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Good morning, everyone. It's an honor to be, for me to be here with you today. I'm gonna talk about clinical challenges in the diagnosis of spinal CSF leak. And Dr. Friedman will go into its differential diagnosis.

Every patient with headache complaints deserves a total history and the clinical examination with the goal of trying to identify red flags for a potential underlying condition. Most of the time we find none, we end up diagnosing a primary headache condition. But five or ten percent of the time, we do see a secondary headache such as the one that we are talking about today. A SNOOP mnemonic is something that is commonly used to look at those red flags. We're not going to go into details of that today, but I do want to spend a minute on some P red flags.

Somebody can have a pre-existing headache condition, but now something has changed or we are dealing with a new headache altogether. We're going to hear a lot today about positional headache, meaning the headache it's worse or only comes with upright or laying down position.

If the headache is precipitated by sneezing, coughing, or exercise, this is a red flag, as is a progressive headache in a typical presentation. Post-traumatic onset of headache is a red flag for us, as is somebody that takes too many pain medications. According to the current international classifications for headache disorders, headache attributed to low CSF pressure can be of any kind, but you do have to have low CSF pressure, which is less than 60 millimeters of CSF, and you have to have, or you have to have evidence of CSF leak on imaging, or both.

The headache has to develop following the CSF leak or can lead to its discovery. A variant of that is the headache attributed to spontaneous intracranial hypotension, or SIH, as I was, I will call it interchangeably with spinal CSF leak today. So that, that condition, it's everything on the left column, except you don't have a history of trauma or procedures like injections to the spine.

Keep in mind that this kind of history can sometimes be forgotten by the patient. I took this information table from a large meta analysis was published 2021 of 144 studies, which shows that SIH age is about 42 or early 40s, similar with, with migraine, by the way. It's predominantly in women.

The time to diagnosis can be as long as 20 years, with an average of 32 days. The headache is present in almost all cases. Low spinal fluid pressure actually was found in no more than about two thirds of patients. And multiple other studies forward have actually—one of them is cited today—showed much lower percentages, and multiple or many headache specialists, including us or CSF leak experts, won't recommend this spinal tap to diagnose the CSF leak because it's a test with low sensitivity and can potentially be harmful.

The incidence of SIH or spinal CSF leak. It's seemingly low, but we may be finding out that might be higher if we are doing better at diagnosing it. I put this slide very quickly just to show three recent publications very different in methodology on CSF leak diagnosis and treatment. One, it's a large meta analysis.

The other one, it's an expert consensus on 22 topics of interest. And the other one, the newest one, it's a pioneering qualitative study on patients with confirmed CSF leak by CSF leak experts. These are a few aspects that I think might be relevant that can help or hinder the diagnosis of SIH. The most important one, probably it's orthostatic headache, which means the headache is there or gets worse as, as you get up, gets better when you lie down.

There are multiple nuances of this, which I'm going to go over. Then we'll see some so-called atypical presentations. We already spoke about CSF pressure. And then it's probably recommended to rather speak about CSF volume rather than pressure. We need to pay attention to precipitating and predisposing factors as well as to some cognitive biases we as providers might have.

A headache that is there upright, it's present almost everybody, but not everybody, 92 percent according to that meta analysis. How long it takes for the headache to come when you, when you stand up, or how long it takes to peak to the maximum intensity. Two hours appears to be a reasonable time, but maybe one hour is a better time.

And some patients, as many of us know, can't stay upright more than four to ten minutes. Typically and classically, the headache improves quickly with laying down in patient with SIH, but that improvement might not be complete. And the longer it takes, the less certain we are about SIH being the culprit for these conditions.

Many of our patients reported the headaches gets worse as the day goes by. And the second half of the day pattern, if it's consistent, is pretty suggestive. And we oftentimes have to go back months or years in time for the patient history to elicit the orthostatic feature that might have been present before.

For example, if you have just 10 weeks as a cutoff, there is a probably 30 percent drop in the sensitivity of this, this test. Some patients have paradoxical worsening of the headache with laying down, or in the morning. And one another example would be many patients complain of back of the head pain or upper neck pain with standing, and that can be CSF leak, but many patients are asking about other cervicogenic type of causes.

It's not, it's an open question what a typical presentation of SIH is. In the, in the headache clinics, the most common problem that we deal with is the entity of chronic daily headache with or without orthostatic feature. Most of the time, it does end up being a chronic migraine, but I have low threshold to test for CSF leak in such patients because the leak can make a headache worse or refractory.

NDPH or newly persistent headache is a variant of that, and SIH can start out of the blue and may not go away for at least three months. That would be the definition of NDPH. Some patients develop a headache that goes from 0 to 10 out of 10 within a minute, like a thunderclap, and they may show up in the emergency room thinking they have a bleed.

Another instance is that patients with SIH can have their leak fixed, but then can leak again and we have to be constantly re-evaluating those patients, especially if they have a predisposing condition. Yes, children and older people can have CSF leaks, so can pregnant ladies, which can pose diagnosis and treatment challenges.

There are a lot of non-headache symptoms that patients with SIH can experience, which may lead the providers to look for alternative diagnosis, which may seem, seem like lower hanging fruits. But if somebody with SIH has headache and on top of that dizziness, it makes it more difficult for them to function.

Many patients with SIH have decreased hearing or ringing the ears, but some may have actually increased hearing. And the cognitive impairment or brain fog is really common in patients with SIH, but the minority of patients can have a well established mimic of behavioral variant frontotemporal dementia, which is due to brain sagging.

Fatigue can happen in maybe a quarter of patients and can be the main complaint. Another extreme is worse atrophy of both upper extremities, which is rather a complication. Severe light sensitivity is another common symptom which can be really, really disabling. Patients with SIH can have seizures, maybe more in kids, or movement disorders, but some of them are diagnosed, are labeled as functional neurological disorder by a provider not used to have SIH in the differential diagnosis, and that can be frustrating and stigmatizing for the patients.

This is the study that was recently published with 15 patients confirmed CSF leak interviewed by CSF leak experts, and all these 15 patients reported headache, prompted or non prompted. And all of them who reported head pressure reported it just volunteering. The other non headache symptoms that you can see here, which are similar with what the other slide has shown, had to be asked specifically by the interviewers. So speaking about the need for education of both providers and the patients on the complexity of CSF clinical diagnosis, the authors of the same paper list here—and I'm sorry, it's a, it's a busy slide—some useful patient reported outcome tools that can be used in general to look at the pain, to look at functioning, physical, emotional functioning, cognitive functioning, headache characteristics.

But the authors report or suggest the need to develop SIH specific symptom battery with headache characteristic and all the others characteristic, and I think we all may, may agree for that. There are some predisposing conditions to SIH or CSF leak, and I would mention bone spur or other spine changes may be the most common, but keep in mind that many patients with those changes don't have a CSF leak.

Connective tissue diseases make the dura weaker. Another condition, I'll just exemplify here the dramatic weight loss that may happen with bariatric surgery. It's another predisposing factor. Patients with CSF leak can be very well at some point, can stand up or sit up and exercise and do some straining.

Or some patients report to me that just living feels like it's giving, uh, changing the life because re-leaks. There is a huge gap between a 10, 15 percent rate of failure of diagnosis in medicine and the 90 percent gaps 90% misdiagnosis of CSF leak that was reported in this study, older, about 20 years ago.

But some of that gap may be due to our own cognitive biases. And an example Dr. Chris Gottschalk puts in this paper is about Chiari malformation being misdiagnosed, or CSF leak being misdiagnosed as Chiari malformation based on the way brain MRI looks. And in his examples some of the biases include availability.

So you have a clear diagnosis, confirmation or outcome bias. There is a malformation and I can fix it. And a zebra retreat: patients unfamiliar with this presentation of SIH thinks that it's a

diagnosis less desirable, desirable to pursue, and it's costly, and it's not worth the effort. There are consequences to not diagnosing a CSF leak, including severe complications like subdural hematomas, superficial siderosis, cerebral vein thrombosis, and coma.

But that coma can be reversible if you diagnose it and treat it appropriately. Underdiagnosis of CSF leak can lead to poor outcomes as well as over diagnosis can. Clinical over diagnosis of CSF leak can lead to unnecessary invasive testing and eventually waste of resources and time and suboptimal outcomes.

This was said before by Andy, and I totally subscribe to that. We have to. We need to do better at identifying CSF leak to improve the quality of life of our patients. I appreciate your attention. And then we'll hear from Dr Friedman. Thank you.