Medical devices that should be prescribed to patients with cataplexy to reduce their risk of injury

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Abstract

People who suffer from cataplexy have a significant risk of injury. This paper discusses (manufacturer-independent) medical devices that should be prescribed to any patient with moderate to severe cataplexy.

Cataplexy

Cataplexies are the most specific symptom of narcolepsy type 1 and therefore its most important clinical diagnostic sign. It is a brief loss of tone of the striated muscles, usually in the legs, arms, hands, and neck/facial muscles. Eye coordination and speech may also be affected. The duration of the loss of tone in cataplexy is short, usually a few seconds, and longer than one to three hours in only about 10-20% of cases. Especially the loss of internal musculature such as the vocal organs or the stomach and intestines may last longer, while symptoms in the arms, legs and head, which are most visible to the layman, last only for a very short time.[6,10] Cataplexy can affect "partially" single muscle groups or generalized the whole holding musculature (then associated with falling and its consequences). About two thirds of cataplexies are partial and hardly recognizable for laymen or inexperienced physicians. Also, many patients themselves do not necessarily immediately consider the partial episodes to be pathological, since their consciousness is not disturbed. Therefore, a physician should specifically ask about brief slackening of the facial muscles, difficulty articulating words, dropping of the head, and buckling of the knees or an unsteady gait. In children, cataplexy presents atypically, especially at the onset of the disease. The children stand out due to a hypotonic ("floppy") or ataxic gait pattern or hyperkinetic-dystonic movement patterns. Repetitive tongue protrusions, bilateral ptosis, bizarre grimacing of facial features with perioral automatisms are also typical. In contrast, the respiratory and smooth muscles are never affected. Nonspecific symptoms such as blurred vision and a feeling of suffocation may occur, as may vegetative symptoms such as tachycardia, blood pressure elevations, or sweating. A cataplectic seizure is often triggered by the biological correlates (neurotransmitters) of emotion. Prolonged episodes, i.e., longer than 30 min, are termed status cataplecticus and occur especially after abrupt discontinuation of anticataplectic medications. Despite relatively typical characteristics such as preserved consciousness, cataplexy can be confused with epileptic episodes or strokes.[1,2,4] In particular, partial cataplexies of the arms must be distinguished from bilateral arm myoclonias in juvenile myoclonus epilepsy (JME), especially since the age of onset of both disorders is similar and consciousness during myoclonias is also preserved in JME patients. Although cataplexy often occurs bilaterally, unilateral manifestations are not uncommon with up to 42% of cases. Especially at the onset of the disease, cataplexy in children presents with many positive (tonic and dystonic-dyskinetic) and negative (hypotonic) movement abnormalities, which only fade into the background as the disease progresses. Sleep paralysis and hallucinations are sometimes seen as

symptoms of narcolepsy. Sleep paralysis refers to brief episodes of complete immobility when falling asleep or waking up, often in combination with dream-like perceptions (hallucinations). However, the hallucinations can sometimes also occur in the waking state, as if the visual perception of the catapletic and a dream in the waking state were superimposed like two films. These hallucinations, in reality actually dreams, are easily distinguishable from schizophrenic hallucinations. A cataplectic knows exactly what is reality and what is dream. The schizophrenic, on the other hand, considers his inner experience to be reality. However, cataplexies and sleep paralysis share many clinical similarities. As in cataplexy, sufferers are consciously aware of their surroundings but cannot interrupt the state of paralysis by voluntary effort. Unlike cataplexy, the muscle atonia in sleep paralysis is usually complete, prolonged. In the course of the disease, a disturbance of night sleep becomes more and more prominent and is experienced as similarly distressing as excessive daytime sleepiness. Patients wake up frequently at night, sometimes for hours.Depression, impaired concentration and memory, and obesity (despite a low-calorie diet) are disproportionately clustered in narcolepsy. Other comorbid sleep disorders are commonly associated with narcolepsy, especially sleep apnea syndrome and REM sleep behavior disorders of all types.[7-8]

Medical aids in cataplexy

Given this symptomatology, it is clear that people suffering from cataplexy are at a significantly increased risk of injury. Nevertheless, unlike in the case of epilepsy, multiple sclerosis or Parkinson’s, this is not taken sufficiently seriously. Our working group has therefore, for the first time worldwide, examined and evaluated medical aids that are intended to safeguard the patient's physical integrity in the case of cataplexy. At this point, we make no claim of completeness. Nevertheless, we are publishing the evaluation already now, because every day without sufficient protection for cataplexy patients represents a medium to very high risk of injury. The order of the aids to be presented for people with cataplexy is oriented according to the potential severity of injuries caused by the failure of certain muscle groups.[1,4,5,6,8]

Knees:

Aids in the case of a dominance of cataplexies in the legs. The most important aspect in this case is to compensate for the complete loss of knee functions. Of all the assistive devices studied, knee orthoses with a range of free motion have the greatest positive effect. On the one hand, largely normal walking remains possible, on the other hand, slumping is prevented. The prerequisite is appropriately robust models that include the upper and lower legs as much as possible. These orthoses can and should be worn permanently. They are suitable for this purpose, and muscular atrophy is not a relevant risk due to the possibility of walking at a normal pace without restrictions. (Fig. 1)

Fig. 1

Arms:

Special attention should be paid to the protection of the arms and wrists, because the tool to be used here must provide two things. Firstly, to replace the muscles which become slack during a seizure, and secondly, to prevent the head and body from hitting the ground in the event of a fall. (Fig. 2)

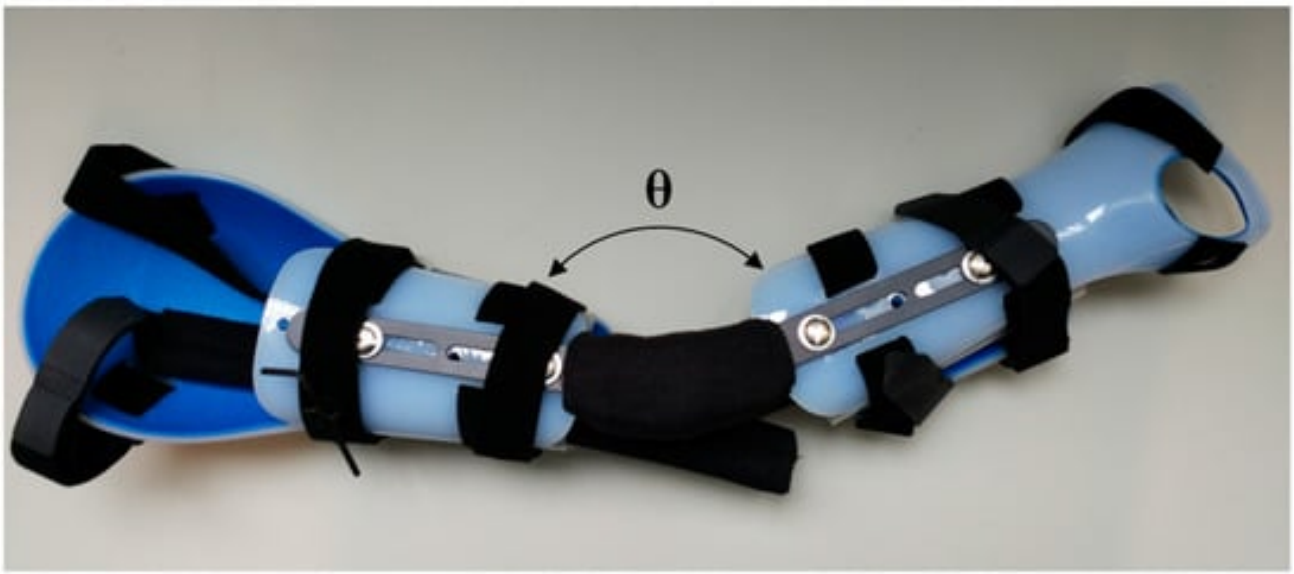


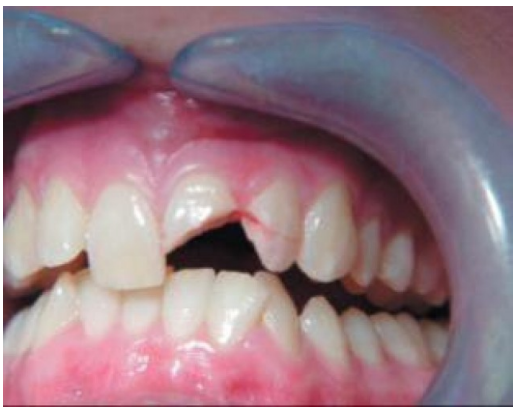
Fig. 2

On the arms, the degree of movement to be adjusted at the elbow has to be a compromise between maintaining a certain degree of movability and sufficient protection in case of falls, because the entire load of the body has to be absorbed by the orthoses to be worn on both sides without breaking in case of a fall. Therefore, only robust, medical or even custom-made orthoses can be considered. There is no question that bilateral arm and hand orthoses are a significant restriction for many

patients, especially younger ones. However, this must be accepted if one wants to avoid severe injuries such as fractures of the nose, knocked-out teeth, etc. as a result of a severe cataplectic seizure. Wearing the orthoses only temporarily on both arms is useless in severe cases, because it is impossible to predict when a cataplectic seizure will occur. Therefore, these appliances should be worn from morning until bedtime.

Teeth:

Dental protection should be provided after three falls to the face at the latest. This must be able to reliably cushion force impacts on the teeth. Therefore, the mouthpiece should be made of rather soft and flexible material (Fig. 3.2), in most severe cases combined with a brace that keeps the teeth in place. Difficulties in speaking due to a mouthguard are an unavoidable side effect and must be accepted. If this solution is unbearable for the patient, he/she should be informed in detail about the possible consequences (Fig. 3.1) for the teeth and mandible/jaw joint before discontinuing wearing a mouthguard.



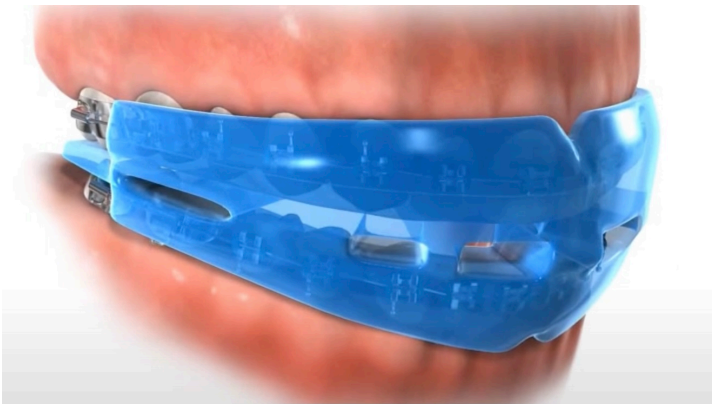


Fig. 3.1 and 3.2

Head: As with all seizure disorders, protection of the head is of paramount importance in cataplexy. A wide range of advanced protective devices is available in the form of epilepsy helmets (Fig. 4). In the case of cataplexy, the indication for this is after one severe or three mild falls with injury to the head. Cosmetic criteria should not play a role for the prescription; only the appropriate degree of protection is decisive.

Fig. 4.1 and 4.2

The range of medical safety helmets has become very broad. It ranges from classic helmets (Fig. 4.1) to lightweight helmets masquerading as baseball caps (Fig 4.2) for less vulnerable patients. Due to the particular vulnerability of the head, head protection should be prescribed even if the indication is debatable in a specific case.

Falls:

If the number of extensive cataplectic seizures is >2/month and/or injuries of any severity have already occurred, there is no way around the use of a wheelchair, even with the ability to walk being otherwise preserved, for protection reasons. There may be a psychologically explainable inhibition to make patients use a wheelchair at all times. But if falls and injury cannot be prevented by medication, prescribing wheelchair use is an absolute necessity and should never neglected due to emotional aspects. These patients can easily exercise their leg muscles in physical therapy or in a gym which will ensure their ability to walk despite being in a wheelchair at all times. The type of wheelchair to be prescribed depends on the patient's stability while sitting during a cataplectic seizure. In less severe cases a classic model (Fig. 5.2) is normally sufficient, in severe cases more protection is necessary (Fig. 5.3).

Fig. 5.1 and 5.2

Discussion

There is no question that, to date, people with cataplexy have not generally been prescribed the aids that are common in other neurological disorders. This is probably due to multifactorial reasons. For example, an epileptic seizure is perceived as more "dramatic" than a cataplectic one. This factor makes a patient with cataplexy seem less at risk, although this is often not the case. It may also play a role that there are

inhibitions about protecting people with cataplexy from injury by taking away some of their personal freedom. There is also the problem of stigmatization with some medical aids, such as head protection. Also, the idea of permanently placing a walker in a wheelchair seems counterintuitive, unless one is fully aware of the risks that cataplectic falls pose. Most physicians are more likely to accept wounds and scars in their cataplectic patients than to do the obvious and prescribe a wheelchair. With all these aids, the indication should be generous, even if the aids mean some limitation for the cataplectic patient.

Conclusion

Patients with cataplexy would benefit in terms of greater protection from injury if they were given the same protective devices as patients with other neurological disorders. Especially orthoses, helmets and/or wheelchairs should become routine in patients suffering from cataplexy.

Since the patients, especially the younger ones, are unlikely to actively ask for medical devices such as orthoses, helmets or a wheelchair, the neurologist should actively suggest these protective measures and prescribe them, better too early than too late.

Conflicts of interest

None.

Illustrations

From the manufacturers, all labels removed.

Ethical standards and patient's rights

This paper is in accordance with the Declaration of Helsinki.

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