Dysautonomia and Osteoporosis in 2300 EDS type III patients, as defined by the Brighton criteria



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

<u>Method</u>: We have studied 2300 EDS-III patients. The BSc and the BC were applied to all patients. Age: range 16 - 87, average 44.43. Females 78.7%. Patients were grouped in < 30 y/o (A), and over 30 y/o (B). Dysautonomia was clinically evaluated in 2283 patients. Densitometries were done in 1060 patients (OMS criteria). Patients younger than age 16 were excluded since the BC has been validated only for ages 16 and over.

<u>Results</u>: 2300/2868 patients had EDS-III (80.2%). BSc negative: 51.0%. Dys total group: positive: M 30.14%, F 56.16%; Group A: M 60.8%, F 84.04%; Group B: M 21.6%, F 60.1%. BMD total group: normal 31.6%, Osteopenia 47.9%, Osteoporosis 20.5%; **Osteopenia**: <u>Group A: M 45.7%, F 50.4%</u>, Group B: M 53.5%, F 48.0%; **Osteoporosis**: <u>Group A: M 8.6%, F 9.8%</u>, Group B: M 20.0%, F 22.9%.

Beighton Score in EDS-III (EDSH)



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 (1164) in 2283 EDS-III patients evaluated

Positive	Total Positive Group $n = 1164$		Group A $(< 30 \text{ y/o})$ n = 382		Group B ($\geq 30 \text{ y/o}$) n = 782	
Males	148	30.4 %	65	60.8 %	83	21.6 %
Females	1016	56.2 %	317	84.0 %	699	60.1 %
	1164		382		782	

Bone Mineral Density studied in 1060 EDS-III patients

	Total Group	Group A (<3	0 y/o) n = 89	Group B ($\geq 30 \text{ y/o}$) n = 741	
	%	Males	Females	Males	Females
Normal BMD	31.6	45.7	50.1	26.5	29.1
Osteopenia	47.9	45.7	40.1	53.5	48.0
Osteoporosis	20.5	8.6	9.8	20.0	22.9

Conclusions:

• EDS-III is extremely frequent in our Clinic (80.2%), referral Center.

- Significant percentage of EDS-III patients are not hypermobile (BSc negative in 51.0%). In some studies, many EDS-III patients are excluded when only applying the BSc rather than the BC, this is our reason for preferring to use the term Ehlers-Danlos type III, instead of Ehlers-Danlos Hypermobile.
- Dysautonomia is very prevalent in young EDS-III females (84%). Low BMD is frequent, even at young ages, including Osteoporosis, 8.6% in males and 9.8% in females.