Low Bone Mineral Density (BMD) and Dysautonomia in Adolescents and Young Patients with Ehlers-Danlos Type III: Clinical Study of 90 Patients

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Abstract:

Introduction: The Hypermobile Ehlers-Danlos (HEDS), or Joint Hypermobility Syndrome (JHS) is a forme fruste of the Hereditary Diseases of the Connective Tissues (HDCT). Osteoporosis is frequent in HDCT (EDS, Marfan syndrome and Osteogenesis Imperfecta, reaching 100% in the latter and in EDS type VI).

Purpose: Describe the frequent alteration of bone mineral density (BMD) and Dysautonomia (Dys) in adolescents and young adults with HEDS.

Methods: The diagnosis of HEDS was made using the Brighton Criteria (BC). Marfanoid Habitus was diagnosed using the BC guidelines. In the last 4 years, of the patients younger than 30 years old, in which densitometries were done, we have found 90 patients with low BMD. Secondary osteoporosis was ruled out with the clinical picture and laboratory tests. Dysautonomia was clinically studied; Tilt Test was usually not done.

Results: Mean age 23.4 years old (16-29). Females 78%. Marfanoids 33%. Dysautonomia 66%. normal BMD in only 36%. Mild Osteopenia 17%, Moderate 21%, Severe 13%, and Osteoporosis 13%. Findings include that 70% of males and 63% of females had low BMD. No pathological fractures were observed in any of these patients.

Conclusions:
Patients younger than 30 years old with Hypermobile Ehlers-Danlos (HEDS), especially Marfanoids, presented a frequent and significant decrease in BMD that was very prevalent both in young male and female patients. Since this BMD alteration is so common in this young group of patients, we recommend getting densitometries in all patients regardless of age, in this frequent and usually undiagnosed condition. Also of note was the very high frequency of Dysautonomia in young patients with HEDS.