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Pediatric Narcolepsy: Addressing the Challenges in Its Recognition, Diagnosis, and Management

Announcer:

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Dr. Thorpy:

This is CME on ReachMD, and I'm Dr. Michael Thorpy. Today, Dr. Anne Marie Morse and I are discussing the diagnosis and management challenges associated with pediatric narcolepsy, both with and without cataplexy.

Welcome, Dr. Morse.

Dr. Morse:

Thank you, Dr. Thorpy. I'm so glad to be here today.

Dr. Thorpy:

So we'll start off by talking about the narcolepsy symptoms in children versus adults. Anne Marie, as you're aware, there are differences between adults and children in the symptoms, and we know that most patients with narcolepsy develop narcolepsy at a median age of 16, so there's 50% of patients, again, who develop narcolepsy in the pediatric age group. So it's most important to recognize the symptoms in that age group. So can you tell us about those differences?

Dr. Morse:

Sure, so I think it's important to recognize that the condition is the same condition, right, medically. However, the symptoms may have a very different look and feel to them. And what we know is that when you talk about narcolepsy, the pentad symptoms include excessive daytime sleepiness, sleep-related hallucinations, sleep paralysis, cataplexy, and disturbed nocturnal sleep. Now when we see the pediatric onset of narcolepsy, there are not only those pentad symptoms, but there also may be some other medical clues that can be helpful. So first and foremost, it's important to recognize excessive daytime sleepiness. What's the nuance in pediatrics? What I mean by that is we most typically are looking for sleepiness as a person who's just inappropriately falling asleep. However, we know that in the prepubescent population especially, that they may actually look more like an ADHD [attention-deficit/hyperactivity disorder], hyperactive type.

Now the flip side can be true when we're talking about postpubescent and a teenager where they may look like more of an ADHD, inattentive type or perhaps a moody teenager who's very irrational or having a lot of arguments with their parents.

Now what's interesting is when you look at the adult phenotype of an individual with narcolepsy, we generally are saying that they actually are not a hypersomnia disorder; they're a hypersomnolence disorder. So they're not having an increased number of hours of sleep in a 24-hour period. In pediatrics, the pediatric onset, we are seeing that sometimes a longer duration of sleep in 24 hours can be

a clue.

Now when we're looking at other features that may be present in pediatric narcolepsy, cataplexy is an important one to distinguish. The current definition of cataplexy is actually wrong. We generally are describing it as these transient episodes of loss of tone, and the reason I say it's wrong is because we know in pediatrics that they can have active motor phenomena as one of their manifestations, including things like eyebrow rising, tongue thrusting, even dyskinetic movements with a persistent hypotonia. And so they can also have those loss of tone and those drop attacks, as well as head drops and their knees buckling, hands dropping from their sides. But it is important to recognize that, and sometimes those can also be independent of any type of emotion, as opposed to that laughter-induced type of experience.

The other pieces I think are important is when talking about the sleep-related hallucinations, very frequently these get confused with just nightmare disorder, and so these children can have both. They can both have very recurrent nightmare, but also describe these really clear depictions of this occurring in their room and being a part of their every single day.

Now outside of this pentad symptoms of narcolepsy, one of the other pieces that is very, very striking and an important one to recognize is that they can have a very abrupt onset of a significant weight gain and really go from a normal weight percentile to going into the category of obesity. So really understanding these other factors that contribute to what the manifestations look like can also help in preventing them from going down that diagnostic delay pathway.

Dr. Thorpy:

Good. Thank you, Anne Marie.

Dr. Morse:

Dr. Thorpy, now that we have a better understanding of these varying symptom presentations between adults and children, how do we get to a diagnosis of pediatric narcolepsy?

Dr. Thorpy:

Well, yes as I mentioned earlier, the diagnosis is often missed in children, and so people have to be really attuned to thinking about narcolepsy in a child that's exhibiting some abnormal behavior. And this really begins with the parents and the teachers, because very often it's these symptoms that you just mentioned that occur that the parents have to pick up, because if they don't bring the child in to see the pediatrician, then obviously, the child's not going to get diagnosed. And also in school, if a child's falling asleep in school, that has to be something that needs to be brought to the attention of the parents so the parents can then, again, seek help from a professional. And you have to think that there has to be a cause for it, and if there's no clear cause, then you have to go to the physician and ask for a more intensive investigation of it. And of course, if there's any evidence that there's any atypical movement disorder associated with this, then that has to be brought to the clinician.

And the clinician needs to be aware of narcolepsy. So very often the clinician is missing it, so it's most important that the physician is aware of these differences in cataplexy between children and adults so that he can pick up on it and then get the child referred to a sleep specialist who can do the appropriate testing and make sure that the child gets an accurate diagnosis.

Dr. Morse:

I think some other things that are important to recognize is that based on their developmental age, there's going to be differences in terms of what are expected. We know that the duration of sleep changes over time. We also know that the current testing that we utilize – the polysomnography and a Multiple Sleep Latency Test – that those diagnostic criteria are based on adult information. And in fact, when we look at the average sleep latency, that is heavily influenced by Tanner staging, and so what might be viewed as not pathologic, based on their Tanner stage and developmental age, may actually be pathologic. And so I do think it's really important to utilize all of this information as you're approaching not only collecting the history, but also as you're thinking through what the diagnostic workup may look like and what those results may yield.

Dr. Thorpy:

For those just tuning in, you're listening to CME on ReachMD. I'm Dr. Michael Thorpy, and here with me today is Dr. Anne Marie Morse. Our focus today is challenges associated with diagnosing and managing pediatric narcolepsy with and without cataplexy.

Dr. Morse, as you know, there are a number of conditions that tend to occur around the time that a child develops narcolepsy, and these comorbidities, they can be either medical disorders or they can be psychological factors. So can you tell us a little bit about these comorbidities and how they can make the diagnosis a little more difficult to make?

Dr. Morse:

So as we had mentioned earlier, we do recognize that there can be a significant weight gain, and so therefore that's something to

definitely be on the lookout. In addition, there can potentially even be things like precocious puberty or impacts on how someone is experiencing their pubertal changes. We also describe that some of these symptoms can look like things like ADHD, depressive syndromes, anxiety. However, those can also occur as true comorbidities. It's important to recognize, as in any chronic medical condition, that there is a major adjustment that goes along with this. And so therefore, not being able to perform at your optimal is going to lead to true feelings of depression or self-esteem issues. The big piece that we want to make sure that we're always focusing on, also, is that quality of life and how it's impacting them socially in regards to not only their self-image, but how are they socializing with other children, and are there any feelings of self-harm, or suicidal thoughts? The final piece that I do think is important is also recognizing that this can have significant implications on school performance and academic achievement.

Dr. Thorpy:

Yes, the obesity and the precocious puberty are particularly interesting features that we see in the patient with narcolepsy, particularly the type that's associated with cataplexy.

Dr. Morse:

So, Dr. Thorpy, how do we tailor pharmacologic therapy for our patients diagnosed with pediatric narcolepsy, recognizing that we're limited in options, and we've already just covered all the range of impacts that the condition can have?

Dr. Thorpy:

Well, that's right. It's quite a different process in really treating the child compared with the adult because there are so many medications that are not approved for children that are approved for adults. Now of the approved products, the traditional stimulants – some of them tend to be approved, like methylphenidate and some of the amphetamines, but not all of them. And so if the clinician's going to stay on label, he should check and make sure which one is approved and which isn't. And then we have the other medications, which are typically used in adults, such as solriamfetol, modafinil, armodafinil. But again, these are not FDA-approved in children and so only should be used in special circumstances when there's very little other options available to the child.

And then when it comes to the treatment of cataplexy, we know that the methylphenidate and amphetamines don't do much for cataplexy, so the child with cataplexy, you either use one of the newer forms of oxybate or antidepressants, but the antidepressants are going to be used off-label, because there's no antidepressant FDA-approved for children. But the antidepressants can be very effective in treating the cataplexy, and so it may be an option that physicians need to consider.

But the most effective agents and that are FDA-approved for children for treating narcolepsy are the oxybate agents. And these forms of oxybate treat all the symptoms of narcolepsy. So we have 3 different forms of oxybate available to us now. There's the sodium oxybate form which is known as Xyrem. And this is taken twice at night. That may be a limitation for many children, because children have great difficulty in waking up at night. So that means that the parent has to get up and try to wake the child to give them the second dose, and that's not ideal.

Now sodium oxybate has a large content of sodium in it, so there is a formulation called Xywav, which is a low-sodium formulation – has 92% less sodium. But again, it needs to be given twice at night – once at bedtime, once 2.5-4 hours after the first dose – and again, somebody may have to get up and give that to the child.

Now there is a third form of oxybate available, but unfortunately it's not FDA-approved for children. The first 2 forms are approved for children 7 years of age and up, but Lumryz, a form of sodium oxybate which is a once-nightly form, is not FDA-approved for children, only in adults. And the advantage of Lumryz, obviously, is that it's only given once at night, so it can be given to the child before going to bed and the child doesn't need to wake up and take that second dose. So it can be an effective agent for children, but it's unfortunately not FDA-approved for children at this time, although the active agent is exactly the same as seen in the other 2 forms of oxybate.

So those are the options that we have available to us, and under the current circumstances, if it's possible for children to receive oxybate, then that is the medication that's going to treat the main symptoms – the sleepiness, the abnormal REM sleep phenomena such as cataplexy, the sleep paralysis, the hypnagogic hallucinations, the vivid dreaming that you mentioned, Anne Marie.

Dr. Morse:

So when looking at the landscape of treatments, it's important to recognize that we might be limited by some of the medications that may be available on label. However, we can go beyond medications. We always want to make sure we're also incorporating behavioral strategies, such as cognitive behavioral therapy for hypersomnolence disorders, as well as making sure that we're offering things like a 504 plan, or an individualized education plan, to help them be more optimal in school.

Dr. Thorpy:

Well, this has certainly been a fascinating conversation, but before we wrap up, Anne Marie, can you briefly share one take-home

message for our audience?

Dr. Morse:

I would say that it's incredibly important to recognize that sleepiness has many faces in pediatric narcolepsy, and making sure that you're doing a detailed evaluation of whether or not sleepiness is present and whether or not it's being masked by a missed diagnosis or medication that may be making it more difficult to recognize the symptoms of narcolepsy in children and adolescents.

Dr. Thorpy:

Yes, I think the most important thing is that if there's any suggestion that there's something atypical going on with a child's sleep, then it's most important that that child gets referred to a sleep specialist, and ideally a pediatric sleep specialist, who can investigate it and come to a final diagnosis.

Well, that's all the time we have today, so I want to thank our audience for listening in and thank you, Dr. Anne Marie Morse, for joining me and for sharing your valuable insights. It was great speaking with you today.

Dr. Morse:

Thank you, Dr. Thorpy. This was an incredible opportunity to discuss a very important topic, and I hope our listeners have gained some clinical pearls to utilize today.

Announcer:

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