

# A Pediatric Case Report: Three-Year-Old Girl Diagnosed with Narcolepsy with Cataplexy

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## Abstract

Narcolepsy with cataplexy is a lifelong disorder of the central nervous system characterized by excessive daytime sleepiness, cataplexy, hypnagogic or hypnopompic hallucinations, sleep paralysis and disrupted nocturnal sleep [1]. We present a case report of a three-year-old girl diagnosed with narcolepsy 7 months after symptoms surfaced, successfully treated with methylphenidate and venlafaxine.

The patient underwent a Polysomnography (PSG) followed by a multiple sleep latency test (MSLT). She underwent a brain MRI and a full montage video- electroencephalogram (EEG). Laboratory tests were performed, including human leukocyte antigen (HLA). Actigraphy was also obtained as was the modified Epworth Sleepiness Scale.

## 1. Introduction

Narcolepsy is a lifelong disorder of the central nervous system characterized by excessive daytime sleepiness, cataplexy, hypnagogic or hypnopompic hallucinations, sleep paralysis and disrupted nocturnal sleep [1]. Excessive daytime sleepiness that occurs with narcolepsy results in significant daytime impairment, mood or behavior changes, making functioning at school or work challenging. Diagnosing narcolepsy can be difficult due to the variability of symptoms. The HLA-DQB1\*06:02 gene variant has a strong relation to narcolepsy, though the absence of this gene cannot exclude narcolepsy as a diagnosis [2]. An early diagnosis of narcolepsy is essential, in order to establish early intervention, thus leading to a better-quality of life.

In general, narcolepsy with cataplexy is rare in children before the age of 5 years old. Narcolepsy may start early in age but may be more noticeable in later childhood and adolescents with symptoms peaking around 14 years of age,

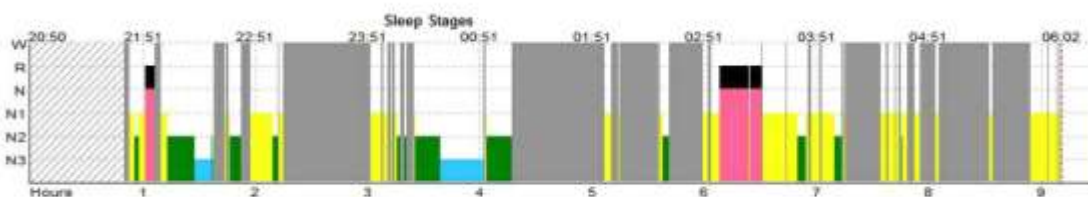
though there is often a delay in diagnosis [3]. A database review of over 1200 cases revealed the onset of symptoms of narcolepsy before the age of 5 years old is 2.1% with 1.1% exhibiting cataplexy [3].

## 2. Report of Case

A 3-year old female was referred to the pediatric sleep disorders clinic for evaluation of excessive daytime sleepiness, aggressive behaviors, sleep terrors and frequent nighttime awakenings. She is developmentally appropriate with no significant past medical history prior to the onset of symptoms 5 months ago. She experienced behavior changes and significant weight gain along with daytime somnolence. Symptoms were not preceded by influenza, influenza vaccine, strep infection and/or head injury. The patient has been difficult to arouse, extremely aggressive when awoken and with sluggish movements. Mother reports the patient sleeps around 20 hours a day. She has been known to fall asleep in school, the bathtub, grocery stores, restaurants, while eating, using the bathroom and during conversations. She experiences muscle weakness in her shoulders, neck and legs when laughing. She has fallen to the ground with laughter. There were reports of nocturnal awakenings due to extremely vivid dreams. Physical exam was remarkable for obesity without narrowing of upper airway.

The polysomnography (PSG) demonstrated no evidence of obstructive or central sleep apnea. No evidence of periodic limb movements was seen. Episodes of muscle weakness, staring spells and aggression were present during the study with no EEG abnormalities noted. The MSLT followed the overnight study showing a mean sleep latency of 6.9 minutes. Sleep was observed in 4 out of 5 naps with sleep-onset REM periods (SOREM) noted in 3 naps. Narcolepsy HLA DQB1\*0602 was positive. Modified Epworth sleepiness scale was 24 demonstrating severe excessive daytime sleepiness. Actigraphy was attempted though no data was collected due to patient noncompliance. MRI brain, routine laboratory tests and EEG revealed no abnormalities.

With these results, she was diagnosed with narcolepsy with cataplexy. Parent was educated on her diagnosis and possible treatment options. In order to minimize daytime somnolence, she was started on methylphenidate 5 mg in the morning. Venlafaxine 37.5 mg was started daily to treat cataplexy. Lifestyle modifications were discussed at length, including routine sleep/wake time, avoiding sleep deprivation and schedule short naps when most sleepy during the day in order to be more productive [1]. Safety concerns were addressed, and school accommodations were mentioned. Patient continuously improved on subsequent clinic visits, returning to optimal daytime functioning over the next year.



**Table 1**

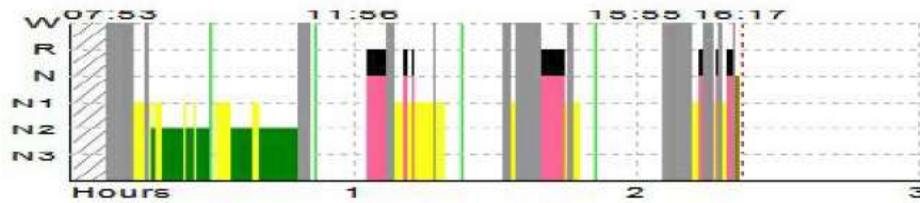


Table 2

### 3. Discussion

Diagnosing narcolepsy with cataplexy is often delayed as it is a rare disorder. Typically patients are referred to multiple specialists before embarking upon a correct diagnosis of narcolepsy. The diagnosis in children is challenging as their presenting symptoms are usually not straightforward [2]. The average time between onset of symptoms and diagnosis of narcolepsy is 10-15 years [2]. Studies have suggested that most cases of narcolepsy have symptom onset after 10 years of age and only 5% of cases showing an onset of symptoms prior to 10 years of age [2]. The American Academy of Sleep Medicine, proposes diagnostic utility of the MSLT in school age children age 5 years and up [4]. The Epworth sleepiness scale has been used in several research studies and shown validity in assessing sleepiness in children 6-16 years of age [5].

### 4. Conclusion

In conclusion, early identification and intervention is key for children with narcolepsy with cataplexy. Stimulants in combination with an anti-cataplexy agent, significantly reduces daytime somnolence, cataplexy as well as improved mood and behavior. Assuring adequate sleep opportunity and education of the care providers result in optimal outcomes.

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