

Dr. Jason Barnes:

Hey, there, welcome to ENT in a Nutshell, my name is Jason Barnes. And today, we are joined by Dr. Neil Patel and we will be discussing superior semicircular canal dehiscence. Dr. Patel, thanks for being here.

Dr. Neil Patel:

Thanks for having me.

Dr. Jason Barnes:

When we talk about superior semicircular canal dehiscence, what might you expect from a patient who presents with this syndrome?

Dr. Neil Patel:

Patients with superior semicircular canal dehiscence syndrome often come in with a wide spectrum of complaints. In our experience, typically they are audiologic more than vestibular. Autophony, or hearing one's voice loudly in an ear, is probably the most common. Somatasounds, or hearing your eyeballs move, or your heartbeat loudly in an ear, or hearing your feet hit the floor when you walk, is probably the second most common. Pulsatile tinnitus is also quite common.

From a vestibular standpoint, vertigo with loud sounds or pressure are the classic symptoms associated with superior semicircular canal dehiscence syndrome. However, chronic imbalance, brain fog, mood and learning deficits, are also commonly endorsed by these patients, but we don't have a good way of assigning these specifically to the dehiscence.

Dr. Jason Barnes:

When you see these folks in clinic, you perform a physical exam. Do you see anything that tips you off that this might be what's going on?

Dr. Neil Patel:

In general on physical examination, you don't usually see much on ear exam. Otoscopy should be more or less normal. These patients typically don't have a history of chronic otitis media or prior ear surgery. A few maneuvers can be performed in the clinic to sort out what's going on.

One of those uses a tuning fork placed on the lateral malleolus, not the malleus, to determine whether they have super threshold bone conduction. Sometimes patients will be able to hear that sound from the tuning fork, typically a 128 or 256 Hertz tuning fork, in the effected ear.

The other maneuver you can try is with pneumatic otoscopy, movements of the drum can sometimes trigger vertigo. This is akin to Hennebert sign cited symptoms, such as objects moving on a clock face when patients would whistle or hum, or oscillopsia, or the room bouncing up and down, induced by loud noises, or vertigo with straining while digging up a root in one's garden. These are obviously strange symptoms and we didn't have a great unifying diagnosis for this.

Dr. Jason Barnes:

We've been calling this superior semicircular canal dehiscence, but can you speak directly to what the pathophysiology is?

Dr. Neil Patel:

I think that's a great point in terms of distinguishing a radiologic finding of superior semicircular canal dehiscence and superior semicircular canal dehiscence syndrome, which is the collection of symptoms that goes along with this finding.

Dr. Jason Barnes:

You mentioned that people generally have conductive hearing loss, but this is an inner ear disease because it's affecting the superior semicircular canal. How does that work?

Dr. Neil Patel:

The third window phenomenon is confusing, so I think it's worth spending a little time on this. Basically there are typically two windows to the inner ear, the oval and the round window. Sound pressure comes in through the oval window, through the stapes and is decompressed or dissipated at the round window. When there's a third window present, some of the sound energy is dissipated through that third window. That produces the conductive hearing loss we see in these patients.

Secondly, super threshold bone conduction, or better than normal bone conduction, is by sound directly entering the inner ear through that third window, or that's at least what we think.

Dr. Jason Barnes:

When someone comes into your clinic, they have a possible conductive hearing loss. Some of these somatasounds, autophagy, the stuff you've been suggesting, superior semicircular canal dehiscence is on your mind, but what else is on your differential diagnosis when you're talking about this?

Dr. Neil Patel:

Really any cause of conductive hearing loss can come up in the setting of a normal ear exam. It's typically otosclerosis is the primary differential diagnosis for the hearing loss. Meniere's disease comes up really in any discussion of patients with vertigo, but the vertigo experienced by superior semicircular canal dehiscence patients is quite different from in Meniere's. It's typically not spontaneous and it's usually evoked by a loud sound or pressure.

Really in lots of otology neurotology differential diagnoses syphilis and spontaneous perilymphatic fistulas come up, but these are really rare. Something worth mentioning that is quite a common diagnosis and increasing in incidents, we believe, is migraine associated vertigo or vestibular migraine.

Finally, one thing to sort out when it comes to autophagy is patchouli eustachian tube dysfunction. These patients will have similar auditory findings, but usually not much in the way of vertigo and their CT scan will show normal findings of the superior canal.

Dr. Jason Barnes:

You suspect superior semicircular canal dehiscence, you've performed your physical exam, you've seen folks in clinic. What are your next steps in terms of workup? We've talked a little bit about the audiogram, but can you speak a little bit more to that?

Dr. Neil Patel:

That's right, the audiogram will show typically a super threshold bone conduction in some conductive hearing loss and that super threshold bone conduction is from sound pressure or sound entering the

inner ear through this third window. The conductive hearing loss is from sound pressure being dissipated through that third window, as opposed to being confined to the inner ear by the oval and round windows.

If the audiogram is suggestive of this, then you'll look at the stapedial reflexes. In somebody with conductive hearing loss, in whom you want to rule out otosclerosis, you would expect normal stapedial reflexes in a patient with superior semicircular canal dehiscence, and the syndrome. If somebody has absent reflexes, you would be more suspicious of otosclerosis or other causes of conductive hearing loss.

Dr. Jason Barnes:

The audiogram is suggestive of this, and I assume you obtain a CT scan.

Dr. Neil Patel:

That's right. A lot of this is patient driven. If the symptoms are bothersome enough and they want to know what's going on, we obtain a plain non-contrast temporal bone CT scan with views perpendicular to and parallel to the superior semicircular canal. Those views are typically referred to as the Stenver and Poschl projections. The Stenver being perpendicular to the superior canal and Poschl being parallel to the superior canal.

These specific views at sub millimeter resolution are usually enough to determine whether the superior canal is completely covered in bone, the location of the dehiscence and the size of the dehiscence.

Dr. Jason Barnes:

Where does vestibular testing fit in this?

Dr. Neil Patel:

If a patient has the typical signs or symptoms, an audiogram that fits the diagnosis, and a CT scan, vestibular testing is very helpful in determining whether the dehiscence is functional. If somebody has bilateral superior semicircular canal dehiscence on imaging, whether only one of the sides is functional and corresponding to their symptoms. Some of these symptoms are difficult to localize for patients, so I think that it provides helpful your specific functional information.

The vestibular testing typically involves VEMP testing, or vestibular evoked myogenic potentials. There are cervical VEMPs and ocular VEMPs, two different ways of assessing function. Cervical VEMPs, test the saccule and ocular VEMPs test the utricle, which also shares innervation by the superior vestibular nerve, the same as the superior semicircular canal.

On ocular VEMPs we would look for increased amplitudes and on cervical VEMPs we looked for decreased thresholds. Different institutions use different criteria, but based on some recent studies, a combination of the two provides probably the highest sensitivity and specificity for the syndrome.

Dr. Jason Barnes:

If a patient lacks objective vestibular testing, does that preclude them from having superior semicircular canal dehiscence?

Dr. Neil Patel:

It may preclude them from having superior semicircular canal dehiscence syndrome based on the overall objective picture. I don't believe that vestibular testing that supports the diagnosis is mandatory prior to surgery, but I do think that it helps to counsel patients that their symptoms may or may not be related to the dehiscence if their vestibular testing is normal.

Dr. Jason Barnes:

Sometimes we talk about official diagnostic criteria. From what I gather from what you're saying, there isn't a specific diagnostic criteria per se, but rather you take in all of this information, both subjective and objective, to decide whether or not this patient has superior semicircular canal dehiscence, that can be treated.

Dr. Neil Patel:

That's absolutely right. I think it's a combination of history, physical exam, ruling out other causes, CT findings, and vestibular testing, particularly if you're going to operate.

Dr. Jason Barnes:

You've done all of this workup and the patient wants treatment. How do you approach treatment in this patient population?

Dr. Neil Patel:

Patients often have many other auditory or vestibular complaints that may or may not be attributed to superior semicircular canal dehiscence. For example, some patients will endorse aural fullness, chronic imbalance that doesn't go along with pressure or sound induced vertigo, mood and learning deficits, brain fog, and a litany of other complaints.

I think the important point here is patient selection. Patients who have lots of audio vestibular complaints, and really can't pin down the typical signs or symptoms of superior semicircular canal dehiscence syndrome, are probably poor surgical candidates, as you're not likely to improve chronic imbalance, aural fullness, non pulsatile tinnitus, and symptoms such as those. However, if a patient has the classic symptoms and understands that intervention is only going to address those symptoms, then I think that could be a reasonable patient for surgery.

Dr. Jason Barnes:

In considering treatment, are there any medical options for therapy?

Dr. Neil Patel:

There aren't. There are some studies that posit whether osteoporosis or low bone density could have something to do with thinning of bone over the superior semicircular canal. This comes up in terms of CSF leaks in the temporal bone and encephalocele as well. But overall, there's nothing that we recommend medically to prevent somebody who has a radiographic dehiscence from developing findings of the syndrome or to prevent the problem altogether.

Dr. Jason Barnes:

What happens if you choose not to treat this?

Dr. Neil Patel:

Observation is probably our most popular treatment approach for patients with superior semicircular canal dehiscence. It is safe to observe these patients. We've determined that hearing does not decline at a more rapid pace than the normal ear, or then the general population.

Patients may still have the conductive hearing loss, which is certainly aidable, as most of these patients have normal or near normal inner ear function. It's very safe to observe these patients. Many of them will come in with a CT scan showing superior semicircular canal dehiscence and really only few complaints or only auditory complaints. When the risks and benefits of surgical intervention are discussed, often patients will choose to observe until they develop more severe symptoms or particularly vertigo.

Dr. Jason Barnes:

Now you discussed surgical intervention, which is what we've been getting to. Once you've selected the correct surgical candidate, what are surgical options?

Dr. Neil Patel:

Much of it is based on imaging. For unilateral superior semicircular canal dehiscence, you want to look at the location of the dehiscence. Typically these are in the middle cranial fossa, but sometimes they can be associated with a dural venous sinus called the superior petrosal sinus that runs along the petrous ridge. If there's a dehiscence adjacent to the superior petrosal sinus, it would be difficult to treat through a middle cranial fossa approach.

If somebody has bilateral superior semicircular canal dehiscence, you need to consider the function of the superior canal. Essentially, if you operate on both sides in sequence and eliminate the function of the superior canal by plugging it, for example, patients can have disabling oscillopsia or when they're going up and down stairs, the room is bouncing up and down. There isn't a very good treatment for that. We consider location of the dehiscence, unilateral or bilateral, and then the two main surgical approaches, which are the middle cranial fossa approach and a trans mastoid approach.

Trans mastoid approach is great because it's not a craniotomy and accesses the superior canal through a very familiar surgical corridor, the mastoid, but it provides the poorest visualization of the actual dehiscence.

The middle cranial fossa approach, on the other hand, does involve a small temporal craniotomy and elevation of the temporal lobe, but it's extradural, quite safe, and provides a direct view of the dehiscence.

The other question that comes up is resurfacing or plugging. Plugging the canal involves theoretically compressing the membranous labyrinth within the bony labyrinth and often yields lack of function of the superior canal. Resurfacing, on the other hand, simply shields that canal from the overlying dura and pulsations of the dura, and can be very effective as well, but is a little more indirect as whatever you use to resurface that area can potentially move, and if you're using a trans mastoid approach, you don't see the dehiscence directly.

Different surgeons have different preferences regarding approaches, but I would say that the majority of cases that we do our middle cranial fossa.

Dr. Jason Barnes:

When you're counseling patients about surgical intervention, what do you tell them they should expect in terms of improvement in symptoms and outcomes?

Dr. Neil Patel:

I think one thing to think about is what their symptoms were before surgery and which we could attribute directly to the dehiscence. We expect that autophagy improves, pulsatile tinnitus improves, somatasounds improves, and pressure or sound induced vertigo would improve.

Brain fog is quite variable. Aural fullness is quite variable. Chronic imbalance usually does not change. Headaches usually do not change either. The recovery from the surgery is challenging for some patients who have a lot of vertigo or chronic imbalance complaints going in, but I would say that most patients do quite well.

Dr. Jason Barnes:

Do you follow up with these patients long-term?

Dr. Neil Patel:

Most of the followup is based on whether their symptoms resolved or not. Some centers will obtain vestibular testing at a 6-12 week time point to confirm that the dehiscence was treated. At our institution, we reserve that testing for patients who have persistent symptoms in spite of surgical intervention.

Checking a hearing test would be reasonable to see what happens to the conductive hearing loss. In a lot of patients it improves. Otherwise, no imaging is required in follow up.

Dr. Jason Barnes:

when you consider surgery, what are some possible complications involved with these interventions?

Dr. Neil Patel:

When you're operating on superior semicircular canal dehiscence, you're operating on a labyrinthine fistula, so the inner ear is placed at some risk. I would say the risk of sensorineural hearing loss is typically quoted at about 5%, but is fortunately quite low. This occurs in either resurfacing or plugging procedures through middle cranial fossa or trans mastoid approaches.

Patients can sometimes develop benign paroxysmal positional vertigo after these procedures as well, as if their vertigo picture wasn't confusing enough, this could be added to the mix.

Almost all patients have some transient disequilibrium. I wouldn't consider that a complication necessarily, but it's certainly something you should counsel patients about following repair.

The facial nerve is also at some risk during middle cranial fossa procedures. The facial nerve can be uncovered at the geniculate ganglion or the greater superficial petrosal nerve and be at some risk during elevation of the temporal dura. In trans mastoid procedures the facial nerve risk is somewhat lower, but in general, we estimate it at 1% overall.

Dr. Jason Barnes:

Well, Dr. Patel, I think we've discussed a complex topic pretty comprehensively. I want to thank you for being here and did just want to summarize once more what we've discussed.

Superior semicircular canal dehiscence often presents with a mixture of symptoms, including hearing loss, autophagy, pulsatile tinnitus, somatasounds, and possible dizziness. When this is suspected, an audiogram should be obtained, which will demonstrate a conductive hearing loss and possible super threshold bone conduction.

A CT scan after obtained will demonstrate dehiscence of the superior semicircular canal and vestibular testing will demonstrate increased oVEMP amplitudes and decreased cVEMP thresholds.

Surgery involves either a trans mastoid or middle fossa approach to plug or resurface the superior semicircular canal and patients undergoing this procedure will likely experience improvement in their vertiginous symptoms, pulsatile tinnitus, autophagy, and somatasounds. is there anything you'd like to add?

Dr. Neil Patel:

I think that's everything in a nutshell.

Dr. Jason Barnes:

As we bring this episode to a close, I did want to end with some final questions. I'll ask a question, wait a few seconds for you to press pause or ponder the answer and then give the answer.

The first question is, what are the typical audiologic findings in superior semicircular canal dehiscence? Patients with superior semicircular canal dehiscence will often have conductive hearing loss with super threshold bone conduction due to the third window phenomenon. These patients will have normal stapedial reflexes.

Our second question is, what is Hennebert sign? Hennebert sign is vertigo induced by pneumatic otoscopy.

Our third question is, what will VEMP testing show in superior semicircular canal dehiscence? In these patients, we will check both oVEMPs and cVEMPs. oVEMPs will have increased amplitude and cVEMPs will have decreased thresholds.

Finally, our last question, what are symptoms that are not likely to be improved by surgery in these patients? When we consider surgery in these folks, we do want to counsel them that they will likely not experience improvement in several symptoms. These include headache, chronic imbalance, and brain fog, to name a few.

Thanks so much for joining us and we'll see you next time.