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DIAGNOSTIC ASPECTS OF INTRACRANIAL HYPERTENSION WITH CONCOMITANT BRAIN DAMAGE

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Annotation: Idiopathic intracranial hypertension is caused by a buildup of cerebrospinal fluid (CSF) in the skull. Intracranial buildup of CSF can cause symptoms like pressure, pain, and vision changes over time. At first, you might not have any symptoms. Then, you may experience headaches. As the condition worsens, you might experience vision changes due to the CSF putting pressure on your optic nerve, which is responsible for your brain's vision processing. Unfortunately, this condition's origin remains mysterious to doctors and researchers. The word "idiopathic" signifies that there is no obvious cause of this condition — or at least, not one that has been identified. This condition is also called "pseudotumor cerebri" because the symptoms of IIH can mimic those of a serious brain tumor. While IIH is not a brain tumor, it can cause serious symptoms, including vision loss, if it's not treated in time.

Key words: Idiopathic intracranial hypertension, pseudotumor cerebri, brain, blood vassels.

Idiopathic intracranial hypertension occurs in only about 1 of 100,000 people, usually in women during their reproductive years. However, among young overweight women, it is 20 times more common. As more and more people are becoming overweight, the disorder is becoming more common.

What causes pressure within the skull to increase is unknown. However, certain people are more susceptible to idiopathic intracranial hypertension because the large veins (venous sinuses) that help drain blood from their brain are smaller than in most people. In these people, blood drains from the brain more slowly, causing a back-up of blood, which increases pressure in the brain and/or within the skull.

Exactly how being overweight contributes to increased pressure within the skull is unknown. But excess fat in the abdomen may increase pressure in the chest, and blood may not flow from the head to the chest as it normally does. Thus, pressure within the skull may increase.

In idiopathic intracranial hypertension, the increased pressure does not result from other identifiable disorders, such as tumors, infections, blood clots, or blockages that prevent the fluid that surrounds the brain (cerebrospinal fluid) from draining as it normally does.

In most people, the development of idiopathic intracranial hypertension cannot be traced to any particular event. In children, this disorder sometimes develops after corticosteroids are stopped or after growth hormone is used. Sometimes the disorder develops after people take tetracycline antibiotics or large amounts of vitamin A.

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Idiopathic intracranial hypertension usually begins with a daily or almost daily headache, which affects both sides of the head. At first, the headache may be mild, but it varies in intensity and may become severe. The headache may be accompanied by nausea, double or blurred vision, and noises within the head that occur with each beat of the pulse (called pulsatile tinnitus). A few people do not have any symptoms.

Increased pressure within the skull may cause the optic nerve to swell near the eyeball—a condition called papilledema. Doctors can observe the swelling by looking at the back of the eye through an ophthalmoscope.

The first sign of vision problems is loss of peripheral (side) vision. People may not notice this loss at first. As a result, people may bump into objects for no apparent reason. Vision may be blurred briefly, sometimes triggered by changing position, and blurring may come and go. Late in the disorder, vision is blurred, and people may quickly become blind. As many as one third of people lose their vision, partially or completely, in one or both eyes. Once vision is lost, it usually does not return, even if the pressure around the brain is relieved.

In some people, the disorder becomes chronic and progressively worse, increasing the risk of blindness. Doctors closely monitor and treat such people to prevent loss of vision.

Idiopathic intracranial hypertension recurs in about 10 to 20% of people.

- A doctor's evaluation
- Imaging tests
- A spinal tap

Doctors suspect idiopathic intracranial hypertension based on symptoms and results of a physical examination. Sometimes doctors suspect it when they detect papilledema during a routine examination with an ophthalmoscope.

If doctors suspect idiopathic intracranial hypertension, they check the field of vision (the entire area of vision that is seen out of each eye), including peripheral vision. They also examine the interior of the eye with an ophthalmoscope if they have not already done so.

Magnetic resonance venography is done to evaluate the large veins (called venous sinuses) that carry blood from the brain. This test enables doctors to determine whether the venous sinuses are narrowed or blocked. Narrowed venous sinuses may be the only abnormality that imaging tests detect in people with idiopathic intracranial hypertension. Magnetic resonance imaging (MRI) of the brain is also done to check for other abnormalities that could increase pressure within the skull.

A spinal tap (lumbar puncture) is done to measure the pressure of cerebrospinal fluid and to analyze the fluid. In idiopathic intracranial hypertension, the pressure of the fluid is increased, often to very high levels. The content of the fluid is usually normal. As soon as spinal fluid is removed during the spinal tap, the pressure inside the head decreases, the venous sinuses may widen, and more blood may flow from the brain. As a result, the headache lessens.

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These and other tests can help identify or rule out other possible causes of increased pressure within the skull (such as a brain tumor blocking the venous sinuses).

Reduction of pressure within the skull

Overweight people with idiopathic intracranial hypertension should lose weight because doing so may reduce pressure within the skull. Idiopathic intracranial hypertension may resolve when as little as 10% of body weight is lost. However, weight reduction programs are often unsuccessful.

Doctors often prescribe acetazolamide or topiramate, taken by mouth, to help reduce pressure within the skull. Acetazolamide and topiramate are effective because they reduce the amount of cerebrospinal fluid produced in the brain.

The usefulness of doing spinal taps daily or weekly to remove cerebrospinal fluid is debated. This treatment may be used when people are at risk of losing their vision while they are waiting for more effective treatment (such as surgery to relieve the pressure within the skull). If regular spinal taps are done, people are closely monitored to determine whether pressure is decreasing. CT venography or magnetic resonance venography may be done again to determine whether the spinal taps widened the venous sinuses. If these veins have widened, more blood can flow out of the brain, and pressure within the skull is reduced.

If people with idiopathic intracranial hypertension have been unable to lose weight, bariatric surgery may help. If people maintain the weight loss after surgery, idiopathic intracranial hypertension may be cured.

Preserving vision

Permanent loss of vision is the main concern in people with idiopathic intracranial hypertension. Because peripheral vision is lost early, before people notice it, an ophthalmologist periodically checks vision, including peripheral vision, using an automated device (a test called perimetry). The device produces a visual stimulus (such as a flash of light) and records the person's responses. Results are very accurate. This periodic testing enables doctors to identify problems with vision as soon as possible.

If vision deteriorates despite these measures, surgery to reduce pressure within the skull may be needed and may be able to save vision. Procedures include

- Optic nerve sheath fenestration
- A shunt
- Stenting

In **optic nerve sheath fenestration**, slits are cut in the covering of the optic nerve behind the eyeball. These slits allow cerebrospinal fluid to escape into the tissues around the eye, where the fluid is absorbed.

A **shunt** is a permanent drain made of plastic tubing. It can be surgically placed so that excess cerebrospinal fluid can be removed. The shunt is placed in the spaces within the brain or in the

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space just below the spinal cord in the lower back. The tubing is run under the skin, usually to the abdomen, where excess fluid can drain.

Stenting involves placing a wire mesh tube (stent) in the narrowed venous sinus. The stent is then opened up to widen the venous sinus.

Weight-loss surgery

If people with idiopathic intracranial hypertension are obese and other measures are ineffective, surgery to help with weight reduction (bariatric surgery, such as a gastric bypass) may be done. If successful, it may cure the disorder.

Low-pressure headaches are intense. They occur when people sit or stand and may be relieved by lying flat. People usually also have a stiff, painful neck and nausea and may vomit.

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