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Monitoring Disease Activity in Sjögren's Disease

Announcer:

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This episode of Living Rheum, titled "Monitoring Disease Activity in Sjögren's Disease," is sponsored by Novartis US Clinical Development and Medical Affairs. The host and speaker have been compensated for their time. This program is intended for health care professionals.

Here's your host, Dr Ethan Craig.

Dr Craig:

Unlike diseases such as rheumatoid arthritis, Sjögren's disease has no calculable disease activity scores or established guidelines for disease activity monitoring. On this episode, we'll focus on the monitoring of disease activity in both research and clinical environments for this disease.

This is ReachMD, and I'm Dr Ethan Craig. Joining me to discuss disease activity monitoring for Sjögren's disease is Dr Sara McCoy. Dr McCoy is an associate professor of rheumatology at the University of Wisconsin. She also runs the Sjögren's Clinic at the University of Wisconsin and serves on the board of directors for the Sjögren's Foundation. Dr McCoy, thanks for being here today.

Dr McCoy:

Thanks for having me.

Dr Craig:

Dr McCoy, let's begin with a look at disease activity monitoring in clinical practice. Can you describe your clinical workflow when you're working with Sjögren's patients and the types of measures that you're using and thinking of in a clinical setting?

Dr McCoy:

Yeah, I'd be happy to. So, at the initial visit, I perform a Schirmer's test and an unstimulated whole salivary flow, which sounds pretty complicated but it's actually pretty easy. So, the Schirmer's test is just a little strip of paper you pop in the outer third of the lower lid of someone's eye, and after 5 minutes you can measure how much tear flow they've had. And then simultaneously I do an unstimulated salivary flow, which is essentially having the patient empty their mouth and then drool in a cup for 5 minutes. And because both are 5 minutes, to save time I do them both at once. And that gives me an idea of whether or not the patient has Sjögren's disease, so diagnostically, and clinically, to see if their disease is progressing in terms of their xerophthalmia or xerostomia.

I think that the workflow with the Sjögren's patient is pretty unique. So, what I do first is I have a set of questionnaires I have my patients fill out. And this is actually in the process of evolving. So right now, I have them complete an ESSPRI, a xerostomia index, and EAT-10. And this is to screen overall for how they're feeling, what I can target, and then also how their dysphasia is doing.

So, the ESSPRI—E-S-S-P-R-I—comprises 3 visual analog scales from 0 to 10, and the 3 scales are in the domains of pain, dryness, and fatigue, so it's usually pretty fast to fill out. And I like having these available because there's the cardinal symptoms of Sjögren's disease, and they provide a global view of how the patient is feeling. And then I have them fill out the xerostomia index and the EAT-10, to grasp how their oral dryness is impacting their life and ability to swallow. And I can use the results of the questionnaire pretty quickly to sort of tailor the review systems and steer our initial discussion. And I do think the dysphasia is frequently overlooked, and so I use the

questionnaire as a quick screening tool, and I can do things like swallow therapy or speech therapy referrals based on some of the answers to their questions. I will say that I'm really interested in the PROMIS as another index of determining how the patient is doing, and that's something that I'll be doing in the near future.

As another example of how I might use these results, if a patient has really high fatigue, then I might ask them to describe the fatigue, and if it's a physical tiring, I can provide them tips on sleep optimization exercise regimens that might help. And if the fatigue seems more mental, akin to a brain fog, then I provide tips on cognition and brain health. So, in general, once we get the patient seated, I already have a quick snapshot into how they're feeling.

Dr Craig:

That's great. And what types of lab and imaging assessments do you perform to monitor patients with Sjögren's?

Dr McCoy:

So, when I see a patient who's diagnosed with Sjögren's disease, and without overlap with other diseases like lupus, I perform a lab evaluation that encompasses disease activity measures and I also evaluate for organ involvement. So usually, I order a CBC with differential, a CMP—and this is important because it has liver tests to screen for things like PBC or autoimmune hepatitis, it gives us our electrolytes which help us screen for things like RTA or renal tubular acidosis, and also kidney tests to make sure that they aren't getting, you know, pretty rare glomerular nephritis or more common tubular interstitial nephritis. Akin to that, I get a urinalysis and a urine protein-to-creatinine ratio. And the list goes on, because I also get immunoglobulins, cryoglobulins, and rheumatoid factor.

And part of the reason why I get these tests is they're part of our ESSDAI, which I'll talk about a little bit later, our disease activity index. I make sure all patients who I'm seeing for the first time don't have HIV or hepatitis C testing. If you listen to the first episode of the podcast, these viruses may serve as a trigger, but also can mimic Sjögren's disease. At the initial visit, and intermittently on follow-up, I'll order a serum or a urine protein electrophoresis, and a β 2-microglobulin or an LDH.

And then I also do want to say, in 2021 there was a really nice guideline published in CHEST for how to evaluate and manage Sjögren's patients with lung involvement, and in that guideline, it was recommended to perform a chest x-ray and pulmonary function tests as screening on all Sjögren's disease patients, even if they're asymptomatic. So, I now perform that at the initial visit.

I recommend, but don't require, a labial salivary gland biopsy or a minor salivary gland biopsy. And this helps us in diagnosis of Sjögren's disease, but also can help in a diagnosed Sjögren's disease patient for prognosis because we know that a focus score greater than 3 can be associated with lymphomagenesis and with extraglandular involvement of Sjögren's. And then finally, I quickly pop on the ultrasound, and do a nice bedside salivary gland ultrasound. And I do this at least at baseline, and we know that the ultrasound can sort of help us also prognosticate things like lymphoma and also help us diagnose causes of glandular enlargement.

Dr Craig:

That's a lot for a visit. So, if we shift gears to the research side and the research component of disease monitoring, can you tell us about the current measures that are being used in clinical trials and in other research in Sjögren's disease?

Dr McCoy:

Thanks for asking that. Right now, one of our major tools is the EULAR Sjögren's Syndrome Disease Activity Index, or the ESSDAI. And it's a disease activity index which includes 12 domains developed by a consensus of worldwide experts. What are these 12 domains? So, the 12 domains include things like constitutional symptoms or signs, and this is fevers, night sweats, or unintentional weight loss. It includes lymphadenopathy or lymphoma, glandular involvement (of note, the glandular involvement here is really glandular enlargement), articular, cutaneous, pulmonary, renal, muscular, peripheral or central nervous system, hematologic, and biologic involvement. And that biologic domain includes low complement, cryoglobulinemia, and hypergammaglobulinemia, which again feeds back to why I get some of these labs at some of the visits. Each domain is weighted, and the final score reflects the addition of each weighted affected domain.

So, the ESSDAI has the benefit of capturing the multiorgan and multifaceted nature of Sjögren's disease, but it does have some weaknesses. So, it fails to differentiate acute versus chronic features, or things that might be more or less responsive to treatment. And it also doesn't really capture the glandular symptoms that are characteristic of Sjögren's disease, right? So, we know that Sjögren's disease symptoms have significant dryness as one of the primary things that drives reduced quality of life, but that actually correlates inversely with ESSDAI. So, the greater the symptom burden of Sjögren's disease, we actually tend to find lower ESSDAIs.

To try to address some of these shortfalls, there have been 2 additional disease activity indices that have been proposed. The first is CRESS, or the Composite of Relevant Endpoints for Sjögren's Syndrome, and the second is STAR, which is the Sjögren's Tool for

Assessing Response.

The CRESS incorporates the ESSPRI which, if you remember, is that sort of quick, you know, how bad is their pain, dryness, fatigue symptom. And it also includes glandular tear and saliva involvement by incorporating the ocular staining score or the salivary flow. But it doesn't require improvement of symptoms, and that is really important, right? A patient should feel better if a drug's effective.

And so, here comes the STAR, and the STAR requires improvement in ESSPRI and in 1 of 4 other domains. So, you have to feel better and either have systemic improvement; improvement in lacrimal gland function, so, tear formation or surface staining; improvement in salivary gland function, like the unstimulated salivary flow; and improvement in the biologic domain. So, you can see that we've sort of had these 2 burgeoning disease activity indices that are coming to the forefront, and I think we'll be hearing a lot more about them in the context of clinical trials.

Dr Craig:

So, Dr McCoy, looking to the future then, are there any measures that you're particularly excited about when it comes to monitoring disease activity for Sjögren's disease? Anything new on the horizon you can think about?

Dr McCoy:

Yeah, so as I mentioned earlier, I'm changing my workflow, and I'm going to try to simplify my questionnaires for my patients and I'm planning on using the PROMIS-29. And that's based on a great recent study by Dr Dana DiRenzo, who's at your institution. And her recent study shows PROMIS provides a broad picture of healthcare-related quality of life in Sjögren's disease.

Dr Craig:

Well, thank you for shouting out Dr DiRenzo and my institution. I appreciate that.

Dr McCoy:

No problem.

Dr Craig:

Well, with these thoughts in mind, I really want to thank our guest, Dr McCoy, for helping us better understand how we can monitor Sjögren's disease activity and how we can think of doing this in the clinic and in research. You know, Dr McCoy, it was a great pleasure to speak with you today and thank you for your time.

Dr McCoy:

Thanks for having me.

Announcer:

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