Dr. Jason Barnes:

Welcome back to ENT in a Nutshell. My name is Jason Barnes, and today we are again joined by Dr. Matt Carlson and we'll be discussing CSF leaks of the temporal bone. Dr. Carlson, thanks again for being here.

Dr. Matt Carlson:

Thanks for having me.

Dr. Jason Barnes:

When we have patients with temporal bone CSF leaks, how do they typically present?

Dr. Matt Carlson:

So most of the time, patients will present with unilateral effusion. So they'll have a conductive hearing loss and by the time they've reached my office, frequently they'll already have had a PE tube placed. And what I always say when I am talking with our residents is a person who continuously drains clear fluid out of their ear tube you should be suspicious for it, but really the history that I think is really almost diagnostic for it is the person who says every morning they wake up and their pillow is completely saturated by clear fluid. Otitis media, acute otitis media, chronic otitis media, those sorts of diseases, really don't produce that sort of fluid, and that really should make you have a high suspicion for CSF leak of the temporal bone. You can also present with rhinorrhea specifically, and that would be through an intact drum and you're leaking through your nose. And again, that might be more common after you've been sleeping all night and you stand up for the first time in the morning. And then lastly, uncommonly you may present for the first time with meningitis or an intracranial complication of the CSF leak, and I would say that that probably occurs in less than 10% of cases.

Dr. Jason Barnes:

And can you tell us what the Dandy maneuver is?

Dr. Matt Carlson:

So a Dandy maneuver is a maneuver that's used to provoke egress of CSF out of the nose. So typically you have a person lay supine for a period of time and then you have them lean forward, and a Dandy maneuver to be positive, or at least to be indicative of a temporal bone CSF leak, it should be unilateral and it should be ipsilateral to the side of the CSF leak. I'm sure it can happen, but I've never seen a person with a left-sided temporal bone CSF leak have bilateral or unilateral right-sided rhinorrhea. So it should be on the same side and it should be very clear fluid.

Dr. Jason Barnes:

And I want to then move on to pathophysiology of what causes this. We won't discuss too much trauma-induced CSF leak, but can you speak briefly about that?

Dr. Matt Carlson:

Yeah. So the etiology of CSF leak is varied. Traumatic etiology can be both from penetrating or blunt trauma, such as motor vehicle accidents or gunshot wounds or other sorts of events that could happen from an altercation. Also under traumatic, you can put iatrogenic in there and that can be postoperative. So a postoperative CSF leak after a translabyrinthine tumor removal, for example, would be within the

bin of traumatic. And then on the other side, you have non-traumatic, and within that are multiple categories.

There's a growing number of spontaneous CSF leaks that are occurring. That is, there's no history that would predispose that you could elicit, at least that would predispose the patient to developing a CSF leak. You can also have congenital CSF leaks, and there are sometimes situations where you have a congenital anomaly that results in a CSF leak later in life, so even a congenital defect might not mean that you're going to get a CSF leak when you're a child but it could predispose you to later. So examples of that are a Hyrtl's fissure, persistent tympanomeningeal canal, for example. Inner ear malformations such as X-linked gusher, for example, can predispose you to meningitis because essentially the inner ear becomes a common cavity with the subarachnoid space, and so if you get an infection of the inner ear that can transmit to the subarachnoid space in the brain and cause meningitis.

Outside of our talk today, but you can also have lateral sphenoid recess leaks that are from Sternberg canal, for example. So you can have some defects that are related probably at least to some level of congenital development that can present later in life.

Those would be the most common different categories of CSF leaks.

Dr. Jason Barnes:

And when we focus less on congenital and traumatic or iatrogenic CSF leaks, what is the cause of CSF leaks in these patients?

Dr. Matt Carlson:

So if you're specifically alluding to spontaneous CSF leaks, historically we would call these idiopathic or as the name implies spontaneous, meaning we don't have an underlying cause that's known. But the more we learn about these, the more likely is the case that this is related to a predisposing condition of elevated intracranial pressure, and this can be more commonly related to somebody who is obese, overweight, and has findings consistent with idiopathic intracranial hypertension. There are other conditions where you can have elevated pressure that's not related to idiopathic intracranial hypertension. That could be from a dural sinus thrombosis, for example, or dural sinus stenosis, which is less common. You could also have a tumor or something else that's resulting in hydrocephalus, for example. But the theory is that at least with idiopathic intracranial hypertension, people with central obesity have increased abdominal girth which pushes up on your thoracic cage, your thoracic center, and that reduces your venous return, which is largely passive. It cycles with respiration but it's a largely passive process, and that basically creates elevated back pressure on your whole system.

CSF is produced through an active system through choroid plexus and the ependymal cells, but its resorption is through a passive process that's primarily mediated by the differential pressure between your subarachnoid space and your venous system out of your head. And so going back to that, if you're overweight you might have reduced venous return, which backs up pressure inside your head. In the short term that's not a big issue, but over time it just can erode and lead to defects that can cause CSF leaks. So the dura is constantly pulsating, and the erosion you might get with elevated pressure is kind of likened to the idea of erosion on the shore of a lake, for example, with the constant lapping of the waves.

Some people can develop thinning of their skull base over time. Naturally, everybody develops thinning of their dura over time. When you're over the age of 60 or 70, your dura is definitely more fragile and more thin. Then less commonly, there are some conditions that result in abnormal bony metabolism resorption that might predispose you to develop CSF leak.

Dr. Jason Barnes:

Now, you mentioned idiopathic intracranial hypertension, and you kind of described the typical patient that presents with idiopathic intracranial hypertension. I thought it would be worthwhile to talk a little bit more about CSF physiology. You mentioned that CSF is produced in the choroid plexus and ependymal cells. What's a normal CSF opening pressure and what is the definition of idiopathic intracranial hypertension?

Dr. Matt Carlson:

So those are really very pertinent questions. Of course, a typical opening pressure is less than 15 cm of water, and I think there's a couple important things to mention. The first is a person with an active leak, your opening pressure might not be accurate, particularly if it's a high-volume leak. So for example, if you did a lumbar puncture on somebody that had an active CSF leak and their pressure was 25, that should make you think that the pressure is probably a lot higher than that if they had that area sealed. And so even if you have a normal opening pressure on somebody you suspect of having underlying idiopathic intracranial hypertension, for example, as a cause, normal pressures should not lead you to believe that they will be normal after you do a repair.

If we talk about CSF production in general, the average adult... So CSF production changes over a person's lifetime as does pressures. The average CSF production for an adult is about 500 cc and at any given time your body contains about 150-200 cc, and so if you understand that you'd understand that you cycle about two or three times your volume throughout the day. There are some situations where you have more production or less production, but that's typical.

So when we talk about pressures in general I said, again, 15 or less is normal. We kind of have this buffer zone between 15 and 20. We tolerate it. It's not normal, but we say that's not really that elevated. But once you start getting over 20 and particularly when you get over 25, you start to say that this is abnormally elevated.

The condition of idiopathic intracranial hypertension, as the name implies having idiopathic tagged to it, means that you shouldn't be able to find a physiological abnormality causing it other than the things we just described of being overweight. So for example, when you get an MRI scan, you shouldn't see a tumor. You shouldn't see an ependymoma creating more CSF, for example. You shouldn't see a dural sinus thrombosis. You shouldn't see a tumor causing hydrocephalus. You should have no localizing features on your examination except for papilledema, and there is a possibility of getting sixth nerve traction from idiopathic intracranial hypertension, and that's permitted to still have that diagnosis. Your CSF taps should be essentially normal and clean. You shouldn't see anything else that would make you think that there's another underlying process. So basically everything on a scan looks pretty normal. Your CSF is clean.

When we talk about findings on a scan for idiopathic intracranial hypertension and elevated pressures, there are some features that do suggest chronic elevated pressure in the head. And the ones that are most commonly talked about are empty sella. So for example on an MRI, if you look at a sagittal film, particularly on t2s, the degree in which the pituitary gland is pushed down within the sella might be indicative of BIH or idiopathic intracranial hypertension.

There's different definitions used. The one I've heard most commonly is that if the height of the pituitary gland is anything less than about a third of the total height of the sella, then you have to start really thinking that this is an empty sella syndrome that could be related to idiopathic intracranial hypertension.

You can also get optic nerve canal kinking, and the optic nerves are unique compared to other cranial nerves in that the CSF will actually travel down the sheath of the nerve all the way to the end, and that's why you can get kinking from elevated pressure and that's also why you can get papilledema on examination.

Less commonly, you can see a subtle herniation of the tonsils, and you can also see dilation of some of the openings of the cranial nerve. So for example, you could have a dilated Meckel's cave, for example, and less commonly you could have a fallopian canal meningocele or something like that.

Dr. Jason Barnes:

And more about the pathophysiology, what happens if these patients go untreated? What are some complications that they would experience?

Dr. Matt Carlson:

It's interesting. A lot of the patients we see in clinic, when they're diagnosed with a CSF leak, we'll tell them, "You should get this fixed relatively soon or you're at risk for getting meningitis." And they say, "Well, what's funny, doc, is I've been leaking like this for 15 or 20 years." And they say, "What's the rush? Do I really need to get this fixed?" And I think their question is very valid and I don't have a good answer to reply, to be honest. In the medical field, we believe you should have it fixed unless there's a risk of meningitis, and that's what basically the standard is to recommend repair of a CSF leak. So we would tell the patient that if you did not get it fixed, you'd have an increasing risk of developing meningitis or an intracranial complication, intracranial infection that increases with time. So the longer you have it, the more likely you are to develop a leak.

In the future, we might find out that certain types of leaks are higher risk and other leaks are lower risk. For example, it might be that a person with a high output leak and elevated intracranial hypertension actually has a physiological shunt that's helpful for them. And if it's high enough flow, you'll only have unidirectional flow and you might not get meningitis. But to be clear, that's not our recommendation right now. Our recommendation is to fix any diagnosed CSF leak.

Dr. Jason Barnes:

When you suspect a CSF leak in a patient in your clinic, what else is on the differential diagnosis?

Dr. Matt Carlson:

So they'll typically... Again, though, if you're talking about a temporal bone CSF leak, by far and away the most common presentation is ear fullness with a little bit of conductive hearing loss, and that's related to the clear effusion. It's not uncommon to go undiagnosed on otoscopy at least initially because CSF is clear and your drum is semi-translucent, so it's very easy to miss it. In fact, you usually see bubbles, air bubbles more than anything else or a meniscus rather than actually seeing the CSF itself. It's kind of like looking into a fish tank with perfectly clear glass and perfectly clean water. You wouldn't know if there's actually water in there or not from a little distance. It's the same idea. But certainly the audiogram showing a mild conductive hearing loss and type B small-volume tympanograms ultimately leads most people to getting a tympanostomy tube. And upon myringotomy, you'll get clear fluid drainage and it's often pulsatile. You'll see it pulsating within the PE tube. And then again, I find almost all of my patients will always say, is they'll say, "I'll wake up and my pillow will be completely drenched in almost a puddle of what looks like water", which to me separates it clinically from otitis media or other causes. That would be the main differential.

Of course, anybody with a unilateral effusion, you should look for a nasopharyngeal or an infratemporal fossa tumor or something that could be obstructing the eustachian tube, although it's an uncommon scenario but possible. And then just otitis media, of course, is in the differential, too.

Dr. Jason Barnes:

And when you see these patients in clinic, what are the typical audiogram findings that you see?

Dr. Matt Carlson:

So most of the time... So if they haven't had a tube before, they'll have a mild conductive... As long as they're filling up their ear with CSF, they'll have a mild conductive hearing loss, usually nothing greater than 20 or 30 dB for air bone gap and they'll have type B tymps, low volume if they've never had tympanostomy tube. If they have a tube or a hole, then they'll have large volumes, of course. And that would be the most indicated audiometric findings.

Dr. Jason Barnes:

So you see a patient in clinic. You suspect a CSF leak. They have a PE tube in place. What's your next step in terms of diagnostic workup?

Dr. Matt Carlson:

So the best way to make the diagnosis of a CSF leak is to get Beta-2 transferrin. Beta-2 transferrin is highly sensitive and specific for a CSF leak. You only need really a drop or two of fluid to be able to test it. It is done through gel electrophoresis, so the actual assay takes a little bit to get the results back. It's not like some of these other rapid assays that you'll get back in 20 minutes or something like that. It's usually at least a day or two until you get the results back. Beta-2 transferrin is a protein that's really only expressed in the subarachnoid space, vitreous humor, and the perilymph of the inner ear, and so it's very sensitive. There are other assays people have used. Beta trace protein is used in Europe and it's also very sensitive and specific, a little bit less so than Beta-2 transferrin.

Of historical relevance but not used today is you can look at things such as glucose, for example. It should be about 60 or 70% of what you see in your blood levels. There are some other historical tests that have been used, but by far and away Beta-2 transferrin is the most useful assay today.

Dr. Jason Barnes:

And what imaging workup do you pursue for this?

Dr. Matt Carlson:

So if they present with rhinorrhea, so unilateral rhinorrhea, you don't know if it's a... And you get a positive Beta-2 transferrin or you believe it's CSF, you don't know if it's an anterior cranial base CSF leak or a paranasal sinus CSF leak or a temporal bone CSF leak. Any part of the skull that has pneumatized air cells that communicates the external world can result in a CSF leak, of course. And so sometimes one of my colleagues in rhinology will have a patient come in with a CSF leak. They'll look on otoscopy and they'll actually see fluid in the ear, and then they send them over to us because they know it's not or they don't think it's related to a paranasal sinus CSF leak. So that's the first thing.

So then we typically will go to imaging, and a fine-cut temporal bone or fine-cut skull-based CT scan with 0.4 mm cuts with dedicated axial, coronal, and sometimes sagittal cuts, is by far and away the most valuable initial test after your Beta-2 transferrin test. And you're looking for a couple of different things.

The most obvious thing is you're looking for a natural dehisce or you're looking for dehiscence in the bone, and it's important to recognize that even 20 to 40% of everyone in the world has these natural dehiscences, and so you're looking for opacification of some air cells around the dehiscence. You want to see if it's multifocal.

People with idiopathic intracranial hypertension are more likely to have multifocal CSF leaks or encephaloceles. I'm also looking for what we call an encephalocele or a meningocele, or a meningoencephalocele. People with idiopathic intracranial hypertension are more likely to actually have a concomitant encephalocele or meningoencephalocele with their CSF leak and not just have a leak by itself. So you'll see a mass like projection into some pneumatized air cells in the paranasal sinuses or in the temporal bone. If it's a high-flow leak or if it's more voluminous leak, it's possible to get surrounding pneumocephalus. That would be more common in a postoperative setting, iatrogenic or traumatic, but I suppose it could happen with idiopathic intracranial hypertension as well.

And then the CT scan is probably the most valuable after Beta-2. You can also get an MRI. MRI is valuable to make sure you don't have something around it. There have been cases where a person will get a CSF leak around a tumor, for example. It can also give you an idea if there are signs of elevated chronic intracranial hypertension, so those things we talked about, empty sella, kinking of the optic nerves, sag of the brainstem with subtle herniation of the tonsils, or dilated openings of your cranial nerves. So it's valuable for that. You can also distinguish just opacification from fluid of CSF from meningocele, for example, so it helps you distinguish is the lining of the brain coming down or is it just fluid in that area? So I think MRI is useful to get in most of these situations. If you didn't get an MRI with it, it wouldn't be considered malpractice by any means, but I would say more and more people are getting MRI with it.

Those are the most common diagnostic things, but there are many other tests that can be done if these initial tests are non-diagnostic or more information is needed.

Dr. Jason Barnes:

So if you have a patient who you are highly suspicious has a CSF leak but you haven't been able to prove it to yourself or to the patient, what are some additional testing or imaging workups you can do?

Dr. Matt Carlson:

I think there's kind of two or three different scenarios. When you mentioned that there's two or three different scenarios you can think of, the first is the person who has recurrent meningitis. So you'd know if something is abnormal and you're trying to figure out the source for it. The second one is the person who reports symptoms that are highly suspicious of it but they can never reproduce it in clinic, and they're very sure they have something. So a person who has an intermittent leak that you can't reproduce or collect fluid in clinic, I'll do this diagnostic workup. If my suspicion is very, very low, I tend to send them home with a Beta-2 transferrin kit to collect fluid and send it back in because my pretest probability in my mind is very low, meaning I don't think it's very convincing. So no history of meningitis, their symptoms aren't really suggestive of it, but I don't want to just dismiss it and say they don't have it. I'm giving them an opportunity to produce some fluid that we can test.

If my suspicion is pretty high based on their symptoms or a history of recurrent meningitis, then I'm really going to do a lot of digging to try to find out if we can get the leak. So I will send them home with a Beta-2 transferrin assay kit so they can send in a sample, but there are some other tests you can do. You can do CT cisternography. So that's a test where intrathecal contrast is given and you'll get scans at different time periods on the CT and see if there's extravasation of the contrast. The important thing for

that is you have to have a relatively active leak to be able to see it, so if it's only leaking very intermittently you might not see it. There are radio-nucleotide scans where you look for radio-nucleotide tracers, but the localization for these tests, even with the newer ones, is quite poor and you can have false positives and false negatives. And I will say that at least for temporal bone CSF leaks, that's very infrequently used.

There is a situation where a person will say they have intermittent leakage through a PE tube, and you're suspicious for it as well. One clinical trick that we'll use sometimes is we'll place a cotton ball right in their medial ear canal right next to their PE tube. We'll tell them to go home, maybe lay down for a little bit or even lay on that ear to put more drainage into that middle ear space, and then we'll have them come back the next day and we'll take the cotton ball out and we'll send that for Beta-2 transferrin. So that can catch those intermittent cases as well.

Dr. Jason Barnes:

So we talked about workup, and I next wanted to move to treatment. If a patient comes to your clinic, you diagnose them with a CSF leak. Maybe you have a scan that demonstrates where the dehiscence is. What is the first step? And I'll more specifically ask is there a requirement for antibiotics in folks who have a CSF leak?

Dr. Matt Carlson:

So there's very limited evidence to support one direction or another. I would say the best evidence comes from some studies that were done by Dr. Hilary Brodie, but those studies were done both at his institution and also in a meta analysis where he looked at the risk of developing meningitis after having a traumatic CSF leak. So it's hard to cross apply that or extrapolate that information onto spontaneous CSF leak patients. Essentially always, if we have a patient who has a lumbar drain in, we are using antibiotics at the same time. If they've had a history of meningitis or if they have recurrent otitis media or something that you might think would predispose them to getting it, we might start antibiotics. But if they have a totally clean ear and they came in and they were tested and it was positive, and say we scheduled surgery for three weeks away, I typically wouldn't prescribe antibiotics at that time, because most of the time these patients have said that they've had this for several years. Again, those statements aren't data driven and some people might put a patient on antibiotics in that situation and I wouldn't fault them for it, but that's how I would approach that specific situation. So any increased risk of getting infection or history of meningitis, I would probably do it. But most of the time, I don't.

I think that brings up one other important point and technically it's a CDC recommendation to anybody that's at risk of meningitis and somebody who does have an active CSF leak. Technically you're supposed to offer and talk to them about pneumococcal prophylaxis. Pneumococcal meningitis is the most common sort of meningitis that you would acquire from a temporal bone CSF leak, and that can reduce your risk. It parallels the same vaccination schedule you'd get with cochlear implants.

Dr. Jason Barnes:

And is there any other kind of medical treatment that can be used for these patients or do they require surgery?

Dr. Matt Carlson:

So traumatic CSF leaks will often resolve spontaneously. There's a lot of different types of CSF leaks. The idiopathic intracranial hypertension patients and those with spontaneous CSF leaks are unlikely to just

stop on their own because they have an underlying physiological process that made it start. And so they have this driving differential pressure across the defect, and it just makes it very unlikely that it will ever resolve on its own. You could try placing the person on Diamox, do conservative measures, bed rest, head-of-bed elevation, stool softeners, antiemetics, all those sorts of things, but frankly it's basically never going to work, I think. So a person with a spontaneous CSF leak is not only more likely to need surgery, but they actually do have increased risk of having surgical failure and that wouldn't be just at the site of repair, but also developing another leak somewhere else because they often have multifocal involvement of their skull base. And again, for many of these patients, it's a vent for them. They're developing elevated pressure and it's a way for their body to take some of this pressure off.

Dr. Jason Barnes:

So in patients with spontaneous CSF leaks, you're going to recommend surgery. What are the surgical approaches and how do you counsel patients on what they should expect?

Dr. Matt Carlson:

Just to say one last thing about the last question you asked. One additional thing that we would put in the realm of medical therapy is diet. So a lot of these patients have very, very elevated BMIs. It's not uncommon that we'll repair a CSF leak in somebody with a BMI of over 40 or 50. And so just not in addition to their general health, but also weight loss in particular having a non-failure after surgery or another one open up, it's beneficial for these patients to go through diet programs and bariatric surgery potentially if they're very, very obese.

So for surgical repair, most of the literature shows that there's a lot of different ways that you can do the operation, and in most situations it's actually going to be successful. So most of the literature identifies that at least in temporal bone CSF leaks that there's an over 90% chance that your first operation will provide a durable repair at that site.

I want to mention one thing related to surgery for CSF leaks in people with idiopathic intracranial hypertension. These patients also have an increased risk of having concomitant superior semicircular canal dehiscence. So when you're doing a middle fossa, for example, you should look at your scans and scrutinize that to make sure you're not lifting up the temporal lobe and seeing an open labyrinth and not recognizing initially and causing problems. In a recent study we did, we found that 15% of patients who had temporal and CSF leaks also had concomitant superior canal dehiscence on the same side, so something worth noting.

The repair, most of the literature suggests that the more autologous the tissue and the more layers you do, the better. Larger defects typically benefit from having soft and also some sort of bony or rigid repair. The soft part of the repair like fascia, for example, will create your watertight seal, but the rigid barrier will be what basically counteracts the continued pulsations from the dura around it. So certainly for anything bigger than 2 or 3 mm, I think it's good to put some sort of rigid material in there. We tend to take part of the craniotomy from middle fossa so we can use autologous bone. At least at our center, we don't like using methylmethacrylate or bone cement for this because it's partially exposed to the middle ear and we want to not have to take it out five years later because of infection, for example.

So most of our patients with temporal bone CSF leaks, most of the time it's involving the middle fossa. Posterior fossa CSF leaks, spontaneous, that is, is extremely rare. I don't know the exact percent, but I would say it's certainly less than 5% of all temporal bone CSF leaks that are spontaneous in nature. So for a middle fossa CSF leak, at our center we'll either do a middle fossa alone or a combined mastoid and middle cranial fossa. Some centers will primarily perform transmastoid repairs, which is also another

way to do it and that is very reasonable. In our experience, the ones that are more amenable to or benefit from a combined or just having a middle fossa approach are the ones that have multiple defects in the temporal bone, ones with very large defects, ones that have very medial defects, and ones that are in the anterior epitympanum anterior to the ossicles. It's just hard to get to those areas without taking out the ossicles in patients. So those are the situations where we tend to favor the middle fossa.

The conditions we would be more likely to just do a mastoid only or transmastoid are maybe the much older patient, a person who is a little bit more infirm, a person who's failed before. If they have a very large defect and say they have had one or two failures, which is very uncommon but can happen, in those situations we're more likely just to close the ear off completely and pack the eustachian tube directly which causes a conductive hearing loss that can be rehabilitated with the bone conduction device if the patient desires. So that's a multiple recurrent scenario, which I would say is less than 5% of cases.

Dr. Jason Barnes:

And you've mentioned that basically the CSF leak is a release valve for someone who might have increased pressures. So if you close that valve that could therefore increase the pressure. So the question I'm getting at is what is the role of a lumbar drain postoperatively after you repair a CSF leak?

Dr. Matt Carlson:

That's a great question. I think that most people who have been in practice for a while maybe started off by doing a lot more lumbar drains and over time moved away from using them. And the reason is in a lot of situations, I don't think it really provides a lot of benefit, but it certainly does increase the risk of having certain complications from it. I'd say the one concern you're mainly worried about is developing pneumocephalus or tension pneumocephalus or sucking air in and causing an infection just by having that directional flow into the head. Those are the main risks I'm concerned about when we talk about lumbar drain post-operatively. On that same subject, I think the question of do you get a lumbar puncture afterwards for opening pressure after you repair on somebody you're worried about idiopathic intracranial hypertension, and I'll say that I think that, and some people might disagree with me, but my perception is that the otologists, temporal bone surgeons, lateral skull-based surgeons, tend to not get taps unless a person has multifocal involvement or if they're symptomatic postoperatively.

So if they're experiencing headaches and things like that or for the person with recurrent leaks I think rhinologists, or endoscopic skull-based surgeons are, at least based on my experience, much more likely to proactively just get a lumbar puncture with opening pressure afterwards just to see what it shows. From our standpoint, even in an idiopathic intracranial hypertension patient who has a surgical repair in the temporal bone has a pretty high chance of cure without a large risk of recurrence, and so that's probably why we're not so keen to do a postoperative lumbar puncture in everybody.

You're presented with a situation where you have a good durable repair. You don't see any other defects. They're not having headaches. You get a lumbar puncture and it shows 25. What do you do with that? I mean, I don't know. I would say you probably don't do anything with it if they're not symptomatic. So it just gives you extra information that you might not use. But I think that, again, the rhinologists look at it from a different angle and maybe we'll do that in the future, too.

Dr. Jason Barnes:

What's the role for Diamox postoperatively?

Dr. Matt Carlson:

The answer is it depends on who you talk to. We don't typically use Diamox postoperatively unless we think it's a high risk of surgical failure. If a person is still leaking postoperatively and you're trying to use that to try to improve your odds of having to do a repair, it's probably not going to work. But it's not a crazy idea to use a Diamox or acetazolamide. The idea behind Diamox is that it reduces CSF production, but at least most people anecdotally believe that there's some level of habituation, meaning it only has a transitory benefit, maybe two weeks, four weeks, or a couple months, and not much longer than that. And so most of the time, we typically don't use it postop.

Dr. Jason Barnes:

And finally, discussing treatment, what complications do you counsel patients on when you are going to surgically manage this?

Dr. Matt Carlson:

So the same complication that you're trying to avoid by having surgery is a complication you can have with surgery, so you can have infection or meningitis. I'd say the risk of having meningitis with the operation is less than 1%. If you're doing a middle fossa approach, you're doing temporal lobe elevation so it's under their dominant temporal lobe. In particular, there is a slight increased risk of having complications, and that would typically be aphasia or stroke from temporal lobe elevation or seizures. And I would put all of those. Also, if you're doing an extradural repair, meaning you're lifting up subtemporally and staying outside the dura, the risk of an intracranial event or intradural event or a stroke is very, very low, certainly less than 1%. In a lot of these patients in about 10 or 15% at least, these patients will have a dehiscent geniculate ganglion when you're elevating, and so I always say that there's a 1% chance of a temporary [inaudible 00:33:30] paresis and a one in a thousand risk of permanent facial weakness because I suppose that's a possibility, although it would be very, very rare.

Sensorineural hearing loss from doing this would be very, very rare, and it would probably be the only thing I could even think of is if you were doing a middle fossa approach and you had somebody who yanked the ossicles by accident, and that would be very rare, or if there was an undiagnosed superior canal dehiscence that wasn't seen preoperatively and somebody was suctioning vigorously over it, for example. Those would be the main complications. Infection, hematoma, bleeding, things like that, but all quite rare.

Dr. Jason Barnes:

And how do you counsel patients on outcomes and expectations following surgical repair?

Dr. Matt Carlson:

So in our experience, 90 to 95% of patients only need one operation for that site to have a durable repair and not have it recur. If patients are very heavy, we tend to tell them that they're probably going to need some sort of medical therapy afterwards. When I say that, I mean weight loss. So either we'll often refer them to see a dietician if they're morbidly obese, or if they tried diets before and they haven't done well with them, we'll send them off to a bariatric consult for consideration of surgical intervention for weight loss. We'll ask them to let us know if they're developing symptoms of elevated pressure, and that's typically first headaches and less commonly you can have visual disturbances. That would be quite rare as the first presentation of elevated pressure right afterwards. Depending on their surgical approach, we'll see them back at around three months, make sure everything looks good, see

them back in a year, and then usually we manage them expectantly. We ask them to call us if they're having any problems thereafter.

Dr. Jason Barnes:

Well, Dr. Carlson, thank you so much again for being here. I think that was a great review of CSF leaks, specifically spontaneous CSF leaks. And I just wanted to quickly review what we've been through. So someone with a spontaneous CSF leak will likely present with a middle ear effusion and more often than not will have had a PE tube placed. In clinic, you might have them Valsalva to elicit a leak either from the ear or from the nose. In terms of pathophysiology, this is often related to idiopathic intracranial hypertension, which is the increase of CSF pressure often due to a patient's being overweight and they have a dehiscence in the skull base. The reason we treat this is because there's an increased risk of meningitis and the differential diagnosis just includes other things that could cause middle ear effusion, like an otitis media.

When we consider the workup, our audiogram findings are going to be consistent with a conductive hearing loss. We'll test the fluid with a Beta transferrin protein test which will be positive because this is a very specific and sensitive test. CT findings would show a dehiscence in the bone, and an MRI might also show an encephalocele or a meningocele. Other things can be considered such as cisternography.

When we consider treatment, there's no real medical therapy for someone with a spontaneous CSF leak. We do talk about weight loss and diet, but often these CSF leaks will not resolve on their own in the setting of spontaneous CSF leak. Antibiotics are not unreasonable, but certainly aren't required and might be somewhat controversial. Lumbar drains are also somewhat controversial and probably don't need to be considered preoperatively.

When we discuss the surgical approach, we often consider a middle fossa approach which can be combined with a transmastoid approach, and some centers might do a transmastoid approach by itself, especially if it's a recurrent CSF leak. Postoperatively is when things become a little more tricky and considering Diamox and lumbar drains, but often these are not required. And the complications involved with surgery are pretty limited, but include meningitis. This is a very successful procedure with more than 90% of folks having resolution of CSF, but those who are more susceptible to this might have recurrence.

Dr. Carlson, is there anything else you'd add?

Dr. Matt Carlson:

No, I think that's great. I think that's CSF leaks in a nutshell.

Dr. Jason Barnes:

Thank you.

That brings our episode to a close, though I did want to go over just a few questions before we finish. I'll ask a question, wait for a few seconds so you can think or press pause, and then give the answer.

So the first question is what is a normal opening pressure for CSF and what is typically seen in idiopathic intracranial hypertension?

The normal opening pressure for CSF is 10-15 cm of water, though if it goes up to 20 that's still considered a gray area, and those with idiopathic intracranial hypertension will have a CSF opening pressure of greater than 25.

Our second question is what is the Dandy maneuver? The Dandy maneuver is when you ask a patient to lean forward and Valsalva, and this will oftentimes elicit a CSF leak if a patient has one.

Our third question is what is the test used to confirm CSF fluid? The test that we often get to confirm CSF fluid is Beta-2 transferrin, though beta trace protein is another one that can be used. They both have high sensitivity and specificity, and Beta-2 transferrin is more often used in the United States.

Thanks for listening, and we'll see you next time.