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EHLERS-DANLOS/HYPERMOBILITY SYNDROME: CAN DIET HELP WITH SYMPTOMS?

As a sufferer of Ehlers-Danlos syndrome - hypermobility type 3, Janet Dennis was finally diagnosed after 30 years of various symptoms. Both of her children have inherited the same genetic type of this disorder. In this article, Janet explains the symptoms, the risks and dietary management of Ehlers-Danlos.

Ehlers-Danlos syndrome (EDS) is a group of genetically inherited disorders of the connective tissue or collagen, which is made out of a protein that provides support to skin, tendons, ligaments, blood vessels and bones and makes up one third of all the protein used in the body. People with EDS inherit errors in their genetic makeup, possibly involving more than one gene, which makes their collagen unusually weak or fragile. Although there are now at least six types of EDS which have been identified, they have many features in common such as joint hypermobility or unstable joints, stretchy skin and tissue fragility which can cause profound biomechanical changes and instability in joints and weakness in muscles leading to pain and injury. This can lead sufferers to seek medical attention for arthritis or arthralgia and the possible requirement for a referral to a pain management clinic. Other issues that may be seen in these patients include dysmobility of the gut and oesophagus, gastroparesis, possible intestinal failure and IBS symptoms, some of which can lead to progressive disability without treatment.[1]

For full article references please email info@networkhealthgroup.co.uk

HOW COMMON IS EHLERS-DANLOS DISORDER?

The first known description for hypermobility was in the 4th century BC describing atony/hyperlaxity of the elbow and shoulder joints found in warriors from India. These warriors were unable to shoot arrows against the enemy effectively due to their lax joints and were, therefore, defeated against the enemy. Joint hypermobility and its related disorders were not fully recognised officially as a clinical condition until the 20th century.[10] It used to be thought that this was a very rare disorder, but research into this complex condition suggests that, out of a recent study of 12,800 participants, 3% had this genetic disorder, so it is not quite as rare as the medical professionals first anticipated. It is often an under-diagnosed, not understood and poorly managed condition.

SYMPTOMS

There is a range of possible symptoms seen across the spectrum of the disorder with pain being the most common symptom. Individuals with this condition may have some or many of the following:

- Joint hypermobility
- Dislocations/subluxations
- Impaired proprioception and alteration of musculoskeletal reflex
- Joint pain/fatigue
- Easy to bruise/scar
- Asthma
- Gastrointestinal symptoms/reflux/swallowing issues/food intolerance
- POTS (postural Orthostatic Tachycardia Syndrome) can cause a fast heart rate, dizziness and fainting when standing
- Mitral valve prolapse - a heart valve abnormality
- Partial or complete failure of local anaesthesia
- Varicose veins/prolapses and hernias /urinary incontinence

- Osteopenia (weakened bones)
- Curvature of the spine
- Poor wound healing
- Premature osteoarthritis
- Gum and teeth problems
- In the vascular type, the walls of blood vessels can rupture easily proving to be fatal

DIETARY MANAGEMENT OF EHLERS-DANLOS SYNDROME

Currently, there are no evidence-based guidelines for the nutritional management of a patient with EDS and symptoms vary widely. However, the involvement of nutrition and the gut is becoming more widely recognised, including poor motility, poor absorption and poor eating habits due to pain, along with the belief that dietary restriction helps with the symptoms. Further research is needed and symptom management is continuing to be updated as more information is discovered about this condition.

A patient may present with one or several of the above symptoms listed as a hospital admission or in an outpatient's clinic. Exclusion or allergy diets are frequently followed with patients eliminating wheat, dairy products and sugar, believing that this helps control fatigue as well as gut symptoms such as bloating and constipation. Currently there is no conclusive evidence to support the benefits of excluding these foods, but many sufferers of EDS report multiple food intolerances, so they would need to be advised on a well-balanced diet, restricting intake of foods if there is evidence of benefits and nutritional adequacy assessed by a dietitian. The importance of a well-balanced diet based on the Eatwell plate, including adequate amounts of calcium and vitamin D (DOH/Food Standards Agency), providing a wide range of essential nutrients, should be reinforced,[2] which should include good quality sources of protein, such as lean meat, poultry, fish, nuts and beans to provide optimum nutrients for soft tissue repair.

SPEECH AND LANGUAGE THERAPY

A patient with EDS may need to be referred to a speech and language therapist as they may have delayed speech problems from a younger age, a weaker swallow and struggle with swallowing denser foods such as apples, potatoes, bread and boiled eggs caused by weaker muscles. Support and guidance may be required with managing their swallow correctly as well as further assessment if their swallow is compromised further.

FOOD PREPARATION/COOKING

Difficulties with buying, preparing and eating food should not be underestimated with joint conditions.

The patient with EDS may find it difficult to use their hands to peel and chop and cut food, it may be difficult for them to stand or sit for long periods, which may have an adverse effect on appetite.[2] They may also be suffering from extreme fatigue, so it is essential for them to plan balanced meals, which are very simple to prepare, or pre-frozen meals are a good alternative. Advice from a dietitian can be useful on preparing easy and quick meals that are nutritional balanced. An occupational therapist may help with advice on using suitable cutlery that maybe easier to hold as well as implements and equipment, which can enable the patient to prepare more of their own food with less pain

MANAGEMENT OF POSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME (POTS)/DYSAUTONOMIA

Dysautonomia is a description of different medical conditions that cause malfunction of the autonomic nervous system. Postural Orthostatic Tachycardia syndrome, or (POTS) for short, is one of the medical conditions identified under the umbrella term of Dysautonomia. It causes a range of possible symptoms in the patient, such as low blood pressure, fainting, dizziness, palpitations and fatigue when the patient stands.

Aim of treatment is to reduce symptoms by increasing fluid volume and salt intake. Best advice for a patient is to have a drink before getting out of bed in the morning and eat small meals and avoid alcohol. Exercise has also been shown to be beneficial. Those with severe symptoms should be referred to a specialised POTS or cardiology clinic.[11]

GASTRO-INTESTINAL SYMPTOMS

Attention to hydration, exercise, sleep and regular mealtimes may need reinforcing. There may also be a need to follow current recommendations for IBS and probiotics may be useful with diarrhoea, constipation and bloating and abdominal pain. Advice may be needed for coping with reflux. Motility disorder is

very common in these patients too. The FODMAPS diet may be useful in some patients, as some food is thought to ferment if it has a slow transit through the gut. If still severely symptomatic, the patient will need referral to a GI specialist.

WEIGHT MANAGEMENT

It is important that people with EDS try to maintain a healthy weight, as any excess weight can exacerbate joint pain and stiffness and further restrict mobility and weight bearing. Exercise can be very difficult at times due to injury, dislocations, pain and fatigue. It is also important for the patient not to become underweight, as this can make maintaining muscle strength more difficult.

OMEGA-3

There may be benefits from the anti-inflammatory effects of omega-3 for painful or inflamed joints.[2] Rich natural sources include sardines, salmon, mackerel and pilchards.[3]

OSTEOPOROSIS

Some studies have examined patients with joint hypermobility and have discovered that patients with Ehlers-Danlos/hypermobility have a lower bone mineral density, as hypermobile joints increase the risk for low bone mass and fractures. In one particular study, hypermobile joints were shown to increase the risk of low bone mass by 1.8 times.[4] This could be due to a number of factors such as reluctance to stand, walk or exercise due to pain levels and injury, immobility, or due to food restriction due to gut disturbances such as chronic constipation or even coeliac disease.[8]

COELIAC DISEASE SYMPTOMS

In Italy, up to 1% of the population has been found to have coeliac disease. A recent study suggests that, in Italy, coeliac disease is 10/20 times more common in patients with confirmed EDS compared to the rest of the population. It is still unclear from this research how an autoimmune condition such as coeliac disease is apparently linked to the genetic condition of EDS. Further studies are needed to confirm the evidence in different geographical areas of the world.[6]

In clinical practice, a patient can present with chronic fatigue, bloating, constipation, nausea, diarrhoea and abdominal pain, some or all of these symptoms can be identified in EDS, as well as coeliac disease. No systematic study has yet been undertaken.[5] These preliminary studies have shown an increased rate of coeliac disease in EDS.[7] Coeliac disease testing may be useful to rule it out as a possible cause of symptoms.

LIVING WITH HYPERMOBILITY

Going to work or looking after a family can be difficult, as life gets more painful. Just everyday repetitive activities, such as walking, using a phone or a computer, sitting, going shopping and getting dressed, can become difficult to manage and cope with. The patient may refuse to participate in family activities because of pain and fatigue. In some patients, even being touched can be painful and some can prefer to sleep alone rather than risk pain and sleeplessness from sleeping with a partner. Activity can be restricted, leading to further pain, fatigue and injury and a general deconditioning of muscles and joints become apparent. A multi systemic approach is needed to deal with the management of various symptoms.

Health professionals need to recognise the impact that this degenerating, deteriorating condition has on every aspect of a patient's life, as well as on their families and help to educate, empower and support the patient. Encouraging self-management should be the aim in order for strength and independent function to be the ultimate goal. A well balanced diet, regular exercise and activity and getting enough sleep is recommended. It is recognised that periods of inactivity exacerbate symptoms, but activity needs to be controlled to prevent further pain, injury and fatigue.

Patients often complain that they have not been listened to and many have lived without a diagnosis for years. The health professional needs to provide education and support, as getting a diagnosis can be difficult and referral to a specialist can take many years. There is no cure for this condition, treatment includes managing symptoms, stabilising joints, getting appropriate support, as well as avoiding complications maintaining a healthy weight and a balanced diet.[9]

Research into this condition remains limited, but there is a charity that helps to raise awareness of the condition and give advice and support to sufferers. This is run by people who suffer from the same condition: The Hypermobility Association HMSA Helpline: 03330 116 388. Visit their website: www.hypermobility.org