

Dr. Jill Rau
Bridging the Gap conference
November 11, 2023

Navigating Spinal CSF Leak Care When You Are Not at a Major Referral Center

Hi, everybody. Thank you for inviting me. I'm really honored to be here. This is a super unique conference and I think it's amazing. So, without further ado, I work at an institute where imaging is possibly goes out to multiple different places. We don't have any particular dedicated CSF interested groups.

So that's kind of what this is about. I don't have any financial disclosures relevant to CSF leak. And I am a member of the advisory board for the foundation. So this is where I work. It's a neuroscience institute. We have sub-specialties of neurology. I'm the only headache specialist. We have neuroradiology, but they're not no longer in-house.

So we can't speak to them and we don't have any imaging facility. They're just the radiologist and I think an x ray machine. We have physical medicine and rehab spine. And we have some neurosurgeons who are actually contracted as their own consortium that work in here, but they don't have the same insurance as us,

so all of our patients that might need neurosurgery may not be able to see the spine surgeons that we actually can speak to readily about this. So what do I do when I, when do I consider intracranial hypotension? How do I consider, how much do I consider it? Like how deep do I get into this and think, Oh, this must be it,

I'm going to go all the way. And when do I recommend or refer my patient to see a specialty center. That's a big question, because from where we are to get to that, it's a big cost. It's a big emotional jump. And if they don't have a spinal CSF leak and are going down that trail, it's a lot of risks financially and emotionally.

As well as some of the physical risks to think or to consider having a leak and commit to that with, if your, if your suspicions are incorrect, and that I think is especially large when you have to go out of state to get that care. So, when do I consider it? Pretty much all the time it's in the back of my head.

When a patient has orthostatic headache or other symptoms that comes, obviously, quite, quite to the forefront. Obviously, when there's findings on the brain or spine imaging that are consistent with that. Occasionally, I will not be thinking of it and I'll open up and look at the brain and I read all my images myself.

I mean, I read the other people's opinions as well, but I also review them myself. Or if they have a low opening pressure, if they had a leak for some—I mean, had a LP for some reason, and then you know, you get a report, oh, the radiologist, I mean,

the, the person who did my LP said, you know, they had to pull back to get any imaging, I mean, to get any fluid out.

I consider it when the patient asks. And you know, sometimes we're going along and they're like, what about spinal CSF leak? If I hadn't been thinking about it, then I think, you know, they know something that I haven't thought about. There's something that I haven't extracted in this history that's making them think about it.

Let's reconsider this. Cough and valsalva headaches, always consider it. New daily persistent headaches. They're, they're a bucket of lots of different kinds of diagnosis, so it should always be considered there. A headache that never got better or changed or got worse after any kind of spinal procedure, I always revisit that as a possibility.

Refractory headaches: they've been ticking along, ticking along, nothing is working. Could there be something else going on? Could that thing be mechanical? Could it be a spinal CSF leak? Same thing with intractable headaches that never go away. If they have progressive neurologic symptoms that are more consistent, but not always, but the things that start to come out and they keep going and I have no other indication of what other underlying condition it might be, it'll pop back up in my head:

could this be consistent with a spinal CSF leak? And then all these weird constellation of symptoms, you know, not everybody presents with your typical orthostatic headache. And so when people are having symptoms that just don't mesh with everything else, and I, I come back and I think, is there something else that I should be considering like an inflammatory disease, or should I be thinking about something more along the lines of spinal CSF leak?

And then the question is like, what are the other things I think about? Like, how much do I think of it as likely to be spinal CSF leak versus how much do I think of this as being some other condition. Dr. Friedman talked about all these other things or a lot of these other things that may be also presenting similarly.

So obviously, migraine and cough headaches. POTS, Chiari, cervicogenic headaches, that's a big one, craniocervical instability, and I'm sure that tons of patients have both CSF leak and craniocervical instability because of that link with the connective tissue disorders. But it's possible to have one and not the other, and so when do we consider that, and what do we do about it?

That's a huge question. I've seen tethered cord present like a CSF leak. Inflammatory disorders tend to have another inflammatory component, something else that hints towards inflammatory issues. But nonetheless, that's what could take me down one road versus the other. TMJ will sometimes have a lot of neck pain that can be orthostatic as well.

Sinus congestion can be orthostatic. Idiopathic intracranial hypertension can sometimes be confusing and patients can have had intracranial hypertension and blow out a leak and then it becomes a very confusing picture. And so when do I think, oh, could there still be a leak even though they have you know, previously had intracranial hypertension. MCAS is its own rabbit hole, but certainly something we see comorbid with intracranial hypotension, but also something that can confuse the picture or stand on its own. And then Eagles syndrome, we often see. It tends to present more like idiopathic intracranial hypertension, but I never want to rule out these things that can cause pressure changes.

So if I see things like this do, that is, that help me go one direction towards intracranial hypotension, um, versus one of the, the diagnosis on its own or in comorbid with something else. And what do I order? Like, what do I do? Because I can't just say all the time, Hey, let's just send it for, you know, my neuroradiologist specialists to look at, you know, if I consider it, I, I have to take a little step back and say, wait, let's see what I can do first to, to help me decide whether or not I'm going to send it to someplace special or not.

Obviously we get a brain MRI, preferably with contrast for the same reasons earlier specified, and I'll get a full spine. But my radiology will go to different locations depending, because I'm in a community hospital. So I will put comments on that I'm asking about CSF leak, please protocol it this particular way.

I've tried different ways to do that with very specific protocols versus more generalized statements. I have one friendly neuroradiologist. I don't mean they're not friendly people. I mean that there's one that's interested in spinal CSF leak, and I will say if it goes to that institution, or I just put it on all of them in case it goes to that institution, "please have Dr. So-and-so protocol this." That being said, even though she's, uh, cognizant of spinal CSF leak, she's not an interventionist. She used to be, but the way they contract the spinal in the neuroradiologists is such that none of them are interventionalists for a very long, complicated reason.

I'll do autonomic testing. Obviously Deb talked about POTS and, and spinal CSF leak and, and how they're related. So I will look for POTS. I'll try and treat the POTS. Sometimes I'll do flex/ex imaging to see if there's any indication of a change there. That may be something we can treat before we have to send to a specialty area.

I'll get MRVs. I'll do inflammatory testing, triptase. That'd be for the MCAS, if there's a hint that maybe there's that component. Um, I try not to do lumbar punctures in patients that I'm highly suspicious of CSF leak, but sometimes there's a reason to get them. And then, I know it's going to sort of make everybody cringy who's in radiology, but sometimes I will order a cisternogram.

If I'm really on a fence, I've tried everything else and it's hard for my patient to get anywhere else, to see if it'll help me really, if you see it there, then I would do more effort to convince my patient.

So we just talked a little bit about, you know, what's happening in the imaging aspect at a non-specialist center. You know, imaging is currently the gold standard for diagnosis. Clearly, if you see a leak or if there's lots of signs on the brain imaging, you can say, okay, I think there's, you know, a leak. It's pretty definitive here. But if you don't have that, you know, imaging can actually take you the other way where people are like, no, there's no sign of it.

So it can't possibly be, you know, a leak and I'm sure you're all familiar with that. Plus we get imaging that's kind of not helpful in many instances because it's not protocolled in such a way. And so then you've spent all this time and effort, not from my perspective, but from patient's perspective, gotten this, insurance has paid for it and then they won't pay for the proper imaging until we get them to a specialty center.

So these are the types of imaging we order, except for I never order CT myelogram at my center because it won't be protocolled in such a way that it will be beneficial for the patient. The CT myelos are done very specifically so that the fluid is on board when the patient is on the table. In an institute that you're not a specialist, it'll be done, they'll sit there for a little bit, walk across the hall. It's, it's a, a puncture into the, of the dura that is not going to be a high yield at an institute that doesn't usually have spinal CSF leak protocoling for that. So we discussed the Bern score earlier today. And so those signs are there, but there were lots of signs looked at for the Bern score that didn't make the algorithm.

And the reason for that is statistical and practical in many instances, but there are other signs that you can look for that may help lean you one direction or another. So for instance, you know, tonsillar descent isn't on the Bern score again for a variety of reasons. But if I have a patient that I'm suspicious of that for, and they, their Bern score might be, you know, moderate, but then they also have that, or some of the other signs that might be out there, I might be more inclined to say, Oh, this really is starting to push me towards

sending this patient to a specialty center. So, we may have, there's diffuse calvarial hyperostosis. It's not a common thing to see, but if you see it, it's something I'm more inclined to say, Oh, look, that's something that is seen more frequently in spinal CSF leak. The optic nerve is something that can be telling. If it's a very straight optic nerve with a very thin sheath that might be more telling or more additional information to help lean me to or away from — probably not away from just more to—sending a patient to a specialty center.

There are changes that you can look at on optic nerve. I am pushing hard at my institute to get the ability to do this myself, both from a research perspective, but then to be able to use that as another hint. So patients with POTS do not see a change in the optic nerve sheath diameter with orthostatic changes, but patients with CSF leak do. In this particular study, anyway.

And then there's some spine imaging that sometimes can be helpful. If you see this ventral spinal cord displacement, then I would again, maybe push me more directional towards the specialty center. Some venous engorgement is often seen. And then, like I said, I will sometimes, not frequently, but sometimes order radioisotope cisternography.

I can think of in one particular patient that I have, he has very, very orthostatic headaches. He's also been diagnosed with POTS based on autonomic testing. Treatment for POTS has not relieved his symptoms even an iota. But he's in debt from all the testing, even the autonomic testing, and he doesn't want to do any more testing unless I'm sure this is going to fix him.

And so we were able to get radioisotope cisternography covered for him, and it was negative. There was no sign, none of the subtle signs on that, and so, you know, so for him, I'm not sending him because financially he doesn't want to make that change unless there's more evidence because he's done so much already.

So if I'm on the fence, what I will do is I'll first treat as many of the underlying conditions, comorbidities, and suspicions that I have to see if we can make any impact. So, you know, migraine is present in 15 percent of the population. So that means a lot of people with spinal CSF leak also have migraine and, and the irritation of the nerves from the CSF leak are probably going to provoke migraines.

We'll try and optimize migraine treatment and see much, how much improvement we can get. If the patient also has POTS, we'll try and optimize the treatment for POTS. I'll send to PT for neck, I'll do nerve blocks. Sometimes these are relieving even in patients with CSF leak just to help get them through. Or trigger point injections, sometimes sent to spine intervention to see if those make any difference because cervicogenic headaches can often present similarly. I'll refer to subspecialists within different fields that they have comorbidities in. And then sometimes I phone a friend. So I have neuroradiologists that I will call and say, can you look at this imaging or talk to about, you know, the presentations and say,

you know, what do you think? Do we think it's worth doing this investigation? The answer is always yes, it's worth it. But it is a big trial, as I'm sure many of you know, to go to these subspecialty centers, and sometimes the subspecialty centers, depending on where you go or just that particular patient, it may be a cursory look and said, no, it doesn't look like a leak to me.

And that can be very disheartening for a patient to think, and I'm sure you guys know this, to think that they have a leak and then be dismissed by a another institute. I have one patient who has low CSF pressure, she has all the symptoms. She went to a big institute, thought that was going to be her, the end of her, her journey that she was going to get fixed.

They told her they didn't see any signs of it. Didn't offer any treatment. She came home and tried to kill herself. We have subsequently sent her someplace else and she's feeling much better. So, you know, it can, it can not always be the best response then. And I think one of the important things is prepping patients to what can happen.

When do I, when do I refer my patients to these centers? Well, like I said, it's a, it's a conversation. It's a very considered conversation with the patient and often the supporting family members, and also setting up expectations for what might happen, what might not happen, what can happen if they don't get the treatment they're expecting, or if the treatment doesn't work.

I also have some local academic institutes that have various people who are sometimes helpful. And so sometimes in cases where it's hard for the patients to travel or we want to try some things locally first, I do have places that I can sometimes get a little bit of help. And then obviously the out-of-state referrals to the big centers like here, Cedars-Sinai, Duke, Mayo, Rochester, Stanford, Jefferson. I'm sure there are others, but these are kind of the ones highest on my list. And if you guys have other ones, please tell me because we're always looking for them. I try and have a conversation with the patient. They will often self-refer to multiple institutes.

And there's a resource issue. I'm not opposed to it, but you know, I try and say, look, well, let's try one first. And then if, you know, come back, see what the results were, have another conversation. Is it worthwhile to go to a second? But I think we need to be cognizant of resources and, you know, having multiple input from different physicians at the same time, that can be confusing for the patient.

So locally, what can I do? Obviously the conservative treatment, which nobody likes to hear about. I use it sometimes. You can also try the, you know, reverse Trendelenburg if that's helped diagnostically. We can do non-targeted epidural blood patches, but at my institute, I've sent someone who definitely had a CSF leak,

I thought, okay, well, clearly it's there. Let's get a non-targeted patch. It was a chronic leak. They gave him five cc's at L2 and I thought, you know. Might as well lick the patient, send him home. I've had sent for blood patches and had neuroradiologists sorry, interventionists, not neuroradiologists say, well, they didn't have an LP recently.

Can that really happen? I was like, yes, yes, it can. Here's some literature. And that, that often actually is well received. People then become very interested. Wow, I didn't know. So that's kind of cool when you get that response. And we have potential for other interventional and surgical treatments but usually not with people who are well versed in the area.

So I tend to be very conservative about using those resources. So I will utilize sometimes non-directed blood patches, if I've been able to have a conversation with the person doing it beforehand. I'm trying to make connections, find the people who know what's going on best, and try and refer on those specific days or to those specific providers.

I'm trying to educate those people, different groups, the interventionists who do the patches, the neuroradiologists who read the reports, but there are multiple institutes that these get sent to, so it's difficult to do. I'm trying to build the system and build a program, but it's... In a program where that's not compensated, people have to donate their own time and they weren't previously interested in that.

It's quite hard to do. You can use the local Maverick if you have one. So I have a guy who's a private practice, owns his own; interventional radiology, he's been trained in neurorads and in pain. He'll do whatever I ask, but. He's not well versed in the field, um, but he's very, very good at what he does.

He's got good hands, as we would say. So, you know, there's always that toss up: when do you use that facility and when do you not? I've been using it more and more just because there are so many patients who really want to give it a try, and it is so challenging. Long waits, plus, you know, financial burden, emotional burden, physical burden of getting to a more specialty center.

So I always want to discuss before I send patients to a specialty center, the expectations, recommendations, and adverse reactions to treatment. In particular, sometimes people will get patches, but they are not told afterwards that they might get rebound intracranial hypertension. So they come back.

My wait list is in six months, so I'll get these messages coming through, you know, my headaches not change or is worse or it's different and they don't get any advice about bending, lifting, and twisting. So I try and tell my patients beforehand what my recommendations are. I say, don't bend, lift, and twist for two months.

Everybody's got a different thing. There's no right answer to that. And I try and talk about rebound intracranial hypertension, and I give them a script for acetazolamide, give them instructions. And if they have questions about whether they should take it or not based on my instructions later, they can contact me.

And I'm often giving out my cell phone because going through the medical records, it's hard to get in touch. I have patients that have been to multiple different centers and then end up coming, getting referred to me, or come to me, or were, had already seen me, and they're still having problems, or actually sometimes referred to me from one of those centers saying, okay, well, nothing's helped now.

Can you help their headache? And what do I do then? Right? They've usually come to me with a list of interventions and medications they've tried. Well, sometimes we

just start over. We redo a whole interview. We redo a whole history. Is there something else we're missing? Also, could you have had patched or surgical treatment and improved, but still have, you know, chronic sensitization or underlying chronic migraine or something, and maybe one of the meds that you tried before didn't work because you had a leak.

Maybe it'll work now. So it's kind of a whole new process for those patients to start over. And sometimes we end up sending them back because maybe they just couldn't find the leak then and they'll find it next time. So I feel like my role is to, you know, see the patients. Consider CSF leak, educate people, educate people near, near me, learn.

I want to learn from you guys, like, the patients. I'm learning today, already learned something new from patients. So, I, it's a process we're all learning. And I appreciate the opportunity that everybody has here and that, we're getting more and more information. The amount of research in this area has gone exponentially up.

We're building lots of new tools and resources and learning more. So I think in the future we'll have better answers, but it's still a maze to be navigated at the moment.