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Chronic Fatigue in Ehlers-Danlos Syndrome Hypermobile Type

FOR NON-EXPERTS

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Persistent tiredness (chronic fatigue) contributes to poor health-related quality of life in the Ehlers-Danlos syndromes (EDS) and has overlapping symptoms with a condition called chronic fatigue syndrome (CFS). Some people with CFS likely have EDS that has not been identified. Checking for chronic fatigue in EDS needs to include a careful examination and testing. Many issues can contribute to fatigue in EDS such as: difficulty sleeping, long-term pain and the body becoming used to inactivity. While there is no single drug treatment for fatigue, many medications can help. Treatment should include checking for physical problems common in EDS, and can involve skilled therapy with an attention to methods that prevent the body becoming used to inactivity. In addition to managing symptoms, treatment of fatigue in EDS also needs to focus on maintaining existing ability, providing support, checking for new problems, and reviewing possible new treatments.

Introduction

Tiredness (fatigue) is called long-term (chronic) if it has continued for more than six months. Persistent fatigue, and its effect on activity and quality of life also describe a condition called chronic fatigue syndrome (CFS), also known as myalgic encephalomyelitis (ME). Fatigue may be a major symptom in hypermobile type Ehlers-Danlos syndrome (hEDS). Doctors can misdiagnose people with CFS that actually have hEDS, leading to treatment not being as good as it could be. There are no large high-quality trials looking at the management of fatigue in EDS. The few publications that offer advice are based on either small studies or expert opinion. While there is no specific definition for chronic fatigue in hEDS, it could be defined as: persistent or recurrent fatigue, present for more than six months, unexplained by other conditions and not the result of ongoing exertion, not substantially alleviated by rest, and resulting in a difficulty engaging in normal levels of activity.

Causes of Fatigue

Notes should be made on things that make fatigue worse or better, sleep disturbance, things that cause stress, and how the patient sees these impact on their well-being. Mental well-being should be thought of as both a cause and a result of the patient's problems. Because fatigue is such a common symptom in illnesses not related to hEDS, it is very important that all relevant information is taken and a thorough physical examination is performed.

The following causes of fatigue are common findings in hEDS: poor sleep quality, chronic pain, the body becoming used to inactivity, problems associated with standing (fainting, low blood pressure or fast heart rate), digestive system issues (being unable to take up enough nutrients from food), night time urination, anxiety and/or depression, headaches/migraines. Chronic fatigue may be caused by something else. Signs of a serious, different condition that needs attention include: weight loss, enlarged lymph nodes (felt as lumps under the skin typically in the neck, armpits, and groin), high temperature and night sweats, red swollen joints, skin color changes, and later age of onset for the condition.

Management and Care

There is no single ideal way for the assessment of patients with fatigue. Fatigue may be due to another problem, and it is the impact of that problem that is the issue. Questionnaires are one tool used to investigate fatigue but perhaps more important is the information gained from patients recording their own daily activities, their general

function, and disability; logging a list of activities. Activity logs can give a starting point for patients to set goals, and judge improvement by achieving these goals. Personal electronic devices are now available to measure activity and these can be useful for monitoring physical exertion.

Advice and Treatment

The doctor needs to work together with the patient and their carers. Engagement with the family is particularly important for young people and those with severe fatigue. The patient and their doctor should share decision making about causes, impact, and stages of management for fatigue. This might include: understanding a need to exclude other disorders from diagnosis, recognizing the reality and impact of the condition, setting realistic goals for improvement, and being prepared for setbacks. Other aspects include exploring the range of treatments and approaches available. The doctor may have a supporting role in applications for help such as financial benefits and social care, by providing medical evidence or answering assessor's questions.

Treatment is based on addressing underlying issues. These might include medications directed at different problems, as well as lifestyle changes. Doctors and patients should be aware that some people do not show meaningful responses to therapy. Patients with lengthy disabling conditions are prone to feelings of abandonment and may be vulnerable to unconventional therapies or exploitation. The long-term support and guidance of the doctor can be very important.

Maintaining Independence

Equipment and adaptations (including, if needed, a wheelchair) should be considered as these can help gain more independence, and improve quality of life. Disruption of education or work can be harmful and should be addressed early. The doctor should assist, following consent from their patient, by advising on fitness for work and education, and the adjustments or adaptations needed.

Treatment Methods

Sleep management, rest, and relaxation are key approaches. Sleep problems can make fatigue worse. Good approaches include: avoiding caffeine or nicotine close to bedtime, exercise during the day, avoiding large meals, and avoiding emotional upset/dwelling on problems before sleep. Calm music or reading may help, and one should avoid screen-based activities like watching TV. The bed should be comfortable, and

the room dark and quiet. During the day, exercise and exposure to natural light can help. Long-term pain should also be managed, as should other medical concerns that can disturb sleep such as fast heart rate, breathing problems, and anxiety disorders.

Prescribed drugs as part of pacing and rest may assist sleep. Rest periods can be introduced but a level of activity should exist that avoids overexertion. Relaxation techniques may help in the management of pain, sleep problems, and stress or anxiety. It is important that patients rest when tired, and not to “push through” periods of fatigue. Common relaxation techniques include: progressive muscle relaxation (focusing on slowly tensing and relaxing each muscle group), and visualization (imagining a peaceful setting and then focus on controlled, relaxed breathing, slowing the heart rate). Other techniques include: massage, meditation, yoga, music and/or art therapy. Graded exercise therapy (GET) and management of daily activities are important, but joint problems can limit this. Gradual exercise may be beneficial, improving physical and mental wellbeing. The main objectives in hEDS are to prevent physical deterioration without causing injury, and provide pain control. A suitably trained therapist or instructor should deliver GET. Recommendations such as “exercise more” without supported advice are not helpful and unstructured or unsupervised exercise may worsen symptoms, as can a strict increase in activity. It should be based on the current activity and goals.

Planning and reviewing activity can avoid “boom and bust” cycles. Exercise must be sustainable as it can take weeks, months, or even years to achieve goals. Symptoms can increase for a few days (e.g., stiffness and fatigue), but this is normal. Activity management can include: spreading out difficult or demanding tasks over time, and planning the day to allow for a variety of activity, rest, and sleep. Using a method called cognitive behavioral therapy (CBT) the patient can aim to sustain or improve abilities and manage the physical/emotional impact of their symptoms. An individualized program should be offered to people with fatigue, but should only be delivered by someone with proper training in CBT.

What We Need To Know

There is not enough information about fatigue in hEDS, it is also unclear how many patients diagnosed with CFS actually really have EDS. Clinical trials are needed to improve healthcare and to assess the effect of treatment. The influence of mental health on treatment is also not clear.

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