

CASE REPORTS

Two sides of a coin: differential response to COVID-19 distancing measures in children with narcolepsy

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Narcolepsy is a chronic neurological sleep disorder, debuting before age 15 years in one-third of patients. Narcolepsy has a negative influence on quality of life, with daily functioning being affected by concomitant cognitive, behavioral, and social problems. In December 2019, a new coronavirus emerged worldwide, causing the severe respiratory disease COVID-19. In the Netherlands, a partial lockdown was implemented that included the closure of schools. Here we present 3 illustrative case reports that teach important lessons for the treatment of pediatric narcolepsy. We observed significant consequences of the partial lockdown measures on daytime functioning, well-being, and school performance, both negative and positive. The consequences of the lockdown led to valuable insights for further treatment, substantiating the importance of personalizing education. Involvement of specialized student counselors, flexibility in the way that education is delivered, and enhancement of environmental factors could help in guiding young patients with narcolepsy through challenges at school.

Keywords: narcolepsy, COVID-19, homeschooling, education, lockdown

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INTRODUCTION

With a prevalence of approximately 25–50 per 100000, narcolepsy is a relatively rare but chronic and debilitating sleep disorder. Narcolepsy is classically characterized by excessive daytime sleepiness, cataplexy, sleep-related hallucinations, sleep paralysis, and fragmented sleep.¹ Narcolepsy is caused by a deficiency of the hypocretin neuropeptides produced in dorsolateral hypothalamic neurons, most likely through autoimmune destruction of hypocretin cells.² Although narcolepsy has traditionally been thought of as a disease of early adulthood, at least one-third of patients with narcolepsy develop symptoms before age 15 years. The diagnostic process in children with suspected narcolepsy is complex, and these children are at increased risk of their condition being unrecognized, misinterpreted, or misdiagnosed.

In addition to the mentioned core symptoms, patients with narcolepsy very often report other symptoms and complaints, such as psychiatric symptoms, fatigue, and obesity.³ In a more functional context, patients report a perception of having a more difficult academic course load and less satisfaction at work compared with healthy control patients.⁴ In the pediatric population, these symptoms are often very prominent and clinically relevant. Specific examples include internalizing and externalizing problems, depressive symptoms, significant levels of attention-deficit hyperactivity disorder symptoms, and problems in social functioning.⁵ Although intellectual functioning seems to be within the average range (IQ scores between 90 and 110), educational problems in terms of poor attention span, cognitive underperformance, hyperactivity, and distractibility are reported by both students and teachers.⁶

The basis of narcolepsy treatment should rest on nonpharmacological interventions, often supplemented with pharmacological treatment. Nonpharmacological interventions aim to ensure a well-balanced nighttime sleep, adequate sleep hygiene habits, and planned naps during the day. More specifically, cognitive-behavioral therapy has proven its effectiveness in several studies, and psychological counseling can be helpful for both the patient and the parents.⁷ There is no general guideline on how to assist young people with narcolepsy in school, but it is evident that there is a need for some sort of guidance and individualized education plans, which allow the implementation of behavioral strategies to optimize school results.

In early December 2019, an outbreak of a pneumonia debuted in China, later identified as being caused by a new coronavirus, SARS-CoV-2. The related disease is COVID-19, which showed a rapid rise in the number of patients. The accelerated spread of COVID-19 soon garnered worldwide attention, and the World Health Organization declared the outbreak an international public health emergency on January 30, 2020. To slow the spread of the virus, governments around the world began to issue public policies, varying from social distancing measures and self-quarantine when experiencing symptoms to almost complete lockdown situations. As a consequence, millions of individuals around the world were being told to stay at home. Schools were closed, and many children and their families have had to face the challenge of homeschooling.

In March 2020, the Netherlands implemented what the government colloquially termed an “intelligent lockdown,” strongly advising people to stay at home as much as possible and keep a 1.5-meter distance from each other. Schools, colleges, and universities were shut down and needed to adapt their

way of educating young people to the new circumstances. This adjustment included less social interaction, more independent learning, and large amounts of screen time. These changes had an enormous impact on how families structured and executed activities during the day. Soon after the measures were implemented, the effects on the functioning and symptomatology of young patients with narcolepsy started to become evident in our outpatient clinic. Interestingly, we observed both negative and strikingly positive effects. Here we present 3 illustrative case reports, showing the various ways that the COVID-19 public health crisis and the associated safety measures were affecting young people with narcolepsy. These experiences also teach important general lessons for the treatment of narcolepsy, which may improve (school) functioning and ultimately the quality of life for these patients.

REPORT OF CASES

Case 1

A female patient aged 16 years diagnosed with narcolepsy type 1 contacted the sleep clinic by video consultation because of deterioration of sleepiness and concentration problems 6 weeks after homeschooling began because of COVID-19 lockdown measures. She had experienced excessive daytime sleepiness, vivid dreaming, and cataplexy since age 11 years. A diagnosis of narcolepsy type 1 was made 3 years later, based on a Multiple Sleep Latency Test (MSLT) showing a mean sleep latency of 2.8 minutes and sleep-onset rapid eye movement periods in all 5 scheduled naps. An earlier nocturnal polysomnography was normal, and actigraphy revealed sufficient sleep 2 weeks before the MSLT. There was no history of psychiatric disorders. Treatment was initiated based on lifestyle advice regarding scheduled naps and a regular sleep-wake pattern. In addition, methylphenidate 5 mg twice a day was started.

After the patient received the narcolepsy diagnosis, she adapted very well with no signs of psychological distress. Before the lockdown, she was a high school student with A grades, played tennis every week, and had a satisfying social life. Follow-up consultations by the treating physician took place every 6 months. After the lockdown, the patient received online education through Microsoft Teams, supplemented with PowerPoint presentations. This new way of teaching demanded continuous attention on the patient's part. She noticed that her ability to concentrate diminished and that she needed other students in her vicinity to remain focused. The fact that the teacher was not visible added to the patient's increased distractibility. Even with the use of methylphenidate and scheduled naps, her attention and concentration significantly deteriorated, and sleep attacks occurred during prolonged plenary online lessons. She made no changes in her sleep routine or sleep duration.

The patient's experiences during this period made her realize that social interactions during tasks, diversity in tasks, and an active and lively environment were pivotal for her to remain concentrated. Fortunately, she was then able to adjust her routine to introduce more changes in tasks, more (online) social interaction with her friends, and more physical activity. These adjustments had a clear effect, and she did not need to increase the stimulant medication. The patient also reported that this period taught her that the type of learning program will

influence her choice of future education, with a preference for group tasks and active learning programs.

Case 2

A male patient aged 9 years with a clinical diagnosis of narcolepsy type 1 had a regular follow-up visit using video consultation 6 weeks after the COVID-19 lockdown period started. His excessive daytime sleepiness became apparent at age 4 years and included falling asleep at school and clear-cut partial cataplexy of his face, neck, and upper body. He was referred to our sleep center at age 5 years. Polysomnography showed no abnormalities. The MSLT showed a mean latency of 11 minutes with 2 sleep-onset rapid eye movement periods out of 5 naps. The full pediatric evaluation showed no other cause for his sleepiness. There was no history of psychiatric disorders. Cognitive function was normal. The presenting complaints with excessive daytime sleepiness and unambiguous cataplexy, combined with the 2 sleep-onset rapid eye movement periods on the MSLT, led us to a clinical diagnosis of narcolepsy. Scheduled naps were started 4 times daily, which improved sleepiness. After some changes in medication because of adverse effects (aggressive behavior on methylphenidate and modafinil), the patient was given a combination of sodium oxybate and dexamphetamine, resulting in the adequate suppression of both sleepiness and cataplexy.

In addition to sleepiness and cataplexy, behavioral problems were the most important problem for this boy and his family. He displayed challenging behavior and emotional outbursts, both at home and at school. These issues seemed partially related to difficulties accepting his diagnosis but also to fatigue and sleepiness. The problems tended to increase during school weeks. Specialized guidance at school was started along with pedagogical home counseling for the parents to find a way to address the behavioral problems. In addition, individual psychological treatment was initiated, focusing on improving acceptance of the disease. These treatments did improve daily functioning and behavior, although the patient was not capable of attending full school weeks.

School closure as a result of COVID-19 measures yielded a strikingly positive effect on the patient's general well-being. His school provided educational material, with his parents guiding him. With this one-on-one homeschooling, frequent short breaks, and scheduled naps, the patient's school performance improved and he was able to finish all his schoolwork. His total sleep duration may have been increased slightly, with his bedtime delayed 30–60 minutes and a spontaneous awakening 60 minutes later than usual. His fatigue disappeared and did not build up during the week anymore, the number of tantrums diminished, and his mood improved. Frequent alternation of tasks, physical activity in-between tasks, and work in a quiet environment seemed to be the most important reasons for the patient's improvement. Being able to live just like his peers and not having the social burden of taking naps at school contributed as well.

It seemed that the classroom setting, with all its social and environmental challenges, had been more distressing than we previously realized: It should be noted that we learned later on that the patient was being bullied at school. The withdrawal from school and thus from the bullying clarified its impact

on his general well-being, opening the way for adjustments in his school setting. Our student support counselor, a specialist in narcolepsy, has taken the experience of the COVID-19 lockdown into account when making a personalized plan for this patient to improve both his school performance and his general well-being and behavior.

Case 3

A male patient aged 14 years diagnosed with narcolepsy type 1 had a follow-up visit through video consultation 5 weeks after the start of the COVID-19 lockdown period. He had experienced symptoms of excessive daytime sleepiness, weight gain, fragmented nighttime sleep, vivid dreams, and cataplexy since age 8 years. When he was age 9 years, he was diagnosed with narcolepsy based on an MSLT with a sleep latency of 3.7 minutes with sleep-onset rapid eye movement periods in 3 of the 5 naps. There was no history of psychiatric disorders. With modafinil 200 mg twice a day and 2 doses of 3,000 mg sodium oxybate per night, his excessive daytime sleepiness, cataplexy, and sleep fragmentation seemed to be suppressed the best. He refused to take naps at school, although he would likely have benefited from them. He only reported excessive daytime sleepiness when his coping strategies (such as changing his activities or cognitive stimulation in the form of using a Rubik's Cube, talking, and so on) were failing.

Since the transfer to high school, the patient had worked hard; nevertheless, his grades were dropping. Eventually, he changed the level of education from pre-university to higher general secondary to relieve some of the burden. In the year before the COVID-19 pandemic, the patient reported further deterioration of his excessive daytime sleepiness; in particular, staying awake during passive classes could be a big struggle. After evaluation, it was concluded that this deterioration seemed to be related to delayed bedtime. In the past year he had changed his bedtime to 10:00 PM to meet up with his peers. The patient was advised to go to bed earlier and finally agreed to go to bed 30 minutes earlier. Rising in the morning was still problematic, indicative of sleep deprivation, but he could not be convinced to go to bed earlier, because of the social impact.

After the COVID-19-related school closure, educational material was provided without further guidance. After 2 weeks, the patient's school program was conducted partially online, although only in short lessons of 30 minutes. He noticed a significant improvement in learning and mood. Two main reasons were identified. First, the patient had shifted his nighttime sleep schedule to 10:15 PM until 8:30–9:00 AM instead of 7:00 AM. He felt no need to take naps during the day, unless he had participated in strenuous exercise. Second, his increased autonomy to structure and plan his days made him more efficient. Looking back, the patient reported that before the lockdown, he had spent much of his energy on staying awake during “boring” classes. Because he could now choose his own method of studying, replacing plenary lessons with self-study together with asking questions when necessary, he had more opportunities for effective studying. The COVID-19 lockdown gave both him, his teacher, and the specialized student support counselor insights into his optimal learning strategies and a balance between optimal sleep duration, sleep timing, and

social consequences. These aspects were all considered when developing a personalized school program (as far as possible within the confines of the school) after the schools reopened.

DISCUSSION

In response to the unprecedented COVID-19 health crisis, restrictions all over the world have been implemented, with an enormous effect on people's daily lives. In the Netherlands, an “intelligent lockdown” was imposed in March 2020, which allowed people to still go outside but widely introduced working at home and the closure of schools, colleges, and universities. The 3 patients with narcolepsy described herein illustrate the significant and varying effects of this new situation on the well-being of young people with narcolepsy and provide insights into the possibilities of optimizing their situations.

Because there is no cure for narcolepsy, the goal of treatment is to reduce symptoms. Pharmacotherapy is an important element in reaching this goal, with increasing literature on this topic in the past decade. However, pharmacological treatment in children with narcolepsy is often “off-label” and is not entirely effective for all symptoms. In this context, nonpharmacological interventions, such as behavioral therapies and influencing social factors (eg, a support network), play a pivotal role in disease management.

With education shifting from a school environment to these children's homes, routines were changed, offering more space for differences in the ways that their education was shaped. In addition, being at home influenced family dynamics in either a positive or a negative way. The lockdown measures influenced their interaction with peers, with differences in individual needs and the interpretation of the rules. Our case reports illustrate how narcolepsy symptoms of young patients are affected not only by their narcolepsy treatment, but also by the changes in the environment and social interactions.

School provides a setting where children learn many basic skills in a structured way. Not only does going to school bring academic, cognitive, psychological, and social benefits, but it is also a child's right. Young patients with narcolepsy can very well blossom in this day-to-day structure, especially when the environment is safe enough to exercise a sound balance between being involved and the demands of the narcolepsy. However, attending school can also be quite challenging for young patients with narcolepsy. Studies on intelligence report pediatric patients with narcolepsy scoring within the average range (IQ, 90–110), with disharmonic, inconsistent profiles.⁶ Despite their total IQ seeming normal, research has reported these patients experiencing academic difficulties and a perceived heightened effort to achieve educational goals.^{4,8} Social interaction is another challenge that young people with narcolepsy may face during normal school attendance, with social skills being more impaired.⁹ These cognitive and social aspects may lead to additional burdens compared with those of healthy children attending school and require personalized educational programs.

The effects of the lockdown have proven quite variable and cannot be translated into a set of general advice for every patient. Whereas in the first case report, the patient needed social interactions to stay concentrated, the patient in the second case

report seemed rather distracted by them, with the busy environment probably costing him lots of energy. However, the consequences of the lockdown do make clear that the way we educate children with narcolepsy can have an enormous influence on their levels of concentration and—consequently—their school outcomes. The effect of factors such as breaks, physical activity, variation in topics, and the method of learning (eg, online/class, individual/plenary) is different for each individual and must be adjusted to one's personal needs.

Delaying school start times to decrease sleep deprivation in adolescents has been a topic of research and debate for at least a decade. Adolescents are at great risk of sleep deprivation, with just 8% reporting 9 hours of sleep a night.¹⁰ In patients with narcolepsy, enough nighttime sleep is even more important. Flexibility in school start times and thus an increase in the duration of nighttime sleep could very well result in a decrease of narcolepsy symptoms.

The consequences of the COVID-19 health crisis illustrate how major life events can change an individual's balance. The patient described in the first case report seemed to have her narcolepsy symptoms under control with medication and behavioral adaptations. There were no signs of psychological imbalance whatsoever. However, homeschooling led to an entirely new situation with reoccurrence of her narcolepsy symptoms as a result. This finding emphasizes that narcolepsy is a chronic disease and should be monitored throughout life because all kinds of changes may lead to new challenges in treatment.

Some patients have significantly improved during the lockdown. For these vulnerable children, being able to plan their own schooldays and respecting the boundaries that their illness puts on them has made them more balanced. The feeling of “not being an exception,” having the same privileges and rules as the other students, can be experienced as a relief and can be very helpful to them in coping with their conditions.

The situation in which homeschooling was not optional but mandatory made everybody realize that much more is possible than previously expected. Teachers are now trained and experienced in teaching online, which could result in a more flexible attitude when it seems that a student may benefit from online teaching. More than before, educators and physicians realize that learning is a highly personalized process and that factors that were not considered before the pandemic can affect concentration, school results, and general well-being. Because of the characteristics of narcolepsy, the learning process in young patients is vulnerable and additional guidance is indicated. Teachers should be educated about narcolepsy, and specialized student counselors should be involved in creating a personalized school program that takes cognitive, social, and environmental factors into account.

The COVID-19-related lockdown implemented in the Netherlands affected young patients with narcolepsy in various ways, both negative but also very positive in some instances. Environmental and social factors were determinative in the increase or decrease of narcolepsy symptoms; however, the effects were unique for each individual. The consequences of the lockdown led to valuable insights and substantiates the hypothesis that teachers and specialized student counselors have to be involved in adapting education to the needs of young

students with narcolepsy. This finding implies being flexible in the way the education is delivered, optimizing environmental factors, and guiding young patients through the social challenges at school.

ABBREVIATION

MSLT, Multiple Sleep Latency Test

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