

Ehlers-Danlos Syndrome (Shoulder Instability)



You may have noticed you have flexible joints, but does this mean you have Ehlers-Danlos syndrome? Read on to find out.

“Ehlers-Danlos syndrome is a group of inherited connective tissue disorders.”

Ehlers-Danlos syndrome (EDS) is an inherited disorder that affects connective tissues—primarily the collagen in skin, joints, organs, and blood vessel walls. Healthy collagen provides strength and elasticity to the structures of the body. There is no known cure for Ehlers-Danlos syndrome, but there are a number of treatment options available to help people cope with this condition.

Symptoms

People with EDS usually have over-flexible (loose or hypermobile) joints and stretchy skin. They may also have other organs in their bodies that are affected, like the heart, blood vessels, bones, bowels, muscles, eyes, and gums. Flexible joints can be unstable and may slip partly out of place (subluxation) or completely out of place (dislocation). These joint separations can occur with little force or can sometimes happen spontaneously. With recurrent episodes of subluxation or dislocation, early-onset arthritis is possible, as well as painful tearing of ligaments, tendons, and muscles.

The two most affected joints in patients with EDS are the knee joint, especially the patella (knee cap), and the shoulder. The hips, ankles, spine, fingers, and joints around the clavicle can also be loose and uncomfortable. If the shoulder is dislocated, it can stretch or tear the capsule and damage other parts of the joint. Once the shoulder has dislocated, there is an increased probability of future subluxations and dislocations.

Causes

Ehlers-Danlos syndrome is a group of inherited connective tissue disorders. In other words, it comes from the genes received from your parents. It is important for patients to understand which subtype of this condition they have (there are now thirteen!)

as each subtype has unique features, including some subtypes that can affect the vascular system (blood vessels), intestinal system, spine, and heart.

Although the overall condition cannot be cured, the various outcomes of the syndrome can be managed. Dr. Romeo recommends an evaluation by a medical geneticist to understand your condition and the potential risks.

Diagnosis

Oftentimes, signs of fragile or stretchy skin, loose joints, and a family history of EDS are enough to make a diagnosis. As part of his assessment, Dr. Romeo uses the BEIGHTON score. Some types of EDS can only be diagnosed in the clinic, but family doctors will usually refer patients to a geneticist for further testing. Genetic tests can confirm the diagnosis and rule out other conditions.

X-rays may also be done to examine the degree of joint separation or arthritis, and MRIs can examine the soft tissues, like tendons, ligaments, and muscles.

Nonsurgical treatment options

The first line of treatment for EDS-related shoulder instability is a dedicated physical therapy program to teach good posture, improved positioning of the scapula during shoulder and arm movements, better

activation of the rotator cuff to help compress the humeral head into the socket, and supportive bio-feedback (which includes the use of shirts or blouses designed to help the patient understand the overall position of the shoulder better). No fewer than six months, and preferably one year, of physical therapy treatment is required before surgical options are considered.

Additionally, people with EDS may use special splints, braces, or compression outfits to keep joints in place and stable throughout day-to-day activities and to prevent painful accidental dislocations. The fewer dislocations, the less damage to the joints and surrounding tissues.

Pain medications are often used by people with EDS to manage the chronic pain of their dislocations, however, many pain medications can come with unwanted side effects, especially when used long-term. Commonly used medications include acetaminophen, nonsteroidal anti-inflammatories (NSAIDs), muscle relaxants, neuropathic pain (nerve pain) medications, and cannabinoids. Sedatives and opioids should be used sparingly and cautiously. Massage, myofascial release, electrotherapy, and heat therapy can also be helpful for pain. It is also advisable for people with EDS to be screened for mental health conditions like anxiety and depression and to receive appropriate care (like medications and therapy) for these conditions when needed.

How surgery is performed

EDS requires a customized approach for each patient. Often, a more ambitious effort to stabilize the shoulder is required than in patients without EDS. Additional efforts to tighten the capsule, combined with patient or donor tissues, may need to be added to increase the likelihood of success. There are multiple shoulder stabilization surgeries possible, including:

- » **Bankart Repair.** A Bankart repair is a procedure in which the torn tissue (labrum and capsule) is sewn back to the socket rim. This procedure is done arthroscopically (via keyhole surgery), using special instruments, stitches, and tiny anchors to connect the torn tissue back to the socket. It is often used in younger athletes with their first dislocation, where there is minimal bone damage.
- » **Latarjet Procedure.** The Latarjet procedure is used when there is damage to the bone of the shoulder socket. It involves taking a small piece of the shoulder blade or scapula bone (specifically, the coracoid) and fixing it to the front edge of the



“It is not unusual for Dr. Romeo to see a patient with EDS who has already had two or more procedures for an unstable shoulder.”

socket to restore the missing bone from this area. The bone is fixed with two screws and sometimes a tiny metal plate. Attached to this piece of bone are two small tendons that reinforce the stability of the shoulder. Finally, the labrum and capsule in this area are repaired with stitches.

- » **Distal Tibia Allograft Procedure.** In some patients with recurrent shoulder instability, the use of a bone and cartilage graft (a distal tibia allograft) may be the best choice. This procedure works well when the bone loss is significant or when patients have had a Latarjet procedure that did not alleviate their symptoms. The graft is fixed to the socket with two screws and, occasionally, a tiny metal (titanium) plate.

Recovery time

Because there are many different types of treatment that can be performed for patients with Ehlers-Danlos syndrome, each patient’s recovery process will be unique to their situation.

Generally speaking, people with Ehlers-Danlos syndrome require extra time to heal after surgery due to their impaired collagen production and higher rates of complications.

Rehabilitation and pain control after surgery have to be in line with a patient’s particular situation and should be developed in coordination with doctors and physiotherapists.

Results

Surgery to treat an unstable shoulder in a patient with Ehlers-Danlos syndrome has a high rate of failure, even in the hands of experts. It is not unusual for Dr. Romeo to see a patient with this condition who has already had two or more procedures for an unstable shoulder. Even if a surgeon tightens the loose shoulder more than usual, the patient may return within nine to eighteen months reporting the repair site stretched out to the same level or worse than it was before surgery.

More advanced and complex procedures may provide longer-lasting outcomes. These include the use of bone to provide more stability since it cannot

stretch out after the procedure, and the use of tendon tissues from a cadaver (allograft material) to reinforce the surgical repair.

Unfortunately for patients who undergo multiple procedures, when the joint becomes too arthritic or when the shoulder remains subluxed or dislocated to the bottom (inferior), a shoulder fusion may be the only remaining treatment to provide some improvement in comfort and function, even though it results in permanent loss of motion. After surgery, some patients still report a slipping sensation because the muscles fail to create compression of the humeral head in the socket.

FAQs

How can EDS affect a patient's shoulder stability?

Due to the impaired elasticity of connective tissue in EDS, the most common problem with the shoulder is too much looseness in the shoulder capsule and ligaments that cannot be controlled by the muscles and tendons, resulting in the shoulder repeatedly shifting partially or completely out of its socket. Over time, or if multiple surgeries are ineffective in controlling the loose shoulder, there is an increased risk of arthritis, pain, and loss of function.

Want to learn more? Find relevant videos, animations, and research material related to this procedure at romeoorthopaedics.com. →



For more information about Ehlers-Danlos syndrome, please request an appointment with experienced Chicago orthopaedic surgeon Dr. Anthony Romeo.

Please visit our website to find out how to schedule your appointment.