

Chiropractic Management of patient with Ehlers-Danlos syndrome, Type III, Hypermobility type

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Ehlers-Danlos Syndrome

EDS is a group of inherited disorders that affect connective tissues — primarily skin, joints and blood vessel walls

Wide variety of symptoms throughout different body systems; each unique in presentation to the individual.

The variety of symptoms and presentations proves challenging to the medical community in diagnosing and treating patients; many of whom are misdiagnosed and suffer as a result.

Ehlers-Danlos Syndrome Facts

- Ehlers-Danlos Syndrome (EDS) is classified as a rare disorder affecting 1:5000 people.
- Based upon recent research, the prevalence of EDS exceeds this number and could be as high as 1:100 to 1:200 people (Collins, 2015; Nielsen, 2013) or even higher

How do you get EDS?

- Most cases of EDS seem to follow the autosomal dominant pattern of inheritance.
- In Autosomal Dominant inheritance, only one gene from the parent needs to be functioning improperly for an individual to be affected.
- When an affected person has children, there is a 50% chance with each pregnancy that the parent will pass on the changed gene to his/her children.
- EDS types that are inherited in the autosomal dominant fashion include the Classical Type , Hypermobility Type, Vascular (VEDS) Type
- Types of EDS which are inherited as autosomal recessive are the Kyphoscoliosis Type and the Dermatosparaxis Type, but are uncommonly seen types of EDS.

Ehlers-Danlos Syndrome History

- The probable first description of EDS was by Hippocrates in 400 B.C.
- In 1657 a Dutch surgeon noted a case history of a boy with hyperextensible skin.
- The first association of hypermobile joints to skin was published in 1892 in Moscow by A.N. Chernogubov.
- Edvard Ehlers in 1901 defined it as a distinct disorder in a case history that included lax joints, hyperextensible skin and a tendency to bruise.
- In 1908, Henri-Alexandre Danlos published a second case history.
- In 1936, Frederick Parkes-Weber suggested the disorder be named Ehlers-Danlos syndrome.

Ehlers-Danlos Syndrome Facts

The first classification of types of EDS originated in the late 1960s and was formalized in 1988;

The Berlin nosology used a numbering for seven EDS types, some with lettered subtypes. Experience proved it to be too complicated, as the classification did not discriminate adequately between the types or between the Ehlers-Danlos syndromes and other related conditions.

In 1997, the revised nosology was written at the Villefranche International Conference that redefined EDS into the current six named types.

Ehlers-Danlos Syndrome Facts

- There are six major types of EDS. The different types of EDS are classified according to their manifestations of signs and symptoms.
- Classical Type
- Vascular Type
- Hypermobility Type
- Other Types

Ehlers-Danlos Syndrome Facts

Each type of EDS is a distinct disorder that "runs true" in a family.

Ehlers-Danlos Syndrome Facts

Classical Type

- Marked skin hyperextensibility (stretchy) with widened atrophic scars and joint hypermobility are found in the Classical Type of EDS.
- The skin manifestations range in severity from mild to severe. The skin is smooth and velvety along with evidence of fragility and a tendency to bruise easily

Ehlers-Danlos Syndrome Facts

Vascular Type

- Generally regarded as the most serious form of EDS due to the possibility of arterial or organ rupture.
- The skin is usually thin and translucent with veins being seen through the skin, which is most apparent over the chest and abdomen.
- Some have facial characteristics include large eyes, thin nose, lobeless ears, short stature and thin scalp hair. Also evident is a decrease in subcutaneous tissue, particularly in the face and extremities.

Ehlers-Danlos Syndrome Facts

Hypermobility Type

- Joint hypermobility is the dominant clinical manifestation.
- Affects large (elbows, knees) and small (fingers, toes) joints is evident in the Hypermobility Type.
- Recurring joint subluxations and dislocations are common: the shoulder, patella and temporomandibular joint dislocate frequently.
- The skin involvement (smooth velvety skin with or without hyperextensibility) as well as bruising tendencies in the Hypermobility Type are present but quite variable in severity.

The Beighton Criteria

Journal of Rheumatology (2000; 27: 1777-1779)

Brighton Diagnostic criteria for the Joint Hypermobility Syndrome (JHS)

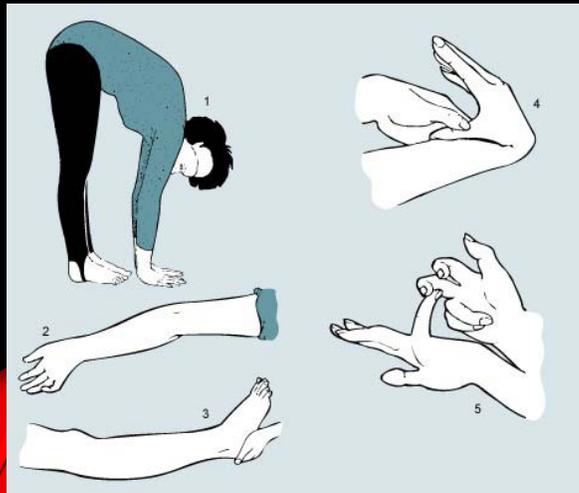
Joint hypermobility, defined as a more-than-normal range of movement (ROM) in a joint, is either localized (increased ROM of a single joint) or generalized.

Joint hypermobility depends on age, gender, family and ethnic background. A score of 5/9 or greater defines hypermobility.

The total score is obtained by results of 5 exercises:

Palms to floor, elbow hyperextension, knee hyperextension, apposition of thumb to forearm, pinkie past 90 degrees

Beighton Criteria: 5 of 9



- 1) Forward flexion of the trunk with knees fully extended so that the palms of the hand rest flat* on the floor – one point
- 2) Hyperextension of the elbows beyond 10 degrees* – one point for each elbow
- 3) Hyperextension of the knees beyond 10 degrees* – one point for each knee
- 4) Passive apposition of the thumbs to the flexor aspect of the forearm* – one point for each hand
- 5) Passive dorsiflexion of the little fingers beyond 90 degrees* – one point for each hand

Ehlers-Danlos Syndrome Management

While there is no cure for EDS, there is treatment for symptoms, and there are preventative measures that are helpful for most.

Our findings suggest that pain is a very common and severe symptom in this group of EDS patients. It is related to dislocations, sleep disturbances and moderate-to-severe impairment in daily functioning. Therefore, treatment of pain should be a prominent aspect of clinical management of EDS."

[Nicol C. Voermans, MD, Hans Knoop, PhD, Gijs Bleijenberg, PhD, and Baziel G. van Engelen, MD, PhD. "Pain in Ehlers-Danlos Syndrome Is Common, Severe, and Associated with Functional Impairment." *J Pain Symptom Manage*. 2010 Sep;40(3):370-8.]

MULTI-MODAL MANAGEMENT FOR PAIN IN EDS

ANALGESICS (if needed)

- *May be ineffective as single agents or require excessive doses. Short-acting opioids Muscle relaxants*

PHYSICAL THERAPY

- Reconditioning Exercise (*gradual toning for stability, not strength*)
- The key is slow improvement in muscle tone, **not strength**. Tone is the degree of muscle contraction at rest; strength is voluntary force that can be invoked at will. Build tone with non-resistance exercise that gradually increases repetitions. Avoid hyperextension, resistance and impact. Elastic bands may make things worse because they combine increased resistance with joint hyperextension. Toning is a life-long strategy; if exercises are stopped, pain usually recurs. (*Howard Levy, MD, PhD*)
- Flare-up management by:
 - Distraction techniques; Trigger-point/myofascial therapy Heat/cold application Ultrasonic/electrical stimulation

PSYCHOLOGICAL THERAPY

- Relaxation, Stress management (*incl. Mindfulness-Based Stress Reduction*)
Group therapy, Cognitive restructuring (*incl. hypnosis*)

MULTI-MODAL MANAGEMENT FOR PAIN IN EDS

PSYCHIATRIC THERAPY

- Medication for mood/pain Individual counseling

SLEEP THERAPY

- *Sleep disturbance is common in EDS (contributes to poor pain recovery; incl. apnea)*
- Tricyclic antidepressants (TCAs)

COMPLEMENTARY/ALTERNATIVE

- Meditation Acupuncture Massage Yoga

MAINTAIN GENERAL HEALTH

- Avoid weight extremes & smoking
- Quit smoking (pain, depression and addiction are all modulated by the same nicotinic and muscarinic receptors). Maintain normal weight; obesity or thinness seem to present problems. (*Mark Lavalley, MD, CSCS, FACSM*)

MULTI-MODAL MANAGEMENT FOR PAIN IN EDS

CHRONIC PAIN

- *(constant & disturbing pain)*
- **MANAGE ACUTE EVENTS**
- *Treat acute events quickly and aggressively to defend against central neuronal plasticity ("learned" chronic pain)*

ANALGESICS FOR CHRONIC PAIN

- Use combinations of medicines on a schedule to keep pain under control
- *if needed*
Short-acting opioids (*breakthrough pain*)
- Long-acting opioids (*baseline pain*)

PREVENT RECURRING INJURY

- *Hypermobile joints and fragile connective tissues endure chronic acute injury*
- Bracing may be helpful (*maintain toning exercise*)

Orthopedic surgery should be avoided

- *Almost never provides long-term improvement*
Added risk of soft tissue fragility and poor wound healing, esp.

COX® DISTRACTION MANIPULATION

- High velocity Low amplitude thrust adjustments would not be well received by these patients
- EDS patients, especially Hypermobility type, show great improvement, with Cox® Technic:
 - Protocol 1: 3x5x4; for Radicular symptoms
 - Protocol 2: 3x10x1; for Non Radicular symptoms

CASE STUDY:

- 37 year old female
- Chronic Widespread pain: cervical, shoulders, thoracic, ribs, lumbar, hips, butt, calves, Intensity of a constant 4-6/10, with more severe breakout episodes
- She describes significant fatigue
- Feels/hears "cracking/popping" of her joints often
- Previously diagnosed with Fibromyalgia and Degenerative Arthritis
- She had past treatment with:
 - Medication
 - Massage
 - Physical Therapy
 - Exercise

CASE STUDY

ARROW
 Sports & Physical Therapy

Please use the following key to accurately mark the areas in which you feel the described sensations. Use the appropriate symbols and include all affected areas.

Dull Numb	//// ==	Stabbing/Cutting Tingling (Pins & Needles)	/// // // // =====	Burning Cramping	XXX SSS
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CHOOSE THE FACE THAT BEST DESCRIBES HOW YOU FEEL

0 No Hurt	2 Hurts Little Bit	4 Hurts Little More	6 Hurts Even More	8 Hurts Whole Lot	10 Hurts Worst

Using the scale of 0-100, with 0 = no pain and 100 = worst possible pain, please write the number indicating your present pain level in the box at the right:

50

Please indicate any changes in your condition in this space:

CASE STUDY

Vitals: Height: 5'3.5"

Weight: 172

BP: 117/84

Pulse: 21

Reflexes: +2/2 in Biceps, Triceps, Brachio, Patella, and Achilles

Dermatome/Skin Sensation: Within Normal Limits

Muscle Strengths: Upper and lower extremity: all graded +5/5

CASE STUDY

- Range Of Motion:
- Cervical
 - Flexion: 42/60
 - Extension: 36/75
 - Right Lateral Flexion: 22/25
 - Left Lateral Flexion: 20/25
 - Right Rotation: 68/80
 - Left Rotation: 60/80
- Lumbar
 - Flexion: 60/60
 - Extension: 22/25
 - Right Lateral Flexion: 22/25
 - Left Lateral Flexion: 22/25

CASE STUDY:

- BEIGHTON SCORE:
 - 8/9
 - Could not get palms to the floor
- She states that she used to be able to perform that move easily, as well as splits.
- I believe that adaptive shortening has taken over as a protective mechanism

CASE STUDY

- Cervical Orthopaedic Testing
 - Positive Kemps at C2-5
 - Locally Tender on Compression testing but not indicative of Discal complaint
 - Negative Distraction
 - Negative Soto Hall
 - Negative VAS
- Trigger points located in Suboccipitals, Cervical paraspinals, Upper Trapezius, Levator Scapulae, bilaterally, left greater than the right

CASE STUDY

- Thoracic Orthopaedic Testing
 - PA compression increased pain at T3-4
 - Positive Thoracic Kemps: T5-8, left greater than the right
 - Posterior Rib Compression increased pain at left T5
 - Trigger Points noted in thoracic paraspinals, and Rhomboids, left greater than right

CASE STUDY

- Lumbar Orthopaedic Testing:
 - Negative Minors Sign
 - Negative Slump
 - Positive Kemps-Localized pain at L3-5
 - Negative SLR, WLR, and Braggards
 - Positive FABRE- Each side created Hip and Back pain
 - Positive Hip Compression bilaterally-created hip pain
 - Trigger points noted in Lumbar Paraspinals, Quadratus Lumborum, Glut Complex, Gemelli, Obturator

CASE STUDY

- Treatment utilized:
 - High Voltage Galvanic coupled with Ultrasound to her Cervical and Lumbar regions (2 separate areas)
 - Manual Therapy: trigger point therapy and cross fiber to aforementioned trigger points
 - Cox® Decompressive Manipulation: Protocol 2: 3x10x1 (full spine)
 - Extrapinial Manipulation to hips/ankles
 - Therapeutic Exercise: Basic Spinal Stability and Core

CASE STUDY

- Coordinated Care with her Integrated Medical Physician: OB/GYN who converted practice
- Began anti-inflammatory diet
 - Lost 20 pounds
 - Gluten Free
 - Increased Whole foods
 - Reduced/Removed Processed foods

CASE STUDY

- Her integrative medical physician sent her to a Physical Therapist, for evaluation of pelvic floor trigger points
- At the appointment, this therapist decided to provide Long Lever, High Velocity, manipulation.
- Without proper explanation, the therapist forcibly twisted the patient, causing great pain and tissue injury
- I was made aware of this situation at my next visit. I wrote to both providers about the Cox® Technic as the treatment of choice. The MD felt sorry and apologized profusely to the patient.
- We are able to control the acute exacerbation quickly.

CASE STUDY

- After phone consultation with her integrative medical doctor, she was referred to Northeast Indiana Genetics, to confirm the diagnosis of Ehlers-Danlos Syndrome, Type 3.
- She is still under care.
- She is Congruent. She understands that there is no cure to her condition, but there is control over her symptoms.

CONCLUSION:

IF YOU CAN'T CONNECT THE TISSUE,
THINK CONNECTIVE TISSUE! (AUTHOR UNKNOWN)

EDS MAY BE MUCH MORE PREVALENT,
THAN CURRENTLY REPORTED

EDS RESPONDS WELL TO CHIROPRACTIC CARE,
UTILIZING COX® DISTRACTION MANIPULATION,
PHYSIOLOGICAL THERAPEUTICS, AND REHAB