**Impact of Pseudotumor Cerebri (Idiopathic Intracranial Hypertension) in Pregnancy: A Case Report**

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This case report reviews anesthetic management and medical considerations for a pregnant patient with a history of pseudotumor cerebri (PTC). The 24-year-old woman, gravida 2, paragravida 0, spontaneous abortion 1, was in active labor at an estimated 38 weeks’ gestation and had been given a diagnosis of PTC 4 years earlier. This patient first experienced global headaches and blurred vision at age 20 years. At the time of onset of her headache symptoms, she underwent a full diagnostic workup and detailed neurologic examination, including magnetic resonance imaging (MRI) of the brain and a lumbar puncture. The MRI was normal. Her lumbar puncture showed elevated cerebral spinal fluid (CSF) pressures and normal CSF composition. The patient’s initial symptoms of headache and blurred vision were managed with medication and serial lumbar punctures. The patient was free of PTC symptoms on admission for labor. A lumbar epidural was placed for labor analgesia. The patient delivered a healthy infant after approximately 10.5 hours of patient-controlled epidural analgesia.

**Key words:** Idiopathic intracranial hypertension, pregnancy, pseudotumor cerebri.

Pseudotumor cerebri (PTC) was described over a century ago and is characterized by elevated intracranial pressure (ICP)—pressure in the skull—and normal brain ventricle size. It has been referred to by many names. Other terms include benign intracranial hypertension and idiopathic intracranial hypertension (IIH). The term benign intracranial hypertension has fallen out of use because this syndrome can result in blindness, a condition that is obviously far from benign. Idiopathic intracranial hypertension is the term often used for PTC in contemporary medical literature. This article will use the term pseudotumor cerebri when relating to the case study, because this is the medical diagnosis of record.

As the word pseudotumor implies, PTC is a process affecting the brain that appears to be—but is not—a tumor. Pseudotumor cerebri is a syndrome of increased ICP without hydrocephalus or mass lesion and elevated cerebrospinal fluid (CSF) pressure but otherwise normal CSF composition. It has been found that pregnancy occurs in patients with PTC at about the same rate as in the general population. Symptoms can worsen during pregnancy due to hormonal changes and weight gain, and PTC can occur in any trimester. Patients with PTC have the same spontaneous abortion rate as in the general population. Pseudotumor cerebri with IIH occurs in 19 to 24 of 100,000 obese women of childbearing age compared with 0.8 to 1.7 of 100,000 in the general population.

Pseudotumor cerebri was first described by Quincke, who called it “meningitis serosa.” This definition was then modified by Dandy and Smith to describe a condition of unknown cause, characterized by elevated CSF pressure and papilledema without hydrocephalus or abnormal CSF composition. However, these criteria do not give justice to the numerous other conditions causing papilledema. The term idiopathic intracranial hypertension and updated diagnostic criteria have been proposed for this disease.

Pseudotumor cerebri is diagnosed by exclusion of other causes of intracranial hypertension. The diagnosis of the patient with IIH is defined by the following criteria (Table 1). The patient has signs and symptoms attributable to increased ICP or to papilledema; ICP is elevated when measured during lumbar puncture in the lateral decubitus position; CSF composition is normal; there is absence of imaging evidence of ventriculomegaly or a structural cause for increased ICP; and there is absence of other causes of intracranial hypertension.

The major symptom caused by increased ICP is a headache, often worse in the morning. The etiology of the PTC is unknown, and, as already noted, the diagnosis is made after excluding other health conditions. Some examples of conditions mimicking these symptoms include venous sinus thrombosis, infection, hydrocephalus, migraine, or any intracranial mass.
lesion (eg, tumor).\textsuperscript{3,5} The mechanism causing increased ICP for patients with PTC is not well understood. Possible causes of PTC include a defect in CSF absorption, increased cerebral swelling (edema), or increased blood volumes. This condition occurs more often in women than men and in people who are obese.\textsuperscript{7}

Papilledema, bulging of the optic disc in the retina of the eye, is an important sign of PTC. Papilledema is not a diagnosis, but rather a sign of an underlying disorder. Other tests, listed in Table 2, facilitate the diagnosis of PTC. These tests include magnetic resonance imaging (MRI), computed tomography (CT), lumbar puncture for CSF pressure and composition analysis and visual field testing.

**Case summary**

The patient was 24 years old, gravida 2, paragravida 0, spontaneous abortion 1. On her admission, the estimated delivery date placed her at approximately 38 weeks’ gestation. The patient presented in active labor and requested epidural analgesia for diminution of labor pain. Two other factors complicated the care of this patient. The patient was a Jehovah’s Witness, and she possessed a clearly written durable power of attorney elucidating her firm beliefs regarding blood product administration. She informed the staff that she would refuse all blood products, even if it meant that administration of such products could potentially save her life. During the preanesthetic assessment, the patient stated that she would refuse an “epidural blood patch” if she developed a headache as result of inadvertent dural puncture. She also insisted that no blood products be administered to her newborn. The second complicating factor was that she was morbidly obese. The patient was 58 inches (147 cm) tall, and weighed 307 pounds (138 kg). Her body mass index was 46.7 kg/m\textsuperscript{2}.

Borderline hypertension had developed during pregnancy, which did not require additional treatment. She was not receiving antihypertensive medications during her pregnancy. Her daily medications included 1 prenatal vitamin; acetazolamide (Diamox), 250 mg; and esomeprazole (Nexium), 40 mg. The patient was treated with 4 lumbar punctures to control headache symptoms before this pregnancy. The last lumbar puncture was performed a few months before this pregnancy. No lumbar punctures were performed during this pregnancy.

The patient began to experience global headaches at age 20 years. She fainted during an exacerbation of an early headache. The patient stated that she had experienced blurred vision intermittently during her headache periods. At the time of onset of her headache symptoms, she had a detailed neurologic workup, including brain MRI and a lumbar puncture for evaluation of CSF pressure and composition. Her CSF pressures were elevated at 420 to 450 mm H\textsubscript{2}O (normal range, 100 to 200 mm H\textsubscript{2}O). However, her CSF composition was normal. Her MRI was normal. She also underwent a thorough ophthalmologic evaluation, including visual field testing. She stated that her ophthalmologist reported that she had bilateral papilledema but no visual field loss. The written results of her ophthalmologic evaluation were unavailable. Admission laboratory work included a

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**Table 1. Diagnostic criteria for idiopathic intracranial hypertension (IIH)\textsuperscript{3,5,6}**

| 1. Symptoms and signs attributable to increased intracranial pressure (ICP) or papilledema |
| 2. Elevated ICP as demonstrated by lumbar puncture in the lateral decubitus position |
| 3. Normal cerebral spinal fluid (CSF) composition |
| 4. No imaging evidence of ventriculomegaly or a structural cause for increased ICP |
| 5. No other cause of intracranial hypertension |

**Table 2. Diagnostic tests for pseudotumor cerebri\textsuperscript{3,5,6}**

| 1. A magnetic resonance imaging (MRI) study with magnetic resonance venography is the preferred imaging study, or a computed tomography (CT) scan. Neither study will show a tumor. |
| 2. Lumbar puncture usually confirms an elevated intracranial pressure in the range of 250-400 mm H\textsubscript{2}O cerebral spinal fluid (CSF). Normal intracranial pressure is 100-200 mm H\textsubscript{2}O. Composition of the CSF is chemically normal. |
| 3. Visual field testing is performed to detect early vision loss. |
complete blood cell count and comprehensive metabolic panel (CMP). Results of both tests were within normal limits.

After we obtained informed consent, we placed an epidural catheter, on a single attempt, between the third and fourth lumbar vertebrae, using sterile technique. We threaded the epidural catheter, confirmed a negative aspirate for CSF or blood, and then administered a test dose of 3 mL of 1.5% lidocaine with epinephrine (1:200,000). Once inadvertent intravascular or intrathecal injection was ruled out, a bolus dose of 1.5% lidocaine totaling 10 mL was administered over 5 minutes. The patient demonstrated a spread to the eighth thoracic dermatome bilaterally from the initial epidural administration of local anesthetic. Thirty minutes after the initial bolus, a patient-controlled epidural analgesia (PCEA) infusion was initiated. The PCEA solution consisted of 0.1% bupivacaine with fentanyl, 2 µg/mL. The PCEA parameters were as follows: 7 mL/h basal rate, 5 mL every 15 minutes as a programmed patient-controlled bolus, and a maximum infusion volume of 20 mL/h. The PCEA infusion continued for approximately 10.5 h during the course of labor. The patient received an additional epidural bolus containing 6 mL of 1.5% lidocaine at the time of complete cervical dilation. She delivered a healthy infant without sequelae. The epidural catheter was removed after delivery of the placenta. The patient recovered free of complications.

Discussion
The literature suggests that pregnant patients with PTC should be managed the same way as any non-pregnant patient with PTC except for drug contraindications. Pregnant patients are advised to avoid excessive weight gain. The 2 major treatment goals are to preserve vision and to improve symptoms.

Although the mechanism of action is not completely understood, most researchers agree that the disorder results from abnormal absorption of CSF. The subsequent increase in extracerebral volume leads to an increase in ICP. Because the process is slow, there is time for the brain's ventricular system to compensate, which explains why there is no dilation of the cerebral ventricles in patients with PTC. Increased ICP places additional stress on the peripheral areas of the brain. Decreased flow in the optic nerve (cranial nerve II) results in papilledema and vision changes. When the abducens nerve (cranial nerve VI) is involved, diplopia occurs.

- History and examination. The most common presenting symptom of PTC is headache, either intermittently or permanent, usually worse in the morning and with recumbent position. Pseudotumor cerebri may aggravate preexistent migraine. Monocular or binocular transient visual obscurations, varying from slight blurring to total loss of light perception, may accompany the headaches or occur independently. Lasting only seconds, they may be provoked or exacerbated by changes in posture from supine to upright, physical exertion, or Valsalva maneuvers such as straining during the active pushing during delivery. Intermittent or constant horizontal binocular diplopia usually indicates a palsy of the sixth cranial nerve, which is the only cranial nerve palsy to commonly occur in PTC.

Some patients do not have any subjective complaints, and papilledema may be discovered on routine eye examination. Occasionally, patients who are not aware of any symptoms are found to have profound loss of vision at that time. Contemporary neuro-ophthalmologic literature reveals that most patients have some evidence of optic nerve disease, such as slightly reduced visual acuity, color deficiency, a visual field defect, or an afferent pupillary defect. Sixth cranial nerve palsy may be uncovered on motility examination. Dilated fundus examination will disclose papilledema. It is often possible to grossly determine the duration and severity of the PTC from the appearance of the optic nerve.

Ophthalmologic examination is a necessary part of the diagnostic workup for patients with PTC. The most sensitive tool to assess the severity of optic nerve involvement in PTC is visual field testing. Visual field testing involves evaluating the total area in which the patient can see objects in the peripheral vision while focusing the eye on a central point. Ideally, static Humphrey perimetry should be used to assess the amount of optic nerve damage on presentation and to follow up the course of PTC. The visual field defects of PTC are similar to those found in patients with glaucoma. The visual field test detects loss in peripheral vision and provides a map of that loss, which is helpful in diagnosis and tracking of further vision loss. In patients for whom visual loss is not an issue, treatment is directed at headache control.

Hypertension can be an underlying cause of severe headaches and swollen optic nerves. An acute hypertensive crisis represents a medical emergency, and the patient should receive prompt medical attention. Caution needs to be taken to lower the blood pressure in a controlled fashion. A sudden drastic decrease of the mean arterial blood pressure in a patient with papilledema may cause optic disc infarction, a finding noted in some patients with PTC.
• **Diagnostic studies.** Conditions other than PTC have been reported to be associated with papilledema. These may include intracranial tumors, subdural and epidural hematomas (secondary to trauma), subarachnoid hemorrhage secondary to a bleeding intracranial aneurysm, hydrocephalus, arteriovenous malformations, meningitis, and arachnoid hemorrhage secondary to a bleeding intracranial aneurysm.

The patient described in this case study had a thorough workup after the onset of headache symptoms. Investigators suggest a systematic approach to the PTC workup. The first goal is to exclude an intracranial mass lesion. This is best done with MRI. If MRI is not readily available, CT of the head will disclose intracranial lesions exerting a mass effect. Ultimately, however, an MRI needs to be performed to exclude neurologic causes of PTC. In particular, venous sinus thrombosis must be ruled out by means of magnetic resonance venography. If MRI is not possible, CT angiography or conventional angiography may be necessary to complete proper imaging. An MRI with magnetic resonance venography is the preferred examination.

If the MRI is otherwise normal, the next diagnostic step is to perform a lumbar puncture. This serves to establish the diagnosis of increased ICP as well as yielding CSF for laboratory analysis to rule out infection, inflammation, or tumor cells. Except for elevated opening pressure and slightly reduced protein levels, no CSF abnormalities should be found. Repeated lumbar punctures are not necessary for follow-up of PTC. Opinions vary regarding the treatment of PTC with serial lumbar punctures to reduce ICP. The pressure-lowering effect of a lumbar puncture is short lived. The technique of repeated lumbar punctures has been effective at relieving the headache symptoms. The patient in this case study obtained sustained relief of headache symptoms after serial lumbar punctures.

Individual symptoms vary, so the course and consequences of PTC are highly variable. In one study of 57 patients, the authors found that PTC is a self-limiting condition, lasting between 1 and several years. There may be recurrences at any time. While present, elevated ICP can cause severe optic nerve damage with irreversible loss of vision.

• **Treatment.** Weight loss has been found to decrease symptoms associated with PTC. Although weight gain is a normal occurrence during pregnancy, the treatment plan outlined for pregnant patients with PTC recommends careful weight management. Obese patients may benefit greatly from a calorie-controlled diet and exercise program. Most patients will be more successful at weight management if they seek professional help. This need not be a commercial weight loss program. Drastic measures, such as surgical weight reduction, have been shown to help treat PTC in morbidly obese patients, although this would not be a treatment option during pregnancy. However, since PTC is a self-limited condition, additional long-term health benefits of such a dramatic weight loss need to be weighed against the risks of major abdominal surgery. If the diagnosis of PTC is incidental, with no subjective symptoms or visual deficits present, weight loss and periodic follow-up with visual field testing are sufficient. When symptoms or visual deficits are present, introduction of weight loss alone will not be sufficient to reduce the ICP in a timely fashion.

For medical intervention, acetazolamide, a carbonic anhydrase inhibitor, is the preferred agent. In one author’s opinion, the dosage needs to be high (1,000 to 2,000 mg/d), to exert a sufficient decrease of ICP. Other dosing intervals have been suggested. This patient required 250 mg/d of acetazolamide to keep her symptoms in remission. Side effects of this treatment include diuresis, as would be expected. Besides the diuretic effect, which usually decreases after the first weeks of treatment, fatigue, loss of appetite, paresthesias of fingers and toes, and metallic taste of carbonated beverages are the most frequent complaints. Dietary supplementation of potassium and magnesium (eg, bananas and orange juice) will help prevent depletion of these minerals. Severe side effects that have been reported include renal stones, acute tubular necrosis, hepatic dysfunction, and aplastic anemia.

Other diuretic agents have been tried with varying success. Furosemide also has been used to treat PTC. It appears to work by both diuresis and reducing sodium transport into the brain. Furosemide is a recommended second-line agent, as efficacy is reported anecdotally.

Recently, successful treatment of PTC with topiramate has been reported. Topiramate is a carbonic anhydrase inhibitor, and its use is frequently associated with weight loss. A larger controlled study will be necessary to confirm the effectiveness of topiramate in patients with PTC.

Oral corticosteroids can be considered as a short-term treatment option in a patient presenting with severe headaches, marked acute disc edema, and very high ICP with or without acute diplopia. Corticosteroids are not advocated for routine or long-term management of PTC. Withdrawal of corticosteroids may lead to a rebound increase in ICP and the side
effects (weight gain, fluid retention, and hyperglycemia) are problematic in patients with IIH. In a Web-based review of medical treatment of IIH, the author noted the following: “Paterson first reported the efficacy of corticosteroids for treating IIH in 5 of 6 consecutive patients. Weisberg has documented prompt beneficial initial responses to corticosteroids. Corticosteroids are still used to treat this disease but their mechanism of action remains unclear.” Although patients treated with corticosteroids often respond well, there may be recurrence of papilledema with rapid tapering of the dosage. This may be accompanied by severe worsening of visual function. A prolonged tapering may prevent return of symptoms and signs in some patients. Use of corticosteroids to treat patients with IIH has largely been abandoned by most neuro-opthalmologists. Long-term corticosteroid treatment of PTC should not be considered. Aside from the potential serious side effects of steroid therapy, the fact that weight loss is almost impossible with corticosteroid use makes this treatment option counterproductive. High-dose intravenous corticosteroid treatment may occasionally be necessary in the presence of rapidly progressive vision loss while the patient is awaiting surgery.

The constellation of PTC with severe systemic hypertension appears to be particularly vision threatening, and aggressive medical treatment and rapid surgical intervention may be necessary to avoid blindness. Such cases are rare, as PTC usually takes a slowly progressive course. But some patients who have longstanding elevation of ICP present with profound vision loss. Even a small amount of further visual decline could take away all remaining useful vision. In such a case, rapid and aggressive treatment, using intravenous corticosteroids and surgical intervention as soon as possible, is also necessary.

Aside from such drastic cases, surgery is performed when a new visual field defect develops or an existing visual field defect enlarges despite medical treatment. Less commonly, severe headaches not responding to medical treatment may warrant surgical intervention. Two surgical procedures are commonly employed to treat visual loss associated with PTC: optic nerve sheath fenestration (ONSF) and lumbar peritoneal shunt. The former is preferred to treat substantial visual symptoms. Lumbar peritoneal shunt is performed when headache is a major complaint. In lumbar peritoneal shunt surgery, a drainage device is implanted connecting the spinal subdural space and the peritoneal cavity. The technique of ONSF employs a medial or lateral orbital approach to expose the optic nerve sheath. Then multiple linear incisions or a window is made into the anterior dural covering of the optic nerve sheath, creating a CSF drainage outlet. Both procedures are effective to rapidly reduce ICP with resolution of papilledema. Shunts require long-term neurosurgical follow-up.

Pseudotumor cerebri occurring during pregnancy is not an indication for termination of the pregnancy. Careful weight management, acetazolamide, and corticosteroids have been used to treat PTC in the pregnant patient. Persistent headache during pregnancy in the patient with PTC requires close follow-up. There is a 10% risk of permanent visual impairment. Bed rest and acetaminophen for treatment of headaches may be sufficient temporizing measures until delivery. If optic nerve damage is occurring, ONSF is usually well tolerated and should not be delayed.

Anesthetic options. General anesthesia, epidural anesthesia, and subarachnoid block (spinal anesthesia) have been implemented on behalf of the patient presenting with PTC. Pregnant patients requiring placement of a lumbarperitoneal shunt and ONSF during pregnancy for treatment of progressive visual loss require general anesthesia. Pregnant patients are at increased risk of complications related to general anesthesia, such as difficult airway complications, aspiration, and surgical awareness. Abouleish et al reported on 2 anesthetic regimens employed for pregnant patients requiring cesarean delivery, in whom “benign intracranial hypertension” (IIH) had been previously diagnosed. In this report, 1 patient received a spinal anesthetic and the other received a general anesthetic. Both patients had uneventful anesthesia.

In 1999, Bedard and colleagues wrote a case report describing epidural anesthesia for a parturient with a lumbarperitoneal shunt placed to control symptoms related to PTC. In this report, an epidural catheter was placed to manage labor pain and then to provide successful anesthesia for cesarean delivery. In 2002, Kaul et al described a case of inadvertent spinal analgesia for a parturient in labor that had a lumbarperitoneal shunt placed for symptom control from PTC. The patient had an epidural catheter placed for labor analgesia. A 3-mL test dose of 1.5% lidocaine with epinephrine indicated intrathecal placement of the catheter. The catheter remained in place until after delivery. The patient experienced a dense block below the T5 dermatome level.

Active labor does not generally place the woman with PTC at high risk of visual loss. Increased ICP
occurring during labor is transient and not harmful.\textsuperscript{6} Vaginal delivery is not contraindicated. A cesarean delivery is not required, and no special precautions are necessary for anesthetic agents at the time of delivery.

- **Follow-up.** Each patient will require an individual follow-up schedule. Because visual changes occur frequently with PTC, fundus photographs are often taken at presentation. These photographs prove to be a helpful follow-up tool as part of scheduled ophthalmologic care. They provide easy documentation and enable comparison independent from drawings and different examiners.

Visual field testing is the most reliable test for progression of optic nerve damage. Progression of current visual field defects or new visual field defects are a strong indicator of progressive damage and commonly indicate the need for surgery. Visual field tests can fluctuate and, if the clinical examination does not concur with a worsened visual field test, it is prudent to repeat the test shortly before surgery is considered.\textsuperscript{5} Successful treatment of PTC will lead to rapid resolution of symptoms and visual field defects, unless permanent optic nerve damage has occurred. In such a case, the visual deficit is irreversible and may be large. Persistent optic nerve swelling can often be observed long after resolution of symptoms. Patients who have become asymptomatic need to be followed up on a regular basis and instructed to promptly report new or recurrent symptoms.

**Conclusion**
The parturient diagnosed with PTC requires close monitoring of symptoms related to increased ICP. Pregnancy can exacerbate the symptoms of headache and contribute to further visual field deficits. There is no reason to terminate a pregnancy for someone diagnosed with this condition. Treatment of pregnant patients with PTC is the same as for nonpregnant patients with this condition. Other than careful weight management, and certain medication precautions, obstetric care is no different from that of normal pregnant patients.\textsuperscript{3,9,23} Worsening symptoms that are related to increased ICP require ophthalmologic and neurologic consultation. Anesthetic management of the asymptomatic pregnant patient with PTC is no different than that of a normal parturient. The patient in this case study received an epidural for labor analgesia. The medications administered via the epidural catheter during labor successfully ameliorated the discomfort associated with labor contractions.

**REFERENCES**

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