

SECONDARY BONE SIZE DEFICIT IN PATIENTS WITH EHLERS-DANLOS SYNDROME

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Background & Aims

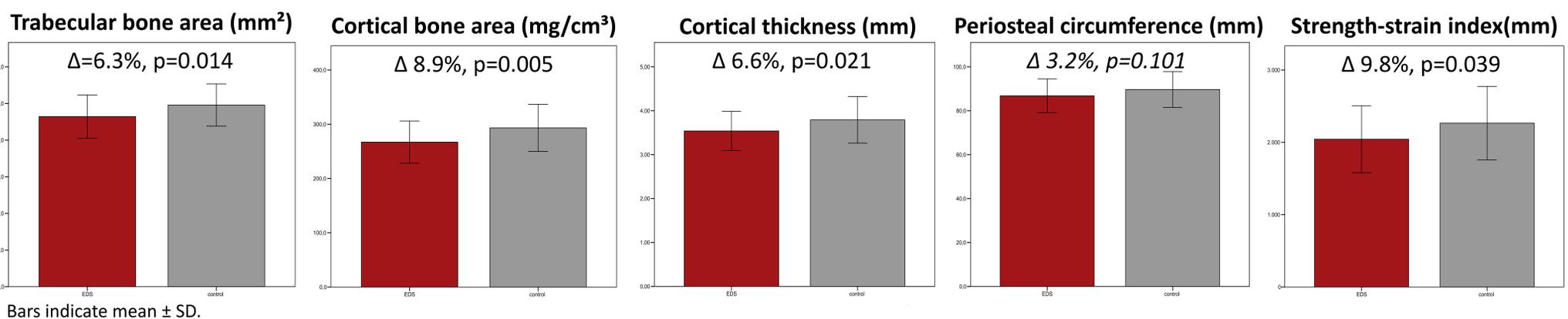
- Ehlers-Danlos syndrome (EDS) comprises a group of inherited connective tissue disorders, caused by various defects in the biosynthesis or secretion of fibrillar collagens. As collagen represents a major constituent of the bone matrix as well as of tendons and muscle, bone strength in EDS patients might be impaired both via direct and indirect pathways.
- Although decreased muscle strength, decreased areal bone mineral density (BMD), and increased fracture risk have been reported, no studies have investigated volumetric bone parameters in these patients. We compared volumetric BMD (vBMD) and cortical bone geometry in patients with hypermobility EDS (EDS-HT) and age- and sex-matched controls.

Subjects & Methods

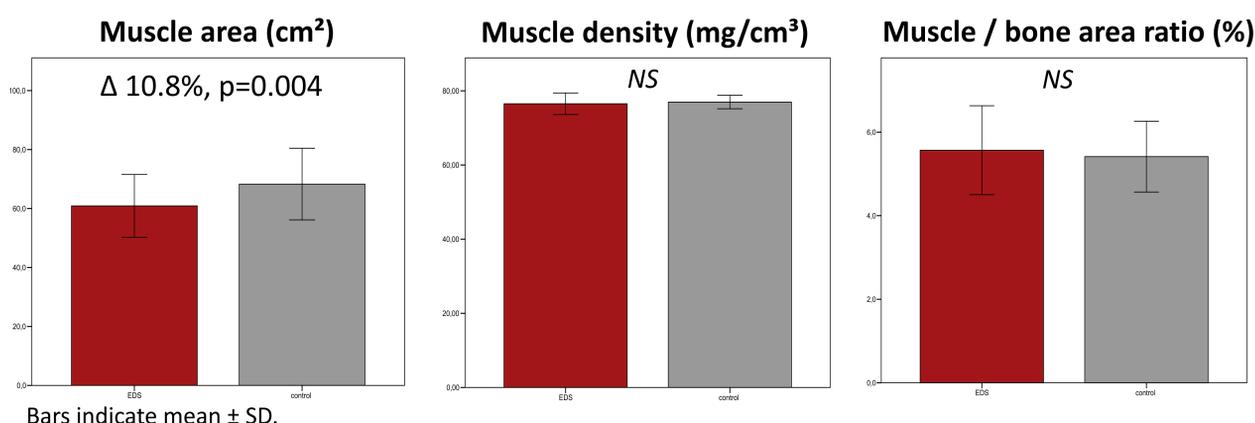
- 42 female EDS-HT patients (age 40.0 ± 10.8 years) and 42 age-matched controls recruited in a cross-sectional study.
- vBMD and bone geometry at the tibia (4% and 66% region) as well as lower leg muscle area (66% region) measured using pQCT.

Results

- Figure 1** shows that EDS-HT is associated with **smaller trabecular bone area** and with **smaller cortical bone area, smaller cortical thickness**, and a trend towards **smaller periosteal (but not endosteal) circumference**. As a result, EDS-HT patients had **lower strength-strain index** as compared to controls. No differences were observed regarding trabecular or cortical vBMD.



- Figure 2** shows that EDS-HT patients have a **lower muscle area** but **similar muscle density and bone/muscle area ratio**.



Conclusions

- EDS-HT patients show preserved bone mineralization but present with both a trabecular and cortical bone size deficit as compared to controls, which might contribute to their increased fracture risk.
- As indicated by the decreased muscle area and normal bone/muscle area ratio, this bone size deficit is probably secondary to decreased mechanical loading in these patients with known muscle dysfunction.