

Ehlers Danlos Syndrome

Ehlers Danlos syndrome is a grouping of inherited disorders affecting the connective tissues, such as that of the skin, blood vessel walls and joints. Connective tissues are a complex mixture of various substances and proteins that provide elasticity and strength to all of the underlying structures within the body.

In the simplest of terms, EDS is a hypermobility condition, making the joints hypermobile and lax. Prevalence is around 1 in 5000. However, 5 other forms of EDS exist including “Classical, vascular, kyphoscoliosis, athrochalasia, and dermatospraxis”. The last three are extremely rare.

For those who have Ehlers Danlos syndrome, they have joints that are overly flexible and skin that is stretchy and fragile. This can become bothersome if the individual has a wound requiring stitches because the skin isn’t capable of holding them.

In severe cases of the disorder, the walls of the intestines, blood vessels and uterus can rupture. It is important to speak with a genetic counselor before you make the decision to start a family. Eyes can also be affected.

Ehlers Danlos Syndrome Anatomy

This syndrome is a rare group of inherited disorders, caused by a genetic mutation, affecting the proteins supporting the bones, skin, joints, internal organs and blood vessels. The syndrome is often grouped into six main types of the condition. Even though there isn’t a cure for the condition, the symptoms can be managed and prevent any further injury to the joints, internal organs and skin.

As most EDS is categorized by hypermobility, symptoms typically include being double jointed, or having very loose joints. Typically these joints are prone to sprain, such as repeated ankle sprains, or other subluxations or possibly even dislocations. Skin can be fragile, susceptible to bruising, or hyper-elastic. People with EDS tend to be tall, skinny with long arms and legs, and spidery fingers.

Beighton’s criteria is used to assess the degree of hypermobility:

1. Can you bend your little finger backwards beyond 90 degrees? Score one point for your left little finger, and another point for your right little finger. Total = 2 points.
2. Can you straighten your elbow beyond 10 degrees backwards? Score one point if your left elbow bends backwards beyond 10 degrees, and another point for your right elbow. Total = 2 points.
3. Can you bend your thumb back onto your forearm? Score one point for each side. Total = 2 points.
4. Can you lock your knees out and hyperextend them beyond 10 degrees? Score one point each side if you can. Total = 2 points.
5. Can you bend forwards and place both hands flat on the floor? Score one point.

Total points = 9.

Depending on how many points you score will show you the level of hypermobility in your body, with a 0 or 1 being generally quite stiff (stable), and 7-9 being very mobile (hypermobile/unstable). Any score above 4 is a sign of hypermobility but may not necessarily mean you have EDS.

How to Treat Ehlers Danlos Syndrome:

1. Medication

Discuss with your doctor any medication that you can take. Anti-inflammatories should only be used short term to help relieve flare-ups. Low dose anti-depressant medications may be used to reduce pain, but the individual should look to natural methods as much as possible as this condition is incurable, and taking medication indefinitely may be of concern.

2. Lower Blood Pressure

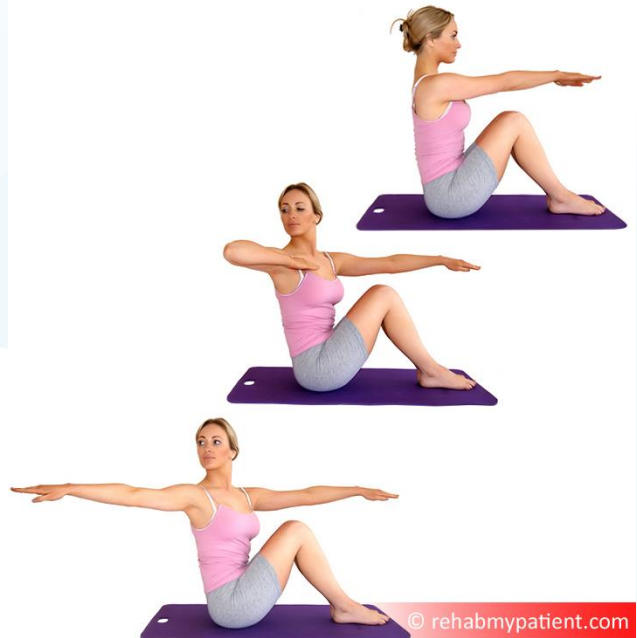
Since blood vessels are extremely fragile in certain forms of the syndrome, you might need to reduce stress on all of the blood vessels by making sure your blood pressure is at an all-time low and within a healthy range.

3. Physical Therapy

Joints that have weak connecting tissues are far more likely to end up dislocated. Exercises for strengthening the muscles that surround the joint will help to stabilize the joint. The physical therapist can recommend a specific brace to help prevent dislocations of the joint. Pilates can also be very beneficial at strengthening the muscles and joints.

4. Surgery

In rare situations, you might have to go through surgery to repair all of the joints that were damaged from the dislocation. Skin and connecting tissues of the joints that are affected may not be able to heal correctly following the surgery.



Tips:

- For those who have a family or personal history of Ehlers Danlos syndrome and are thinking about beginning their own family, you might want to take the time to speak with a genetic counselor to assess your risks.
- Avoid contact sports and various other activities that can leave you at an increased risk of injury.
- To prevent any falls and injuries inside of the home, keep doorways and walkways free of clutter.
- Avoid having a bunch of electric cords and loose rugs, which will lead to an increased risk of you falling and tripping.
- To protect skin that is easily damaged and prevent premature aging, wear sunscreen when outside and use a mild soap.
- For those who have the condition, there is a 50 percent chance that it can be passed on to your children.