Preoperative bisphosphonate treatment in patients with neuromuscular scoliosis improves bone strength of vertebral body

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Introduction
• Boys with muscular dystrophy as presented by Duchenne muscular dystrophy (DMD) lose muscle strength and are usually confined to a wheelchair by the age of 12-14.
• Glucocorticoid (Deflazacort) therapy is widely used to extend the ambulatory periods and to prevent scoliosis. If myogenic scoliosis develops after wheelchair-bound life, scoliosis surgery is necessary to acquire the sitting balance.

Osteoporosis is one of the major concerns to perform scoliosis surgery. Patients with DMD or congenital muscular dystrophy (CMD) have fragile bones due to loss of ambulation, glucocorticoid therapy and DMD itself.

Result 3. As the duration of treatment was longer, BMD increased further. There were no incident osteoporotic fractures during BP treatment.

Result 4. All patients could continue to use BP without any adverse effects. Adherence: 100%, no adverse effects such as gastrointestinal tract disturbance

Result 5. As spine BMD increased, higher fixation strength of pedicle screw was obtained.

Subjects & Methods
Non-ambulatory boys with muscular dystrophy who had scoliosis surgery were preoperatively administered oral BP (Alendronate 35mg) once a week. BMD and bone turnover markers were measured before and after BP treatment.

Patients demographics
Number of the patients 12
Age (years) 14.4 ± 1.6
Body weight (kg) 31.1 ± 9.0
Type of muscular dystrophy (cases) DMD: 9, CMD: 3
Presence of prevalent fracture 25% (3/12)
Type of fracture Femur: 2 cases, Humerus: 1 case, Vertebrae: 0 case
Previous history of therapy with glucocorticoid 0% (0/9)
Duration of BP treatment (days) 160 ± 53 (85-280)

Clinical assessments
BMD L2-4, Whole body (T-spine, L-spine, Pelvis)
Bone turnover markers Bony ALP, P1NP, TRACP-5b
Pedicle screw fixation strength Pedicle screw insertion torque during operation

Case Presentation
15-years-old DMD boy
• Height: 156cm, weight: 25kg
• Non-steroid user
• Non-ambulatory at the age of eight
• Difficult to keep sitting at the age of fourteen
• Duration of treatment: 183 days

Discussion
• Several studies showed the efficacy and safety of BP for osteoporosis associated with DMD.

Results
Result 2. BP treatment significantly decreased bone turnover.

Result 1. Non-ambulatory boys with muscular dystrophy had severe bone loss, and BP treatment significantly increased spine BMD.

Conclusions
• Twelve patients with muscular dystrophy had severe osteoporosis with high bone turnover, and BP treatment significantly increased spine BMD and decreased bone turnover without any adverse effects.
• Improvement of bone fragility by preoperative BP treatment will secure success in surgical treatments for myogenic scoliosis.

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