

Speaker 1:

Hey there! Welcome back to ENT in a Nutshell. Today, we are joined by rhinologist, Dr. Garret Choby, and we will be discussing CSF leaks. Dr. Choby, thanks again for being here.

Dr. Garret Choby:

Thanks for having me. It's a pleasure.

Speaker 1:

Today we'll be talking about CSF leaks, and I do want to say that there are different types of CSF leaks. There are some in the temporal bone, which are discussed in a different episode in the otology section, and there's also iatrogenic and traumatic CSF leaks, which we'll touch on in just a bit, but today our focus will mainly be spontaneous CSF leaks. So Dr. Choby, when a patient comes to your clinic and they have a spontaneous CSF leak, how do they usually present?

Dr. Garret Choby:

These patients are classically going to present with unilateral rhinorrhea. This may have been missed over the years and mistaken for things like allergic rhinitis or drainage, but when you ask carefully, they may say, "It's always from my left side," or, "Always from my right side." When it's unilateral, it's always a tip off to you that you got to think about CSF rhinorrhea.

Speaker 1:

Are there any other symptoms or ways they describe this fluid that can tip you off as well?

Dr. Garret Choby:

Some patients will tell you that it tastes salty. When it drips in the back, it might taste a little bit salty. Classically, they say it is mostly all the time and also when they lean forward, it can be a higher flow, that is classically is referred to as the Dandy maneuver, just leaning forward. I also find that if it's a sphenoid leak, it may come out sort of as a rush because the accumulated net sphenoid sinus, when they lean forward, it comes out as a large volume, if you will, maybe a little less so in an ethmoid or cribriform leak, but more so with the sphenoid.

Speaker 1:

When you're in clinic, maybe you'll scope a patient. Can you usually find the CSF leak?

Dr. Garret Choby:

If it's not localized on a scan before they come in to see you, it can be a little bit difficult. What I often times will do is use a 30 degree scope looking upwards in the sphenoid ethmoidal recess and have them lean forward for me and take a close look there. You can also visualize the olfactory cleft pretty well if it happens to be a cribriform leak, but it can be challenging times to see the actual site on scope exam.

Speaker 1:

And as I mentioned earlier, there is maybe some overlap in otology and rhinology here with CSF leaks, because it can come out of the nose even if it's not coming from the sinuses, right?

Dr. Garret Choby:

Yeah. Great point. It's always very important if someone presents with CSF rhinorrhea, to make sure you look at both their tegmen on their CT scan, as well as do a thorough ear exam. Because some folks with lateral skull-based leaks, they will come through the eustachian tube and into the nose, and they may present with leakage from their nose, but you really don't want to mistake that for a CSF rhinorrhea. Makes sure you get over your otology partners, if it is a lateral skull based leak.

Speaker 1:

Sure. So what's the pathophysiology involved with these CSF leaks? Why do they happen?

Dr. Garret Choby:

Classically these patients have elevated intracranial pressure. The thought is that over time that it thins the skull base until they eventually form a leak. CSF is produced in the choroid plexus and by the ependymal cells and produce up to 500 milliliters a day in total. Now, when you check-in opening pressure in these patients, when their system is sealed off, normal pressure is about 10 to 15, in general terms, and these pressures are classically very elevated. Some of them in my practice I've seen elevated 30, 40, even 50 in some cases.

Speaker 1:

And what other patient characteristics will you see in these folks?

Dr. Garret Choby:

Classically these patients are obese and more commonly are female than male, usually in the middle aged, so thirties, forties, fifties, in that age range. Many patients also have long-term headaches and may have vision changes like blurry vision as that pressure is transmitted across the optic nerves. And lastly, if you pick it up on a scan you've just seen, you can also look for an empty sella, which is a classic finding of elevated intracranial pressure.

Speaker 1:

These folks sometimes present with months, maybe even years of dripping from their nose, why do we treat it? What happens if we don't treat these CSF leaks?

Dr. Garret Choby:

The main rationale to treat these is for long-term prevention of meningitis in most cases. That's really, reasons, one, two, three, and four. As a secondary reason, they're coming to you with a symptom, usually it's drainage, so you are also treating that symptom for them. The primary reason is to prevent ascending meningitis.

Speaker 1:

In rhinology clinic, which is going to be a little bit different from otology, what else is on your differential diagnosis for someone who presents with a "CSF leak" from their nose?

Dr. Garret Choby:

Other things to think about, it could be a routine reason for rhinorrhea like allergic rhinitis or non-allergic rhinitis. Vasomotor rhinitis is also a fairly common thing you may see in a rhinology clinic, and that's clear drainage from the front of the nose, something to be thought about or entertained. But

again, classically the spontaneous CSF rhinorrhea is unilateral, and there's usually some fairly characteristic patient findings, like the obesity, more commonly in females in the middle age range.

Speaker 1:

So someone's in your clinic, they have unilateral rhinorrhea, maybe it's an overweight female, maybe they have vision changes, headaches. So your suspicion is pretty high. Let's say that they don't have any imaging or anything else. What's your workup for this patient?

Dr. Garret Choby:

So if I have a high suspicion, my initial step is usually testing the fluid. There's a number of ways you can test it, but for sure, the most definitive way is a beta two transferrin test. If they're leaking enough, this can be collected in a small cup and sent off as a specimen. Alternatively cotton pledget can be placed in nose and then collected for testing. Now, beta two transferrin is tested here at our institution so it's a very quick test so we can send it right down, but most institutions need to send it in via the mail. It needs to be kept cold so the protein does not break down. So there's some kits out there that help to keep it cold while sent in, which should be noted when you think about doing the testing in your office.

Speaker 1:

Do you get beta two on all your patients that you suspect CSF leaks?

Dr. Garret Choby:

I do. So in general terms want to have a positive beta two before I'll consider treating them.

Speaker 1:

What type of imaging will you obtain in these patients?

Dr. Garret Choby:

As my first step in imaging, I usually will just get a fine cut, a maxillofacial non-contrast CT scan. This is helpful because in many cases show the site of the leak, especially the sinus next to it is somewhat opacified, classic in the sphenoid. You can often just pick out that thin area of bone and pinpoint the leak, in many cases. You can also look for other things like fitting the tegmen, or scalloping the skull base, or even the empty sella you can see in many cases, which are a tip off for the elevated pressure.

Speaker 1:

What are some of the more common sites that you'll see, certainly in the operating room, but also on imaging for folks with CSF leaks?

Dr. Garret Choby:

Classically with spontaneous CSF leaks, the two most common sites are the sphenoidal, classically, the lateral recess of the sphenoid. And then either the ethmoid should be either the cribriform or the ethmoid roof, and more commonly the cribriform. There can be leaks other places like the frontal sinus and elsewhere, but for sure, the two most common are the sphenoid and then the cribriform.

Speaker 1:

Just to dig a little bit deeper, because I've been asked this more times than I care to share, from my understanding there's an anatomic structure in the lateral sphenoid recess. What is that? And is it real?

Dr. Garret Choby:

Are you referring to Sternberg's canal? The myth of Sternberg's canal? It is fairly debated I would. I'm not a big believer in Sternberg's canal, to be quite honest with you. Although, there are some anatomic studies that suggest it. I think more commonly, this is an area of thin bone, usually lateral to V2. There can be just some thin parts of the bone there that an encephalocele or CSF will sneak around, but if you get asked about it, there, there is that the mythological Sternberg's canal.

Speaker 1:

You mentioned encephalocele, how often do you see that? Or what does it look like on scan?

Dr. Garret Choby:

Typically on a CT scan and encephalocele will show just a pooching through of those dural contents with fluid inside of it, more easily to discern on MRI than a CT scan. Occasionally I, especially with the sphenoid encephalocele, it may fill the entire spheroid and it's hard to tell it's an encephalocele on a CT because the whole thing just looks so pacified. More common those are going to occur in that lateral recess of the sphenoid, but certainly could poke through in the ethmoid as well.

Speaker 1:

So you brought up MRI, is an MRI necessary for these patients? When or when don't you obtain these?

Dr. Garret Choby:

I don't think an MRI is necessary, unless they come with one, we don't usually order one. They can be helpful to discern an encephalocele. You can look for that T2 signal throughout, showing a fluid-filled SAC with the encephalocele. It may be more definitive to look for an empty sella as well, but I don't routinely get them unless I think for some reason they would need it.

Speaker 1:

Some patients who present with a CSF leak, it's a slam dunk, they are leaning over to tie their shoes. They have rhinorrhea on the CT scan. You see the dehiscence where you kind of expect it, but what do you do in patients or for patients who you suspect a CSF leak, but you can't really find on a CT scan and you don't have positive beta two?

Dr. Garret Choby:

Again, I think in my practice, we have many patients who may come thinking they have a leak. Until they show me a positive beta two, I always have a little degree of reservation, whether they're actually leaking. So I really like to get a positive beta two to know for sure. To further work this up, especially they have a positive beta two, but you can't localize it yet. One helpful adjunct can be a CT cisternogram, is where a lumbar puncture is performed with a radio nucleotide injection, if you will. Then a CT scans are obtained usually immediately then a few hours afterwards, which can help to show if there's fluid in the sinus cavities and help to localize that leak in some cases. Other options would be to go directly to the operating room and perform a lumbar puncture with fluorescein. That's an off-label use and some things that you need to talk about to the patient, like things like risk of seizure, et cetera. There are

some pretty well-defined protocols for a fluorescein, which are helpful. Then an endoscopic exam can be performed looking for the fluorescein in the nose. Ideally you want to localize that ahead of time, either with a CT scan or a CT cisternogram before you get to the operating room.

Speaker 1:

So once you've identified a patient with a CSF leak, and you have an idea of where it is, we'll want to start talking about treatment. Is there a medical therapy that can save patients from going to the operating room?

Dr. Garret Choby:

The short answer on that is not really. There was a fairly controversial paper published this year about using acetazolamide in spontaneous leak patients to try to get them to seal without operative intervention. However, there's also other published data showing that the longer you wait to fix a leak, the higher rates of meningitis there are. So in general, we treat these patients not as an emergency, but in a relatively short time period to prevent the risk of meningitis. In general, surgery is the definitive treatment for them.

Speaker 1:

When you see a patient in clinic, who you have high suspicion of CSF leak, or you've confirmed it with your beta two transferrin, the risk is meningitis. So do you immediately start treating them with antibiotics?

Dr. Garret Choby:

That's also a fairly controversial thing. In my practice and in most rhinology practices, we don't routinely treat them for meningitis. The risk of meningitis is actually fairly low in this population to be quite honest with you. There's also some protective mechanism probably of the positive intracranial pressure, pushing things into the nose, as opposed to in reverse, which may protect them. We think that it's probably not worth treating them due to risk of antibiotic complications compared to the risk of meningitis in most of these patients preoperatively.

Speaker 1:

So we've been suggesting through this time that surgery is going to be the definitive cure for this. When you have a patient with a CSF leak, what is your surgical approach? How can you fix a CSF leak? What kind of exposure do you need and what tools do you use?

Dr. Garret Choby:

So there's been a paradigm shift over the past 20 to 30 years. We treat the vast majority endoscopically in my practice and in most practices. I can't think of the last one, besides a surgically created leak, that we needed to a repair via an open approach. So, endoscopically for the vast majority. Again, I think it's really important to do your very best to pinpoint the exact site preoperatively. We'll talk a little about differences in treating both sphenoid versus cribriform leaks. There are different mechanisms to do this. Some folks will give a lumbar puncture at the beginning of the case and do fluorescein in every case. We don't do that routinely, again, unless we need help in localizing the site of the actual leak. A number of things can be discussed both in regards to, inlays putting things through the defect, as well as onlays. A number of options are out there depending on the size of the leak and what your approach may be.

Speaker 1:

So when you approach a CSF leak, what are the layers that you feel like are necessary to create a good repair?

Dr. Garret Choby:

I think for the majority of cases, the most important layer and the thing we focus on really is the intranasal layer, the mucosal layer, and that can fix the vast majority of leaks. Depending on the size and the site of the leak, we will occasionally put something through that defect as well, things like DuraGen or occasionally fat. A free mucosal graft, or a nasal septal flap, will fix the majority of cases without doing a formal inlay for these spontaneous leaks.

Speaker 1:

So you said free mucosal graft and nasal septal flap. Can you tell us what these are and how you choose when to use them?

Dr. Garret Choby:

So we'll start with free mucosa grafts are a little bit easier. You can think of it along the lines of a skin graft, if you will. These can be harvested from a number of areas. My preferred spot is the nasal floor. There's very little morbidity there. They can also be harvested from things like the inferior, middle turbinate, or even the nasal septum. This is a fairly hardy layer of mucosa along with the periosteum below it and that's important to keep its orientation. So when you place it in the defect to make sure it's placed properly so the mucosa is not facing the bone. It's also very important that in the site of your defect, you have that bone cleared 360 degrees around the defect, such that when you put in your free graft, it's really secured against the bone and not against other mucosa.

Speaker 1:

Can you describe the nasal septal flap?

Dr. Garret Choby:

Yeah. The nasal septal flap is our work horse for skull-based reconstruction, for most cases. It's a flap of the nasal septum mucosa based on the posterior cerebral artery, which is a branch of the sphenopalatine artery, as it comes across the rooster of the sphenoid below the natural os of that sphenoid sinus. We typically harvest this with a needle tip Bovie cautery and have a nice wide pedicle, and then widen that distal part on the flap. Once you reach the head of the middle turbinate, in order to diminish the risk to the olfactory, I fill in that area. It is going to rotate into a number of areas, including the intercranial base, like the cribriform or ethmoid roof or more posteriorly along, the sphenoid sinus, the sella lateral recess, et cetera. I typically will select a septal flap if there's a larger defect like for instance, a large encephalocele. If I know the patient has very elevated intercranial pressure, in revision cases, or when there's multiple defects, for instance, along the cribriform.

Speaker 1:

Are there different success rates between the free mucosal graft and the nasal septal flap?

Dr. Garret Choby:

So for spontaneous leaks, both are actually quite successful, in most cases. A greater than 90 to 95% success rate, in most cases. When you look at the surgical literature, it has been shown with high flow CSF leaks, [00:15:35] are much more effective than free mucosal grafts when you look at those large defects created by big tumor surgery. In spontaneous leaks, both are pretty effective. The main reason to think about a septal flap, in some cases, is it's much more robust and there's again with really elevated pressure, less likelihood for a repeat leak later on with more hardy vascularized tissue than a free mucosal graft, especially if someone is not perhaps going to manage their elevated intracranial pressure postoperatively, which we'll discuss in a bit, which is a very important part of their treatment paradigm.

Speaker 1:

Before going to the operating room, what are the potential complications you counsel patients on?

Dr. Garret Choby:

I talk to them about a number of things. The first thing I talk about is the potential for failure of the repair. Again, that in our experience, that failure rate is pretty low, probably 5% or so in general terms, but does occur on occasion. I also talk about the risk of meningitis, by sealing this off you potentially put the patient at risk for meningitis. So we do get some perioperative antibiotics for most of these cases. I at least a dose IV in the operating room and occasionally post op, depending on what their nose may look like. If it's a cribriform leak, I do talk to them about their risk of diminished olfaction long-term, or, at least unilaterally in some of those cases. Then lastly, I do talk to them that in some cases they may have a worsening headache, immediately postoperatively or change in their vision because as assumingly, their pressure is going back up and that transmission may cost them those symptoms, as we discussed.

Speaker 1:

Is there morbidity related to the flaps that you harvest?

Dr. Garret Choby:

There is. So with needle septal flap harvesting, there is definitely some morbidity. There is long-term cresting of the septum, which can occur for four to six weeks postoperatively. In some cases there's an increased risk of that to the cartilage in that area, things like septal perforation or healing issues in that area at the donor site. It's also important to note that, especially in those sphenoid areas, they may be at risk for injury to the median nerve, which could give them long-term dry eye or possibly V2 numbness, especially in those, in that "Sternberg's canal" region. You could get into issues with V2, numbness long-term as well. So there is some morbidity to doing these repairs.

Speaker 1:

After surgery, you've already alluded to this, but they're not out of the woods yet. What other therapies or recommendations do you make to these patients?

Dr. Garret Choby:

Again, the underlying issue in most of these patients is elevated intracranial pressure, and that needs to be addressed postoperatively. There's been some published data showing that success rates with spontaneous, or excuse me, with endoscopic repair of over 90%, when patients do not manage their elevated pressure, post-operatively that success rate goes down to about 80%. So it is important to

manage that longterm. A number of options are available. We typically in our practice, will get a lumbar puncture about a month after surgery to check that pressure. If you check it when they have ongoing leak, it's not a very accurate measurement because they have a pop-off valve, if you will, where their pressure is probably artificially low. So we check about a month afterwards, and if it's elevated, we may give them a acetazolamide therapy, which is a diuretic therapy to lower pressure. If it's a repeat leak or they're really elevated, a shunt maybe entertained as well. We also counsel all patients that weight loss is very important. It's been shown that about a 10% weight loss reduction may have a significant improvement in their elevated pressure. We often refer them to our nutrition clinic or our weight loss clinics as well.

Speaker 1:

You've already kind of touched on this, but the outcomes seem quite successful over 90%. So what's your followup with these patients afterwards?

Dr. Garret Choby:

So I do tend to see these patients long-term for at least a year or so after surgery. That's to make sure that they've healed well, there's no donor site morbidity as well as to be quite honest, make sure they follow through with the recommendations for managing their elevated intracranial pressure. I also have to make sure to get plugged in with ophthalmology to get a nice eye exam because sometimes they can have issues with their optic nerve, often a long-term elevated pressure. It's also important to make sure they see neurology long-term for help in managing this, whether it's, again, the acetazolamide or with our neurosurgery partners treating with a shunt, et cetera.

Speaker 1:

I feel like we've covered a lot today. Is there anything else you'd want to add?

Dr. Garret Choby:

The last thing I'll mention is that in my practice and in many others, I treat these with my neurosurgical partner. We do a lot of skull-based surgery together, but I also think it's important for co-managing these patients as there can be a lot of morbidity to these procedures, as well as again, assistance with their long-term elevated intracranial pressure and potentially shunt management as well. It is nice to do this as a tag team from both services. So we both have a lot of good input for these patients.

Speaker 1:

Sure. So just to summarize what we've talked about, anterior skull-based spontaneous CSF leaks often present with unilateral rhinorrhea, which is worsened with things like leaning forward or producing a Valsalva maneuver. This is often seen in a patient population with idiopathic intracranial hypertension and is caused by chronic pressure at an already weak point, which can be either at the lateral sphenoid sinus or at the cribriform. Work up includes analysis of fluid via beta two transferrin, a CT scan to identify where the leak might be, and if required more scans like a cisternogram. Surgical correction is basically the definitive therapy and is offered, yes to improve quality of life, but more importantly to prevent meningitis. Surgery includes visualization of the area endoscopically and could be sealed with a free mucosal graft or a pedicle flap, depending on the situation. Postoperatively it's important to manage these high pressures with either medication like Diamox, possibly a drain, and also diet and weight loss. Anything else?



Dr. Garret Choby:

I think that's great. Thank you.

Speaker 1:

Thank you.

Dr. Garret Choby:

This episode is coming to a close, but I did want to end with a few questions. I'll ask a question, wait a few seconds for you to either pause and give the answer a thought and then give you the answer. So the first question today is what are the two most common locations for anterior skull-based spontaneous CSF leaks?

The two most common locations for anterior skull-based spontaneous CSF leaks are the lateral sphenoid, this can sometimes be correlated with Sternberg's canal, and the cribriform plate.

Second question. What are the two types of repairs that can be offered for anterior skull-based spontaneous CSF leaks?

The two most common types of repairs for spontaneous CSF leaks is either a free mucosal graft, which can be obtained from several sites, including the floor of the nose, and also the nasal septal flap, which is a vascularized pedicle flap off the posterior septal artery.

My final question for today is after you operate on patients to surgically, correct spontaneous anterior skull-based CSF leaks, what postop recommendations do you give patients?

After successful surgical correction of the CSF leaks patients should be educated to change their diet and try to lose weight. And furthermore, they can be treated with Diamox or acetazolamide that's all for today. Thanks so much for joining and we'll see you next time.