Medical Care of Patients with a Joint Hypermobility Disorder

This article is intended for medical generalists caring for patients with the commoner joint hypermobility conditions. I encourage patients with these conditions to read it too, so as to better collaborate with their physicians. This material is based on much input from national and international experts in the field, plus my own clinical experience of working with these patients for over 15 years.

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Introduction

This document aims to help doctors provide good medical care to people with the two most common conditions in which joints are abnormally loose, or "hypermobile." Family doctors, internists and pediatricians can do this, and do it well. No one else will do it at all. Since most of the tools for helping people with these conditions are already familiar to doctors in relation to other illnesses, this is not hard. Most patients are young, and highly motivated to overcome their illness. Until medical generalists step in to diagnose them and treat them, they will continue to be largely undiagnosed and wrongly treated. Helping these young people through their challenges to find a fulfilling life can be both satisfying, and inspiring, for the generalist physician.

Why primary care practitioners should treat people with joint hypermobility syndromes

These conditions are not rare. My estimates suggest a prevalence of at least one in five hundred: so most high schools include one, or several, adolescents struggling with a hypermobility syndrome. But no medical specialty takes responsibility for their treatment. (Geneticists diagnose, but do not treat.) Typically, patients are either not diagnosed, or misdiagnosed, by rheumatologists, orthopedists and others, whose treatment is often fruitless, or positively harmful. But since these conditions affect several body systems, the doctor best equipped to manage them is the primary care generalist. A family doctor, general internist or pediatrician can provide much, and sometimes all, the medical care needed by most people with a hypermobility syndrome. This article, which is on my website at *AlanSpanosMD.com*, summarizes what the primary care practitioner needs to know, based on the consensus of national and international experts.[1]

Diagnosis of the common joint hypermobility syndromes

The companion article to this one is *Joint Hypermobility: Diagnosis for Non-Specialists.* It explains a protocol for diagnosing "hypermobility spectrum disorder" (HSD) and its relationship with "hypermobile-type Ehlers Danlos Syndrome (hEDS). These two terms denote adjacent areas on a theoretical spectrum, rather than two biologically-distinct medical conditions. Treatment is the same for HSD and hEDS.

The generalist can diagnose HSD with reasonable confidence, but adding "possible hEDS" will allow for the possibility of future revision by a geneticist. If that occurs, it will not materially alter treatment, nor invalidate the benefits of the treatment already given for "HSD." For these reasons, I will refer to "HSD/hEDS" as the condition this article deals with. Since most patients are female, I will refer to patients as "she."

Clinical Presentation of HSD/hEDS

Main features of HSD/hEDS are listed below. The patient may have any or all of them. Onset is in childhood, with symptoms often worsening in adolescence and early adulthood. Most patients are female. Some only come to medical attention when they are already parents, if a child is found to have a hypermobility condition, and it then becomes clear that the parent is also affected.

Main features of HSD/hEDS[2]	
Multiple hypermobile joints	Some of these subluxate or dislocate, and some are painful.
Widespread pain	Pain is not restricted to joints. It often has neuropathic features, i.e. is burning, jolting, or tingling, perhaps with altered tactile sensation.
Orthostatic intolerance	On prolonged standing the patient is dizzy, mentally "foggy", or just feels a strong need to sit or lie down. Heart rate may be very rapid.
Gastrointestinal disorders	A wide variety of functional and structural GI conditions are common in EDS. Two articles on this are at *AlanSpanosMD.com*.
Reduced stamina	This may be due to hypersomnia, orthostatic intolerance, or it may resemble the profound prostration of post-viral fatigue syndromes, lasting for days or even weeks after exertion.
Hypersomnia	Long sleep time, and often a need to nap during the day, are typical. Sleep may be light and restless, or unusually deep, so that the patient may sleep through alarms.
Anxiety +/- panic attacks	These symptoms may overlap with those of orthostatic intolerance, i.e. episodes of extreme anxiety, faintness, tachycardia, sweating, and/or other autonomic symptoms.
Easy bruising +/- bleeding	Clotting studies are generally normal except for bleeding time. The cause is probably fragile or unusually-permeable small blood vessels.
Slow healing of injuries	Sprains, fractures, lacerations and surgical incisions heal slowly and may be wide with depressed, thin skin in the scars.
Family history	One or more family members usually have at least some of the patient's problems.
Other features	Many other features including distinctive facial structure, migraines, dysmenorrhea, bladder dysfunction, and various neurologic syndromes are seen in HSD/hEDS. Recently, suspicion has risen that mast cell activation syndrome (MCAS) may also have a relatively high prevalence in this group.

Treatment for HSD/hEDS: General Principles

I suggest the following four general principles, for effective management of HSD/hEDS in primary care.

1. Doctor-patient collaboration

People with HSD/hEDS are used to dealing with doctors who know nothing about their condition. Typically, these patients have themselves learned a lot about it, mainly through online sites. These include videos of lectures by leading experts, often at a level appropriate for doctors as well as patients. So this is a topic on which doctors should expect to be educated by their patients, and should encourage them in this. Information presented by the patient is generally credible, and relevant to her care. Taking this seriously is helpful in itself, and strengthens the therapeutic alliance with the patient. A starting point for physician-oriented information online is the website of the Ehlers Danlos Society at *ehlers-danlos.org.* Another is my own website at *AlanSpanosMD.com.*

2. Deal with one problem at a time

Most HSD/hEDS problems can be managed separately, allocating a visit to each. This greatly reduces the feeling of overload every doctor experiences when confronting a patient with a complex disorder affecting several organ systems. But the full problem list should be at least glanced at, at each visit, since medications (or nondrug treatments) for one problem may have effects on others.

3. Precautions for surgery and anesthesia

When unconscious, the HSD/hEDS patient can easily suffer ligament and joint injuries while being moved before, during and after surgery. Also, as all connective tissues tend to heal more slowly in HSD/hEDS, surgical technique should take account of this. Post-operatively, allowance should be made for the prolonged healing time expected. Such concerns apply also to procedures like endoscopy, if they require anesthesia. These issues are spelled out in a one-page handout for the surgeon and/or anesthesiologist, available on my website at *AlanSpanosMD.com.* I advise that patients themselves have a copy and follow its guidance on how to get better, safer treatment during surgery or anesthesia.

4. PCP-specialist collaboration

When the PCP refers an HSD/hEDS patient to a specialist, it's most helpful if a referral letter is sent, with a copy for the patient to take to the appointment. This should let the specialist know the patient has HSD/hEDS and that this should be considered in the specialist's assessment and treatment. The patient should be encouraged to take to the appointment any printed materials she can find on these items. Handouts at *AlanSpanosMD.com* may be a useful starting point. Also, the website of the Ehlers Danlos Society, at *ehlers-danlos.com,* is adding to its collection of handouts for specialists and generalists.

Treatment for HSD/hEDS: Specific Symptoms

Pain

Pains in HSD/hEDS have multifactorial causes. Some joints hurt when they dislocate, though many do not. Also, people with HSD/hEDS often have pain and tenderness in areas without joints: for instance, in the hands at around the midpoints of the metacarpal bones. Sometimes these pains are referred from distant joints, but sometimes no joint can be incriminated. Pains commonly have the burning, tingling or shock-like features of neuropathic pain. These may be due to deformation of nerves around subluxing joints, or to unknown processes in the central nervous system. Pains often respond to work with a physical therapist, whether the mechanism is clear or not. Given these uncertainties, the following suggestions seem prudent, and are supported by clinical practice and expert consensus.

Pain medications that commonly help in HSD/h EDS are listed in the table on the next page. Note that for **Migraines** (which are common in HSD/hEDS), the usual abortive and preventive medications may be tried. It is not known whether HSD/hEDS alters the success rate with any of these. However, their side effects may affect other EDS symptoms. Propranolol may prevent migraine and also help orthostatic hypotension and anxiety; nortriptyline may reduce migraines and also help sleep and pain.

Physical therapy can greatly help people with an HSD/hEDS. Therapists need to meet three criteria:

1. They must have training and experience in at least one technique of manual therapy.
2. They must work one-on-one with clients for the full duration of each treatment session.These patients are complex: treatments need repeated adjustments in the light of unpredictable responses, both good and bad.
3. The PT must be prepared to do some study of methods useful in HSD/hEDS. A handout on my website at *AlanSpanosMD.com* provides downloadable materials and references on this, for physical therapists.

Swimming is often uniquely helpful for people with HSD/hEDS, if a convenient, and suitably warm, pool is available. A PT can give directions on helpful workout routines, but many patients also benefit from just getting in the water and moving around in it, doing whatever feels good.

Braces can be very helpful, especially for knees, ankles, elbows, wrists and fingers. The physical therapist should be in charge of recommending and fitting them. Some commonly-used brands don't work well for people with HSD/hEDS. The best range is by Bauerfeind. For fingers, Silver Ring Splints are excellent, and can be fitted at home via an online Skype interview with one of the company's orthotists, accessed at *silverringsplint.com*.

Surgery is often considered, to stabilize painful joints in HSD/hEDS. However, the operations often fail. Indeed, a common way that people with HSD/hEDS get diagnosed is when several joint stabilizations have been done, and none have succeeded. The patient herself should be made aware of this. She should deal with only those orthopedists who are prepared to modify their surgical techniques, and their recommendations, when the patient has HSD/hEDS. The main review of this topic is summarized and critiqued in the article, *EDS and Orthopedic surgery*, at *AlanSpanosMD.com*.[3] Spinal surgery presents especially formidable dilemmas, discussed in my article, *EDS: What Your Specialists Need to Know*.[4]

Main useful pain medications for HSD/hEDS patients over age eighteen[5]

This table reflects experience in one clinic. There are no comparative studies, or placebo-controlled trials, of any analgesics for HSD/hEDS pain. Since symptoms shift unpredictably, all medications should be reviewed every few months. Medications should be trialled one at a time, at an adequate dose, for the minimum time needed to establish benefit and adverse effects. The commonest error in prescribing for HSD/hEDS is to leave the patient on a drug for months or years without ever having established what it is, and isn't, doing for her. See product literature for details of possible adverse effects.[5]

Medication	Typical starting regimen	Comments
NSAIDS	Specific for each agent	Some help for some patients, but GI side effects often limit use. Renal dysfunction is a special concern if patient has orthostatic intolerance. [5]
NORTRIPTYLINE 10mg	1 QHS an hour before bedtime. Increase by 1 every 5 days, but not over 5 QHS (50mg). Titrate to maximum benefit vs adverse effects.	Use daily, not episodically. Helps sleep as well as pain. For most patients the only side effects are dry mouth and mild constipation, and these often subside with use. But may worsen tachycardia in patients with orthostatic intolerance. [5]
GABAPENTIN 300mg	1 QHS x 5 days, increase by 1 QHS every 5 days, up to 4 QHS. If tolerated, consider adding 1 QAM for 5 days then BID for 5 days, then double those doses if tolerated.	Use daily, not episodically.Helps sleep as well as pain. Unsteadiness and morning sedation are main initial adverse effects. Once optimum day and night regimens are established, further adjustment of any of the doses may be tried. [5]
TRAMADOL 50mg	1 QD for 3 days, then 1 BID for 3 days, then 1 TID for 3 days, then 1 QID. Some patients may tolerate 2 TID or QID.	For daily or episodic use. Adverse effects and interactions limit its value. But has the advantages of episodic or daily use, and avoidance of the issues surrounding Schedule 2 opioids. [5]
TIZANIDINE 4mg	1 QHS for sleep disruption due to pain. Can be titrated up, e.g. 2 QHS, and 1-2 up to TID PRN if daytime analgesia also needed.	Common adverse effects are chest pain, dysuria, agitation, drowsiness, exhaustion, feverishness. Main advantage is brief action, so if taken at night should not cause morning sedation. It can be used routinely or episodically. [5]
OTHER MUSCLE RELAXANTS	See above for tizanidine. Other muscle-relaxants seem only rarely helpful for EDS, and are dosed as for other conditions.	Some patients get more subluxations after taking muscle relaxants at night: they may impair reflex holding patterns so that hypermobile joints sublux more than usual, during sleep. [5]
OPIOIDS	Use standard dose titrations as for other conditions.	Usual adverse effects and risks apply. Despite them, some patients insist that "I can only move" because of a daily opioid regimen. Be alert for unusual allergic or inflammatory effects due to mast cell activation by opioids. Beware serious skin damage risk from transdermal preparations. [5]
BENZODIAZEPINES	Use standard dose titrations as for other conditions.	Some patients report improved neuropathic pain, not just better sleep and reduced anxiety. This may reflect their effect as mast cell stabilizers. [5]

Hypersomnia

This can be a major handicap, since several productive hours may be lost every day to the unusually long sleep times many HSD/hEDS sufferers exhibit, and also due to a need for daytime naps. Sleep studies are rarely helpful unless there are clear symptoms of sleep apnea or narcolepsy. Often, treatment of pain reduces total sleep requirement and improves daytime alertness and energy. A small dose of an alerting agent daily may help, preventing sleepiness during a segment of the day when the patient needs to be fully alert, for instance a student who needs not to nap during an afternoon class, or a mother who needs to be alert so as to drive and go shopping. This simple measure can add a worthwhile amount to the useful time the patient has during the day.[6]

Exhaustion

Exhaustion, i.e. poor stamina limiting normal activities, should be distinguished from daytime sleepiness. When not due to poor sleep, this may be due to circulatory instability (see below). Whether this is so or not, it may improve markedly with low doses of amphetamines. Main options are 5-10mg of dextroamphetamine (Dexedrine), mixed amphetamine salts (Adderall) or methylphenidate (Ritalin) QAM, repeated if necessary a few hours later. Armodafanil (Nuvigil) may also help, especially if daytime sleepiness is also present.

Circulatory instability

Most HSD/hEDS patients have some degree of circulatory instability, typically manifesting as orthostatic intolerance (OI), i.e. dizziness or faintness on staying upright, and/or reduced stamina with physical exertion. Typically the patient gets cold, pale or blue hands and feet on staying upright, and may have marked tachycardia, which by definition would warrant a diagnosis of POTS (postural orthostatic tachycardia syndrome). This field is confused and confusing. Different categories continue to be proposed and argued over, including POTS, neurally mediated hypotension (NMH) and others. To date there is not a single prospective, randomized controlled trial of any treatment of these conditions. So treatment protocols mainly reflect the habits and biases of individual providers or institutions. Treatments are essentially trial-and-error, and involve pharmacologic and also nonpharmacologic interventions. The interested generalist can often do at least as well as a cardiologist in this area, and perhaps better, since the cardiologist may be focused on titrating treatment against measurements (pulse and blood pressure changes) rather than patients' symptoms, and these are often discordant. Also, cardiologists' follow-up of these patients tends to be at long intervals, thus preventing them from doing the closely-monitored, symptom-based, brief treatment trials that seem to be most helpful.

GI symptoms

Many, perhaps most, HSD/hEDS patients have GI symptoms. These may be caused by relatively rare conditions such as gastroparesis, or malabsorption syndromes, as well as any of the common gastrointestinal conditions. Clinical clues to the diagnosis may be scanty, so a wide differential diagnosis should be considered. If a clinical diagnosis and symptomatic treatment are not helpful, then early workup is warranted. My website includes an article for the gastroenterologist on these conditions,[7] as well as a critique of a recent literature review on this subject.[8]

Bleeding and bruising

Multiple bruises, which may be spontaneous, are commonplace among people with HSD/hEDS. The phenomenon itself rarely needs specific treatment. Claims have been made that Vitamin C supplementation may reduce this, but without supporting evidence. Clotting factors are normal, so the bruising is assumed to be due to fragility, or leakiness, of small blood vessels. Prolonged bleeding is common, perhaps due to failure of blood vessels to contract adequately after injury. This is relevant for the surgeon. Menorrhagia should be specifically enquired about, and treated in the usual ways, in EDS patients. Note that life-threatening catastrophic bleeding, especially in pregnancy or delivery, should prompt consideration of whether the patient has Vascular EDS (vEDS), rather than the much more common Hypermobility type (HSD/hEDS). A geneticist can make this determination.

Anxiety and panic attacks

Assessment and treatment are the same in HSD/hEDS as in its absence, with one important exception. This is, that instability of autonomic function may cause symptoms of anxiety and panic attacks in HSD/HEDS patients. Thus, episodes of panic may respond to low-dose propranolol (10-20mg) as well as to other anxiolytics.

Surgery and anesthesia

See page 3, above.

Other symptoms

Various other symptoms may be associated with HSD/hEDS. In general, if any chronic symptoms have unusual features, or fail to respond to their usual treatments, an online search should quickly reveal whether there's a relevant connection with HSD/hEDS. Patients themselves can access such information, thereby helping the doctor and further strengthening the collaborative doctor-patient relationship that is a key to success with these patients.

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[1] The International Consortium on Ehlers Danlos Syndrome and Related Conditions reported the findings of its various committees in a special issue of the American Journal of Medical Genetics in 2017. Specific reports are referenced in the following endnotes.

[2] Only rather recently, since the 1990s, has attention been paid to extra-articular manifestations of the hypermobility syndromes. The mechanism of these is largely unknown. However, it has become clear that much of the symptom burden of these conditions arises from these conditions, rather than from joint pain and displacements.

[3] Ericson Jr. WB, Wolman R. 2017. Orthopaedic management of the Ehlers–Danlos syndromes. Am J Med Genet Part C Semin Med Genet 175C.

[4] Henderson Sr. FC, Austin C, et al. 2017. Neurological and spinal manifestations of the Ehlers–Danlos syndromes. Am J Med Genet Part C Semin Med Genet 175C.

[5] See product literature for possible adverse effects and interactions.The usual cautions apply when considering these medications for patients under 18. Experience with several of them is limited in this age group, and lower starting doses may be appropriate. Similar cautions apply to the elderly.

[6] By "alerting agents" I refer to the amphetamines used for ADHD, i.e. dextro-amphetamine, Adderall and Ritalin (methyphenidate), as well as Provigil (modafanil) and Nuvigil (armodafanil).

[7] See article, *EDS for the Gastroenterologist* at *AlanSpanosMD.com.*

[8] Fikree A, Chelimsky G, Collins H, Kovacic K, Aziz Q. 2017. Gastrointestinal involvement in the Ehlers–Danlos syndromes. Am J Med Genet Part C Semin Med Genet 175C.