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Narcolepsy

Narcolepsy is a disorder of which most have heard but few understand. It is relatively rare and disrupts the ability both to be awake and to sleep. In essence the brain does not control sleep or wakefulness and the result is falling asleep during the day, together with other symptoms. It is serious and disruptive of normal life but it does respond to treatment, allowing sufferers to adapt to the condition.

Simply, the disorder results in the usually irresistible urge to fall asleep during the day. It is very uncommon but can impact on those people associated with the narcoleptic individual.

It can be very damaging to work and social relationships because of the disruption it causes.

The condition may present with four principal symptoms but not all sufferers have all four:

- **Excessive Daytime Sleepiness**, which affects everyone with narcolepsy and which may be called '**sleep attacks**'.
- **Hallucinations and excessive dreaming**, which can occur at the onset of sleep or just before waking.
- **Cataplexy**, which is partial muscle weakness, which may only affect one side of the body
- **Paralysis**, Where the individual wakes and finds themselves temporarily unable to move or speak.

Narcolepsy may be classified into one of two types:

- **Type 1**, involving cataplexy and occurring in about one in five cases
- **Type 2**, does not involve cataplexy and is seen in about 4 out of 5 cases.

Incidence of Narcolepsy:

Narcolepsy is thought to affect about one in 2,500-3,000 people, with about 30,000 people experiencing the condition in the UK. The figure may be an underestimate because it is thought that some cases go unreported and it may take several years to diagnose. It is normally diagnosed in young adults, and occurs more commonly in males than females.

Features

People with cataplexy have sudden muscle weakness. It may affect the face or neck resulting, for example, in sudden jaw dropping.

Sometimes it is one side of the body and, in the most severe cases, the person may fall to the ground, which may cause injury. When they occur, the individual may not be able to speak or move. It may also be triggered by emotions such as humour-related activities such as laughing, fear or anger. In children it may be associated with facial contortions or tongue protrusion.

Other features may include:

- **Automatic movements**, moving body parts whilst asleep.
- **Amnesia** of events prior to an episode
- **Sudden outbursts** whilst asleep

The Cause of Narcolepsy.

Narcolepsy is associated with the **hypothalamus** in the brain which regulates sleep and wakefulness.

In Type 1 Narcolepsy, a chemical called **hypocretin** (also known as **orexin**) manufactured by some neurons (nerve cells) is very low or absent in type 1 individuals. It is suspected that the cause is associated with an autoimmune disorder.

In Type 2 Narcolepsy it is less clear that the disturbance is with neurons making hypocretin, although it may well be so.

Narcolepsy may be a feature of unrelated inherited conditions such as:

- Autosomal dominant cerebellar ataxia, narcolepsy and deafness
- Autosomal dominant type 2 diabetes

Other events may **trigger** narcolepsy by causing an autoimmune problem. These include:

- Pubertal or other hormonal changes
- Infections such as streptococcal throat or some viruses.

Diagnosis of Narcolepsy

The history may clearly suggest the diagnosis but it is important to eliminate other conditions which include:

- Restless leg syndrome
- Sleep apnoea (stopping breathing or breathing shallowly)
- Hypothyroidism
- Depression
- Head Injury
- Other sleep disorders

Tests for diagnosis of narcolepsy include:

- **Sleep studies**, which include the use of an EEG (electroencephalogram) which records brain waves to monitor sleep activity. It is valuable to identify the pattern and nature of sleep and also to identify periods of sleep apnoea.
- **Multiple Sleep Latency Test**, which identifies likelihood of daytime sleeping
- **Maintenance of Wakefulness Test**, assesses the likelihood of daytime ease of sleeping
- In some cases, **a lumbar puncture** may be done to measure hypocretin levels.

Treatment of Narcolepsy

The management of the condition is able to treat the symptoms but narcolepsy itself is not curable.

Narcolepsy should never be self-diagnosed or self-treated.

1. Lifestyle modifications

Once diagnosed, a review of daily routine and the activities and lifestyle may help to reduce any incidents.

Activities that make episodes more likely should be identified and addressed, and potentially

dangerous activities such as driving or swimming should be avoided.

2. Medication

There are a variety of medications which may reduce the disruption that symptoms can cause and which target daytime sleepiness and some of the other symptoms. Some drugs are not licensed for narcolepsy itself.

- **Stimulants**, also referred to as **Wakefulness medications** may be prescribed and include drugs such as modafinil and armodafinil. They may help reduce the severity or frequency of daytime sleepiness, but have side effects including headache, irritability, insomnia and nervousness.
- **Sodium oxybate** helps sleep and reduces the incidence of cataplexy. It commonly causes headaches, nausea and diarrhoea, weight loss, anxiety and bedwetting.
- **Histamine affecting drugs**, such as pitolisant, interact with histamine receptors, blocking some cellular actions
- **Antidepressants**, such as venlafaxine or fluoxetine, are used to control sudden loss of muscle control and sleep paralysis. They may lead to nausea, dry mouth, blurred vision and drowsiness.

The Outlook for Narcolepsy

The condition is not usually dangerous of itself, but sudden irresistible sleeping is disruptive. It compromises the ability to drive or use moving machinery.

Some organisations allow or provide appropriate accommodations, to allow people to follow their occupations whilst managing the constraints of narcolepsy.

For the individual, there are actions which can help to manage the condition including:

- Following a sleep schedule
- Avoiding bright lights and electrical devices, particularly around bedtime
- Avoiding alcohol, caffeine, smoking or eating too close to bedtime
- Maintaining a good level of activity
- Identifying the time when sleepiness is most apparent and organising a nap.

With care, awareness of the constraints and medical support as required, it should be possible to maintain a reasonably normal lifestyle.

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