Skeletal Disproportion in Glucocorticoid Treated Boys with Duchenne Muscular Dystrophy

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Background

- Short stature is common in boys with Duchenne Muscular Dystrophy (DMD). Little is known about body proportions in DMD.
- Use of DXA to assess body proportion and measure bone is feasible in children with chronic conditions .

Objective

To compare body proportions and bone lengths in boys with Duchenne Muscular Dystrophy (DMD) treated with glucocorticoids with healthy controls using dual energy absorptiometry (DXA) images.

Methods

Participants

- 30 boys with DMD who had DXA performed on the Lunar iDXA were recruited.
- Excluded: not current treated with Glucocorticoids (GC), metal instrumentation or severe scoliosis (Cobb angle >20°).
- 30 healthy age matched boys who had DXA performed as part of a DXA study of bone mineral density was the comparative group.

Variables and analysis

- Total height (Ht), sitting height (SH), leg length (LL) and bone lengths (femur, tibia, humerus, forearm) were measured using DXA.
- Ht, SH, LL, SH:LL ratio (SH:LL) and bone lengths in DMD were compared to controls, adjusted for age and puberty (multiple linear regression).

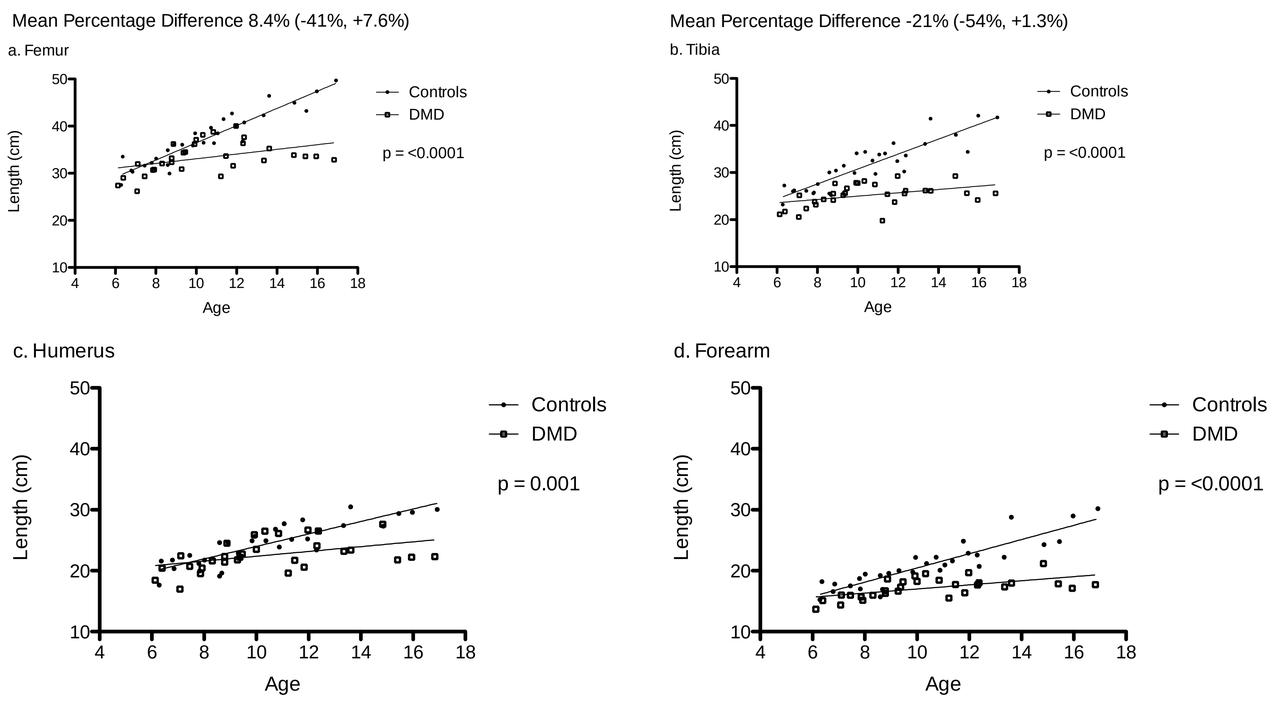
Results

Table 1: Baseline demographics of boys with DMD and healthy controls.

	Controls	DMD	p-value
	(n, 30)	(n, 30)	
Age	10.2 (6.3-16.9)	10.0 (6.1-16.8)	0.97
Pubertal stage (%)			0.02
- Prepubertal	17 (57%)	26 (87%)	
- Pubertal	13 (43%)	4 (13%)	
Steroid duration (years)		7.1 (1.3 to 15.2)	-
Testosterone treatment		2 (7%)	-
Bisphosphonate		10 (33%)	-
Vertebral fractures		14 (47%)	-
Non-ambulant		10 (33%)	-
Bone age (years)		9.5 (5.0-16)	-

Descriptive results were expressed as median (range).

Figure 2. Boys with DMD showed greater deficit in distal limb (tibia, forearm) length than proximal limb lengths when compared to healthy boys.



p-values are for differences between DMD and control β slopes
p-values remained significant after adjusting for age and pubertal status.

Figure 1. Boys with DMD showed greater deficits in leg length than sitting height, resulting in higher SH:LL than controls.

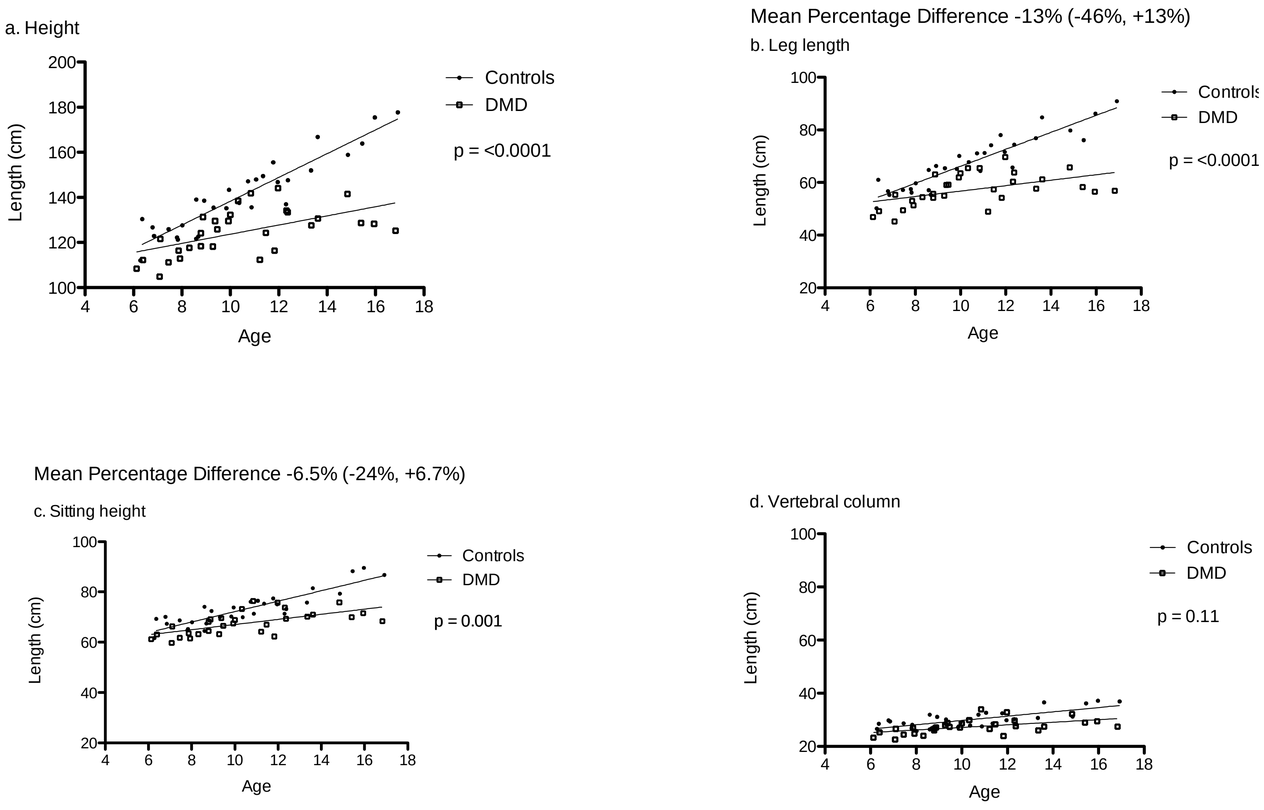


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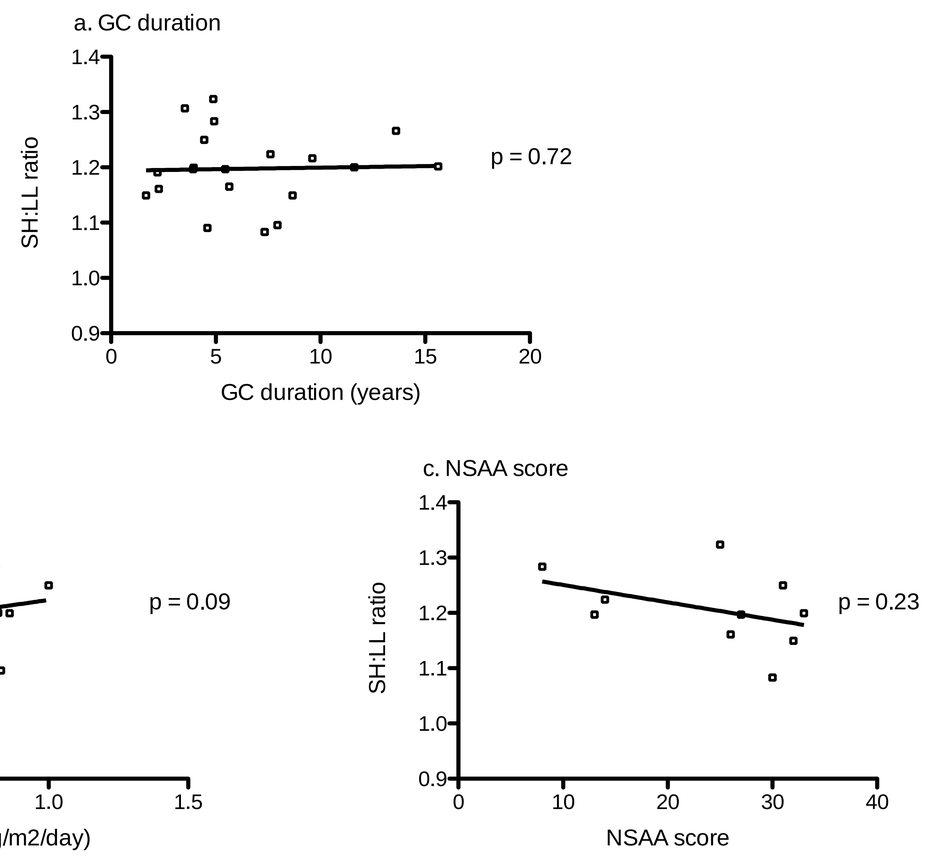
Figure 3. Body proportion of a subset of boys with DMD without contractures is not associated with glucocorticoid duration, dose or ambulatory score.



GC: Glucocorticoid; NSAA: NorthStar Ambulatory Assessment

Conclusion

1. **Growth impairment in GC treated boys with DMD was associated with skeletal disproportion.**
2. **Lower limbs and distal long bones were more affected than the spine.**

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