The muscle-bone interaction in Turner syndrome.

Abstract

OBJECTIVES: Turner syndrome (TS) is associated with an increased fracture rate due to reduced bone strength, which is mainly determined by skeletal muscle force. This study aimed to assess the muscle force-bone strength relationship in TS and to compare it with that of healthy controls.

METHODS: This study included 39 girls with TS and 67 healthy control girls. Maximum muscle force (Fmax) was assessed through multiple one-legged hopping with jumping mechanography. Peripheral quantitative computerized tomography assessed the bone strength index at the tibial metaphysis (BSI 4) and the polar strength-strain index at the diaphysis (SSI polar 66). The effect of TS on the muscle-bone unit was tested using multiple linear regression.

RESULTS: TS had no impact on Fmax (p=0.14); however, a negative effect on bone strength (p<0.001 for BSI 4 and p<0.01 for SSI polar 66) was observed compared with healthy controls. Bone strength was lower in the TS group (by 18%, p<0.01, for BSI 4 and by 7%, p=0.027, for SSI polar 66), even after correcting for Fmax.

CONCLUSIONS: Similar muscle force induces lower bone strength in TS compared with healthy controls, which suggests altered bone-loading sensitivity in TS.
Abstract

OBJECTIVE: Although hypogonadism and SHOX gene haploinsufficiency likely cause the decreased bone mineral density and increased fracture rate associated with Turner syndrome (TS), the exact mechanism remains unclear. We tested the hypothesis that muscle dysfunction in patients with TS contributes to increased fracture risk. The secondary aim was to determine whether menarche, hormone therapy duration, positive fracture history and genotype influence muscle function parameters in patients with TS.

DESIGN: A cross-sectional study was conducted in a single university hospital referral centre between March 2012 and October 2013.

PATIENTS: Sixty patients with TS (mean age of 13.7 ± 4.5 years) were compared to the control group of 432 healthy girls.

MEASUREMENTS: A Leonardo Mechanograph® Ground Reaction Force Platform was used to assess muscle force (Fmax) by the multiple one-legged hopping test and muscle power (Pmax) by the single two-legged jump test.

RESULTS: While the Fmax was normal (mean weight-specific Z-score of 0.11 ± 0.77, P = 0.27), the Pmax was decreased in patients with TS (Z-score of −0.93 ± 1.5, P < 0.001) compared with healthy controls. The muscle function parameters were not significantly influenced by menarcheal stage, hormone therapy duration, fracture history or genotype (linear regression adjusted for age, weight and height; P > 0.05 for all).

CONCLUSION: Fmax, a principal determinant of bone strength, is normal in patients with TS. Previously described changes in bone quality and structure in TS
are thus not likely related to inadequate mechanical loading but rather represent a primary bone deficit. A decreased Pmax indicates impaired muscle coordination in patients with TS.


Artificially low cortical bone mineral density in Turner syndrome is due to the partial volume effect.


Abstract

We aimed to show that the decrease in the cortical bone mineral density (BMD) in the radius in Turner syndrome (TS) is artificially caused by the partial volume effect. We confirmed that the partial volume effect-corrected cortical BMD is not decreased in TS compared to in the healthy controls. Other factors are responsible for the increased fracture rate in TS.

INTRODUCTION: Decreased cortical bone mineral density (BMD) has been reported in Turner syndrome (TS), using peripheral quantitative computerised tomography, and it is perceived as one of the major factors leading to increased fracture risk. We tested the hypothesis that low cortical BMD in the radius is caused artificially by the partial volume effect.

METHODS: A cross-sectional study was conducted at the university hospital referral centre between March and October 2013. Thirty-two participants with TS who consented to the study were included (mean age 15.3 ± 3.2 years). We assessed the cortical BMD in the radius as well as the tibia, where the cortex is thicker compared with the radius.

RESULTS: Whereas the cortical BMD was decreased in the radius (mean ± SD Z-score –0.6 ± 1.5, p = 0.037), it was increased in the tibia (mean Z-score 0.83 ± 1.0, p < 0.001). After correcting the cortical BMD for the partial volume effect, the mean Z-score was normal in the radius in TS (0.4 ± 1.3, p = 0.064). The corrected cortical BMD values were similar in the radius and tibia (1108 ± 52 vs. 1104 ± 48, group difference p = 0.75).

CONCLUSIONS: The cortical BMD is not decreased in TS. The partial volume...
effect is responsible for previous findings of decreased cortical BMD in the radius. Altered bone geometry or other factors rather than low cortical BMD likely play a role in the increased fracture risk in TS.


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