



COMMENT

Family reflections: raising kids with chronic pain

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It's hard enough to raise teenagers, but how do you raise teenagers facing complex medical conditions? This is the question and challenge that we undertake every day in our house.

In 2015, our then 9-year-old son Cam, was diagnosed with Hypermobile Ehlers Danlos Syndrome (h-EDS) after many years of strange and progressing symptoms. His most common symptom was pain, but it changed day by day—minute by minute. His whole body seemed to be like a whack a mole game of hurting. He would wake in the morning and maybe his foot or back hurt severely. By the end of the day I would receive a call from him in tears because his legs hurt so badly that it took him 20 min to make the short walk three blocks home from school. I watched him walk across the room and witnessed his knee dislocate. We were drinking tea one night and he cried out and grabbed his head as a sudden headache came on. I looked at my fourth-grade child and realized he was having pain every single day. Bad pain. This was not normal. And then I remembered. Four years ago we had seen a doctor who mentioned a condition called Ehlers Danlos Syndrome because Cam, at age 5, complained often of knee pain and he was also very flexible. I dug out the article and my heart sunk as I read about this genetic condition that so aptly described my child.

Armed with a diary of his daily pain and the article, I headed to Cam's primary care physician. I remember the sadness in his doctor's eyes as I mentioned the article and handed him the pain diary to review. He just nodded and said he thought it was appropriate to send us for genetic testing for a formal evaluation. My heart, already low from sinking before, dropped even lower. It's never really come all the way back up.

So, we headed for genetic testing and, after a physical exam and many questions, Cam was officially diagnosed with h-EDS. Everything changed that day. Our child, our ridiculously smart, precocious, funny, child was handed a diagnosis that recognized he was on a very difficult journey into an unknown future of potential disability, medical complexity, and pain. Pain and children shouldn't be in the same sentence. We also learned that day that h-EDS is rarely the only diagnosis someone has—that this is an “umbrella” condition with many other conditions under or associated.

Ehlers Danlos Syndromes (there are at least 13 known types) are connective tissue disorders in which the body does not create appropriate connective tissue. Connective tissue literally holds, connects, the body together. It is found in skin, organs, and joints. So, pain and disorder are rarely limited to one joint, organ, or system. Folks with EDS live with chronic pain due to daily dislocations throughout their bodies, but often also have other “odd” pain or dysfunction. Each person is different, but for Cam it's wide-spread body pain (especially in the knees, feet, and back), headaches, celiac disease, chronic constipation, acid reflux, anxiety, chronic pain, and cold urticaria—yes, he lives in Wisconsin

but is allergic to the cold, with sudden sensation like when in cold pools or cold air! EDS means lots of doctors and appointments. In the first year, we saw a pain doctor, physical therapist, cardiologist, gastroenterologist, psychologist, occupational therapist, orthopedist, neurologist, and primary care. We look back at it now and call it the “diagnosis storm.” Little did we realize at that time another storm was coming.

In 2018 Cam's older sister Mary was also diagnosed with h-EDS. Her onset was different. It was a bit less dramatic, so it took more time for us to see what I now feel like should have been obvious. On Halloween of 2016 Mary twisted her ankle trick or treating. We iced it and elevated it and sent the other kids out to get candy for her. We assumed that after a few days the ankle would get better and it did, for the most part. The problem was it kept getting re-injured. Gym class became a nightmare for her ankle because it was a lot of running. But still, we didn't think EDS because it was just one joint and one pain. We got a brace and worked out some alternate exercises which helped. But the ankle continued to bother her on and off. Then, by 2018, the headaches started. As did the other aches and pains. Suddenly Mary was having daily pain too. Headaches, back aches, and rib pain. Also, a lot of fatigue as well as feeling dizzy often. My heart dropped even lower the day she was also handed an h-EDS diagnosis. Because this time I knew more about what it meant and how unfair it was. Mary went on to also be diagnosed with migraines, chronic fatigue, anxiety, and Postural Orthostatic Tachycardia Syndrome. My wise, quiet, intelligent girl was facing an equally daunting and unfairly hard future as her brother.

So now it is 2020 and we've been in the battle, as a family, for four years. What does a household with two chronically ill teenagers look like? Well, a lot like anyone else's on the surface. Homework is done daily, friends come over, siblings argue, the dog needs walking, there's a random sock on the living room floor. But when you take a closer look and listen you find out that it's not like other homes. The kitchen counter is filled with medication, school calendars are constantly interrupted by medical appointments, mornings aren't just hard because teenagers don't like waking up but because they are waking up with dislocations and migraines while preparing to go to school. We have more doctors' appointments than sleepovers. Planning a day trip to the mall or to any destination involves assessing if we need knee, ankle or wrist braces. Do we have enough energy to walk half a mile? Do we need to bring a wheelchair? We've gained a lot of patience and wisdom.

We freely share what has helped us so others can try it too. In a nutshell, we talk about our approach to pain management as a triangle; it has three sides:

- The first side is medical—our doctors who we see for a combined total of more than 100 appointments per year. This is

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where we find the medicines that help, or sometimes don't help. Getting the right medications on board helps even things out a bit. This is where we get referrals to other specialists. This is where we brainstorm.

- The second side is body. This is where we engage with physical and occupational therapists. Keeping our bodies strong and focusing problem areas helps reduce injury and helps increase stamina.

- The third side is mind. My kids each see a therapist, and this helps in many ways. First, it gives them safe place to just vent about how hard this battle is. Furthermore, the therapists help devise strategies for coping with this life-long burden as well as setting them up for success in things like school and social situations.

My kids will tell you that when all three sides of the triangle are supported, they do their best.

We've started a support group, traveled to conferences, made friends with other pain warriors, and dedicated ourselves to being advocates. There's a certain empowerment that comes with helping others when you're faced with a medical condition that currently doesn't have a cure. Recently, Mary had a special opportunity to present at a medical conference for pediatric pain providers. She shared with them some of the daily challenges she faces, her perspective, and what she felt they needed to know from a kid with chronic pain. Her messages focused around honesty, validation, and continued dedication to improvement.

Our request to medical professionals, be they doctors, nurses, or researchers, is that you recognize that EDS, and chronic pain in any condition, is real. That you are honest with us when you explain what you know and don't know about EDS and its associated condition. That you tackle pain as a team, like the triangle mentioned previously, by setting your patients up for medical, mind, and body care. And that you will continue to learn

about and study this disorder to find new therapies to treat it. Specifically discovering the gene for h-EDS is at the top of our wish list.

EDS is not well known in the medical community. Some medical professionals have never heard of it, while others say things along the lines of "oh, yeah, that's the bendy disorder," assuming it just means people are double jointed. As with so many other rare disorders, education is lacking while need is growing. Recently, an organization called Chronic Pain Partners has created an accredited CME course about EDS for medical professionals found at <https://www.chronicpainpartners.com/new-ehlers-danlos-syndromes-physician-cme-education-program/>.

We understand that EDS is not widely understood or studied, but it is real and the pain can be truly devastating. My kids deserve to have a doctor who says they believe them and will try their best to help. The thing about humans is they don't always believe what they can't see. And you can't see pain on a body or on an X-ray. So, hearing someone say that they believe you and they want to help goes a long way. We know from our experience that when one provider can validate this pain and share it with another provider, the integrity of care improves.

We hope, wish, and pray for the advancement of research in the area of chronic pain as well as EDS. Why does pain become chronic? Why do my kids become sensitized to pain? Is there a gene for h-EDS? Is there a therapy that could help reduce or eliminate chronic pain? Is there a better approach to chronic pain management? Bottom line, what more can be discovered to help my children feel better?

My kids are amazing. They are smart, mature, capable, and wise now beyond their years. They will do amazing things with their compassion, wisdom, and intelligence. I have no doubt that their futures are bright. Imagine what more they could do if they were freed of the weights of h-EDS.