

Dysautonomia in Hypermobile Ehlers-Danlos Syndrome (or EDS type III)

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Presados amigos:

Eu lamento muito não poder estar com vocês hoje, eu estou nesse momento num Congresso em Murcia, Espanha. Por isto eu os envio este Powerpoint. Eu espero que seja produtivo, e perdão por telo feito em Inglês.

Um abraço pra todos

Dr. Jaime Bravo

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None

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What is Dysautonomia

- It is a condition in which patients tire easily, after activity, after a big lunch or at the end of the day, they feel sleepy and usually feel “like their batteries have become discharged” and have no energy.
- Due to the chronic fatigue, and episodic sensation of weakness is that in many cases the wrong diagnosis of Depression, Fibromyalgia, Chronic Fatigue, Hypothyroidism or Hypoglycemic crisis is made.
- Family and friends usually see them as being lazy and unsociable, since they lack the energy to participate in social meetings or to interact with other people.

Dysautonomia in Hypermobile Ehlers-Danlos Syndrome (hEDS)

- It is a frequent condition that can cause poor quality of life and usually goes undiagnosed. It is particularly frequent in people with hEDS. These people may not even know that they have hypermobile joints, and physicians usually pay no attention to this condition and do not make the diagnosis.
- hEDS can produce musculoskeletal complaints (joint pain, recurrent tendinitis, joint sub-luxations, etc) and problems derived from weak tissues (Dysautonomia, hernias, varicose veins, uterine and mitral valve prolapse, myopia, spinal disc disease, early osteoarthritis and early osteoporosis).
- Note: I prefer the term **EDS-III** rather than Hypermobility type EDS, since in my study of 2300 pts, 51% had a Beighton score of 3/9 or less.

Symptoms of Dysautonomia

- Chronic fatigue, with somnolence.
- Pre-syncope or syncope.
- Low blood pressure.
- Cold intolerance.
- Poor memory, concentration and disorientation.

Why is Dysautonomia associated with EDS-III

- The reason is the weak collagen of the vein wall in the lower extremities. In a patient with normal veins the blood progresses up to the brain, but if the vein dilates, due to poor collagen, then the blood pools in the lower extremities and has more difficulty to get back to the heart and brain, causing brain hypoxia , and the symptoms of Dysautonomia.

Symptoms of Dysautonomia

Sudden changes, such as getting out of bed rapidly, or standing in line or while walking slowly in shopping malls or supermarkets, or after eating a big meal, specially with alcohol, the blood return to the heart diminishes, and as a result, the blood pressure drops suddenly and the brain oxygenation decreases.

When someone stands up suddenly, 300 to 800 ml of blood stay in the veins of the abdomen or lower extremities and in patients with Dysautonomia the body is incapable to compensate rapidly and symptoms appear (dizziness, headaches, severe fatigue, syncope).

Causes of Dysautonomia in EDS-III

A.- Autonomic Nervous System dysfunction, characterized by:

Orthostatic hypotension or
Postural orthostatic tachycardia (POTS).

Symptoms: Palpitations, lightheadiness, tiredness, dizziness, somnolence, poor thermostat regulation, chronic fatigue, pre-syncope or syncope.

B.- Increased venous pool in the lower extremities, derived from collagen weakness of the venous wall.

Since most physicians erroneously believe that a low blood pressure is beneficial, Dysautonomia frequently goes undiagnosed. Thus, these patients usually have a poor quality of life.

In EDS-III the veins in the legs and abdomen act as a blood pool

A good analogy is what happens to fluids inside half-filled bottles. When moved from a horizontal to a vertical position, the fluids remain close to the bottom of the bottle. Something similar occurs to people with Dysautonomia, reason for which, when they stand up abruptly, they can develop dizziness and even syncope, due to lack of oxygen to the brain.



Causes that aggravate Dysautonomia

- Dehydration, due to:
 - In hot weather
 - Fever
 - Vomiting and diarrhea
 - Diuretics
- High altitude, in cities like Mexico City, and in mountain climbing.
- Standing at church or in a line for too long, or when getting up suddenly from bed or from a sitting position.
- Phobic reaction to close surroundings (Claustrophobia).
- The sight of blood, especially if from a relative or close friend.
- Frightening situations.
- Severe pain or pain associated with anxiety, such as when getting an injection.

Causes that aggravate Dysautonomia (Cont.)

- Sexual relations.
- Menstrual periods.
- Pregnancy.
- Prolonged hot bath (bathtub, shower, sauna, jacuzzi) or with Bikram yoga, which is done at 42° C.

Causes that aggravate Dysautonomia (Cont.)

- With a strong emotion or with nervousness, such as when taking an exam.
- Acute anemia.
- Walking slowly, like when walking in Malls or at the super market.
- With certain BP medicines that can produce orthostatic hypotension, as a secondary effect.
- Standing for a long period of time, without moving.
- After a big meal or excess of alcohol.

Dysautonomia in EDS-III in a Chilean Study

In our study of 2300 patients

In the group younger than 30 y/o (382 pts):

Dysautonomia was present in:	Males	60.8%
	Females	84.0%

Ref. .-Bravo JF. Presented as a Poster. First International Symposium on Ehlers-Danlos.
Ghent. Belgium. Sept. 2012

Treatment of Dysautonomia

A.- General recommendations:

- Avoid standing or sitting without moving arms or legs.
- Avoid walking slowly in shopping malls or supermarkets, for prolonged periods of time.
- When sitting in a bus or an airplane, it is necessary to move the knees and ankles frequently and to get up and walk.
- After a big meal or heavy drinking it is necessary to lie down for 15 minutes or more. Do this also, when noticing early signs of Dysautonomia.

Treatment of Dysautonomia (Cont.)

- We recommend drinking liquids until the urine is clear like water.
- Use of elastic panty hose or stockings, with a pressure at the ankle of at least 20 mm of Hg, to increase blood return from the lower extremities.
- Tight body garments are very useful (Hamonet).
- In the absence of hypertension or kidney damage, we recommend adding to the diet 6 grams of salt a day (measured).
- Moderate aerobic exercises are useful.

Treatment of Dysautonomia (Cont.)

B.- Medications.

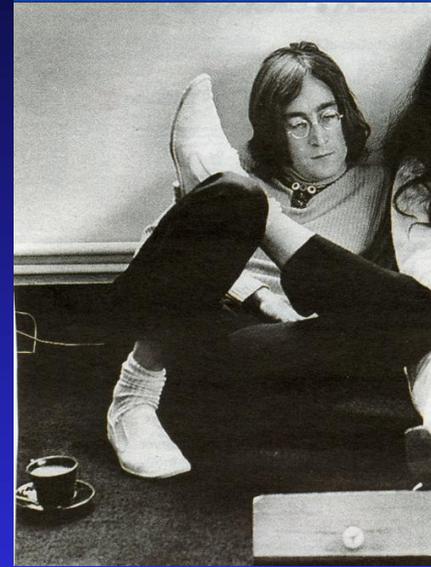
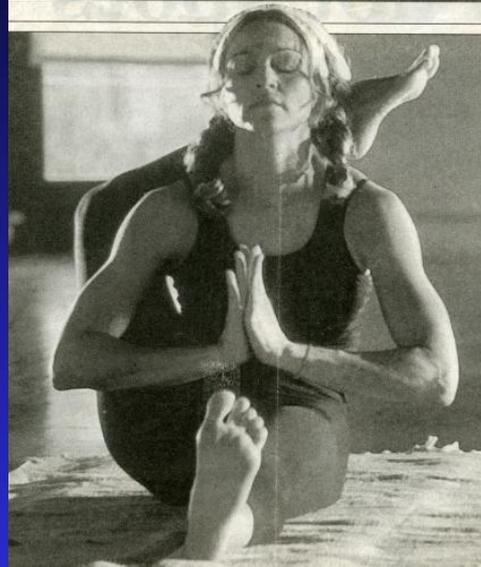
- 1.- Fludrocortisone (Florinef), Is a mineral-corticoid. 0.1 mg/day. Effect lasts 24 to 36 hours.
- 2.- Midodrine (Gutron). 2.5 to 5 mg 3 times a day. Effect lasts only 4 hours.
- 3.- Atenolol can be added to reduce tachycardia in POTS, keep in mind that it causes a slight reduction of the blood pressure.

Treatment of Dysautonomia (Cont.)

- Anti-depressants: Fluoxetine
Sertraline
Citalopram
Escitalopram
Paroxetine
- Anxiolytics. Alprazolam or others

Anemia and Sleep Apnea aggravates Dysautonomia , it is necessary to treat them.

Hypermobility can be an Asset







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