

MEASURING QUALITY OF LIFE IN CHILDREN WITH NARCOLEPSY

By Regina Patrick, RPSGT, RST

The sleep disorder narcolepsy is characterized by excessive sleepiness, sleep paralysis, cataplexy (i.e., the temporary partial or total loss of skeletal muscle control, which is typically triggered by a strong emotions), vivid realistic hallucinations that occurs with sleep onset (i.e., hypnagogic hallucination) or on awakening (i.e., hypnopompic hallucination), and disturbed nocturnal sleep (e.g., difficulty staying asleep).¹ All symptoms of narcolepsy do not have to be present for a person to be diagnosed with the disorder. The impact of these symptoms on the quality of life (QOL) varies as a person with narcolepsy ages. Because of the rarity of narcolepsy (its prevalence is 0.03%–0.06%), no questionnaire has been developed that specifically measures QOL in children or adolescents with narcolepsy. Several questionnaires exist that measure the QOL in healthy children or children with other illnesses such as cancer or depression. Administering these questionnaires to children with narcolepsy reveal that these children have a lower QOL compared to healthy controls. However, the unique psychosocial issues faced by children with narcolepsy at different ages may affect how a child perceives his or her QOL. A recently developed a questionnaire, called the NarcQoL-21, shows promise in more accurately assessing QOL in children with narcolepsy.

The onset of narcolepsy symptoms usually begins at 15–25 years old. Approximately 30 percent of people with narcolepsy experience symptoms before 15 years old; 16 percent of people, before 10 years old; and 4 percent of people, before 5 years old.³ However, because symptoms of narcolepsy are often mistaken for other disorders, only 4 percent of children with narcolepsy are accurately diagnosed before the age of 15 years.³ In children, symptoms of narcolepsy are often mistaken for a neurological disorder (e.g., epilepsy) or a psychological/psychiatric disorder (e.g., depression, attention deficit-hyperactivity disorder [ADHD]). Some ways that symptoms of narcolepsy may affect a child's QOL are described below.

EXCESSIVE SLEEPINESS

Excessive sleepiness affects a child's ability to retrieve previously learned information, to pay attention, and to concentrate when learning new information. Teachers and parents may consequently view the child as "lazy." If a child is frequently caught napping, adults may perceive the napping as a psychological coping skill—for example, as a way of avoiding school work. A child may consequently be punished.

However, excessive sleepiness in children can also manifest as excessive activity for which child may receive a diagnosis of ADHD. Teachers and parents frustrated by the hyperactive behavior may punish the child.

SLEEP PARALYSIS, CATAPLEXY, AND HYPNAGOGIC/HYPNOPOMPIC HALLUCINATIONS

The symptoms sleep paralysis, cataplexy, and hypnagogic/hypnopompic hallucinations last from a few seconds to several minutes and end spontaneously. Of these three symptoms, cataplexy has the greatest impact on QOL.

SLEEP PARALYSIS

During a sleep paralysis episode, muscle atonia of rapid eye movement (REM) sleep continues to manifest just as a child is awakening or going to sleep. Other sensations that may also be experienced during an episode are numbness, tingling of the limbs, sweating, a sensation of pressure on the chest, and hallucinations. Based on a child's or parent's description of sleep paralysis symptoms, a physician may treat the child for a psychological or neurological problem.

CATAPLEXY

An episode of cataplexy occurs when the muscle atonia of REM sleep suddenly intrudes into wake when a person experiences a strong emotion. In a mild episode of cataplexy, only a few muscles such as the jaw, arm, or leg muscles suddenly weaken, which may cause the child to have a slack jaw, drop objects, or have difficulty in walking. In a severe case of cataplexy, all skeletal muscles suddenly weaken and the child may fall to the ground as if in a faint. Adults may think a child is "play-acting" on seeing a child stumbling or falling down in an apparent faint and may accordingly scold or punish the child.

Episodes of cataplexy may expose a child to ridicule by classmates or to bullying (e.g., children may intentionally induce episodes by causing the child to laugh or by frightening the child). As a result, a child may try various means to avoid going to school or being around peers. Such withdrawal can cause a child to feel isolated.



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Depending on the severity of cataplexy, a child may not be able to participate in sports, allowed to drive, or participate in school activities that may trigger episodes of cataplexy and expose a child or others to danger. Being unable to participate in activities with classmates or friends may cause the child to feel isolated.

To prevent episodes of cataplexy, a child may try to control emotions and consequently develop a flat affect. Adults may mistake the flat affect for “sullenness.”

Based on descriptions of cataplexy symptoms by a child or the child’s parents, a physician may diagnose and treat the child for a neurological problem (e.g., seizures) or a psychological problem.

HYPNAGOGIC/HYPNOPOMPIC HALLUCINATIONS

Hypnagogic/hypnopompic hallucinations result from REM sleep dream imagery occurring during wake. On describing the imagery, parents or a physician may believe the child is having delusions or some other psychological problem. If a child is frightened by the hallucinations, a parent or physician may believe that a child is simply having night terrors, nightmares, or panic attacks and not take the child seriously.

DISTURBED NOCTURNAL SLEEP

Disrupted nocturnal sleep affects memory consolidation (i.e., incorporating information into long-term memory after learning).⁴ A child with narcolepsy may consequently have trouble with schoolwork, and teachers and classmates may label the child as “not smart.”

Struggles with sleepiness remain throughout life in people with narcolepsy. However, episodes of cataplexy, hallucinations, and sleep paralysis can decrease with aging.⁵⁻⁸ This reduction may be related to the decrease in the amount of REM sleep or changes in other mechanisms of REM sleep that occur with aging.⁵

Changes in the severity of these symptoms as a child ages may affect the child’s assessment of QOL. However, assessing self-reported QOL in children with narcolepsy is difficult because of factors such as language (e.g., young children may be unable to answer or understand questions) and age.

In research studies^{9,10} of QOL in children with narcolepsy, investigators typically administer several types of questionnaires to the children and/or their parents to measure different features associated with narcolepsy such as depression, insomnia, narcolepsy symptoms, sleepiness, and ADHD. The premise of using several questionnaires is that the information collectively

derived from the questionnaires would approximate a child’s QOL more accurately than either questionnaire alone. However, the collective information provides incomplete information on a narcoleptic child’s QOL.

For example, Gregory Stores¹⁰ and colleagues administered several questionnaires to narcoleptic children (4–18 years old) and their healthy counterparts such as the Ullanlinna Narcolepsy Scale, the Strengths and Difficulties Questionnaire, the Child Depression Inventory, and the Child Health Questionnaire. The Child Health Questionnaire, which was used to measure QOL, revealed that narcoleptic children had poorer QOL than healthy children. However, the QOL was assessed by parents and not reported by the children. Hence, some issues important to children with regard to their QOL may have been missed.

Clara Inocente and colleagues similarly administered several questionnaires to children and adolescents with narcolepsy and normal controls to assess sleepiness (Adapted Epworth Sleepiness Scale and Pediatric Daytime Sleepiness Scale), depression (Children’s Depression Inventory), insomnia (Insomnia Severity Index), ADHD symptoms (Conners Parents Rating Scale-revised), narcolepsy symptoms (Cataplexy Severity Rating Score), and health (VSP-A [Vecu et Sante Percue de l’Adolescent], a French self-report health-related QOL questionnaire for teens). Inocente found that depression had a major negative impact on QOL in children with narcolepsy, and that delay in diagnosis, the presence of cataplexy, or treatment had no effect on QOL in children. However, Inocente cautions that their findings may have some bias: their study did not distinguish between the impact of different levels of severity of the disease on QOL. Thus, the narcoleptic patients may have had more severe symptoms and may not fully reflect what occurs among narcoleptic children.

A questionnaire that is commonly used to assess general health in children is the KIDSCREEN questionnaire. The KIDSCREEN questionnaire contains questions concerning a child’s health and exercise level (e.g., “How would you describe your health in general?”); feelings (e.g., “Do you enjoy your life?”); mood (e.g., “Do you feel you do everything wrong?”); leisure time (e.g., “Have you had enough time for yourself?”); family and home (e.g., “Do your mother/father understand you?”); money (e.g., “Have you had enough money to do the same as your friends?”); friends (e.g., “Do you do things with other children or young people?”); and other relationships (e.g., “Do other children or teenagers make fun of you?”); However, no question on the KIDSCREEN is specific for children with narcolepsy.

For this reason, a Swedish research team, headed by John Chaplin,² developed a questionnaire, the NarQoL-21, that is sensitive and specific in measuring the QOL of children with narcolepsy. Similar to the KIDSCREEN questionnaire, the NarQoL-21 asks a broad set of questions about a child's emotions and social interactions. It also asks questions about school and concentration. A unique feature of this questionnaire is that it asks children about their expectations for their future (e.g., "There will be good opportunities for me", "Everything will be fine for me in the future", "I think I will get a driving license", "I see obstacles for me in the future").

To develop a questionnaire with a focus on health-related QOL issues that are of most importance to children with narcolepsy, the researchers conducted semi-structured interviews with a focus group of children and adolescents (ages, 8-18 years). Their responses initially revealed seven themes—emotional support, school performance, social image, concern about the future, being limited by the condition, personal energy, and disturbed sleep—with 135 questions related to the concept of their health-related QOL. The researchers dropped questions having to do with sleep and physical problems since these issues are already covered by existing validated questionnaires or dropped questions for other reasons. After this reduction, a pilot questionnaire containing 40 questions was developed and administered to the children. Their responses on this questionnaire was subjected to further statistical analyses, after which the researchers rejected more questions. This elimination resulted in the final 21 questions (hence, "NarQoL-21").

In another aspect of their study, Chaplin and colleagues compared the KIDSCREEN-10 Mental Health Index (KIDSCREEN-10) and NarQoL-21 with regard to distinguishing between healthy individuals and patients. After subjecting the questionnaires to several statistical analyses, they found that both questionnaires had good to excellent specificity (i.e., high true-negative rate) and sensitivity (i.e., high true-positive rate). However, the NarQoL-21 was more sensitive than the KIDSCREEN-10 index in detecting children with narcolepsy.

Symptoms of childhood narcolepsy and the impact of the disease on a child's QOL are often not recognized by healthcare professionals. As a result, many children will not be diagnosed correctly until adulthood. A tool such as the NarQoL-21 could potentially provide more accurate information on the impact of the disease on a child and provide better discrimination between children with and

without narcolepsy. However, the NarQoL-21 questionnaire was developed in the Swedish language and for Swedish children. Before it can be used on a widespread level, it will need to be translated into other languages and validated in other countries. If future studies corroborate its validity, the NarQoL-21 could have a role in reducing the negative psychosocial impacts of the disease and shortening years of misdiagnosis often experienced by children with narcolepsy.

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