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Pediatric Narcolepsy: A Primer for Clinicians Who Manage Children

Announcer:

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

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Dr. Brown:

Hi everyone, this is CME on ReachMD and I'm Dr. Brown. Today we're gonna be talking about management of narcolepsy in children. Now with narcolepsy you have this uncontrollable urge to sleep that is associated with specific features and really it's about the intrusion of sleep, specifically REM sleep into wakefulness. So you can have symptoms like excessive daytime sleepiness, cataplexy, hypnagogic hallucinations, as well as sleep paralysis. In addition, you tend to have fragmented sleep because there is a disruption in normal sleep architecture. The two main subtypes of narcolepsy, there's narcolepsy type one where you have excessive daytime sleepiness associated with low muscle tone, which is cataplexy. And then this has been shown to be related to low levels of hypocretin or orexin in the brain. For narcolepsy type two, you have excessive daytime sleepiness without cataplexy and many times these patients have normal levels of hypocretin. Now with hypocretin deficiency as mentioned, about 90 to 95% of these patients have as the seen in two with hypocretin deficiency.

This is seen in about 90 to 90% of narcolepsy type one and really is a as a result of destruction of the hypocretin producing neurons. There's also been some genetic susceptibility as there have been mutations in the HLA DQB oh 1 0 2 gene that has been found to contribute to development of narcolepsy. The other piece is when we talk about epidemiology of narcolepsy, it's somewhere around 0.02 to 0.06% in incidents. But we believe that this is a gross underestimation because there are so many mimics of narcolepsy, especially in children and there's often a delay in diagnosis and there's been a wide variability of that incidence based on the ethnic group as well as geographic region. So for instance, it's been shown to be lower in countries like Israel but higher in places like Japan. And this is found to be related to probably the difference in the distribution of that HLA DQB oh 1 0 2 the peak onset occurs in adolescents.

So about 50 to 65% occur about 20 years of age. There've been some studies that show that there might be a bimodal peak at about 15 years and 35 years of age. So what are the typical symptoms of narcolepsy? Excessive daytime sleepiness is a hallmark and oftentimes you have this irresistible urge to sleep, which can impact school performance as well as social interactions. Secondly, cataplexy especially in type one, which is the sudden loss of muscle tone or weakness that is triggered by strong emotions such as laughing or anger. And then you may notice that it's more subtle in children and may actually present as clumsiness or jaw dropping. Many times you often have a disrupted night's sleep and this is characterized by frequent awakenings, vivid dreams as well as restless sleep. Finally, with sleep paralysis and hypno hallucinations, you have this temporary inability to move upon waking or upon falling asleep.

And the hallucinations can either occur as you fall asleep, which is hypnagogic hallucinations or as you're waking up, which is hypnopompic hallucinations. You have some atypical symptoms in narcolepsy as well, like behavioral issues including irritability, hyperactivity and ADHD like symptoms. You might also have mental health issues in children such as anxiety dis disorders or panic attacks or depression. Weight gain is something that we've seen as well and this may seem to be unexplained. They may tend to have

these automatic behaviors and really being unaware of their tasks with poor executive functions. Now when it comes to evaluating narcolepsy, we use a detailed history and evaluation, but you also use objective measures such as a sleep diary, actigraphy to get a sense of their sleep patterns and overnight sleep study to get a sense of their sleep architecture as well as rule out any underlying medical sleep disorder like sleep apnea.

And then a multiple sleep latency test, which basically is multiple nap tests that occurs following the overnight sleep study where the patient is given five nap opportunities about two hours apart and you are really looking at how quickly they fall asleep and if they enter into REM sleep or rapid eye movement sleep. So if you have a mean sleep latency or time to sleep onset of about less than eight minutes and you have two or more sleep onset REM periods, you have your diagnosis of narcolepsy. In some situations where you have a mean sleep latency that's less than eight minutes but you don't, you have less than two or more ramp periods. We are really talking more about diagnosis such as idiopathic hypersomnia. Now this was a really micro discussion of narcolepsy in children and our time is up and I hope to see you next time.

Announcer:

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