

muscular metabolic disease. The main risk factors associated with the development of rhabdomyolysis are lack of physical conditioning and the performance of extreme muscular exertion.⁸ Our patient was a young woman who was used to physical activity. However, in our opinion, the use of a muscle electrostimulator while performing physical activity may generate tensile strengths capable of provoking the disruption of the muscle fibre's integrity with the well-known consequences. Due to lack of more cases, we cannot establish a universal recommendation advising against the use of electrostimulators. Nevertheless, common sense suggests that their use should not be made while performing active exercises.

References

1. Walsworth M, Kessler T. Diagnosing exertional rhabdomyolysis: a brief review and report of two cases. *Mil Med.* 2001;166:275–7.
2. Parmar S, Chauhan B, DuBose J, Blake L. Rhabdomyolysis after spin class? *J Fam Pract.* 2012;61:584–6.
3. Peña Irún A, Pérez del Molino Castellanos A, González Santamaría AR, Santiago Ruiz G. Rhabdomyolysis after a spinning session. Tests to rule out metabolic myopathy. *Semergen.* 2014;40:109–10.
4. Firestein GS, Budd RC, Harris ED, McInnes IB, Ruddy S. *Kelley's textbook of rheumatology.* 2 – Volume set, expert consult: online and print. 8th ed. Philadelphia: Elsevier Saunders; 2008.
5. Macdonald R, Rosner Z, Venters H. Case series of exercise-induced rhabdomyolysis in the New York City jail system. *Am J Emerg Med.* 2014;32:466–7.
6. Guarascio P, Lusi EA, Soccorsi F. Electronic muscular stimulators: a novel unsuspected cause of rhabdomyolysis. *Br J Sports Med.* 2004;38:505 [discussion 505].
7. D'Orazi L. Passive gymnastics. *Med Secoli.* 1990;2:269–92.
8. Lin H, Chie W, Lien H. Epidemiological analysis of factors influencing an episode of exertional rhabdomyolysis in high school students. *Am J Sports Med.* 2006;34:481–6.

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“Benign Hypermobility”-Hyperlax Ehlers–Danlos Syndrome. Other Comorbidities[☆]



«Hiper movilidad benigna»-síndrome de Ehlers-Danlos hiperlaxo. Otras comorbilidades asociadas

Dear Editor,

Nowadays, there are 11 different types of Ehlers–Danlos syndrome described. Types I and II (classic Ehlers–Danlos) and mainly type III are classified as benign hypermobility.

When studying this syndrome, we noticed that the word “hiperlaxitud” (hypermobility) does not exist in the dictionary of the Real Academia de la Lengua (Royal Spanish Academy). The concept of “hypermobility” refers to the articular hypermobility observed in these patients, causing painful symptomatology. As shown in the recent article of Pantoja et al.,¹ a score higher than 4 in Beighton's test² is the essential criterion for the diagnosis of this pathology, along with the presence of pain.

In the first International Symposium on Ehlers–Danlos syndrome, which took place in Ghent in September, 2012, an attempt was made to establish new diagnostic criteria that would offer higher specificity than the old criteria of Brighton and Villefranche.³ These new criteria must allow for the inclusion of truly hiperlax forms and the obtainment of universally accepted instruments for the measurement of laxity, not only articular but also from other tissues, avoiding professional subjectivity in the assessment of mainly cutaneous involvement⁴ (more or less flexible, silky, etc.).

The first inclusion criterion to be able to diagnose a patient of this pathology is that the patient is over 16, because, physiologically, tissue laxity is greater during childhood. Furthermore, it is important to point out that, physiologically, tissue laxity is higher in women than in men. This pathology usually affects women, as occurs among main connective tissue diseases.

The most interesting data we have gathered in our hypermobility/Ehlers–Danlos syndrome unit during the last 3 years is the following:

- Out of 23 patients diagnosed with articular laxity, 3 of them were men and 20 were women. The average age of our patients was 33 years. Out of the 23 patients, 18 (78%) were subjected to at least one musculoskeletal surgical intervention or due to peripheral nerve entrapment. Of those 18 patients, 11 needed reintervention of the same pathological process. The surgical interventions average was 3 per patient, and there was one specific case of a patient who was subjected to 13.
- Affectionate-emotional disorder was observed in 14 patients (60%). Of those, 3 had attempted suicide.
- In 5 patients (22%), low tension figures were observed and/or there were episodes of vasovagal syncope, in relation to the dysautonomia. In 6 patients (26%) a clear Raynaud's phenomenon with its 3 phases was observed. Autoimmune diseases were discarded for all of them. We observed livedo reticularis in 2 patients. Vitamin D3 deficit was identified in 17 patients (74%). We have not performed densitometric studies on our patients; therefore, we cannot provide data about the potential greater incidence of osteoporosis in subjects with this pathology, as pointed out in studies.^{5,6} We have not studied how many patients presented fibromyalgia or chronic fatigue syndrome criteria⁷ either.

As well-indicated in the section “Clinical Rheumatology in Images”, it is of interest to rheumatologists and other experts who assess musculoskeletal pathology to know the necessary information in order to diagnose “benign hypermobility” syndrome⁸ and rule out the possible presence of a vascular form or type IV, which is associated to visceral aneurysmal malformations.^{4,9} This vascular type is rare and difficult to diagnose and requires confirmation through genetic tests. Likewise, it is important to know the presence of comorbidities and the high rate of failed surgical interventions that are observed in patients with benign hypermobility, aspects to be taken into account in case of requiring management in surgical units.

References

1. Pantoja L, Diez C, Alexis D. Hyperlax Ehlers–Danlos syndrome: benign hypermobility? *Reumatol Clin.* 2014;10:189–90.
2. Beighton P, de Paepe A, Steinmann B, Tsipouras P, Wenstrup RJ. Ehlers–Danlos syndromes: Revised nosology, Villefranche, 1997. Ehlers–Danlos National

[☆] Please cite this article as: Turrión Nieves AI, Moruno Cruz H, Martín Holguera R, Sánchez-Atrio AI. «Hiper movilidad benigna»-síndrome de Ehlers Danlos hiperlaxo. Otras comorbilidades asociadas. *Reumatol Clin.* 2015;11:263–264.

- Foundation (USA) and Ehlers–Danlos Support Group (UK). *Am J Med Genet.* 1998;77:31–7.
- Bravo JF. Ehlers–Danlos syndrome, with special emphasis in the joint hypermobility syndrome. *Rev Med Chil.* 2009;137:1488–97.
 - Bravo JF, Wolff C. Clinical study of hereditary disorders of connective tissues in a Chilean population: joint hypermobility syndrome and vascular Ehlers–Danlos syndrome. *Arthritis Rheum.* 2006;54:515–23.
 - Yen JL, Lin SP, Chen MR, Niu DM. Clinical features of Ehlers–Danlos syndrome. *J Formos Med Assoc.* 2006;105:475–80.
 - Gulbahar S, Sahin E, Baydar M, Bircan C, Kizil R, Manisali M, et al. Hypermobility syndrome increases the risk for low bone mass. *Clin Rheumatol.* 2006;25:511–4.
 - Ofluoglu D, Gunduz OH, Kul-Panza E, Guven Z. Hypermobility in women with fibromyalgia syndrome. *Clin Rheumatol.* 2006;25:291–3.
 - De Wandele I, Rombaut L, Leybaert L, van de Borne P, de Backer T, Malfait F, et al. Dysautonomia and its underlying mechanisms in the hypermobility type of Ehlers–Danlos syndrome. *Semin Arthritis Rheum.* 2014;44:93–100.
 - Pepin M, Schwarze U, Superti-Furga A, Byers PH. Clinical and genetic features of Ehlers–Danlos syndrome type IV, the vascular type. *N Engl J Med.* 2000;342:673–80.

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Fibromuscular Dysplasia and Coronary Heart Disease[☆]



Displasia fibromuscular y enfermedad coronaria

Dear Editor,

We submitted information for a woman of 43 years, with a medical history of systemic arterial hypertension secondary to bilateral stenosis of renal arteries (Fig. 1A) and cerebral ischaemic attack

due to dissection of left carotid artery (Fig. 1B). She was sent for assessment due to effort angina. Coronary catheterization showed proximal occlusion of the anterior descending artery (Fig. 1C) with collateral flow from the right coronary artery (Fig. 1D). Aortography of supra-aortic trunks was normal. There was no evidence of anaemia, thrombocytopenia, or alterations in acute phase reactants or autoimmunity.

Coronary disease in patients younger than 45 years can be classified into atheromatous, non-atheromatous, hypercoagulability states or due to drug consumption. Even though atheromatosis is

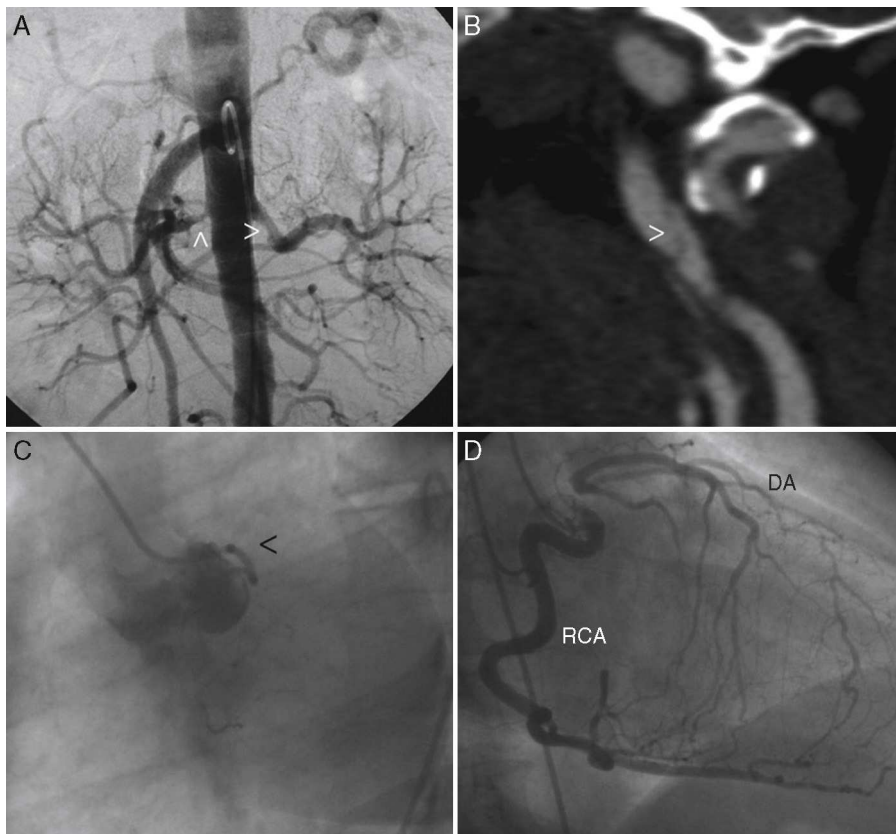


Fig. 1. (A) Abdominal aortography evidencing proximal stenosis at both renal arteries level (arrowhead). (B) Angiography by computed tomography where it is possible to observe left carotid artery dissection (arrowhead). (C) Left coronary catheterization (left anterior oblique) where only a poor-developed circumflex artery can be seen (arrowhead). (D) Right coronary catheterization (left anterior oblique) that evidences a dominant right coronary artery (RCA) that fills for collateral circulation of descending artery (DA), which is occluded at proximal level.

[☆] Please cite this article as: Martínez-Quintana E, Rodríguez-González F. Displasia fibromuscular y enfermedad coronaria. *Reumatol Clin.* 2015;11:264–265.