Thoughts about Ehlers-Danlos and Dysautonomia

- Patients with Ehlers-Danlos are not always hypermobile. In my study of 2,300 Ehlers-Danlos type III (EDS-III), now also known as Ehlers-Danlos Hypermobile (EDSh), presented as a Poster in the 2018 EDS Congress in Paris, I showed that 63% of these were not hypermobile (using the new Brighton 2017 Criteria). It is necessary to think that there are very hypermobile, little hypermobile and nonlax EDS-III patients, thus this last group should never be discarded. This brings us to the paradox that in a high percentage there are "nonlax hypermobile patients". Personally I am one of these patients and had Dysautonomia, and while living in the US, it was never diagnosed, nor treated.

- Of all the patients with Dysautonomia who I have treated, 100% of them had Ehlers-Danlos, most of them Hypermobile type (SEDh). I prefer to call them, EDS type III, since not all are hypermobile. The cause of the Dysautonomia resides in that the venous insufficiency of the legs (caused by the abnormal collagen of its walls) produces "a venous blood pool", resulting in cerebral hypo-perfusion and hypo-oxygenation, causing the symptoms.

- A sequel of a non treated Dysautonomia is Adult Attention Deficit Syndrome, because in Dysautonomia there is cerebral hypo-oxygenation, which is obvious harmful. This could suggest the appearance of cerebral damage in later stages of life. Due to this, it is very necessary to diagnose and treat it as soon as possible. It is my belief that the Attention Deficit Syndrome in Children is due to an arterial hypotension and since pediatricians generally do not check their blood pressure, this condition is not recognized as Dysautonomia. These children most likely also have EDS. Their way to compensate the lack of cerebral oxygen (which causes attention deficit) is with hyperactivity, which results in normalization of the blood pressure. If the blood pressure is low, perhaps an increase in salt intake could be more beneficial than prescribing medications, such as Ritalin or similar. It is necessary to make scientific studies to clarify these impressions.

- With experience in the diagnosis of Dysautonomia, it seems to me unnecessary to ask for a Tilt Test for its diagnosis, since its symptoms are very characteristic: chronic fatigue with somnolence, lack of energy, migraines,

dizziness, cold intolerance, with or without fainting spells. The Tilt Test is very uncomfortable, sometimes dangerous (arrhythmias, syncope, etc.) and expensive. The worse thing is that in case of a negative result, it does not discard the diagnosis, since it is known that it can give false negatives. Therefore, generally I do not ask for it. The diagnosis should be suspected in young women, especially if they have joint hypermobility. In a lower percentage, these symptoms are also present in male EDS patients with or without hypermobility. It is also important to keep in mind that there is a dominant inheritance.

- During my 30 years working as rheumatologist in the U.S. I treated thousands of patients with Fibromyalgia (FM), reason why I know the disease well. Now with experience in Ehlers-Danlos, I have come to the conclusion that people diagnosed with FM, probably are EDS-III. In both cases they can have chronic fatigue and arthralgias, that can be invalidating. Generally medical examinations and laboratory tests are normal. When I see a patient which has been diagnosed with FM, I use the Brighton Criterion (a positive test is diagnostic of EDS-III), which comes out positive in the majority. If somebody tells me, "my aunt has FM", I tell them that their aunt probably has EDS-III and I suggest them that she needs an evaluation. Most doctors do not have the EDS diagnosis in mind, and the closest they get is to diagnose these patients with FM.

- When a patient has seen many doctors (medical pilgrimage), of varied specialties, with normal physical findings and have not arrived to a correct diagnosis, it is necessary to suspect EDS, whether the patient is or isn't hypermobile. These patients have generally suffered the following for years: arthralgias, joint cracking noises, sprains, tendinitis and sometimes subluxations. Sometimes associated to chronic fatigue, lack of energy, migraines, dizziness, with or without fainting spells (Dysautonomia). Many usually present depression and anxiety.

- Patients with EDS must know that it is not immunological, but a hereditary genetic disease, and is not associated with sinovitis (joint inflammation). Generally it does not destroy the joints, except when there is associated Osteoarthritis, that usually appears early in these patients. There can be found an association with EDS in a low percentage of patients with

Rheumatoid Arthritis or Lupus Erythematosus. A patient with arthritis can have added symptoms, due to EDS, such as arthralgias, tendinitis, subluxations and joint cracking noises, either hypermobile or not. In these cases, it is important to add the diagnosis of EDS-III to decide which of the two diseases the symptoms belong to and thus not to increase the medicine for arthritis, which can have an indirect effect, and add analgesic and physical therapy, if necessary.

As time passes I see more association of allergies and EDS. The majority of these patients have had Asthma or Allergic Rhinitis as a child, which in many cases it has persisted thru adulthood. This allergy is usually Spring Allergy and specially allergy to insect bites. It is for this reason, that studies relating Mast Cells with EDs are important.

- It is time for doctors to stop thinking that hypermobility is an advantageous condition and know that this is a hereditary, dominant disease, very frequent and that causes pains and problems that can severely affect the patients quality of life. In spite of not having a curative treatment, the advantages of a precise diagnosis are undeniable. EDS-III affects 40% of the population and causes articular (including muscular tear and tendon rupture) and extra articular problems. Among the latter ones, the most common are: Dysautonomia, arrhythmias, tachycardias (POTs, Postural Orthostatic Tachycardias), gastric reflux, irritable bowel syndrome, hiatus hernia, diverticulitis, fragile skin with bad healing, bruises, infertility, premature membrane rupture, pelvic floor problems, such as prolapse and recurrent cystitis. In addition, it can be associated to Depression, Anxiety, Panic Crisis and Phobias and to propioception alterations, which consists in a bad perception of the position of the extremities, such as not knowing if the big toe is looking up or down, when examined by the physician. It is known that these patients can also have the following at an early age: Osteoarthritis, Erosive Osteoarthritis, Osteopenia or Osteoporosis, Hallux valgus, Varicose veins, Hernias and even Spinal Disc disease. We suspect that an EDS patient that smokes has more probabilities of developing Emphysema. The awareness that the person has Vascular EDS, can save his life. These patients must know that they could have high risk pregnancies, cerebral aneurism or arterial ruptures. Also invasive procedures or examinations must be avoided, such as endoscopies, because of the possibility of organ ruptures.

- With respect to the Dysautonomia, it is important that doctors, specially cardiologists, pay more attention to arterial hypotension, even slightly low pressures, such as 120/80 in the office, associated to the above mentioned symptoms. It is known that the blood pressure rises at the doctor's office, by the white apron phenomenon. This is why I believe that the disautonomic symptoms are more important that the blood pressure measured only once in the office. I ask my patients to measure their blood pressure at home occasionally and write it down, adding a commentary in relation to the energy they presented at that moment.

Patients with Dysautonomia should alert the anesthesiologist about their diagnosis, in case of an operation and specially if they are taking Fluodrocortisone, that is a mineraloccorticoid.

The myth that a low blood pressure is a good condition, must disappear. Both hypotension and hypertension are diseases. How can hypotension be normal if it causes migraines, dizziness, chronic fatigue and syncopal episodes, which disappear with the proper treatment, substantially improving the patient's quality of life.

For more information, refer to **www.reumatologia-dr-bravo.cl**

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September 27th, 2018