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# Orthopedic Management of the Ehlers-Danlos Syndromes

## FOR NON-EXPERTS

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**T**he role of orthopedic surgery in the Ehlers-Danlos syndromes is controversial and difficult to discern from available medical literature. Non-surgery treatment is preferable, but for carefully selected patients, specific joint stabilization and nerve release procedures can provide symptomatic relief when conservative measures fail.

### **Introduction**

Orthopedics is the treatment of muscles, bones, and joints. The Ehlers-Danlos Syndrome (EDS) is a connective disorder that involves overly moveable joints (joint hypermobility, JH). JH is not always painful, but if so, (1) is difficult to diagnose without highly specialized training, (2) does not show on standard diagnostic tests, (3) does not respond to standard treatment, (4) lowers the threshold for associated joint injuries, (5) causes premature wearing of joints, and (6) results in a higher failure rate for

treatment, both medical and surgical. The following is a brief summary, describing a general approach to patients with EDS and JH. The authors do not specifically endorse, approve, recommend, or certify any specific procedure or technique, and provide these opinions for general information only.

The medical term for partial dislocation of a joint is “subluxation,” and EDS patients have frequent subluxation and occasional dislocation of large and small joints. The asymmetric loading of the joint surfaces as the joint subluxes contributes to the early wear of the joint surface, and it takes very little injury to make a “loose” joint “loose and painful.” EDS patients often have nerve pain. This type of nerve problem does not typically damage the nerves but causes pain where the nerves end, not where they are compressed, and unfortunately does not show on diagnostic tests, and can be resistant to treatment. Pain from nerve problems can mimic joint pain from instability, and this feature of EDS/JH seriously complicates the lives of EDS patients and their doctors.

EDS patients tend to show multiple complaints, specifically vague, on/off pain involving the limbs or spine. Doctors have a tendency to seek a simple, single diagnosis. In EDS patients, it tends to be much harder to determine the exact cause or causes of the patients’ pain, expectations may be unrealistic, and technical difficulties can have serious consequences. In spite of this, for patients, surgery may be the only treatment that reliably results in persistent pain relief.

Pain relief is a clear goal of every EDS patient. Surgery is often the last resort for EDS patients and may be the only reasonable option for some conditions. EDS patients have a higher incidence of bleeding complications, and wider scars, and less predictable healing. This does not mean they should not have surgery, but optimal treatment would include involvement of a surgeon with knowledge and experience specifically with EDS patients.

## **Non-Surgical Treatment Options**

Generally speaking, non-surgical options for treatment of joint pain should be exhausted prior to recommending surgery. The following is a partial list.

### **Acute Pain**

Pain may be from a sudden (acute) event or a long-term (chronic) pattern. In the acute setting, the standard orthopedic “R-I-C-E” (Rest, Ice, Compression, Elevation) treatment is safe and can be effective. It is not particularly effective or practical in the

chronic setting. The usual treatment options for any patient with an acute injury are appropriate for most EDS patients.

## Chronic Pain

In the chronic setting, there are multiple options that may be effective. Patients and physicians would both appreciate an “oral medication” that results in effective pain relief. Unfortunately, oral medication for EDS patients is problematic: Medications do not change the underlying cause of the pain, and often have problematic side effects.

Due to various aspects of EDS, non-steroidal anti-inflammatory drugs (NSAIDs), acetaminophen, and opioids can be harmful. Gabapentin and pregabalin may help and reduces anxiety but are associated with weight gain. Naltrexone has been used off-label for chronic pain with some success. Splints can be quite helpful for specific types of joint instability, particularly as part of a coordinated treatment program. Splints limit joint motion, and can limit pain, but may or may not result in increased stability, and if used consistently can make muscles weaker through disuse.

Physical therapy and exercise programs are essential components to successful pain relief in patients with EDS. Exercises that emphasize low-impact strengthening, position sensing, and improved posture can be extremely helpful. Physical therapy can be used effectively to increase core muscle strength and to stabilize specific joints. Local anesthesia injections can be helpful in determining the source of pain. It should be noted that the most common forms of local anesthesia are now known to be toxic to cartilage producing cells, and ropivacaine should be used preferentially for these injections. Anecdotally, carbocaine tends to work better in EDS patients. Dietary considerations are becoming more important. There may be clearer recommendations in the future. Weight control is a major imperative for any patient with EDS. Bone health, with adequate calcium intake and appropriate vitamin D levels, is very important. Exercise is also an important component of bone health but can be problematic.

## Surgical Treatment Options

EDS patients are at increased risk from any form of surgery. The decision to recommend an orthopedic operation needs to be carefully considered. Surgery is an option for a select number of specific conditions in EDS patients, but there remains very little in the surgical literature to support this approach. Normal diagnostic tests and a higher failure rate should not prevent surgical intervention in the EDS population, but caution is advised.

**Cervical Spine:** instability in the head and neck and cases of the lower brain pushing down into the spine may absolutely require surgical intervention. Upright MRIs are advisable. Degeneration of the supportive cartilage discs between bones of the spine is common. Fusing these bones may be necessary, but holds risks for the health of adjacent bones. Minimally invasive techniques, when appropriate, are preferred.

**Thoracic Outlet:** the thoracic outlet is the space where nerves and blood vessels to the arm pass from the neck/chest area into the arm. Symptoms are often related to Thoracic Outlet Syndrome (TOS). The nerves in this area are subject to forces, particularly in the case of subluxations. Compression and/or tension on nerves cause symptoms where the nerves end, not where they are pinched or pulled. The result is vague hand/arm pain that unfortunately overlaps with the other areas that tend to be painful in patients with loose joints. Physical therapy is essential for this condition. Botox injections into muscles can give tremendous relief. Surgery may be indicated but can make patients worse. TOS is a complex and controversial topic in the medical community and seriously complicates the lives of many patients with EDS.

**Shoulder:** shoulder instability responds well to physical therapy in most patients. The goal of therapy is to increase the resting tone of muscles, without overpowering others. Imaging is typically normal. Surgery (Neer Inferior Capsular Shift) can be extremely helpful in stabilizing the shoulder. Possible complications include recurrent instability, and joint stiffness. In patients with very, very loose shoulders this procedure has a high failure rate and should be approached cautiously. Tears around the shoulder are not uncommon and are more likely in patients with excess joint motion. Surgery is indicated some cases. Possible complications include repeated tears and joint stiffness.

**Elbow:** these problems often resolve spontaneously or with physical therapy or other modalities, such as Platelet Rich Plasma (PRP) injections, but when persistent and uncontrolled, surgery can be a reasonable option.

**Wrist:** EDS patients tend to have unstable ankles, knees, and hips, and frequently fall on their outstretched hands. This wrist damage can convert loose wrist joints into painful loose wrist joints. Physical therapy and hand exercise often make this type of wrist pain worse. Surgical stabilization of the wrist works reasonably well in some cases. Nerve entrapment causes severe wrist pain and can be treated successfully with surgery if the diagnosis is made.

**Thumb:** many thumb issues can be successfully treated with surgery.

**Fingers:** hyperextension of the finger joints may not produce symptoms. If painful, or if the fingers catch or lock, figure-of-eight splints are extremely helpful. Surgery is an option if the splints fail, but this type of surgery is technically challenging and has a higher failure rate. Tendon problems respond well to surgery, if necessary and the diagnosis is correct.

**Lumbar Spine:** extreme pressure and swelling of nerves in the lower spine is a concern for any patient with EDS or JH who shows severe back pain, particularly with leg weakness or numbness between the legs, incontinence or sudden onset of sexual dysfunction. This can require emergency surgery to prevent permanent paralysis and loss of bladder/bowel control.

**Hip:** subluxation can lead to inflammation, which makes it difficult for patients to sleep on their sides. This may show up on MRI and usually responds to physical therapy and steroid injections (avoided if possible). In difficult to treat cases, surgery can give tremendous relief, if the diagnosis is correct. Tears are much more common in EDS patients, and removal or repair can give relief, although long term evidence for this procedure is lacking. Joint instability (of the SI, sacroiliac joint) is very common in EDS patients and shows as vague lower back/ pelvic pain. This often responds well to physical therapy. Injection of an irritant (prolotherapy) for isolated SI joint instability can help but remains controversial. Braces to stabilize the SI joint can be helpful. Surgery for SI joint instability is rarely necessary but can give immediate and permanent relief of pain. Hip pain may also be from disk failure in the lower spine.

**Knee:** knee instability usually responds to physical therapy and occasionally requires a knee support. Surgery can be utilized in cases of tearing. Physical therapy can be quite helpful in creating stability of the knee joint. Advances in implant design have made surgery a more viable option for arthritis.

**Ankle:** the ankle tends to give out on uneven ground, and often causes falling. The ankle may also be injured by the fall and can become more unstable. Ankle braces and orthotics work reasonably well in many patients but are cumbersome. Procedures around the ankle have a high failure rate, and wound problems are common. Physical therapy and orthotics are the mainstays of treatment, but stabilization surgery can be helpful.

**Foot:** bunions that aren't painful should best be left alone. Pain in the ball of the foot is also common. Steroid injections may make this problem worse. Orthotics are the mainstay of treatment for foot deformities.

**Nerve:** peripheral nerve problems are common in EDS patients. Surgery for peripheral nerve compression is extremely reliable if the diagnosis is correct. Unfortunately, EDS patients often present with multiple, simultaneously overlapping nerve complaints, and sorting out the cause can be tedious, time-consuming, and resource intensive. Electrodiagnostic studies are often ordered to assess for nerve damage but are not helpful when the results are normal, which is common. Understanding the contribution of the patient's upper spine is advised as a starting point.

## Summary

EDS results in a tendency toward premature wear of all the major joints in the body, without causing diagnostic tests to become abnormal. Painful joint instability usually responds to conservative treatment. If this is unsuccessful, surgical intervention may help. Successful treatment of EDS patients requires extensive knowledge of anatomy and physiology, as well as treatment options, including surgery, and extensive resources in terms of diagnostic testing, physical therapy, and consultation/coordination of treatment with knowledgeable providers.

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*This article is adapted from:* 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. <http://bit.ly/2Fe5em0>