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The Evaluation & Treatment of Thumb, Wrist, and Arm Pain & Weakness in Hypermobile Patients with EDS and HSD

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Webinar – August 7, 2019

Video https://youtu.be/vOmY_RgbmOc

[00:00:00]

[00:00:07] **Sarah Jo Ritchie:**

Hello everyone. I'd like to welcome you to today's webinar: "The Evaluation & Treatment of Thumb, Wrist, and Arm Pain & Weakness in Hypermobile Patients with EDS and HSD". My name is Sarah Jo Ritchie, and I am the Volunteer Coordinator at The Ehlers-Danlos Society, and I am your moderator today. This webinar is a part of our ongoing series, "Living with EDS and HSD."

This is how today's webinar is going to work. Webinar attendees will be muted at all times during the webinar. However, you are able to type any questions you may have throughout the presentation into the question box at any time. Dr. Erickson will not be able to see or respond to any of these questions until the Q&A portion at the end of the presentation.

Please do not send your questions more than once. It will not increase your chances of having your questions answered; it will only make it harder for us to sift through the questions and make sure we are able to get to as many as possible.

Dr. Ericson is an ABOS board-certified orthopedic surgeon with hand fellowship training, who has been practicing hand and upper extremity surgery since 1990. Dr. Ericson has been a fellow of the American College of Surgeons since 1993 and a fellow in the American Academy of Orthopedic Surgeons and a member of the American Society for Surgery of the Hand since 1994. Dr. Ericson is a past president of the Massachusetts Orthopedic Association and was the Chief of Hand Surgery at Winchester Hospital in Winchester Massachusetts, from 1994 to 2005, where he was in private practice for 15 years.

After 30 years in the Boston area, Dr. Ericson was recruited to Seattle in 2005, and worked briefly in Woodinville at Northwest Hand Specialists before opening his own practice in 2006. Dr. Ericson is an orthopedic hand surgeon with extensive training in upper extremity surgery with a focus on painful conditions that do not show on standard diagnostic tests, which includes joint instability and peripheral nerve problems. He diagnosed and treated his first EDS patient in 1990 and has been actively involved with EDS community nationally for several years.

He is the co-author of “The Orthopedic Management of Ehlers-Danlos Syndromes” article published in the *American Journal of Medical Genetics* in 2017. Alrighty, thank you so much, Dr. Ericson, for being with us today.

[00:02:26] **Dr. William Ericson:**

Thank you. It’s a pleasure to be here. Can you hear me?

[00:02:29] **Sarah Jo Ritchie:**

Yes, we can absolutely hear you.

[00:02:31] **Dr. William Ericson:**

Okay, so I’m going to speak about thumb, wrist and arm pain, as well as weakness, particularly in hypermobile patients, and everything in this talk applies to hypermobile patients as well as Ehlers-Danlos patients. I’m in the Seattle area, and my practice is skewed towards this particular pattern problems, and I operate on two to four Ehlers-Danlos patients a week, which is more than anybody else I know. The problems are subtle, and they’re just not easy to sort out, but surgery can work under certain circumstances.

- [00:03:11] I have reasonable credentials in terms of background in training, and I got an excellent education and have belonged to numerous organizations, and tried to further my knowledge, particularly in these areas that are difficult to diagnose and treat. I got an award from the Association of Extremity Neurosurgeons in 2009 for some of this work and was a guest speaker at two international hand meetings in the last year speaking about this particular pattern problems. I was also an invited co-author for two book chapters in the last year or two on peripheral nerve compression, and as Sarah mentioned, I was the invited author of this review article, which you can obtain either from me, or through the Ehlers-Danlos Society.
- [00:04:03] I got an award this year from the Arthritis Foundation for knowledge that I have found with regard to the treatment of some arthritis, which has to do with much of this talk today, and just to share with you, the Harvard alumni magazine asked people like me what they liked about what they were doing, and my response was that I had to bring up everything I learned in, my medical training and residency and outside of medical profession and applied simultaneously to patients with Ehlers-Danlos syndrome to come up with a plan like an explanation for the symptoms and a plan for how to improve them, and it has been, a great career.
- [00:04:55] These problems are inherently complicated. This is a painting by Paolo Mascagni, a physician in the 1700s, and it's anatomically correct. And it is amazing how complicated the upper extremity is, and if anything, actually works. And trying to figure these things out on your own, it's impossible. It's not that easy for doctors either. There's no instruction manual from God about how these things work. The orthopedic doctors in general have a tendency to sort of reduce things to simple things like, "Do you need an operation or not?" And if the answer is no, then just move on.
- [00:05:38] If you go to a doctor who has a photographic memory, and they read this review article in *The Journal of Hand Surgery* in 2014, it's an excellent article, but it doesn't offer any tools for how to treat these patients, what operations work or don't work. It just describes what patients with hypermobility are suffering from in general terms. So, I'm going to start at the top of the head and sort of move down the arm and give an explanation, and I urge physicians to look at patients at the whole picture, not just where it hurts, but the context of where it is hurting and how it affects a person's life.

- [00:06:19] So, one of the most useful things I've come up with is this template, which I used to treat people with joint hypermobility, and you see this skeptical woman with the arm that's killing her. And first of all, what happens in Ehlers-Danlos patients, they look fine no matter how much they're suffering. There's no visible indication, but there's a problem, and men in general, tend to be visual. So, this, creates a mis-impression on people examining patients with Ehlers-Danlos syndrome.
- [00:06:52] The neck is usually a problem starting early and becomes progressive, and people start seeking medical care early for this, but the X-rays and MRI tend to be normal. The name for arthritis ankylosing spondylosis and when there's nerve root impingement is called radiculopathy, and antero – and retrolisthesis are when there's excess movement in the frontal plane between the vertebrae, and myelopathy is when there's spinal cord compressions.
- [00:07:21] And then taking care of Ehlers-Danlos patients with vague arm complaints, you have to start at the neck because when you press on nerve roots and because they come out of the neck, you get referred pain where the nerve ends, not where it's pinched. The neck itself may or may not be sore. As you move down the arm, the next issue is the thoracic outlet, which is where the arteries, artery from the heart and the veins from the arm connect, and the nerves from the neck go into the arm, and this area is tight, and it's tighter in women and women who have nervous shoulders, and these problems hurt more in women than men. At least that's my observation.
- [00:08:02] The pressure on the nerves or tension on the nerves and the thoracic outlet causes vague arm pain where the nerves end, not where they're compressed. So, when evaluating a patient with inherently vague intermittent complaints, it would be important to examine them for thoracic outlet issues. The shoulder tends to be unstable and subluxes inferiorly, and it partially dislocates, and the nerves go with the arm, and you can put tension on the nerves as they come out of the neck and go into the arm in a way that causes the nerves themselves to become sore, and patients get burning pain where the nerve ends, not where it's pulled on.
- [00:08:40] Breast weight complicates the situation in women's, in women more so than men and tends to pull the chest forward and down. As you go around the arm, on the outside of the arm, tennis elbow and forearm pain are very common in patients with joint hypermobility issues, and I'll explain my

theory about why that's the case. Proximity to nerve entrapment gives carpal tunnel symptoms that's not carpal tunnel syndrome. It's the same nerve, but when you compress the nerve at the elbow, you get more vague pain that tends to be more in the carpal tunnel where the nerve ends, and the weakness it causes is a very specific weakness that is peculiar in that there are several ways to adapt to it, and the – I believe this is painless.

[00:09:29]

The adaptations cause problems in other joints in patients who are hypermobile. A lot of these complaints get labeled as carpal tunnel syndrome or atypical carpal tunnel syndrome, and then people sometimes have surgery, and it doesn't work. This explains why carpal tunnel syndrome is probably the least common problem the patients with Ehlers-Danlos syndrome have, but it's probably the one that they're labeled with the most. The same physiology that causes carpal tunnel syndrome causes pain along the flexor tendons in the palm, and that tends to be activity related and improves with rest.

[00:10:07]

The thumb is unstable in patients with connective tissue disorders, and it can become painful long before there's arthritis, and untreated instability leads to arthritis, which affects one-third to one-half of women by age 55. It is a very common problem. It is completely underestimated, underappreciated by the medical profession, particularly men where this is a relatively rare problem. The wrist is unstable in patients with hypermobility issues, and it's very easy to make an unstable wrist unstable and painful by falling, and arthritis tends not to happen unless there's actually a ligament tear, but the ligaments are stretchy, and they tend to stretch more rather than actually tear.

[00:10:55]

The weakness caused by proximity to nerve entrapment is extremely frustrating. It makes it difficult to open eggs in the kitchen, turn keys, pull zippers up, put buttons through small holes, and fold laundry. The pinching is impaired, and it doesn't harm people. It just causes frustration. The frustration tends to affect posture eventually. These problems are all worse if people are overweight. The problems are worse as you get older. The blood supply to the nerves diminishes, and the tissues around the nerve, I believe, become tighter, and patients who are under sustained stress, and it can be physical or emotional stress or financial stress get tired or depressed, and these feelings when expressed physically, tendon affects the posture, and the posture tends to affect the nerves by the shoulder dropping forward and down and pulling the nerves with it.

- [00:11:47] So, with this pattern problems people with vague intermittent symptoms that come and go seemingly at random, but it can be damned inconvenient if they're all acting up at the same time. They can cause enough pain that it is distracting, so people have a hard time concentrating or performing tasks for employment or caring for kids, and other times the symptoms are minimal if any. If you're well-rested, and your posture is good, and your palms are up, the patient may have minimal symptoms.
- [00:12:23] So, a lot of patients with Ehlers-Danlos syndrome, probably almost all of them, and many patients with hypermobility issues have headaches, and it would be important to get an MRI at some point because the headaches can be caused by a Chiari malformation, and that's something that needs an operation, and the headaches can be one of those additional problems that affect the posture and make the nerve symptoms worse.
- [00:12:48] The nerves that come out of the neck and go into the arm and specific nerve roots, there's a distribution, and when you pinch the nerves in the neck as they come out, you get pain where the nerves end, not where they're pinched. So, the disks between the vertebrae: this is an MRI of the cervical spine, lateral view. There are shock absorbers between each of the vertebrae, and those shock absorbers get cyclic loading, and they flatten, and they don't flatten like a beer can. They flatten like a basketball being stepped on, and they bulge, and the bulging discs can press on the spinal cord and cause myelopathy, or if they're off to the side, they can press on the nerve roots and cause big arm complaints.
- [00:13:30] These are the distributions of where the arm pain tends to be from a neck source. So, when evaluating patients with joint hypermobility issues with vague intermittent complaints that include some wrist/arm pain, it's important to keep the neck in mind as a source of pain because these are often overlooked, and patients with thumb problems often have neck problems around the same time for the same reason that the loose joints wear prematurely, and it is very common to have a person have overlapping symptoms in this pattern.
- [00:14:10] So, the thumb is sore because it's arthritic, and the neck is stiff because it's arthritic, and the most common nerve root impinging into the sixth cervical nerve root, and it causes overlapping pain. It can constantly pain in the same distribution of the thumb, and if you can, and especially for this, and explain all the symptoms, the prognosis with intervention when

you know what you're going to actually get out of the surgery is much clearer, and patient satisfaction is higher.

[00:14:43] The thoracic outlet is a big issue in patients with hypermobility issues, and that is the space between these muscles where the nerves come out of the neck and go into the arm, and they are accompanied by an artery from the heart and the vein coming back from the arm, and this area is tight, and it's tight in everyone. But patients with loose joints tend to have tighter muscles, and if you pinch the nerves in the region between the shoulder and the neck or pull on them, it causes vague arm complaints.

[00:15:16] So, generally speaking, by the time a patient comes to my office, almost 100% of them have easily provoked numbness in the arm with tests for thoracic outlet syndrome, and the first treatment for patients, generally speaking, is to try to improve their posture and eliminate this particular source of arm pain from their pattern of problems. This patient on the left has no arm pain, and I have a model in my office, which is the base of the skull and the neck and the thoracic outlet, and on the left side when he has good posture, this is what the nerves look like coming out of the neck going into the arm.

[00:15:55] And on the right side, this is how this patient sits when he's playing video games and not paying attention to his posture, and after a while, this causes vague discomfort in the arms that radiates proximally, and this particular pattern complaints is ignored or missed by most of my colleagues, but it is a very common problem particularly in patients who have chronic wrist, thumb or arm pain and are frustrated seeing physicians without answers.

[00:16:28] Thoracic outlet surgery is inherently risky and is generally speaking to be avoided. The main techniques for doing this include rib resection or removing the interscalene muscles. Rib resection is probably a better operation, but it's hazardous, and it can cause death, as well as a minor complication such as hemothorax or pneumothorax, and the nerves are at risk for injury also. The scalenectomy can make the head unstable, and cutting stabilizing muscles in loose-jointed people is generally speaking, probably not a good idea. It may be necessary, but it's a last resort type of approach.

[00:17:16] We have this other problem where we have gone from being upright and leaning forward and twisting our arms, and these nerves as they go from the neck to the hand are not designed to stay in this position. There's also

DNA evidence that the human form came from the sea, and if you look at sea lions, they actually have a hand and a wrist and a forearm with radius and ulna and humerus and scapula, and the shoulder joint in that animal is designed to be loaded partial weight-bearing and pushed inward, and we have become upright, and the shoulder's become an attraction joint, and we tend to lean forward and twist our arms, and that causes the pattern problems and pain in the picture on the right.

[00:18:03] The shoulder itself in Ehlers-Danlos patients is always loose, and there are specific exercises that can be done to make the shoulder stable, but it's my current hypothesis that patients with loose joints have soft nerves, and there are nerves that go through tunnels that are compromised by the tunnel in patients with loose joints, and one of those nerves is probably the suprascapular nerve that goes to the supraspinatus and infraspinatus muscles, and if my theory is correct, this would explain why some patients can go to therapy for years and still not be able to stabilize their shoulders because the nerve to the shoulders stabilizers may not be working correctly.

[00:18:43] Capsular stabilization, the surgery is a viable operation, but it's difficult in my experience to find an orthopedic surgeon who will willingly operate on multi-directional instability in a patient with a collagen disorder, and I'm not sure why that is, but it is, it's definitely a problem trying to find a provider who's willing to take the risk of a stiff joint or a failure. There are tissue augmentation devices now available that are likely to make this operation more predictable in patients with poor connective tissues that weren't available until relatively recently.

[00:19:22] As you go down the arm, elbow pain is a fairly common problem in Ehlers-Danlos patients. When I gave the talk in Baltimore, I think there were 500 people in the audience, and when I asked how many people had pain in this general area, about 500 people raised their hand. The extensor carpi radialis brevis tendon attaches at the elbow, and it's possible to make that muscle large enough that it actually pulls off the bone.

[00:19:50] This is what it looks like on the MRI on the right-hand side when it does pull off the bone, and it can be reattached. The reason it falls off the bone is something that is part of what I figured out. This is what it looks like to make an incision and reattach the muscle to the bone. The ECRB, extensor carpi radialis brevis muscle, hypertrophies, and it can get so big it actually pulls off.

- [00:20:20] My theory is that the radial tunnel and lateral epicondyle pain is caused indirectly by compression of the median nerve at the elbow. The median nerve comes across the elbow and pierces the pronator teres muscle, which you see in the bottom drawing. This compromises a small branch called the anterior interosseous nerve, and that nerve goes to the thumb and index finger flexors. That weakness can be adapted to by wrist extension, and the extensor carpi radialis longus and brevis are the muscles that pull that wrist up.
- [00:20:56] The extensor carpi radialis longus attaches directly to bone, but the brevis attaches to a layer of fascia, and the fascia rips, and when it rips, it swells, and when it swells, it hurts, and unfortunately, it hurts right over the radial nerve, which becomes the posterior interosseous nerve, and pain in this area generally speaking, when treated by an orthopedic surgeon, gets treated as a nerve problem when it's actually mostly tendon problem.
- [00:21:23] The fact that it's treated as a nerve problem makes people order nerve tests, and when the nerve test is normal, then they say, "Well, it's not that," but tendon problems don't show on a nerve test, and it can be a tendon problem and a nerve problem but not something that shows up on a test. If you cut the ligament that presses on the median nerve, that part of the nerve works normally, immediately, and then patients don't have to adapt.
- [00:21:49] And if you cut the deep fascia of the extensor carpi radialis brevis, it stops hurting, and probably 95% of patients have 95% relief of pain if you cut that fascia and decompress the nerve. If you just decompress the nerve, which is what most of the books about this particular topic describe, the success rate in [inaudible] series is between 50 and 60%. That is not something to look forward to if someone proposes a surgical procedure.
- [00:22:25] You can expose the radial tunnel through a lateral forearm incision, which I used to do this incision. You can just hack through the extensors themselves. This is a patient who's operated by somebody famous in Seattle that didn't work, and she was hypermobile, and she came to me off working on narcotics and was told she was a bad healer. On hypermobile patients, if you make this incision, you will always get a disfiguring scar, and this was done the way the book's described is compress the nerve but not addressing the extensor carpi radialis brevis fascia. I redid her surgery through this incision with relief of pain, off narcotics, and back to work, and sending me all her friends and relatives.

- [00:23:12] This is in a patient who had the nerve decompressed, which did not work. The patient had the same symptoms in the recovery room that they went to sleep with, and I redid his surgery through an anterior incision. You know he's hypermobile because of the width of the scar. The median nerve goes across the elbow, and as the anterior interosseous branch come off, and it goes to three muscles and a large sensor branch to the wrist bones. So, pinching this nerve at the elbow causes weakness, particularly opening things in the kitchen, peel packs, potato chip bags.
- [00:23:49] If it gets worse, it causes aching pain in the carpal tunnel because that's where the nerve ends, and this gets mistaken for carpal tunnel syndrome because the pain is in the carpal tunnel, not where the nerve is pinched. This is a drawing from the Mayo Clinic about the terminal branch of the anterior interosseous nerve, and it's quite large, and unfortunately, it is in the carpal tunnel, and pinching the nerve at the elbow causes aching pain in the carpal tunnel that can be severe at times. So, one of the hazards being hypermobile is getting your carpal tunnel operated on for intermittent, severe wrist pain in the carpal tunnel when the problem's at the elbow.
- [00:24:34] Carpal surgery done in my hands endoscopically has almost 100% success rate and a very little scar. The proximity nerve entrapment is probably a safer operation with a quicker recovery, but cutting that fascia restores the strength of those three muscles and eliminates the referred pain in the recovery room.
- [00:25:00] So, these are some patients with weakness of the muscles innervated by the anterior interosseous nerve who had electrical tests, which were completely normal, and this video demonstrates the weakness.
- [00:25:13] [*crosstalk*]
There's no strength here, and this one, I can't budge.
- [00:25:18] This is outside the operating room, and this is a half hour later in the recovery room.
- [00:25:22] A bit hard.
- [00:25:22] **Patient:**
Yeah.
- [00:25:24] **Dr. William Ericson:**
Oh, hard as you can.

[00:25:24] **Patient:**

[inaudible]

[00:25:25] **Dr. William Ericson:**

What do you think?

[00:25:29] **Assistant:**

Very good.

[00:25:29] **Patient:**

Yes.

[laughs]

I think it hurts.

[00:25:30] **Dr. William Ericson:**

Push the thumb down like that.

[00:25:34] The thumb comes back also. So, the first person was operated for intermittent severe [crosstalk]. This patient has pain in the radial tunnel.

[00:25:38] [crosstalk]

Okay. Pull down hard with your fingers. Just try to pull down hard as you can.

[inaudible]

This finger though, I can't budge it. About hard as you can. Your thumb too. Pull your thumb down like this.

[inaudible]

[00:26:09] Good. Let's do it one more time, and pull as hard as you can pull, the whole strength.

[00:26:16] **Patient:**

Very much.

[00:26:17] **Dr. William Ericson:**

Good, and the thumb. Pull your thumb down.

[00:26:18] **Patient:**

Excellent.

[00:26:23] **Dr. William Ericson:**

So, this patient, watch what he does with this thumb. I have to pull his thumb straight down.

Oh, let me straighten that one out.

Look how weak he is.

This middle finger is quite strong. Shake your thumbs.

See what he did with his thumb?

Okay. Hold out your hand. Push down your thumb just like that.

This is an adaptation, which I had to block.

[crosstalk]

In the recovery room, I can lift him off the table.

[00:26:48] **Off-Screen Speaker:**

Oh, I found my, the big – my biggest center.

[00:26:48] **Dr. William Ericson:**

Okay. Okay. Relax your hand. Make a fist for me. Make it tight first and pull down hard as you can. Good.

[00:27:03]

The weakness people adapt to, and one of the ways to adapt is to bite pill containers. So, prescription pill bottles are extremely difficult to open. So, people leave them open and get the non-childproof containers. The weakness is profound in patients who are hypermobile. So, this problem affects virtually 100% of patients with hypermobility issues. The compression of the nerve doesn't damage it. I think it's a combination of low pressure over large surface area and actually deflection of the nerve, and as soon as you cut that fascial band, the nerve is perfused normally and conducts normally.

[00:27:48]

The technique involves cutting the bicipital aponeurosis, which is the structure attached to the biceps that pushes the pronator teres muscle into the median nerve and then cutting the fascia of the ulnar origin of the pronator. And nobody really needs either of these structures, and there's no healing, and the nerve works normally as soon as this is done. In the operating room, this is what it looks like.

[00:28:13]

So, at the top of the image, there's the artery and then the venae comitantes and then the median nerve, and that weight triangular thing

in the middle there is the fascia of the ulnar origin of the pronator teres muscle to wrapping around the median nerve, which is the thing that looks sort of like a shrimp colored to, to the right. That little band gets lifted off the nerve and cut, and the ends pop apart, and then the anterior interosseous nerve works normally immediately.

[00:28:44]

You don't have to make a big incision to expose this. Right now, it's about one inch. The patient's right arm has had the surgery. You can see that there's no dent in the forearm from the bicipital aponeurosis where there's a large indentation on the left side, patient's left side. This is a patient with Ehlers-Danlos syndrome who had an operation by somebody else that didn't really work and was – the doctor would probably conclude that they shouldn't operate on Ehlers-Danlos patients because the surgery doesn't work, and you get a big ugly scar. But this is me redoing the surgery with relief of your symptoms and sending me all her friends and neighbors.

[00:29:29]

This is the person who had an anterior interosseous nerve palsy with a problem above the elbow but got an operation in the forearm and is a hypermobile person who has a hypertrophic scar which – and it was unnecessary. This is an Ehlers-Danlos patient who had their left side done a couple months ago and then the right side. Scars tend to be a little bit wider. They also had their wrists operated on for wrist instability that was painful. People who have this weakness may look like this when they write.

[00:30:11]

The person shown here had their nerve fixed on the right-hand side, and, and in recovery room, pinched like this. Previously, before surgery, they pinched like it's shown on the left side with both hands, but this happens when you restore the strength of this, these muscles by fixing the nerves. The results are immediate. Changes have people pinched in the recovery room. There are four ways to adapt to the weakness, and you don't get to pick which way you adapt. You can use the next muscle down in which case people get arthritis at the end joint of their thumb on the outside, bone spur or synovitis, and sometimes arthritis at the index finger end joint.

[00:30:59]

People who pinch sideways either stretch the ligament at the knuckle of the thumb or cause increased instability and then subluxation at the base of the thumb. To adapt by pinching sideways against the index finger, get a bone spur at the index finger metacarpal phalangeal joint. That can become painful, and patients who pull up the wrist, get tennis elbow in a non-tennis player. The looser the joints are and the weaker the nerve and the more discreetly the person adapts by one of these four ways, the more

likely they are to have a soft tissue failure, and it's my observation that the looser the joints, the weaker the nerve, and it seems to make sense to me that the nerve is going to be softer with stretchier collagen and more vulnerable to compression.

[00:31:51] So, these things are not linear processes, and I have a device that I'm working on to quantitate this, which I think will show this with hard numbers that the degree of laxity is in fact correlated with the degree of weakness, and that is one of those confounding things if you're the patient, why these things would be so different in use on other people.

[00:32:17] I took a graphics course and came up with this flowchart. The main problem, and this affects all patients with hypermobility issues is the median nerve at the elbow, and there are sensory consequences and motor consequences. The sensory consequence is that it hurts where the nerve ends, which is really the carpal tunnel and wrist, not where it's pinched, and it hurts with a palm in a down position, particularly mouse or keyboard or driving or writing.

[00:32:45] The motor consequence is two muscles that one bends the end joint of the thumb, and the one that bends the end joint of finger, and it changes how people pinch without telling people why they're doing it that way, and you can pull the thumb in or use ligaments to give resistance or pull the wrist up. In the dark box on the right-hand side, the four things on the left-hand side are all painful and don't show on tests and tend to be universal in patients with Ehlers-Danlos syndrome, and the items on the right showing X-rays and tend to get treated.

[00:33:22] Items on the left tend to get ignored or test ordered and then patients reassure that there's nothing wrong because the test is normal. You can download this. Some of these images are available on my website, which is <http://ericsonhand.com/>. This is an unstable thumb. I am almost dislocating this with minimal pressure, and on an X-ray, it would look like this, where the thumb metacarpal is not lined up with the trapezium.

[00:33:53] And if this is painful, it will typically progress to arthritis. This is the Eaton classification of thumb arthritis, but the bone starts out located and loose, and then it migrates, and as it subluxes, the load increases and then a cartilage wears off prematurely, and this is a problem that affects at least one-third of women and perhaps 50% by age 55, but it, it is an issue in almost all patients with Ehlers-Danlos syndrome.

- [00:34:27] If the patient is evaluated prior to cartilage damage at the base of thumb, it is possible to stable the thumb using a tendon to create a new ligament, and this is the Eaton-Littler ligament reconstruction that was invented in the '60s and published in 1973, and I do about 150 of these a year, and, but I don't know anyone else in the Seattle area doing this operation regularly, and I'm not sure why that is, but it is a good operation even in patients with connective tissue disorders with very few failures.
- [00:35:06] The ligaments are not strong enough, but the tendon in a non-weight-bearing joint can be used as a ligament even in patients with connective tissue disorders. This is what the scars look like. They're a little bit smaller these days. This is an Ehlers-Danlos patient who also had a small bone in the wrist movement called the pisiform. This is what the scars look like later on. Sometimes, a ligament at the knuckle...
- [00:35:42] Okay. Sorry for the interruption. I had a slide that showed technical failure here, but I took it out.
- [*laughs*]
- I should have left it in. So, I started to say the ligament at the thumb knuckle can just stretch, and if you try and repair this, the failure rate is about 100%. A fusion would be necessary for that, and there are multiple ways of performing that particular operation, but generally speaking, we try to avoid fusions in Ehlers-Danlos patients because they tend to cause increased load at either end of the fusion site.
- [00:36:18] If you come in with a joint destroyed like this, you obviously can't do a ligament reconstruction, and there's a salvage procedure that I do where I take out the bone that removes the interface between the thumb metacarpal and the trapezium, and use a tendon to suspend the bone, so it can't migrate, and that works reasonably well. It does not work as well as the thumb that god gave you, but it's reliable for probably 80% relief of pain in most patients, at least 80% relief of pain, and probably at least 80% of the pre-arthritis strength of the thumb. And if that sounds good, it can be a great operation.
- [00:37:02] This is what the X-ray looks like when the bone is removed, and that's a titanium bone anchor holding the tendon to the bone to keep it from migrating. You get a slightly smaller incision without operation. Patients who pinch sideways against an exterior can get a large bone spur at the index finger MP joint radial collateral ligament, and this can be painful. If

you operate on this just by itself, and the orthopedic literature describes an extremely high failure rate, basically advise us not to operate on it.

[00:37:35]

If you take the bone spur off and reattach the ligament, that's the collateral ligament is attached to the spur, and fix the nerve, the anterior interosseous nerve at the same time. This operation works fine. Sometimes, the end joint of thumb just bends backwards all the way, and that can lead to arthritis on the top of the thumb near the thumbnail or arthritis on the bottom of the thumb where there's a small bone inside the tendon.

[00:38:06]

If there's arthritis at the top of the thumb, and it's sore when you push on it but not when you load the joint and move it, you can just make a small incision through one of the creases and remove the bone spur and the swollen lining of the joint with a quick recovery and immediate relief of that pain. If the pain is on the bottom side, and there's clicking, there's a little tiny bone called a sesamoid. It doesn't always show on X-rays, and it should have cartilage on it.

[00:38:33]

It's about the size of Arborio rice, and it can be quite a nuisance, and it causes painful clicking at the end joint thumb, which almost always gets mistaken for a tendon problem at the next joint down. Doesn't leave much of a scar to take that, that bone out. This is the pisiform, which is a bone inside a tendon, and it's loose in patients with hypermobility issues, and it's very easy to make it loose and painful.

[00:39:00]

There's no treatment for this other than taking the bone out. It is an accessory bone that you do not need, and you can make a little incision over it and then peel the tendon off. It's a simple operation in this sense, but the ulnar nerve is right by the bone, and you have to be extremely careful doing this operation if you're going to use a small incision.

[00:39:23]

At least almost no scar if you do it this way. This is a patient, an Ehlers-Danlos patient with their pisiforms removed, and this is probably about two months out on the right and maybe six months out on the left. The small finger, the index finger, and the thumb all have two extensor tendons going across the joint, and these tendons can stretch away from each other, and they can snap back and forth, and when the tendons go sideways, it can cause surprisingly strong pain in that area but look normal, and there would be no swelling, and it's a very subtle diagnosis, but there's no non-surgical treatment for this.

[00:40:05]

You can expose the two tendons and tie them back together, and that is a reliable operation, and in Ehlers-Danlos patients, I've done all the tendons

for painful snapping, and the recurrence rate is extremely small when done this way. It is a bit of a MacGyver-type operation where you actually have to move the fingers and make sure that the tendons are in balance, and it's just not a simple cookbook thing, just go in there and trim something.

[00:40:38] These tendons are all connected sideways, and they interact with each other. So, when you tie one down, you affect the adjacent tendons, so it's not something to just take lightly even though it's a small operation, but these are attendants that when this patient made a fist before surgery, they all popped off into the grooves, and no casts, no therapy. There are special techniques – related issues to try and decrease the amount of adhesions people get when you do this operation.

[00:41:15] In the thumb, the extensor pollicis brevis gets moved to the next bone over, so it pulls the bone up instead of hyperextending the joint, and then the extensor pollicis longus gets moved over into the central location. And this is an Ehlers-Danlos patient that had a previous thumb base of the thumb ligament reconstruction that then later on a couple years developed painful subluxation of the extensor pollicis longus tendon, and this is what the scar looks like.

[00:41:50] Flexor tenosynovitis is pretty common in patients that often do hand exercises because the weakness and in patients with scar tissue, it causes aching pain along the tendons, and these can be exposed through a small incision in one of the creases, and the sheath can be opened up and the lining of the tendon removed with appreciable relief of symptoms with that.

[00:42:14] Instability between the radius and ulna is a less common problem but can be treated surgically. This is an Ehlers-Danlos syndrome patient with wrist instability and radial ulnar joint instability treated surgically. The preferred method of the first time around is to take the extensor retinaculum that goes over the joint and to cut it and then refit the pants over best fashion, and that works most of the time.

[00:42:43] If that stretches out and the person gets recurrent instability, there's a more invasive operation where you can take a tendon from someplace else, and drill holes through the bones, and pull the tendons through the holes, and reestablish the ligamentous constraints of the radioulnar joint, and that is a reliable operation even in Ehlers-Danlos patients.

[00:43:06] This is my first Ehlers-Danlos patient who I operated on for thumb instability in the early '90s, and I did one thumb and then the other thumb, and then I operated on her wrist instability and did one side and

then the other. Then I took out her pisiform on one side and then the other, and then I did her median nerve in the forearm in one side and then the other, and then I did her radial tunnel, and she did fine with each of these operations.

[00:43:31] Eventually, she felt painful instability at the CMC joints of her ring and small fingers, and these days, I would treat them with prolotherapy, but back then, I didn't know very much, and I fused them which – and she did fine until she went to a funeral, and then someone grabbed her hand and broke off the fusion masks, which I then had to fuse to the next one over, and she did fine with that.

[00:43:56] But these days, the patients tend to come in with all these things bothering them at once, and back in those days you know, we just, I just operated on the thing that was the most painful at the time, but she had each of these operations that I'm describing. Getting back to this template, the treatment I recommend is to look at the whole person and to evaluate them for where they are in the arc of life and try and assess for what kind of stress they're having in their daily life and not tell them to have less stress but offer ways to cope better with the stress that they're going to have because sometimes, people may just have two bad choices, and they have to figure out which of the choices is less painful or less stressful, and that may be the best they can do, and I would try and assist them with that.

[00:44:54] The surgery that I do on patients in these circumstances works reliably if patients have good posture and their shoulders are not unstable. It is less reliable in terms of changing functional status if the patients have poor posture, and their shoulders are loose. Sometimes, these problems below the elbow are so painful that patients can't do the exercises they need to, to correct their posture, and in those occasions, we do operate on people without good posture and without stable shoulders, but we try to address the thoracic outlet part first.

[00:45:35] And the main goal in patients who have significant thoracic outlet issues is to try and make them as manageable as possible, which generally speaking would be as strong and pain-free in hand as possible from the elbow down. So, typically, anything that was sore from the elbow down that could be fixed, I would try and fix so that the patient had the best chance of maintaining the posture.

[00:45:59] The majority of patients who still have significant functional limitations in terms of employment or using keyboards or working with their arms

is typically the thoracic outlet, so we try and identify that early. Another problem is that anxiety is a core finding in patients with loose joints and hypermobility, and if they start seeing doctors, they have what I would describe as a post-traumatic stress disorder where they don't – they get to the point where they don't want to go see a doctor because they know it's going to end badly that they're going to sit and wait, and then they're going to explain their vague intermittent complaints, and then they're going to get challenged or not respected for what's going on.

[00:46:51] Then may have some tests ordered, and then the doctor may come in smiling, "The MRI's normal." The patient, his fears are then realized, and they know they're going to have to find some other solution to this, and that it was a waste of time and money, and the doctor just thinks they're nuts, but the doctor is making them nuts. So, this is the patient population tends to wind up in my office, and this explains the majority of symptoms that can be addressed.

[00:47:23] And patients, generally speaking, do well when they are motivated and they have adequate support. The indication through surgery in general would be when things are too painful to go on with whatever has been done so far. It's helpful if the diagnosis is clear, and that is rarely the case in Ehlers-Danlos syndrome patients. They usually have more than one problem, and they often overlap in terms of where the symptoms are expressed.

[00:48:01] Not every doctor knows about these options. There are, I think, about 1,000 hand surgeons in the U.S. with my level of training, and they did not all get the same education that I got. So, I don't know what else doing capsular stabilizations of the wrist for the wrist instability that other Ehlers-Danlos patients have, and I don't know anyone doing as many thumb ligament reconstructions as I do, and some people, some hand surgeons are just not aware that these are our options.

[00:48:33] The other problem is the surgery. It's just an inherently risky thing to do, and you may not know where you are on this thing called Scott's parabola, but surgical techniques come and go, and some new idea comes out, and then bunch of people do it, and the results are good, and then with careful follow-up, the results turn out not to be so good, and then it turns out maybe it's not a good idea to do at all.

[00:49:00] For example, tuberculosis wasn't really understood in terms of being an infectious disease, and there was a period of time where if you had tuberculosis in your lungs, they would remove your lung, which got rid of

the tuberculosis but also got rid of your lung, and these days, no one would remove a lung for tuberculosis. They would give you antibiotics, but every surgical procedure you know is at risk for being on this curve somewhere.

[00:49:26]

So, if someone comes out with some new idea, I would generally wait and see if it really is a good idea because the history of surgery is full of operations that people thought were great and then turn out not to be so great. Sometimes, the goals are very clear and realistic, and that happens when you have a good doctor who's able to communicate and a patient who's able to understand, and if you are not communicating well with your doctor, it's probably not a good idea to have surgery.

[00:50:08]

The risks of surgery are clearly higher in patients with Ehlers-Danlos syndrome. The wounds, in my experience, you know, and any problems with healing, scars tend to be a little bit wider, but I did show you some examples of huge scars with surgery that didn't work, and the problems – there are causes that are multiple in patients with Ehlers-Danlos, and they're not the patient's fault. If you have one of these problems that's straightforward, that part can be fixed, but the doctor and the patient may be under the impression that that was the only problem, and it is very easy to undergo surgery, and it just has marginal improvement, and it frustrates everyone.

[00:50:52]

The other issues with Ehlers-Danlos syndrome is they tend to get dizzy when they stand up quickly, and the ankles are unstable, so people fall routinely, and it's very easy to make the thumb and the wrist that were already loose to make them loose and painful, and the normal things that people take for this like aspirin tend not to be an option for patients with Ehlers-Danlos syndrome because they almost all have gastroesophageal reflux.

[00:51:17]

There are also hormones that circulate to make the joints looser at different times of the month, and things swell differently at different types of the month. Just makes it very complicated if you're the patient. And even simple things like having a trigger finger released, patients with cognitive disorders are often resistant to Xylocaine and bupivacaine. So, they go to the dentist, and the local anesthesia doesn't work. That's a clue that they might have a connective tissue disorder and worrying about all these things and facing them when they are invisible to everyone else gets people to be pretty tired and after a couple months, people get grouchy.

- [00:51:54] It's hard on couples. Surgical procedures that are fairly routine that should be approached with caution particularly in Ehlers-Danlos patients, carpal tunnel release. You know, carpal tunnel syndrome is common, but that's usually not the problem in patients with carpal, with Ehlers-Danlos syndrome. It's usually the median nerve at the elbow, and they tend to manage the symptoms by changing how they do their activities, and that works reasonably well until they get carpal tunnel syndrome on top of it.
- [00:52:27] Then the carpal tunnel gets recognized and operated on, but the problem that has been there the whole time intermittently is still there, and then patients and doctors are disappointed. The thumb collateral ligament repair tends to fail unless you fix the nerve at the same time, and the forces are high enough that the ligament reconstructions are not reliable, and fusion is probably better.
- [00:52:49] The index finger collateral ligament bone spur I talked about where the failure rate in most literature is extremely high. Stabilization procedures for hyperextension of the proximal interphalangeal joints are just fraught with hazard. The tissues stretch and recurrent instability's a problem. The risk capital stabilization works well. In this patient population, I've had very few recurrences, but patients do get tendons operated on, like de Quervain's tendonitis. The nerve from the elbow, the median nerve, causes pain at the wrist, and this gets mistaken for de Quervain's, and people have surgery that can easily make them worse.
- [00:53:29] Taking bones out can cause more instability in patients with connective tissue disorders that cause other problems. The radial tunnel surgery is predictable if you know the person's posture is good, and they don't have thoracic outlet symptoms, but that is not how most of my colleagues look at this. They look at it as a nerve problem when it's actually mostly a tendinosis.
- [00:53:51] Ulnar nerve surgery inherently hazardous in Ehlers-Danlos patients. Most of the patients with Ehlers-Danlos syndrome has have ulnar nerve symptoms, but they tend to be from the thoracic outlet and operated on the ulnar nerve. The elbow can easily make the nerve problem worse. The – we talked about the shoulder stabilization is just inherently difficult for the doctor than the patient, and thoracic outlet surgery can cause many more problems. So, it's just not something to take lightly. So, it has been my pleasure to take care of patients with these problems, and I feel fortunate to have figured out a few of these issues.

- [00:54:32] And you're not alone if you're hypermobile, that these problems affect a huge number of people, particularly women. The – there are treatment things. This is a video of a patient of mine. Typical hypermobile patient.
- [00:54:49] So, [*inaudible*] patient today with joint hypermobility issues such as wrist pain and thumb pain, and the thumb pain is at the base of the thumb, and this is an example of the base of joint instability. You can see the bone can move almost a centimeter, and it also moves outwards a centimeter, and this is loose and painful. The other thing she has is wrist instability, and she has full motion in the wrist, and this is the scaphoid. And if I move her wrist into radial deviation and push under scaphoid, you can see the wrist dislocate, and that is painful.
- [00:55:28] Full motion, but a mechanical instability, and this would have a normal X-ray, a normal MRI, a normal bone scan, a normal arthrogram, a normal arthroscopy, but abnormal motion under load. So, let's check the other side. So, she has had surgery to correct these problems on her right side, and you can see there's not much in the scar. Part of the tendon is taken here and passed underneath the skin at this point and then passed underneath these muscles, and with the same exam, the doesn't really move this way, and this plane moves just a little bit, which is normal, but quite snug, and where the joint is reduced and loaded, the, the motion is smooth and painless.
- [00:56:17] The wrist was stabilized for a small incision here. You can barely see her scar, and by the operation, she has basically full motion. I'm going to push on the scaphoid here. The wrist does not dislocate. It is stable and solid. She also had a hand pain and had a weakness secondary to proximity to nerve entrapment, and that gives you weakness in these two muscles if we put her fingers like this. Pull down as hard as you can. I can't budge this. These two are the same strength.
- [00:56:51] And her thumb. Put your thumb down like this. I can't budge. And if we examine her other side – make a fist for me. You can see a little indentation in her arm, which is where the bicipital aponeurosis pushes the flexor pronator muscles in. If we put her hand [*inaudible* 00:57:13] hand in this position, these two should have the same strength, and her little finger I can't budge. Pull down hard as you can. Here's another strength on her index finger and then her thumb.
- [00:57:18] Put your thumb down like this. Pull it hard as you can. There you go. No strength at all. Now, this is a confusing weakness because people could

adapt. You have to understand that to adapt, really, the adaptation is on the wrist up, and with her wrist up, now, again, this finger is quite strong, and the thumb is quite strong, but people who do this all day long who are loose-jointed gets sore right here. You can say, "Ow."

[00:27:50] **Patient:**

[*laughs*]

Ow.

[00:57:23] **Dr. William Ericson:**

And she had this operated on this side and resolved the pain, and she had [*inaudible*], which was treated at the same time, and she has the same problem on this side. Here's the [*inaudible*] right on this side.

[00:58:08] **Patient:**

This is just so – I mean, sturdy, and, and reliable, and this one just depends on what I'm doing.

[00:58:14] **Dr. William Ericson:**

So, I want you to...

[00:58:19] **Dr. William Ericson:**

Anyway, that's the conclusion of my presentation. These are common problems. They can be treated. The surgery is predictable if you operate on patients with good posture and stable shoulders. It is less predictable, but it can work if you operate on patients who don't have good posture and don't have stable shoulders. Recurrence is though the – this requires a knowledgeable surgeon, a good anesthesiology department and special equipment in the operating room to do these operations through small incisions and an assistant who has good endurance with their owners.

[00:59:06]

It can hold very still to do these operations through small incisions, and unfortunate to have those circumstances where I am right now, and I'm happy to share this information with anyone. We would like to hear about it. So, that concludes my talk.

[00:59:23] **Sarah Jo Ritchie:**

All righty. Thank you so much, Dr. Ericson, for a fantastic and informative webinar. Now, we already have a few questions in, but just as a reminder, you can still type any questions you may have for Dr. Ericson in the question box, and we'll try to get to as many of them as we can. Our first

question, “is the anchovy surgery appropriate for EDS patients whose thumbs continuously sublux?”

[00:59:48] **Dr. William Ericson:**

Well, the anchovy procedure would be done when there's cartilage damage, and it's similar to the operation I do. I actually used to do the anchovy procedure, but the last couple that I did were in hypermobile patients, and they totally fell apart. So, I stopped doing the anchovy procedure at approximately 2003 or 2004. Let's say, 2004, I stopped doing the anchovy procedure.

The – you have to look at the whole picture, and if the person has arthritis in their neck, and they have gastroesophageal reflux, and they have bunions, and they're falling apart, the tissues around that joint are not likely to be reliable for holding the anchovy in place. The – when I moved to Seattle, about the time I moved here at, a well-known, well-respected professor by the name of Roy Meals, in LA, reported a 20-year follow-up on enlargement patients where all he did was take out the trapezium and pin the thumb metacarpal to the index finger metacarpal in distraction for six weeks and then let it heal.

So, he called it a hematoma distraction arthroplasty, and his results were as good or better than anyone else's reported results for thumb arthritis surgery. So, about 15 years ago, I tried doing his operation, but my patients had problems with the pins. The pins, if you leave them out, would get infected. If you left them under the skin, you had to go back to the operating room and take them out. There are sensory nerves that could get wrapped up by the pin.

My patients would do things like elliptical exercise machines and fall off them and bend the pins. So, I stopped doing the resection arthroplasty with the distraction after this, you know, and all kinds of problems with the pins, and I chose this other operation where you take a tendon and split it in half and use part of the tendon to keep it from migrating, and that works reliably.

More recently, I've wound up, you know, placing the tendon underneath the other half. It's for making a U-turn, and that gives you a stiffer joint that's less likely to fall apart. So, I'm into – I recommend, you know, mechanically stabilizing the base of the thumb, and I think that can be done with the anchovy procedure, but I also think that, you know, if you go to a teaching hospital and the residents are not supervised, and they

take out the trapezium and just stuff a loose tendon in the space, that that is just not going to work out well for a person who's hypermobile.

So, it's a complicated way of saying, "Yes, the anchovy procedure is a reasonable option to do, and I have seen it work in patients who are hypermobile." It's not my choice, but I've certainly seen it work, but I also have seen it fail through circumstances that the patient wouldn't necessarily be aware of, and they're not necessarily good for the patient.

So if you know your surgeon, and they're smart and well trained, and they do the surgery, and they follow their patients. They see the patients pre-op, and they see them post-op. Not a PA, or an MA, or a nurse, and they're aware of every failure, those are doctors that you're likely to get better results from. So, I would say if you trust your surgeon, you're probably going to be okay.

[01:03:35] **Sarah Jo Ritchie:**

Alrighty. Thank you so much. To continue with surgery questions, what about thumb CMC stabilization with an Arthrex TightRope implant in lieu of ligament reconstruction?

[01:03:50] **Dr. William Ericson:**

That is a technique that I used to augment my repair when tissues are really crappy. If you use it by itself, it will work for a while, but it's my opinion that that thread, it's a fiber wire suture that goes between the two buttons that that thread will eventually break, and depending on how much scar tissue you've made it, it may or may not make any difference because if you hold the bone in place for a while in some patients, the tissues around the joint will contract and can stabilize the joint.

So, I have seen patients have a soft tissue capsulorrhaphy, where the tissues just got tightened around the joint, and then had it supplemented with an Arthrex endobutton technique, and they've done fine, but I've only seen them out like one or two years, so long term, I don't know. I have used it. I do use it. It's a great technique to add more stability to the situation.

I tend to be a belt and suspender type doctor, so I will add it to a case where I think the soft tissues are not adequate, or if they've had a previous surgery, and let's say that patient's particularly pain tolerant, particularly strong-willed, and the weak link for them is their soft tissues that their, their soft tissues could never just to stand up to their willpower. That's a person where I would tend to use something like that also.

As a solitary device, I don't think it's a good idea, but I haven't seen anybody in my geographic area have it used as just a solitary device, but it is a very useful technique. There are small incisions, and [inaudible] tends not to be a problem unless it's, you know, where you can feel it, and someone who's thinking about using supplemental tissues, I think generally speaking, is probably a good idea for patients with Ehlers-Danlos syndrome.

[01:05:55] **Sarah Jo Ritchie:**

Alrighty, and another surgery question. "Do you find there is an increase in the incidence of developing CRPS for patients with EDS following surgery on the hand and wrist?"

[01:06:07] **Dr. William Ericson:**

No. I've had almost no CRPS in any of my patients in the last 15 years since I moved to Seattle. It really hasn't been an issue in my patients unless they had CRPS from somebody else's surgery. CRPS is a real thing, but some of my colleagues don't believe it, but they, if they don't believe it, they've never actually seen the real thing. It's an idiosyncratic thing. It doesn't really have anything to do with the operation itself. You know, it's not like one operation causes it.

In my experience, it tends to be in patients who are anxious and who don't have social support, and are worried, and in that patient population, I try and reassure them and also use ketamine as a part of their anesthesia, and ketamine helps interrupt chronic pain cycles. So, it helps to establish a rapport with the patient, where you understand the patient's goals, and if the patient doesn't have goals, or if their goals are to be on drugs and be on disability, that you don't get in the way of those goals and try and stop them from that, but it really hasn't been an issue in my patients at all.

But we do a number, I do a number of things that are specifically geared towards patients with connective tissue disorders. For example, my assistant is the best assistant I've had in 30 years, and he is extremely strong with his arms, but the strength isn't what matters. It's his endurance. So, he is able to hold the tissues apart gently as if the patient's awake, and we make these little incisions, but he doesn't just crank on the tissues. He just holds still, and it's much harder for someone to assist me with these operations with these little incisions, but he could ride a motorcycle all day long and not have his arms be tired.

So, like, it helps to have the right diagnosis, and it helps to have just one procedure, so you could see CRPR in patients who have an injury like a wrist fracture, and then a PIN injury like a closed reduction where the fracture is manipulated under anesthesia, and it falls apart, and then they have like another closed reduction in PINs that there – some patients, there's a window of vulnerability where if you re-injure them as the body's trying to heal, the normal sensory input gets misdirected to stimulate a pain center in the brain, and it can be a disaster.

But going to the operating room once and having one injury and having the patient with reasonable expectations, so I think establishing reasonable expectations of the patient and then trying to exceed them makes them much less likely to have anxiety or anxiety-related problems, which I put CRPS in that category. So, the answer is no. The CRPR really has not been an issue in almost any of my patients with Ehlers-Danlos syndrome.

[01:09:10] **Sarah Jo Ritchie:**

Alrighty. Thank you so much. We have a question on tourniquet use with extremity surgery. "Should the incision can be closed while the tourniquet is still up or is it best to let it down to check for bleeding before closure?"

[01:09:22] **Dr. William Ericson:**

Well, I personally feel pretty strongly about letting the tourniquet down and waiting till the tissues perfuse, so the arm's warm and see what bleeds, because particularly Ehlers-Danlos syndrome patients where they have capillary fragility, and it's pretty easy to just put a retractor in, and if you're just a little bit rough, you can poke a hole or rip a vein, and it may not bleed until things are dilated and warmed up.

So, if I had a student, my recommendation to them would be rigidly to do the surgery with the tourniquet up and not cut any blood vessels at all and then when you're done, let the tourniquet get down and wait a few minutes and wash the wounds out and make sure that there is no bleeding. And once the, once you're sure there's no bleeding, then close the skin.

Now, doing that, I still have an occasional hematoma, and it tends to be a stitch in the [inaudible] tissue that rips a vein, and then it bleeds, and it bleeds, you know, like a golf ball-sized hematoma, and those tend to resolve spontaneously without any major complications, but I have some colleagues who I know close when the tourniquet's still up, and personally,

I don't like phone calls middle of the night. So, I cover all my patients unless I'm out of town, but a lot of my colleagues have PAs or MAs they're signed up to, and they don't get called in the middle of the night when things swell up.

But I don't like surprises, so I would not almost ever close an incision while the tourniquet's still up. I would want the tourniquet down. In addition to having the tourniquet down, I have the anesthesiologist remove it immediately from the arm because if it's applied properly, it causes a venous tourniquet, and if you let the arterial blood flow in and don't let the venous blood flow out, you get a lot of bleeding immediately into the tissue and that causes swelling and pain.

If you take the tourniquet off immediately, the – you do not get as much bleeding, as much swelling, or as much pain. So, some of the anesthesiologists don't like it because when they let the tourniquet down, they're supposed to chart that time in their chart, and they're supposed to, they're getting an acid load and have to be aware of the pulse and the pressure and so forth. They tend not to want to do the tourniquet down and off the arm as part of one step.

So, but that's okay. Those anesthesiologists don't have to come to my room. We, I want the tourniquet down and off the arm immediately, and I would recommend not closing the incisions with the tourniquet up because you're going to have an occasional bleeder, and you're going to find out late when it's inconvenient, and it's going to be particularly inconvenient to the patient.

[01:12:27] **Sarah Jo Ritchie:**

Alrighty. Thank you so much. We have quite a few questions on finger, thumb, and wrist dislocations, and if there are any braces or splints that you suggest, both for daytime use and to sleep in.

[01:12:40] **Dr. William Ericson:**

Well, you know, we're not supposed to give specific, you know, commercial, advice about these things, but I have no conflict of interest, but Silver Ring Splints are by far the most effective device for proximal interphalangeal joint issues. They're comfortable, and they're, you know, almost pretty. The thumb, I think the ligaments, generally speaking, you know, need to be repaired surgically. The splints are okay, but they're, they're just never going to be great.

If you have Marfan syndrome, where everything is extremely loose, those patients actually don't usually come to surgery because they can't really generate enough force to wear out the joint. So, they're even looser than Ehlers-Danlos patients, and splints in those cases probably aren't – I'm not sure they're even helpful because they're so loose. But the thumb, you know, Silver Ring Splints in general has a whole bunch of products for the thumb and wrist.

There's another company, MetaGrip, a push orthotic for the thumb that's probably the most useful splint for thumb instability issues. It's designed by hand therapists. It fits the majority of patients' hands with their sort of normal range of size and proportions, and they're relatively inexpensive compared to seeing hand therapists. If you have relief of symptoms with this splint, a short opponens splint, the surgery, the Eaton-Littler ligament reconstruction basically does the same thing as a slip-up, but it's a split inside.

So that would, that would be a therapeutic test if you have someone with vague hand pain, and you think it's mostly the thumb. If you put them in a splint for a short opponens splint on their thumb, and their pain gets better, then you can be pretty well assured that the ligament construction will do the same thing. The wrist, you know, there are splints that are sort of, U-shaped that wrap around the radioulnar joint, and those can be helpful for mild to moderate instability wrist as, but itself, I don't think the splints are particularly affected in terms of changing the instability, but they can let other people know you're having a problem, and they can reduce the number of times that the joints unlocks, but I don't think they're going to make the wrist stable.

So, the thumb splints, I'm a big fan of. The finger PIP joint splints, a figure of eight splints for the PIP joints I think are really helpful. I don't think the splints at the end joints are, are as helpful, but the people with these joints all the way at the end of their fingers tend to be more in the vascular EDS category rather than just hypermobile, and they have, you know, other issues that tend to be a bigger problem. So, I hope that information is helpful.

[01:15:47] **Sarah Jo Ritchie:**

Yes, thank you so much. Another question: "It sounds like neck problems are related to the arm and hand issues. Do you have any suggestions on how to address the combination of issues, especially if the neck becomes worse?"

[01:16:00] **Dr. William Ericson:**

Well, when a patient comes to me, I usually let them talk uninterrupted for several minutes and trying to figure out what is their highest, priority for fixing what is stratifying their complaints, what is bothering them the most, and the patient may start out saying one thing, but you find out when you're actually examining them that they've changed their behavior because there's some other problems. There's a bigger problem, but they're just working around it, and they're just focused on that one problem, and that's probably a more realistic approach to try to get medical care, to try and focus on the thing that's bothering you the most when you're seeing a physician.

And when you're seeing a physician, if you're not convinced the patient that you're being listened to as a patient, and you're not convinced that your interests are the center of the doctor's attention and focus, you probably should be going someplace else for care. A more successful approach that I advocate is to look at the whole person and try and start figure out where they are on their art of life and what's bothering them the most.

In my geographic area, there are neck surgeons, and as a group, they tend to say, "You need surgery. If you do, I'm your guy, and if you don't, you know, see ya." That isn't universal, but there is a strong tendency for them to just say, "You know, I'm a technician, not an investigator." There are physiatrists who are orthopedic doctors who don't do surgery, that are trained in physical medicine and rehabilitation, and they are typically the most helpful doctors to patients with connective tissue disorders, and they are trained in typically in neck issues and thoracic outlet issues.

And some of them really enjoy seeing Ehlers-Danlos patients, because they can almost always help, and some of them just want to do nerve injections or spine injections. Some of them just want to do nerve tests, but if you can find someone who actually wants to listen to patients and disambiguate their problems and send them off to specialists for each of the individual problems they have, that can be a great partnership with a patient.

So, for that patient with their questions, these things overlap, and you have to see someone who understands the overlapping nature of these problems, and it wouldn't be unreasonable to start with a physiatrist, which would be an orthopedic doctor. Physiatrists, uh, are also called physical medicine and rehabilitation doctors in some areas and have them

look at the spine. Start with, you know, your posture and get X-rays of your neck.

And with hypermobility issues, the X-rays should be done with you upright and also with the neck in flexion and extension, and if you're having nerve symptoms that go down the arm, you'd probably be advised to get an MRI of the neck to look at the nerve roots also and start with posture and neck and then sort of work things down. The surgery that I do works predictably if the neck and the thoracic outlet are stable and understood, and that would be the first step for most of my patients is not if they come in with thumb pain, it's not to operate on the thumb but to look at the whole person and sort of figure out how much of their problem can be changed by changing their activities as opposed to changing their arm anatomy.

And working on posture and weight loss is a delicate topic, but my patient population, not particularly heavy, but nationwide, the country's become obese, and weight loss would be one thing that patients would have under their direct control that would benefit every aspect of their musculoskeletal system if their body mass index was in a reasonable range, but very hard to do with our food chain, processed foods, and the expensive inconvenience of eating a healthy diet.

So again, I would urge using that template that I've come up with to try and figure out what's bothering patients the most and incorporate evaluation of the neck and posture as the first steps to that.

[01:20:29] **Sarah Jo Ritchie:**

Alrighty. Thank you so much. We have time for maybe two more questions. Any questions we don't get to, we will forward on to the helpline, and if you have any questions that don't get covered today, please reach out to our helpline. Email them, call them. They are amazing, and they could help you find the answers you need. But, for Dr. Ericson, "what are your recommendations for sutures and closing to minimize the stretched scars and avoid poor outcome?"

[01:20:54] **Dr. William Ericson:**

Well, the way I do it, is I make small incisions, but small incisions means big risk. So, you know, I could not have done these operations earlier in my career with these little incisions, and I couldn't do them without certain equipment, and I couldn't do them without an excellent assistant. So,

it's a team effort. I try and use all internal stitches and have done so for probably 20 years. I've had a few of these operations myself, and one of the things I really hated more than anything else was suture removal. So, unless we're doing low rotation flaps, I will use internal stitches.

I used to use Vicryl, which is a common subcutaneous suture that orthopedic doctors use, but my patient population reacts very strongly with scar tissue to Vicryl. So, I switch to monochrome about 15 years ago with much less tissue reaction. I have particularly a wound closure technique that I use that I've developed that involves one small knot underneath the skin at one end of the incision and then going through the dermis, back and forth in a zigzag fashion, trying to align the edges perfectly, and I usually do three layers.

And then if it's over a joint, do a fourth layer, and then I will put glue on the skin, something called mastisol or benzoin, but you have to be careful with Ehlers-Danlos patients, because a lot of them have mast cell activation disorder and a lot of contact allergies. So, you have to make sure they're not allergic to tape or adhesives to do this, but you put some glue on the skin, and then you put tape on the glue and then cover that with a bandage that makes it so there's very little stress on the incision.

And if you do that, the incisions heal pretty nicely. They're a little wider than non-Ehlers-Danlos patients. But big stitches and big incisions, especially over big joints, will always give you a big wide scar in a particularly least connective tissue. So – and the closure that I recommend, you probably wouldn't let the intern do the closure, because if it falls apart, you'd lose the whole incision as opposed to interrupted stitches, whereas you screw up one of the stitches, there's multiple other stitches, but these incisions were just to use one stitch.

It just has to go in perfectly every time, and I feel pretty confident doing that with every single incision. And I think I've had one or two incisions open up in the last, 10 years, and that it healed fine in spite of that. This is just not for the casual surgeon to do things this way, and you really have to have someone who is seriously interested in every single detail about how to get a reasonable scar in addition to fixing the problem.

The small incisions are not something that just anyone should try and do. Like I wouldn't expect other hand surgeons to see this video and then go make a one-centimeter incision to decompress the proximity nerve. That's just not...

[laughs]

...likely to be a good idea. But you know, when I did this operation in the first time, I was the assistant, and I helped somebody decompress the median nerve from the wrist to the armpit, and it took us an hour too to close the incision. It left a huge ugly scar. It's, that's not necessary.

So small incisions, that's one way, but that does increase your risk, and you have to really know what you're doing surgically to do these things safely through these little incisions. Monochrome, much less reactive than Vicryl. Nylon stitches put in tight, cause pain, and don't hold the tissues that well, and wind up with big scars. You can tape the scars for a couple weeks afterwards, if you don't get an allergic reaction.

There's also a steroid impregnated tape called Cordran. If you get a hypertrophic scar, meaning it's raised and pink, and it might be itchy or sore, the steroid impregnated tape can make that raised scar softened and flattened within a couple weeks. So that's another approach, is if you have an incisional issue, you can put a steroid on the incision, then it will flatten and lighten, especially with these incisions near joints that when you call in the scar tissue, it tends to contract, and you get a bigger, thicker scar around a joint.

[01:25:45] **Sarah Jo Ritchie:**

Alrighty. Our last question for the day comes from somebody that's newly diagnosed. They were just wondering if you could recommend a good place to start in terms of getting a diagnosis with issues, imaging, and *et cetera*.

[01:25:59] **Dr. William Ericson:**

So, I would start with the Ehlers-Danlos website, chronicpainpartners.com, and I would go to my webinar.

[laughs]

If that wasn't the probably – the most useful thing I'm aware of for the upper extremity issues. The previous webinar that I did after the talk I gave in Baltimore, I thought, was a reasonable size to start for upper extremity. The other webinars are on the chronicpainpartners.com website, are all done by people who are well-known in the Ehlers-Danlos community, and are leaders in their little areas. So, if you are having a specific focus of your problem, I would go to the chronicpainpartners.com website and try and find webinar information about that particular issue.

The article, like the volume of the *American Journal of Medical Genetics* that had a special volume on Ehlers-Danlos syndrome in 2017, is also a wonderful place to get information, but it tends to be more medical, like it'd be more helpful if you had a medical degree to read it. But if you want to share something with a physician, that supplement to the American Journal of Medical Genetics that was arranged for by Brad Tinkle was extremely helpful, as a – you know, if you want to give a doctor something to read, that volume that Brad Tinkle created was fantastic.

Brad Tinkle has also written a couple books on joint hypermobility issues that I think are extremely helpful, and I think there are additional resources at the chronicpainpartners.com website that there are some other books written by patients that divide problems up by organ systems. So, they talk about dysautonomia, GI problems, musculoskeletal problems by joint, but I think the, that those are the most useful things.

Brad Tinkle's books on joint hypermobility, I thought, were extremely well-written and helpful just in general, and the other resources available through the Ehlers-Danlos Society and chronicpainpartners.com I think have been extremely helpful.

[01:28:22] **Sarah Jo Ritchie:**

Alrighty. Thank you so much, Dr. Ericson, but that's all the time we have for our webinar today. As I mentioned earlier, if your question did not get answered, please reach out to us on our helpline, and we will get back to you as soon as we can. If you would like more information about anything that was presented today, please check our website for more resources and information and give our helpline a call. Also, sign up for our newsletter if you haven't it already. It's a fantastic source for the most up-to-date information and upcoming events.

Our next webinar will be on August 21st. We have Jason Perry giving a talk on "Dislocation Management in EDS and HSD." You can look out for the signup on our website for the next webinar shortly. Thank you again, Dr. Ericson. Your presentation was extremely informative and very helpful. Technology permitting, this webinar should be available on our YouTube channel within the next week or so. If you found this webinar helpful in any way, please consider hitting that like button once it's available on our channel and subscribing, so that you can be alerted to when we're uploading our videos.

Also, there will be a donation button on our main YouTube page along with a link to donate in the description of the uploaded webinar. It's thanks to donations that allow us to continue to provide programs like our webinars along with other great projects and research we're undertaking. Once again, Dr. Ericson, thank you so much, and I hope that everyone that joined us today has a wonderful remainder of your day.

[01:29:45] **Dr. William Ericson:**

Thank you very much for the opportunity to discuss these issues.

[01:29:50] **Sarah Jo Ritchie:**

Have a great day.

[01:29:51] **Dr. William Ericson:**

Thank you. You too.
