MSG 2009 Abstract

Mechanostat theory used to understand bone complications in children with Duchenne muscular dystrophy

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Abstract

Introduction: To date, there has not been an adequate way to look at bone density in children with chronic disease. These children are compared to age-matched controls, which is inadequate considering many have been on chronic glucocorticoid therapy or non-ambulatory which alters growth, lean muscle mass and bone composition. Such is the case in Duchenne Muscular Dystrophy. These patients are at risk for bone complications such as scoliosis and fragility fractures, however, predicting these complications remains difficult.

Objectives and Hypotheses: This study aims to better understand the relationship between bone density and bone complications in DMD using the mechanostat theory. We predict that children with DMD will have secondary osteopenia (low bone mass due to low muscle mass) and those that have experienced bone complications will have more severe forms of osteopenia.

Methods: This is a cross-sectional study of 7 patients with genetically or muscle-biopsy diagnosed DMD or Becker's MD. The bone density results were obtained from the DEXA scans of total body, proximal hip and lumbar spine. The mechanostat theory was applied to create ratios of bone mineral content to lean tissue mass (BMC/LTM) and compared to normal control values. A chart-assisted interview was carried out in to acquire patient's demographic information and orthopaedic history.

Results: The children were ages 10-16 (mean 13.6). The height for age in all patients was less than -0.5 s.d. (range -0.5 to -3.9). The LTM/height ration was less than 2s.d. for 4/7 patients suggesting secondary osteopenia. The BMC/LTM was less than -2 s.d. (-2.0 to -5.4) in 5 of the 7 patients. These 5 patients are demonstrating a low BMC even when adjusted for their LTM, or mixed osteopenia. Fragility fractures occurred in 2/7, one of whom demonstrates severe secondary osteopenia (LTM/ht of -4.7 s.d.) and the other demonstrating a mixed-type osteopenia (LTM/ht of -3.9 s.d and BMC/LTM of -5.4 s.d.).

Conclusions: Duchenne muscular dystrophy patients with severe osteopenia, as assessed by the mechanostat theory, are at higher risk for suffering fragility fractures.