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hEDS patient-management booklet

MARC-EDS Consortium

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Genetic Testing and Hypermobility

What is Ehlers-Danlos syndrome (EDS)?

Ehlers-Danlos syndromes are actually a group of 13 inherited connective tissue disorders. Some symptoms are common to all 13 subtypes, including:

- Joint hypermobility (meaning that joints can move further than expected),
- Skin hyperextensibility (meaning that skin is stretchier than usual), and
- Tissue fragility (which can look like easy bruising and poor wound healing).

What are the different types of Ehlers-Danlos syndrome?

The most common subtype of EDS is **hypermobile EDS**, which consists of generalized joint hypermobility, joint instability, and chronic pain. This type occurs in about 1 in 500 people. Although we know that this type of EDS is inherited, we **do not** know which set of genes cause this condition.

The other subtypes of EDS (such as vascular or classical EDS) are much rarer. In all of the 12 other subtypes of EDS, we **do** know which set of genes causes those conditions.

How do we diagnose the different types of Ehlers-Danlos syndrome?

It depends on what type of EDS we think you may have. If we think you have the hypermobile type of EDS, which does **not** have a confirmatory genetic or lab test, we use a set of international criteria that looks at a number of signs and symptoms, including how flexible your joints are, how soft/stretchy your skin is, and more. If we think you have one of the other, rarer types of EDS, we can perform genetic testing to identify the genetic change that may have led to your specific condition. Because we do not know which set of genes causes the hypermobile type of EDS, we generally do not perform genetic testing if we think you have the hypermobile type of EDS.

What are diseases that may look like Ehlers-Danlos syndrome, but aren't? How can we diagnose them?

There are other conditions that mimic Ehlers-Danlos syndrome; some of these include conditions like Marfan syndrome. Genetic testing is more useful when we suspect one of these rarer conditions for which the genetic basis is well-understood.

What is the role of genetic testing in people who have or are suspected to have a diagnosis of EDS?

If we suspect that you have a subtype of EDS that is **not** the hypermobile type, or if we suspect that your symptoms could be due to another, rarer condition like Marfan, then we may pursue genetic testing in order to understand what is causing your symptoms.

Possible outcomes of genetic testing include: positive, negative, or variant of uncertain significance (VUS). If the genetic testing result is positive, it means that we found a genetic change which is known to cause a particular disease. If the result is negative, it means that we have not found any genetic changes that are known to cause disease. If the result is a variant of uncertain significance, it means that we have found that you have an ambiguous genetic change. In these cases, it is not known whether the change causes disease or not. With variants of uncertain significance, there is a possibility that future research will be able to confirm whether the genetic change in question does or does not cause disease.

How is genetic testing done?

Genetic testing is performed by sending a saliva or blood sample to a lab, which will perform next-generation sequencing (a way of reading your entire DNA sequence) and return results in about 3-4 weeks. Collecting a saliva sample involves spitting into a test tube or rubbing a swab on the inside of your check, while collecting a blood sample involves going to a lab to get blood drawn. With the saliva tests, we can often send a kit to your home so that you can collect and mail the sample yourself. Once we have received the results from the lab, we will discuss the results with you either in-person or over a video or telephone visit.

What is next-generation sequencing?

Next-generation sequencing (NGS) is a way of reading DNA sequences more quickly. It reads many short segments of DNA at the same time and then figures out how those short sequences connect in order to reconstruct the entire sequence. You could think of it like this –your DNA sequence is like a book and want to copy every page in that book; one person could just go straight through the book and copy every page, but that would take a while. With NGS a thousand people each copy just a few pages of the book (in NGS, they're called "short reads") and then the computer puts all those few-page copies together at the end (called "assembling"). As you can imagine, this saves quite a lot of time.

Who pays for genetic testing?

Payment for genetic testing depends on your age, why you are receiving the testing, and what kind of insurance you have. Different insurance companies vary in their willingness to pay for genetic testing. Certain types of genetic testing are generally not covered by insurance in people older than 21. Other types of genetic testing

(usually panel tests which check a subset of genes associated with a particular disease) are more likely to be covered by insurance but will require prior authorization by your insurance company. The prior authorization process requires that we demonstrate the medical need of this testing to your insurance company. This process can take a few weeks. If your insurance does not cover a particular panel, there is the option of paying for that testing out-of-pocket; many panels cost around $250.

Are there other implications of genetic testing?

If your genetic testing is positive (meaning that it shows a change which we think is causing your symptoms), we may decide to test other members of your family to figure out if they share that particular genetic change in order to their level of risk. Additionally, a positive result may change your medical care, usually by increasing the frequency of monitoring evaluations such as echocardiograms or ophthalmology visits. Regardless of whether your genetic testing is positive or negative, we will work together with you to come up with a strategy to treat any symptoms you may be experiencing.

Are there protections against genetic discrimination?

Under the Genetic Information Nondiscrimination Act (GINA), you are protected against discrimination based on your genetic information with regards to health coverage and with regards to employment. In other words, employers and insurance providers are legally prohibited from denying you a job or insurance coverage on the basis of your genetic testing results. Importantly, GINA **does not** cover new life, disability, or long-term care insurance policies.

For more information:

https://www.genome.gov/about-genomics/policy-issues/Genetic-Discrimination#:~:text=The%20Genetic%20Information%20Nondiscrimination%20Act%20(GINA)%20of%202008%20protects%20Americans,and%20employment%20(Title%20II).

Patient-Provider Relationship

A common complaint for patients is that their concerns are dismissed, their symptoms discounted or they are simply told they need better physical and mental health routines. You are the expert in how you are feeling and what concerns you but oftentimes patients struggle with communicating these things or feel like their providers are not listening to them.

Fact: No matter how altruistic their reasons for serving in a health-related role, practitioners still need to make money. In our current system, that seeing patients quickly.

Some providers have more time allowed than others, but no provider is truly given the amount of time they would need to do their job well without the pressure of time. As someone with EDS your list of concerns probably covers at least one symptom in every body system plus navigating multiple providers and self-care processes. We will help you navigate this process and feel like your voice is heard while protecting your relationship with your practitioners using the National Patient Advocate Foundation guidelines through an EDS lens.

Step 1: Be Prepared

Have a list that you carry at all times and can amend on the fly as things come up. This list will include:

1) Things you have questions about or do not understand
2) Symptoms you are having (just list them all – most of the time we don't associate our symptoms with EDS but the likelihood is that they are associated in some way).
    a. Describe the symptom – try to use descriptions that are used in the health field to specify symptom type (See Table)
    b. When did it start?
    c. How often do you experience this?
    d. Indicate on a scale the worst it's ever been and what's it like usually – for the worst, think about what happened to trigger it on its worst day ever. Understand the pain scale.
    e. Is it constant or intermittent?
    f. List things that make it worse or better (if any)

3) Choose the items that are of biggest concern to you and review them before your appointment.
    a. Primary Care Physician (PCP) – your PCP can truly only deal with one complaint per visit especially if you are still working through how to manage it.
    b. Specialty Care Physician – your specialist can possibly handle one or two complaints. Often your symptoms are interrelated so if you have noticed a pattern even if the physical symptom differs, mention them together as a pattern.
    c. Therapists and allied health – similar to specialists they can likely only deal with two complaints at a time but again mention the patterns then indicate your top two concerns that you would like to address
4) Identify any symptom patterns—this will help make your conversation with your provider worthwhile.

Step 2: Communicate succinctly and clearly.

Make sure you have limited your list to what is manageable by your physician. Think through how you will describe the symptoms related to your concern. Have you noticed that you will describe an issue in one part of your body only to have the practitioner address a different part? This is why it's best to describe to them what is happening in conjunction with where it hurts. There are a number of ways the issues can manifest in the body and the body part with the problem may not be the root cause. Think of how you might tell this in a story as an example – **a short and sweet story** – so they can understand better what you are experiencing.
Practice telling the story in advance. Think of this as a speech class presentation. Similar to that scenario, you will be feeling anxious, especially since your EDS diagnostic odyssey has led you to many dead ends and negative experiences with practitioners. For PCPs and therapists you can "shop" around until you find someone who you have a comfortable relationships. Specialists are more rare so you will need to find a way to work with your specialist. The better your ability to lay out a clear and informative story the better. Take the emotions out of your story and maybe even vent to someone else before your appointment so you can stay focused and objective.

Step 3: Advocate for yourself and never be ashamed to speak about any issues

Research shows that women and minorities are much more likely to receive delayed care or have their symptoms dismissed by their physicians. This is often not intentional but due to implicit bias that all people have about other groups of people. Being prepared to communicate information in as direct a manner as possible will help with this. But there is a fine line between self-advocacy and becoming the "problem patient." Some key responsibilities you have as a self-advocate:

1. It is your responsibility to present your issue in a clear and informative way that makes it a compelling concern needing to be solved. Too much emotion and detail without facts will put you down the road of having concerns dismissed as a mental health issue
2. Own your issues and present anything that is of concern – don't shy away from voicing your lack of understanding of procedures, treatments, insurance and other benefits navigation. Inability to pay or financial concerns are a valid concern to address, and you should never feel unable to voice these concerns.
3. If you have done some research and would like to try a treatment path please do the following in advance of your conversation with your doctor:
    a. Find the original source of the information or other articles that have demonstrated the validity of the treatment in a non-biased setting.
    b. Make sure you understand what the research says. If you have not been trained to do this through your career ask a friend or family member to work through it with you or contact your local university to see if a faculty member would be willing to help.
    c. Keep in mind that not all information sources are equal and just because people are talking about something doesn't mean they understand it completely or it actually will amount to anything. Another thing to keep in mind is whether or not that research also applies to people with EDS who experience many symptoms that are not unique to EDS but occur for very different reasons.
    d. Present the information to your provider as follows:
        i. I am experiencing THIS. I found this article stating TREATMENT works on THIS. Can we discuss if you consider TREATMENT to be a worthwhile strategy for me?

For more hints and strategies, see:
https://www.npaf.org/blogs/how-to-talk-to-your-doctor/

Irritable Bowel Syndrome (IBS)

What is irritable bowel syndrome(IBS)?

Irritable bowel syndrome is a common gastrointestinal problem that affects patients diagnosed with Ehlers-Danlos Syndrome. It is characterized by abdominal pain or discomfort associated with altered bowel habits, diarrhea or constipation and/or bloating. There is no significant inflammation, infection or blockage in the bowel. IBS symptoms can come and go over time as well as vary in severity.

What are the symptoms of IBS?

Individuals with IBS report recurrent abdominal pain or discomfort along withone or more of the following symptoms:

- Change in bowel habits, such as diarrhea, constipation, or both
- Abdominal pain or discomfort improves after a bowel movement
- Excessive gas or bloating

The symptoms usually occur for several months and range from mild to severe.

What are the types of IBS?

Constipation-predominant IBS (IBS-C), Diarrhea-predominant IBS (IBS-D), Constipation and diarrhea or mixed symptoms (IBS-M)

What causes IBS?

The bowel is a long muscular tube that contracts (squeezes) and relaxes in an organized manner in order to move food while it is digested and absorbed and to remove waste from the body. Most of us do not feel these muscles move. When the bowel contents move too quickly diarrhea results; if they move too slowly constipation happens. If one area of the bowel squeezes and the next area fails to relax there may be a temporary spasm as contents do not move, which causes pain or cramping of the bowel. In people diagnosed with IBS, normal intestinal activity results in pain. This increased perception in IBS is called visceral hypersensitivity. Since eating causes the bowel to contract and move food, many individuals with IBS experience symptoms like urgency, cramps or diarrhea after eating.

What tests are used to diagnose IBS?

There are no specific diagnostic tests for IBS. If you have typical symptoms of IBS, your physical exam is normal and there are no warning signs then it may not be necessary to do any further testing. However, IBS can mimic other common GI disorders and further work up may be recommended.

How is IBS treated?

IBS cannot be cured and symptoms vary for individuals. Treatment involves diet, lifestyle changes, and often medicines. Add fiber to your diet to help your intestines function normally. Fiber increases intestinal motility and reduces constipation by creating a bulkier bowel movement. Fiber decreases intestinal motility in diarrhea by absorbing excess waterfrom the stool. Dietary fibers are found in complex carbohydrates, grains, fruits, vegetables, and beans, whole grain breads and cereals. Supplements such as Metamucil, Citrucel, Benefiber are natural fiber and maybe a helpful addition for fiber. *When adding fiber to your diet, do it slowly to allow your intestines to adjust.*

- Avoid fatty, greasy foods in orderto decrease production of the hormone called cholecystokinin. Cholecystokinin strongly stimulates contractions of the colon and aggravates the colon.
- Avoid gastric irritants such as caffeine, alcohol and nicotine.
- People with IBS do not produce more gas than people without IBS,but they often experience more cramping. Monitor your own response to specific foods and adjust accordingly. Some people with IBS cannot tolerate certain foods such as broccoli, cauliflower,onions, beans and cabbage. Consider a low FODMAP diet.
- Keep a food diary in order to find out which foods are easier for you to digest.
- Large meals can cause cramping and diarrhea in people with IBS. If this happens to you, try eating four or five small meals a day. Or, haveyour usual three meals, but eat less at each meal.
- Avoid swallowing excessive amounts of air by chewing and eating slowly in a relaxed environment. Avoid chewing gum, mints, and tobacco products which increase air swallowing.
- Wear loose comfortable clothing.
- Do regular exercise to strengthen abdominal muscles; stronger abdominal muscles are better ableto handle gas problems.
- Reduce stress. Stress is any stimulation that requires adaptation or change. It affects people in different ways. Recognize the cause; learn what triggers your stress; learn different coping skills (exercise, share concerns, biofeedback, meditation, and counseling).

Type of medicines available fortreatment of IBS.

Spasms or pain: hyoscyamine (Levsin),dicyclomine (Bentyl) or Enteric coated Peppermint oil capsules.

IBS-C: magnesium compounds (such as magnesium oxide, milk of magnesia); polyethylene glycol (Miralax); Lubiprostone (Amitiza); linzess (Linaclotide) or prucalopride (Motegrity)

IBS-D: loperamide (Imodium); diphenoxylate/atropine (Lomotil); alosetron (Lotronex)

What are alternative treatments?

Your provider may prescribe low doses ofantidepressants to relieve pain, anxiety and altered bowel habits. There is evidence that antibiotics and probiotics may help to reduce IBS symptoms. Cognitive behavioral therapy may help some people. Talk with your provider about the available treatment options.

Constipation

What is constipation?

Constipation is one of the most common digestive complaints in the United States affecting up to 12% of the general population. Constipation is defined as infrequent bowel movements, usually fewer than threespontaneous per week.

How does EDS affect the digestivetract system?

Connective tissue is found throughoutthe human body including the digestive tract including around the nerves of the digestive tract. Any abnormality in the connective tissues by EDS results in a disruption of the bowel function. The connective tissue of the digestive tract plays a vital rolein the ability of the muscles of the digestive tract to undergo contraction (motility) in order to expel waste from the digestive tract. This results in sluggish or slow movements of stool through the colon and/or difficulty passing stool.

What are the symptoms of constipation?

Individuals with constipation have oneor more of the following symptoms:

- Need to strain when having abowel movements
- Dry, hard or large stool thathurts when passed
- Feeling of incomplete emptyingof stool
- Need to use fingers to assist with bowel movements (manual disimpaction)
- Infrequent bowel movements
- Abdominal bloating ordiscomfort or pain

What are the complications of constipation?

Individuals can experience bloating, gas, pain, hemorrhoids, anal fissure (tear in the skin that lines the anus),rectal bleeding, rectal prolapse (rectal tissue that pushes out through the anus), stool impaction (a mass of dry stool that can’t be passed), and stool leakage.

What are ways to treat constipation?

The first treatment for constipation is to eat a diet high in roughage (fiber). Dietary fiber holds water, which keepsthe stool soft and adds bulk to stool. An intake of 20-25 grams of fiber from sources including fresh fruits and vegetables per day is recommended. Dried prunes or prune juice can be useful.

Other sources of fiber include bran, psyllium (Metamucil) and synthetic bulking agents such as Citrucel, Benefiber, Fiberal chewable tablets, and Fiber one. It is important to take the fiber supplements at mealtime with a full glass of water. This will help to restore and maintain regularity. Add fiber slowly to your diet so your digestive system can adjust to the increased fiber. If you need to supplement fiber, begin by taking 1 or 2 rounded teaspoons of powder 1-2 times a day with meals. These supplements are very safe. If you are not getting the desired results after 5 days then increase the daily dose.

What are some steps to take everyday?

- Establish regular daily bowel habits; don't ignore the urge to have a bowel movement. Allow15 minutes after breakfast or after your main meal of the dayto sit on the toilet. Do not strain.
- You may find that elevating your feet on a footstool or "squatty potty" in front of the toilet helpful or bending forwardhelps stool to pass.
- Daily exercise can stimulate themuscles of the colon to contract.
- Constipation often occurs duringtravel or vacation or in stressfulsituations, so pay special attention to your bowel regimenduring this time.
- Avoid routine use of laxativesthat contain cascara, senna, and castor unless a medical provider has recommended them. Prolong use can be habit-forming and may damage your intestine.
- Over the counter laxatives or stool softeners such as milk ofmagnesia, magnesium oxide, sodium docusate (Colace) andpolyethylene-glycol (Miralax) are safe and effective options for occasional or mild constipation.
- Enemas or suppositories canalso be used as needed.

What are other available treatment options?

There are several medications available to treat constipation that promote better bowel movements such as lubiprostone (Amitiza), linaclotide (Linzess). There are also tests that can be performed to furtherevaluate the cause of your constipation.

- Probiotics have also been shown to be helpful with treating constipation and symptoms of abdominal bloating. There are various probiotics available on the market. When choosing a probiotic, look for one that contains bifidobacterium and lactobacillus.

Abdominal Bloating

Abdominal bloating is a common bothersome symptom that is often more troublesome than abdominal pain. Symptoms typically worsen as the day progresses, especially after eating a meal and tend to improve overnight. Bloating can be associated with functional gastrointestinal disorders or organic diseases.

The pathophysiology of bloating remains unclear but there is some evidence suggesting that gut hypersensitivity, impaired gas handling, altered gut microbiota, food intolerances, and abnormal abdominal-phrenic reflexes may contribute to bloating.

What is bloating and distention?

Bloating refers to a subjective sensation of excessive gas and/or flatulence, fullness, abdominal hardness or tightness, or the feeling of abdominal inflation or swelling. This occurs in response to a buildup of gas or food contents within the digestive tract leading to an upper body fullness feeling known as bloating.

Distention is an actual increase in size of one's waist (girth).

What are potential causes of bloating?

- Constipation
- Problems with how the intestinal muscles are working
- Gastroesophageal reflux disease
- Inflammatory bowel disease such as Crohn's or ulcerative colitis
- Peptic ulcer disease
- Functional dyspepsia
- Irritable bowel syndrome (IBS)
- Lactose intolerance or other food intolerances
- Overeating
- Gastroparesis
- Small Intestinal Bacterial Overgrowth (SIBO)
- Medicines or foods that contain lactulose or sorbitol
- Celiac disease
- Dumping syndrome
- More serious disorders: ovarian cancer, ascites or tumors
- Pancreatic insufficiency

How is bloating diagnosed?

Performing tests in patients with bloating can be challenging since tests often don't show any abnormalities, but some of the commonly obtained tests are:

- Abdominal x-rays
- Endoscopy
- Ultrasound
- CT scan of abdomen and pelvis
- Gastric emptying tests
- Hydrogen breath test

What treatment options are available for abdominal bloating?

First line of treatment is dietary changes. Research has shown that a FODMAP diet, which is low in fermentable oligosaccharides, disaccharides, monosaccharides and polyols can reduce the symptoms of gas. The small intestine does not always fully absorb these carbohydrates, so they can be passed along into the colon where they are fermented by bacteria resulting in gas production. A low FODMAP diet avoids fermentable, gas producing food ingredients such as:

- Oligosaccharides, which are found in wheat, onions, garlic, legumes and beans
- Monosaccharides, including fructose (a type of sugar found in honey), apples and pears
- Disaccharides, such as lactose in milk, yogurt and ice cream
- Polyols or sugar alcohols found in foods such as apricots, nectarines, plums and cauliflower, as well as many chewing gums and candy

Other treatment options include:

- Probiotics that contain bifidobacterium infantis and lactobacillus acidophilus
- Enteric coated peppermint oil capsules
- Simethicone pills or liquid, activated charcoal
- Consider an empiric trial of antibiotics such as rifaximin or perform hydrogen breath test
- Treatment for constipation (lubiprostone or linaclotide)
- Antidepressants such as amitriptyline, nortriptyline, duloxetine
- Prokinetics such as metoclopramide, domperidone
- Avoid eating greasy/fatty foods
- Avoid chewing gum or drinking carbonated drinks
- Avoid eating too quickly
- Fiber supplements such as psyllium, methylcellulose or 100% bran can exacerbate symptoms

Pain scale	1 – 3 You have pain but you can ignore it and it doesn't disrupt your able to do the things you normally do 4-6 Moderate pain, you could ignore it somewhat but it keeps reminding you that it's there – this is when you would consider taking an as-needed pain relief approach 7-10 Pain the prevents you from doing the things you normally do the higher up you go the more debilitating (10 typically results in other physical symptoms like vomiting, crying, inability to move)	
Pain and physical Symptoms	• Aching • Cramping • Fearful • Gnawing • Heavy • Hot or burning • Sharp • Shooting • Sickening • Splitting • Stabbing • Punishing or cruel • Tender • Throbbing • Tiring or exhausting • Pain or tenderness. • Redness or bruising. • Limited motion. • Muscle spasms. • Swelling. • Muscle weakness. • soreness, cramping, aching, stiffness or burning	Description of symptom
GI Symptoms		
Autonomic Symptoms	Dizziness Tunnel vision/fainting Tachycardia Urine leakage vs loss of bladder control Dyspnea Body temp regulation	
Everywhere	How to describe widespread…	

Gastroparesis

What is gastroparesis?

The word gastroparesis means paralysis of the muscles in the stomach. Gastroparesis is a disorder when food empties slowly from the stomach,resulting in bothersome symptoms that interfere with daily life. Gastroparesis may occur when the nerves to the stomach are either damaged or not working properly. There are other conditions that can cause similar symptoms, so careful evaluation must be done before making the diagnosis.

What causes gastroparesis in EDS?

Connective tissue is present throughout the body including the digestive tract. It must stretch and be squeezed in order for food to move through and be digested. It has been proposed that abnormalities in the connective tissue in the digestive tract might alter how the digestive tract moves. Further research is needed to better understand the role of connective tissue within the digestive system. It is thought that autonomic dysfunction may also be a contributing factor to upper digestive tract symptoms in EDS. **Other causes of gastroparesis are**: diabetes, idiopathic, post-surgery (reflux/fundoplication, ulcer or hiatal hernia repair, bariatric surgery), scleroderma, systemic lupus erythematosus, Addison's disease, severe hypothyroidism, amyloidosis, Parkinson's disease, viral illness or paraneoplastic, medications such as narcotics/opioids and some antidepressants.

What are the symptoms of gastroparesis?

- Feeling of fullness after eating a small amount of food
- Nausea
- Vomiting
- Abdominal bloating
- Vague stomach pain
- Weight loss due to a decreased appetite

What are the complications of gastroparesis?

- Dehydration
- Weight loss
- Malnutrition
- Undigested food that hardens and remains in your stomach (bezoar)
- Decreased quality of life

What are the usual ways to treat symptoms of gastroparesis?

- Dietary changes: eat small meals up to six times a day of low fat and low fiber, rather than three large meals. Liquids are often better emptied than solid food. Forsome patients switching to liquid, pureed diet and taking a multivitamin may help.
- Avoid medications that slow stomach emptying such as **opioids**.
- Promotility agents are medications prescribed to help the stomach to empty more quickly such as **metoclopramide** (Reglan) a promotility agent, **Erythromycin** is an antibiotic that can speed up stomach emptying. Another promotility agent, **Domperidone**, is used in several countries outside of the UnitedStates, which is not approved by the FDA in the US but can be obtained by special arrangements with the FDA.

Medications used for the treatment of nausea and vomiting.

These medications are called antiemetic agents such as **ondansetron** (Zofran), **prochlorperazine** (Compazine) and **trimethobenzamide** (Tigan). These canbe used on an as-needed basis.

What other options are available if treatments fail?

If the dietary modifications, useof promotility and antiemetics agents not effective in treating gastroparesis then there are other options, which would be further discussed with a gastroenterologist.

POTS and Hypermobility

What is Postural Orthostatic Tachycardia Syndrome (POTS)?

POTS is a type of dysautonomia, or autonomic nervous system dysfunction, in which the heart rises dramatically with a positional change – such as moving from laying or sitting to standing upright. Along with heart racing (tachycardia) many other symptoms are observed (described below).

How is POTS diagnosed?

POTS is diagnosed according to an increase in heart rate with positional change. One form of autonomic testing is a tilt table test. Here, vitals such as heart rate and blood pressure are measured as a person is strapped to a table that moves from a lying flat position to 60 degrees head up. Additional tests may include QSART, or sweat output at four different locations, and response to Valsalva, or breathing out against resistance for 15 seconds. Testing may also be done simply with an "active stand test" where vitals are taken from laying at rest and again in standing at two-minute intervals. For a POTS diagnosis, the heart rate must increase by 30 bpm in adults (40 bpm in adolescents) in the absence of sustained orthostatic hypotension (prolonged drop in blood pressure).

What are the subtypes of POTS?

Neuropathic	Hypovolemic	Hyperadrenergic
Impairment in nerves controlling return of blood to the heart.	Low blood volume, or the amount of blood in the body	Elevated levels of norepinephrine (aka: adrenaline) in upright position

Note: Individuals may have more than one subtype. All subtypes of POTS lead to an increased "fight or flight" response of the body, or activation of the sympathetic nervous system, which leads to heart racing.

What are the symptoms of POTS?

POTS includes both symptoms that coincide with positional changes (orthostatic) and symptoms that may occur in both upright and laying down positions (non-orthostatic).

Primarily Orthostatic	Not primarily orthostatic	
Tachycardia Lightheadedness Palpitations Syncope or near-syncope (note: only about 1/3 of POTS patients fully "pass-out")	Fatigue Cognitive dysfunction (such as brain fog) Headache Visual symptoms Anxiety Nausea	Chest pain Gastrointestinal symptoms Sleep disturbance Dizziness Leg weakness Shortness of breath

Why do people with hEDS or hypermobility have POTS?

POTS and autonomic dysfunction is more common in hypermobility but the cause is not fully understood. One potential proposed cause is the connective tissue differences in hypermobility may lead to changes in blood vessels and impaired blood return to the heart. This may contribute to excessive blood pooling in the lower body and more sympathetic nervous system activation.

What may make POTS symptoms worse?

- Exercise or overexertion
- Heat
- Valsalva maneuvers (hold breath and strain)
- Dehydration
- Large meals
- Abrupt postural changes
- Prolonged standing
- Prolonged bedrest
- Caffeine
- Alcohol
- Traveling to high altitudes
- Infection
- Menses
- Stress

What are treatment options for POTS?

• Discontinue medications that make symptoms worse (e.g., beta blockers, tricyclic antidepressants, diuretics, etc.) • Increase sodium to individualized amount • Increase fluid to appropriate level • Increase specific movements and exercises. Suggestions include strengthening and cardiovascular activities while laying down, reclined, or in seated positions and gradually building to an upright position.	• Compression garments, especially lower body and abdominal • Avoid exacerbating factors (see above) • Medications • Nervous system regulating activities • Breathing practices • Mindfulness and meditation practices • Energy conservation and pacing techniques • Sleep hygiene • Nutrition and dietary interventions

Resources:

Dysautonomia International
Dysautonomia Support Network
Exercise Guideline:

http://www.dysautonomiainternational.org/pdf/CHOP_Modified_Dallas_POTS_Exercise_Program.pdf

Joint Dislocations, Subluxations and Instability

What are joint dislocations and subluxations?

People with hypermobility have joints that move beyond a typical range of motion. This is called 'hypermobility'. Sometimes it results from having ligaments (the structures that link bones to other bones) that are too loose, which is called 'ligamentous laxity'. Sometimes, it might be because tendons, which link bones to muscle, are loose or because the muscles themselves are weak (muscular hypotonia). Occasionally, a person's bones are made differently (called 'dysplasia') so that all the right parts are not in the right places. Any of these problems can lead a joint to be unstable, or to move beyond a typical range of motion. This often leads to 'subluxations', which is when the bones of a joint stay generally in its appropriate area, but are not quite where it belongs. Sometimes, though, it leads the bones of a joint to move entirely out of a normal location, called a 'dislocation'.

What happens if a joint dislocates or subluxes?

If the bones in a joint separate entirely, this will be visible from the outside and generally causes intense pain and limitation of movement, when then leads to swelling and signs of tissue damage in the joint. When a joint is dislocated, especially the first time it happens, an expert usually needs to put it back into place.

Subluxations may not be visible from outside the body and cause pain that is less intense (but can still be significant). Subluxations will often return to the correct location on their own or with minor manipulations.

What tests are used to diagnose joint instability?

There are no specific diagnostic tests for unstable joints and most of the time an unstable joint is evident after talking with and examining a person. Sometimes imaging like X-rays are used to identify a bony dysplasia that puts a person at risk for instability. X-rays can also identify bony damage or MRI can find tendon or muscle swelling and damage that can occur after a traumatic dislocation.

What are the treatments of joint dislocations and subluxations?

Initial and emergency treatment includes the RICE protocol (rest, ice, compression and elevation). Anti-inflammatory medications (like ibuprofen or naproxen) can also be used to treat pain and swelling. Occasionally, the joint may return to the normal location on its own. More often, an expert will have to manually reposition the joint. Generally, the joint will then be immobilized and RICE will be recommended to help the joint to heal. Sometimes surgery may be helpful immediately or if the joint continues to dislocate or to remain unstable.

Subluxations will generally return to their normal position with rest or minor manipulation. RICE can be helpful here, as well.

How can dislocations and subluxations be prevented?

People with hypermobile joints may have unstable joints that move even without accidents or trauma. So, preventing dislocations and subluxations may be impossible. However, maintaining good body positioning and good muscle tone can help to stabilize the joints so they have less chance of moving out of position. Physical therapists can help to identify muscle weakness that puts someone at risk of instability and can prescribe exercises to improve that weakness. There are some movements that are also highly likely to cause problems—for example squatting is particularly challenging for unstable knee caps—and these should be avoided. If a joint has become particularly unstable, then a brace can be used when doing an activity that is very likely to cause it to move out of position. Sometimes surgery is needed to stabilize a joint.

Resources:

For more information about specific conditions, see:
www.orthoinfo.org

EDS and Physical Therapy

What is Physical Therapy?

Physical Therapists are trained in the study of human movement. Evaluation and treatment techniques are used to preserve, enhance, and/or restore movement and physical function. Physical Therapy strategies are taught to preserve and protect joints during activities and with the use of correct ergonomics and body mechanics.

How does EDS affect the musculoskeletal system?

Connective tissue is found throughout the human body including the ligaments, tendons, muscles, and joints. This can result in symptoms that are isolated to certain joints or occur more globally throughout the body.

Symptoms include:

- Joint subluxations
- Joint dislocations
- Ligament sprains
- Muscle strains and tears
- Joint instability
- Low energy level or muscle fatigue
- Feelings of muscle tightness or spasm
- Poor balance or clumsiness
- General fatigue

What are the complications if symptoms are not addressed?

Individuals can often experience increasing number of injuries. Injuries can be both acute (new) or chronic (repeated). If not addressed, this typically leads to gradual reduction in the number of activities the individual participates in. The decreased activity leads to more deconditioning which results in decreased endurance (causing more fatigue), worsening balance (causing more falls and injuries), worsening coordination, and general decrease in quality of life. The limitations typically impact both work and home life.

What are ways to treat joint pain, instability, and deconditioning?

Physical Therapy is best used on an individual basis to provide guidance tailored to each patient's specific needs. Pain prevention can be optimized by practicing good posture and computer ergonomics, wearing supportive shoes with good arch support, and avoiding injurious activities. What are some steps to take everyday?

- Take time to improve set up of every-day activities; position high-use items to avoid reaching overhead or low to the ground.
- Use cushions to help support posture and to decrease stress and the work of the body in holding sustained positions.
- Find an enjoyable way to regularly exercise. Exercise that is repeatable and allowing consistency is more important than an activity an individual can only do occasionally.
    - Core and balance work are very important.
        - A Physical Therapist will advise on exercises/techniques to stabilize joints and work towards a home program that meets the patient's needs and is doable and repeatable.
        - Pilates exercises can work well. Look for workouts or practitioners referencing experience or a program for hypermobility when able.
            - Typically safest to do with an experienced practitioner first to correct form.
        - Yoga is cautionary for this population. There are versions of Yoga for bendy bodies which, for some not all individuals, can be safely performed. Look for workouts or practitioners referencing experience or a program for hypermobility when able.
            - Typically safest to do with an experienced practitioner first to correct form.
            - Focus should be stability and slow movements versus stretching or flexibility.
- Stretching should generally focus on moving the body through normal ranges in a slow and controlled manner instead of lengthening tissues further, unless a Physical Therapist has recommended otherwise.
- Options, other than stretching, to address the sensation of tightness:
    - Massage
    - Stick rollers
    - Massage guns
    - TENS units

Resources:

www.ehlers-danlos.org/information/physical-therapy-for-hypermobility/
www.ehlers-danlos.com/2017-eds-classification-non-experts/evidence-based/rationale-physical-therapy/treatment

EDS and Occupational Therapy

What is Occupational Therapy?

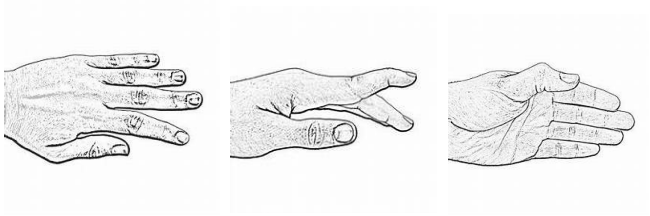
Occupational Therapists focus on the things you want and need to do in your daily life. OT intervention uses daily activities (occupations) to allow participation in the important activities to improve quality of life. Occupational therapy strategies are aimed at problem solving and adapting so clients can fulfil their daily routines, roles and goals.

How does EDS affect daily routines, work, leisure and social participation?

- General fatigue – difficulty with completing basic (bathing, dressing) and instrumental (cleaning, cooking) daily tasks
- Joint subluxations, dislocations, instability – difficulty with writing, typing, music instruments, gaming, lifting and/or carrying objects, difficulty with extended periods of sitting or standing
- Hand pain – difficulty with cooking, opening jars/bottles, household chores and activities such as yard work, gardening

What are some common presentations seen with EDS?

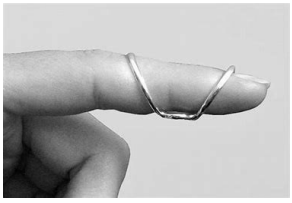
- Hyperextension tendency/deformity is the extension of the joint beyond the neutral position.
    - In fingers – may result in a painful joint, decreased power when pinching and a delay in being able to bend the finger/hold a sustained posture.



    - In wrist – may make weight bearing through open hand position painful.
    - In elbows – may result in painful elbow or locking when lifting/carrying heavy objects

What are ways to address hand pain, joint laxity and instability?

Orthoses – wrist and hand/finger orthoses (soft wrist wrap, wrist brace, wrist widget, custom made thumb splints/orthosis, oval 8 splints, silver ring splints, buddy loops/taping, elbow compression sleeves, compression gloves)




Isometric exercises - exercises involving muscular contractions against stationary resistance (without movement of the involved part of the body). Short arc strengthening exercises are exercises working on last few degrees of range of motion against resistance.

Proprioception - conscious and unconscious awareness of sense of movement and position of body. Educating client on safe and optimal position of joints when lifting/carrying objects or grasping or pushing down on surfaces.

What are some steps to make daily routines easier?

Joint protection techniques – use larger stronger joints instead of smaller joints, when lifting or carrying objects hold items close to your body. Example, avoid holding onto a book, instead support book on a book rest or prop it up on a cushion.

Adaptive devices – good grip utensils, foam tubes to build up handles of spatula, tooth brushes or pencils, spring loaded scissors, right angled knives or gardening tools





Energy conservation – plan your calendar on a weekly basis and chart out days with moderate amount of activity such as doctor/therapy appointments, grocery store – can be color coded – similar to setting your pill box for the week. Then add rest days on days following activity days to build in time for recovery. On activity days, try to keep your daily routine short/manageable (example avoid shampooing and styling hair on the day of appointments/travel, have prepared meal/prepackaged meal on moderate activity days).

Energy Budgeting and Hypermobility

Energy and Hypermobility

Fatigue, or an overwhelming sense of exhaustion, is one of the most debilitating symptoms in EDS. Most people with EDS report significant fatigue and those with POTS or MCAS in addition to EDS may be even more likely to report fatigue. Fatigue is different from drowsiness, in that drowsiness is primarily feeling the need to sleep, whereas fatigue may involve overall energy and even affect motivation. Some describe it as feeling "weighed down" or like they are lifting a ton of bricks when trying to move arms or legs.

What might affect fatigue (both increasing and decreasing)?

- Physical activity, muscle strength (or lack thereof) and potential fears associated with movement
- Sleep disturbances
- Self-efficacy concerning fatigue, or a sense of ability to do certain things
- Pain severity
- Participation in community and social interactions
- Breathing dysfunction
- Sensory processing including proprioception or body awareness
- A person's beliefs about their joint hypermobility and potential limitations
- Orthostatic intolerance (dizziness and symptoms with positional changes)
- Satisfaction with treatment options and medical management
- Anxiety or depression
- Chiari malformation
- Managing chronic illness and demands associated

How does fatigue affect daily function?

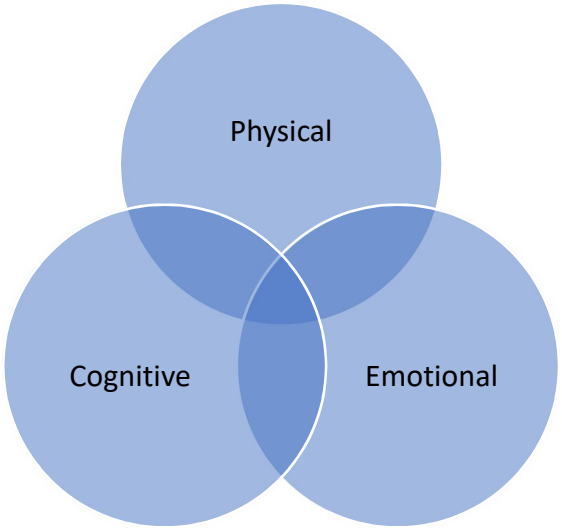
Fatigue has potential to impact psychological distress such as irritability, depression, and frustration; difficulty with concentration, decision-making, reaction speed, memory, and productivity; challenges with communication; appetite; and overall task performance. This in turn affects ability to work or go to school, social participation, completing hygiene and self-care, mobility, running errands, completing house or yard work, driving, engaging in hobbies, and potentially even sleep, despite overall feeling exhausted.

How do you manage fatigue?

- Identify factors that may contribute to fatigue and learn early-warning signs
- Manage available energy through compensation and adaptation
- Implement pacing while introducing new activities using modified approach

What are the types of energy?

Physical	e.g., walking, lifting, bending, reaching, climbing, standing
Cognitive	e.g., thinking, sensing. This may include sensory overload, social interactions, and demands on your brain for processing information.
Emotional	e.g., feeling. Note: this includes extremes of emotions including "positive" types of feelings such as excitement, as these can also be depleting.



What is energy budgeting?

There are many terms used in the world of fatigue management and working to increase function through saving energy.

Energy conservation: saving energy through modifying the way that you complete a task or the routine in which you conduct daily activities. This may include using assistive devices such as a shower chair to sit for bathing or a long-handled item to avoid reaching or bending. It may also mean considering ergonomics in the kitchen to decrease the load on joints or asking for help and delegating certain tasks to others.

Pacing: it is highly recommended that you plan your day and week strategically to minimize wasting energy. For example, break up larger tasks into smaller, component parts and spread them out throughout the day. Other examples include: stop to rest before you feel tired, remembering your three types of energy and considering all in your total expenditure, alternate using upper and lower body muscles to give the other muscles a break, chopping vegetables for a dinner recipe in the morning and assembling in the afternoon. In the case of individuals with significant fatigue, this may include using a mobility device to enable them to participate in activities that require more endurance or taking breaks by sitting/laying down to enable performance of upright tasks. Some refer to this as managing their "spoons" (See: Spoon Theory)

Who can help?

Consider working with a professional to maximize energy levels. Individuals such as occupational therapists, psychologists, physical therapists, exercise physiologists, and health coaches may all be appropriate.

Resources:

Spoon theory: www.butyoudontlooksick.com

The MTHFR gene

What is the MTHFR gene?

The MTHFR gene provides instructions for making an enzyme called methylenetetrahydrofolate reductase. This enzyme plays a role in processing amino acids, the building blocks of proteins. Methylenetetrahydrofolate reductase is important for a chemical reaction involving the vitamin folate (Vitamin B9) and uses riboflavin (Vitamin B12) as a cofactor.

Why is MTHFR important in individuals with EDS?

Everyone has two copies of the *MTHFR* gene, one copy from each parent. There are two common variants in the *MTHFR* gene sequence, called C677T and A1298C. About 1 in 3 people have at least one *MTHFR* variant and 1 in 10 people have variants in both copies of the *MTHFR* gene. In some populations, more than half of individuals have at least one of these common variants.

A person with two copies of the common C677T *MTHFR* variant can have slightly decreased levels of folate and a slightly higher homocysteine level than someone without any C677T copies. It is less clear whether having the A1298C variant affects folate and homocysteine levels, but it seems to have less impact than the C677T variant.

What are the known associations between MTHFR and health?

Whether there is a relationship between the common *MTHFR* variants and many medical problems has been questioned for decades. Many online communities and websites claim that the two common variants in the *MTHFR* gene may cause different health issues including autism, glaucoma, and hypothyroidism, among many other conditions. Also, products are marketed that claim to correct deficiencies caused by having one of the common MTHFR variants. Most often these products are not developed based on scientific evidence and those marketing them prey on patients to make a profit.

There are hundreds of published articles in scientific literature exploring the relationship between the common variants in *MTHFR* and associations with many conditions. Previously, some small but statistically significant connections have been made between *MTHFR* polymorphisms and neural tube defects, stroke, serious mental illness, heart disease risk, and many other conditions. However, later studies have not confirmed these relationships.

The GWAS (Genome Wide Association Study) Catalog catalogs genetic association studies that have used proven analysis methods. As of writing this section, the following are validated associations between the C677T variant and medical conditions: slight changes in folic acid, homocysteine, glycine, serine, mean reticulocyte volume (red blood cell size), and glomerular filtration rate (kidney function) measures; increased risk of multiple sclerosis schizophrenia; differences in age at menopause and BMI-adjusted hip circumference. The A1298C variant has only been associated with a change in BMI-adjusted hip circumference.
There may be more associations in the future as more population genetic data is collected, especially associations relevant to individuals of currently underrepresented ancestries.

Should I have genetic testing for the common MTHFR variants?

In 2013, the American College of Medical Genetics and Genomics (ACMG) and the American College of Obstetricians and Gynecologists (ACOG) advised against routinely testing for *MTHFR* variants. Since then, more professional medical groups have made the same recommendation. Despite this, testing is still widely available. Therefore, if someone chooses to have testing for the common *MTHFR* variants, they should interpret the health implications of those findings cautiously.

What are available treatment options?

If someone is known or suspected to have a common MTHFR variant, a healthcare provider may check folic acid, Vitamin B12/MMA, and homocysteine levels. If any of these are abnormal, then supplementation and continued monitoring may be recommended. This should be done by healthcare provider who is knowledgeable about the patient's history and specific deficiency.
Conditions associated with MTHFR are generally thought to be avoided if a person's diet includes enough folic acid, which is found in many fruits and vegetables.

Additional information:

It is worth noting that some VERY rare variants in *MTHFR* can result in complete methylenetetrahydrofolate reductase deficiency, which can cause a severe neurological disorder evident at birth or early in childhood. This severe condition is not caused by the common variants discussed in this section.

Resources:

Below are a few helpful sites for further information about *MTHFR* variants:

- www.cdc.gov/ncbddd/folicacid/mthfr-gene-and-folic-acid
- www.blog.23andme.com/articles/our-take-on-the-mthfr-gene

Summary statistics were downloaded from the NHGRI-EBI GWAS Catalog (Sollis et al., 2022) on 07/19/2023.

Mast Cells and Hypermobility

What are Mast Cells?

Mast Cells are a kind of white blood cell that help to fight infection in the body. When a mast cell is activated by seeing an infection, it releases histamine and other chemicals including the enzyme alpha-tryptase. These recruit other white blood cells and make a local inflammatory reaction that helps to fight the infection.

Why are mast cells relevant to hypermobility and EDS?

People with a variety of conditions, including hypermobility spectrum disorders and EDS, can experience abnormal or excessive mast cell activity. This is sometimes called mast cell activation syndrome (MCAS). While the symptoms overlap, this is different than "mastocytosis" disorders where people make too many mast cells. Hereditary alpha tryptasemia (HaT) is another condition with similar symptoms. In HaT, people have an extra copy of the alpha-tryptase gene (*TPSAB1*) causing high levels of the alpha-tryptase enzyme.

What are the symptoms of mast cell activation syndrome?

Symptoms of MCAS affect multiple organ systems and can include:

a) Skin: flushing, itching, swelling, and hives
b) GI: diarrhea, nausea, vomiting, abdominal pain
c) Lungs: wheezing and shortness of breath
d) Heart: fast heart rate, low blood pressure, passing out

What are the triggers of mast cell activation syndrome?

There are many different triggers for mast cells. Different people have different triggers and people may not always respond to a trigger. Life factors, like stress levels and medications, may make a person more likely to respond to a trigger. Triggers for mast cells to release histamine and other chemicals include (but are not limited to):

- Extreme temperatures or sudden temperature changes
- Emotional, physical, or environmental stress
- Exercise
- Fatigue
- Food or beverages, including alcohol
- Medications (opioids, NSAIDs, antibiotics, local anesthetics) and contrast dyes
- Strong odors
- Venoms (bee, wasp, mosquito, etc.)
- Infections (viral, bacterial or fungal)
- Mechanical irritation, friction, vibration
- Sunlight

How is mast cell activation syndrome diagnosed?

A consensus conference in 2010 laid out three criteria for diagnosing mast cell activation disease:

1) Typical symptoms in two or more organ systems (see above) should recur and not have another clear cause or explanation
2) Mast cells should be directly implicated in the symptoms
3) Symptoms should improve when the actions of histamine are blocked

What are the biochemical tests for mast cell activation syndrome?

When mast cells are activated, the released histamine is most directly responsible for many symptoms, but it is extraordinarily unstable and difficult to measure. The enzyme alpha-tryptase is also released and this can be measured from blood, but is also variable and unreliable. Alternatively (or in addition), breakdown products of histamine, leukotriene or prostaglandin can also be measured in urine collected for 24-hours, and refrigerated as collected. However, a person needs to have multiple levels tested, including a baseline and a second level within 2 hours of a significant mast cell activation event. Even with this, the change in levels is not always enough to be diagnostic.

Should I have testing for mast cell activation syndrome?

Testing with multiple blood and urine samples may be done by a knowledgeable specialist at a major medical center. Most often, allergists and immunologists will order this testing. Since testing for the chemicals released by mast cells is not widely-available, most people use criteria 1 & 3 (above) to make a "likely MCAS" diagnosis.

What are treatment options?

If there is a known trigger, this may be able to be avoided. Medication treatments for known or suspected mast cell activation syndrome rely on blocking histamine responses using a combination of medications. For preventing symptoms, these generally include antihistamines (often cetirizine, fexofenadine, or loratadine) and gastric H2-receptor blockers (such as famotidine). Ranitidine used to be a widely-used H2-receptor blocker, but it is unfortunately no longer available. If an attack is occurring, diphenhydramine or hydroxyzine may be needed. Additional medications include cromolyn and steroids for acute reactions can be added by a specialist if these don't fully control symptoms. But, steroids should be avoided unless absolutely necessary since they have serious acute and chronic side effects.

Should I treat mast cell dysfunction?

Sometimes, avoiding triggers is sufficient to limit symptoms. For many people the symptoms of mast cell activation syndrome are more of an irritant than a danger. In this case, treatment is useful if the symptoms are bothersome or frustrating. For a few people, symptoms can be dangerous such as if they cause difficulty breathing or swallowing. If this is the case, that person should be treated and under the care of an expert immunologist/allergist.

Resources:

Below is a helpful site for further information about mast cell activation syndrome:

www.aaaai.org/conditions-treatments/related-conditions/mcas

Dietary Supplements

What are dietary supplements?

Supplements are meant to help a person to get enough vitamins and minerals when the diet does not. Unless a specific deficiency is indicated, and supplementation is the specific treatment, they are not meant to treat, diagnose, prevent or cure disease. If supplementation is the treatment for a disease, that supplement would be prescribed by and monitored by a physician.

While the Food and Drug Administration (FDA) approves prescription and over-the-counter medication, their role in supplement regulation is very different. Medications (prescribed or over-the-counter) go through rigorous clinical trials before they can be sold, while supplements do not have this rigorous testing or approval process. Supplements are considered to be the "wild wild west" of the pharmaceutical industry. Companies that supply them are not required to prove that they can actually do whatever their advertising claims or to ensure that their ingredients (active and inactive) are actually safe.

What supplements are recommend for people with hypermobility?

Some patients with Ehlers Danlos Syndrome may find it helpful to use supplements in their diet. There are a few supplements that can typically be universally helpful for patients with EDS such as Vitamin D3 and Calcium to help with a risk of decreased bone density. However, most supplementation should be done with care and to treat specific problems.

What about supplementing collagen?

Collagen supplements are not recommended and will not help your underlying collagen issues caused by EDS. Collagen supplements are immediately broken down by your body into their most basic parts-- amino acids. These amino acids are then directly incorporated into your faulty collagen processing. So, taking collagen supplements won't fix your collagen, it's just an expensive way to give your body more materials to make more faulty collagen.

The rare exception to this is if a person has a very limited diet or has poor absorption. A few of the amino acids in collagen are rare, so, occasionally, someone might need to supplement collagen if their diet can't get them enough of these amino acids.

How can testing help?

Because some of the conditions that often come with EDS can interfere with getting enough nutrition (Gastroparesis, Celiac Disease, Irritable Bowel Syndrome, Mast Cell Activation Disorder, and others), many people with hypermobility and EDS think about adding supplements. It is a good idea to have testing before doing any supplementation. This may include testing blood chemistries regularly and possibly a check of specific micronutrient levels. Many supplements benefit from being balanced within your body. In addition, supplementation can be expensive, so you don't want to pay to supplement things you already get enough of.

Are there risks to dietary supplements?

Supplement manufacturers are required to follow good manufacturing practices to make sure that the product they are selling the consumer is the correct compound, is pure, is at the strength advertised, and contains the ingredients advertised.. After a product is on the market, the FDA practices what is called relies upon the free market to find any safety or accuracy issues. If the FDA is alerted to an unsafe dietary supplement they can remove the product from the shelves or ask the manufacturer to recall it.

While supplements are oftentimes advertised as natural products, that does not lessen their potential effects on the body. Taking supplements at higher than recommended doses or in the place of a prescription medication could lead to complications. It is also possible to experience 'polypharmacy' affects from taking a multitude of different supplements. Just like with prescription medications, supplements can interact. This is especially problematic for supplements since a doctor or pharmacist is often not aware of the supplement or of a potential interaction.

Supplements can have intense effects on delicately balanced body systems and processes such as clotting, responses to anesthesia, liver and bone health, and metabolism of birth control or other medications. Always consult a pharmacist about your current medications and possible interactions before starting any new supplement. This caution is even more important in children as their systems are more delicately balanced and therefore safe and effective doses of supplements are usually quite different for them. Consult a pediatrician before giving any supplement to a child.

As with a prescription medication, take care before starting any supplements if you are pregnant or nursing. Most supplements (if they are studied at all before being marketed and sold) are not studied in pregnant or nursing people and their safety and transference to a developing fetus or newborn are not documented.

What form of supplement might be best?

It is also important to consider the route of administration for these supplements; if you have gastrointestinal conditions that require you to have a feeding tube or cause major issues with absorption, non-oral supplementation may be a good idea. Some compounds cross the skin easily and can be administered through a vitamin patch, others can be taken in sublingual form.

Summary

In summary, please consult your doctor before administering any forms of your own supplementation since some require more careful planning and monitoring. Consider getting micronutrient testing to confirm the supplements you need be taking before spending a lot of money on things you do not need. Watch for interactions between supplements and prescription medications.

For more information: https://ods.od.nih.gov/factsheets/WYNTK-Consumer/