HDC Podcast Series

TITLE	Understanding Huntington's Disease Chorea: Functional Impact and Best Practices
LENGTH:	17:04 min
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LEARNING OBJECTIVES:

- Define Huntington's disease chorea (HDC) and understand its functional impacts
- Understand the value of assessing HDC, and learn key questions health care providers (HCPs) can ask to evaluate the effect of HDC on a patient's daily life and function
- Explore strategies and advice to help patients and care partners effectively communicate the impact of HDC with their HCPs

SCRIPT

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Opening

Dr Klepitskaya: Hello, and welcome to today's podcast! I'm Dr Olga Klepitskaya, a director in Medical Affairs at Neurocrine Biosciences and a movement disorders neurologist. Today I'm thrilled to be joined by 2 highly respected colleagues, both of whom are experts in Huntington's disease.

First, Dr Seeberger, could you please introduce yourself?

Dr Seeberger: Hi everyone! I'm Dr Lauren Seeberger, and I'm also a movement disorders neurologist. I currently work at the VA Medical Center in Boise, Idaho. I was formerly the Director of the Huntington's Disease Society of America, or HDSA, Center of Excellence at the University of Colorado.

Dr Klepitskaya: Wonderful! And now, Dr Sung?

Dr Sung: Hi everyone! I'm Dr Victor Sung, and I too am a movement disorders neurologist. I serve as the Director of the University of Alabama at Birmingham Huntington's Disease Center of Excellence.

Dr Klepitskaya: It's an honor to have both of you here. Let's dive into our discussion about Huntington's disease chorea and explore its impact on patients and families.

Overview of HDC

Dr Klepitskaya: Let's start with a foundational question for our listeners. Dr Sung, what exactly is Huntington's disease, and what do we mean by Huntington's disease chorea?

Dr Sung: So, Huntington's disease is a pure neurodegenerative disorder. It manifests as a triad of symptoms, which we call the "Huntington's disease triad," that includes motor, cognitive, and behavioral symptoms. But the hallmark symptom of Huntington's disease is chorea. The word "chorea" comes from the Greek word for "dance," so it describes the irregular, nonrhythmic, and abnormal involuntary movements that show up in more than 90% of patients with Huntington's disease.

This movement is so characteristic that Huntington's disease was once called "Huntington's chorea." So, although the disease involves more than chorea, this history highlights the importance of chorea in its diagnosis.

Dr Seeberger: Absolutely. And to add to what Dr Sung said, these movements are erratic and irregular and can involve the arms, legs, trunk, and even the facial muscles. These are excessive, irregular movements that are superimposed upon the patient's own natural body movements, so they can really disrupt everyday function.

Dr Klepitskaya: Thank you both for clearly explaining chorea in Huntington's disease. Now that we've defined Huntington's disease and chorea, let's discuss how chorea progresses over time.

Dr Seeberger: Of course! For many patients, Huntington's disease chorea begins with very subtle involuntary movements, like small, twitchy movements in their toes or fingers. These subtle movements often mark the onset of chorea.

Over time, these movements worsen and become more noticeable, so patients may experience both physical discomfort and psychological stigma.

Dr Sung: I agree with Dr Seeberger that chorea often starts subtly and, because it progresses slowly over the years, it can be almost imperceptible at first. Because of this, patients may not even notice the day-to-day changes because they've learned to live with it over the years.

Over time, the chorea tends to progress in both severity and amplitude of the movements. And then in the later stages of the disease, it often plateaus and is replaced by other symptoms, like rigidity, bradykinesia, and dystonia. It's important to note, though, that chorea doesn't always go away completely, but it might not continue to worsen as rapidly as in the mild and moderate stages of the disease.

Dr Seeberger: Exactly. Some patients can develop severe, excessive involuntary movements even after being admitted to a nursing home, so we may need to use specialized chairs to help accommodate their movements and keep them from getting injured.

Something we often see, as chorea worsens and the movements become more exaggerated, is that patients start having difficulty handling silverware or doing other routine daily life activities. They might have an involuntary jerk that takes them off course. It can make eating take longer, and some patients may abandon using utensils altogether if their chorea is severe. Oftentimes, care partners will just cut up the food for them, so that they only need to use a fork to eat.

Understanding the Functional Impact of HDC

Dr Klepitskaya: Those are great points. Dr Sung, could you explain how Huntington's disease chorea affects patients' daily functioning, starting with the early stages?

Dr Sung: One of my big "soapbox" points is that chorea shouldn't be discarded as "just an involuntary movement."

For instance, involuntary movements of the mouth and face can affect speech and swallowing. Chorea in the hands affects fine motor tasks, which can impact daily activities like work. Chorea in the trunk and legs can affect gait and balance. So everyday functions like walking, using their hands, and talking can all be affected by chorea.

Some of the early impacts of chorea are first noticed at work, with tasks that require high physical function. For example, I had a patient who was a mechanic, and his mild hand chorea caused him to drop and crack an engine block. This incident cost his auto shop $10,000 to replace. So that small twitch had a big impact, putting both his job and his Huntington's disease under a lot of scrutiny. Many patients have jobs that require coordination in using their hands or feet or speech articulation, and chorea can affect all of those.

Dr Seeberger: I agree. For me, 1 example that comes to mind is a patient who was an attorney. He began noticing mild involuntary movements in his hands and, since he had a family history of Huntington's disease, he watched closely and identified it very early. He sought treatment right away to avoid being perceived as impaired in the courtroom. That's an example of stigma from others seeing involuntary movements. For him, it was more about the emotional impact and stigma of visible movements than physical interference.

Dr Klepitskaya: These are powerful stories. And as the disease progresses, how does chorea impact daily functioning, Dr Seeberger?

Dr Seeberger: I think it's an intensification of previous issues, where they are more apt to knock over a glass or a bottle, which can be embarrassing out in public or even in their own home. I had a patient who worked on a factory line and had leg movements that made it difficult to stand in place, so other people worried that he could get caught in the machinery. They ended up asking him to take time off from work until he could get help to control the movements.

Dr Klepitskaya: That is quite a serious impact of having visible chorea symptoms. As chorea progresses and becomes more visible, what other issues might arise?

Dr Sung: Chorea can make patients look intoxicated, so I've had multiple patients who were arrested because they were mistaken for being intoxicated when they weren't, and I've had to write letters to get them out of jail. The HDSA provides cards that say, "I have Huntington's disease, a brain disorder," so patients can carry them in their wallet for this exact reason.

Dr Seeberger: I had a patient with a similar experience who was walking down the street because they were no longer driving, and they got picked up by law enforcement who thought they were intoxicated. I also had a patient who took her car to the dealership and when she went back to pick up the car, they wouldn't give her keys back and they called the police. That was a dramatic example—they were withholding her car and judging her even though she had brought the car in the day before.

I also recall another patient who struggled to apply her eyeliner due to chorea. She always liked to look put together for work but, with the involuntary movements, even doing her hair was onerous and time consuming.

Dr Klepitskaya: Yes, everyday tasks like applying makeup can become challenging, and even simple things like buttoning shirts may require changes to their wardrobe. Dr Seeberger, have you seen this?

Dr Seeberger: For sure—they tend to wear slip-ons.

Dr Sung: One question we often ask to assess the function of a patient is if they can dress themselves. Care partners usually say that the patient can manage with slip-on shoes and pull-on clothing, but struggle with buttons, zippers, and shoelaces. This speaks to the impact that chorea has on fine motor tasks and a patient's level of independence.

The Importance of Assessing the Functional Impact of HDC

Dr Klepitskaya: That really shows the significant functional impact that Huntington's disease chorea can have on patients, even early in the disease. With that in mind, can you also discuss why it is important to ask patients and their care partners about the functional impact of chorea?

Dr Sung: Yes. As we've discussed, chorea develops slowly, so patients often get used to it. Additionally, anosognosia, or disease unawareness, is common in Huntington's disease. This means patients often don't have a lot of insight or awareness of their chorea, so they may not bring it up to their health care provider. This makes it our responsibility as clinicians to ask about and assess for chorea.

One approach is to ask the care partner because they often notice the movements when the patient does not. You know, we all have these stories where you ask the patient, "Have you noticed abnormal movements like chorea?" and the patient says, "No, I haven't." And then the care partner is in the background, shaking their head vigorously, like, "Yes, they definitely have that." So, involving the care partner is a key piece of the assessment.

A structured assessment is helpful for both initial evaluation and tracking the progress of treatment. Having a structured way to document it—say, by labeling it as mild, moderate, or severe—gives you an objective way to see how chorea is changing over time. In multidisciplinary Huntington's disease clinics, like mine and Dr Seeberger's, we are familiar with using the Unified Huntington's Disease Rating Scale, or UHDRS, especially for chorea. In my clinic, I assess the maximal chorea score on every patient at every visit. I also train residents and fellows to do the assessment. I know this is not feasible for everyone to do, but that's 1 of the things I use, and I like having that numerical score that I can see change over time.

Dr Seeberger: I agree, and I'll add that since most patients are cared for by primary care physicians, it's helpful for physicians to monitor chorea, documenting it as mild, moderate, or severe. They may never end up using the UHDRS or a total maximal chorea score, but this scale helps them to consider how chorea progresses over time and the impact of chorea on functionality. Then they might start asking more detailed questions like “Is your chorea affecting your ability to type or write by hand?”

And like Dr Sung mentioned, sometimes patients don’t actually accept that they have chorea. So instead of labeling it as "chorea," we can focus on function, like asking, "Do you drop things? Can you get a button through a buttonhole?”

It's also important to teach primary care physicians how to ask these practical questions to make patients more comfortable and the topic more relevant to their everyday lives.

Questions to Assess the Impact of HDC

Dr Klepitskaya: I think we can agree that the common clinical question "Is it bothersome?" might not fully assess the functional impact of chorea. In your practice, are there any particular functional domains or specific questions you might ask that you feel are best at assessing the functional impact of Huntington’s disease chorea?

Dr Sung: I focus on 3 areas: face and mouth, arms, and legs. For face and mouth, one of my go-to questions is about speech articulation, which can often go unasked. I’ll ask, “Does anyone ever ask you to repeat yourself, whether in a face-to-face conversation or on the phone?” This 1 question can reveal a lot, especially with patients who have mild chorea. Sometimes, this brings out a bit of frustration, like, “Yes, every time I call my wife, she’s asking me to repeat myself and it’s driving me crazy.” Then I’ll follow up by asking, “Well, if we could improve that, would you be interested in treating it?”

For arms, I’ll ask, “Do you ever spill or drop things?” We also often ask if the patient can dress themselves, and care partners usually say that the patient can manage with slip-on shoes and pull-on clothing, but struggles with buttons, zippers, and shoelaces. This highlights how chorea affects fine motor tasks and a patient’s level of independence.

For legs, I ask about things beyond falls, which they often deny. Instead, I ask, “Do you ever trip or stub your toe on things?” That subtle balance issue can often be tied back to chorea, and we know it’s something we could potentially treat.

Dr Seeberger: We’ve discussed this a lot at my clinic. We ask questions like, “Can you still cut your own food?” and “Are you still able to handle a fork and knife?” I also ask, “Have you had to switch from a regular glass to something with a lid or something plastic?” These questions highlight the importance of primary care physicians quantifying chorea as mild, moderate, or severe and address its impact on daily living.

Dr Klepitskaya: I think our listeners now understand why it's important to assess Huntington's disease chorea both clinically and for its functional impact, and what different questions we can ask to truly identify this impact.

Advice for Patients and Care Partners to Communicate Impact to HCPs

Dr Klepitskaya: Can you summarize your advice for patients and care partners on how to communicate the impact of Huntington's disease chorea to their providers?

Dr Sung: It's interesting—patients often don't recognize the impact of chorea, so the first step is helping them to see it. However, the burden often falls on the care partner who is going to notice these things more easily than the patient will.

If the patient is in denial or doesn't see it as clearly, the care partner can find a way to voice concerns to the health care provider. The tricky part with that is they're trying not to fracture their relationship with their loved one who has Huntington's disease. Not everybody can find that voice easily. If you feel like your relationship can handle it, as a care partner, the advice is just to speak up as much as you can.

It's important for health care providers to create opportunities for care partners to share their thoughts and concerns in a supportive environment. For example, setting aside time for private conversations with care partners ensures that they can express themselves openly without feeling uncomfortable discussing sensitive topics in front of the patient.

Dr Seeberger: I agree, and we should ask detailed questions, like: Is the person with Huntington's disease comfortable sitting in their chair at home or if there's a certain chair they can sit in that they won't fall out of? How are they sleeping at night? Are they comfortable? Are they able to lie fairly still as they're going to sleep? As you start to ask these detailed questions about motor function, standing, sitting, transitioning, and getting in and out of the car, the care partners are going to start to pay attention to that. Those with early Huntington's disease may start to pay attention too and be able to give us feedback.

Conclusion

Dr Klepitskaya: Before we wrap up, thank you both for sharing your insights and recommendations. Any last thoughts for our audience?

Dr Seeberger: We've discussed the progressive nature of Huntington's disease chorea and how the movements can really disrupt everyday function. The examples we shared are real patient stories from our clinics about the true functional impact and burden of chorea. I think it's important for health care providers to ask detailed questions like the ones we mentioned today. I don't think we've been as detailed in asking about chorea as we should have been in the past and as we should be going forward. While we measure it well, we haven't been as rigorous in how we ask about the functional impact, which is a gap. We need to improve how we ask questions about the impact of chorea in several areas of life.

Dr Sung: I agree. We mentioned that chorea develops slowly, so patients often get used to it. This makes it our responsibility as clinicians to ask about and assess how chorea is impacting the patient and their care partners. When asking about the functional impact of chorea, some points to consider include talking to the care partner when possible, using structured assessments to

monitor how chorea is changing over time, and focusing on 3 areas: the face and mouth, the arms, and the legs.

Dr Klepitskaya: Absolutely.

Thank you for joining us on today's podcast. As we explored Huntington's disease chorea and the value of understanding its functional impacts on patients' daily life, we also discussed strategies and specific questions that health care providers, patients, and care partners can ask to help identify the impacts of chorea. We hope this conversation provided valuable insights to better support those living with Huntington's disease chorea. Thank you for listening.