Hypermobility and Ehlers Danlos Syndrome Handout

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Q: What is hypermobility?

A: Joint hypermobility is a condition that features joints that easily move beyond the normal range expected for that particular joint. Hypermobile joints tend to be inherited. Symptoms of the joint hypermobility syndrome include pain in the knees, fingers, hips, and elbows.

Q: What is EDS (Ehlers Danlos Syndrome)?

A: The Ehlers-Danlos syndromes are a group of connective tissue disorders that can be inherited and are varied both in how they affect the body and in their genetic causes. They are generally characterized by joint hypermobility (joints that stretch further than normal), skin hyperextensibility (skin that can be stretched further than normal), and tissue fragility. The Ehlers-Danlos syndromes (EDS) are currently classified into thirteen subtypes. Each EDS subtype has a set of clinical criteria that help guide diagnosis; a patient's physical signs and symptoms will be matched up to the major and minor criteria to identify the subtype that is the most complete fit. There is substantial symptom overlap between the EDS subtypes and the other connective tissue disorders including hypermobility spectrum disorders, as well as a lot of variability, so a definitive diagnosis for all the EDS subtypes when the gene mutation is known—all but hypermobile EDS (hEDS)—also calls for confirmation by testing to identify the responsible variant for the gene affected in each subtype. For those who meet the minimal clinical requirements for an EDS subtype—but who have no access to molecular confirmation; or whose genetic testing shows one (or more) gene variants of uncertain significance in the genes identified for one of the EDS subtypes; or in whom no causative variants are identified in any of the EDS-subtype-specific genes—a "provisional clinical diagnosis" of an EDS subtype can be made. These patients should be followed clinically, but alternative diagnoses and expanded molecular testing should be considered. Please remember that an individual's experience with an EDS is their own, and may not necessarily be the same as another person's experience. Diagnostic criteria are meant solely to distinguish an EDS from other connective tissue disorders, and there are many more possible symptoms for each EDS than there are criteria. (https://www.ehlers-danlos.com/what-is-eds/)

Q: What are areas in which you might be affected?

A:

- Joints (painful joints, flexible joints, "double-jointed", etc)
- Headaches
- Gastrointestinal (GI) (slow moving, diarrhea, constipation, abdominal pain, Medial Arcuate Ligament Syndromes, etc), skin (easy bruising, scarring, etc)
- Allergic-like reactions (Mast Cell Activation syndrome, etc)
- Low blood pressure or dizziness with standing (POTS – postural orthostatic tachycardia syndrome)

Q: What can I start to do to help my symptoms?

A: #1 Start learning more about your symptoms and disease! Often this is inherited and at times genetic testing will be ordered. If not, that's ok. I want you to learn more about hypermobility than anyone else. You are the driver of your own ship and health! School me in what you know :)

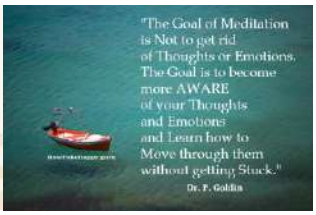
- Lidocaine ointment or patches:
  - These are found over-the-counter over the by pharmacy section of Smith's, Walgreens, Walmart, etc. The brand name is Aspercreme but you do NOT need the brand name. Just make sure the ingredients say "lidocaine 4%" when you flip the box over.
  - For Ointment: may use 4-5x/day on worst 2-3 areas
  - For patches: place on 12 hours and off 12 hours (best on backs, necks, - somewhere flat)
- Voltaren gel (This is a prescription anti-inflammatory cream/ointment)
  - Place on no more than 2 sets of joints (ex: knees and wrists OR knees and ankles OR fingers and knees)
  - MUST use 4-5x/day!!! If you only use 1-2x/day you will not get the full anti-inflammatory effect and the inflammation will continue
  - ***May mix with lidocaine ointment if you want.
- Physical Therapy (PT)
  - This will likely be ordered on your first visit as obtaining a physical therapist to help treat your muscle pain, stabilize your joints, and strengthen the muscle around the joints and ligaments will be important! Make sure the PT you have understands hypermobility and/or EDS and chronic pain.
- Joint splints
  - These also may be ordered on your first visit or after your visit with your physical therapist. This is not to prevent your joints from overextending but a reminder to yourself to prevent yourself from over extending your joints. Remember, constant cracking, over extending your joints can have an affect down the line as your are forcing those joints past where they should be. **Have good body awareness** (meaning, know where your joints are, and keep them in a neutral position!)

(examples of a finger splint)

- Meditation
  - This can be very helpful for refocusing yourself, reducing pain, improving sleep, and overall mental health and physical wellness. Some apps that can be downloaded for free on your phone are:
    - Calm

    - Headspace

  - Meditate for at least 10-15 min/day. But remember, if you can't seem to get that in, even 2-3 minutes is great! It's amazing how taking 10 min at the beginning of your day can affect the next 1440 minutes (or 24 hours) of your day!



- Referrals:
    - You likely will be referred to a Geneticist for formal diagnosis. If you also have GI (stomach/intestinal) issues you will likely be referred to a GI specialist. The same goes for skin issues (Allergist/Immunologist), or blood pressure issues (POTS specialist)
- Other Pain treatments:
    - We will likely try other medications for your pain. Ultimately, opioid medications are NOT the answer and will usually cause worsening of GI (stomach/intestinal) issues! May patients come off their opioids and feel better or the same off of them.
        - Oral medications: Some medications that have helped people with hypermobility are NSAIDs, acetaminophen (Tylenol), Pregabalin (Lyrica), Gabapentin, Cymbalta
        - Lidocaine infusions:
            - These are an intravenous (IV) infusion of the local anesthetic (numbing medication) directly placed into your vein and infused over a 30 min period. Some patients who do not respond to other oral medications respond to this type of treatment as it acts almost like a nervous system reboot. It is by no means a magic wand but can possibly reduce your pain to a more manageable level! This will likely be done in your physician's office with close monitoring. If you have seizures, arrhythmias (irregular heartbeat), or allergy to lidocaine you might not be a candidate. So, let your doctor know if you have any history of these.

Remember, you are not alone! There are many people out there with hypermobility and often similar pain and symptoms that you are going through. Often keeping track of your symptoms, what makes it better or worse, and then understanding your body can be very helpful. Hang in there! You got this!