Bone markers and bone mineral density in Duchenne muscular dystrophy

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Duchenne muscular dystrophy (DMD) is an inherited X-linked recessive disorder and is the most common muscular dystrophy in childhood. To date, there exists no specific causal treatment, but glucocorticoids (GC) have been shown to slow the progression of the disease and improve muscle function. Few studies have been conducted concerning bone health in DMD boys.

This cross-sectional study examined bone mineral density (BMD) and biochemical markers of bone turnover in 24 boys with DMD (2.3-19.7 years), most of whom were being treated with prednisolone, and 24 age-matched healthy boys (2.7-19.6 years). According to Tanner staging no significant difference in puberty level was seen between the groups.

BMD for total body (TB), TB head excluded \( (\text{TB}_{\text{HE}}) \), spine, heel, and hip, were lower in the DMD group compared with the control group \( (p<0.001) \), as were Z-scores for TB and spine BMD \( (p<0.001) \). BMD, at all sites, increased with increasing age in the control group. BMD values obtained from the hip and the heel decreased within the patient group with increasing age while values obtained from the TB, TB\(_{\text{HE}}\), and spine remained at the same level during childhood and adolescence. The differences in BMD values between patients and controls increased significantly with increasing age \( (p<0.001) \).

All bone formation markers were significantly lower in the patient group compared with the control group; BALP \( (p=0.002) \), PINP \( (p<0.001) \) and OC \( (p<0.001) \). Significantly lower values for the bone resorption markers CTX \( (p<0.001) \) and TRACP5b \( (p=0.002) \) were also observed, however, not for ICTP \( (p=0.087) \). The CTX/TRACP5b quotient (a suggested index of the osteoclast activity in relation to the number of osteoclasts) ranged between 0.09-0.38 for the patient group and 0.15-0.85 for the control group. Matched-pair analysis, which could be performed in 16 pairs, showed that 11 patients had a lower quotient than their corresponding control \( (p=0.025) \).

In conclusion, BMD was generally decreased in DMD patients compared with healthy matched controls and these differences in BMD values increased with increasing age. Both bone formation and resorption were suppressed in the DMD boys. BALP appeared to be the marker that better differentiates the pathological state in DMD, at least during puberty, which could be due to the highly significant relation between longitudinal growth and BALP. The mechanisms for these findings could be a combined action of GC treatment, low muscle mass and reduced mechanical loading in DMD patients, thus resulting in a lower degree of bone accrual and less longitudinal growth during childhood and adolescence.