Ehlers-Danlos, an Unknown and Disturbing Syndrome. Free Comments

Claude Hamonet* and Lucette Ducret
Department of Medicine, University in Créteil, France

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*Corresponding author: Claude Hamonet, Department of Medicine, University in Créteil, Emeritus Professor, 94010, Creteil, France, Email: pmhamonet@wanadoo.fr

Abstract
Ehlers-Danlos syndrome is a frequent, inherited disorder which is rarely diagnosed. This exposes these fragile patients to a long medical wandering and the risks of an iatrogenic that sometimes endangers their lives. It is a hereditary disease, without genetic testing in the most frequent clinical pictures that is transmitted to all the children of a parent who is affected. A definitive diagnosis is possible on a grouping of clinical signs. Cognitive disorders are common. The risk of aneurysms is possible due to arterial fragility. Cohabitation with autism and Asperger's disease exists and needs to be deepened.

Keywords: Ehlers-Danlos syndrome; Hyper mobility; Aneurysms; Proprioception disorders; bleeding; Asperger disease; Autism; Iatrogenic; Chronic pain; Cognitive disorders; Hereditary disease

Introduction
This frequent hereditary connective tissue disease has great difficulty in being admitted by the medical community. Its very existence is often questioned by some doctors who see in it the expression of a psycho-affective disorder. They thus deprive the concerned patients of diagnosis and adapted treatments. These patients are fragile and exposed to a long medical wandering and iatrogenic which can go as far as putting their lives in danger. We have diagnosed and often monitored and treated more than 5,000 people with this disease in 23 years. This intensive clinical experience has led us to many findings about this syndrome in medical practice, including neurological and psychiatric signs.

Identifying Ehlers-Danlos syndrome
It is possible today and we think that it is easy, for any knowledgeable clinician, on clinical symptoms alone. This is how two dermatologists: Tschernogobow, a Russian in Moscow and Ehlers [1,2] in Copenhagen, initially described it. They mentioned symptoms that have retained their strong relevance today: hemorrhages, joint instability and coetaneous fragility, motor control impairment (ataxic gait) wrote Ehlers. On the other hand, Danlos [3] was mistaken and described another illness: pseudoxanthoma elastic. He insisted on stretch ability and elasticity of the skin, which he describes as “extraordinary” [3]. This is the origin of many rebuttals in front of a skin, although a little stretchable, but not as stretchable as described by Danlos [3]. Other mistakes in the identification of this disease, polymorphic in its expression, will seriously compromise its identification, the most obvious error being that women complaining of pain, fatigue, sleeping and digestive disorders, precisely the characteristic of Ehlers-Danlos disease, were misdiagnosed with fibromyalgia instead of Ehlers-Danlos syndrome, which was unknown to the first descriptor [4].

The diffuse nature of the connective tissues involvement with damage to all human tissues is at the origin of the very large number of manifestations which confuse the clinician trained in semi logical analysis organ by organ. Moreover, this symptomatology is variable from one day to the next, posing, in the minds of doctors, the question of the organicity of the symptoms. The influence of internal factors (hormonal in the woman mainly), external (trauma, physical activity, climate variations) appears very clearly. Family sickness, it varies in expressions between members of the same family, including between heterozygous or homozygous twins as we have observed. Its evolution is impossible to foresee and one can, in the same family, observe severe forms with many disabilities and paucisymptomatic forms. In the absence of genetic testing for most Ehlers-Danlos syndrome’s cases [5], it is possible to establish the definitive diagnosis on the basis of a grouping of clinical signs suggestive by their frequency and/or specificity. Twelve signs have been selected in a recent study [6] that overlaps the descriptions of other clinicians [7,8]. Clinical manifestations include arterial
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anterior to the cerebral circulation which is the consequence of shock waves on poorly shielded brains by pathological tissues which does not dampen them as well. Elsewhere, images of neurovegetative disorders, involving the supraventricular region by venous stasis may be confused with those seen in multiple sclerosis.

Cognitive difficulties are very often associated (neurology%): memory disorders, attention, concentration, orientation in time and space, learning disabilities with dyslexia, dysorthographia, dyspraxia with negative consequences on schooling and working life. They usually contrast with observation, judgment and reasoning abilities that allow for compensation. The association with autism and Asperger’s disease is manifested in two ways: by the behavior of certain children in a family with Ehlers-Danlos syndrome. They are hyperactive, gifted but having difficulties in contact with others. Elsewhere, it is the psychiatrists, trained in the diagnosis of the syndrome, who identify them.

Conclusion

Ehlers-Danlos syndrome is a clinical entity currently identifiable with certainty by the clinical inputs. The genetics contribution remains very modest in everyday practice. Advances in the clinic are sufficient to identify the disease, to inform about the risk of transmission to avoid certain dangerous therapies (antiocoagulants and surgery) and to implement effective treatments (clothing with proprioceptive ortheses and oxygen therapy).

References


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