Transcript Details

This is a transcript of a continuing medical education (CME) activity. Additional media formats for the activity and full activity details (including sponsor and supporter, disclosures, and instructions for claiming credit) are available by visiting:
https://reachmd.com/programs/cme/challenges-in-recognizing-and-diagnosing-idiopathic-hypersomnia/17908/

Time needed to complete: 1h 02m

ReachMD
www.reachmd.com
info@reachmd.com
(866) 423-7849

Challenges in Recognizing and Diagnosing Idiopathic Hypersomnia

Announcer:
Welcome to CME on ReachMD. This episode is part of our MinuteCE curriculum.

Prior to beginning the activity, please be sure to review the faculty and commercial support disclosure statements as well as the learning objectives.

Dr. Rodriguez:
Hi, I'm Dr. Alcibiades Rodriguez. I'm a Neurologist and a Sleep Physician with NYU Grossman School of Medicine. Today we're going to talk about Challenges in Recognizing and Diagnosing Idiopathic Hypersomnia.

Idiopathic hypersomnia, as you may know, is one of the central nervous system hypersomnias. The idiopathic essentially means that we don't know the origin and may well be a group of diseases, is characterized by the excessive daytime sleepiness, no cataplexy, because otherwise it would be narcolepsy type 1. And we should do testing, including a PSG showing 6 to 7 hours of sleep with no sleep-disordered breathing or the sleep disruption that may cause the sleepiness, and an MSLT with less than 8 minutes of mean sleep latency characterized hypersomnia or diagnosed hypersomnia, right, and have at less than 1 SOREMP in the MSLT and of course no SOREMPs in the PSG. And then naturally, you can use a total 24-hour with a sleep time with more than 660 minutes, around 11 hours, which you can document by PSG out for – with actigraph; however, for practical purpose, I probably would not do that. And you need to be sure there's no other issues involved like a medication, friction, trauma to the brain, or insufficient sleep, which is the most common cause of excessive daytime sleepiness. This is the diagnostic criteria for idiopathic hypersomnia.

With shorter, close supported criteria we can have in this case severe and prolonged sleep inertia that we're going to explain a little bit in the next slide. We'll call it sleep drunkenness; un-refreshing naps, either long or short, but un-refreshing; autonomic instability, including headaches, orthostatic disturbances, perception of temperature dysregulation, called Raynaud's phenomenon; memory and attention difficulties; and some depressive symptoms in around 15 to 25% of patients. Of this, probably the sleep drunkenness is the most important because it seems to be a little bit more common in these patients.

Usually starting in adolescence around 16 to 21 years old. And sleep drunkenness is characterized by prolonged difficulty waking up with repeated returns to sleep, irritability, and autonomic behavior and confusion. It could be very long, more than 1 hour is an extreme form of sleep inertia, seems to be present in around 33 to 66% of patients. It can be present in other CNS hypersomnia, including narcolepsy, seems to be more common in IH. Naps are long and unrefreshing. And when you see a PSG with a sleep efficiency of more than 90% in an adult, it may be suggestive of idiopathic hypersomnia.

Why so difficult? As I mentioned, it could be a group of diseases, right? But the issue is really rare. It's still rare, right? It starts in the teenager years, so many things are happening at that age. For example, most of the teenagers I see are deprived of sufficient sleep. Some of them may have delayed sleep phase, so they may sleep late, late, late and catch up on weekends and look like they're sleeping very long. Well, they may have both. At this point, may have some mood disorders, medication may be involved, and also recreational drugs. MSLT is inconsistent in children, is not that reliable. And even in adults, the 14 to 25% have been reported in these patients to shame diagnosis with narcolepsy, IH to normal so pay attention to that. Clinical suspicion is important. It's likely a heterogeneous condition that overlap with other CNS hypersomnia. So that's what makes it so confusing. It could be other sleep

disorder signal, for example, patients that get sleep apnea, you may treat them and they continue to be sleepy, and they have a history when they were younger and they were sleepy, most likely they have IH, but it's difficult to prove. What makes it difficult too is that there's no biomarker involved. So, traditionally of being a diagnosis of exclusion, even when we have our criteria.

So how do we clarify the diagnosis? So have a detailed clinical history, testing with appropriate clinical correlation, right? If patient has sleep drunkenness, the extreme form of sleep inertia, may be a clue. Other sleep disorder may be present or it doesn't mean that the patient doesn't have IH. There are new tools that we have like the Idiopathic Hypersomnia Severity Scale, which may be helpful as a measure of burden of symptoms and also as a follow-up and treatment response tool. So, all those things can help us to elucidate IH.

This is a nice graphic where we see the overlap between IH and narcolepsy type 2. We have many similar things including sleep inertia; however, you find this most likely will have IH, also they have long unrefreshing naps. So little clues that can help you to elucidate the mystery of idiopathic hypersomnia. Most of the patients are underdiagnosed, which is what you need to think about.

Hopefully that was helpful for you and have a good day. Thank you.

Announcer:
You have been listening to CME on ReachMD. This activity is jointly provided by Global Learning Collaborative (GLC) and TotalCME, LLC. and is part of our MinuteCE curriculum.

To receive your free CME credit, or to download this activity, go to ReachMD.com/CME. Thank you for listening.