Muscle-bone unit in beta-thalassemic major females: A pQCT study

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Purpose

Beta-thalassaemia major represents a haemoglobinopathy caused by a hereditary defect in the synthesis of beta chain in adult haemoglobin. The aim of this study was to evaluate the bone structure and relationship between bone strength and muscle force in beta-thalassemic female patients by peripheral Quantitative Computed Tomography (pQCT), with a purpose of confirm the existence of musculoskeletal interactions at the tibia of these patients.
Methods and Materials

Twenty six females with beta-thalassemia major (26 female, age 22-41, body-mass index/BMI: 21-23) with history of fragility fracture and 25 healthy subjects matched for age and BMI underwent pQCT (XCT 2000 Stratec, Pforzheim, Germany) at the non dominant tibia. The following pQCT parameters were measured: cortical bone mineral density (CBD), marrow cavity area (MC), strength strain index (SSI), bone cross-sectional area (BCA), muscle cross-sectional area (MCA), fat cross-sectional area (FCA) and BCA/MCA ratio at the 14%, 38% and 66% of the tibial length.
**Results**

SSI was significantly reduced in beta-thalassemic major female patients compared with normal subject, although CBD was significantly elevated in female thalassemic patients (p<0.01). MC of female patients were significantly higher compared with healthy female controls (p<0.01). Compared with healthy subjects, BCA, MCA, FCA and BCA/MCA ratio were statistical significantly reduced in females with beta thalassemia major (p<0.01).

Table I.

<table>
<thead>
<tr>
<th>pQCT Parameter</th>
<th>CBD (14%)</th>
<th>SSI (14%)</th>
<th>MC (66%)</th>
<th>BCA (66%)</th>
<th>MCA (66%)</th>
<th>FCA (66%)</th>
<th>BCA/MCA Ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients</td>
<td>1193.47 ±36.12</td>
<td>1068.5 ±228.07</td>
<td>153.72 ±52.84</td>
<td>221.56 ±55.56</td>
<td>4654.3 ±25.32</td>
<td>2407.25 ±63.28</td>
<td>3.65 ±1.23</td>
</tr>
<tr>
<td>Controls</td>
<td>1165.61 ±16.6</td>
<td>1384.58 ±217.74</td>
<td>109.10 ±7.23</td>
<td>308.12 ±59.05</td>
<td>5529.8 ±75.8</td>
<td>3087.56 ±25.61</td>
<td>5.13 ±2.7</td>
</tr>
<tr>
<td>p value</td>
<td>&lt; 0.0001</td>
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</table>
Conclusion

Bones in beta-thalassemia major patients are exposed to a variety of abnormal influences. Previous studies have reported that osteoporosis is an important cause of morbidity in these patients. The pathogenesis is multifactorial and includes factors such as hormonal deficiency, especially gonadal failure, bone marrow expansion, increased iron stores, desferrioxamine toxicity, calcium/vitamin D deficiency and reduced physical activity; all seem to have a serious impact on the impaired bone metabolism of this disease. The "bone-muscle-unit" concept suggests that muscular strength mechanical effects possibly control changes in whole bone strength and mass under biological mechanisms.

Although dual X-ray absorptiometry (DXA) is the most commonly used technique for bone mineral density estimation, it provides a measure that does not account for changes in the quantitative and geometric distribution of trabecular and cortical bone tissues. Peripheral quantitative computed tomography (pQCT) permits the volumetric density of cortical and trabecular bone to be assessed separately as well as the noninvasive evaluation of the geometric properties of the tibia. Muscle cross-sectional area, which is known to be correlated to muscle force, can be non-invasively measured by means of pQCT.

In this study we used pQCT to determine cross-sectional bone geometry and volumetric bone mineral density in beta-thalassemia major females with fragility fracture history. Our results showed bone fragility (low BCA and SSI), an increased bone quality material (high CBD) and a marrow cavity expansion in the female patients compared to the controls subjects.

It is already known from the literature that beta-thalassemia patients present a lower body composition in comparison with healthy subjects. Our results showed statistically significant reduced muscle mass (MCA) and fat mass (FCA) in female patients compared to healthy subjects. The relationship between cross-section of cortical bone and muscle (BCA/MCA ratio) was weaker in the patients than in the controls, suggesting the presence of interfering pathogens on the bone-muscle unit. The bone marrow expansion of appendicular skeleton is due to increased and ineffective haemopoiesis occurring in these patients. Perhaps this bone expansion has an interactive effect with early estrogen deficiency in bone of these female patients. Probably it potentiates the resorptive effect of early estrogen deficiency at the trabecular level (cortico-endosteal and trabecular envelopes), while promotes stimulating bone formation and antagonize the resorptive effect of estrogen deficiency at the cortical level (periosteal and intrahaversian envelopes).

In conclusion beta-thalassemic major females present fragile bones (low BCA and SSI), weak muscles (low MCA) and low fat mass (low FCA). The increased marrow cavity and elevated cortical density in correlation with reduced BCA/MCA ratio and nutrition stunting suggest an estrogens related disturbance in bone metabolism in these patients.
Fig. 0: pQCT images at the 14% and 38% of tibia length in a 32 years old female beta-thalassaemic patient show a tibia marrow cavity expansion.

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Fig. 0: pQCT images at the 14% and 38% of tibia length in a healthy 30 years old female, demonstrates a normal tibia marrow cavity.

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Fig. 0: pQCT image at the 66% of tibia length in a 39 years old beta-thalassaemic female patient with BMI 22, shows reduced muscle cross-sectional area, fat cross-sectional area and tibia bone cross-sectional area.

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**Fig. 0:** pQCT image at the 66% of tibia length in a healthy 37 years old female with BMI 21.5, demonstrates normal muscle cross-sectional area, fat cross-sectional area and tibia bone cross-sectional area.

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References


