Dysautonomia and Osteoporosis in 1751 Hypermobile EDS patients, as defined by the Brighton criteria



Jaime F. Bravo San Juan de Dios Hospital. Santiago, Chile



Background/Purpose: To highlight the high frequency of Dysautonomia and Osteoporosis in Ehlers-Danlos Hypermobile type (EDSH), complications that are very frequent and usually undiagnosed. Identify the percentage of EDSH that are not hypermobile (negative Beighton score (BSc) with positive Brighton criteria (BC)). Stress the need to make the diagnosis with the BC and not using only the BSc.

Method: We have studied 1751 EDSH patients since 2002. The BSc and the BC were applied to all patients. Age: range 16 - 87, average 45.4. Females 78.6%. Patients were grouped in < 30 y/o (A), and over 30 y/o (B). Dysautonomia was clinically evaluated in 1734 patients. Densitometries were done in 830 patients (OMS criteria).

Results: 1751/2189 patients had EDSH (80%). BSc negative: 55.1%. Dys positive: M 26.5%, F 52.7%. Group A: M 56.1%, F 79.5%. Group B: M 20.1%, F 46.8%. BMD total group: normal 29.5%, Osteopenia 48.2%, Osteopenias 22.3%. Osteopenia group A: M 50.0%, F 46.3%, Group B: M 55.4%, F 46.8%. Osteoporosis: Group A: M 13.6%, F 14.9%. Group B: M 21.5%, F 23.6%.

Beighton Score in EDSH



Positive Brighton Criteria in 100% (per definition)

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.





Positive Dysautonomia (825) in 1734 EDSH patients evaluated

Positive	Total Positive Group n = 825		Group A (<30 a) n = 235		Group B (≥30 a) n = 590	
Males	99	26,5%	37	56,1%	62	20,1%
Females	726	52,7%	198	79,5%	528	46,8%
	825		235		590	

Bone Mineral Density studied in 830 EDSH patients

	Total Group	Group A (<	30 a) n = 89	Group B (≥30 a) n = 741	
	%	Males	Females	Males	Females
Normal BMD	29,5	36,4	38,8	23,1	29,6
Osteopenia	48,2	50,0	46,3	55,4	46,8
Osteoporosis	22,3	13,6	14,9	21,5	23,6

Conclusions:

- EDSH is extremely frequent in our Clinic (80%), referral Center.
- Significant percentage of EDSH patients are not hypermobile (BSc negative in 55.6%). In some studies, many EDSH patients are excluded when only applying the BSc rather than the BC.
- Dysautonomia is very prevalent in young EDSH females (79%). Low BMD is frequent, even at young ages, including Osteoporosis, circa 14% in both sexes.