

# LOOSE CONNECTIONS



**POETRY ISSUE**

**2016 Donors**

**Ehlers-Danlos Syndrome—Hypermobility Type:  
A Much Neglected Multisystemic Disorder**



The  
**Ehlers  
Danlos**  
Society.

Your Magazine About Living With EDS  
**Winter 2016**





## TABLE OF CONTENTS

<b>Hope Begins With You</b> .....	3
<b>From the Editor's Desk: "And acquainted with grief" / Mark C. Martino</b> .....	4
<b>So Beautiful, It Hurts / Amanda Lynn Woodrum</b> .....	7
<b>Substitute / Fiona Branson</b> .....	8
<b>My EDS Journey / Chelsie Hedges</b> .....	10
<b>Mirrors / Sophie Niall</b> .....	11
<b>The Last Time I Climbed a Mountain / Gwen Miller</b> .....	12
<b>Coping With Chronic Illness: A Teenager's Two Cents (Part Two) / Dinah Everton</b> .....	13
<b>I WANT TO BREAK OUT (Trapped by EDS) / Sean O'Brien</b> .....	15
<b>The Push Puppet / David Doukas, MD</b> .....	16
<b>When Hugs Hurt / Lorna Stewart</b> .....	17
<b>I Am Real, So Do I Exist? / Michelle Hardy</b> .....	18
<b>I Wake Up Before My Alarm / Kaitlyn Brennan</b> .....	19
<b>You Don't Look Sick / Linda Wilson</b> .....	20
<b>Our Donors in 2016</b> .....	21
<b>Ehlers-Danlos Syndrome—Hypermobility Type: A Much Neglected Multisystemic Disorder</b>	
Yael Gazit, MD, MSc, Giris Jacob, MD, PhD, and Rodney Grahame, CBE, MD, FRCP, FACP .....	27
Photography Credits .....	36
Board of Directors, Medical & Scientific Board, Publisher Information .....	37

## 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 and related disorders.

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

**WWW.EHLERS-DANLOS.COM**

General Email: [info@ehlers-danlos.com](mailto:info@ehlers-danlos.com)  
Submissions: [stories@ehlers-danlos.com](mailto:stories@ehlers-danlos.com)

P.O. Box 87463  
Montgomery Village, MD 20886 USA  
Phone: 410-670-7577

The Ehlers-Danlos Society – Europe  
Office 7, 35-37 Ludgate Hill, London EC4M 7JN UK  
Phone: +44 203 887 6132



# Hope Begins With You

**O**UR INAUGURAL YEAR AS THE EHLERS-Danlos Society has been one of the most productive in over three decades of history as an organization.

We have come a long way since our 1985 founding by Nancy Rogowski as the Ehlers-Danlos National Foundation. From our first biannual Ehlers-Danlos International Symposium—where over 200 medical professionals gathered to refine the nosology, diagnostic criteria, and treatment protocols for the entire range of Ehlers-Danlos syndromes—to our annual Global Learning Conference and establishment of our new eminent medical and scientific board, we've made great strides in the advancement of Ehlers-Danlos research, awareness, advocacy, and patient care.

With investments of thousands of donors throughout the world, we are building a new infrastructure for discovery, treatment, and community—globally.

As we strengthen, our future brightens. Together, we envision a day where early diagnosis means better management and even better patient outcomes. Across a not-so-distant horizon, we picture ever more productive, energetic lives, enveloped by the light of mutual support; steeled against pain; and steadied by the help of medical professionals fully steeped in their knowledge of Ehlers-Danlos and all of their comorbidities.

Join us. Help build the first-ever International Ehlers-Danlos Patient Registry. Help unlock the genomic secrets of Ehlers-Danlos and their multi-systemic effects in areas ranging from gastrointestinal, autonomic, mast cell, and neurological complications, to fatigue and chronic pain.

Together, we pray for the day when no child of Ehlers-Danlos comes into the world without the full support of well-informed, properly trained health professionals providing a world class standard of care.

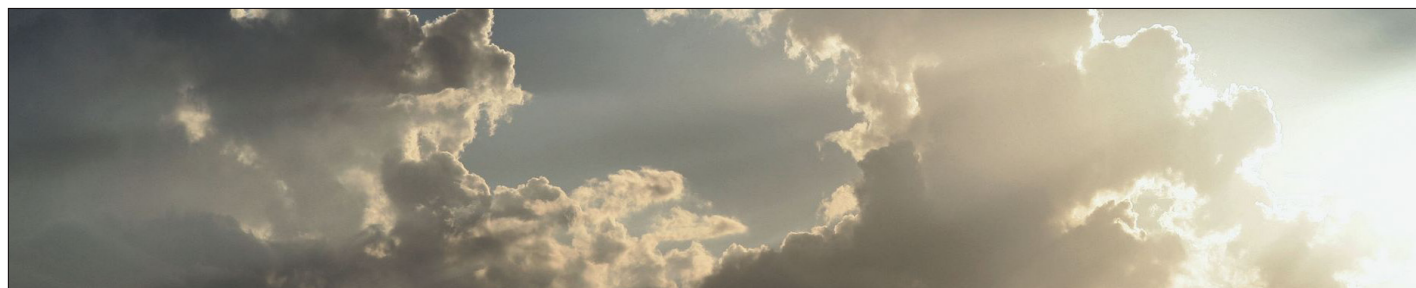
**Our strength begins with hope.**

[Join us.](#)

**Our hope begins with you.**

**Thank you.**

**LARA BLOOM AND SHANE ROBINSON**  
CO-EXECUTIVE DIRECTORS



# From the Editor's Desk: "And acquainted with grief"

**A**LL OF US ARE ACQUAINTED WITH GRIEF.  
Every one of us mourns, each in our own way.

I've tried to think of a less personal way to write this, but it seems to be impossible. Grief is a personal experience for each of us, and all I can really do is discuss my own. What should be remembered as you read this is I've survived all of it, that's ultimately the point; none has erased the good times or any of the joy I had, each has woven into the fabric of my life.

**"Good times and bum times, I've seen 'em all  
And, my dear, I'm still here."**

*Stephen Sondheim, "I'm Still Here"*

I lived through the AIDS era, during which I lost ex-lovers and a long list of friends I had hoped to have the rest of my life. I still don't know how I survived myself, except pure chance—I spent decades not planning for a future because I was sure I was going to follow them. I haven't.

I have survived the deaths of grandparents and parents. My father's father retired and went on

a road trip from Ohio almost immediately; he stopped to see us in Virginia, then died a couple of weeks later in Lubbock from sudden, "galloping" leukemia. My father died from Alzheimer's and vascular dementia after a long descent. My mother had multiple heart attacks and went into a coma during her Mother's Day dinner in 1993, and we had to decide to follow her wishes and take her off life support. I just had my 60th birthday, and as of January 4 have lived longer than she.

Perhaps the best thing said to me after Mom's death came from an acquaintance who had dealt with the early death of her mother; she told me she hadn't gotten over it, that time hadn't reduced the wound, and she didn't expect it to ever disappear, although it had become easier to hold the grief. I was grateful to hear that. I didn't want to forget. Ever. Rose Kennedy said it perfectly: "It has been said, 'time heals all wounds.' I do not agree. The wounds remain. In time, the mind, protecting its sanity, covers them with scar tissue and the pain lessens. But it is never gone." People we love are too important to us to heal completely from the wound of their leaving us behind.

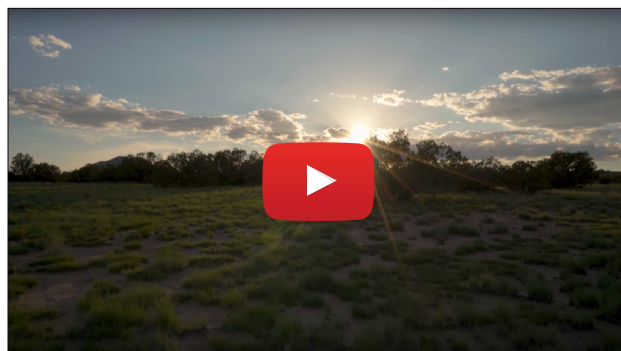
*Lascia ch'io pianga  
mia cruda sorte,  
e che sospiri  
la libertà.*

**Allow me to weep  
for my cruel fate,  
and to mourn  
my lost freedom.**

*Il duolo infranga  
quests ritorte,  
de' miei martiri  
sol per pietà.*

**May my sorrow break  
these chains,  
if only out of pity  
for my suffering.**

G.F. Handel's *Rinaldo*  
Libretto Giacomo Rossi; translation Susanna Howe.



**You**Tube

Joyce DiDonato & Il Pomo D'oro  
<https://youtu.be/PrJTmpt43hg>





**"So get hit in your head  
And there may be a few things  
you can't recall at all  
But you get hit in your heart  
And you're in pieces and parts  
Pieces and parts."**

*Laurie Anderson, "Pieces And Parts"*

There are other forms of grief, and some of these are familiar to us chronically ill. Some are surprised by their diagnosis; grief over the loss of their expected life and dreams is immediate and sharp. Serious illness is a major event; we mourn the loss of independence, of friends we held dear, of activities we loved.

For others like me, diagnosis is the confirmation of a life-long struggle, the next in a seemingly infinite sequence of farewells to parts of life.

My earliest memories are of the family joints, and of the things I wanted but couldn't manage. No being able to ice-skate while I was a kid in Québec. Struggling to find how I could manage to swim without dislocating my shoulders. As a young teenager not taking the offer of a European tour as a pianist—because even then I couldn't see success coming to me with the body I had. Discovering parts of the Army ROTC training program for my scholarship were impossible for me to perform at all.

Diagnosis gave me answers, but no closure. I realized with hypermobile fingers it was no wonder I couldn't rid myself of wrong notes, no matter how much I practiced—and why was I surprised when my back began to resent the abuse I'd placed on it? So, I gave up totally on piano, then on music altogether because not performing hurt too much to think about. Then on theater, too, when I started realizing I couldn't rely on my body enough to safeguard other actors onstage. Finding a new profession in graphic design, to understand eventually I could no longer respond quickly enough to maintain success. And all of that change, all the upheavals, become the ordinary, so one is constantly on watch for the next. But that's grief.

In 1969 psychiatrist Elisabeth Kübler-Ross wrote *On Death and Dying* about her experiences with

terminally ill patients. She described a series of stages people go through when confronted by loss. While widely accepted, it is not consistently supported by further research, and it was only a model of common experiences with grief. The five stages provide a framework for talking about grief, and I suspect most of us know of them: denial, anger, bargaining, depression, and acceptance. Not everyone visits all the stages in sequence or even at all; sometimes they're helpful to help work through a loss, but sometimes they can be a hindrance, because grief isn't a straight road that we all travel identically.

**"The grief we feel over the life we lost may re-emerge now and then...indefinitely. Grief comes in waves, and can arrive unexpectedly. One moment, we can feel accepting of the changes in our lives. The next minute we can be overcome by sadness. A simple interaction can trigger it...The grieving process I've gone through as a result of chronic illness has been one of the most intense of my life."**

*Toni Bernhard, JD, <https://www.psychologytoday.com/blog/turning-straw-gold/201405/3-things-the-chronically-ill-wish-their-loved-ones-knew>*

We are visited in mourning by many emotions more than the five stages suggest—emptiness, loneliness, helplessness, disconnectedness. But it's not just our emotions that get overwhelmed. Grief affects us in every way, including physically and cognitively. It can be hard to find the energy to take care of ourselves, even though that is essential to truly come to terms with living with losses. When our world changes so completely, when what we cherished and loved is taken away from us, our minds pull us inward. Our sleep patterns go awry—we sleep too much, or not at all. We can be tired all the time, even right after waking up; or we might become hyperactive, unable to sit. And almost anything EDS has forced on us can get worse: aches and pains, digestive problems, headaches, allergies, increased sensory sensitivity. You begin to feel like stranger in your own life. You begin to think your future is lost.

Let yourself mourn. Don't try to be strong, to force yourself to move on before you're ready. Mourning



cannot be denied, it can only be delayed. Listen to your mind; healing from grief starts inward. Listen to your body. Ask for help if you need it; accept help when it's offered.

**"When my husband, Fred, died, my father told me that time does not heal all wounds but gives us the tools to endure them. I have found this to be true in the greatest and smallest of matters."**

*Patti Smith*

Be sure you're eating well, and drinking enough water. Breathe. Rest when you can, find ways to get outside yourself, like meditation, or music, or art, or theater and movies. Breathe. Get some exercise, even if it's simply walking; movement will help remind your body what normal used to be. Breathe. Don't cut yourself off from human contact, don't be afraid to touch. Breathe. Make your body feel more normal by keeping clean, wearing decent clothes, getting your hair cut. Breathe. If you think you need the help, find a counselor either through your doctor or perhaps your church; and if you feel in danger, find your doctor. Breathe. Keep a journal. Write, paint, play. Breathe.

Forgive yourself for surviving. Forgive yourself for living. It will get easier. You won't forget the people you want to remember. You won't forget the good times you had before your life changed, and you won't stop having good times afterward. But it will get easier to remember and easier to choose to live. Love. Take each moment as it comes, and each moment, choose as well as you can. As we all do.

The bad news is: Nothing lasts forever.

The good news is: Nothing lasts forever.

**MARK C. MARTINO**

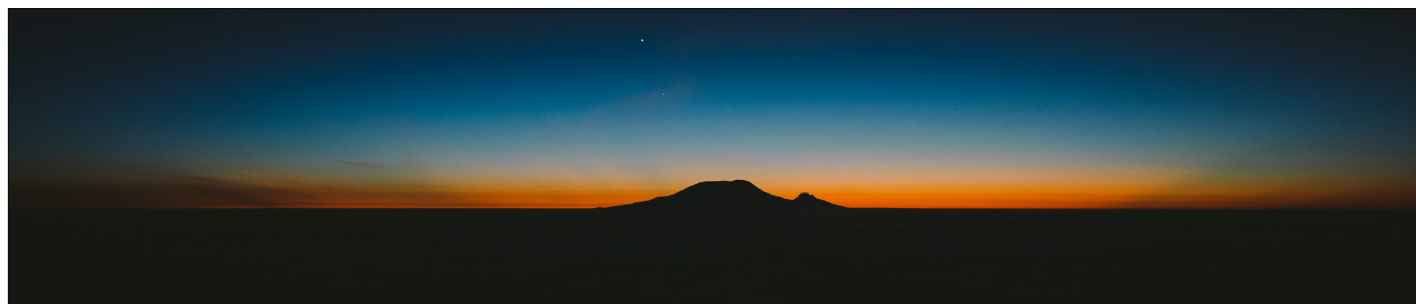
**Sure on this shining night  
Of star made shadows round,  
Kindness must watch for me  
This side the ground.  
The late year lies down the north.  
All is healed, all is health.  
High summer holds the earth.  
Hearts all whole.  
Sure on this shining night  
I weep for wonder wand'ring far alone  
Of shadows on the stars.**

The poem comes from a book by James Agee entitled "Permit Me Voyage" published 1934 by Yale University Press © by owner, provided at no charge for educational purposes.



**You Tube**

Music by Morten Lauridsen, words by James Agee.  
Sung by Conspirare.  
<https://youtu.be/-R67jhPhXuk>





# So Beautiful, It Hurts

They always told me,  
 "You're so beautiful, it hurts."  
 They have no idea  
 about my pain  
 about what jerks and jars  
 and pulls my bones apart.  
 They don't know about this smile  
 this plastic face  
 this constant grieving gracious grace  
 they way I fake and fake and fake and fake.  
 The way I break.

If I had a dollar for each time  
 they told me I am beautiful,  
 I would be rich.  
 I would climb mountains and travel far!  
 Why am I so poor?  
 How much more?  
 Will it ever end?  
 Will it ever begin?  
 Is this the middle?  
 Is this my total life?  
 I know this;  
 This is my wasted waist.  
 This is the nauseous taste.  
 This is my beautiful, bitter face.

This is how I face the world  
 To be beautiful and brave and bold  
 When this body feels ancient  
     and decayed and old.  
 How will it carry me  
 through age, through time, through years?  
 I'll ride the tide of my tears.  
 Baptize this beautiful self  
 Cleanse these scars that mark  
 the passage of time  
 Marks on this existence of mine  
 This life I hate to love  
 As this face fills with lines

from smiles or frowns—  
 I cannot remember.  
 I can't remember the last time I felt joy.  
 Or love  
 that wasn't for a child  
 who was pure, and innocent  
 and naive  
 And didn't care that inside I felt so so so ugly.

They always told me  
 "You're so beautiful, it hurts."  
 They didn't know how they hurt  
     me with their words.  
 How their expectations coerced me  
 to live beyond my means  
 How their demands turned  
 this innocent soul mean.  
 How everyone who ever fell in love with me  
 just wanted something;  
 (A piece of me)  
 (The peace inside me)  
 How my heart churns  
 and my body bursts at the seams.  
 WHAT DOES IT MEAN?!  
 Why does this beautiful existence  
 feel so cruel?  
 What must I do  
 to turn this death into life  
 this darkness to light  
 this small shallow shell  
 into a force that can fight?  
 To travel forward;  
 in knowing,  
 and understanding,  
 and accepting,  
 this beautiful face  
 this terrifying  
 fickle fate.

**AMANDA LYNN WOODRUM**



# Substitute

**T**ILLY SHIVERED AS THE CHILL WIND blew across the back of her bare, slightly bow-backed legs, as she stood on the edge of the Netball pitch. She was hoping desperately on the one hand, that she wouldn't get picked for anyone's team, and wishing on the other, that she could just go indoors and get warm. Her muscles were tightening in the icy wind, and she feared a muscle spasm, whilst an aching pain crept from the cold ground through to her feet.

"I pick Tilly," a confident voice piped, and miserably, Tilly duly crossed the pitch to join Rachel's team. "You're in goal," said Rachel. Seeing Tilly's alarm at this prospect, she said, "Don't worry – we'll wallop 'em. You'll never see them down your end."

Nodding her head, Tilly trudged to the goal area. She hated netball. The number of times her shoulder had dislocated during the game had already made it past the score – yet they just kept making her play. She sighed. It wasn't that she hated sport; she just hated the kind of sport that resulted in a casual blow to the shoulder, perhaps with a ball, perhaps as an accidental collision, which would leave other girls laughing and maybe rubbing their shoulders ruefully, but would leave Tilly with an entirely dislocated shoulder.

It is hard to explain what it feels like, when your shoulder—or indeed any other joint—in your body, dislocates, if you are lucky enough never to have had the misfortune to experience it.

The best way Tilly could describe it was to ask you to imagine her arm, like that of an old jointed china doll, where the stringing of the joints has been stretched to the point where now each limb falls floppy from its socket. Sometimes she would lose all sensation in her arm when her shoulder dislocated.

Always though the pain in the shoulder, and in all the muscles around the shoulder, stretched now almost to breaking point, nerves trapped and screaming, would be immense. It is reckoned by doctors that the pain of a dislocation is far greater than the pain of a broken bone.

Yet Tilly loved gym, loved acrobatics, loved dance, for these were controlled movement, controlled exercise, in which the risk of being accidentally thumped by a girl twice your size was significantly reduced. Such were Tilly's thoughts as she trudged down the pitch. Once in goal, she turned to survey her team, and was reassured that she was likely on the winning side. The goalkeeper always got chosen last and the one person who loved that position





more than any other was Denny Malone, a seriously well-built girl who towered above the rest of the class, and was always picked first for goalie whatever the sport. Denny waved to Tilly from the other end of the pitch, and Tilly waved back. Both she and Denny shared the dubious honour of “freak” status amongst the girls, Denny for her height and size, Tilly for her fragile dislocating joints.

The whistle blew and the game began. Rachel's predictions proved accurate for nearly all of the first ten minutes, with all the action centered on the opposing goal. Tilly was left to stand and stamp, trying desperately hard to keep warm as the bitter easterly wind blew across the school playground.

She wished she could just go indoors and get warm. In every single one of her joints that had ever dislocated, she now felt that old aching pain intensify. To take her mind off the pain, Tilly was thinking of the hot showers that would follow this nightmare.

A voice yelled, “Tilly!” She spun around, to get hit hard on her left shoulder so that with a sickening crunch it dislocated. Tilly collapsed onto the freezing ground.

The whistle parped *peep-peep* sharply, and Miss Jones strode over to Tilly. She barked, “Are you alright?” The other girls crowded round.

“It’s just my shoulder, it’s dislocated again,” said Tilly.

Miss Jones clucked in a disapproving manner. “Such a nuisance,” she said. “Let’s get you off the pitch.”

“No! Wait!” said Tilly. The girls watched horrified and mesmerised as, with a sickening sharp turn and crunch, Tilly re-located her shoulder into its socket. Some one cried, “Yeuch.” Another said, “That is *disgusting*.”

With her shoulder back in place, Tilly stretched out her right hand seeking help from the crowd, but all turned their heads away – save for Denny, who stepped forward and grasped Tilly's slight hand in a paw-like embrace, pulled her to her feet, and walked slowly with her to the edge of the pitch.

“Right,” said Miss Jones. “Rachel, pick a substitute.”

### **FIONA BRANSON**

ILLUSTRATION BY THE AUTHOR

© Fiona Branson 16/11/2016

Fiona Branson is a disabled actress with Ehlers Danlos on Equity's Disabled Artistes Register, she has appeared in *East Enders* and *Silent Witness* on BBC TV. In 2016 she was shortlisted for London Old Vic 12 as a playwright, for her script *Fox!*, which she has been developing at the National Theatre.



# My EDS Journey

---

12 years old  
I'm feeling cold  
The pain starts to spread  
A tear I shed

Now even in heat,  
Or sitting in a seat  
Discomfort I feel  
Can this be real?

Tell of the pain  
While filled with shame  
"That's what happens when you grow"  
Put on a smile for show

18 now,  
with asthma in tow  
Scoliosis explains  
The constant back pains

Dizziness  
Sleeplessness  
The constant dull ache  
None of which I've learnt to shake

22 and blooming  
You're fine people are assuming  
But behind closed doors  
Uncertainty is for sure.

The future unclear,  
Dislocations are a fear,  
Now heart valves have a leak  
Things are looking pretty bleak.

Be positive they say  
Heart monitors on my lap, they lay  
Vulnerability shows  
And independency slows.

Normal tasks become a chore  
Mind fog makes all seem a bore  
Receiving pitiful eyes are the norm  
But is this just the calm before the storm?

What will the future bring?  
Bad reputation will cling  
Stop or change the things you love  
And start wearing compression socks and gloves

Assessments here and there  
True emotions never shared  
Loneliness takes hold  
Pride and stubbornness become bold

But Ehlers my friend,  
Although we'll be together until the end,  
You will not win, no  
For I will fight to grow

The fear to overcome  
And to the pain, become numb  
Your respect to earn  
And to understand and learn

To develop and progress  
And to not from my dreams digress  
To live life to the full  
But with patience and always being careful.

**CHELSIE HEDGES**







# Mirrors

---

Thin brittle bone paper,  
Listing and wilting in the sun.  
Why must you fall down?  
Why must you scramble to shade.

Your edges are dusted and reeking,  
Crying for what you've become.  
What is underneath your material?  
What is underneath that blanket sheet.

Held together by a whistling stitch,  
Bursting at the brim with teeming bubbles.  
How are you still in existence?  
How are you still in existence.

Desperate and aching but with no lust,  
Aching like teeth and tusks and troubles.  
Where has your heart left you?  
Where have your limbs gone without you.

Distant memories like ink on the whiteness,  
Now raised and pink like fresh tattoo pain.  
When did this happen to your gleam?  
When did you become so pale.

Palliative treatment of creaking birch,  
Still silver peals away to reveal an off grain.  
When did this happen to me?  
When did this happen to me.

**SOPHIE NIALL**



# The Last Time I Climbed a Mountain

My God, I touched the sky today  
as land flowed out beneath me,  
clouds skimming that sweet break  
between earth and existence.

Sheer cliffs hover near a thousand feet  
of air and hope of strength.  
A wind blows through that could  
catch you up and carry you away.

Can it take me to a place of bodies  
that stay whole?  
Of hearts that let the blood flow right,  
of pieces of this form that move  
smooth-and-strong?

My God, I touched the sky today  
and I swear,  
I swear it,  
I swear I could feel these disjointed bones  
plant themselves into the grassy mountaintop,  
a stalwart beacon to the past,  
an echo of a body that  
tore through this life.

Oh, for the trouble of being alive,  
where nothing gold can stay  
and frosty nights kill off the harvest;

rot sneaks in through the dark (ness)  
blighting these wandering bodies.  
Is there more to it all than this?  
Will there be freedom beyond?

My God, I touched the sky today,  
and oh, it was the last time.

**GWEN MILLER**





# Coping With Chronic Illness: A Teenager's Two Cents (Part Two)

**I**T'S DAYS LIKE THESE WHEN I CLOSE MY eyes and hear the geneticist's words in my head: "You have a very clear case of Ehlers-Danlos syndrome – but you should go on to live a full life with no complications."

The first part was true; the latter was not. My official diagnosis came in October of 2014 after nearly two years of diligent research and frustration. The EDS diagnosis joined scoliosis and POTS diagnoses; my Chiari Malformation diagnosis came in March 2015.

Now, in the last days of 2016, I am searching for answers for the not-so-new symptoms that continue to plague me. Perhaps the struggle will never end. Every year it seems to be something new, but I suppose that that is the tune of living with EDS. My attempts to console myself always include, "There is no progress when there is no struggle," but that's not so helpful when I don't know for what reason I am working to make progress.

It's been over two years since I was diagnosed with EDS on that absurdly sunny October day. It's been nearly four since I heard "Ehlers-Danlos syndrome" for the first time. Even after being a part of the community for all this time, I feel like an outsider. I know that I have EDS, but still it shocks me at times. I hope the shock dissipates soon, because my conditions and disability do not halt for my quasi-denial.

It wasn't the EDS that put me on crutches; the Chiari Malformation is mostly responsible for that. I was prescribed these crutches at the age of 13 and have been using them for over a year. Some of my classmates still inquire whether or not my broken leg will heal. I joke that since I've been on crutches for over a year, it must be a particularly

nasty fracture; but there are only so many times one can use a joke, so I say I have a neurological condition and try to scamper away.

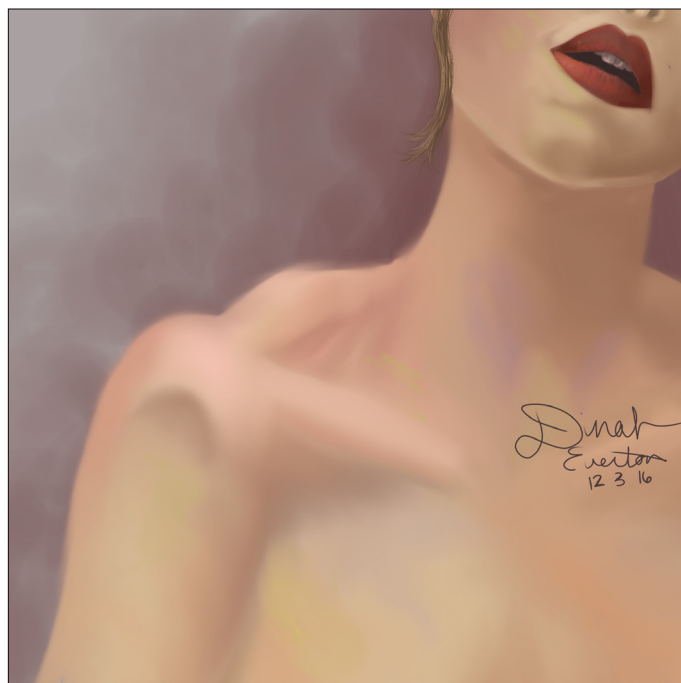
My teenaged classmates are not the only ones guilty of making assumptions about my disability. Hospital registrars tell me that they hope that my leg gets better soon. Some teachers think they've witnessed a miracle when they see me ambling around the classroom without crutches (even with the ample support of desks and other furniture). Family members have told me I'd be off crutches before I knew it. I am told I don't look sick, but no one can see that my collagen is defective.



I realize that, by being visibly disabled, I serve as a mirror for those that imagine how they would go on in life if they were to end up with a disability. The transition from being invisibly to visibly disabled was not easy for me. I thought at first I would only need a cane, but a physical therapist told me they don't offer much support. Right they were.

I vividly remember hobbling around a store and being intimidated the inquisitive stares. It seemed as though everyone else knew I was new at being “visibly disabled.” What still irks me about my crutches is how they are an inaccurate representation of my condition. My crutches are meant to help only one facet of my conditions; no one else can see the war that still rages inside my body. There is such a stigma shrouding invisible disability.

Being on crutches could certainly have been harder, but that isn’t to say that there haven’t been tribulations. Last year, one of my peers would steal my crutches and run down the hallway – or he’d kick my crutches out from under me. This year, another boy would loudly address me in the hallways as “cripple.” His idea of an apology when I finally had the principal intervene was to tell me that he didn’t see why what he was saying was such a big deal. Recently I was told that for a disabled girl I’m “really pretty.” Anyone told this knows how frustrating this is, because the person saying it thinks it’s the best compliment.



At a wedding, an older woman eyed me up and down, looking at my shoulder brace and crutches, and said, “I’m picturing something really painful, like a car accident.” I informed her politely that I have a neurological condition, true enough and easier than the puzzled looks when I say that I have a genetic disorder. Her reply? “Oh, well that’s good – well, not good – uh, God, you know what I mean.”

I’d be lying if I said that I still don’t get flustered at times. When I come across someone that is using some form of an assistive device, I hope that they also notice mine and that we possess some form of a mutual understanding. That way, no opportunities are presented to me to mortify either myself or the other person. I never know what to say, even though I have been playing this game for a year and a half; I’m still learning the unspoken rules. My experience is that often it’s best to say nothing at all.

I try to beat the odds and defy the stereotypes. Some people rush to open doors for me, but are astounded when I open the doors for them instead. Others are stupefied when they see that I am a consistently straight-A student. My work ethic has not suffered much as my conditions have declined; in fact, I use my worsening health as motivation to work harder. It is probably better that I burn out doing something worthwhile rather than wither away doing nothing. I follow this philosophy until I am nearly blind with exhaustion each and every day. I know that in order to ever be considered an equal as a disabled woman, I will have to work much harder. And work harder I will.

I have found some of the strongest people in the EDS community, the chronic illness community, and the disabled community. We are not the superheroes of action movies, but we are strong in our own way, and my story is only one of many. I firmly believe that one of the best ways to help each other – even if it isn’t the easiest – is to be more vocal about our struggles through art, writing, and educating others. I struggle with being open, but I know it can indeed make a difference. Our futures may sometimes appear to be bleak, but I believe that there is hope for us out there.

Hope is a fragile thing, though.

## **DINAH EVERTON**

ILLUSTRATIONS BY THE AUTHOR

(*Coping With Chronic Illness: A Teenager’s Two Cents [Part One]* is available in the Spring 2015 issue of [Loose Connections](#).)





# I WANT TO BREAK OUT (Trapped By EDS)

---

I want to break out  
I want to be free  
Free from these chains  
That are holding me

Crippling pain  
From my outer skin  
To all the way down deep,  
Deep within

It has me imprisoned  
Holding me tight  
Taking my hopes, my dreams  
My sleep at night

So many things  
I wanted to do  
To live my life  
The way I choose

But now I'm trapped  
Within these four walls  
Staring out a window  
That beckons me with calls

Stuck here I am  
As the days become years  
Unable to shed this curse  
That plagues me with pain and tears

**SEAN O'BRIEN**



# The Push Puppet

---

This toy I once played with,  
Danced and moved at my whim  
With only a thumb-push to make it wobble  
I laughed each time it stood

When I look back on my push puppet  
I see that its falls both amused and taught me  
With the disintegration and  
    reintegration of its form,  
It reassured me: we fall down, and get up again

But as I grew  
My body's joints started to dislocate  
I did not want them to—each time  
    praying it wouldn't happen again  
Why couldn't I just normally walk, run, and play?

My joints disobeyed my will at their whim,  
With me falling, or joints popping out in my sleep  
Now Nature—my own DNA—pushes the button  
And I am the push puppet

**DAVID DOUKAS, MD**

(FAM MED 2016;48(9):731.) REPRINTED WITH  
PERMISSION FROM THE SOCIETY OF TEACHERS OF  
FAMILY MEDICINE, [WWW.STFM.ORG](http://WWW.STFM.ORG).







# When Hugs Hurt

---

Nothing can alter or ever replace,  
My Dad's horror, as he tried to embrace,  
Me, like always, he just wanted a hug,  
My reaction however, made him feel like a thug.

I shrieked and recoiled, as his arms held me tight,  
His alarm was apparent, this wasn't a fight.  
There was no violence, no action, no tone,  
But the damage was done, to my joint, to my bone.

He'd only wanted to love me, to greet me as such,  
He wasn't a small man but neither too butch.  
You don't have to be strong, hench, nor great,  
To damage my bones, make me dislocate.

The lightest of touch, heck, even the air,  
Can cause me such pain that will never repair.  
My bones are so brittle, my joints are too frail,  
As my Dad looked in horror, I started to pale.

My shoulder had come out, was protruding quite far,  
Confusion, clouded the face of my poor, suffering Da',  
The pain on his face, competing with my own,  
How had this happened, 'Don't touch me' I moan.

Jumping back, like I'd accused him of attack,  
My Dad looked heart broke, as a rib popped  
in my back,  
'What just happened?' – He tried to entail,  
'What did I do, how did I fail?'

After putting myself together, I tried to explain,  
The loose joints, weak bones, the constant injury  
and pain.  
How my attention is needed, before contact is made,  
So I can prepare and position, more attention is paid.

'I canny hug my daughter' my poor Dad cries,  
His head hung low, with watery eyes.  
'Hug me still', I try to explain,  
But do it in a manner that limits my pain.

Shout my name, gain my attention,  
Don't ask for a hug, you don't need to mention,  
Just open your arms and let me come to you,  
And when we embrace, be gentle too.

**LORNA STEWART**



# I Am Real, So Do I Exist?

---

I am blood and bones and nerves  
Defined by the physical, bearing pain and limits most palpable

I am hurt, yet do not bleed  
Somehow that is hard for others to believe

My body can only do its best, but that is not where my limits rest:  
Their narrow minds refuse to grasp that which a familiar name not yet has

I am as tangible as my pain  
I feel our existence in blood and bones and nerves

But since they cannot comprehend or classify it,  
Then of course it cannot exist.

**MICHELLE HARDY**





# I Wake Up Before My Alarm

---

I wake up before my alarm,  
 Thinking to myself "what now? My arm?"  
 I look down to see 4 joints astray,  
 I try not to yell out in disarray.  
 This causes so much pain for me,  
 This is the pain that never leaves me be.  
 I stumble out of bed,  
 And to my dismay, I dislocate a rib head.  
 I must keep moving though,  
 All I can possibly do is push through the pain and go.  
 As I trip on a stair,  
 I realize few people care.  
 My reality has become that of a rumor,  
 But those people have a really bad sense of humor.  
 Just because you cannot see my disease  
 Does not mean I can make it go away with a simple "please".  
 I try to control this pain all day,  
 But all I want to do is go out and play.  
 I will never be like the other kids on the field,  
 I will never be fully healed.  
 My invisible disability has led to my eternal fragility,  
 My body has lost its credibility.  
 Although I may look the same,  
 Chronic pain is no game.

**KAITLYN BRENNAN**



# You Don't Look Sick

---

You don't look sick, they will say  
Unaware of the struggles I face each day  
Have you tried this and that, they will ask  
I nod as I re-adjust my invisible mask  
She'll just cancel her plans, they state  
Already determining and sealing my fate  
It can't be that bad, they will claim  
Like somehow i am the one to blame

Its not easy being different, I will cry  
When you use all your strength and try  
Have a little patience with me, I'll plead  
Just because you don't see me hurt and bleed  
Don't judge me so harshly, I'll request  
But know this, I'll always try my best

It's hard when you body just breaks  
With every small step and stumble it aches  
I'll listen and take it on the chin  
But today's failure will be tomorrow's win

**LINDA WILSON**





We gratefully acknowledge the invaluable support of our donors.



## Company Gifts

### Over \$100,000

Amerector Inc.

*EDS Center for Clinical Care & Research  
and EDS International Symposium*

### \$50,000–\$100,000

Bauerfeind USA Inc.

The Ehlers-Danlos Support UK

*EDS International Symposium*

### \$10,000–\$50,000

First Giving

The Saint Paul Foundation

### \$5,000–\$9,999

Friends for EDS Foundation

*EDS Center for Clinical Care & Research*

Neumann Family Trust

TRUIST

### \$2,500–\$4,999

Amazon Smile

The Benevity Community Impact Fund

Crushed Red

Louis C. Barber Estate

*Donation to Annual Conference*

### \$1,000–\$2,499

A.M.S.E.D Genetique

Community Foundation For Southeast  
Michigan

The David and Karen Walter Charitable  
Giving Fund

Give With Liberty Employee Donations

The Josh Stevens Foundation

Kohlberg Kravis Roberts and Co.

*EDS International Symposium*

North Scott Community School District

Northrop Grumman Corp. Charity Trust

Reading Blue Mountain & Northern R.R.

Robin and Bennett Greenspan Fund

Z/G Foundation

*Research Fund*

### \$500–\$999

Bravelets LLC

Cambridge University Hospitals

*EDS International Symposium*

Clara Lemarr Foundation

Club General Motors

Crushed Red-Clayton

Crushed Red-Kirkwood

Everydayhero

Fidelity Charitable Gift Fund

IBM Employee Services Center

Jay and Pam White Family Fund

*EDS Center for Clinical Care & Research,  
in memory of Adria Anderson Austin Cook*

Marshall Repairs LLC

*EDS Center for Clinical Care & Research*

National Mah Jongg League, Inc.

Silicon Valley Community Foundation

Schwab Charitable Fund

The Sosland Foundation

United Way of Greater Philadelphia and SNJ

Wells Fargo Community Support Campaign

YourCause

### \$250–\$499

The Davis Funeral Home, Inc.

*EDS Center for Research & Clinical care,  
in memory of Robin Huelle*

Jewish Community Federation

JustGive

Nuclear Electric Insurance

SOS & Co.

State Employees' Credit Union

### \$100–\$249

Aetna Foundation

Argent Credit Union

*Research Fund*

ASNA Beauty Salon, Inc.

*EDS Center for Clinical Care & Research*

Battelle Always Giving

Brenda Bunbury Realty, LLC

Bristol-Myers Squibb Foundation

Cardinal Health Inc. Matching Gift Program

CitiClosing Services, Inc.

*EDS Center for Clinical Care & Research*

The Community Foundation of Louisville

Depository Inc.

*EDS International Symposium,*

*in memory of Amy Bordogna Price*

Friedland Moravian Church New Life Class

*In memory of Jennifer Brinegar*

Lawrence Livermore National Laboratory

HOME Campaign

Moonan Family Fund of the Greater Lowell

Community Foundation

*Marketing Fund Donation*

Ozone Park First Class Laundry LLC

*EDS Center for Clinical Care & Research*

Polytech Education Association

Square Inc.

TERC

University of Pittsburgh Bradford Campus

### \$50–\$99

Bright Funds Foundation

DonateWell

Everyone Givvs, Inc.

Fidelity Charitable Gift Fund

Oargyris OT PC

*EDS Center for Research & Clinical Care*

St. Paul's Episcopal Church

Thomas R. Brown DDS INC.

*EDS Center for Research & Clinical Care*

Thrivent Choice

### \$25–\$49

The Huretta W Dobbs Trust DTD

*In memory of Adrian Douglas Brown*

R and S Cohen Family Rev Trust

The Peddler's Cottage

### \$1–\$25

America's Charities

Goodshop

Lake Cumberland Funeral Home

*In memory of Donald Mincey*

Mt. Airy Mattress



## Individual Gifts

### \$10,000+

Linda Neumann-Potash

### \$5,000–\$9,999

Raymond Augustin

*EDS Center for Research & Clinical Care*

Mandir Shaanti Bhavan

*EDS Center for Research & Clinical Care*

J. Donald Best

*CINK Medical Study Donation*

Peter O. Johnson

*CINK Medical Study Donation*

Lois Luallin

Tibor Neumann

John Scheller

*CINK Medical Study Donation*

### \$2,500–\$4,999

Marvin L. Bellin

*Research Fund*

Jerrice Ann Fritzlen

*Research Fund,*

*in memory of Morgan Amanda Fritzlen*

Richard M. Taffet

### \$1,000–\$2,499

Thomas R. Barmore

Marcus B. Bisram

*EDS Center for Research & Clinical Care*

Patrick Black

Jeffrey Michael Blilie

Richard Campbell

Rosemary Carroll

Eliot Chack

*EDS Center for Research & Clinical Care*

Gail K. Derick

*Conference Scholarships*

Margaret Egan

*In memory of Stephanie Egan*

Mark Elliott

*EDS Center for Research & Clinical Care*

Ellen Folts

*Circle of Hope*

Mark D. Garten

Vanessa Hampton

*CINK Medical Study*

Neeatranie Mohanlall

*EDS Center for Research & Clinical Care*

Teresa Murray

*EDS Center for Research & Clinical Care*

Carol L. Offield

*In memory of Heidi Moore*

Bonnie Osterwalk

Judith Thatcher

*EDS Symposium, EDS Center for Research  
& Clinical Care, Circle of Hope*

Laura Wilson

### \$500–\$999

Anne Breslin

Robin Broder

*EDS Center for Research & Clinical Care*

Jane Corley

*EDS Center for Research & Clinical Care*

David P. Cyr

Raymonde Gentile

Steven Greenbaum

*EDS Center for Research & Clinical Care*

Rhonda Kometer

*EDS Center for Research & Clinical Care*

Ravindra Mahabir

*EDS Center for Research & Clinical Care*

Lettice Rhodes

*EDS Center for Research & Clinical Care*

Renee Schibler

*EDS Center for Research & Clinical Care*

Catherine Styza

*EDS Center for Research & Clinical Care*

Chris & Traci Via

*EDS Center for Research & Clinical Care*

Barbara Warner

*Circle of Hope*

### \$250–\$499

Holly Allen

Angelique Alexander

Lori Altman

*EDS Center for Research & Clinical Care*

Jose Beria

*EDS Center for Research & Clinical Care*

Julie Berman

*EDS Center for Research & Clinical Care*

John R. Bielski

Patricia Cavanaugh

*EDS Center for Research & Clinical Care*

Raymonde Gentile

*EDS Center for Research & Clinical Care*

Wayne Heinmiller

Chris Huelle

*EDS Center for Research & Clinical Care,  
in memory of Robin Huelle*

Kelly Jonelis

*Shinji & Konrad's Birthday Fundraiser*

Theresa M. Keyes

Jennifer LaSpina

Gerard Lerman

*Circle of Hope*

Kathleen Lion

Mike MacDougall

Mary Pickering

*In memory of Christy Pickering*

Margie Priestle

Jennifer Rodgers

*Research Fund*

Ravina Natasha Vibart

*EDS Center for Research & Clinical Care*

Barbara J. Washburn

John T. Weber

### \$100–\$249

Kathleen Andruschat

Debbie Bachman

Kathie Baldadian

*EDS Center for Research & Clinical Care*

Wanda Barlow

*EDS Center for Research & Clinical Care*

Mary Kate Barnes

Kathryn Barton

Rebecca Bascom

*EDS Center for Research & Clinical Care*

Elayne Baumgart

*EDS Center for Research & Clinical Care*

Betty A. Bishop

Kathleen Blandford

Emmett Bonner

Debra Boogaard

John F. Braconnier

*In memory of Stephanie Braconnier*

Abby Brody

*EDS Center for Research & Clinical Care*

Brian Brooks

Gary Buchmann

Dulcie Bull

George Caesar

Fred D. Campbell

Nigel Casserley

Kathy Cohen

*EDS Center for Research & Clinical Care*

Peggy Coleman

*EDS Center for Research & Clinical Care*

Anita Colletti

*EDS Center for Research & Clinical Care*

Alan Coltri

Kathleen Conlin

*EDS Center for Research & Clinical Care*

Richard Connors

*EDS Center for Research & Clinical Care*

J. William Cotter

*In memory of Stephanie Braconnier*

Susan M. Cyr

Louise Cyr

*EDS Center for Research & Clinical Care*

Mark R. Czichotzki

Andrew Czudak

Lisa Dalton

Paula Dameika





Paul Dasilva

*EDS Center for Research & Clinical Care*

Lisa Dembo

*EDS Center for Research & Clinical Care*

Megan Devaney

Alison Doucette

Rose Drennen

*EDS Center for Research & Clinical Care*

Leigh Edgar

*EDS Center for Research & Clinical Care*

Matthew Egan

Roberta Evans

Lynn Faulkner

*Vascular Research Fund,*

*in memory of Frank A. Faulkner*

Roseann Fekete

*EDS Center for Research & Clinical Care*

Janice Feurtado

*EDS Center for Research & Clinical Care*

Lisa Florez

*EDS Center for Research & Clinical Care*

Scott Frey

*EDS Center for Research & Clinical Care*

Sheryl Geltner

*EDS Center for Research & Clinical Care*

Alicia R. Gilbert

Donna Gleason

Rosalie Greenspan

*EDS International Symposium*

Robin Greland

*EDS Center for Research & Clinical Care*

Edward Grose

*EDS Center for Research & Clinical Care*

Ronald Harlan

Cynthia Harrison

*EDS International Symposium*

William K. Hausbeck

*In memory of Geraldine Brown*

Vern Hines

Brent Hofmann

*EDS Center for Research & Clinical Care*

Brent Hofmann

Geoffrey Howson

Sarah Hung

*EDS Center for Research & Clinical Care*

Blake Hurtik

*EDS Center for Research & Clinical Care*

Michael Jacobs

*EDS Center for Research & Clinical Care*

Suzanne Judge

Leslie Kays

*EDS Center for Research & Clinical Care*

Ameer Kayum

*EDS Center for Research & Clinical Care*

Rachel Kerbrat

*EDS Center for Research & Clinical Care*

Jeanette Kuiphof

Rolland L. Lavallee

James W. Leonard

*In memory of Aren Wish*

Chris Lighthall

Enid Lythgoe

*EDS Center for Research & Clinical Care*

Carolyn MacLeod

*EDS Center for Research & Clinical Care*

Tina Miller

Jacquelyn Minnick

*EDS Center for Research & Clinical Care*

Neetranie Mohanlall

*EDS Center for Research & Clinical Care*

Leah Monger

*In memory of Douglas Adrian Brown*

Richard Morgan

Marcy Radler Moyer

*EDS Center for Research & Clinical Care*

Allison Muth

*EDS Center for Research & Clinical Care*

G. Nassour

*EDS International Symposium*

Julie M. Newton

*Research Fund*

Steven Nootbaar

*EDS Center for Research & Clinical Care*

Kim M. Ogle

*Vascular Research Fund,*

*in memory of Frank A. Faulkner*

Donna Olivero PT

Steven Ostroski

*EDS Center for Research & Clinical Care*

Debra K. Partridge

*In memory of James Dunn and Paul Shupe*

Nadira P. Persaud

*EDS Center for Research & Clinical Care*

David Polzien

*In memory of Jennifer Brinegar*

Heather Purdin

Anne Rafalo

Seenauth Rai

*EDS Center for Research & Clinical Care*

Anjanie Rajpaul Ramsaywack

*EDS Center for Research & Clinical Care*

Nicholas Ridgely

Linda Ritchie

*EDS Center for Research & Clinical Care*

Brian Rooth

*In memory of Brandon Rooth*

Eugenia Rooth

*In memory of Brandon John Rooth*

John Roper

Irving B. Ross

*In memory of Glenn D. Ross*

Joseph R. Rossi

*In memory of James Paul Shupe*

Kathryn Roth

Michael Rubin

*EDS Center for Research & Clinical Care*

Taly Rutenberg

Damon Rutland

*EDS Center for Research & Clinical Care*

Pooran Samlall

*EDS Center for Research & Clinical Care*

Debra Scully-Wagner

Lisa Seecharan

*EDS Center for Research & Clinical Care*

Nancy Seeley

Paul Sell

*EDS Center for Research & Clinical Care*

Loaknauth Sobhai

*EDS Center for Research & Clinical Care*

Rosemary Soldiviero

*EDS Center for Research & Clinical Care*

Bruce G. Spengler

*In memory of Pastor Adrian Brown*

Dennis C. Spitz

*In memory of Stephanie Marie Braconnier*

Jason Stasiulewicz

*EDS Center for Research & Clinical Care*

Roger Stevenson

David Stone

*EDS Center for Research & Clinical Care*

Richard A. Stone

*In memory of Mr. William C. Nyberg*

Peter J. Summers

*Research Fund*

Richard Taffet

*EDS Center for Research & Clinical Care*

Janet Tangney

Lynn A. Thompson

*EDS Center for Research & Clinical Care,*

*in memory of Robin Huelle*

Kerry Upperton

Laura K G Utterback

*EDS Center for Research & Clinical Care*

Lynn VerHelst

*EDS Center for Research & Clinical Care*

Craig D. Vernon

*Vascular Research Fund, in memory of*

*Frank A. Faulkner*

Sue Volkman

Waller Walker

Jimmie Parks Watkins

*EDS Center for Research & Clinical Care*

Philip Weinberg

Allyson Weir

*EDS Center for Research & Clinical Care*

Rosetta Wester

Catherine White

*EDS Center for Research & Clinical Care*

Pearl White

*EDS Center for Research & Clinical Care*



Elise Whitley  
*EDS Center for Research & Clinical Care*  
 Gloria Willis  
*In memory of Elmer Fischer*  
 Lynne Wohlner  
 Nanette Yeager

## \$50–\$99

Deborah Adams  
 G. N. Allen  
 Noah Baerman  
 Geraldine Brown  
*In memory of Geraldine Brown*  
 Maggie Buckley  
 Dulcie Bull  
 Jack Warren Burnam  
 Patricia Camayd  
 Laurel Carr  
 John Chaffey  
 Barbara Chambers  
 Sharon Cohen  
 Kimberly Day  
 Maribeth Donaldson  
*EDS Center for Research & Clinical Care*  
 Terry Durkin  
 Stephen Edgcumbe  
 Barbara Egelko  
 Susan Egelko  
 Louis B. Eichhold  
*In memory of Daryl Miller*  
 Dorothy F. Elicker  
 Maureen Enmark  
 Barbara Faze  
 Brandon Fields  
*Research Fund*  
 Nancy Florida  
*EDS Center for Research & Clinical Care*  
 Lourdes Font  
*EDS Center for Research & Clinical Care*  
 Christian A. Gertsch  
*In memory of Ramona Bukowski*  
 Neil Gibson  
 Jay Goldberg  
 Camille Gray  
*EDS Center for Research & Clinical Care*  
 Megan Greenholt  
 Nancy Grieve  
*EDS Center for Research & Clinical Care*  
 John Hackett  
*Vascular Research Fund*  
 Janet Hamilton  
 Libby Hanaway  
*EDS Center for Research & Clinical Care*  
 Kristine Hastings  
*EDS Center for Research & Clinical Care*  
 Jane Herbert  
 Nicole Herlands

Stuart Herlands  
 Stephen Hershey  
 Amy Hopkins  
 Lorene Hughes  
 Robert Jacobs  
 Jessica Jameson  
 Wanda L. Jared  
 Paula J. Johnson  
 Palma Joslin  
 Mary Joyal  
*EDS Center for Research & Clinical Care*  
 Elise G. Kaplan  
*EDS International Symposium, in memory of Ed Lovenwirth;*  
*EDS International Symposium, in memory of Mrs. Eileen Levine;*  
*In memory of Steven Blase*  
 Marvin D. Kautz  
*In memory of Joanne Elizabeth Townsend*  
 Jane Kohuth  
*EDS Center for Research & Clinical Care*  
 Carly Kostakos  
*EDS Center for Research & Clinical Care*  
 Hazel Lambert  
 Stan Landowski  
*EDS Center for Research & Clinical Care, in memory of Robin Huelle*  
 Charlotte Lee  
 Gundel Lee  
*EDS Center for Research & Clinical Care*  
 Steven Letnes  
 John Lorah  
*In memory of Jeen Humphrey*  
 Aileen Lott  
 Ann Macedo  
*EDS Center for Research & Clinical Care*  
 Mary Macedo  
 Judith Madden  
*EDS Center for Research & Clinical Care*  
 Joni Mann  
*In memory of Scott Evan Siegel*  
 Cordae Mattson  
 Adrienne Mayer  
 Mary Lou McCormick-Howar  
 Renee McWhorter  
 Debby Mctaggart  
 Michael Meade  
*EDS Center for Research & Clinical Care, in memory of Robin Huell*  
 Lynne Merritt  
*EDS Center for Research & Clinical Care*  
 Terry L. Meyers  
*In memory of Lee Byrd*  
 Elizabeth Mills  
*EDS Center for Research & Clinical Care*  
 Susan Morris  
*EDS Center for Research & Clinical Care*

Robin Muth  
*EDS Center for Research & Clinical Care*  
 David Null  
 Carol Porosoff  
 Mary Louise Raymond  
*In memory of Mrs. Stephanie Marie Braconnier*  
 Shane Robinson  
 Heidi Schmedes-Shook  
*EDS Center for Research & Clinical Care*  
 Joanna P. Scholz  
*EDS Center for Research & Clinical Care, in memory of Stephanie Braconnier*  
 Constance Schoppe  
 Janet Schramm  
 Robert Schur  
*EDS Center for Research & Clinical Care*  
 Scott Scrivner  
 William C. Segal  
 Joan M. Serviss  
*In memory of Robin Huelle*  
 Holly S. Shupe  
*In memory of James Paul Shupe*  
 Larry J. Shuttlesworth  
*EDS Center for Research & Clinical Care, in memory of Robin Huelle*  
 Chester Simcoe  
*In memory of Kenneth Burch*  
 Amy Smith  
 Lauren Smith  
*EDS Center for Research & Clinical Care*  
 Corinn Sparks  
 Kellyanne Stewart  
*EDS Center for Research & Clinical Care*  
 Sarah Strauss  
*EDS Center for Research & Clinical Care*  
 Rosemary Sullivan  
*EDS Symposium*  
 Caroline Teal  
 Neil Thomson  
 Virginia Tiedemann  
 Kiara Walker  
*EDS International Symposium*  
 Christopher Ward  
*EDS Center for Research & Clinical Care*  
 Richard J. Weed  
 Kimberly Weiner  
*EDS Center for Research & Clinical Care*  
 Jane Wharton  
 Jeffrey Wilczewski  
*EDS Center for Research & Clinical Care*  
 Elizabeth Wilkinson  
 David L. Witt  
*In memory of Geraldine Brown*  
 Catherine T. Womelsdorf  
*In memory of Stephanie Marie Braconnier*  
 Jan Wragg



Stefanie Yurus  
 Sandra Zickrick  
*EDS Center for Research & Clinical Care*  
 Jeff J. Zweiback  
*EDS Center for Research & Clinical Care*

## \$25-\$49

Anonymous  
 Russ Ackerman  
*EDS Center for Research & Clinical Care, in memory of Robin Huelle*  
 Diana Archer  
*EDS Center for Research & Clinical Care*  
 Beth Baird  
*EDS Center for Research & Clinical Care*  
 Sylvia K. Baird  
*In memory of Geraldine Brown*  
 Michelle Barber  
*EDS Center for Research & Clinical Care*  
 Nancy Berna  
 Mary Boutin  
 James L. Braciak  
 Anne Bradley  
*EDS Center for Research & Clinical Care*  
 Sandra J. Brogan  
*In memory of Pastor Adrian Brown*  
 Janice Brown  
 Jennifer Brown  
 Daniel G. Bumgarner  
 Olivia Christmann  
*EDS Center for Research & Clinical Care*  
 Shirley J. Clement  
*Research Fund*  
 Lindsay Cropp  
 Thomas J. Dearing  
 Jacqueline Ann DiMauro  
 Nancy Doyle  
*In memory of MaLisa Porter*  
 Paula Eury  
*EDS Center for Research & Clinical Care*  
 Timothy Fox  
 Georgianne Ginder  
 Theresa Grabiner  
 Clifford Grand  
*Vascular Research Fund, in memory of Frank A. Faulkner*  
 Diane Hackett  
*Vascular Research Fund, in memory of Meghan*  
 S. Hamelund  
*In memory of Adrian Brown*  
 Mike & Andrea Hannigan  
 Carol B. Heap  
*In memory of Gloria Grabert*  
 Kristen Heinlein  
 Carolyn M.R. Hill  
 Charlotte Himmelfarb

Arthur M. Hohendorf  
*In memory of Gerri Brown*  
 Gary Hojdila  
*In memory of Deborah Hojdila*  
 Sallie D. Hollar  
 Shauna Iler  
*EDS Center for Research & Clinical Care*  
 Lynette Jansen  
*EDS Center for Research & Clinical Care*  
 Eleanor Kardish-Kamil  
*EDS Center for Research & Clinical Care*  
 Theresa M. Keyes  
 Jane Ann Klowak  
*EDS Center for Research & Clinical Care*  
 Christine Kuckuk  
 Jaime Lamberto  
 Kathleen Lawless  
 Matthew Lees  
*EDS Center for Research & Clinical Care*  
 Maureen LeFevre  
 Linda Leinan  
 Kay Lustberg-Goldbeck  
*EDS Center for Research & Clinical Care*  
 Darlene MacAuley  
 Ann Macedo  
*EDS Center for Research & Clinical Care*  
 Anne Madden  
 Ernest Mathews  
 Shannon McKenzie  
 Dona F. McNeil  
 Eric Meyer  
 John C. Milan  
*In memory of Adrian Brown*  
 Tina Miller  
*Research Fund*  
 Deanne Morse  
 Judith H. Mullen  
*EDS Center for Research & Clinical Care, in memory of Robert J. Caspar*  
 Natalie Nixon  
 Anne O'Brien  
*In memory of Donald Mincey*  
 Valerio Antonio Paciello  
 Stephanie Palumbo  
*EDS Center for Research & Clinical Care*  
 Lucy Pausz  
*EDS International Symposium*  
 Michael Pessler  
*EDS Center for Research & Clinical Care*  
 Marianne Plucienik  
 Alice Praast  
 Renee Quiggle  
 Dorothy Racunas  
 Ada R. Rampersaud  
*EDS Center for Research & Clinical Care*  
 Joe Richardson  
*In memory of Adria Austin Cook*

Nancy Richardson  
*EDS Center for Research & Clinical Care*  
 George H. Schaefer  
*EDS Center for Research & Clinical Care, in memory of Robin Huelle*  
 Kate Schultz  
*EDS Center for Research & Clinical Care*  
 William C. Segal  
 Tori Smith  
 Deborah Ann Sprecher  
 Sarah Strauss  
 Susan T. Teague  
*In memory of Jennifer Brinegar*  
 Christine Tosun  
*EDS Center for Research & Clinical Care*  
 Maria Valles  
*EDS Center for Research & Clinical Care*  
 Rina F. Vassallo  
*In memory of Stephanie Braconnier*  
 Susan Velky  
 Michael Vladimer  
 Donald L. Walsh  
*In memory of Kenneth W. Burch*  
 Gayleen Williams  
 Christopher Wilson  
*EDS Center for Research & Clinical Care*  
 Joe A. Wilson  
*In memory of Hallie Brody*  
 Jane A. Wood  
*In memory of Geraldine Brown*  
 Tina Woolard  
 Haven World  
*EDS Center for Research & Clinical Care*  
 Sarah Yeager  
*EDS Center for Research & Clinical Care*

## Under \$25

Linda Amtsfield  
 Pamela C. Baugh  
 Brent S. Berger  
 Allen and Julie Berman  
 Raymond E. Bivens  
 Denise R. Bixler  
 Jessica Bowie  
*EDS Center for Research & Clinical Care*  
 Karen Boyle  
*EDS Center for Research & Clinical Care*  
 Chelsea E. Bulmer  
 Sandi Caldron  
 Debra A. Caldwell  
 Rosalie A. Clemetson  
*EDS Center for Research & Clinical Care, in memory of Robin Huelle*  
 Kimberlie Clyma  
 Jacqueline A. Coates  
*EDS Center for Research & Clinical Care, in memory of Robin Huelle*





Sylvia Condro

*EDS Center for Research & Clinical Care*

Valerie Coulton

*EDS Center for Research & Clinical Care*

Claire DeGiralamo

*EDS Center for Research & Clinical Care*

James Donaghy

Mary A. Earley

Paul Ehrlich

*In memory of Stephanie Egan Braconnier*

Vincent Falsitta

Sky J. Ferrell

Claire Foote

*EDS Center for Research & Clinical Care*

Michael Goldberg

Carol Goldsmith

Mary Elizabeth Graham

Laurence J. Griffin

*In memory of Shane Contaloni*

Monica Howard

*In memory of Geraldine Ann Brown*

Karen C. Huff

Margretta E. Hurd

Janice Jennings

Victoria Kendall

*EDS Center for Research & Clinical Care*

Elizabeth Knox

Elizabeth Anne Kowald

James W. Kreiser

Lisa Layton-Krout

Laura K. Lloyd

Annette Login

Donna Maltese

*EDS Center for Research & Clinical Care*

Jo-Anne McCutcheon

*EDS Center for Research & Clinical Care*

Julian Mendoza

*EDS Center for Research & Clinical Care*

Chris Meyer

Richard Milone

Mary L. Morett

Cathy Pardiny

Edward Payne

Vijay Prakash

Robert Reiner

Lisa Romie

Bill Rumback

*In memory of MaLisa Porter*

Benjamin Schlechter

Benjamin Schmerler

*EDS Center for Research & Clinical Care*

Richard Schmidt

*In memory of Clarence Earnhart*

Charles R. Sims

Denise Smith

Patricia A. Smith

*In memory of Geraldine Ann Brown*

L. Joshua Sosland

Shirley Spidell

*In memory of Geraldine Ann Brown*

Ronald K. Sterk

Lori Stuermer

Charlene K. Swanson

*Vascular Research Fund, in memory of*

*Frank A. Faulkner*

Nora E. Wages

Susan Wasserman

Rebecca L. White

Teresa J. White

*In memory of Jennifer Brinegar*

Donald E. Wolf

Jacob Wright

*EDS Center for Research & Clinical Care*

Listings as of December 8, 2016. When our office moved this year, our donor database did as well; every effort has been made to integrate both systems, and we apologize for any errors.



# Ehlers-Danlos Syndrome—Hypermobility Type: A Much Neglected Multisystemic Disorder

Rambam Maimonides Medical Journal 2016 October; 7(4): e0034. ISSN: 2076-9172

Published online 2016 October 31. doi: 10.5041/RMMJ.10261.

Special Issue on Rheumatology

Guest Editor: Alexandra Balbir-Gurman, M.D.

Yael Gazit, M.D., M.Sc.,<sup>1\*</sup> Giris Jacob, M.D., Ph.D.,<sup>1,2</sup> and Rodney Grahame, C.B.E., M.D., F.R.C.P., F.A.C.P.<sup>3</sup>

<sup>1</sup>Internal Medicine F and the Institute of Rheumatology, Tel Aviv Sourasky Medical Center, and Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel

<sup>2</sup>J. Recanati Autonomic Dysfunction Center, Tel Aviv Sourasky Medical Center, Tel Aviv, Israel

<sup>3</sup>Hypermobility Unit, London and Centre for Rheumatology, Division of Medicine, University College London, London, UK

\*To whom correspondence should be addressed. E-mail: yaelhms@gmail.com

Copyright © 2016 Gazit et al. This is an open-access article. All its content, except where otherwise noted, is distributed under the terms of the Creative Commons Attribution License (<http://creativecommons.org/licenses/by/3.0>), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

## ABSTRACT

Ehlers–Danlos syndrome (EDS)–hypermobility type (HT) is considered to be the most common subtype of EDS and the least severe one; EDS-HT is considered to be identical to the joint hypermobility syndrome and manifests with musculoskeletal complaints, joint instability, and soft tissue overuse injury. Musculoskeletal complaints manifest with joint pain of non-inflammatory origin and/or spinal pain. Joint instability leads to dislocation or subluxation and involves peripheral joints as well as central joints, including the temporomandibular joints, sacroiliac joints, and hip joints. Soft tissue overuse injury may lead to tendonitis and bursitis without joint inflammation in most cases. Ehlers–Danlos syndrome-HT carries a high potential for disability due to recurrent dislocations and subluxations and chronic pain. Throughout the years, extra-articular manifestations have been described, including cardiovascular, autonomic nervous system, gastrointestinal, hematologic, ocular, gynecologic, neurologic, and psychiatric manifestations, emphasizing the multisystemic nature of EDS-HT. Unfortunately, EDS-HT is

under-recognized and inadequately managed, leading to neglect of these patients, which may lead to severe disability that almost certainly could have been avoided. In this review article we will describe the known manifestations of the extra-articular systems.

**Keywords:** Disability, Ehlers-Danlos syndrome, hypermobility syndrome, joint hypermobility, multisystemic, neglect

## INTRODUCTION

The Ehlers–Danlos syndromes (EDSs) constitute a group of inherited disorders of connective tissue characterized by soft hyperextensible skin and joint hypermobility, distinguished by additional connective tissue manifestations.<sup>1</sup> The Ehlers–Danlos syndrome was first described by Ehlers in Denmark in 1898 and Danlos in Paris in 1908. They published individual case studies with common features of ligamentous laxity and skin hyperextensibility.<sup>2</sup> Ehlers–Danlos syndrome–hypermobility type (EDS-HT) is considered to be the most common subtype of EDS<sup>3,4</sup> and the least severe





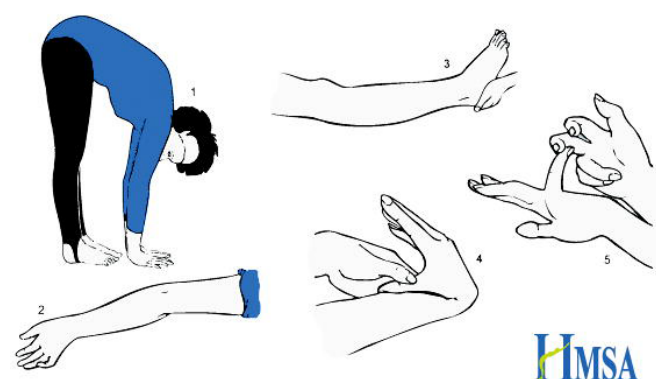
one.<sup>3</sup> It is characterized by joint laxity, soft, stretchy, and often semi-transparent skin, and musculo-skeletal complications, without severe complications of arterial dissection or bowel rupture seen in EDS-vascular type,<sup>1,5</sup> and without hemosiderotic scars and molluscoid pseudotumors seen in the EDS-classical type.<sup>1,6</sup> Ehlers–Danlos syndrome-HT, now considered to be indistinguishable if not identical to the joint hypermobility syndrome (JHS), manifests with musculoskeletal complaints, joint instability, and soft tissue overuse injury.<sup>3,7–12</sup> Musculoskeletal complaints manifest with joint pain of non-inflammatory origin and/or spinal pain. Joint instability leads to dislocation or subluxation and involves peripheral joints as well as central joints, including the temporomandibular joints (TMJ), sacroiliac joints, and hip joints.<sup>7–9</sup> Soft tissue overuse injury may lead to tendonitis and bursitis<sup>10,11,12</sup> without joint inflammation in most cases.<sup>3,11</sup> Although an inflammatory component is rare, EDS-HT carries a high potential for disability<sup>13</sup> due to recurrent dislocations and subluxations and chronic pain.<sup>8,11,12,14,15</sup> Throughout the years, extra-articular manifestations have been described, including cardiovascular and autonomic nervous system,<sup>16–22</sup> gastrointestinal,<sup>19,23</sup> hematologic,<sup>24–26</sup> ocular,<sup>27</sup> gynecologic,<sup>19,28–31</sup> neurologic,<sup>19,25,32,33</sup> and psychiatric manifestations,<sup>7,8,11,19,34,35</sup> emphasizing the multisystemic nature of EDS-HT. Unfortunately, EDS-HT is under-recognized and inadequately managed,<sup>36–38</sup> leading to neglect of these patients which may lead to severe disability that almost certainly could have been avoided.<sup>39</sup>

## GENERAL CHARACTERISTICS AND MANIFESTATIONS

Joint hypermobility (JH), defined as an excessive range of joint movement taking into consideration age, gender, and ethnic background, is inherited<sup>40,41</sup> and may pose no problem. Acquired hypermobility may also result from changes in connective tissue in other diseases such as systemic lupus erythematosus.<sup>42</sup> Joint hypermobility is recognized by the nine-point Beighton score<sup>43</sup> (Figure 1) and includes passive dorsiflexion of each fifth finger greater than 90°, passive apposition of each thumb to the flexor surface of the forearm, hyperextension of each elbow greater than 10°,

hyperextension of each knee greater than 10°, and ability to place the palms flat on the floor with the knees fully extended.

Figure 1: Calculation of the Beighton Score



The Beighton score is calculated as follows:

- One point if while standing forward bending you can place palms on the ground with legs straight
- One point for each elbow that bends backwards
- One point for each knee that bends backwards
- One point for each thumb that touches the forearm when bent backwards
- One point for each little finger that bends backwards beyond 90 degrees

Taken with permission from the Hypermobility Syndromes Association (HMSA) site (<http://hypermobility.org/help-advice/hypermobility-syndromes/beighton-score/>).

Ehlers–Danlos syndrome-HT, now considered to be indistinguishable if not identical to the joint hypermobility syndrome (JHS),<sup>44</sup> is a clinical condition of JH with symptoms of joint instability, arthralgia, myalgia, soft tissue injuries, and arthritis.<sup>4,5,46</sup> Diagnosis relies on the Brighton criteria (Table 1).<sup>47,48</sup> The predominant presenting complaint is pain, which is often widespread and longstanding, with patients reporting pain ranging from 15 days to 45 years.<sup>39,49</sup> Chronic pain may start in adolescence (with 75% of hypermobile adolescents reporting symptoms by the age of 15) or even as late as the fifth or sixth decade of life.<sup>3,39,45</sup> Severity sometimes correlates with the degree of joint instability.<sup>3,15</sup> Fatigue and sleep disturbance, most probably secondary to severe chronic pain, subluxations, and dislocations while



Table 1: Revised Diagnostic Criteria for Ehlers-Danlos Hypermobility Type, a.k.a. Joint Hypermobility Syndrome (JHS).

*Major Criteria:*

- A Beighton score of 4/9 or greater (either currently or historically);
- Arthralgia for longer than three months in four or more joints.

*Minor Criteria:*

A Beighton score of 1, 2, or 3/9 (0, 1, 2, or 3 if aged 50+);

- Arthralgia (>3 months) in one to three joints or back pain (>3 months), spondylosis, spondylolysis/spondylolisthesis;
- Dislocation/subluxation in more than one joint, or in one joint on more than one occasion;
- Soft tissue rheumatism, >3 lesions (e.g. epicondylitis, tenosynovitis, bursitis);
- Marfanoid habitus (tall, slim, span:height ratio >1.03, upper:lower segment ratio less than 0.89, arachnodactyly (positive Steinberg/wrist signs);
- Abnormal skin: striae, hyperextensibility, thin skin, papyraceous scarring;
- Eye signs: drooping eyelids or myopia or antimongoloid slant;
- Varicose veins or hernia or uterine/rectal prolapse.

JHS is diagnosed in the presence two major criteria, or one major and two minor criteria, or four minor criteria. Two minor criteria will suffice where there is an unequivocally affected firstdegree relative.

Taken with permission from the Hypermobility Syndromes Association (HMSA) site (<http://hypermobility.org/helpadvice/hypermobilitysyndromes/thebrightonscore/>).<sup>50</sup>

Table 2: Multisystemic Nature of EDS-HT.

System	Manifestations
Cardiovascular	Aortic regurgitation, aortic root dilatation, mitral valve prolapse, mitral regurgitation, tricuspid regurgitation, Reynaud phenomenon
Autonomic Nervous System	Palpitations, dizziness, pre-syncope, syncope
Gastrointestinal	Gastroesophageal reflux, dyspepsia, gastritis, delayed gastric emptying, irritable bowel syndrome
Hematologic	Easy bruising, bleeding tendency, prolonged bleeding time, oral mucosal bruises, menometrorrhagia
Ocular	Myopia, strabismus
Gynecologic	Dysmenorrhea, menorrhagia, dyspareunia, uterine prolapse
Urologic	Constipation, fecal soiling, urinary tract infections, urinary incontinence, bladder prolapse, rectal prolapse
Obstetric	Short labor and delivery, premature rupture of membranes, pelvic pain, varicose veins, worsening of dysautonomia during pregnancy, postpartum hemorrhage, complicated perineal wounds
Neurologic	Headache, local anesthesia failure, postural instability, increased frequency of falls, impaired proprioceptive acuity, Chiari 1 type 1
Psychiatric	Kinesiophobia, anxiety, depression



changing posture during sleep, are frequently associated.<sup>3,11,12,15</sup> Affected individuals are often misdiagnosed with chronic fatigue syndrome, fibromyalgia, depression, hypochondriasis, and/or malingering prior to recognition of joint laxity and establishment of the correct underlying diagnosis.<sup>3</sup> Over the last three decades it has become apparent that EDS-HT has a widespread distribution and is not manifested solely in the joints (Table 2).

## Cardiovascular and Autonomic Nervous System Manifestations

A mild degree of aortic root dilatation has been found in up to one-third of EDS-HT patients,<sup>20,21,22</sup> necessitating echocardiographic evaluation and surveillance. Raynaud phenomenon was found in 38% of EDS-HT patients.<sup>19</sup> Patients with EDS-HT may suffer from palpitations, chest pain, dizziness, pre-syncope, and syncope,<sup>17</sup> which has been attributed in the past to mitral valve prolapse (MVP). Mitral valve prolapse was originally included in the earlier version of the Brighton criteria in 1986.<sup>47</sup> With more modern evaluation techniques clinically significant MVP has not been found to be more prevalent among EDS-HT patients.<sup>21,22,50,51</sup> For this reason MVP was removed from the Brighton criteria in 1998.<sup>48</sup> The frequency of MVP among EDS-HT patients was found to be 28%–67% in more recent studies,<sup>52,53</sup> but its clinical significance is not clear. Symptoms formerly attributed to MVP are now considered to be related to autonomic dysfunction, which was found to be highly prevalent among EDS-HT patients.<sup>16–18</sup>

## Gastrointestinal Manifestations

Gastroesophageal reflux was found in 57% of EDS-HT patients.<sup>19,23</sup> Chronic gastrointestinal discomfort was reported in 86% of patients with EDS-HT, attributed to dyspepsia, gastritis, or gastroesophageal reflux. Irritable bowel syndrome was found among 62% of patients. Early satiety and delayed gastric emptying are reported and exacerbated by opioids.<sup>3</sup>

## Hematologic Manifestations

Easy bruising and bleeding tendency is common in all EDS types, including EDS-HT.<sup>25</sup> It manifests with prolonged bleeding time,<sup>24,26</sup> oral mucosa fragility with mucosal bruises,<sup>9</sup> and menometrorrhagia.<sup>54</sup> Since coagulation tests are normal,<sup>24–26</sup> the underlying cause is presumed to be mechanically impaired collagen too weak to afford adequate protection to the capillaries. It is important to note that small and large arterial dissections have not been reported in EDS-HT.

## Ocular Manifestations

Myopia has been found in up to 50% of EDS-HT patients,<sup>54</sup> and high myopia of more than –6.0 diopters was found in 16% of patients compared with 0% in the control group.<sup>3,27</sup> Strabismus was found in 7% of EDS-HT pediatric patients<sup>55</sup> (as opposed to only 2%–4% of the general pediatric population), and it is often refractory to surgical correction.<sup>56</sup> Meyer *et al.* found size variations and shape abnormalities of collagen fibrils in the extra-ocular muscles that control the movement of the eye.<sup>57</sup>

## Gynecologic Manifestations

Dysmenorrhea and menorrhagia are common<sup>19,28,29,54,56</sup> and thought to be due to muscle contractions occurring with greater force given the loose connective tissue. Dyspareunia was found among 30%–57% of EDS-HT women,<sup>28,29,58</sup> thought to be caused by small tears in the vaginal surface and lack of appropriate vaginal secretions.<sup>56</sup> Pelvic organ prolapse is common,<sup>19,28,29,56,59–62</sup> including uterine prolapse which was found in almost 40% of women with EDS-HT.<sup>49</sup>

## Urologic Manifestations

In children with hypermobility constipation and fecal soiling were found to be more common in boys, and urinary tract infection and urinary incontinence more common among girls.<sup>63</sup> In another pediatric series 13% of girls and 6% of boys suffered from urinary tract infections.<sup>64</sup> Stress urinary incontinence was found in 40%–



70% of women with EDS-HT,<sup>28,58,65</sup> often earlier in life, thought to be due to a weakened pelvic floor, which may be worsened to bladder prolapse.<sup>56</sup> Fecal incontinence was found in up to almost 15% of EDS-HT patients, as compared to only 2.2% of the general population.<sup>65</sup> Rectal prolapse may also be found among EDS-HT patients.<sup>66</sup> Furthermore, Dordoni *et al.* reported on two EDS-HT family members who suffered from visceroptosis, including bilateral kidney prolapse, gastric ptosis, liver prolapse, and ovarian and heart prolapse.<sup>67</sup>

## Obstetric Manifestations

While labor and delivery might be rapid (shorter than 4 hours),<sup>19,29</sup> and premature rupture of membranes is common,<sup>54,68,69</sup> pregnancy in women with EDS-HT is generally normal with good maternal and neonatal outcome.<sup>30,70</sup> However, joint laxity and pain may increase during pregnancy.<sup>3,29,30,54,70</sup> Pelvic pain and instability necessitate the use of pelvic belt, crutches, and/or bed rest in 26% of women with EDS, the majority being EDS-HT (compared to only 7% among non-affected women).<sup>56,70</sup> Varicose veins in the legs and the vulva are more common among pregnant women with EDS-HT.<sup>56</sup>

Dysautonomia, characterized by lightheadedness, dizziness, fainting, etc., may worsen during pregnancy,<sup>56</sup> and when postural orthostatic tachycardia syndrome (POTS) is present a blood pressure fall was reported.<sup>71</sup> Women with EDS-HT are more prone to postpartum hemorrhage (19% versus 7%) and complicated perineal wounds (8% versus none).<sup>70</sup> Premature delivery was found to be more related to EDS-HT of the infant (40%), and was less prevalent if the mother had EDS-HT (21%).<sup>70</sup>

## Neurologic Manifestations

A total of 40% of children with EDS-HT<sup>72</sup> and 50% of adults<sup>14</sup> suffer from headaches, characterized as chronic recurrent headaches in the absence of structural, congenital, or acquired central nervous system lesions that correlate with their symptoms.<sup>73</sup> Many complain of headaches related to the neck or facial pain that might be related to jaw or TMJ problems.<sup>56</sup> Headaches may also be part of dysautonomia, which was found in 78% of EDS-

HT patients versus 10% of controls,<sup>17</sup> characterized by dizziness/ lightheadedness and pre-syncopal episodes, which were found in 88% and 83% of patients, respectively. Partial or complete failure of local anesthesia was described during biopsies and dental or obstetric procedures.<sup>74,75</sup> Hakim and Grahame found local anesthesia resistance in 58% of EDS-HT patients versus 21% of controls.<sup>32</sup> Proprioceptive acuity has been found to be impaired among EDS-HT adult patients<sup>76,77</sup> and pediatric patients.<sup>78</sup> Postural instability and balance and gait impairment, resulting in increased frequency of falls, were found among EDS-HT patients as compared to matched healthy controls.<sup>79</sup> Impaired proprioceptive acuity is thought to influence muscle strength. Therefore, improving muscle strength on the basis of proprioceptive impairment may be more important for reducing activity limitations than just improving muscle strength.<sup>80</sup> Chiari 1 malformation type 1 was found in 4.7% of EDS-HT patients<sup>19</sup> and may be associated with cranio-cervical instability and/or the tethered cord syndrome.

## Psychiatric Manifestations

Fear of joint pain and/or instability may lead to avoidance behavior (kinesiophobia) and exacerbate dysfunction and disability.<sup>3,7</sup> Depression and anxiety are more common among EDS-HT patients<sup>7,19,34</sup> and are exacerbated by fatigue and pain.<sup>11,15</sup>

## GENERAL REMARKS

The multisystemic nature of EDS-HT results in patients having difficulty coping with the syndrome, as well as medical personnel failing to understand the true nature of the condition. This may adversely affect the therapeutic relationship, giving rise to skepticism, resentment, distrust, and hostility on the part of the patient.<sup>3,7</sup>

Although EDS-HT is the most common type and the least severe type of EDS, it tends to be underdiagnosed and mistreated, sometimes leading to severe disability that may have been preventable if diagnosed and treated properly.<sup>64,81,82</sup> A survey among physiotherapists in the UK found that only 32% of respondents received formal training in EDS-HT management.<sup>83</sup> Patients





perceive a lack of awareness of the syndrome among health professionals and describe delays in diagnosis and access to appropriate health care services.<sup>84</sup> Many patients reported lengthy diagnosis trajectories and treatment for individual symptoms rather than EDS-HT as a whole. Receiving a correct diagnosis is necessary in order to access appropriate care pathways, for example, referral for physiotherapy for EDS-HT rather than for an acute single joint problem.<sup>84</sup> A study conducted among military personnel found misdiagnosis of EDS-HT has a disabling impact on military personnel with EDS-HT who are exposed to strenuous physical activities.<sup>85</sup> Significant neuromuscular and motor development problems have been found among a pediatric population, and delay in diagnosis resulted in poor control of pain and disruption of normal home life, schooling, and physical activities.<sup>64</sup> Furthermore, they conclude that knowledge of the diagnosis and appropriate interventions are likely to be highly effective in reducing the morbidity and cost to the health and social services.<sup>64</sup>

## DIAGNOSIS

Diagnosis relies on the revised Brighton criteria, but it is important to rule out other connective tissue disorders, especially Marfan syndrome and other types of EDS. Unfortunately, no genetic defect has been found, and for such a prevalent and complex genetic disorder multiple genes might be involved.

## MEDICAL MANAGEMENT

Treatment requires multidisciplinary co-operation and consulting with a cardiologist with echocardiogram monitoring every 2–5 years, orthopedic surgeon with a follow-up once a year, oral and maxillofacial surgeon for temporomandibular joint involvement, gastroenterologist when gastrointestinal manifestations are present, ophthalmologist to rule out other connective tissue diseases and when ocular manifestations are present, urologist and urogynecologist when urologic manifestations are suspected, neurologist and neurosurgeon when prolonged headache is present to rule out Chiari 1, and psychiatry when anxiety and/or depression are suspected. Allergo-

logic consultation may also be needed when there are multiple drug reactions and/or food allergies. An autonomic nervous system specialist should be consulted when signs and symptoms of POTS or other autonomic nervous system manifestations are present. Management includes physiotherapy and hydrotherapy aimed at symmetric and generalized muscle strengthening and proprioception acuity improvement, including deep connective tissue manipulations after each session, occupational therapy when wrists and fingers are involved, and cognitive behavioral therapy for proper adjustment to the chronic nature of the condition. Nutrition has an important role in treating EDS-HT, and nutritional deficiencies should be sought out and treated.

## CONCLUSION

Ehlers–Danlos syndrome-HT is a complex hereditary disorder which is multisystemic, probably due to the prevalence of connective tissue in all body systems. Its gene defect has yet to be found and might be of multigenetic nature, but until then we have to think about the possibility of EDS-HT in every chronic pain patient, and look for joint hypermobility as well as other multisystemic manifestations of this prevalent syndrome.

## ABBREVIATIONS

EDS	Ehlers–Danlos syndrome
HT	hypermobility type
JH	joint hypermobility
JHS	joint hypermobility syndrome
MVP	mitral valve prolapse
TMJ	temporomandibular joints

## FOOTNOTES

Conflict of interest: No potential conflict of interest relevant to this article was reported.

## REFERENCES

1. Beighton P, De Paepe A, Steinmann B, Tsipouras P, Wenstrup RJ. Ehlers-Danlos syndromes: revised nosology, Villefranche, 1997. Ehlers-Danlos National Foundation (USA) and Ehlers-Danlos Support Group (UK). *Am J Med Genet*. 1998;77:31–7. [https://doi.org/10.1002/\(SICI\)1096-8628\(19980428\)77:1%3C31::AID-AJMG8%3E3.O.CO;2-O](https://doi.org/10.1002/(SICI)1096-8628(19980428)77:1%3C31::AID-AJMG8%3E3.O.CO;2-O).
2. Grahame R. Ehlers-Danlos syndrome. *S Afr Med J*. 2016;106:S45–6. <https://doi.org/10.7196/SAMJ.2016.v106i6.10991>.
3. Levy, HP. Pagon RA, Adam MP, Ardinger HH, et al., editors. GeneReviews [Internet]. Seattle, WA: University of Washington, Seattle; 1993–2016 [accessed October 7, 2016]. Ehlers Danlos Syndrome, Hypermobility Type. Updated March 31, 2016. Available at: <http://bit.ly/2ePdTyk>.
4. De Paepe A, Malfait F. The Ehler-Danlos syndrome, a disorder with many faces. *Clin Genet*. 2012;82:1–11. <https://doi.org/10.1111/j.1399-0004.2012.01858.x>.
5. Pepin, MG.; Murray, ML.; Byers, PH. Pagon RA, Adam MP, Ardinger HH, et al., editors. GeneReviews [Internet]. Seattle, WA: University of Washington, Seattle; 1993–2016 [accessed October 7, 2016]. Vascular Ehlers-Danlos Syndrome. Updated Nov 19, 2015. Available at: <http://bit.ly/2fiszsrl>.
6. Malfait, F.; Wenstrup, R.; De Paepe, A. Pagon RA, Adam MP, Ardinger HH, et al., editors. GeneReviews [Internet]. Seattle, WA: University of Washington, Seattle; 1993–2016 [accessed October 7, 2016]. Ehlers-Danlos Syndrome, Classic Type. Updated Nov 19, 2015. Available at: <http://bit.ly/2dL9wGW>.
7. Branson JA, Kozłowska K, Kaczynski KJ, Roesler TA. Managing chronic pain in a young adolescent girl with Ehlers-Danlos syndrome. *Harv Rev Psychiatry*. 2011;19:259–70. <https://doi.org/10.3109/10673229.2011.614484>.
8. Hagberg C, Berglund B, Korpe L, Andersson-Norinder J. Ehlers-Danlos syndrome (EDS) focusing on oral symptoms: a questionnaire study. *Orthod Craniofac Res*. 2004;7:178–85. <https://doi.org/10.1111/j.1601-6343.2004.00288.x>.
9. De Coster PJ, Martens LC, De Paepe A. Oral health in prevalent types of Ehlers-Danlos syndromes. *J Oral Pathol Med*. 2005;34:298–307. <https://doi.org/10.1111/j.1600-0714.2004.00300.x>.
10. Rombaut L, Malfait F, Cools A, De Paepe A, Calders P. Musculoskeletal complaints, physical activity and health-related quality of life among patients with the Ehlers-Danlos syndrome hypermobility type. *Disabil Rehabil*. 2010;32:1339–45. <https://doi.org/10.3109/09638280903514739>.
11. Rombaut L, Malfait F, De Paepe A, et al. Impairment and impact of pain in female patients with Ehlers-Danlos syndrome: a comparative study with fibromyalgia and rheumatoid arthritis. *Arthritis Rheum*. 2011;63:1979–87. <https://doi.org/10.1002/art.30337>.
12. Rombaut L, Malfait F, De Wandele I, et al. Medication, surgery, and physiotherapy among patients with the hypermobility type of Ehlers-Danlos syndrome. *Arch Phys Med Rehabil*. 2011;92:1106–12. <https://doi.org/10.1016/j.apmr.2011.01.016>.
13. Voermans NC, Knoop H. Both pain and fatigue are important possible determinants of disability in patients with the Ehlers-Danlos syndrome hypermobility type. *Disab Rehabil*. 2011;33:706–7. <https://doi.org/10.3109/09638288.2010.531373>.
14. Sacheti A, Szemere J, Bernstein B, Tafas T, Schechter N, Tsipouras P. Chronic pain is a manifestation of the Ehlers-Danlos syndrome. *J Pain Symptom Manage*. 1997;14:88–93. [https://doi.org/10.1016/S0885-3924\(97\)00007-9](https://doi.org/10.1016/S0885-3924(97)00007-9).
15. Voermans NC, Knoop H, Bleijenberg G, van Engelen BG. Pain in ehlers-danlos syndrome is common, severe, and associated with functional impairment. *J Pain Symptom Manage*. 2010;40:370–8. <https://doi.org/10.1016/j.jpainsymman.2009.12.026>.
16. Rowe PC, Barron DF, Calkins H, Maumenee IH, Tong PY, Geraghty MT. Orthostatic intolerance and chronic fatigue syndrome associated with Ehlers-Danlos syndrome. *J Pediatr*. 1999;135:494–9. [https://doi.org/10.1016/S0022-3476\(99\)70173-3](https://doi.org/10.1016/S0022-3476(99)70173-3).
17. Gazit Y, Nahir AM, Grahame R, Jacob G. Dysautonomia in the joint hypermobility syndrome. *Am J Med*. 2003;115:33–40. [https://doi.org/10.1016/S0002-9343\(03\)00235-3](https://doi.org/10.1016/S0002-9343(03)00235-3).
18. Mathias CJ, Low DA, Iodice V, Owens AP, Kirbis M, Grahame R. Postural tachycardia syndrome--current experience and concepts. *Nat Rev Neurol*. 2011;8:22–34. <https://doi.org/10.1038/nrneurol.2011.187>.
19. Castori M, Camerota F, Celletti C, et al. Natural history and manifestations of the hypermobility type Ehlers-Danlos syndrome: a pilot study on 21 patients. *Am J Med Genet A*. 2010;152A:556–64.
20. Wenstrup RJ, Meyer RA, Lyle JS, et al. Prevalence of aortic root dilation in the Ehlers-Danlos syndrome. *Genet Med*. 2002;4:112–17. <https://doi.org/10.1097/00125817-200205000-00003>.
21. McDonnell NB, Gorman BL, Mandel KW, et al. Echocardiographic findings in classical and hypermobile Ehlers-Danlos syndromes. *Am J Med Genet A*. 2006;140:129–36. <https://doi.org/10.1002/ajmg.a.31035>.



22. Atzinger CL, Meyer RA, Khoury PR, Gao Z, Tinkle BT. Cross-sectional and longitudinal assessment of aortic root dilation and valvular anomalies in hypermobile and classic Ehlers-Danlos syndrome. *J Pediatr*. 2011;158:826–30.e1.
23. Levy HP, Mayoral W, Collier K, Tio TL, Francomano CA. Gastroesophageal reflux and irritable bowel syndrome in classical and hypermobile Ehlers Danlos syndrome (EDS). *Am J Hum Genet*. 1999;65:A69.
24. Anstey A, Mayne K, Winter M, Van de Pette J, Pope FM. Platelet and coagulation studies in Ehlers-Danlos syndrome. *Br J Dermatol*. 1991;125:155–63. <https://doi.org/10.1111/j.1365-2133.1991.tb06063.x>.
25. De Paepe A, Malfait F. Bleeding and bruising in patients with Ehlers-Danlos syndrome and other collagen vascular disorders. *Br J Haematol*. 2004;127:491–500. <https://doi.org/10.1111/j.1365-2141.2004.05220.x>.
26. Mast KJ, Nunes ME, Ruymann FB, Kerlin BA. Desmopressin responsiveness in children with Ehlers-Danlos syndrome associated bleeding symptoms. *Br J Haematol*. 2009;144:230–3. <https://doi.org/10.1111/j.1365-2141.2008.07446.x>.
27. Gharbiya M, Moramarco A, Castori M, et al. Ocular features in joint hypermobility syndrome/Ehlers-Danlos syndrome hypermobility type: a clinical and in vivo confocal microscopy study. *Am J Ophthalmol*. 2012;154:593–600. e1. <https://doi.org/10.1016/j.ajo.2012.03.023>.
28. McIntosh LJ, Mallett VT, Frahm JD, Richardson DA, Evans MI. Gynecologic disorders in women with Ehlers-Danlos syndrome. *J Soc Gynecol Investig*. 1995;2:559–64.
29. Castori M, Morlino S, Dordoni C, et al. Gynecologic and obstetric implications of the joint hypermobility syndrome (a.k.a. Ehlers – Danlos syndrome hypermobility type) in 82 Italian patients. *Am J Med Genet Part A*. 2012;158A:2176–82. <https://doi.org/10.1002/ajmg.a.35506>.
30. Volkov N, Nisenblat V, Ohel G, Gonen R. Ehlers-Danlos syndrome: insights on obstetric aspects. *Obstet Gynecol Surv*. 2007;62:51–7. <https://doi.org/10.1097/01.ogx.0000251027.32142.63>.
31. Dutta I, Wilson H, Oteri O. Pregnancy and delivery in ehlers-danlos syndrome (hypermobility type): review of the literature. *Obstet Gynecol Int*. 2011;2011:306413. <https://doi.org/10.1155/2011/306413>.
32. Hakim AJ, Grahame R, Norris P, Hopper C. Local anaesthetic failure in joint hypermobility syndrome. *J R Soc Med*. 2005;98:84–5. <https://doi.org/10.1258/jrsm.98.2.84>.
33. Milhorat TH, Bolognese PA, Nishikawa M, McDonnell NB, Francomano CA. Syndrome of occipitoatlantoaxial hypermobility, cranial settling, and chiari malformation type I in patients with hereditary disorders of connective tissue. *J Neurosurg Spine*. 2007;7:601–9. <https://doi.org/10.3171/SPI-07/12/601>.
34. Baeza-Velasco C, Gély-Nargeot MC, Bulbena Vilarrasa A, Bravo JF. Joint hypermobility syndrome: problems that require psychological intervention. *Rheumatol Int*. 2011;31:1131–6. <https://doi.org/10.1007/s00296-011-1839-5>.
35. Bulbena A, Duro JC, Porta M, et al. Anxiety disorders in the joint hypermobility syndrome. *Psychiatry Reserve*. 1993;46:59–68. [https://doi.org/10.1016/0165-1781\(93\)90008-5](https://doi.org/10.1016/0165-1781(93)90008-5).
36. Grahame R. Time to take hypermobility seriously (in adults and children). *Rheumatology (Oxford)*. 2001;40:485–7. <https://doi.org/10.1093/rheumatology/40.5.485>.
37. Gurley-Green S. Living with the hypermobility syndrome. *Rheumatology (Oxford)*. 2001;40:487–9. <https://doi.org/10.1093/rheumatology/40.5.487>.
38. Keer, R.; Grahame, R. *Hypermobility Syndrome – Recognition and Management for Physiotherapists*. London: Butterworth-Heinemann; 2003.
39. Simmonds JV, Keer RJ. Hypermobility and the hypermobility syndrome. *Man Ther*. 2007;12:298–309. <https://doi.org/10.1016/j.math.2007.05.001>.
40. Child AH. Joint hypermobility syndrome: inherited disorder of collagen synthesis. *J Rheumatol*. 1986;13:239–43.
41. Beighton, P.; Grahame, R.; Bird, HA. Genetic Aspects of the Hypermobility Syndrome. In: Beighton P, Grahame R, Bird HA, editors. *Hypermobility of Joints*. 2nd ed. Berlin, Germany: Springer; 1989. pp. 55–66. [https://doi.org/10.1007/978-1-4471-3900-3\\_5](https://doi.org/10.1007/978-1-4471-3900-3_5).
42. Beighton, P.; Grahame, R.; Bird, HA. Clinical Features of Hypermobility Syndrome. In: Beighton P, Grahame R, Bird HA, editors. *Hypermobility of Joints*. 2nd ed. Berlin, Germany: Springer; 1989. pp. 67–84. [https://doi.org/10.1007/978-1-4471-3900-3\\_6](https://doi.org/10.1007/978-1-4471-3900-3_6).
43. Beighton PH, Solomon L, Soskolne CL. Articular mobility in an African population. *Ann Rheum Dis*. 1973;32:413–18. <https://doi.org/10.1136/ard.32.5.413>.
44. Grahame, R. Hypermobility and Hypermobility Syndrome. In: Keer R, Grahame R, editors. *Hypermobility Syndrome – Recognition and Management For Physiotherapists*. London, UK: Butterworth-Heinemann; 2003. pp. 1–14. <https://doi.org/10.1016/B978-0-7506-5390-9.50005-8>.
45. Kirk JA, Ansell BM, Bywaters EL. The hypermobility syndrome. *Ann Rheum Dis*. 1967;26:419–25. <https://doi.org/10.1136/ard.26.5.419>.
46. Grahame R. Joint hypermobility: clinical aspects. *Proc R Soc Med*. 1971;64:32–4.
47. Beighton P, De Paepe A, Danks D, et al. International Nosology of Heritable Disorders of Connective Tissue, Berlin, 1986. *Am J Med Genet*. 1988;29:581–94. <https://doi.org/10.1002/ajmg.1320290316>.





48. Grahame R, Bird HA, Child A. The revised (Brighton 1998) criteria for the diagnosis of benign joint hypermobility syndrome (BJHS). *J Rheumatol*. 2000;27:1777–9.
49. El-Shahaly HA, el-Sherif AK. Is the benign joint hypermobility syndrome benign? *Clin Rheumatol*. 1991;10:302–7. <https://doi.org/10.1007/BF02208695>.
50. Dolan AL, Mishra MB, Chambers JB, Grahame R. Clinical and echocardiographic survey of the Ehlers-Danlos syndrome. *Br J Rheumatol*. 1997;36:459–62. <https://doi.org/10.1093/rheumatology/36.4.459>.
51. Mishra MB, Ryan P, Atkinson P, et al. Extra-articular features of benign joint hypermobility syndrome. *Br J Rheumatol*. 1996;35:861–6. <https://doi.org/10.1093/rheumatology/35.9.861>.
52. Camerota F, Castori M, Celletti C, et al. Heart rate, conduction and ultrasound abnormalities in adults with joint hypermobility syndrome/Ehlers-Danlos syndrome, hypermobility type. *Clin Rheumatol*. 2014;33:981–7. <https://doi.org/10.1007/s10067-014-2618-y>.
53. Kozanoglu E, Coskun Benlidayi I, Eker Akilli R, Tasal A. Is there any link between joint hypermobility and mitral valve prolapse in patients with fibromyalgia syndrome? *Clin Rheumatol*. 2016;35:1041–4. <https://doi.org/10.1007/s10067-015-3024-9>.
54. Ainsworth SR, Aulicino PL. A survey of patients with Ehlers-Danlos syndrome. *Clin Orthop Rel Res*. 1993;286:250–256. <https://doi.org/10.1097/00003086-199301000-00037>.
55. Pemberton JW, MacKenzie Freeman H, Schepens CL. Familial retinal detachment and the Ehlers-Danlos syndrome. *Arch Ophthalmol*. 1966;76:817–24. <https://doi.org/10.1001/archophth.1966.03850010819007>.
56. Tinkle, BT. A Guide for the Issues & Management of Ehlers-Danlos Syndrome Hypermobility Type and The Hypermobility Syndrome. Niles, IL: Left Paw Press, LLC; 2010. Joint Hypermobility Handbook.
57. Meyer E, Ludatcher RM, Zonis S. Collagen fibril abnormalities in the extraocular muscles in Ehlers-Danlos syndrome. *J Pediatr Ophthalmol Strabismus*. 1988;25:67–72.
58. Castori M, Camerota F, Celletti C, Grammatico P, Padua L. Quality of life in the classic and hypermobility types of Ehlers-Danlos syndrome. *Ann Neurol*. 2010;67:145–7. <https://doi.org/10.1002/ana.21934>.
59. Al-Rawi ZS, Al-Rawi ZT. Joint hypermobility in women with genital prolapse. *Lancet*. 1982;1:1439–41. [https://doi.org/10.1016/S0140-6736\(82\)92453-9](https://doi.org/10.1016/S0140-6736(82)92453-9).
60. Norton PA, Baker JE, Sharp HC, Warenski JC. Genitourinary prolapsed and joint hypermobility in women. *Obstet Gynecol*. 1995;85:225–8. [https://doi.org/10.1016/0029-7844\(94\)00386-R](https://doi.org/10.1016/0029-7844(94)00386-R).
61. Carley ME, Schaffer J. Urinary incontinence and pelvic organ prolapsed in women with Marfan or Ehlers Danlos syndrome. *Am J Obstet Gynecol*. 2000;182:1021–3. <https://doi.org/10.1067/mob.2000.105410>.
62. Aydeniz A, Dikensoy E, Cebesoy B, Altinadaq O, Gursay S, Balat O. The relation between genitourinary prolapsed and joint hypermobility in Turkish women. *Arch Gynecol Obstet*. 2010;281:301–4. <https://doi.org/10.1007/s00404-009-1103-3>.
63. de Kort LM, Verhulst JA, Engelbert RH, Uiterwaal CS, de Jong TP. Lower urinary tract dysfunction in children with generalized hypermobility of joints. *J Urol*. 2003;170:1971–4. <https://doi.org/10.1097/01.ju.0000091643.35118.d3>.
64. Adib N, Davies K, Grahame R, Woo P, Murray KJ. Joint hypermobility syndrome in childhood A not so benign multisystem disorder. *Rheumatology (Oxford)*. 2005;44:744–50. <https://doi.org/10.1093/rheumatology/keh557>.
65. Arunkalaivanan AS, Morrison A, Jha S, Blann A. Prevalence of urinary and faecal incontinence among female members of the Hypermobility Syndrome Association (HMSA). *J Obstet Gynecol*. 2009;29:126–8. <https://doi.org/10.1080/01443610802664747>.
66. Grahame R. Pain, distress and joint hyperlaxity. *Joint Bone Spine*. 2000;67:157–63.
67. Dordoni C, Ritelli M, Venturini M, et al. Recurring and generalized visceroptosis in Ehlers-Danlos syndrome hypermobility type. *Am J Med Genet A*. 2013;161A:1143–7. <https://doi.org/10.1002/ajmg.a.35825>.
68. Taylor DJ, Wilcox I, Russell JK. Ehlers-Danlos syndrome during pregnancy: a case report and review of the literature. *Obstet Gynecol Surv*. 1981;36:277–81. <https://doi.org/10.1097/00006254-198106000-00001>.
69. De Vos M, Nuytinck L, Verellen C, De Paepe A. Preterm premature rupture of membranes in a patient with the hypermobility type of the Ehlers-Danlos syndrome. A case report. *Fetal Diagn Ther*. 1999;14:244–7. <https://doi.org/10.1159/000020930>.
70. Lind J, Wallenburg HC. Pregnancy and the Ehler-Danlos syndrome: a retrospective study in a Dutch population. *Acta Obstet Gynecol Scand*. 2002;81:293–300. <https://doi.org/10.1034/j.1600-0412.2002.810403.x>.
71. Jones TL, Ng C. Anaesthesia for caesarean section in a patient with Ehlers-Danlos syndrome associated with postural orthostatic tachycardia syndrome. *Int J Obstet Anesth*. 2008;17:365–9. <https://doi.org/10.1016/j.ijoa.2008.04.003>.
72. Mato H, Berde T, Hasson N, Grahame R, Maillard S. A review of symptoms with benign joint hypermobility syndrome in children. *Ped Rheumatol*. 2008;6(Suppl 1):P155. <https://doi.org/10.1186/1546-0096-6-S1-P155>.



73. Jacome DE. Headache in Ehlers-Danlos syndrome. Cephalalgia. 1999;19:791–6. <https://doi.org/10.1046/j.1468-2982.1999.1909791.x>.
74. Kaalund S, Hogsaa B, Grevy C, Oxlund H. Reduced strength of skin in Ehlers-Danlos syndrome type III. Scand J Rheumatol. 1990;19:67–70. <https://doi.org/10.3109/03009749009092623>.
75. Arendt-Nielsen L, Kaalund S, Bjerring P, Hogsaa B. Insufficient effect of local analgesics in Ehlers-Danlos type III patients(connectivetissue disorder).Acta Anaesth Scand. 1990;34:358–61. <https://doi.org/10.1111/j.1399-6576.1990.tb03103.x>.
76. Mallik AK, Ferrell WR, McDons AG, Sturrock RD. Impaired proprioceptive acuity at the proximal interphalangeal joint in patients with the hypermobility syndrome. Br J Rheumatol. 1994;33:631–7. <https://doi.org/10.1093/rheumatology/33.7.631>.
77. Hall MC, Ferrell WR, Sturrock RD, Hamblen DL, Baxendale RH. The effect of the hypermobility syndrome on knee joint proprioception. Br J Rheumatol. 1995;34:121–5. <https://doi.org/10.1093/rheumatology/34.2.121>.
78. Fatoye F, Palmar S, Macmillan F, Rowe P, van der Linden M. Proprioception and muscle torque deficits in children with hypermobility syndrome. Rheumatology (Oxford). 2009;48:152–7. <https://doi.org/10.1093/rheumatology/ken435>.
79. Rombaut L, Malfait F, DeWandele I, et al. Balance, gait, falls, and fear of falling in women with the hypermobility type of Ehlers-Danlos syndrome. Arthritis Care Res (Hoboken). 2011;63:1432–9. <https://doi.org/10.1002/acr.20557>.
80. Scheper M, Rombaut L, de Vreis J, et al. The association between muscle strength and activity limitations in patients with the hypermobility type of Ehlers-Danlos syndrome: the impact of proprioception. Disabil Rehabil. 2016 Jun 24;:1–7. [Epub ahead of print].
81. Grahame R. Joint hypermobility: emerging disease or illness behavior? Clin Med (Lond). 2013;13(Suppl 6):s50–2. <https://doi.org/10.7861/clinmedicine.13-6-s50>.
82. Wolf JM, Cameron KL, Owens BD. Impact of joint laxity and hypermobility on the musculoskeletal system. J Am Acad Orthop Surg. 2011;19:463–71. <https://doi.org/10.5435/00124635-201108000-00002>.
83. Palmar S, Cramp F, Lewis R, Muhammad S, Clark E. Diagnosis, management and assessment if adults with joint hypermobility syndrome: a UK-wide survey of physiotherapy practice. Musculoskeletal Care. 2015;13:101–11. <https://doi.org/10.1002/msc.1091>.
84. Terry RH, Palmer ST, Rimes KA, Clark CJ, Simmonds JV, Horwood JP. Living with joint hypermobility syndrome: patient experience of diagnosis, referral and self-care. Fam Pract. 2015;32:354–8. <https://doi.org/10.1093/fampra/cmv026>.
85. Mullick G, Bhakuni DS, Shnmuganandan K, et al. Clinical profile of benign joint hypermobility syndrome from a tertiary care military hospital in India. Int J Rheum Dis. 2013;16:590–4. <https://doi.org/10.1111/1756-185x.12024>.



#### Loose Connections, Autumn 2016 Photograph Credits

Cover: Paul Itkin

TOC: Johann S.

Page 3: Dallas Sun Rise 15, © July 2008 by richardphotos(Richard Gorden) Mapps (Michael Mapplebeck)

Page 3,12, 18: Teddy Kelley

Page 4, 6: Elliott Englemann

Page 7, 13, 19: Faded Forest

Page 8, 15, 19: NASA

Page 10, 16, 28: Vladimir Kudinov

Page 11, 17: Steve Richey

Page 12: francois-hoang

Page 27: Vincent Guth





## Publisher Information



### FOUNDER

Nancy Hanna Rogowski (1957–1995)

### Co-Executive Directors

Lara Bloom and Shane Robinson

### Board of Directors

Sandra Aiken Chack, *Chair*

Susan Hawkins, *Vice Chair*

Richie Taffet, MPH, *Secretary*

Richard Malenfant, MBA, MPH, *Treasurer*

Peter Cohl

Heidi Collins, MD

Clair Francomano, MD

Fraser C. Henderson, Sr., MD

Fransiska Malfait, MD, PhD

Jane Mitakides

Linda Neumann-Potash, RN, MN, CBN

Jakob Rasmussen

John Zonarich, Esq.

### Medical and Scientific Board

Fransiska Malfait, MD, PhD, *Chair*

Qasim Aziz, MBBS, FRCP, PhD

Peter Byers, MD

Marco Castori, MD, PhD

Raymond Dalglish, PhD

Clair Francomano, MD, PhD

Cecilia Giunta, PhD

Rodney Grahame CBE, MD, FRCP, FACP, FRSA

Alan Hakim, MA, FRCP

Tomoki Kosho, MD

Cathleen L. Raggio, MD

Glenda Sobey, MB ChB, BSc Med (Hons), FC Derm

Jane Simmonds, MCSP, MMACP, FHEA

Brad Tinkle, MD, PhD

*Loose Connections* is published by The Ehlers-Danlos Society. The Society can be contacted by email, [info@ehlers-danlos.com](mailto:info@ehlers-danlos.com), or in writing: The Ehlers-Danlos Society, P.O. Box 87463, Montgomery Village, MD 20886 USA (Phone: +1 410-670-7577); The Ehlers-Danlos Society – Europe, Office 7, 35-37 Ludgate Hill, London EC4M 7JN UK (Phone: +44 203 887 6132).

Copyright 2017 The Ehlers-Danlos Society unless otherwise protected. The opinions expressed in *Loose Connections* are those of the contributors, authors, or advertisers, and do not necessarily reflect the views of The Ehlers-Danlos Society, the editorial staff, the Medical and Scientific Board, or the Board of Directors. The Ehlers-Danlos Society does not endorse any products.

### GUIDELINES FOR SUBMISSIONS TO *LOOSE CONNECTIONS*

1. Attach your text document in either Word (.doc) or Rich Text Format (.rtf) to an email sent to [mark.martino@ehlers-danlos.com](mailto:mark.martino@ehlers-danlos.com) that also tells us how to reach you for more information.
2. For photographs, attach them to an email to [mark.martino@ehlers-danlos.com](mailto:mark.martino@ehlers-danlos.com); please identify the event or cause for the photographs, including any relevant identification (persons involved, date, photographer's name if needed) and how to reach you for more information.
3. Text articles, photographs, or any other submissions to *Loose Connections* are accepted only on condition that publication of that material is not under restrictions on its publication. The Ehlers-Danlos Society reserves all and final editorial privileges, including the right to choose not to print a submitted story; submissions may be edited at the discretion of the editorial staff.

