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Chronic Fatigue in Ehlers-Danlos Syndrome Hypermobile Type and Hypermobility Spectrum Disorder

FOR NON-EXPERTS

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Chronic fatigue contributes to poor health-related quality of life in the Ehlers-Danlos syndromes (EDS) and hypermobility spectrum disorders (HSD), and has overlapping symptoms with a condition called myalgic encephalomyeltis (ME) or chronic fatigue syndrome (CFS)(ME/CFS). Some people with ME/CFS likely have EDS or HSD that has not been identified. Checking for chronic fatigue in EDS and HSD needs to include a careful examination and testing. Many issues can contribute to fatigue such as: difficulty sleeping, long-term pain, and the body becoming used to inactivity. While there is no single drug treatment for fatigue,

medications can help. In addition to managing symptoms, treatment of fatigue also needs to focus on maintaining existing ability, providing support, checking for new problems, and reviewing possible new treatments.

Introduction

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)(ME/CFS). Chronic fatigue is 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. Fatigue may be a major symptom in hypermobile type Ehlers-Danlos syndrome (hEDS) and hypermobility spectrum disorder (HSD). Doctors can misdiagnose people with ME/CFS that actually have hEDS or HSD, 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 or HSD. The few publications that offer advice are based on either small studies or expert opinion.

Causes of Fatigue

Notes should be made on things that make fatigue worse or better, sleep disturbance, things that cause stress, and how the person with fatigue sees these impact on their well-being. Psychological well-being should be thought of as both a possible cause and a result of the person's health concerns. Because fatigue is such a common symptom in many illnesses, 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: 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 to assess 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. 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.

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, maintaining tolerated levels of activity 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. 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, pacing, and rest may help. 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. Physical treatments and management of daily activities are important, but joint problems can limit this. Physical therapies may help with function and daily activities, improving physical and psychological well-being. The main objectives are to prevent physical deterioration without causing injury, and provide pain control. A suitably trained therapist or instructor should advise. 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. 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

Clinical trials are needed to assess the effect of treatment and improve healthcare. The influence of psychological health on treatment is also not clear. Graded Exercise Therapy (GET) is controversial. There is evidence from surveys of people with ME/CFS that GET causes their symptoms to get worse. It is advised that each person with

fatigue works closely with their doctors and/or therapists to identify appropriate and individualized treatments and goals. These might for some people be gentle exercises that are developed over time if tolerated, and changed or discontinued if not. Each person is different.

The content of the 2017 review is being updated in 2020 and will include revisions to advice on GET based on the literature, a recent Cochrane Review update (October 2019), and the anticipated 2020 report of the UK National Institute for Health and Care and Excellence.

This article is adapted from: Hakim A, De Wandele I, O'Callaghan C, Pocinki A, Rowe P. (2017). Chronic fatigue in Ehlers-Danlos syndrome—Hypermobile type. Am J Med Genet Part C Semin Med Genet 175C:175–180. http://bit.ly/2EVvoMF