When a new patient enters your office complaining of visual loss, you have a structured way of assessing the problem. But how should you handle the differential diagnosis when that patient is a pregnant woman?

To a large extent, it should be business as usual. “I think the first thing a neuro-ophthalmologist needs to know is that the approach to evaluating symptoms in the pregnant patient is the same as it is in the nonpregnant patient, with almost no exceptions,” said Kathleen B. Digre, MD, at the University of Utah in Salt Lake City. “That is because the most important thing is to diagnose the problem. Once you have that, then you have a chance of treating the problem.”

**Visual Shifts**

Many women experience visual changes during pregnancy. “Pregnancy itself can cause changes in refraction,” Dr. Digre said. These refractive changes are thought to be due in part to progesterone-mediated fluid shifts in the cornea—and these same fluid shifts may lead to complaints of blurred vision and intolerance to contact lenses.

However, any visual complaint that develops during pregnancy should prompt a thorough workup. “The physiologic changes in pregnancy make vascular conditions more frequent, including intracranial arteriovenous malformation, aneurysm, retinal artery occlusion, spontaneous orbital hemorrhage, and pituitary apoplexy,” said Andrew G. Lee, MD, at the Methodist Hospital in Houston. In addition, he said, “Some tumors—such as pituitary adenomas, meningiomas, and orbital schwannomas—grow during pregnancy because of hormonal shifts. These are not caused by pregnancy but will be brought to medical attention during that time.”

**Leading Suspects**

Pregnant patients who present to a neuro-ophthalmologist are most likely to be affected by one of the three following conditions, the experts said.

**Preeclampsia and eclampsia.** “We must always be on the lookout for severe preeclampsia or eclampsia,” Dr. Digre said. “It’s essential to get a blood pressure check on every pregnant woman who walks in the door. Eclampsia can affect the entire visual pathway.”

The leading systemic signs of preeclampsia are hypertension and proteinuria; eclampsia is defined by the development of convulsions and/or coma in a woman who is already preeclamptic. “Ophthalmic manifestations of severe preeclampsia and eclampsia include optic disc edema; hypertensive retinopathy; choroidal infarction; serous retinal detachment; and disorders of higher cortical function, such as alexia, simultanagnosia, and cerebral blindness due to stroke or posterior reversible encephalopathy,” Dr. Lee said.

**Papilledema.** When seen during pregnancy, papilledema commonly is due to idiopathic intracranial hypertension (IIH). But, said Dr. Lee, “It also may signal underlying cerebral venous sinus thrombosis, as the hypercoagulable state of pregnancy can pro-
duce thrombotic complications.”

“In our practice, we see about one pregnant woman every few months,” most of whom have IIH, Dr. Lee said. “The reason for this is that IIH is a condition of young females of childbearing age, and IIH is associated with weight gain, so you naturally have an overlap between the two conditions.”

“IIH is one of the most common conditions I see, whether the patient is pregnant or not,” said Julie Falardeau, MD, at Oregon Health & Science University in Portland. Management is very similar, she said, but “we want to follow pregnant patients a little more closely; and weight loss, which is usually recommended for treatment of IIH, is not an option” in this patient population.

If a pregnant woman has IIH, this does not automatically make the pregnancy high risk, said Dr. Lee. “The decision for vaginal delivery or cesarean section should not be determined by IIH. In addition, IIH is not a rationale for terminating an otherwise healthy pregnancy.” IIH in pregnancy does highlight the issue of medication and teratogenicity concerns, however (see “Treatment Considerations” and “More Online,” below).

Optic neuritis. As with IIH, optic neuritis (ON) commonly affects women of childbearing age and may present in pregnancy, Dr. Digre said. “It can occur early in pregnancy, but it usually occurs afterward because of the beneficial immune changes that take place during pregnancy,” she said. “Pregnancy is somewhat of an immune-privileged state.”

ON related to sarcoidosis also may improve during pregnancy; however, the opposite may be true in pregnant women with lupus.2

Diagnostic Considerations

When it comes to the diagnostic workup, “We can use almost any diagnostic procedure that we would normally use,” Dr. Digre said. Still, some considerations should be kept in mind.

Shield the abdomen. This should take place with computed tomography (CT) scans and angiograms.

Consider risks and benefits. Magnetic resonance imaging (MRI) is “often used routinely” during pregnancy, Dr. Digre said. However, gadolinium is an FDA class C agent (see “Know the drug classifications,” below); thus, the risk-benefit profile must be considered.

This is also the case with fluorescein angiography, as the drug used is class C and crosses the placenta. “But if you need to use it to make the diagnosis, then use it,” Dr. Digre said.

With regard to MRI and gadolinium, Dr. Lee said, “We use a noncontrast study, although that might limit our ability to make a diagnosis.” One way to increase diagnostic ability is to use fluid attenuation inversion recovery (FLAIR) images.2 But, in general, he said, “If the condition is life- or vision-threatening, then the benefits of gadolinium and FA will outweigh the risks.”

A note on blood work. Given that pregnancy is a hypercoagulable state, a hypercoagulability workup should be performed in addition to standard blood work in patients who experience arterial or venous occlusive events in pregnancy. This may include activated protein C, Factor V Leiden, functional assays of antithrombin III, and homocysteine levels.2

Treatment Considerations

The “business as usual” approach may need to shift somewhat when it comes to treatment, Dr. Digre said.

Know the drug classifications. Every ophthalmologist should know the FDA drug classification system, Dr. Digre said.

Dr. Lee noted, “FDA class A drugs are safe, but we don’t have a lot of those. Most of what we use are class B or C.” For instance, “The immunomodulatory agents used to treat multiple sclerosis [MS] are Copaxone [glatiramer], which is class B, and the interferons, which are class C.”

Technically, said Dr. Digre, class B is defined as “animal studies show risk, but no evidence of risk is apparent in humans,” and class C is defined as “risk is unknown but cannot be ruled out.” In practical terms, she said, these two classes translate to “if the condition warrants the drug’s use, then use it.” However, she said, “Avoid those in the D and X classes—those definitely could present problems.”

Consult a specialist. Dr. Digre recommended partnering with a high-risk obstetrician or a perinatologist. “They’re used to these issues. If you have a partnership, you can work out the best treatment plan.” And you can ask them, “Is this medication going to cause a problem or not?”

Postpartum Notes

Some conditions may take a hiatus or even improve during pregnancy because of increased immune tolerance. “MS and other autoimmune conditions tend to be better during pregnancy, but flare-ups can occur after delivery,” Dr. Lee said. Women with MS, for instance, are more likely to have a demyelinating attack postpartum.

Two other postpartum conditions warrant mention. Horner syndrome has been reported following epidural analgesia, but it is usually a transient condition.1 Chiari-like tonsillar herniation (sometimes erroneously referred to as Chiari I malformation) has also been reported; it may follow epidural analgesia or be caused by intracranial hypotension or intracranial hypovolemia.1


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MORE ONLINE. For a discussion about Diamox in pregnant patients with IIH, see this article at www.evenet.org.
When it comes to pregnancy-related treatment dilemmas, Dr. Digre said, “Most of the calls I get run along the lines of, ‘Can I use Diamox in this patient?’”

**Teratogenic concerns.** Diamox (acetazolamide) is a mainstay of treatment for IIH. However, based on animal studies indicating that the drug could cause limb malformations, the drug was ranked as a class C medication, and ophthalmologists have tended to steer clear of using it in pregnant patients. “Neuro-ophthalmologists and neurologists usually take over the treatment of IIH, as most ophthalmologists don’t feel comfortable with prescribing acetazolamide,” Dr. Falardeau said.

**Reconsidering the issue.** Earlier this year, Dr. Falardeau and her colleagues at Oregon Health & Science University published a study on the issue. “Initially, we just wanted to look at the data and see how many pregnant women had taken Diamox—and then, of those, how many ended up with babies who had birth defects,” Dr. Falardeau said. However, in looking at the published literature, “We couldn’t find a single case of Diamox-related limb malformations in human offspring,” she said.

This prompted the Oregon researchers to take the next step. They sent questionnaires to women with IIH and their physicians; the questionnaires focused on IIH diagnosis, obstetric history and outcomes, and pediatric outcomes.

The researchers received data on 158 pregnancies. Acetazolamide was prescribed during the first trimester in 50 of the 158. There was no evidence of an increased risk of spontaneous abortion and no evidence of an increased risk of birth defects.1

**Teaching points.** Despite the study findings, “I’m still extremely careful,” Dr. Falardeau said. “I would prefer not to use Diamox during the first trimester.” If a patient is already taking Diamox and is doing well, and then gets pregnant, “I may recommend that she get off of the medication,” she said. “Why take a chance?”

If there’s no threat to vision, Dr. Falardeau prefers watchful waiting. However, when that is no longer a prudent approach, “Then it becomes a process of balancing risks and benefits, and I tell the patient, ‘You are obviously at risk of getting worse, and we need to consider this.’”

When this is the scenario, it’s especially important to take the time to walk the patient through the drug’s risk-benefit profile, Dr. Falardeau said. “Often, these patients are young women. They’re scared. We need to be mindful of their concerns.”

Dr. Lee offered an alternative to Diamox: “Sometimes in patients who do not wish medical therapy, serial lumbar punctures can be used as a temporizing measure until delivery.”

With regard to the course of IIH in pregnant patients, Dr. Falardeau said, “Most of my patients had the condition prior to becoming pregnant. Beyond that, I’ve seen a little bit of everything—women who got worse during pregnancy, those who got better, and some whose condition held steady.”

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