

# Hand Therapy and Ehlers-Danlos Syndrome

## HAND THERAPISTS' IMPORTANT, UNDER-RECOGNIZED ROLE IN HELPING PERSONS WITH EHLERS DANLOS SYNDROME.

(from the Keynote Address at the European Federation of Societies of Hand Therapy Congress in Rimini Italy, May 12, 2023)

The objective of article is to provide insights into various manifestations of Ehlers Danlos Syndrome (EDS), the impact on daily functioning and how collaboration between client and therapist helps to achieve pain control, joint stabilization and participation in activities that are valued by the client. I will share some orthotic interventions and adaptive devices developed in collaboration with persons who have with EDS.

An individual with symptoms of EDS may become a client of a hand therapist due to joint hypermobility/instability in the upper extremity, which is how I first became involved with helping persons with EDS. Two such individuals, Nicole and Ariel, have figured predominantly in my education of how EDS can severely impact an individual's quality of life and overall health.

EDS is a complex, inherited condition that affects the connective tissues in the body.

Connective tissue (made up of cells, fibrous material and collagen) is ubiquitous and responsible for supporting the skin, blood vessels, bones, joints and organs. Thus, faulty connective tissue has widespread implications and symptoms tend to increase and get worse as the individual ages.

There are 13 subtypes of EDS and all subtypes have poor collagen production and various forms of hypermobility. Genetic markers have been identified for 12 of the 13 subtypes; the genetic marker for 13th type [hypermobile EDS or HEDS] has yet to be identified.

Definitive diagnosis of EDS requires genetic testing to determine the subtype, as well as consideration of the symptoms and co-occurring conditions.

Joint instability is not restricted to the upper extremity but extends to the spine and lower extremities. Thus an "EDSer" (Nicole and Ariel's term) may be covered from neck to toe with braces and splints and also use ambulatory aids.

To grasp the extent of the joint hypermobility, the Beighton Scoring System is a useful place to start.



From: <https://www.ehlers-danlos.com/assessing-joint-hypermobility/>

A positive Beighton hypermobility score is any outcome greater than or equal to 5/9 points in adults, 6/9 points in children (before puberty), and 4/9 points in adults over age 50. Nicole and Ariel both scored 9/9. The escape artist Houdini is suspected to have had EDS.

The individual with EDS is really the expert on their body and is likely well informed about their condition and may have a widespread network of fellow "EDSers" or "Bendies" that they share information with. Be open to being an equal partner in the process. It's better when the clinician works collaboratively WITH the person with EDS.

Unfortunately, Nicole and Ariel reported that experiences with clinicians have sometimes been negative. They assert that it is not helpful when clinicians think they know more about how the body of a person with EDS works, than that person does. Furthermore, the basic education of the clinician might not be sufficient to address the treatment or care that the EDS client needs. Some clinicians can get scared off.

Nicole stated, "EDS life is consistently full of inconsistent chaos". "Patient-led care is the most amazing thing". She suggested asking your client "how do you want to do this?" Be collaborative.

Ariel found herself avoiding physiotherapists after her physio asserted that "pain is important to build muscles". One size does not fit all and it takes a lot of creativity to help an EDSer. It's important to recognize that the clinician and EDSer need to be partners.

In addition to the Beighton score, there is the Brighton Criteria for Diagnosis which is especially useful if there has not been genetic testing.

### Brighton Requirements for Diagnosis of EDS:

Any ONE of the following:

- Two major criteria
- One major plus two minor criteria
- Four minor criteria
- Two minor criteria and unequivocally affected first-degree relative in family history

### Major Criteria

- Beighton score of at least 4
- Arthralgia for longer than 3 months in 4 or more joints

### Minor Criteria

- Beighton score of 1, 2, or 3
- Arthralgia (3-month duration) in one to three joints or back pain (3 month duration) or spondylosis or spondylolisthesis
- Dislocation or subluxation in more than one joint, or in one joint on more than one occasion
- Three or more soft tissue lesions (e.g., epicondylitis, tenosynovitis, bursitis)
- Marfanoid habitus (tall, slim, arm span great than height (ratio >1.03 ratio), upper segment less than lower segment (.0.89 ratio), arachnodactyly
- Skin striae, hyperextensibility, thin skin, or abnormal scarring
- Ocular signs: drooping eyelids, myopia, antimongoloid slant
- Varicose veins, hernia, or uterine or rectal prolapse
- Mitral valve prolapse

The Brighton Criteria illustrate that EDS is so much more complex than just joint hypermobility and there is a constellation of other symptoms and co-occurring conditions that create challenges for individuals with this syndrome and deteriorating symptoms as the individual ages. Should you suspect you have a client with EDS (even if not yet diagnosed), it is helpful to expand your knowledge of the strange biology of the condition and wide-spread symptoms that extend beyond the joints.

Here is a link to an excellent resource: <https://www.ohsu.edu/sites/default/files/2019-09/CPD%20MSK19-Thu-3-Friedman.pdf>

Nicole and Ariel have a spectrum of strange, interconnected symptoms. Their conditions are seriously life-altering and Nicole's health is often precarious. Nicole also has a disorder of the autonomic nervous system called Dysautonomia (often seen with EDS), affecting many organ systems. To be specific, she has the subtype called Postural Orthostatic Tachycardia Syndrome (POTS).

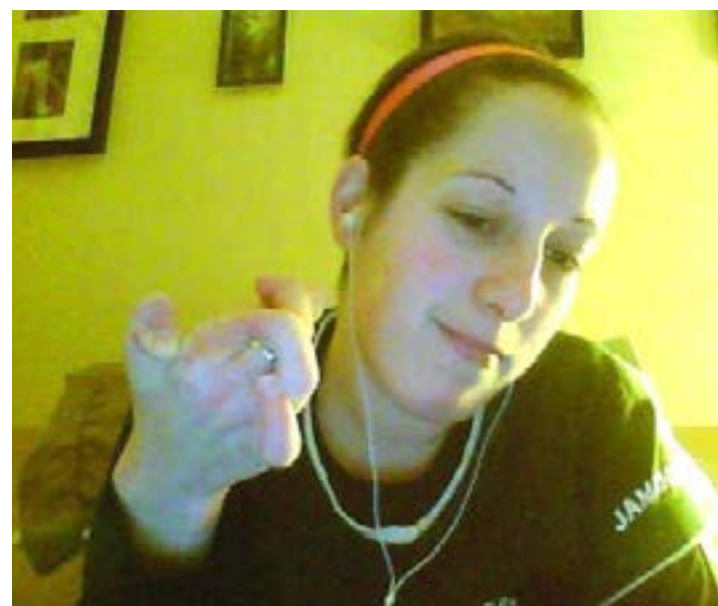
Nicole initially came to me because her occupational therapist thought she would benefit from custom-made silver-ring orthoses for her hypermobile fingers. (Figure 1)



After she tried various samples that I had, she was content with the more economical Oval-8 orthoses that were customized to hold her proximal interphalangeal joints (PIP) in a few degrees of flexion. (Figure 2)



In contrast, Ariel was not satisfied with Oval-8 orthoses and wanted custom-made silver-ring orthoses to control the hyperextension in her PIP and DIP joints.



After trying some sample silver-ring orthoses, Ariel decided to purchase two separate custom-made orthoses – one for the PIP joint and one for the DIP joint. (Figs 3 and 4) Generally, she finds that the PIP joint orthosis is sufficient to limit hyperextension at both the PIP and DIP, however, she wears both orthoses when she is doing a lot of keyboarding.



Demonstrating how just the PIP extension-blocking Silver Ring Orthosis (worn on the index finger) controls both the PIP and DIP joints (in contrast to the unstabilized long finger).

Next, Nicole wanted stabilization of her wrist and thumb, while still allowing wrist motion. "Just putting my hand in my pocket causes my wrist to sublux." In Figure 5, Nicole demonstrates how, with a combination of wrist flexion and passive thumb abduction, her thumb touches her forearm (one of the test motions in the Beighton score).



I cautioned her to avoid stressing her thumb with this demonstration.

Nicole stipulated that she wanted a wrist orthosis that 1) did not extend the full 2/3 up the forearm (as I would normally do for wrist-stabilization) and 2) permitted some active wrist flexion and extension. We worked collaboratively to design and fabricate a circumferential forearm-based wrist-thumb stabilizing orthosis, molded from 1.6 mm (1/12 in.) Orfilight with hook-receptive neoprene straps. (Figure 6) Full thumb interphalangeal (IP) flexion was permitted but there was an IP extension-blocking hood over the distal phalanx. The distal edges in the hand were more proximal than I would normally incorporate in a wrist orthosis, to permit some active wrist flexion and extension, as requested by Nicole. The circumferential forearm-base provided sufficient surface area to compensate for the shorter forearm lever arm.



Permits some active Wrist Flexion







Nicole was “pleasantly surprised” that the orthosis was comfortable, did its job and made life easier and safer.

Nicole, inscribed her orthosis with a Japanese expression meaning “fall down seven times get up eight”. (Figure 7) Years after writing this inscription, she acknowledges that as a non-Japanese person, this might be construed as cultural misappropriation. However, the expression demonstrates her determination. She used the orthosis for 3.5 years until the plastic cracked.



To manage her gastroparesis and cyclic vomiting (symptoms associated with her dysautonomia), Nicole had a soft, plastic jejunostomy tube (J-tube) surgically inserted into her small intestine that emerged through the skin of her abdomen. This tube was hooked up to a pump that dispensed hydration fluid. Nicole used a large syringe to inject enteral nutrition into the tube, ten times daily.

So, the next challenge was to provide joint stabilization so that she could more easily manipulate the syringe. We were confronted with a decision – either make an activity-specific orthosis or modify the syringe. We chose the latter. Thus, we made a two-part adaption – an enlarged head for the plunger, molded from thermoplastic pellets, and an enlarged finger grip on the body of the syringe tube, molded from sheet thermoplastic. (Figure 8 and [Video 1](#)) The next consideration was that any modification needed to be removable and

transferable to other disposable syringes for feeding. When molding the two components, hand cream was applied to the plastic of the syringe to prevent the heated thermoplastic from sticking. The adaptation worked very well. Again, you see evidence of her self-motivation in the phrase that she wrote on the plunger head. “When life smacks you in the face, smack it back.”



Here is another syringe adaptation, based on our design, made by an occupational therapist for a friend of Nicole in the USA. (Figure 9)



After Nicole’s forearm-based wrist-thumb orthosis broke, she chose to focus on stabilization of the thumb only. Ariel had similar needs. Since fragile skin is an important consideration for both, and we decided that lining the circumferential hand-based thumb-stabilizing orthoses with 1.6 mm (1/16 in.) thick neoprene was desirable.





For Ariel, I made two hand orthoses and I utilized a simple loop fastener that was easy to replace when it wore out. Note the thumb IP extension-blocking hood over the distal phalanx. (Figure 10)

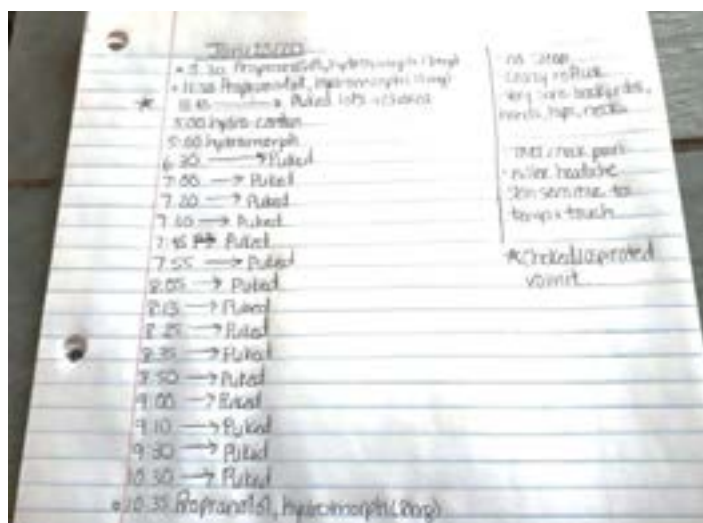


For Nicole, due to her extreme thumb instability, I incorporated more thermoplastic in the design and created a pull-back strap with a hole through one end. (Figure 11 and Video 2) Note the beige neoprene lining seen in the video.



1.6mm thick thermoplastic lined with 1.6mm thick Neoprene

Perhaps the most unusual request from Nicole was to make her, what she called, "Barfy Splints". In case you are unfamiliar with the term "barf" it means to vomit. Another term is "puke". There are often occasions when Nicole has been stuck in cyclic vomiting. Note the frequency of vomiting on one particularly bad day. (Figure 12)



Nicole specifically wanted an orthosis that supported her wrist and digit metacarpophalangeal joints in slight flexion. (Figure 13) Note again the hole in the straps to minimize stress to her fingers when fastening and unfastening the straps.



2019- 2mm thick Orflight Orthosis Supports MCPs of thumb and fingers. Note: wide wrist strap and holes in straps to facilitate fastening to minimize thumb stress - finger to thumb opposition is possible.

On many occasions she has found that the orthosis provides good support to her wrist when she is hospitalized (which unfortunately is often) and has an IV in her hand. (Figure 14)



Ariel has not found any prefabricated orthoses that help her unstable shoulders. She instead uses KT Tape (Kinesiotape) which helps with proprioception. (Figure 15) She needs someone else to apply the tape. While the tape is very helpful, she needs to alternate different brands to reduce the "tape rash" reaction causes by the adhesive. She also uses an allergy relief nasal spray and takes anti-histamines to help with hives.



Unstable Shoulders - KT Tape (Kinesiotape) - helps with proprioception need someone to do taping - irritates skin with prolonged use.

Pillows - Prefab Shoulder Stabilizing Orthosis - didn't work

Individuals with EDS often spend years searching for answers that account for their symptoms. No two people with EDS are identical. They have different symptoms, different subtypes, and different experiences. Hopefully one day all medical professionals will readily consider that someone might have EDS, thus reducing the time to diagnosis and improving pathways to care.

Since orthoses are a key component of intervention, for long-term/permanent use, it is important to consider that these devices are often visible and the aesthetics really matter. However, helping a client with EDS may well extend beyond providing orthoses for unstable joints.

On the part of the hand therapist, one needs to be creative, open-minded to the unusual requests of a person with EDS, and above all, collaborative throughout the assessment and intervention process to optimize outcomes. Partnership is key. I was challenged to think outside the box. I tried to be accommodating and flexible. I listened, learned and allowed my clients to lead as much as possible. They are in fact the experts about their bodies.

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With contributions from Nicole and Ariel, EDS Experts





**IFSHT NEWSLETTER - REACH VOLUME 3, NO. 2**



Issue 2 of volume 3 of the IFSHT newsletter is currently in production and will hopefully be available in August 2023.

Please check out the following link to access it: [https://ifsht.org/publications/?publications\\_category=29](https://ifsht.org/publications/?publications_category=29)

The publication aims to collate Research, Education, Achievement and Clinicians in Hand and upper limb therapy around the world.

In the next edition of REACH we will look to the future; consider how knowledge expands and how hand therapist can move with the tide with our continued new section on how to write and publish research and horizon scanning for ongoing research which may affect our practice in the near future. It seems pertinent then that this issue features clinical pearls on the use of 3D printing, a technology that has now become mainstream in recent years.

This issue's Spotlight On! Section will feature the British Association of Hand Therapists, we also continue our new "Volunteer" section and ongoing profiles of recipients of the prestigious IFSHT Lifetime Achievement Awards.

We call on hand and upper limb therapy clinicians and researchers to submit any contributions for consideration to: [informationofficer@ifsht.org](mailto:informationofficer@ifsht.org)

**UPCOMING EVENTS**



It's now over a year now since the last Joint Triennial Congress in London. Now fast approaching is the next Joint Triennial Congress in Washington in 2025.

The website for this event is launched so please follow for updates! <https://www.ifssh2025.org/s/>



On 6th to 7th October 2023, the British Association of Hand Therapists will hold their annual conference in Bournemouth. Event details at: <https://www.eventbrite.co.uk/e/baht-conference-bournemouth-2023-tickets-57611564197>



# Pearls of Wisdom