

DYSAUTONOMIA AND OSTEOPOROSIS IN 1751 EHLERS-DANLOS TYPE III PATIENTS, AS DEFINED BY THE BRIGHTON CRITERIA

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Background/Purpose:

Highlight the high frequency of Dysautonomia (Dys) and Osteoporosis in Ehlers-Danlos Syndrome type III (EDS-III), awareness that the complications are very frequent and usually undiagnosed. Identify the percentage of EDS-III patients 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 studied 1751 EDS-III patients. The BSc and BC were applied to all patients. Age: range 16-87, average 45.4. Females 78.6%. Patients were grouped in < 30 y/o (Group A) and all ages (Group B). Dysautonomia was looked for, clinically, in all patients. Densitometries were done in 830 patients (OMS criteria).

Results:

1751/2189 patients had EDS-III (80%). BSc neg: 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%, Osteoporosis 22.3%. Osteopenia Group A: M 50.0%, F 46.3%. Group B: M 55.4%, F 46.8%. Osteoporosis: Group A: M 13.36%, F 14.9%. Group B: M 21.5%, F 23.6%.

Conclusions:

EDS-III is extremely frequent in our Clinic (80%), referral Center.

Significant percentage of Ehlers-Danlos type III patients are not hypermobile (BSc negative in 55.6%). In some studies many EDS-III patients have been excluded when only applying the Beighton score, rather than the Brighton criteria.

Dysautonomia is very prevalent specially in young EDS-III females (79%).

Low bone mineral density is frequent, even at young ages, including Osteoporosis, 14% in both sexes, in patients younger than 30 years old.