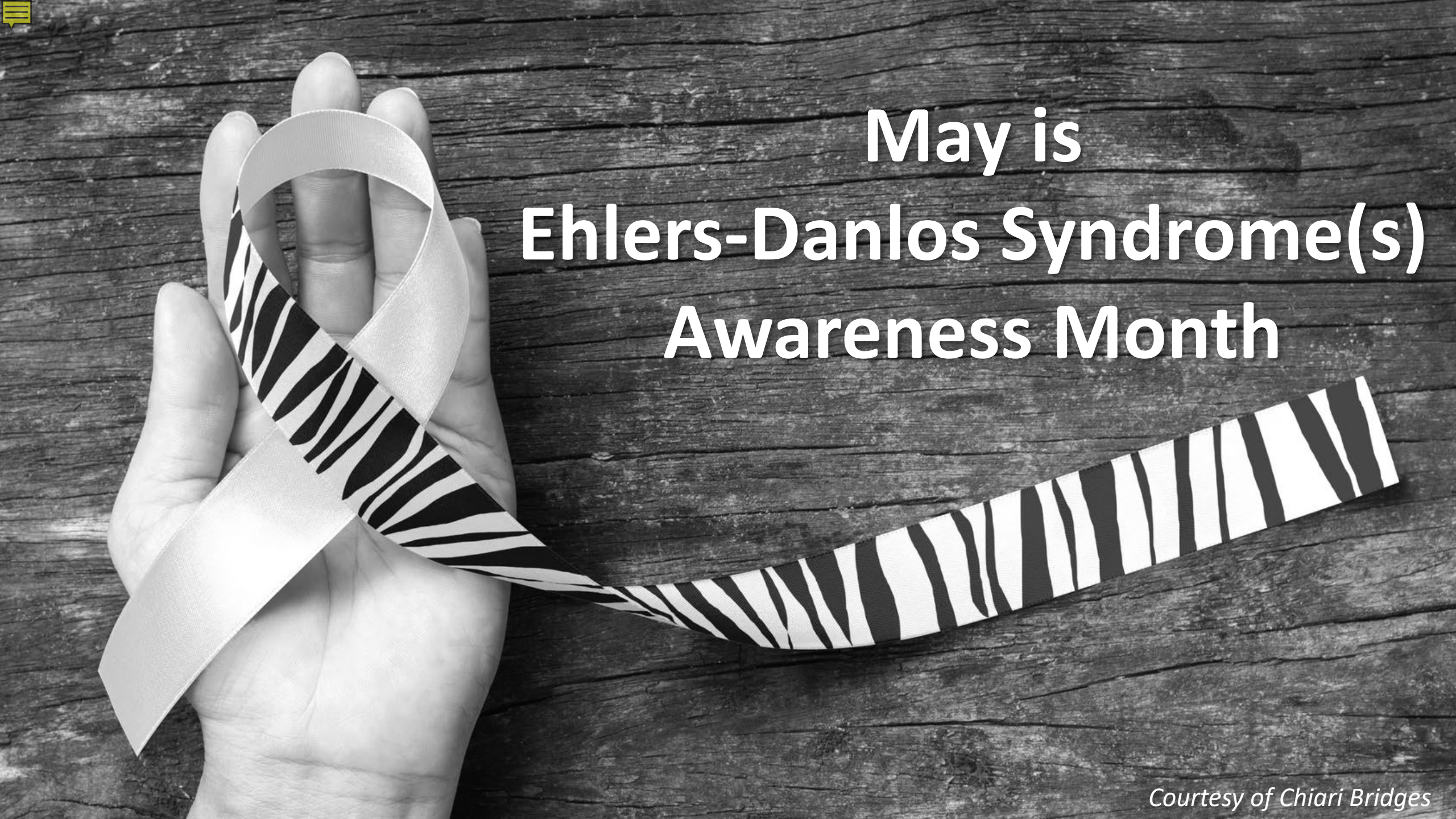

May is
Ehlers-Danlos Syndrome(s)
Awareness Month
Courtesy of Chiari Bridges

Courtesy of Chiari Bridges

EDS Awareness Fact #1

Ehlers-Danlos Syndromes (EDS) are a group of hereditary connective tissue disorders involving defects in our collagen. Collagen is a structural protein that is often described as the "fibrous glue" that holds our bodies together. It is the most abundant protein in the human body, providing structure to our organs, muscles, bones, skin, blood vessels, and connective tissues.

EDS Awareness Fact #2

Courtesy of Chiari Bridges

There are thirteen known subtypes of Ehlers-Danlos syndromes (EDS). While they tend to share certain general characteristics: hypermobility, skin laxity, and tissue fragility, each subtype has its own set of symptoms and criteria that can help narrow down the genetic testing needed.

Courtesy of Chiari Bridges

EDS Awareness Fact #3

Studies have shown that Ehlers-Danlos syndromes (EDS) can have many neurological and spinal manifestations, to include:

- Intracranial Hypertension (IH/IIH)
- Chiari Malformations (CM)
- Atlantoaxial & Craniocervical Instability (AAI/CCI)
- Segmental Kyphosis & Instability
- Tethered Cord Syndrome (TCS)
- Spontaneous CSF Leaks causing Intracranial Hypotension
- Dystonia & Other Movement Disorders
- Neuromuscular Complications

EDS Awareness Fact #4

Courtesy of Chiari Bridges

Ehlers-Danlos syndromes (EDS) are known to cause CSF Leaks (even spontaneous leaks). Much in the same way that it causes skin fragility, it makes the dura thin and fragile as well. In fact, CSF Leak expert, Dr. Schievink estimates that "slightly less than 100 percent of patients with a spontaneous CSF leak have an underlying connective tissue disorder."

Courtesy of Chiari Bridges

EDS Awareness Fact #5

In medical school, students are taught to not neglect the obvious, with the analogy of, "when you hear hoofbeats, think horses, not zebras." This philosophy poses a problem for those with Ehlers-Danlos syndromes, as it's not that our condition is rare, but that it's rare that doctors know anything about it; so we often go years or decades pursuing answers to our numerous symptoms.

EDS Awareness Fact #6

Courtesy of Chiari Bridges

It may seem counter-intuitive, but many hypermobile people also suffer from stiffness and tightness in their joints and muscles. Muscle stiffness and tightness is caused by the extra work muscles have to do to try and keep a hypermobile person's joints stable. We affectionately refer to them as Stiff Zebras!

Courtesy of Chiari Bridges

EDS Awareness Fact #7

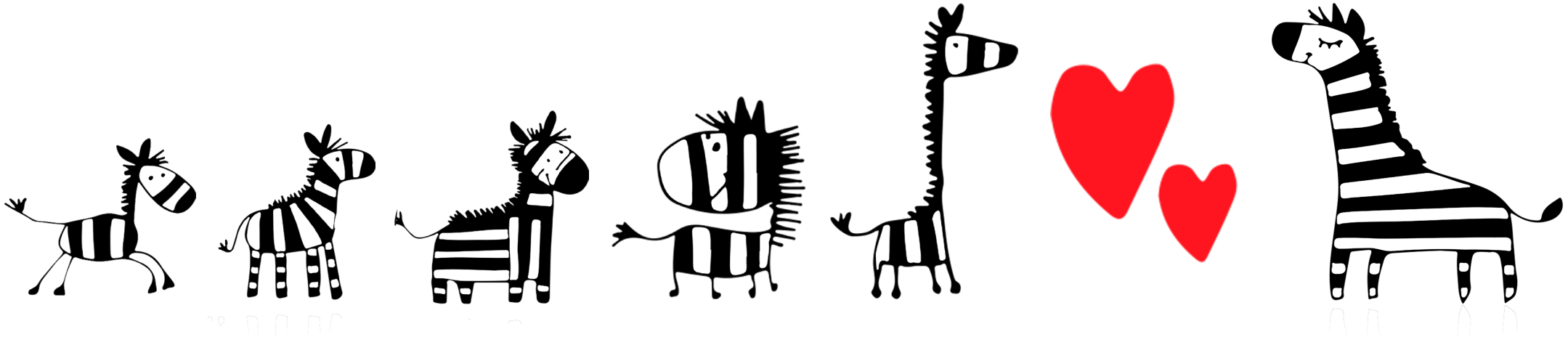
Chiari Malformation Type 1 (CM1) has been reported as a comorbid condition in hypermobile EDS (hEDS). The precise incidence of the CM1 and EDS association is unknown, but the female to male ratio is higher in the CM1 and EDS subgroup (9:1) than in the general CM1 population alone (3:1).

-Dr. Fraser Henderson

EDS Awareness Fact #8

Courtesy of Chiari Bridges

Ehlers-Danlos Syndromes (EDS) are known to be hereditary. Some are known to be autosomal recessive (where both parents must have the mutated gene to pass it down to a child) and others are autosomal dominant (where only one parent need have the mutated gene to pass it down to the child). It's important to remember that you can't control what you pass down to your children genetically, so it should NEVER be allowed to become a blame game!



EDS Awareness Fact #9

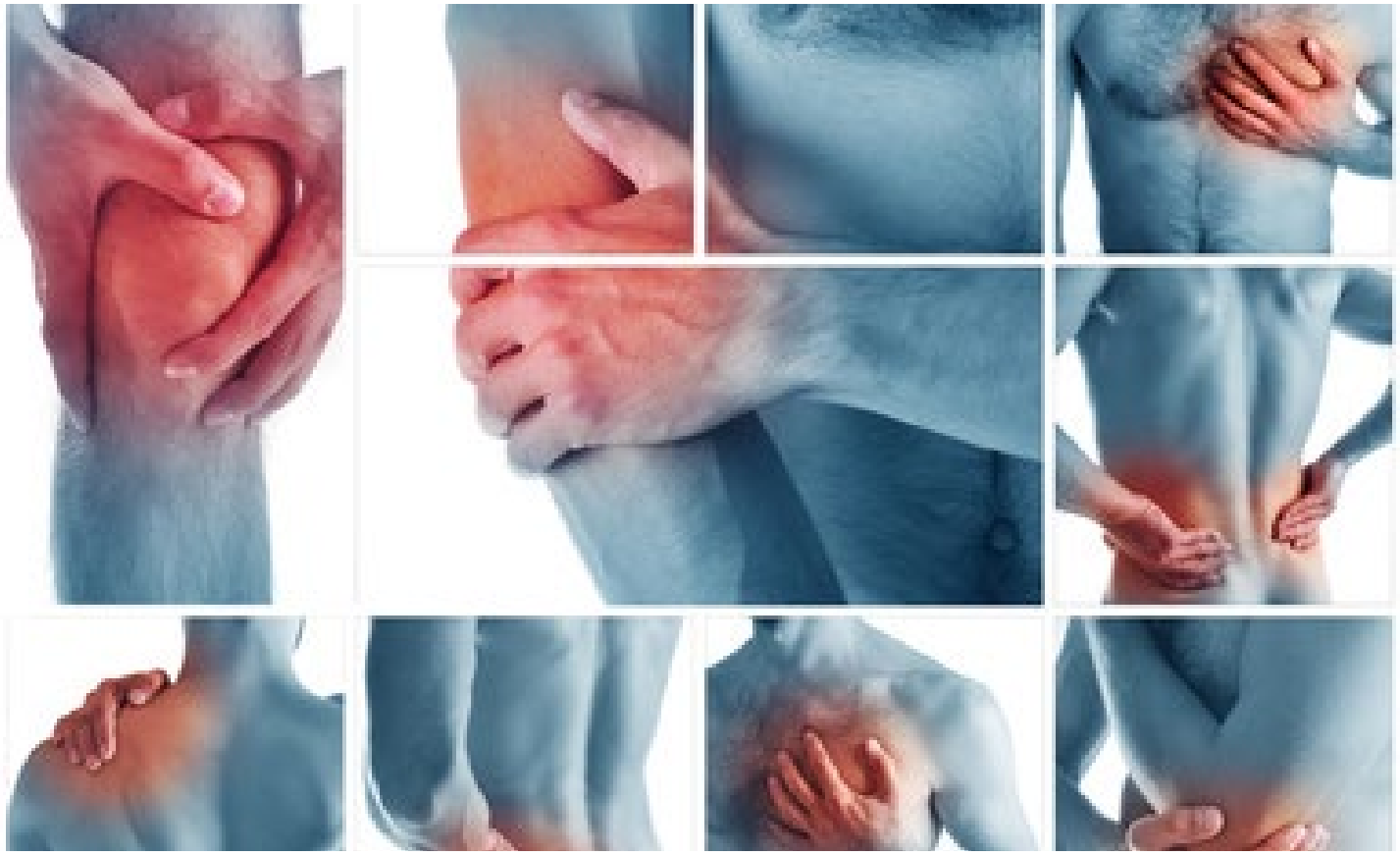
Courtesy of Chiari Bridges

Studies show that those with Ehlers-Danlos Syndromes (EDS) have a higher prevalence of symptomatic Obstructive Sleep Apnea (32% vs 6% in the general population). For this reason, we recommended that EVERYONE with EDS is tested for Sleep Apnea and use their CPAP RELIGIOUSLY!

EDS Awareness Fact #10

Courtesy of Chiari Bridges

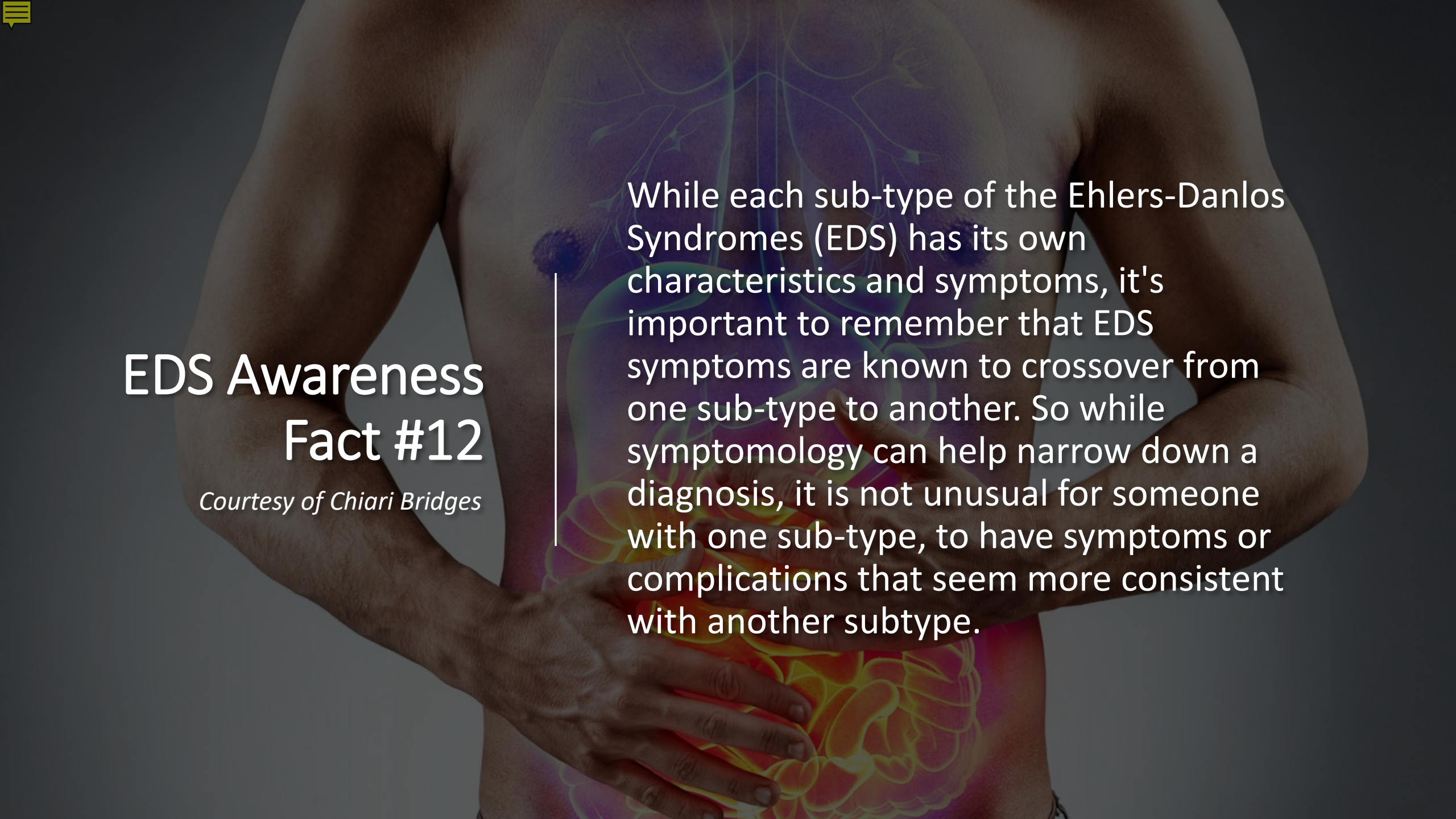
Recent statistics are proving Ehlers-Danlos to be far more common than initially believed (just a few years ago the prevalence was believed to be 1 in 5,000 to 1 in 20,000 depending on the subtype). The most recent numbers published on its prevalence is 1 in 2,500 to 1 in 5,000 people depending on the subtype (1.5-3 million people worldwide).



EDS Awareness Fact #11

Courtesy of Chiari Bridges

There are no known cures for Ehlers-Danlos syndromes (EDS). There is no way to change/repair our mutated collagen. We deal with one symptom at a time as our bodies try to interact with our faulty collagen as if it's normal. We typically have a multitude of medical specialists (for each manifestation) and a box of braces to try and help keep our joints in place.



EDS Awareness Fact #12

Courtesy of Chiari Bridges

While each sub-type of the Ehlers-Danlos Syndromes (EDS) has its own characteristics and symptoms, it's important to remember that EDS symptoms are known to crossover from one sub-type to another. So while symptomology can help narrow down a diagnosis, it is not unusual for someone with one sub-type, to have symptoms or complications that seem more consistent with another subtype.

EDS Awareness Fact #13

Ehlers-Danlos Syndromes (EDS) involve a mutation in our collagen. Collagen is often described as the "cellular glue" that holds everything in our bodies together. Going with that analogy, it is as if what should be Gorilla Glue, was replaced with watered down Elmer's Glue. It just doesn't have the consistency to hold things together as we need it to.

Courtesy of Chiari Bridges

EDS Awareness Fact #14

Courtesy of Chiari Bridges

The connection between "discopathy and early degenerative spondylotic disease in Ehlers-Danlos Hypermobility Type (hEDS) and Classical Type (cEDS), is well established. EDS is characterized by segmental instability, kyphosis, and scoliosis."

Courtesy of Chiari Bridges

EDS Awareness Fact #15

Most specialists recommend an upright MRI for patients that have a problem at the craniocervical junction and EDS. This allows them to see precisely how bad things shift and how low the tonsils go when gravity is factored into the equation while upright. Despite their well-documented recommendations, we still find ourselves having to fight to get this important testing done.

EDS Awareness Fact #16

Courtesy of Chiari Bridges

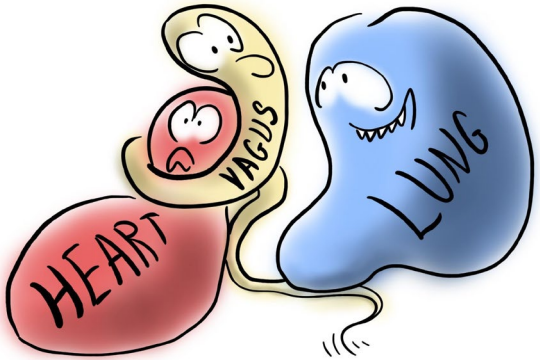
Studies have shown that "a high prevalence of patients with hereditary disorders of connective tissue in their retrospective series of CM post-decompression failures, needed further intervention, including craniocervical fusion and/or tethered cord release, suggesting that 'EDS and other disorders of connective tissue should not be overlooked in CM.'"

Some of these "disorders of connective tissue" are pathological to Chiari Malformations (CM), which is why we recommend testing for all of them before decompression!

Courtesy of Chiari Bridges

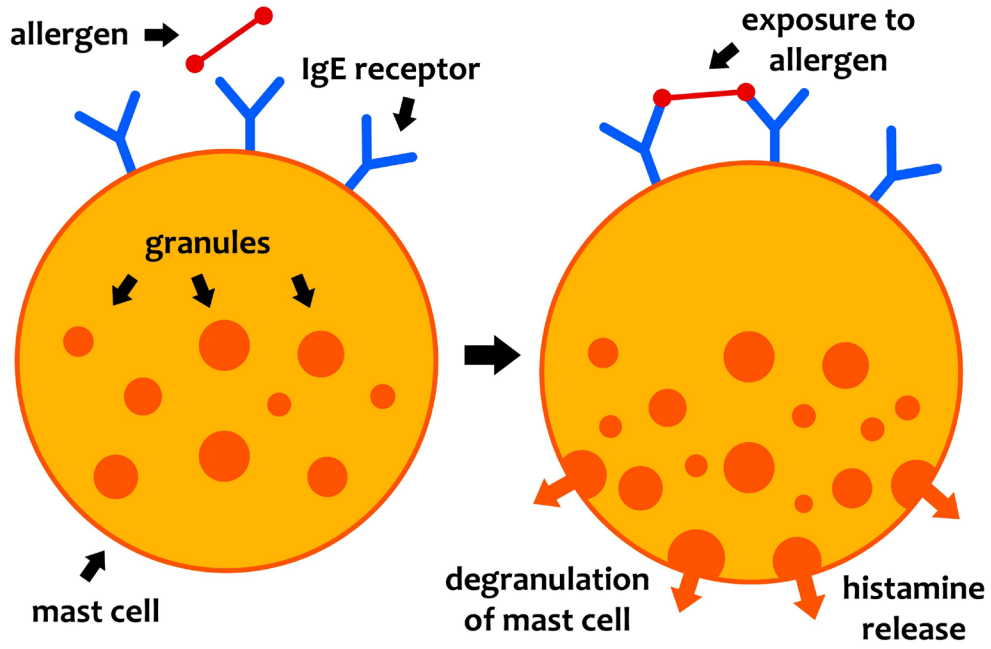
EDS Awareness Fact #17

Ehlers-Danlos syndromes (EDS) tend to cause problems at the cervicomedullary junction, often compressing the medulla (lower part of the brainstem) and Vagus Nerve. This often leads to damage of the Autonomic Nervous System (ANS), which controls your blood pressure, heart rate, respiration, swallowing, body temperature, digestive system, and other symptoms of dysautonomia.



EDS Awareness Fact #18

Courtesy of Chiari Bridges



Mast Cell Activation Syndrome is common amongst those with Ehlers-Danlos syndromes (EDS). Even if you've tested negative for allergies, you still might be having periodic increases in histamine and tryptase that manifest with reactions in your skin, gastrointestinal tract, cardiovascular system, and/or respiratory system.

Courtesy of Chiari Bridges

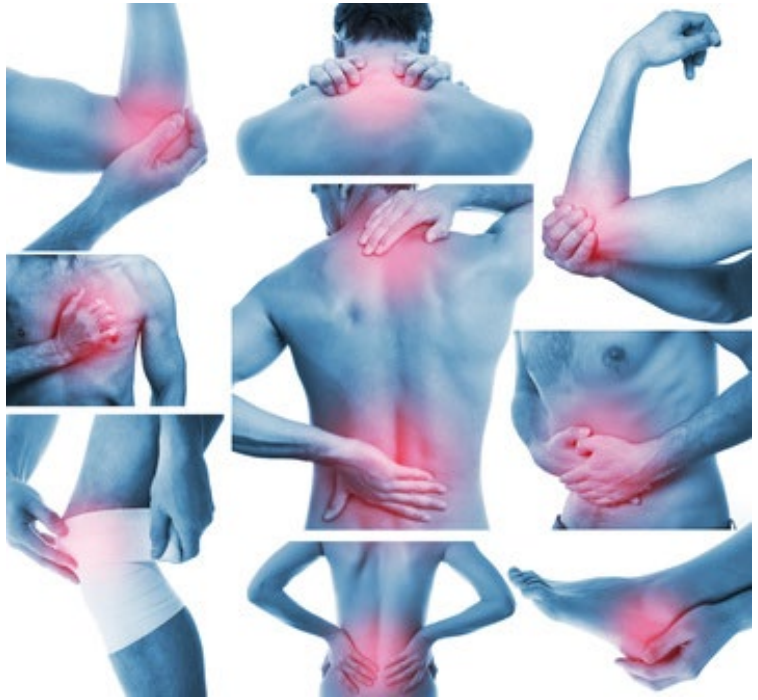
EDS Awareness Fact #19

Some specialists now believe that 50-60% of those with fibromyalgia have Ehlers-Danlos Syndrome (EDS) or at least Joint Hypermobility Syndrome (JHS). If this is true, EDS is far more common than originally thought.

EDS Awareness Fact #20

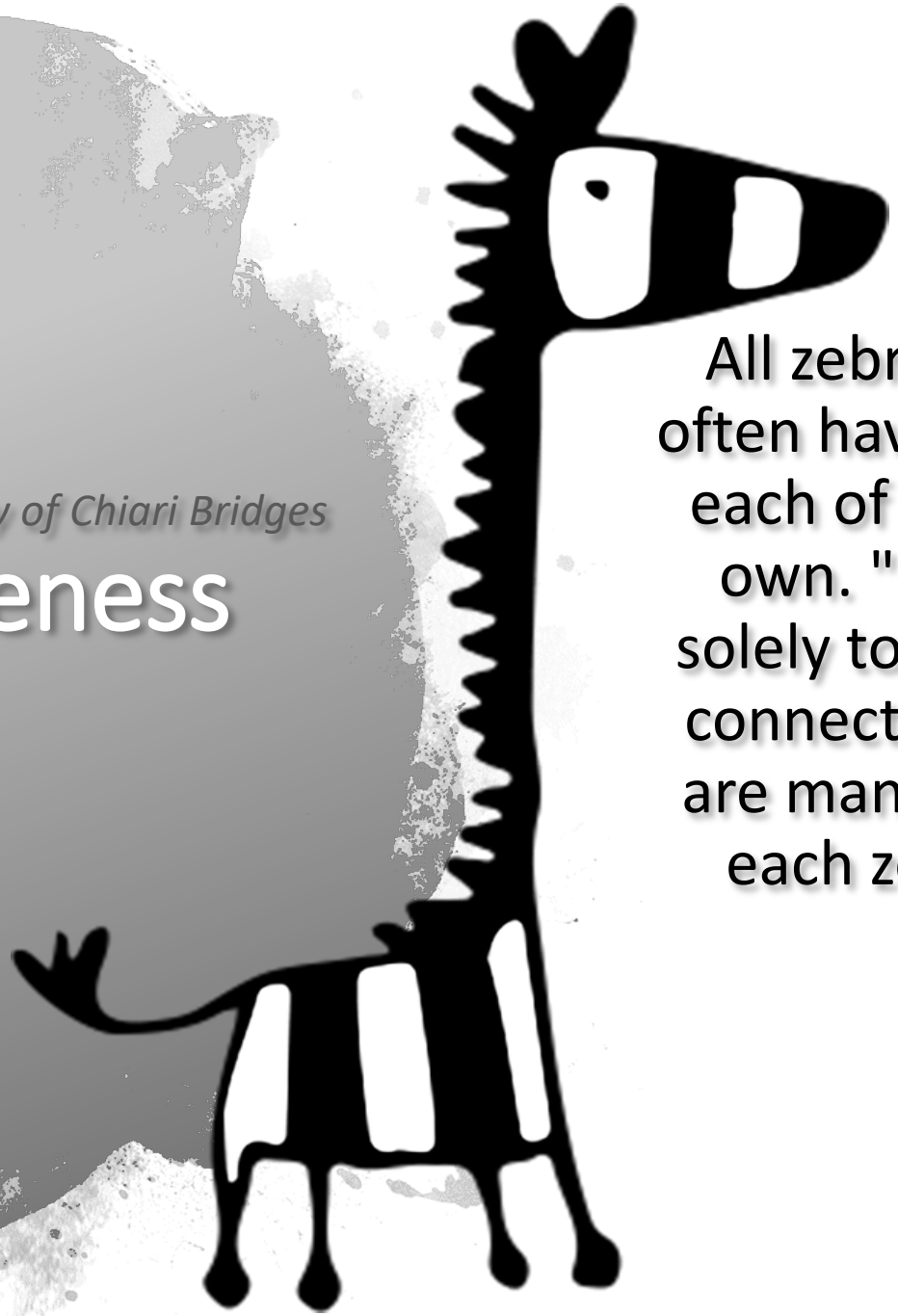
Courtesy of Chiari Bridges

Ehlers-Danlos Syndromes (EDS) are known to involve a mutation in our body's collagen, but it's not as simple as it sounds. EDS doesn't just affect our skin and joints, but everything in our bodies that was designed to interact with that collagen. That is why we tend to have so many problems throughout our bodies that you would think couldn't be related. It really is our connective tissues that connect the issues.



Courtesy of Chiari Bridges

EDS Awareness Fact #21



All zebras aren't the same! While we often have many symptoms in common, each of us fights a battle uniquely our own. "Diagnostic criteria are meant solely to distinguish an EDS from other connective tissue disorders, and there are many more possible symptoms for each zebra than there are criteria."

EDS Awareness Fact #22

Courtesy of Chiari Bridges

Collagen is the "single most abundant protein in the animal kingdom." In the human body, it comprises approximately one-third of the total protein, found in skin, cartilage, muscles, bones, tendons, ligaments, intervertebral discs, blood vessels, organs, gums, teeth, eyes, etc.

loose connective tissue

dense connective tissue

adipose tissue

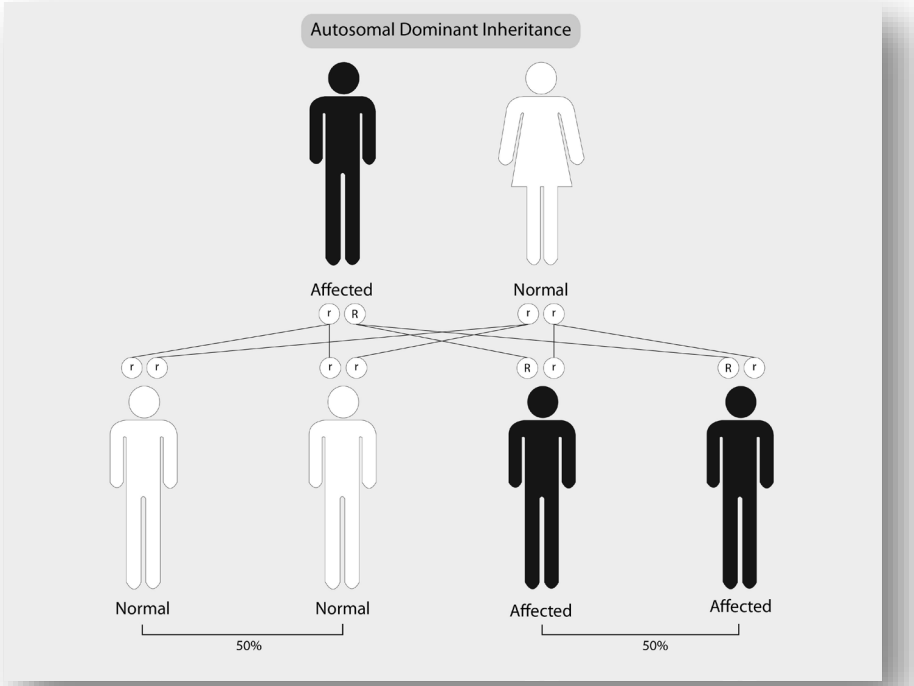
EDS Awareness Fact #23

Our Intervertebral Discs are comprised of collagen, therefore the discs of the Ehlers-Danlos Syndromes (EDS) patient, tend to dehydrate, thin, bulge, slip, and herniate into the spinal cord. The average person might have 1-2 disc problems in their lifetime, EDS patients tend to have many discs bulging and herniated at any given time, often in different levels of the spine. This condition is known as Degenerative Disc Disease (DDD).

Courtesy of Chiari Bridges

EDS Awareness Fact #24

Courtesy of Chiari Bridges



Each EDS subtype is a distinct hereditary disorder. The only time more than one subtype will be seen in a family is if both are genetically inherited from one or both parents. When more than one mutation is genetically confirmed, the most serious subtype will generally be the diagnosis given. It is important to remember that all EDS symptoms are known to cross the subtype lines, so it's quite common to have symptoms from multiple subtypes, but only have one inherited mutation.

EDS Awareness Fact #25

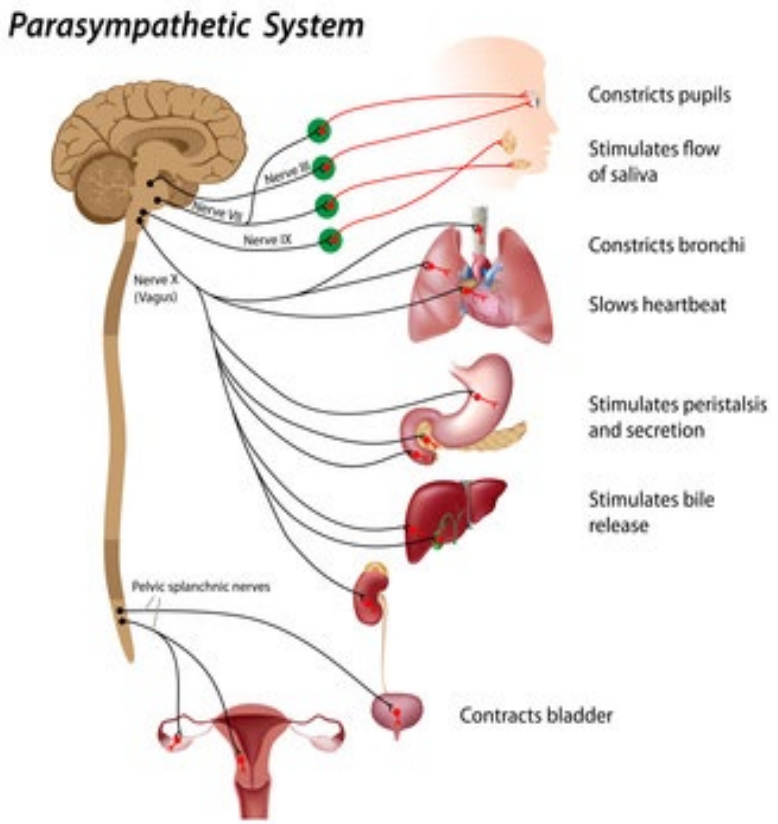
Courtesy of Chiari Bridges

PRIMARY SIGNS OF HYPERMOBILITY

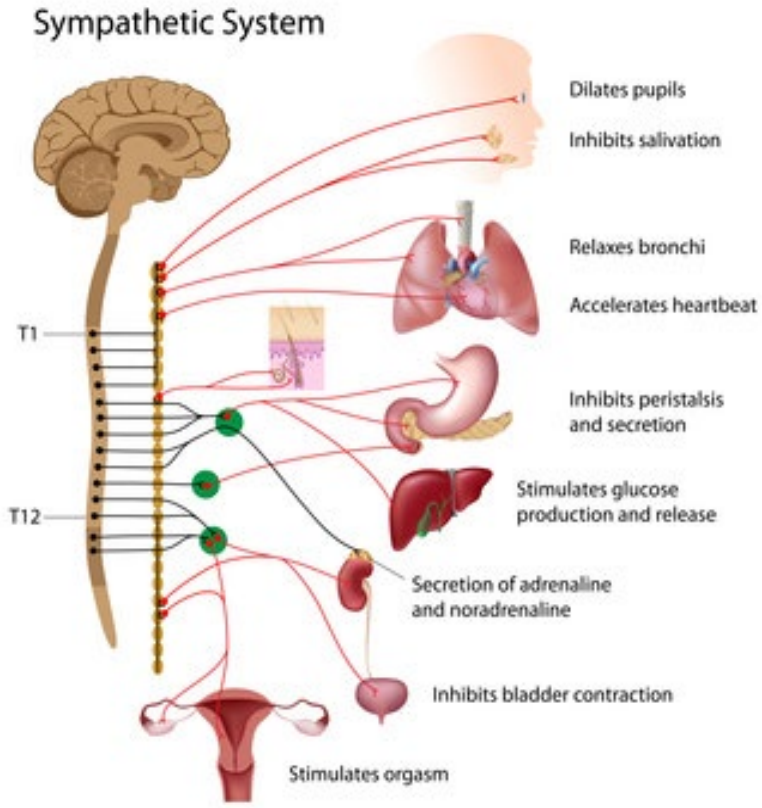
Clicking/Popping of Joints, Neck and/or Jaw (TMJ)
Rolling/Twisting of Ankles
Joint Pain
Neck Pain
Hyperextension of Joints
Tendonitis (Tennis/Golfer's Elbow, Achilles Tendonitis)
Bursitis
Trigger Finger
Multidirectional Instability of the Shoulder
Recurring Sports Injuries
Frequent Dislocations and/or Subluxations
Poor Proprioception
Muscle Stiffness/Tightness
Scoliosis
Other Musculoskeletal Problems

SECONDARY SIGNS OF HYPERMOBILITY

DYSAUTONOMIA



Dysautonomia is a dysfunction of the Autonomic Nervous System (ANS), so it includes everything in your body that happens automatically (without cognitive thought): Tachycardia (rapid pulse); POTS (Postural Orthostatic Tachycardia Syndrome); MCAS (Mast Cell Activation Syndrome); Inability to control basic body functions, such as body temperature, breathing rate, blood pressure, digestion, bowel/bladder function.



EDS Awareness Fact #26

Courtesy of Chiari Bridges

Courtesy of Chiari Bridges

EDS Awareness Fact #27

SECONDARY SIGNS OF EHLERS-DANLOS

GASTROINTESTINAL PROBLEMS

Irritable Bowel Syndrome (IBS); Food sensitivities; Malabsorption; Dysmotility; Esophageal sensitivity/spasms; Esophageal Dysphagia; GERD (Gastroesophageal Reflux Disease); Gastroparesis; Dumping Syndrome and Functional Dyspepsia. Symptoms can range from reflux, diarrhea, constipation, gas, bloating, and vomiting. Additional GI problems associated with EDS include Hiatal Hernias, Organ prolapse & Abdominal aortic aneurysm.

EDS Awareness Fact #28

Courtesy of Chiari Bridges

SECONDARY SIGNS OF EHLERS-DANLOS
OB/GYN

Irregular, Heavy, Painful, Menstruation
Increased Joint Pain Before/During Menstruation/Pregnancy
High Rate of Miscarriages
Preterm Labor
Placenta Previa
Pelvic Floor Instability
Uterine Prolapse
Urinary Incontinence
CSF Leaks From Epidurals

Additionally, we tend to have hormonal problems for differing reasons, these problems should be considered when hormonal therapies (birth control) are introduced.

Courtesy of Chiari Bridges

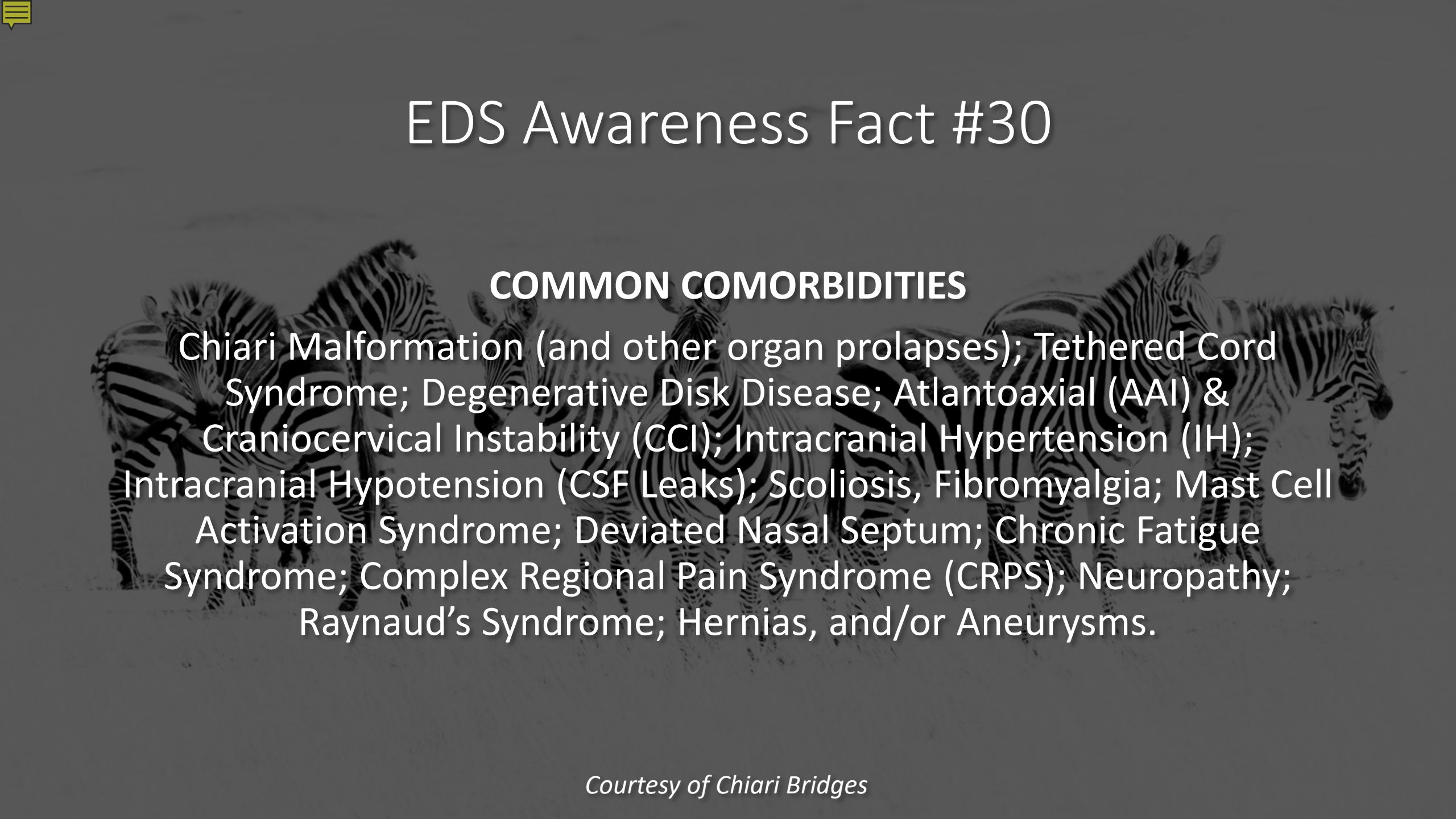
EDS Awareness Fact #29

SECONDARY SIGNS OF EHLERS-DANLOS

SKIN & BONES

Easy Bruising; Stretch Marks; Varicose Veins; Slow Wound Healing; Abnormal Scarring; Elastic/Stretchy Skin; (esp. under upper arms & upper/inner thighs); and/or Translucent Skin (esp. chest/breasts & bottom of arms); Osteopenia; Low Bone Density; Low Bone Mass; Weakened Bone Structure and/or Flat Feet.

EDS Awareness Fact #30



COMMON COMORBIDITIES

Chiari Malformation (and other organ prolapses); Tethered Cord Syndrome; Degenerative Disk Disease; Atlantoaxial (AAI) & Craniocervical Instability (CCI); Intracranial Hypertension (IH); Intracranial Hypotension (CSF Leaks); Scoliosis, Fibromyalgia; Mast Cell Activation Syndrome; Deviated Nasal Septum; Chronic Fatigue Syndrome; Complex Regional Pain Syndrome (CRPS); Neuropathy; Raynaud's Syndrome; Hernias, and/or Aneurysms.

Courtesy of Chiari Bridges

GETTING DIAGNOSED

Ehlers- Danlos Syndromes (EDS) aren't hard to diagnose. What is hard is finding a doctor that knows about the symptoms enough to refer you to a geneticist. Once referred, a geneticist will do a clinical evaluation for hypermobility (known as your Beighton Score). Next, they will evaluate you for major/minor criteria (called The Brighton Criteria). Once those are both assessed, they will determine which genetic tests should be done for your case. All sub-types have known mutations that can be tested for except hEDS.

EDS Awareness Fact #31

Courtesy of Chiari Bridges

www.ChiariBridges.org