2016 Donors

Ehlers–Danlos Syndrome—Hypermobility Type: A Much Neglected Multisystemic Disorder
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Ehlers-Danlos Syndrome—Hypermobility Type: A Much Neglected Multisystemic Disorder
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THE EHLERS–DANLOS SOCIETY

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

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Our 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

Hope Begins With You
From the Editor’s Desk: “And acquainted with grief”

All 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.

Il duolo infranga questi ritorte,
de’ miei martiri sol per pietà.

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

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.

Joyce DiDonato & Il Pomo D’oro
https://youtu.be/Pr1Tmpt43hg
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.

“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”_
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.

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

Tilly 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 as 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)

It’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 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 by 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.
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 hast

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

It's 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
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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

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. 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. Ehlers–Danlos syndrome-hypermobility type (EDS-HT) is considered to be the most common subtype of EDS and the least severe
It is characterized by joint laxity, soft, stretchy, and often semi-transparent skin, and musculoskeletal complications, without severe complications of arterial dissection or bowel rupture seen in EDS-vascular type, \textsuperscript{1,5} and without hemosiderotic scars and molluscoid pseudotumors seen in the EDS-classical type. \textsuperscript{1,6} 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. \textsuperscript{3,7–12} 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. \textsuperscript{7–9} Soft tissue overuse injury may lead to tendonitis and bursitis \textsuperscript{10,11,12} without joint inflammation in most cases. \textsuperscript{3,11} Although an inflammatory component is rare, EDS–HT carries a high potential for disability \textsuperscript{13} due to recurrent dislocations and subluxations and chronic pain. \textsuperscript{8,11,12,14,15} Throughout the years, extra-articular manifestations have been described, including cardiovascular and autonomic nervous system, \textsuperscript{16–22} gastrointestinal, \textsuperscript{19,23} hematologic, \textsuperscript{24–26} ocular, \textsuperscript{27} gynecologic, \textsuperscript{19,28–31} neurologic, \textsuperscript{7,8,11,19,34,35} and psychiatric manifestations, \textsuperscript{7,8,11,19,34,35} emphasizing the multisystemic nature of EDS–HT. Unfortunately, EDS–HT is under-recognized and inadequately managed, \textsuperscript{36–38} leading to neglect of these patients which may lead to severe disability that almost certainly could have been avoided. \textsuperscript{39}

**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 \textsuperscript{40,41} and may pose no problem. Acquired hypermobility may also result from changes in connective tissue in other diseases such as systemic lupus erythematosus. \textsuperscript{42} Joint hypermobility is recognized by the nine-point Beighton score \textsuperscript{43} (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](http://hypermobility.org/help-advice/hypermobility-syndromes/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 by 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), \textsuperscript{44} is a clinical condition of JH with symptoms of joint instability, arthralgia, myalgia, soft tissue injuries, and arthritis. \textsuperscript{45,46} Diagnosis relies on the Brighton criteria (Table 1). \textsuperscript{47,48} The predominant presenting complaint is pain, which is often widespread and longstanding, with patients reporting pain ranging from 15 days to 45 years. \textsuperscript{39,49} 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. \textsuperscript{3,39,45} Severity sometimes correlates with the degree of joint instability. \textsuperscript{3,15} 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 first-degree relative.

Taken with permission from the Hypermobility Syndromes Association (HMSA) site ([http://hypermobility.org/helpadvice/hypermobilitysyndromes/thebrightonscore/](http://hypermobility.org/helpadvice/hypermobilitysyndromes/thebrightonscore/)).

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.\textsuperscript{3,11,12,15} 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.\textsuperscript{3} 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,\textsuperscript{20,21,22} necessitating echocardiographic evaluation and surveillance. Raynaud phenomenon was found in 38% of EDS-HT patients.\textsuperscript{19} Patients with EDS-HT may suffer from palpitations, chest pain, dizziness, pre-syncope, and syncope,\textsuperscript{17} 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.\textsuperscript{47} With more modern evaluation techniques clinically significant MVP has not been found to be more prevalent among EDS-HT patients;\textsuperscript{21,22,50,51} For this reason MVP was removed from the Brighton criteria in 1998.\textsuperscript{48} The frequency of MVP among EDS-HT patients was found to be 28%–67% in more recent studies,\textsuperscript{52,53} 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.\textsuperscript{16–18}

Gastrointestinal Manifestations

Gastroesophageal reflux was found in 57% of EDS-HT patients.\textsuperscript{19,23} 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.\textsuperscript{3}

Hematologic Manifestations

Easy bruising and bleeding tendency is common in all EDS types, including EDS-HT.\textsuperscript{25} It manifests with prolonged bleeding time,\textsuperscript{24,26} oral mucosa fragility with mucosal bruises,\textsuperscript{9} and menometrorrhagia.\textsuperscript{54} Since coagulation tests are normal\textsuperscript{24–26} 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,\textsuperscript{54} and high myopia of more than −6.0 diopters was found in 16% of patients compared with 0% in the control group.\textsuperscript{3,27} Strabismus was found in 7% of EDS-HT pediatric patients\textsuperscript{55} (as opposed to only 2%–4% of the general pediatric population), and it is often refractory to surgical correction.\textsuperscript{56} Meyer et al. found size variations and shape abnormalities of collagen fibrils in the extra-ocular muscles that control the movement of the eye.\textsuperscript{57}

Gynecologic Manifestations

Dysmenorrhea and menorrhagia are common\textsuperscript{19,28,29,54,56} 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,\textsuperscript{28,29,58} thought to be caused by small tears in the vaginal surface and lack of appropriate vaginal secretions.\textsuperscript{56} Pelvic organ prolapse is common,\textsuperscript{19,28,29,56,59–62} including uterine prolapse which was found in almost 40% of women with EDS-HT.\textsuperscript{49}

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.\textsuperscript{63} In another pediatric series 13% of girls and 6% of boys suffered from urinary tract infections.\textsuperscript{64} Stress urinary incontinence was found in 40%–
70% of women with EDS-HT, often earlier in life, thought to be due to a weakened pelvic floor, which may be worsened to bladder prolapse. Fecal incontinence was found in up to almost 15% of EDS-HT patients, as compared to only 2.2% of the general population. Rectal prolapse may also be found among EDS-HT patients. 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.

Obstetric Manifestations

While labor and delivery might be rapid (shorter than 4 hours), and premature rupture of membranes is common, pregnancy in women with EDS-HT is generally normal with good maternal and neonatal outcome. However, joint laxity and pain may increase during pregnancy. 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). Varicose veins in the legs and the vulva are more common among pregnant women with EDS-HT.

Dysautonomia, characterized by lightheadedness, dizziness, fainting, etc., may worsen during pregnancy, and when postural orthostatic tachycardia syndrome (POTS) is present a blood pressure fall was reported. Women with EDS-HT are more prone to postpartum hemorrhage (19% versus 7%) and complicated perineal wounds (8% versus none). 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%).

Neurologic Manifestations

A total of 40% of children with EDS-HT and 50% of adults 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. Many complain of headaches related to the neck or facial pain that might be related to jaw or TMJ problems. Headaches may also be part of dysautonomia, which was found in 78% of EDS-HT patients versus 10% of controls, 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. Hakim and Grahame found local anesthesia resistance in 58% of EDS-HT patients versus 21% of controls. Proprioceptive acuity has been found to be impaired among EDS-HT adult patients and pediatric patients. 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. 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. Chiari 1 malformation type 1 was found in 4.7% of EDS-HT patients 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. Depression and anxiety are more common among EDS-HT patients and are exacerbated by fatigue and pain.

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. 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. A survey among physiotherapists in the UK found that only 32% of respondents received formal training in EDS-HT management.
perceive a lack of awareness of the syndrome among health professionals and describe delays in diagnosis and access to appropriate health care services. 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. 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. 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. 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.

**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. Allergologic 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**

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