

# LOOSE CONNECTIONS



The  
**Ehlers  
Danlos**  
Society™

Your Magazine About Living With EDS and HSD  
**Summer 2017**

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## THE EHLERS-DANLOS SOCIETY

The Ehlers-Danlos Society is a global community of patients, caregivers, medical professionals, and supporters, dedicated to saving and improving the lives of those affected by the Ehlers-Danlos syndromes, hypermobility spectrum disorders, and related conditions.

We support collaborative research initiatives, awareness campaigns, advocacy, community-building, and care for the EDS and HSD population.

Our goals are worldwide awareness – and a better quality of life for all who suffer from these conditions. Research is at the center of what we do, so that one day we will have a cure.

**Our strength begins with hope.**

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# 2017 Global Learning Conference and Continuing Medical Education

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**WE ARE DAYS AWAY FROM THE 2017 EDS** Global Learning Conference. In the midst of our last month of preparation, we received wonderful news, and (for the first time since 2012) the Society in association with University Medical Center of Southern Nevada is delighted to announce Continuing Medical Education credits will be available to registered professionals. The conference is designated a live educational activity, "Revised Nosology for the Ehlers-Danlos Syndromes and Introduction to the Hypermobility Spectrum Disorders." for a maximum of 16.25 AMA PRA Category 1 Credit(s)<sup>™</sup>.

The CMEs help us bring to doctors, nurses, physiotherapists, and other medical professionals, knowledge about the new diagnostic criteria, the associated conditions, and all the connected presentations this year. We look forward to seeing everyone in Las Vegas!

For information about the conference, beforehand and afterward, visit [ehlers-danlos.com/2017-eds-global-conference/](http://ehlers-danlos.com/2017-eds-global-conference/).

For CME information, [ehlers-danlos.com/2017-global-learning-conference-cme/](http://ehlers-danlos.com/2017-global-learning-conference-cme/).

# DAZZLE VEGAS



The Ehlers-Danlos Society  
Global Learning Conference  
Bally's Hotel, Las Vegas  
September 7-9, 2017



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**Rebekah**, Atlanta, GA, Former Chair of the Board,  
Ehlers-Danlos National Foundation



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SUMMER  
2017





# Finding Hope: Help for Those Living with Ehlers-Danlos Syndromes

**CHRONIC PAIN—OR PAIN THAT LASTS** longer than six months—is a difficult and often debilitating diagnosis, and for those living with an Ehlers-Danlos syndrome (EDS), the joint pain is very real. EDS can cause loose joints that are prone to dislocation, hyperextensible joints which are linked to the early onset of osteoarthritis. The associated pain can vastly limit your daily activities, and can wreak havoc on your emotional state.

Luckily, there are still ways you can manage your pain levels so you can continue living your life.

## Stay Positive

As the chronic pain management website [OvercomingPain.com](http://OvercomingPain.com) insists, “Chronic pain is a serious problem but is often made worse by misinformation, negative attitudes and beliefs, outdated ideas [and] negative emotions.” When it comes to pain management, getting the right mindset and maintaining a positive attitude are two key steps that will help set the foundation for your success, even with an invisible illness like EDS.

## Ask for Help

With a condition such as EDS, the risk of further injury is intensified by joints that are easily dislocated, and although you might think that movement would help lessen the pain, it may make things worse. Living with chronic pain means some days will be worse than others, and unfortunately it is hard to predict what each day will be like. It is important to ask for help with [daily activities](#) when you need it, such as hiring a housekeeper to help with upkeep, finding yardwork help, or simply

asking a friend or family member to accompany you to the grocery store to help with reaching and heavy lifting. There is absolutely nothing wrong with asking for help, and your body will thank you in return.

## Acupuncture

Acupuncture is an ancient healing practice that has been used to treat a variety of conditions, including pain, for centuries in Asian societies. It has recently started gaining popularity in Western cultures as well, and even [Harvard Medical School](#) has been supporting the use of acupuncture for pain relief in recent years. [WebMD](#) lists acupuncture as a “painkiller” and an alternative to pills or prescription medications. Multiple studies have shown the healing effects of acupuncture, particularly on pain relief, and the FDA has approved acupuncture needles as medical devices since the mid-1990s. For those suffering from chronic pain, it is worth a try, however, make sure the acupuncturist is aware of your EDS, as one of the symptoms of EDS is stretchy skin that is prone to bruising.



## Acupressure

Interested in acupuncture but scared of needles? Try [acupressure](#) instead. This form of traditional Chinese medicine has the same effect as acupuncture – without the use of any needles. As with acupuncture, let the acupressurist know of your EDS, as you will require a gentler touch.

## Massage

Massage therapy has a role in pain management and has been shown to be effective in pain relief and management. When incorporated into a patient's pain management program, massage has also been shown to reduce the need for pain reliever medications. Rather than stretching your already loose joints, the [massage](#) should focus on strengthening the musculature.\*

Nobody wants to live with long-term or lifelong pain. The techniques listed above are just a few of the proven tools you can incorporate into your life for pain management. Don't be discouraged if these options don't work for you; every body is different and chances are, there is another technique or healing practice that might be better suited for your body. However, it's clear that there are a wide variety of offerings available these days that can have a very positive impact on the quality of life for those living with EDS.

### **JACKIE WATERS**

\* [Editor's note: [Myofascial release massage](#) is particularly beneficial to many of us with EDS, which is a technique of applying gentle sustained pressure into the myofascial connective tissue restrictions to eliminate pain and restore motion.]



# An Open Letter to My New Specialist

*Dear Hand Specialist,*

I am still processing our first important appointment and I thought that it might be helpful for both of us if I am able to break down and articulate my feelings during and after leaving your office and to let you know honestly why I won't be coming back.

I arrived in your office in late winter, on a sunny but cold day. I was dressed as I usually am for most of the year, in long john thermals, thick socks, sturdy shoes, layers of thermals including vest, spencer, long sleeved T-shirt, a warm woollen cardigan and two scarves. I loved your corner office with the winter sun pouring in and I enjoyed that unlike many medical offices where I am often too cold, I appreciated that I could remove both my scarves and my cardigan.

I placed both across my lap as I entered your office and I was a little self conscious that I looked like the Michelin man compared to you in your light layer of clothing. But I am used to that now.

Thank you for your warm welcome and for the gentle way that you asked how you can help me. I truly appreciated that when I explained my condition to you that you responded with what came across as sincerity and honesty that you have never heard of EDS. Thank you also for listening to me as I explained that in winter my aching hands are especially bad and that I have come here today to ask about splints for my hyperextended fingers and compression gloves that can be fitted to my hands. I explained that compression gloves keep my hands warm, the chilblains away and give my tissues the extra support that they need in

winter to stop my blood vessels from bursting. I usually use over the counter gloves and they work wonders but don't fit perfectly.

I explained that I am doing better than expected since diagnosis a year ago, with my small team of wonderful supportive specialists who are managing my energy, my blood levels, my injuries and my hormones. That years of painstaking trial and error, with a large dash of heart ache and agony but a few wins along the way, have resulted in a very precise regime of therapy, rest, and what appears at last to be a winning combination. I explained that I can still drive, go out a little, but that every third day I need to stay in bed, which is fine because then I get to do all my writing.

Thank you for listening to me so well, I felt heard.

But then you answered me and I was not expecting what came next. I felt as though you had listened to my story but then like a judge, jury and executioner, you were now making your ruling on everything about me, all of my body, everything that I do, and everything that I am doing wrong. This is the impact that your ruling had on me:

- When you told me categorically that the first thing that needs to happen is to get me out of bed I felt dismissed and defensive of the specialists who have put me there, with such hard work and such great results. I felt as though you did not hear me that this is a crucial part of my stability.
- When you told me that the only reason that I get chilblains is that I am not dressing properly or warmly enough I felt scolded and invisible before your eyes; the Michelin man not only already doing all the things that you are instructing and assuring will fix me, but wearing the exact brand





you mention. How do you not see this...or me? I want to point out my layers and open my phone and show you the article on Wikipedia with the last couple of lines which mention that regular chilblains may be a sign of a connective tissue disorder, and that EDS is exactly that.

- When you instruct me not to ever use the internet to look up my condition and you tell me that bloggers are liars because if they use a wheelchair ten percent of the time, that they only blog about wheelchairs, I feel kicked in the gut. When you go on to tell me that all bloggers exaggerate and that as a result I will then get a skewed idea of what my future health will be, I feel an impact again. You don't stop for breath for me to tell you that the only reason that I got my diagnosis after 30 years is because I have been doing my own Google research, worked out which kind of Specialist to see who would know about EDS and that I myself actually use a wheelchair 10% of the time...and that I blog full time and not only about wheelchairs.
- When you hand me a brochure of products that I might like to buy and tell me that when I come back next time we can look at my fingers and splints (the main thing that I came here urgently for), I felt robbed because you have not addressed what I brought to you and yet I must still pay you for the privilege.

I came here to ask for help with my hands, I was not asking you to start from scratch and "fix me." I was not asking you to compete with all the other specialist that I have. I am a well functioning proactive human being and even if I didn't study Biochemistry at University, I would still know way more about my body and the thirty years that I have suffered than you do.

I need to tell you these things even though I will never come back and see you because I want you to know that if you treat every patient as a bunch of text book symptoms, each with only a single solution, then not only will some people

leave unhappy as I did, but some, like me, may not be fixed, or worse. I need you to know that as patients we trust you. We need you to help us in our individual and unique issues, and if I didn't have the background, the gumption, the trust in myself and my specialists, then I could well have taken your advice to my detriment.

I believe and agree that there is a ton of information out there that is not accurate, but also that it comes not only from the internet but also from specialists who misinform either through inexperience, lack of sufficient training, or who see things only through limited filters. I am not saying that you are any of these, but I would love it if you could see that there is a whole world of information out there and that in terms of hands alone, it isn't only confined to what you alone have learnt in school. Your patients are people, with individual needs, diseases, inherited conditions, and as a hand specialist, a foot specialist, a heart specialist or a surgeon, you each have the opportunity to be part of a whole bunch of teams who can change people's lives.

This letter is not about what you did wrong, it is about the impact on me, and more importantly, the opportunities that you could and can have for so many people.

*Jennifer Peacock-Smith*



I am in my forties and suffer from EDS. I have a rich story to tell which I am now writing my book about, which spans four decades, nearly a dozen cities in 6 different countries (USA, South Africa, Fiji, New Zealand, Canada, Australia), a bunch of different medical systems, two Medevac experiences, three children who are all now in the process of diagnosis which is so much easier for them now that I have been diagnosed.



# Gina's Story: Part One

**E**VEN AT FIVE YEARS OLD I WOULD GET sick more than the other kids my age, and when I did get sick it hit me harder and always lasted longer than everyone around me.

I got ringworm at six and I lost all the hair on my body; at the same time I hurt my right eye, so that sunlight hurt it. My sight in that eye was always a tad blurry; it never really healed. If going bald for six months and having to wear an eye patch was not hard enough, I started getting hurt in my kick-boxing class: sprains, bumps, and bruises. My mother and I were called to the principal's office each time one of my teachers saw one of my injuries, and we would be questioned separately by police. Those interviews kept me from ranking up in kick-boxing. Mom told me I needed to practice more and stop being lazy: "It's just small pains, push through, it won't kill you." I never got past the third belt and had to stop, it was just too painful. Mom said maybe when I get older I would be ready to try again.

When I was 11 I tried karate. Yet again I'd get hurt, with the smallest mock punch. Mom and Dad called me a "wuss." I tried hard to prove them wrong. I wanted to one day to be a black belt. But after two years we had to stop due to hard financial times; I only made it to fourth belt.

I found I needed to hide my pain from my mom; she saw me as being a sissy, being lazy, or just trying to get out of school. I wouldn't limp around my mom or the bullies at school. The bullies got worse as I got older. They jumped if they saw weakness. I would just wrap up and splint my swollen joints, and hide them under my clothing. In my last year of middle school I wanted to be in a sport again to show I was strong. I joined tennis, and was pushed

by my family to practice. My first game I fell and my right knee popped out of place. I popped it back but ended up losing the game along with my spot in the team. I would not play again.

In high school, I joined FFA and wanted to show livestock. I signed up to show a goat named Grady, a sheep, and Fiona, a pig. With the goat I won third, my first time showing any animal. I didn't place with the sheep.

Some older kids found it fun to let the pig out over and over. Each time took a full team to get her back in her stall. When it was time to show her, she didn't want to cooperate because she was tired and mad. I didn't blame her, I felt the same. She ran at me and hurt my right leg, popping my knee right out of socket.

I ended up with my first surgery, and it was a very hard experience. I had a seizure waking up from anesthesia and was quite ill after waking. They put me in the recovery room while I wasn't fully free from the anesthesia. My mother had to leave to get the jeep, but said she would be right back. As soon as she was gone, the nurses picked me up and put me in a wheelchair. I kept telling them, "I need to wait for my mom still." I was moved out of the hospital, told to stand; they pulled the chair out from under me and handed me the crutches, still wrapped up. They pushed me out into the rain and left me; confused, I tried to walk around into the road. Luckily mom was just driving up and grabbed me. I told her what happened as I cried.

I was less than 100 pounds, but they gave me pain medication at the dosage for a 200 pound person. The wrong surgery was done, making my knee more lax then before. When we confronted the



surgeon and the nurses in the hospital, everyone blamed someone else, even blamed me. I needed a second surgery.

I got hooked on showing animals (I just stayed away from pigs). But several years of goat showing were taking their toll, particularly because of the bracing—when a goat pushes against your knee and shows off their muscles for a judge. I planned to show a boer doe and chickens my last year of high school because they don't brace, they just look pretty.

Before I could pick out my new goat, I wanted to learn to hunt with a bow-and-arrow. My FFA agricultural teachers wouldn't teach me unless I shot in the shooting team. I really was not a fan of guns. It was clear on my first day of shooting this was not a good idea; setting up I popped my right knee out.

I was up first, giving me no time to put my knee back in place. I thought I would just shoot a round and then I could sit and fix it. I could see my knee cap almost on the side of my knee. I took my first shot, but wasn't ready for the kick back. It flew my shoulder out of place, then it slammed right back in. At the same time I felt a very bad pain in my right knee. Looking down I didn't see my knee cap now, and I couldn't straighten my leg.

I look back up and my teacher was yelling, "Shoot again! You are holding up the line!" I had missed the target and he didn't want me to stop until I hit it. I tried to tell my teacher I needed to sit but he talked over me, thought I was scared. He told me to buck up and made me shoot again and again. Each time I felt my shoulder pop out and slam back. I was the only girl. I held my tears because I knew he would just yell more if I did, but I didn't know how long I could hold out before I fainted. Every shot missed by more and more. I stood there shooting for half an hour until finally my teacher and the boys lost patience.

One guy sat by me and asked if I was okay. He was the only one who had noticed my knee was out. When the two of us were setting up, he had seen

it happen. I told him there no way I could get up again. I said, "When my mom gets here to watch, I'm going home. I can't shoot again, I'm going to hurt myself very badly."

He was kind. He just nodded and got up. It was my turn again next, but he asked to take more shots because he was not sure if his gun was sighted right. He missed again and again, on purpose I knew.

I called my mom to bring my crutches and to get there as soon as she could. My new friend bought me the time I needed. I fought to not cry or black out, because if I cried with all the guys around they would never respect me. As soon as my mom got there I waved her over. The boys were watching my friend shoot. I broke down in tears. I showed Mom my swollen knee and my messed up shoulder. She told my teacher I got hurt and needed to go to the doctor.

She got me in the jeep and I almost blacked out before I got home. She felt with ice and rest I would be okay. I went back to school on crutches; when I walked in the FFA classroom I was surrounded by the shooting team and the teacher. "Did you fall or what?"

I told them what happened and they didn't believe me. I showed them my jacked-up colorful shoulder that was the size of a grapefruit, and my knee that was three times its normal size. They stopped laughing and just dispersed without a word. My teacher said, "You made of paper? I don't want to see you shoot on the team any more. You can't hit a thing, better you just stay home and do something else. You're just not cut out to play with the boys." He gave me a pat on the back and walked off. I just stood there staring at the wall.

He never respected me after that. I was just an annoyance and he had no patience for me. A week later, I went to the doctor.

*(Part Two will run in the next issue.)*

**GINA R. COOK**

# From the Editor's Desk: What Now?

**T**HIS YEAR HAS BEEN THE MOST important for the EDS community in two decades, perhaps ever; it's easily been the busiest. March held the release in the *American Journal of Medical Genetics* of the revised diagnostic criteria for the EDS and for the first time articles on associated conditions, as well as a reorganization of joint hypermobility resulting in the hypermobility spectrum disorders; if you haven't already, you can read them all at <http://bit.ly/EDS2017papers>. Anticipation and reaction was intense – and a lot of work had to happen very quickly thereafter. I'm still uncertain how to write about such a personally momentous event. So I'll do what I generally do; tell a story. Or two.

In the 80s I worked construction for a while. One early morning I found myself leveling concrete to create a parking lot for a clothing store—which is hard work, hard particularly on the shoulders, moreso when those joints refuse to stay in place, and wet concrete gets everywhere because you're working pretty quickly to keep the surface smooth. About two hours in, my pants, boots, socks, arms had been soaked through—and I started feeling strange. That morning is far enough away that memory has, mercifully, muted many of the subsequent details. It turned out that I was allergic to wet concrete in some fashion, and had broken out in hives over almost all my legs, my arms, and elsewhere.

At a follow-up appointment with the worker's comp doctor, and because I couldn't afford doctors much in those days, I thought to ask about my "family joints." I showed him my dislocating shoulders, and asked him if he had any idea what might be responsible. My memory's clear here: "Well, it might be 'Ehloss-Danlen' syndrome, something like that. But you don't have the stretchy skin."

That was more than I'd known, so I had him write it down on a piece of paper and put that in my wallet.

That slip of paper stayed in my wallet for more than 15 years, until the day in 2000 when everything physically wrong with me caught up all at once. After I was diagnosed in the emergency room with a panic episode—in fact probably my first transient ischemic attack—I was recuperating but not feeling much better, and felt sure something more than anxiety was going on. I remembered that piece of paper, went to Google, and entered "Ehloss-Danlen" in the search box which produced no result, but a suggestion: "Did you mean Ehlers-Danlos syndrome?" Maybe I did. I found the Ehlers-Danlos National Foundation and the answer to not just me but, as it turns out, almost six centuries of the family joints. I wrote the diagnostic criteria for hypermobility type down the left side of a piece of paper, and on the right side, my symptoms that matched and why they matched. I took the list in to my new primary care physician, and said, "This is what I have, isn't it?" Unlike so many of you, I was lucky enough to have a doctor that could think and took me seriously. He looked it over and said, "You're right."

That was my first step on a path that led from the origins in the 1400s of the mysterious family joints to the foundation which helped me out so much in those first years, and the path to so much new information and study, transforming my life and spirit.

Along that path I came to the NIH/NIA study led by Nazli McDonnell and Clair Francomano. They confirmed my diagnosis, but better, brought two of the more exceptional people I've known into my life. Nazli is a fiercely invested researcher; her work on the longterm study resulted in many of



the papers that are the basis for what we know about the EDS. I miss working with her (our writing collaboration was perhaps the easiest I've ever had the privilege of knowing). I'm happy she found a medical home with the VA. Clair continues the good fight to help as many of us as she can in as many ways as possible.

They were my guides as I learned about EDS through my early years with the foundation. Somewhere in those conversations and meetings (2007 or 2008) I raised the question about an international conference, like the one in 1997, to reexamine the criteria since they were learning so much. Nazli said about five years. But the financial outlook in the states worsened, and so many changes came to the foundation as a result that I put the idea away.

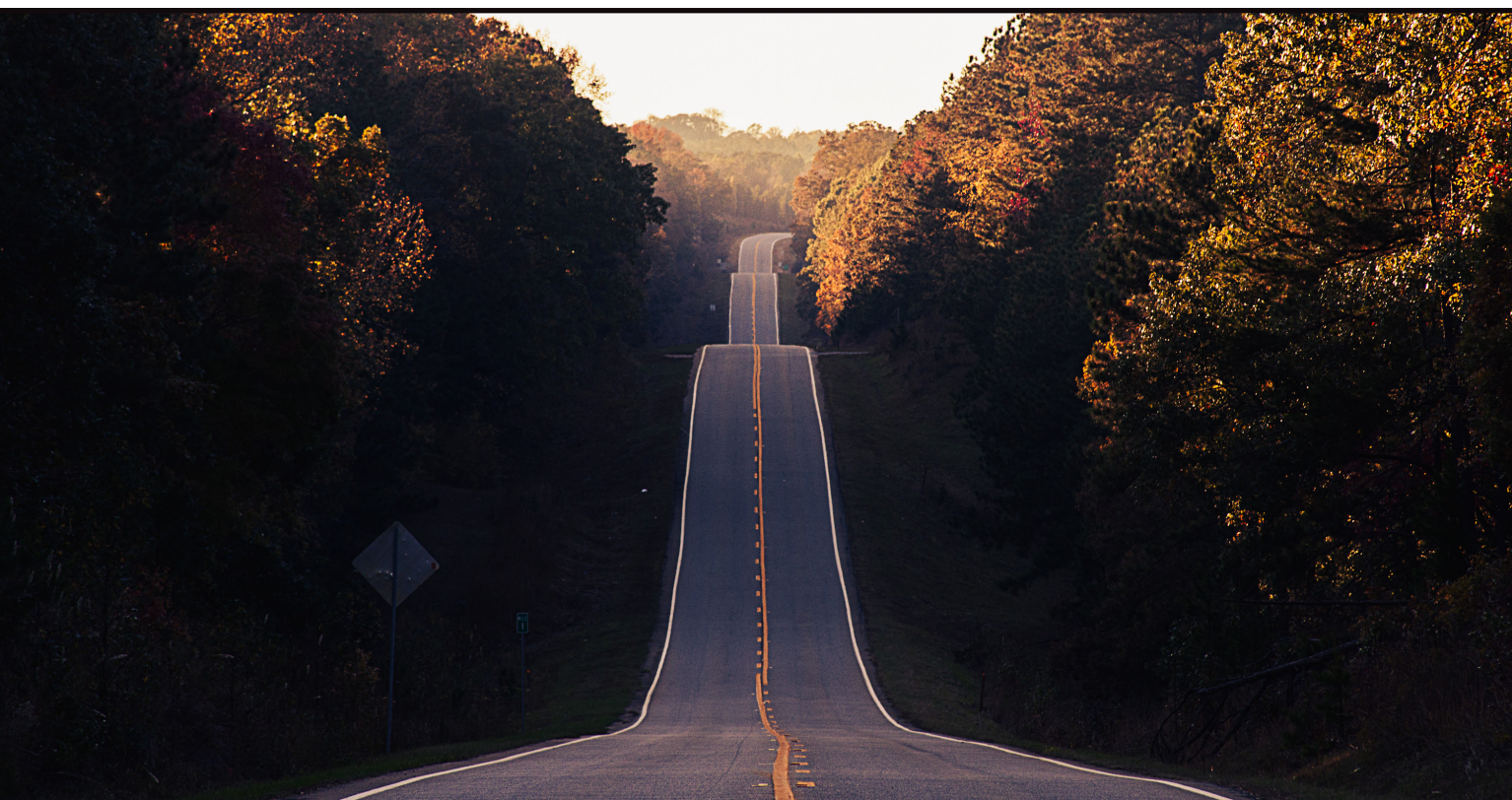
Lara Bloom came to the foundation with the drive to get that done, and the vision to set up a recurring examination every two years. Along with Dr. Brad Tinkle, she brought together so many around the world to bring the EDS International Consortium together, leading to the New York Symposium in May last year where the first drafts of the new criteria and other papers were unveiled. After much more discussion, then peer review, the finals were published in March – leading to much public discussion (with several 18-hour days for me

that month) and some controversy. In two years, we'll do it again.

I started doing my work with the foundation to give back for their help in getting me diagnosed, so I could help others the same way. I continued to do it because someone had to, and for good or ill, I'd learned a hell of a lot about EDS. But this is the longest I've spent with a single organization. The days when the work is physically or emotionally too much are days when I would say to myself, "Just last until the next nosology." The heredity of my matriarchal lineage for almost six centuries has been answered. I've lasted beyond the point I once used as my ultimate goal. I would be satisfied, even happy with that much. But I go on, because I can still help. I go on.

"Perform unceasingly the works that must be done, for the man detached who labors on to the highest must win through. Moreover, you should embrace action for the upholding, the welfare of your own kind. Whatever the noblest does, that too will others do: the standard that he sets all the world will follow." (adapted from the Bhagavad Gita by Constance DeJong for Philip Glass, *Satyagraha*)

**MARK C. MARTINO**





# Joint Hypermobility Syndrome and Postural Orthostatic Tachycardia Syndrome (HyPOTS)

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## Abstract

**Objective:** To evaluate the association between joint hypermobility syndrome associated with postural orthostatic tachycardia syndrome (HyPOTS) and fibromyalgia.

**Methods:** A chart review of patients followed at two outpatient Rheumatology facilities within one hospital system were analyzed. Thirty-seven patients (3 male, 34 female) diagnosed with HyPOTS and fibromyalgia were reviewed.

**Results:** HyPOTS is a chronically disabling musculoskeletal disorder presenting clinically as widespread musculoskeletal pain and/or fatigue with joint hypermobility. However, HyPOTS may be inadvertently diagnosed as chronic fatigue syndrome or fibromyalgia. In fact, the group of fibromyalgia patients evaluated at a rheumatology outpatient clinic were found to meet the clinical criteria for HyPOTS.

**Conclusions:** Patients diagnosed with fibromyalgia and chronic fatigue syndrome may benefit from a further evaluation for HyPOTS. Thus, if HyPOTS is properly defined, a wide range of therapeutic options for these patients become available in order to improve their physical manifestations and quality of life.

## Introduction

Often, the most common complaints voiced by patients in a rheumatology office are that of increased arthralgias, myalgias, and constitutional symptoms. It is not uncommon to receive a referral for evaluation of a patient with complaints of arthralgias, myalgias and other constitutional symptoms but without obvious signs of synovitis or connective tissue disease. It is also common to have these complaints attributed to patient's underlying disease due to their nonspecific nature. However, nonspecific arthralgias, myalgias, and fatigue are just that – nonspecific. Yet, these symptoms can lead to significant decline in a patient's quality of life. As physicians, we consistently strive to treat disease and to improve a patient's quality of life. This may mean prescribing a treatment strategy, including immunosuppressive therapy, and taking on the risk of increased side-effects. We propose that in select patients, joint hypermobility may be contributing to these non-specific complaints and that recognition and intervention can help to improve the patient's quality of life.

Joint hypermobility syndrome (JHS) is a chronic, disabling disorder which manifests as widespread musculoskeletal pain and/or fatigue in the presence of generalized joint hypermobility. Joint hypermobility can exist without any associated musculoskeletal disorder. Although a limited literature is available on joint hypermobility; approximately 5% of the adult Caucasian population has JHS with an increased prevalence noted in Asian and African-American populations [1]. Furthermore, the prevalence of JHS is noted to be approximately 10-30% in the juvenile population which decreases with age [2]. Despite the fact that there is a limited understanding of the pathophysiology of JHS, genetics do appear to play a role. For example, in studies performed on the expression of the tenascin-X gene, the protein product of the gene has been noted to be present in low amounts in patients diagnosed with JHS and the Ehlers-Danlos Syndrome (hypermobility type) [2].

Furthermore, environmental triggers, such as viral illness or extended periods of deconditioning, have been proposed as causes for benign joint hypermobility transitioning into JHS [2].

In addition to musculoskeletal pain and fatigue, a subset of patients with JHS have also been found to suffer from visceral manifestations and postural tachycardia with orthostatic intolerance. Visceral manifestations often center upon gastrointestinal dysfunction. Of note, abnormalities in proprioception have also been observed in patients with hypermobility along with exaggerated blood pooling in the lower extremities, suggesting some sort of molecular defects in blood vessels [3]. Postural orthostatic tachycardia syndrome, (POTS), is also noted in patients with collagen disorders and hypermobility, which is defined as an increase in heart rate greater than 30 beats/minute within 10 minutes of standing despite a lack of decline in blood pressure. Patients often report pre-syncope or syncope events and associated feelings of lightheadedness, nausea and palpitations [4].

POTS has been found to be associated with sympathetic hyperactivity and small fiber neuropathy. Similarly, fibromyalgia has been associated with sympathetic hyperactivity [5]. Although there is minimal research available, case reports of adults diagnosed with fibromyalgia/chronic fatigue syndrome [2] and studies of joint

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hypermobility performed in juveniles diagnosed with chronic fatigue syndrome (CFS) [6] shed light on patients who may inadvertently be diagnosed with fibromyalgia/CFS in the setting of joint hypermobility and POTS, or HyPOTS as we have termed this clinical entity at our medical facility.

## Materials and methods

### Case studies and chart review

Initially, we reviewed a case of a patient diagnosed with lupus and joint hypermobility whose clinical presentation was consistent with HyPOTS. This case review led to us to construct a protocol for review by the University Hospitals Cleveland Medical Center IRB for a review of the charts of patients with joint hypermobility and to enable us to report on clinical manifestations. This chart review study was carried out in compliance with the Helsinki Declaration. Thus, following IRB Protocol #09-13-25, entitled, Joint Hypermobility Syndrome and POTS (HyPOTS) was approved to review the charts of patients seen at two outpatient Rheumatology facilities of University Hospitals Cleveland Medical Center. The charts of 37 patients between the ages of 18 and 58 who were diagnosed with JHS using the Brighton 1998 Criteria for the diagnosis of benign joint hypermobility syndrome were reviewed [7].

All of the patients included in this chart review study had presented with one or more of the following symptoms: chronic fatigue, generalized musculoskeletal pain (arthralgia/myalgia), autonomic dysfunctions (e.g. pre-syncope events, palpitations, lightheadedness, hypotension), gastrointestinal symptoms (i.e. irritable bowel syndrome type), and anxiety.

Additionally, all of the patients carried the diagnostic label of fibromyalgia. Similar to research previously performed on JHS, there was a female predominance. In that regard, males made up only 3 of 37 patients who were evaluated. The mean age of these patients at the time of clinical evaluation was 34.6 years.

The charts of these 37 patients with a diagnosis of JHS were analyzed (Table 1). Twenty-three (62%) of the patients had a positive tilt table test consistent with the diagnosis of postural orthostatic tachycardia syndrome (POTS) whereas 7 (19%) of these patients had a negative tilt table test. Seven patients (19%) did not have a tilt table performed at time of this analysis because of multiple reasons, including financial limitations and non-compliance.

Along with fulfilling the Brighton 1998 Criteria for JHS, the patients included in our case study were noted to have clinical signs of autonomic dysfunction, including reported pre-syncope events and lightheadedness. After applying a standard of care treatment regimen, including education, physical therapy, increased salt and water intake, mineralocorticoid supplementation and compression stockings, a chart analysis documented improvement in the clinical symptoms of all patients analyzed.

## Discussion

The case series study was performed at a Rheumatology outpatient facility after noticing a trend towards joint hypermobility in certain patients with associated reports of fatigue and fibromyalgia-type symptoms. After treating multiple patients with joint hypermobility, often in patients with primary connective tissue disease, the significant disability and decrease in reported quality of life amongst these patients with joint hypermobility was apparent. Although these patients often had adequate control of their underlying disorders they reported significant suffering and symptomatology, which was not relieved with

Abbreviations for patients with concurrent rheumatic diseases; SpA: Spondyloarthritis; UCTD: Undifferentiated Connective Tissue Disease; SLE: Systemic Lupus Erythematosus; RA: Rheumatoid Arthritis; SS: Sjögren's Syndrome.  
-- indicates no diagnosis of concurrent rheumatic disease

**Table 1.** Demographics, Clinical Signs of Autonomic Dysfunction, Tilt Test Results and Concurrent Rheumatic Diseases of Patients Diagnosed with JHS

Patient	Age	Gender	Clinical Signs of Autonomic Dysfunction	Tilt Table Test Results	Concurrent Rheumatologic Diseases
1	22	Female	+	+	SpA
2	39	Female	+	+	--
3	38	Female	+	+	SLE
4	28	Female	+	+	Behcet's
5	32	Female	+	Not performed	UCTD
6	40	Female	+	-	--
7	37	Female	+	+	SLE
8	28	Female	+	+	Discoid Lupus
9	32	Female	+	+	--
10	48	Female	+	-	SS
11	36	Female	+	+	UCTD
12	32	Female	+	+	--
13	38	Female	+	+	SS
14	39	Female	+	+	UCTD
15	40	Male	+	Not performed	--
16	35	Female	+	-	--
17	35	Female	+	-	RA
18	26	Female	+	+	UCTD
19	33	Female	+	Not performed	--
20	22	Female	+	+	SS
21	26	Female	+	Not performed	SS
22	33	Female	+	+	--
23	55	Female	+	+	SS
24	58	Female	+	+	SS
25	28	Female	+	+	--
26	40	Female	+	+	--
27	44	Female	+	+	UCTD
28	27	Female	+	+	Behcet's
29	44	Female	+	-	--
30	31	Female	+	Not performed	UCTD
31	18	Female	+	Not performed	--
32	31	Female	+	+	--
33	44	Female	+	-	UCTD
34	30	Male	+	-	--
35	28	Female	+	+	SLE
36	30	Female	+	Not performed	SLE
37	40	Male	+	Not performed	--

increasing immunosuppression, despite attempts at advanced therapies in some patient cases. The patients were most commonly treated with increasing glucocorticoid therapy for presumed flare and at times, the patient's individualized immunosuppressive regimens were subject to increased dosing.

A systematic literature search was performed when we began to recognize a connection between our patients with joint hypermobility, fatigue, fibromyalgia-type symptoms and autonomic dysfunction. A detailed literature review of the PubMed database retrieved a few case reports of patients with hypermobility and chronic fatigue or fibromyalgia labels, which was more commonly found in adult Ehlers-Danlos Syndrome patients, whereas a few papers discussed the relationship between chronic fatigue and joint hypermobility in children. However, we were unable to find an extensive case series as noted in our facility for evaluating patients for joint hypermobility,

autonomic dysfunction, and the associated and commonly reported poor quality of life in our patients.

We discovered that there was a selective group of patients with JHS who have autonomic dysfunction which contributes to the clinical manifestations with this creating a disorder in otherwise benign joint hypermobility situations. Thus, patients can often be inadvertently labeled at diagnoses with fibromyalgia, chronic fatigue or as having active connective tissue disease. These labels can interfere with treatment decisions. Potential manifestations of autonomic dysfunction can also occur which include cardiac dysrhythmias, postural orthostatic tachycardia syndrome, orthostatic hypotension and orthostatic intolerance. Although the underlying physiological mechanisms leading to such phenomena in JHS have not been clearly identified, they are likely to include reduced vascular tissue elasticity, and impaired peripheral vasoregulation as a consequence of adrenoreceptor or neuronal abnormalities [9].

An evaluation of JHS using the Brighton 1998 Criteria is a tool employed to help identify patients who may be falsely labeled as fibromyalgia and/or chronic fatigue and who may benefit from the interventions previously listed. Patients often fit criteria for multiple diagnoses and recognizing that a patient may coexist in multiple categories is necessary for adequate treatment. White et al. [10] reported on 74 patients labeled with fibromyalgia and found that 58% of the patients also met 1988 CDC criteria for the diagnosis of chronic fatigue syndrome. Branco et al. [11] reported on the prevalence of fibromyalgia in the general population compared to rheumatology outpatients using the London Fibromyalgia Epidemiological Study Screening Questionnaire (LFESSQ). In that study, 32% of rheumatology outpatients screened positive for significant pain and fatigue compared to 13% in the general population. Fourteen percent of these rheumatology outpatients were labeled as fibromyalgia compared to 6.7% in the general population [11]. Accordingly, the US Centers for Disease Control reports of fibromyalgia coexisting 25-65% of the time in connection with other connective tissue diseases have been noted [12]. As discussed by Ahn and Goldman-Ramsey [13], there remain limitations in measurements of fatigue in SLE whereby research showed fatigue experienced in 53-80% of lupus patients and the pain/fatigue combination experienced in 95% of lupus patients. Given the significant impact of HyPOTS on the quality of life, these patients should be screened for hypermobility accordingly.

The diagnosis of HyPOTS is recognized as leading to increased treatment options for patients suffering from the aforementioned manifestations of this disorder. This would involve, a well-rounded approach to managing postural tachycardia and include adequate salt and water intake, compression stockings, mineralocorticoid administration, along with physiotherapy techniques, consisting of joint and core muscle strengthening, orthotics, which taken together can improve these patients symptoms and quality of life [7]. In fact, the main point to be taken away from this retrospective analysis is that many of the clinical symptoms attributed to primary connective tissue disease could, in fact, be related to joint hypermobility. Once made aware of this problem from which a patient's autonomic disturbance is confirmed by a positive tilt table test, future instructions to the patient start with improving support of the lower extremities through the use of compression stockings and adequate salt and water intake. Providing the patient with an overall awareness of the problem empowers the patient to take control of the symptoms.

The patients in our study continue to be followed within our rheumatology division for symptomatic management of HyPOTS,

often in conjunction with a team of specialists, including those practicing cardiology and neurology. We have found that treatment of HyPOTS using the interventions indicated above have led to significant improvement in the quality of life of these patients as we continue to recognize new cases of patients with HyPOTS.

## Significance and innovations

- Joint hypermobility syndrome is a chronic disorder which presents with arthralgia, myalgia and at times in association with visceral manifestations and POTS (postural orthostatic tachycardia syndrome).
- Patients with joint hypermobility syndrome are more likely to be diagnosed with chronic fatigue syndrome or fibromyalgia compared to the general population.
- HyPOTS (joint hypermobility syndrome associated with postural orthostatic tachycardia syndrome) has been underrepresented in rheumatologic literature. Clinical findings of HyPOTS may be considered to be complications of patient's underlying connective tissue disease, resulting in increased treatment of the connective tissue disease.

Interventions, such as maintaining adequate hydration along with use of compression stockings, rather than intensifying the treatment of an underlying connective tissue disease, lead to improvements in symptoms.

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## Conflict of interest

None to be declared.

## References

1. Hakim A, Grahame R (2003) Joint hypermobility. *Best Pract Res Clin Rheumatol* 17: 989-1004. [[Crossref](#)]
2. Folci M, Capsoni F (2016) Arthralgias, fatigue, paresthesias and visceral pain: can joint hypermobility solve the puzzle? A case report. *BMC Musculoskelet Disord* 17: 58. [[Crossref](#)]
3. Gazit Y, Nahir AM, Grahame R, Jacob G (2003) Dysautonomia in the joint hypermobility syndrome. *Am J Med* 115: 33-40. [[Crossref](#)]
4. Haensch C, Tosch M, Katona I, Weis J, Isenmann S (2014) Small-fiber neuropathy with cardiac denervation in postural tachycardia syndrome. *Muscle Nerve* 50: 956-961. [[Crossref](#)]
5. De Wandele I, Calders P, Peersman W, Rimbaut S, De Backer T, et al. (2014) Autonomic symptom burden in the hypermobility type of Ehlers-Danlos syndrome: A comparative study with two other EDS types, fibromyalgia, and healthy controls. *Semin Arthritis Rheum* 44: 353-361. [[Crossref](#)]
6. Barron DF, Cohen BA, Geraghty MT, Violand R, Rowe PC (2002) Joint hypermobility is more common in children with chronic fatigue syndrome than in healthy controls. *J Pediatr* 141: 421-425. [[Crossref](#)]
7. Grahame R, Bird HA, Child A (2000) The revised (Brighton 1998) criteria for the diagnosis of benign joint hypermobility syndrome (BJHS). *J Rheumatol* 27: 1777-1779. [[Crossref](#)]
8. Benarroch EE (2012) Postural tachycardia syndrome: a heterogeneous and multifactorial disorder. *Mayo Clin Proc* 87: 1214-1225. [[Crossref](#)]
9. Bonyhay I, Freeman R (2004) Sympathetic nerve activity in response to hypotensive stress in the postural tachycardia syndrome. *Circulation* 110: 3193-3198. [[Crossref](#)]



10. White KP, Speechley M, Harth M, Ostbye T (2000) Co-existence of chronic fatigue syndrome with fibromyalgia syndrome in the general population. A controlled study. *Scand J Rheumatol* 29: 44-51. [[Crossref](#)]
11. Branco JC, Bannwarth B, Failde I, Abello Carbonell J, Blotman F, et al. (2010) Prevalence of fibromyalgia: a survey in five European countries. *Semin Arthritis Rheum* 39: 448-453. [[Crossref](#)]
12. Fibromyalgia | Arthritis | CDC. Cdcgov. 2016. Available at: <http://www.cdc.gov/arthritis/basics/fibromyalgia.htm#> Accessed August 3, 2016.
13. Ahn GE, Ramsey-Goldman R (2012) Fatigue in systemic lupus erythematosus. *Int J Clin Rheumatol* 7: 217-227. [[Crossref](#)]

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