty centers are already working on these improvements.

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About idiopathic intracranial hypertension

WHAT IS IDIOPATHIC INTRACRANIAL HYPERTENSION? Idiopathic intracranial hypertension (IIH) is a condition most common in women of childbearing age who are overweight. Headaches are a frequent complaint and swollen optic nerves can be seen on examination. Increased pressure in the spinal fluid can often be shown by lumbar puncture (spinal tap). Permanent vision loss is the most feared complication of this disorder, so prompt evaluation and treatment are important.

WHAT CAUSES IIH? The cause of IIH is currently unknown. Some think that hormones may play a role. Being overweight may contribute, because weight loss often puts this illness into remission. Whatever the reasons, the disease causes swelling of the optic nerves leading to eventual vision loss which can be permanent.

WHO IS LIKELY TO HAVE IIH AND WHAT ARE THE SYMPTOMS? Young overweight women are most likely to experience IIH. This does not mean that thin older men do not occasionally develop the disorder, but it is very unusual. Headaches are very common. But some features of headaches are much more suggestive of IIH. These include transient visual obscurations and pulsatile tinnitus. Transient visual obscurations are vision blackouts that occur in one or both eyes, often after standing up quickly or bending over, lasting for only seconds and resolving quickly. Pulsatile tinnitus is a ringing in the ears that is synchronous with the heartbeat. Patients often state that they can hear their heartbeat while sitting up or standing. This symptom has less importance if the patient hears it only in the ear that is lying on the pillow. The patient's headache may also be worse while lying down and get better when standing.

WHAT TESTING NEEDS TO BE DONE TO DIAGNOSE IIH? Patients require a good eye examination. Those with IIH have swelling of one or both optic nerves. All patients require an MRI, which produces a

detailed picture of the brain. Certain findings on MRI help confirm increased pressure in the head. A lumbar puncture performed correctly should show increased pressure but otherwise normal findings.

ARE THERE PREVENTIVE MEASURES OR TREATMENT? Losing weight can help reduce the chance of getting this disorder and help keep a patient healthy. The drug acetazolamide can relieve pressure in the head and allow the swollen optic nerves to return to their normal size. This will prevent vision loss. The drug topiramate has also been used to assist with weight loss. Bariatric surgery has been used in some patients to help with weight loss. IIH cannot be successfully treated with medications alone in some patients. These patients, especially those with rapidly progressive vision loss, may need surgical treatment. Three surgical procedures are used to treat IIH: optic nerve sheath fenestration, shunting procedures, and venous sinus stenting.

The optic nerve has a sheath, or covering, in which a surgeon may cut a small window to allow escape of cerebrospinal fluid to reduce pressure on the optic nerve. There are several types of shunting procedures, but the favored procedure is the ventriculoperitoneal shunt. In this procedure, a piece of tubing is placed from the brain down to the abdomen, allowing spinal fluid to drain and reducing pressure on the optic nerves. Lastly, if these procedures do not work and a severe narrowing of one of the major veins in the back of the brain is found, a stent can be placed in the vein to help keep it open to allow for better drainage of spinal fluid. Patients should be followed by an ophthalmologist who is expert in this area (a "neuro-ophthalmologist") through all of these steps to make sure vision is not deteriorating.

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