Dr. Prem Subramanian
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Rebound Intracranial Hypertension: Clinical Presentation and Treatment

I'm Prem Subramanian, neuro ophthalmologist and orbital surgeon here at CU, and we're going to go to the other side of intracranial pressure, intracranial hypertension, and in particular, rebound clinical or rebound intracranial hypertension, how it presents clinically, and what we might do to manage patients.

These financial disclosures are not relevant to what I will be speaking about today.

As many of us are aware, the relationship between skull base defects and CSF leaks may originate with high intracranial pressure to begin with. So, patients who have typical risk factors for elevated intracranial pressure, being young, female, overweight or obese, or having gained weight recently, are the ones who are at greatest risk for having intracranial hypertension.

This may or may not be symptomatic, and in fact, if it is chronic, if they have perhaps mild papilledema, optic nerve swelling, but no symptoms, it may never be discovered. This chronic ICP elevation leads to thinning of bone at the skull base. and spontaneous CSF rhinorrhea or otorrhea. And interestingly, in my practice at least, and when we look in the literature, it is not typically associated as much with spinal CSF leaks.

In any case, up to half of spontaneous CSF leaks do originate in patients who had high intracranial pressure. This skull based defect develops. The dura ruptures, you get CSF leak, and it's like a built in shunt, right? You tip over into intracranial hypotension, although, again, these patients may not have some of the symptoms that other patients have.

And then they come to our attention, and patients get skull base leak repair. Uh, bony as well as soft tissue repair. And if the leak occurs again, you get low intracranial pressure. But if that repair holds, you have the potential to then get high intracranial pressure. And I already mentioned that patients who are in the demographic for idiopathic intracranial hypertension may be at greater risk for this recurrence of the ICP disorder and tipping over into elevated ICP.

Now, a logical person would think that the signs and symptoms of elevated ICP are going to be profoundly different than those of low infracranial pressure. So it should be trivial to differentiate them, right? Well, we all know that that is not true. And in fact, this slide lists side by side for you symptoms of low intracranial pressure and elevated intracranial pressure.

And you can see so much overlap, right? Positional headache with either. Again, classically, low ICP headache is worse with sitting up, while high ICP headache should get better. Not always true, though. But hearing changes versus pulse-synchronous tinnitus, one must ask really good questions to differentiate those properly.

Diplopia can happen in either one because either high or low ICP may put the sixth cranial nerves on stretch, leading to esotropia, crossing of the eyes, and a distance double vision. And then, accompanying symptoms like light sensitivity, nausea, and dizziness may be seen with either disorder, while transient visual obscurations because of papilledema, swelling of the optic nerve in response to elevated ICP, is much more characteristic of high ICP disorders, while light sensitivity may be more likely to occur with low ICP disorders.

But again, a lot of overlap, and we can't necessarily tell them apart just on the basis of symptoms. Let's talk a little bit about typical pseudotumor cerebri disorders of elevated intracranial pressure and the signs and symptoms that are associated with those, because this is what I am looking for as a neuro ophthalmologist when a patient presents to me with a history of prior intracranial hypotension, skull base repair, and now new symptomatology.

So, this is a case of a typical patient with elevated intracranial pressure. She is 26 years old, has had migraine headaches since age 15, but she now has a new daily headache that's worse in the morning, but it doesn't really seem to be positional. She does report that at times her vision seems to be tunneled, but she has no double vision, and in between these brief episodes of tunnel vision, her vision is normal.

Now, patients who have swollen optic nerves can get all types of visual disturbances, but most classically, they may have a transient blackout of vision that then reverses. It comes on with positional change, often bending down and then standing back up, or getting out of bed very quickly in the morning.

It is very important to discriminate true transient visual obscuration, which has no association with lightheadedness or dizziness, with orthostatic hypotension, which is the constellation of dimming or graying of the vision, visual alteration, along with a sense of lightheadedness, dizziness, even feeling like you're going to pass out upon standing or sit quickly from sitting or lying down. But this patient was light sensitive.

She had medications to treat for migraine headache as well as a birth control pill. She does not smoke, drinks a beer a day, and takes no other supplements. There are supplements like vitamin A and certain medications that are also associated with intracranial pressure elevation.

That's always important for me as a physician to consider when a patient who has, again, a history of a skull base defect, spontaneous CSF leak that has been

repaired. If they are put on one of those medications that could put them at risk for ICP elevation, critically important to be aware of that.

The patient's visual acuity was normal. She had no pupillary defect. Her extraocular movements were full, consistent with her not experiencing any double vision. Her front of her eye was normal, and then when we looked in the back of her eye, we found that she had this optic disc swelling bilaterally. The optic nerves here are swollen, the margins are blurred. The vessels are a little bit obscured, as shown here. This optic nerve is a little bit less swollen on the right side than on the left eye. So this is consistent with the diagnosis of elevated intracranial pressure. This next slide indicates varying severity of optic nerve swelling.

This is a normal optic nerve, and it goes all the way up through obscuration of various features on the nerve up to where we see here a complete obliteration of all of the typical landmarks that we see in the optic nerve when we look in the back of someone's eye. There is a correlation, broadly speaking, between the severity of the optic nerve swelling and the likelihood of a patient having vision loss from it.

So, when I see these more advanced levels of optic nerve swelling, I become quite concerned and will be more aggressive in terms of my intervention to try to treat that intracranial pressure elevation. Visual field testing, for those of you who are patients, you may have had this done if you have experienced optic nerve swelling.

Visual field testing is very critical for helping us to follow the potential visual consequences of papilledema, because we can't tell, I can't tell from looking at an optic nerve, just if the vision is going to be normal, mildly affected, moderately affected, or severely affected. I already mentioned that the severity of the swelling can help me, but it is by no means 100%, and we need these additional tests to help us to know what is going on with our patient's vision.

So, the question becomes, what are we going to do next, right? Do we need to get an MRI scan? When patients are newly diagnosed with ICP disorders, with elevated ICP disorders, they get an MRI to make sure that there is not a structural lesion within the brain, like a tumor or something else, that could support that.

And similarly, there are findings like this, an empty sella turcica, where the pituitary gland sits, that is taken as evidence for elevated ICP. The problem with that is, once the empty sella occurs, it never goes away. So, if a patient had high ICP. They got a skull base defect, a CSF leak, ends up with low ICP, and then it got repaired, that cell stays empty.

It doesn't refill. So, the problem is, is I can't use that as a sign of anything. It just tells me that the ICP is elevated, was elevated at some point in time. An MR venogram is often useful for looking to see if there are clots, looking to see if there is a characteristic narrowing of the distal transverse sinus that can be seen in elevated ICP.

And that is a more sensitive and specific sign in the sense that when ICP goes down back into a normal range, or we don't want it to go into the too low range, but when it goes below an elevated state, that stenosis in the venous sinus may open back up. So, the presence of a distal transverse sinus stenosis is supportive, because remember, again, papilledema is pretty specific for elevated ICP, but all those symptoms that we saw before are not.

So, if the patient comes to me, maybe they haven't developed papilledema yet. Maybe their ICP has started to rise and rebound and go into the high range, and I'm going to do additional things to try to make that diagnosis. Now, what's the next step? Do we need to do a lumbar puncture? I would suggest that that is generally not going to be necessary.

We don't want to do something, especially in patients who may have had spinal CSF leaks in the past, that is going to put them at risk for developing yet another spinal CSF leak. As I mentioned before, in my practice, it does seem that the patients who tip over into a clinically detectable papilledema involving elevated ICP state had skull base defects and not spinal defects, but it's always possible.

I don't think it's necessary to get lumbar puncture in patients under these circumstances because there is clinical evidence, papilledema, that shows that the ICP is a problem. So what are we going to do next? Maybe get an MRI to make sure there's no infection or inflammation or something like that based on the imaging characteristics.

But then I think empiric treatment, medical management with acetazolamide to lower CSF production, to lower the intracranial pressure, is probably the best first step because if we can at least get the ICP to come down and then get the papilledema to start resolving, then we can work on other things.

Other things like lifestyle modifications, for better or worse. These disorders of ICP elevation may be associated still with persistence of those risk factors for elevation, and in particular, body habitus BMI. And there is a good amount of literature that shows that if patients have a idiopathic intracranial hypertension type of picture, and they lose somewhere between 6 to 10 percent of their body weight from where they are at the time that the elevated ICP develops,

then that can put the ICP problem into remission. It can result in the intracranial pressure coming back down to normal, making the papilledema go away, helping the symptoms to go away, and equally importantly, protecting patients from having to have further surgery to repair a new skull base leak that we are always concerned will happen if there is persistent ICP elevation that is not treated.

But sometimes we have to do more surgery. Not, again, not necessarily to repeat, repair that leak, because hopefully it hasn't happened again, but we may need to do optic nerve sheath fenestration if the papilledema is really bad and it is threatening

the vision. Shunting may need to be done, and there are, as many in this conference know, there are some surgeons, when they repair a skull based defect, they will put in a shunt in all patients at that time because they are concerned that they could develop a rebound intracranial hypertension.

That's not my practice. I think that putting a shunt in someone who doesn't necessarily need it is not really indicated, but we will do it if we cannot control the intracranial pressure with medical therapy or lifestyle modification, and of course if the patient is showing us that they are at risk for persistence of their condition, worsening of their condition, and potentially weakening their previous repair and ending up chasing their tail now with a low ICP state again.

Venous sinus stenting may be a good option if there is one of those distal transverse sinus stenoses that is associated with the ICP being elevated, a nice treatment that can then prevent a need for further invasive open surgery. So, ultimately, our long term goals are to prevent breakdown of the original repair of the CSF leak, to address the underlying risk factors, and to minimize our patients' symptoms so that they can get on with their lives and function like they want to.

In conclusion, a CSF leak is often caused by elevated ICP, and rebound may be more common with intracranial versus spinal leaks. The symptoms can definitely overlap, underlying risk factors need to be identified, and ophthalmologic examination to look for papilledema is a really important part of trying to determine If a patient has had a recurrence or a rebound into the high ICP side of things, and treatment is aimed at ICP reduction before a new leak occurs.

Thank you very much.