

Expert Opinion

The Management of Pseudotumor Cerebri During Pregnancy

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Key words: pseudotumor cerebri, pregnancy, lumbar puncture

Abbreviations: PTC pseudotumor cerebri

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The management of pseudotumor cerebri (PTC) in pregnancy, while not a common problem, raises several difficult issues.

CLINICAL HISTORY

This 20-year-old woman developed mild headaches and was found to have papilledema in December 1998. Her height was 5'4", and her weight was 205 lb. A neurologist and an ophthalmologist diagnosed pseudotumor cerebri after an evaluation that included a magnetic resonance imaging (MRI) scan of the brain with normal findings, a lumbar puncture revealing an opening pressure of 31 cm of water, and a normal cerebrospinal fluid (CSF) examination, except for a decreased protein level. She was prescribed acetazolamide (Diamox), 500 mg twice daily. When I saw her in September 1999, she was 5 months pregnant and had continued taking the acetazolamide throughout the pregnancy. She reported having had no headaches for the previous 4 months and no visual symptoms. Her weight was then 220 lb. The findings of the physical examination were normal except for mild optic disk edema and enlarged blind spots.

Question.—How would you recommend managing PTC during pregnancy?

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EXPERT COMMENTARY

The patient was diagnosed with PTC prior to the beginning of her pregnancy, based on the modified Dandy criteria for diagnosis: (1) symptoms and signs of increased intracranial pressure; (2) normal neuroimaging studies, except for an empty sella; (3) CSF pressure of 250 mm of water or greater measured in the lateral decubitus position with the patient relaxed; (4) normal CSF contents; and (5) other causes excluded.

Pseudotumor cerebri is a syndrome of increased intracranial pressure without hydrocephalus. It generally occurs in obese women of childbearing age. The most common symptoms are headache (in more than 90% of patients) and visual disturbances (up to 70% of patients), including transient visual obscurations, visual field loss, and loss of central visual acuity. Many patients experience pulsatile tinnitus, a symptom that must be specifically queried since it is not often volunteered. Other symptoms include neck or back pain, dependent edema of the extremities, diplopia (usually from unilateral or bilateral abducens palsies, a nonlocalizing sign of increased intracranial pressure), facial palsy, ataxia, and paresthesias.

Many types of medication are associated with the development of PTC, some only reported anecdotally. This patient did not use tetracycline or vitamin A-containing medications prior to her diagnosis. A thorough medication history is imperative, since PTC is also associated with various antibiotics, and "PTC without papilledema" may be a manifestation of analgesic overuse headache.¹ Although there is no in-

creased risk with oral contraceptives, cases of PTC have been reported with Norplant use.

The only case-control study looking at PTC in pregnancy showed no increased incidence compared with a population matched for age and parity.² About 15% of women with PTC seek medical attention for symptoms during pregnancy or relate the onset of symptoms to a prior pregnancy. Weight gain, fluid retention, and hormonal fluctuations are common to both conditions. Most patients develop symptoms during the first trimester. Some women relapse with subsequent pregnancies.

There is no contraindication to pregnancy in women with PTC, and most patients can be managed during pregnancy with minimal intervention. There is no demonstrated contraindication to fertility treatments in patients with PTC. The prenatal vitamins should, however, be checked for their vitamin A content. I generally advise discontinuing any nonessential medication and treat patients with occasional lumbar punctures if needed. Weight gain should be limited to 20 lb. Acetazolamide can be used after 20 weeks if necessary, but thiazide diuretics should be avoided.

Since the most worrisome feature of PTC is visual loss, patients should be followed throughout their pregnancy by an ophthalmologist or neuro-ophthalmologist to monitor their vision. Monitoring should include quantitative visual field testing. In a stable patient with no visual loss, visits every 2 to 3 months are adequate. Patients should be told to call if they detect any change in their vision in the interim. Generally, visual loss in PTC occurs dramatically in the presence of papilledema. However, treatment should not be based on the appearance of the optic nerves alone, since many patients have residual optic disk elevation after their intracranial pressure normalizes.

If there is significant visual decline, the patient may be treated acutely with corticosteroids and a surgical procedure performed to relieve the increased pressure. Depending on the local expertise, either optic nerve sheath decompression or lumboperitoneal shunting is acceptable. Optic nerve sheath decompression usually requires less anesthesia time, always a consideration during pregnancy.

Medical treatment of chronic headaches associ-

ated with PTC is particularly challenging during pregnancy. Analgesic rebound headache occurs in patients with PTC, and some women have worsening of previous headaches related to hormonal changes. Thus, not all headaches in pregnancy complicated by PTC are caused by increased intracranial pressure.

I instruct patients to submit a diet and symptom journal and recommend a low tyramine diet.³ None of the commonly used prophylactic medications are considered category A (safe to use during pregnancy). Tricyclic antidepressants must be discontinued 2 weeks prior to delivery, and no serious adverse reactions have been reported using propranolol for the treatment of gestational hypertension. The safest analgesics are meperidine or acetaminophen with codeine, but neither is appropriate for frequent use. Occasional lumbar punctures can often ease headache symptoms. Surgery is not advocated for the treatment of headache alone.

The prognosis for PTC in pregnancy is excellent for both mother and baby. Some patients may continue to have symptoms after delivery, but others will have remission or a fluctuating course. Women with PTC who are considering starting a family should be advised to plan each pregnancy, in order to adjust medications in advance and to arrange appropriate monitoring by a neurologist and ophthalmologist. Communication with the obstetrician throughout the course of pregnancy is imperative, although PTC does not generally place a woman at "high risk." The increased intracranial pressure occurring during labor is transient and not harmful. A cesarean section is not required, and no special precautions are necessary for anesthetic agents at the time of delivery.

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