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Challenges in the Radiographic Diagnosis of Intracranial Hypotension

Good morning, everyone. My name is [Dr. Jessica Houk]. I'm an assistant professor of neuroradiology at Duke University, and I'm really excited to be here today. I wanted to start by thanking Dr. Andrew Callen for the invitation to talk to you all and thanking the [Spinal CSF Leak Foundation] for putting on this symposium.

So today I was charged with talking about the challenges in radiologic diagnosis of intracranial hypotension. Most of you know that's a very broad topic, right? And we heard from Dr. Friedman this morning that the majority of patients with SIH do have a positive brain MRI, but there are a minority, somewhere in the range of 10 to 15 percent, that do have negative brain MRIs at presentation.

And we also know that Chiari malformation— or SIH is often misdiagnosed as Chiari malformation, and we see that not infrequently in the reading room, and so that, that continues to remain a big issue when talking about imaging. I recently took the certifying exam and I was pleased to see that there were actually a handful of questions that wanted future neuroradiologists to be able to distinguish between those two entities.

So they're testing us on that now, which I think is hugely important. Today though, I'm going to focus on the Bern score and how it relates to clinical severity, so, specifically focusing on the paper that we published in AJNR this year, titled "Validity of the Bern Score as a Surrogate Marker for Clinical Severity."

I have no relevant disclosures. A bit of background on the Bern score. I know a lot of you are probably familiar with it, but the Bern score is a quantitative scale characterizing the severity of brain MRI changes in spontaneous intracranial hypotension. And its initial use was to predict the probability of a spinal leak on subsequent spine imaging, so originally using CT myelography.

And this was the landmark publication that came out in 2019 from the Bern group in Switzerland, titled "Assessing Spinal CSF Leaks and SIH with a Scoring System Based on Brain MRI Findings." And at ASNR this year, this group actually mentioned that they didn't term the score "the Bern score" when they wrote this paper, but kind of casually that's become what the scoring system is known as.

And we've seen in recent years since this publication a growing interest in the Bern score and specifically in brain MRI findings and how they relate to patients underlying pathophysiology. So, this was a follow up publication that came out

looking at the diagnostic yield of lateral decubitus DSM and this was looking at CSF-venous fistulas.

And the Bern score also showed utility in stratifying these patients for whether or not they would have a positive finding on DSM. A recent terrific paper came out of Colorado from Callen and colleagues looking at the relationship of the Bern score with spinal elastance and opening pressure. So again, more interest in looking at brain MRI findings and what their association is with the underlying pathophysiology.

And then of course, we're all interested in what the brain MRI looks like following treatment, right? So following closure of the dural leak or treatment for the venous fistulas. So a little bit about the Bern score assessment. So how do we come up with the score? So there's major criteria, for which patients get two points each for these, and then minor criteria, which are given one point for each of these.

So we're looking at engorgement of the venous sinus, pachymeningeal enhancement, the suprasellar cistern measuring less than four millimeters; and the minor criteria are looking at subdural fluid collections and then other manifestations of brain sag to include a decreased prepontine cistern

measuring less than five millimeters, and then a mamillopontine distance measuring less than 6. 5 millimeters. So the total score is used to stratify the probability of a patient having a spinal CSF leak, with low being less than or equal to 2, intermediate being 3 to 4, and high greater than or equal to 5.

So we know that brain MRI changes can reverse after successful treatment. This raises the question, can the Bern score be used to assess success after treatment? Can we look at the brain MRI and decide whether someone has been successfully treated? But in order to say that, we would need to determine whether clinical severity, and that's typically headache severity, correlates with these changes on brain MRI.

And so at the time of this paper and at the time of us talking about this idea and this work, the relationship between clinical headache severity and the Bern score had not yet been evaluated. So the purpose of our paper was to determine the degree of correlation between the pretreatment Bern scores and headache severity in SIH.

And this led to our publication looking at the validity of the Bern score as a surrogate marker for clinical severity in these patients. A little bit about the paper. So, in our methods, this was a retrospective cohort of consecutive adult patients who underwent workup for SIH. All of these patients at Duke complete a headache impact test score or a HIT-6 score as part of our routine evaluation.

And I'll talk a little bit more about the HIT-6 score as we go here. And then brain MRIs were reviewed and HIT-6 scores for each subject were reported.

As far as inclusion criteria, we included consecutive patients who did complete this headache impact testing questionnaire. They all must have had an available brain MRI showing signs of SIH. And then, of course, if patients did not have a pretreatment brain MRI or if their MRI did not have IV contrast on board, we did not include those patients as you cannot calculate an accurate Bern score without IV contrast on board.

And then we really focused on having the brain MRI obtained within a timeframe of zero to six months from the HIT-6 score being collected, because we really wanted to capture what the brain MRI looked at the time point of symptom severity. And then if patients had an incomplete HIT-6, they were also excluded.

So we looked at assessing the correlation between the HIT-6 score and Bern scores. We calculated a correlation coefficient and a 95 percent confidence interval. We also fit a regression line to look at this relationship and then used ROC curves to determine the ability of the Bern score to discriminate between low severity and high severity headache groups using the HIT-6 scores.

We had 57 subjects, 61 percent were female and the mean age was 54 years. So these were our results. And I think these are really important results. So there was a low correlation between HIT-6 and Bern scores in all groups. So in the overall cohort, there was a low correlation, in the patients that had a brain MRI within three months of the HIT-6 questionnaire, there was a low correlation.

And then also those that had a brain MRI between three to six months of obtaining the HIT-6. Further, the Bern score performed poorly at discriminating subjects who fell into the headache severity categories of severe versus not severe. Screen left, the area under the curve was 0. 635, and screen right, in those patients who had a brain MRI between zero to three months, the area under the curve was 0.606. So: limitations for our study. It was retrospective. Duke is a quaternary institution. And then this is important. And this is something we talked about when we were looking at doing this study is that HIT-6 has not yet been validated specifically in patients with SIH. It has been used and has been widely validated to assess the impact of headache severity and patients with episodic and chronic migraine syndrome, and in clinical trials for those patients, it has been used, but really,

when we were looking at this, it was the best measure that we had to look at impact on quality of life and headache severity. The conclusions of the paper were, importantly, that pre-treatment Bern scores show *low* correlation with clinical headache severity in patients with SIH. And so these scores do not reliably reflect headache severity and should not replace clinical outcomes measures when we're looking at these patients and how they're doing post-treatment.

So I wanted to show a couple of imaging findings and examples here from the paper. So this was a patient that presented with orthostatic headache, and on their initial MRI, the only finding of SIH was a venous distension sign. So they had a Bern

score of two. Their HIT-6 survey came back at 67, which puts them in the high severity category from a headache severity standpoint.

They went on to have a CT myelogram, which showed an epidural fluid collection and a calcified disc osteophyte here, causing the dural tear. And just to juxtapose that, so this was a different patient who also presented with orthostatic um, headache. Their Bern score was 4 based on measurements of the suprasellar cistern, the prepontine cistern, and pachymeningeal enhancement, so. Higher Bern score than the patient that we previously looked at, but their headache, their reported subjective headache severity and quality of life impact, was substantially lower than the patient that we saw previously.

This patient also went on to have a CT myelogram, which showed an epidural fluid collection and a disc osteophyte there. So the takeaways here and the things that I want to leave you with, the Bern score is an important tool for standardized imaging analysis in patients with SIH, and I think there's a lot of work to be done about why different pathophysiologies in these patients manifest with different findings on brain MRI.

But importantly, brain MRI following treatment should be interpreted in conjunction with clinical evaluation and with prior spinal imaging. And our group has been working on this for the last two years, working on coming up with a clinical outcomes measure. We saw several of those papers leading into that work by Tim Amrhein and colleagues.

And we're really hopeful that these endeavors will result in helpful management and treatment algorithms for our patients. And with that, I'll thank you for your attention.