EDS, HSD and Orthopedic Surgery

People with EDS[1] and HSD[2] go to orthopedic surgeons for help with their joints, but quickly discover that orthopedists are not familiar with their underlying conditions. Orthopedic procedures do sometimes help such patients, but failure rates, and surgical complications, are high. In this article I describe the large areas of uncertainty on this topic, and suggest how to benefit from orthopedic treatment and reduce its risks.

Introduction

Orthopedics is the surgical specialty dealing with with bones, "and associated structures (such as tendons and ligaments)." Joints are where bones connect by means of tendons and ligaments. So problems with loose joints are fair game for orthopedists, and people with joint hypermobility conditions naturally seek help from them. It should follow, then, that orthopedists are expert at treating these problems. But patients are well aware this isn't so. It's depressingly common for people with hypermobility conditions to go through several orthopedic operations that fail to accomplish their aim, before the diagnosis of a hypermobility condition is even considered. And then, the patient is usually told to have some other doctor – not the orthopedist – look into this possibility to see if it's so. If a diagnosis like Ehlers Danlos Syndrome is then made, the orthopedist often doesn't know how to proceed. Some tell the patient there's nothing further they can help with. One patient was informed, cheerily, that "I can't even pronounce it, so I'm sure not going to try to treat it," and then advised to go elsewhere.

In this article I'll summarize the little that is known about orthopedic surgery for people with EDS and Hypermobility Spectrum Disorders (HSD's), and what the practical implications are for people with these conditions.

Why we know so little about orthopedic treatments for EDS and HSD[3]

There are three main reasons why orthopedists are largely unaware of the hypermobility conditions, and whether their treatments can help with them. The first reason is that most people with loose joints do <u>not</u> have a hypermobility condition, disease or syndrome – they just have some loose joints, that are not giving any serious trouble, if any trouble at all. Many of them are good athletes – especially in gymnastics, where lax joints are a big advantage; dancers also benefit from having joints that are looser than most people's. If you do have a joint hypermobility condition, you naturally assume that anyone with joints as loose as yours has a problem like yours, but this isn't so. For most "double-jointed" people, they have several loose joints but no special medical problems because of that. They may sprain or dislocate a joint playing sports, but it heals normally and they can get back to their sporting activities. So most of an orthopedist's experience of loose joints is in young people who are fit, well and athletic. If they have an injury requiring surgery, the surgery goes fine and the patient recovers as well as anyone else. So, it's very understandable that orthopedists don't think of serious-sounding con-

---

[1] EDS stands for Ehlers Danlos Syndromes. Note this is plural. There are several Ehlers Danlos Syndromes. Their classification, and the abbreviations used for them, were changed in 2017. See the article, *Why the New EDS Classification Matters,* in this series for a discussion of the new classification.

[2] HSD stands for Hypermobility Spectrum Disorder, a new term coined by the committee that developed the new classification of EDS in 2017. See Note #1 above.

[3] For an account of why physicians in general (not just orthopedists) are very ignorant of hypermobility conditions, see the article *Why Are My Doctors Clueless about EDS?* in this series.

ditions like “Ehlers Danlos Syndrome” or “Hypermobility Spectrum Disorder” when they are treating such patients.

The second reason why EDS and HSD are missed by orthopedists is that they’ve not been taught how to diagnose them. An office screening assessment, requiring no tests or x-rays, will identify the great majority of people with these conditions, but orthopedists are not taught how to do this. Instead, they believe that only a geneticist can diagnose inherited conditions like EDS and HSD. This view used to be shared by geneticists, but they are becoming less protective of their turf as the wait time for a genetics appointment has increased till it is now often over a year. Recent deliberations by international genetics experts have resulted in their acknowledgment that non-geneticists can diagnose many EDS/HSD cases, but this hasn’t been made known to the medical profession at large.[4]

The third reason why orthopedists are slow to detect EDS/HSD, is that since they haven’t learned about them, they tend to have misconceptions about them. And some of these misconceptions make it seem justifiable to ignore these conditions. One of these misconceptions is that these conditions are very, very rare: rare enough that you don’t need to consider them in daily practice. But in fact, they are quite common – any high school is likely to have one or two students with an EDS or HSD. Another misconception is that because they are inherited conditions, part of the patient’s fixed genetic makeup, there’s nothing that can be done for them. This belief is quite deeply ingrained, and often told to patients. But a moment’s reflection shows how weirdly wrong it is. My severe short-sightedness is inherited, but no optometrist has told me I can’t benefit from glasses! Plenty of serious maladies are inherited, including sickle-cell anemia, cystic fibrosis, Down syndrome, hemophilia, and muscular dystrophy. “It’s inherited, so there’s nothing we can do,” is not a statement heard about any of these: they all are helped greatly by medical treatment. And orthopedists themselves often operate, with great success, to correct congenital, genetically-determined malformations of the skeleton such as club feet or hip dysplasia.[5] Orthopedists should really pay attention when they find themselves repeating that statement, and notice how silly it is.

What we do and don’t know about orthopedic treatment for EDS/HSD

There seems to be only one published review of the evidence on orthopedic treatment for hypermobility conditions. In itself, this is striking evidence of how little we know. The review is authored by Dr. William Ericson and Dr. Roger Wolman. Dr Ericson specializes in surgery of the hand and arm “and has a large experience with EDS patients.” Dr Wolman is not a surgeon, but specializes in rheumatology and sports medicine. Their review was commissioned by an international body of experts seeking to put together a compendium of expert knowledge on hypermobility conditions. It was published in 2017 as part of a special volume of the American Journal of Medical Genetics (which is read by almost no-one except geneticists). An indication of how slender is the “evidence base” on this topic is the fact that, of the 26 articles referenced in this review, only four actually focus on hypermobility in orthopedic practice. All combined, these four articles total a scant 22 pages. Slim pickings indeed from the world of medical research!

It is worth noticing that this review has only two authors (far fewer than most of the other reviews published with it), that only one of them is an orthopedic surgeon, and that he works in the subspecialty of hand and arm surgery. There is no sign that a general orthopedist was involved at all. Many assertions are made in the review, without referencing the support for them. One does not know if they arise out of Dr Ericson’s personal experience, or a consensus

[4] For much more on this, see the article *Why the New EDS Classification Matters,* in this series.

[5] Indeed, the very word, “orthopedics”, is from the Greek for “childhood straightening”, referring to surgery to correct congenital deformities.

among his colleagues. This is an important uncertainty, because many studies have shown that we doctors can be misled by our personal "clinical experience." One of my medical school professors used to teach that "Clinical experience is just prejudice, repeated with ever-increasing conviction, year after year."

Sadly, copyright rules prevent me from putting the Ericson/Wolman review on this website. It can be obtained online from the publisher.[6] But below, I'll summarize its main points, which you could take and show to your orthopedist. Perhaps you can even persuade him or her to keep it and read it(!). I will summarize it below, then lay out some practical conclusions for people with hypermobility conditions who are considering orthopedic treatment. **To skip the following sections, go straight to the last section below: "Conclusions of a non-orthopedist: what patients need to know."**

Six big points

Ericson and Wolman refer repeatedly to EDS, and often also to "joint hypermobility (JH)," implying that what is true of the one, is also true of the other. These groups are now covered by the terms Ehlers Danlos Syndromes (there are several), and Joint Hypermobility Spectrum Disorders, abbreviated to "EDS/HSD." I'll use that combined acronym for the population referred to in the Ericson/Wolman review.

Ericson and Wolman make six major points in their opening paragraph. Of joint hypermobility in general, they write:

1. It "is not always painful, but if so, is difficult to diagnose without highly specialized training."[7]
2. It "does not show on standard diagnostic tests." Joint hypermobility can't show on the "standard diagnostic tests" used by orthopedists, because these are x-rays and scans done with joints in a normal, resting position. The resultant pictures tell you nothing about where they would end up if the patient moved them to the end of their ranges.
3. It "does not respond to standard treatment protocols." This refers specifically to many of the standard exercise routines prescribed by physical therapists.
4. It "lowers the threshold for associated joint injuries." This means that it takes less force to damage a joint, if the joint is loose to start with.
5. It "causes premature wearing of joints." This refers to osteoarthritis, the "wear and tear" condition that is normal in the elderly, but can develop earlier if joints are lax.
6. It "results in a higher failure rate for treatment, both medical and surgical."[8]

Lest the reader finds this list too disheartening to continue reading, the authors then acknowledge that although "extremely frustrating for the patient as well as the physician … there is much that can be done for EDS patients." They promise to summarize "a general approach to patients with EDS and joint hypermobility." But then they quickly disabuse the reader of any expectation of specific advice: "The authors do not specifically endorse, approve, recommend, or certify any specific procedure or technique, and provide these opinions for gen-

---

[6] Search online for: "Ericson Jr. WB, Wolman R. 2017. Orthopaedic management of the Ehlers-Danlos syndromes. Am J Med Genet Part C Semin Med Genet 175C:188–194." This should lead you to several versions of the article. Some may be freely downloadable, others may require payment.

[7] There is disagreement amongst the experts on how "highly specialized" this training needs to be. The article, at *AlanSpanosMD.com* gives a protocol for diagnosing most cases of HSD and hEDS, that a family doctor or internist should manage without much difficulty.

[8] No support is given for this extreme statement, that seems to imply that any and all treatments for any aspect of joint hypermobility have a "higher failure rate" than in patients without hypermobility.

eral information only." What we had hoped would be a summary of *knowledge* is sounding more like an *opinion piece*. They then advocate treatment by "a Multi-Disciplinary Team (MDT) approach including physicians . . . physiotherapists, occupational therapists, psychologists, and nurses . . . to plan management more effectively." This sounds lovely, but I wonder if any reader has ever come across such a dream team. There were a few in the 1990s (including one I created myself), but doctors collaborating in multi-disciplinary teams have become a critically-endangered species in the 21st century.

The "medical literature" on EDS, hypermobility and orthopedics

Ericson and Wolman dispose of this in only a third of a page, mainly by explaining why the scanty publications on this subject are so unhelpful in clinical practice. Reasons they cite are:

1. High rates of complications and failures of surgery are reported, but they "lack detailed analysis or explanation of why surgery did not go well."
2. Unfamiliarity of practicing doctors with joint hypermobility hampers their research, since they are not using similar definitions of the conditions they are treating, and are largely untrained in how to identify joint hypermobility.[9]
3. When, as often happens, patients have the wrong diagnosis for their hypermobility, this leads to invalid conclusions in the research.

Nevertheless, the authors state with confidence that "the risks of all the known hazards of surgical intervention are distinctly higher in EDS patients." Moreover, "there is sparse information [on] orthopedic surgery in patients with EDS, *particularly successful surgery"* (my italics). Study of the primary sources the authors quote suggests this is an understatement. None of them actually records data to support *any* particular orthopedic procedure as being "successful" in any verifiable way. What they do record is *individual cases* of benefit. These cases are convincing; but there's no way to know how, if at all, one can generalize from such successes so as to guide treatment of other patients.

Basics of diagnosis

Ericson and Wolman then devote one page to diagnosis. No references are cited except for one in support of their remark that rates of "clinical depression" are raised in patients with EDS, and that this complicates diagnosis. Their main points are:

1. Pain in EDS/HSD can have several mechanisms that can be hard to distinguish, including mechanical stretching of ligaments, inflammation, and deformation of nerves by compression or stretching.
2. Nerve deformation "causes pain where the nerves end, not where they are compressed. (It) does not show on electrodiagnostic tests, and can be refractory to treatment." Nerve pain "can mimic joint pain from instability, and this feature . . . seriously complicates the lives of EDS patients."
3. Pain can be present for a long time without causing any abnormalities on x-rays; when normal these "tend to mislead the physician(s). The first clue that there is a JH problem would be a painful joint with normal radiographs."

[9] Perhaps the main failure is that most doctors don't appreciate that people with joint hypermobility have loose joints <u>but tight muscles.</u> The muscle tightness can mask the joint looseness, unless the examining doctor takes specific ways to get around this.

4. "EDS causes premature aging of the musculoskeletal system," which can cause "overlapping symptoms" at a young age.
5. EDS patients tend to have "multiple complaints, specifically vague, intermittent pain involving the limbs or spine" which can lead to a mistaken label of "fibromyalgia."
6. Patients "often drift between different specialists . . . without a firm diagnosis or successful treatment plan." They may become seriously depressed.

Pain relief: non-surgical options[10]

Ericson and Wolman devote over a page to this topic. Most of these points are widely endorsed by doctors, but are unsupported by objective evidence. They are medical hearsay, rather than scientific knowledge. The main points are:

1. Surgery "may be the only reasonable treatment for some conditions . . . but may not be an option at all . . . An unstable joint may be so lax that NO surgical procedure will ever be successful." Non-surgical treatment "should be exhausted prior to recommending surgery."
2. For an acute (i.e. sudden, and recent) injury, "the usual treatment options are appropriate for most EDS patients," beginning with "R-I-C-E (Rest, Ice, Compresion, Elevation)."
3. For chronic (i.e. longstanding) pain, "there are multiple options that may be effective."
4. Benefits from pain medications are often limited by their side effects, especially in EDS/HSD patients.
5. Splints "can be extremely helpful."[11]
6. Exercise programs can be "extremely helpful" but need to be carefully designed. Those "that do not take into account that EDS patients have loose joints but tight muscles are doomed to failure." Exercise regimens "that emphasize 'range of motion' exercises or repetitive, forceful actions such as 'work hardening' are inappropriate and can make patients' joint symptoms worse." (This is important, as these kinds of exercise program are often foisted on patients who have had occupational injuries, with the aim of helping them get back to work.)
7. Often, local anesthetics don't work well; some people do better with carbocaine than the usual lidocaine or bupivacaine. Most local anesthetics are toxic to cartilage cells; for various reasons, ropivacaine may be preferred for injections into joints.
8. There are no established guidelines for diet in EDS patients, but weight control is always important.
9. Adequate calcium and vitamin D is necessary to maintain bone strength. Exercise is also important, but may be hard for people with severe joint symptoms.

Surgical options

Ericson and Wolman devote three pages to short summaries of the role of surgery for various joints, for patients with hypermobility disorders. But they open with a major caution: "Surgery is an option for a select number of specific conditions in EDS patients, but there remains very

[10] See also pages 4-5 of the article *Medical Care of Patients with a Hypermobility Disorder,* in this series.
[11] People with hEDS/HSD are often not offered splints, though these would help them. See the article on *"Physical Therapy . . ."* in this series, for an account of this.

little in the surgical literature to support this approach . . . Normal diagnostic tests and a higher failure rate should not preclude surgical intervention in the EDS population, but serious prudence is advised."

Cervical spine

The authors mention this important topic only very briefly. It's dealt with in detail in another review, part of the same series, authored mainly by neurosurgeons.[12]

Other joints

Each of the following gets a paragraph: thoracic outlet, shoulder, elbow, thumb, fingers, lumbar spine, hip, knee, ankle, foot. However, the text and the references only expound the standard treatments for laxity in these joints: they don't deal specifically with people who have a whole-body, inherited connective tissue disorder like EDS/HSD. Essentially, these very brief vignettes say what orthopedists do for these conditions and that they help some people. They believe that some people with hypermobility conditions are helped by them, but clear evidence to support this belief is lacking. They do not address the fact that many procedures offered to people with EDS/HSD are widely practiced, even though evidence of their benefit is still lacking. A major example is endoscopic surgery for labral tears of the hip joint. The number of these operations done every year in the USA has increased by eighteen times in the last half-decade. But there are no studies showing that this enormous increase has improved outcomes over the nonsurgical treatments used before.

Nerve compression

The authors note that for pain due to nerve compression, "surgery is extremely reliable if the diagnosis is correct." But they caution that making the correct diagnosis is a challenge, and is often not illuminated by special tests, which may be entirely negative even though the patient has a compressed nerve which will respond to surgery. They stress that several other factors can intertwine in causing nerve pain, including problems with the spine and with the thoracic outlet.

Conclusions of a non-orthopedist: what patients need to know.

I am not an orthopedist, but a "generalist" with a special interest in hypermobility syndromes. From that viewpoint, the overwhelming message from this review is that orthopedists are very unsure which of their surgeries do, and and which don't, help in EDS/HSD. There's general agreement that risks of complications are much more common in these patients, and these complications incude failure to achieve the goal of the surgery. But there is a deafening silence on the practical question of when surgery may be worth these risks.

Given the lack of solid evidence, it's not surprising that there are sharp disagreements among orthopedists on these issues. A few believe that orthopedic surgery should never, or almost never, be attempted for patients with hypermobility. Others, like Dr Ericson, assert that "surgery may be the only treatment that reliably results in persistent pain relief."

[12] To find this article, Google "Henderson Sr. FC, Austin C, Benzel E, Bolognese P, Ellenbogen R, Francomano CA, Ireton C, Klinge P, Koby M, Long D, Patel S, Singman EL, Voermans NC. 2017. Neurological and spinal manifestations of the Ehlers–Danlos syndromes. Am J Med Genet Part C Semin Med Genet 9999C:1–17". You should be able to download it from more than one site, though there may be a charge for doing so.

Based on the above, and on my own experience of following several hundred patients with hypermobility conditions, some for over a decade, I offer the following modest suggestions to patients with EDS/HSD who are seeing orthopedists.

- If you have a joint hypermobility condition, or even if you only think you may have one, then **make this clear to any orthopedist you see**. Show the orthopedist your loose joints and demonstrate how "double-jointed" they are. Also make clear which joints are giving you trouble, in addition to the one that's the main agenda for this visit. An orthopedist, who understands that you have generalized loose joints, may be more cautious in recommending surgery, and if surgery is advised, may take care to select a procedure that takes this into account – and should be more likely to work because of that.
- Ask the orthopedist specifically to assess if your pain might involve **nerve compression in your spine, your "thoracic outlet," or elsewhere**.
- **Take non-surgical treatments seriously**, including medications, braces, physical therapy, and an exercise program designed by a physical therapist who has studied EDS. Surgery is the last resort, only if these fail. For non-surgical treatments, see my article, *Physical Therapy, Exercise & Braces for People with EDS*, at *AlanSpanosMD.com*. For pain medications, see the article, *Medical Care of Patients with a Hypermobility Disorder*.
- If surgery is recommended, **remind the orthopedist of your joint hypermobility**, and your specific diagnosis (EDS, HSD etc.) if you have one. Ask if the surgical procedure will be altered out of concern for this, and ask what complications you should anticipate because of your hypermobility.
- Consider getting **a second opinion** on the surgery. The orthopedist, or your primary care practitioner, may be able to suggest from whom to seek one. Some orthopedists specialize in surgery of a particular area, for instance the knee or the shoulder: a second opinion from such a specialist may be particularly helpful. A firm agreement between both orthopedists on what should be done would be reassuring. Divergent opinions might be a reason for deferring a decision on whether to have surgery at all.
- At your pre-operative visit, **give your orthopedist a copy of the article, *Surgical & Anesthetic Precautions for HSD and hEDS***, at *AlanSpanosMD.com*. Follow the instructions with that article, on how to get it into the hands of the other people who will need it before and during your surgery. And read it yourself, before the operation.
- After surgery you will likely see a physical therapist to help your recovery. Be aware that patients have had their surgeries fail because the PT didn't appreciate how fragile their tissues were, and advised an exercise program that undid the surgery and caused further damage. So be sure to **have the physical therapist read the article on physical therapy mentioned above**, and study it before starting you on an exercise program.

I hope you have found something of use in this article. On such a topic there are bound to be different opinions. I welcome feedback from patients or health professionals with other perceptions of these issues.

Alan Spanos MD
March 2019

Dr Spanos is a Clinical Associate at UNC School of Medicine. His practice is in Chapel Hill, NC.
Other articles are on his website at *AlanSpanosMD.com*.