

# Non-pharmacological aids for patients suffering from narcolepsy with cataplexy

## *White Paper 2021*

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### **Abstract**

Narcolepsy with cataplexy is an often underestimated disorder in both its primary and secondary forms. As with epilepsy, narcolepsy and cataplexy pose a high risk of injury to the patient. Yet, narcoleptics have historically received poorer care than epileptics with regard to non-pharmacological aids. This "White Paper" informs both professionals and patients about protective measures that are advisable.

## **Narcolepsy with cataplexy - the illness**

Narcolepsy is a severe and also potentially dangerous (risk of injury) neurological disease, which, due to its drastic symptom pattern, is in no way inferior to the disease burden of epilepsy. Narcoleptics get up rested, but are dead tired again after a short time. This has nothing to do with not getting enough sleep, staying out late, or going to bed late. Narcolepsy is a purely neurological disease, a serious disorder of sleep-wake regulation, whose centers are located in the brainstem and midbrain.<sup>1-3</sup>

The primary form is thought to be an autoimmune process that leads to destruction of the hypocretin-producing brain cells in the hypothalamus. The secondary form occurs in the context of other brain-organic diseases such as Parkinson's syndrome, multiple sclerosis, H63D syndrome or post-stroke conditions. Both forms have an identical symptomatology. Infectious diseases such as streptococcal infections (PANS syndrome) or even the swine flu vaccination from 2009 have also been blamed as triggering factors. However, a precondition for the primary form is a genetic vulnerability, which is characterized by a positive HLA DQB1\*0602. The disease can occur at any age. The symptoms are very variable. Neither intelligence nor life expectancy is affected. Once the disease has occurred, it persists throughout life. Medications help alleviate symptoms in the primary form, however, a cure is not possible. Daytime sleepiness is either constant or occurs in the form of seizure-like attacks of falling asleep, usually lasting only a few minutes. At varying intervals, the drowsiness returns several times a day, sometimes barely noticeable to third parties. This rhythm is reminiscent of the approximately 4-hour sleep-wake rhythm of the newborn. If there is no possibility to sleep for a few minutes, the tendency to fall asleep increases and eventually becomes imperative. If a narcoleptic does not get his required sleep in the prodromal stage of a seizure, for example due to strong stimuli from outside, he falls into a half-sleep for short moments or absent-minded nonsensical actions are performed. A cataplexy is a short-lasting and sudden slackening of the facial, arm or leg muscles, as well as - and this is a new finding of the research of the International H63D Research Consortium - also the gastrointestinal tract. Food in the stomach is hardly transported further, and in the colon there is a massive drop in peristalsis. The body of the narcoleptic thus falls into the REM sleep episode in an apparently awake state, hallucinatory dreams included. This is referred to as "affective loss of tone." Depending on the severity of a seizure, the slackening results in slurred speech or loss of voice, double vision, head dropping, knees going soft, or complete collapse. However, consciousness is not affected in pure cataplexy.<sup>1-9,15,16</sup>

Sleep paralysis can occur during the direct transition from wakefulness to REM sleep. The affected person wants to get up, but the body is trapped in the muscle slackness of REM sleep. This phenomenon may occur upon falling asleep or upon awakening. The narcoleptic wants to wake up, but sleep won't let go. Disturbed sleep at night can be seen as a continuation of the abnormal 4-hour rest-activity rhythm. Abrupt awakening for no apparent reason is typical. The narcoleptic is wide awake and excited, doing some kind of work in the middle of the night or putting his ideas and thoughts on paper, sometimes with amazing efficiency.<sup>3-9,16</sup>

## **Non-pharmaceutical aids in narcolepsy with cataplexy**

With regard to the risk of injury, cataplexies may pose the main problem. Of course, a narcoleptic must make sure not to drive a car, avoid fire hazards, not use deep tub baths, etc.; but apart from that, the risk of injury from an imperative sleep attack is low. Cataplexy is different: spontaneous atony of muscles or whole muscle groups can lead to injuries, sometimes even very serious ones, quite similar to those of epilepsy. Nevertheless, patients with cataplexy rarely receive adequate care. Scrapes and scars are usually considered of no concern by treating physicians, even when such massive injuries are involved as broken bones, nose fractures, tooth damage, skin wounds, etc. This neglect is most likely explained by the fact that patients with narcolepsy, especially the secondary form, are boxed in as "untreatable" and "incurable."<sup>12,14-16</sup>

The International H63D Syndrome Consortium, because secondary narcolepsy can be a symptom of H63D syndrome, has - like some other researchers - compiled a list of useful medical aids. It depends on the specific case which of these utensils should be prescribed:<sup>10,15,17</sup>

- automatic switch-off devices for kitchen appliances, stove tops and the like
- avoiding bathtubs
- seats in shower cabins
- orthoses to support particularly affected joints and limbs
- epilepsy protective helmet for patients with a history of head injury due to cataplexy
- dental protection, preferably also made to measure or otherwise individually fitted
- unbreakable lenses for eyeglasses
- stable footwear
- walking stick
- bilateral crutches (if a walking stick does not provide sufficient protection)
- wheelchair, with seat position fixation (if orthoses and a protective helmet are not accepted by the patient)

Compliance with some of these tools may not be very good initially. Appropriate education and, if necessary, supportive psychotherapy can significantly improve it. Above all, the social environment must be involved, since a patient who walks freely one day and is in a wheelchair the next will otherwise appear suspect to the uninformed public. Above all, the patient's social circle must be involved, since a patient who walks freely one day and is in a wheelchair the next will otherwise appear unjustifiably suspicious to the uninformed public.

## **Conclusion**

The risk of injury from cataplexy is considerable and usually underestimated. Prescribing non-pharmacological medical aids may not eliminate but reduce injuries. By involving the social network of the patient to be cared for, both stigmatization and misunderstandings can be counteracted, which has a positive effect on the patient's compliance.

## Conflicts of interest

None.

## Ethical standards and patient's rights

This article is about scientific facts based on research literature. It is not reporting on a clinical trial, especially not a prospective one. Our research work is always conducted in accordance with the Declaration of Helsinki.

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