

Information on hypermobility (Ehlers-Danlos) and Dysautonomia, for parents and professors.

The Ehlers-Danlos Syndrome (EDS) is a disease and not only a condition, that affects 40% of the population. These people can be very hypermobile, little or may have no joint hypermobility. They may have articular and non articular symptoms affecting the quality of life of the patients. It is not an arthritis (there is no inflammation) and is not an immunological problem. It is a hereditary disease and because of that it can appear in childhood. As it has dominant inheritance, it will affect half of the brothers and sons, as well as one or both parents. In spite of its high frequency, this disease is not well known by the doctors, as much in Chile as abroad.

In this article we want to emphasize the importance of early diagnosis and treatment since childhood and for this reason we think is necessary to inform parents and professors.

A child with EDS can be born with hip dysplasia, which if not treated can give pain and difficulty walking for life. During childhood these children can have hernias (inguinal or umbilical), allergic rhinitis or bronchial asthma or recurrent bronchitis. They are generally allergic, in special to insects bites. It is frequent to find flat foot and scoliosis. Usually they begin to walk late, after the age of 12 months and generally they do not crawl. They can have muscular weakness, requiring physical therapy. In adolescence usually they have "growing pains", especially in the lower extremities.

How to suspect the diagnosis

A good observer, parent or professor, can realize that these children have special characteristics. They can have obvious joint hypermobility, they can be hyperactive or calm, sometimes sleepy, they may seem absent and without energy. They may have chronic fatigue associated to yawning. They can be laying down in a sofa or with the hand holding the head, as if the head weighted too much. They may have difficulty to write, since they do not handle well the pencil, due to the joint hypermobility (sign of the hyperlax scribe). This sometimes is cause of bullying. They seem to be unsociable, since they do not have energy and no desire to participate in activities with others. They feel better when they are active and worse with inactivity (it lowers the blood pressure). For this same reason they are

coldblooded. With these symptoms the doctor suspects hypoglycemia, hypothyroidism or anemia, but this is not the case and the laboratory examinations are normal. These children at times end not attending classes and remaining at home, doing nothing, with which they feel worse, since inactivity lowers their blood pressure. They arrive late at classes or they miss classes frequently. The adolescent females feel bad during the menstrual period. These patients have intolerance to excessive heat and to very hot and prolonged baths. Usually they have bad memory and poor concentration. They suffer from migraines, dizziness and some may have fainting spells, for which they are seen by neurologists, to discard epilepsy. All these symptoms are due to the fact that they have Dysautonomia (low blood pressure). The cause of this low pressure in EDS, is the result of venous insufficiency of the legs due to the collagen alteration of the vein walls. When standing from a sitting position the blood stays in the lower extremities, diminishing cerebral irrigation and oxygenation. They can be somewhat clumsy due to alteration of the propiocepción, that consists of not appreciating the position of the extremities, like not being able to note if the big toe is downwards or upwards, when the doctor examines them. The skin usually is pale and thin letting us see the veins, especially in the forearms. For this same reason the escleras (the white of the eye) can be light blue. Due to vascular fragility there is tendency to bruises, sometimes without apparent cause, which can be confused with battering. There is poor wound healing and striae. Some children are very agile, as if they were made of rubber and others are able to play making exaggerated movements of the fingers and boasting of their capacities to other children ("party tricks"). The ankle sprains are frequent and other injuries like tendinitis and sublaxations (temporo mandibular joint, the base of the thumbs, the knee, etc.). Cracking noises are characteristic of EDS, they are not harmful and is more, usually by cracking the joints the pain and the tension of the joint diminishes. It is frequent to have joint pain, but without arthritis, that is to say, there is no joint inflammation (there is no redness, pain, or heat). The pain is similar to the one seen in Fibromyalgia (FM). It is my impression is that in adults FM is part of EDS.

It is necessary to know that EDS type III is most frequent than type IV, also called Vascular EDS which is more serious, since usually can have arterial ruptures.

In EDS we usually find Myopia and Strabismus. Some children have the typical face of EDS type III: triangular face, pale skin, prominent bony nodule

on the dorsum of the nose, blue esclerae, and sometimes prominent ears. In 15% the children can be tall, with long extremities and big hands and feet. Sometimes they have sunk or prominent chest. This constitutional type is called Marfanoid Habitus, by the similarity to the Marfan Syndrome. This disease, also presents joint hypermobility, but it is more serious, presenting arterial problems. By their high stature they stand out in the sports, especially in swimming, but because of having weak tissues usually they get frequent injuries. Here the necessity that the hypermobile children have a scholastic insurance.

In an hypermobile children with Attention Deficit a possible cause is Dysautonomia (low blood pressure) and in these case, it would be better to treat the hypotension (by increasing the salt) than to give them Ritalin or other medications.

The extra articular symptoms of EDS are:

- Digestive problems, such as nausea, vomiting, irritable bowel syndrome, diarrhea, constipation.
- Depression, Anxiety, Panic Crisis and Phobias.
- Dysautonomia.
- Alteration of the proprioception (clumsiness).
- Arrhythmias, with tachycardia or bradycardia.

It is necessary to give facilities to these youngsters at school

- They should not remain seated, without moving, for prolonged periods. It is necessary to allow them to get up and around during and between classes. We should allow them to be a little restless.
- Some have special needs, such as drinking water and going to the bathroom during classes. The reason for this is that Dysautonomia treatment requires to take abundant liquids and to increase the activity. Students that have difficulty to write could be authorized to use tablets or cellular phones to record the classes in audio.
- Many have cold intolerance, and for this reason they should be allowed to dress warmly and the classroom temperature should be kept warm.

- Some of them have difficulty learning due to bad memory and low concentration and they need more time and understanding. Study renders them less and sometimes in spite of studying a lot the result is not good. Sometimes in spite of knowing the lesson, they get confused and give wrong answers.
- Some arrive late or miss classes due to their disease. Others do not attend because of pain or chronic fatigue and miss tests or exams, so it is necessary to give them facilities to solve these problems.
- Physical Education teachers should not demand these patients more than what they can give and they must avoid exercises and sports that may injure them. If this is not possible, a doctor should give a certificate to exempt them from these classes. The ideal is that they should be active and to build good muscles but without injuries. We recommended Pilates, Yoga, Swimming and Bicycling.
- Hopefully children should not get homework, since they need to play, run and not to continue to be seated for more hours.
- It is necessary to supervise that during breaks the children keep active and are not seated or playing with their cellular phones. At home it is also necessary to foment activity and to avoid using too much time playing with the cellular phone or watching television. They must get up from the chair and move about every half an hour.
- These children suffer pains (it is necessary to believe them), anxiety, depression, and sometimes bullying and for this reason it is necessary to be alert to detect these problems and to support them with affection and understanding.

Need to notify parents and tutors

If a professor suspects that a student could have SED or Dysautonomia, he must notify the parent or tutor to inquire on the matter and we would advise him to see the Page Web www.reumatologia-dr-bravo.cl

If the first impression is corroborated, it is necessary to consult a doctor specialized in the subject. As the disease has dominant inheritance, is probable that half of brothers and one of the parents has the disease and they may not know it.

In summary With a early diagnosis it is possible to initiate an effective treatment and to avoided low grades, bullying, anxiety and depression. Knowing the cause of his child problems, gives the parents great tranquility. With treatment it is possible to reduce articular pain and prevent recurrent muscle-skeletal injuries. By improving arterial cerebral circulation we improve the scholastic yield, the self-esteem and in general, we improve greatly the quality of life. The medical pilgrimage is avoided, visiting different specialists (pediatricians, orthopedists, hematologists, neurologists and psychiatrists, among others. Also we avoid multiple examinations and laboratory tests, which generally give normal results.

The proper diagnose and treatment gives parents tranquility, when they understand their child's multiple symptoms of and the possibility to help him.

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