

Fractures in Boys with Duchenne Muscular Dystrophy and their Relationship To Age



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Objective

To identify the prevalence of fractures and to characterise length of steroid exposure, mobility status, pubertal status, vitamin D level and bone mineral content (BMC) within 1 year prior to sustaining fracture.

Method

A retrospective review of bone morbidity in a contemporary cohort of boys with Duchenne Muscular Dystrophy (DMD) currently managed in a Scottish tertiary neuromuscular centre.

Clinical details and results of bone surveillance were obtained in 47 boys, aged 9 years(2-16).

DXA bone mineral content (BMC) at total body (TB) and lumbar spine (LS) were adjusted for bone area.

Fractures were classified based on radiological confirmation. Results are reported in median (range).

Results

Demographics

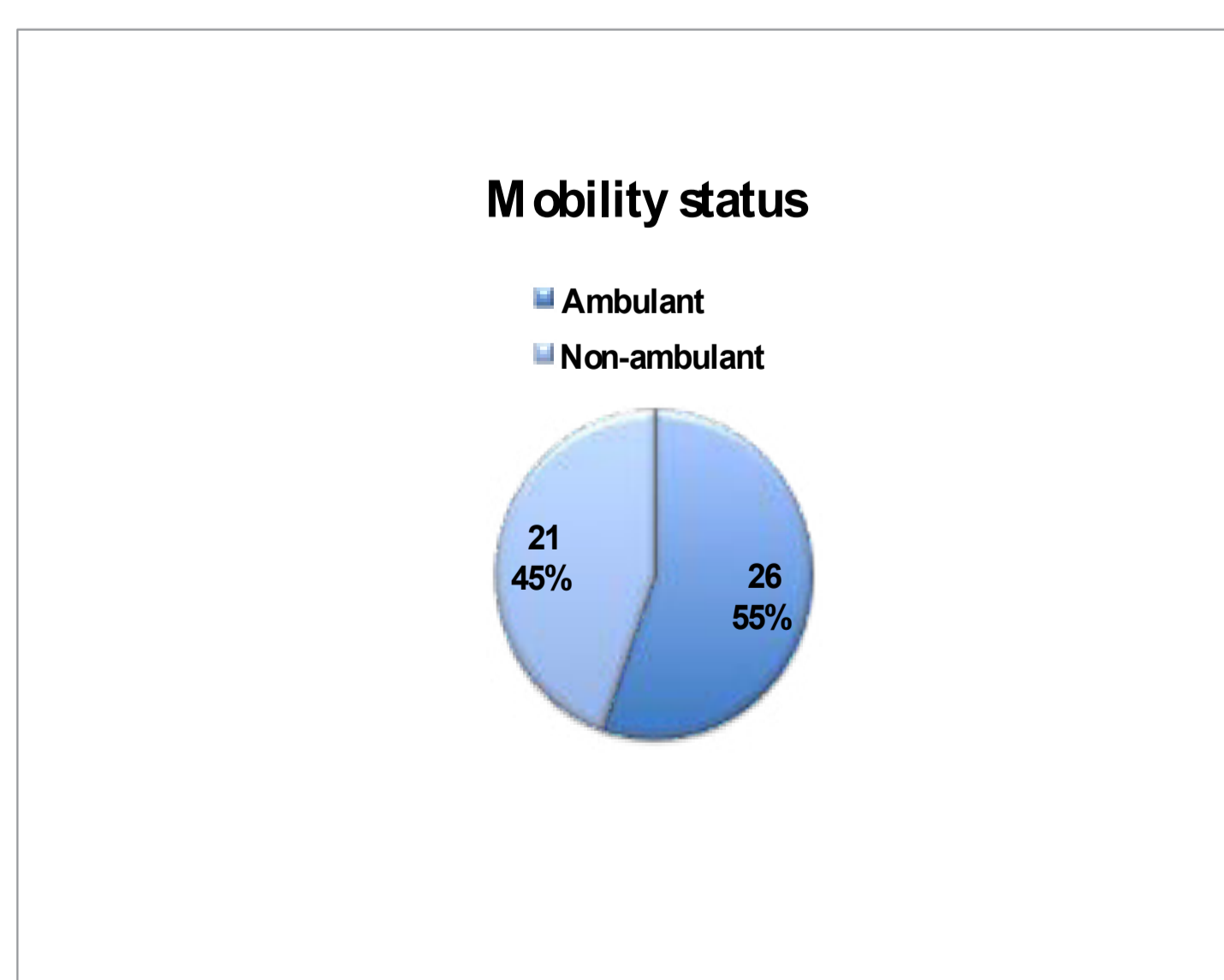
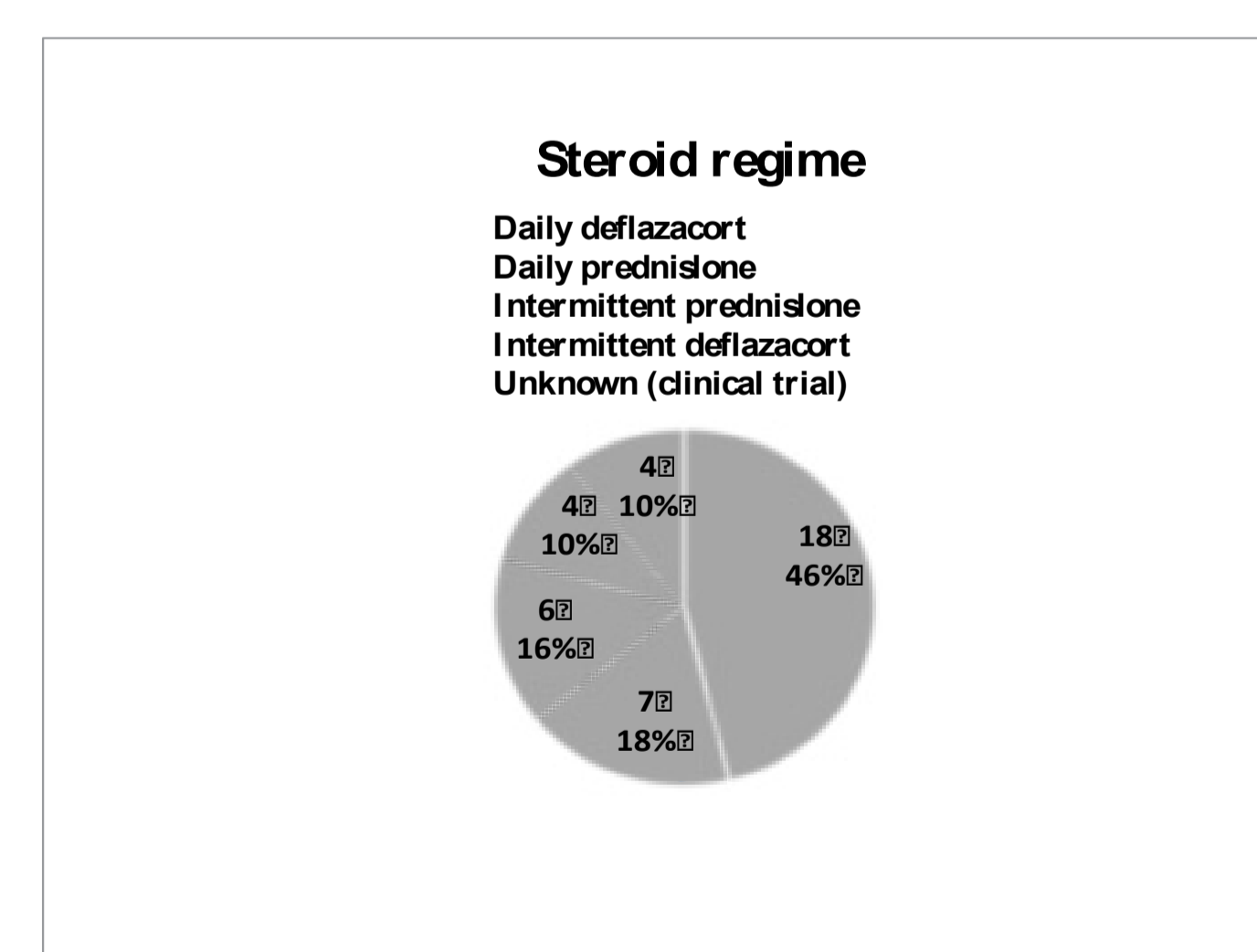
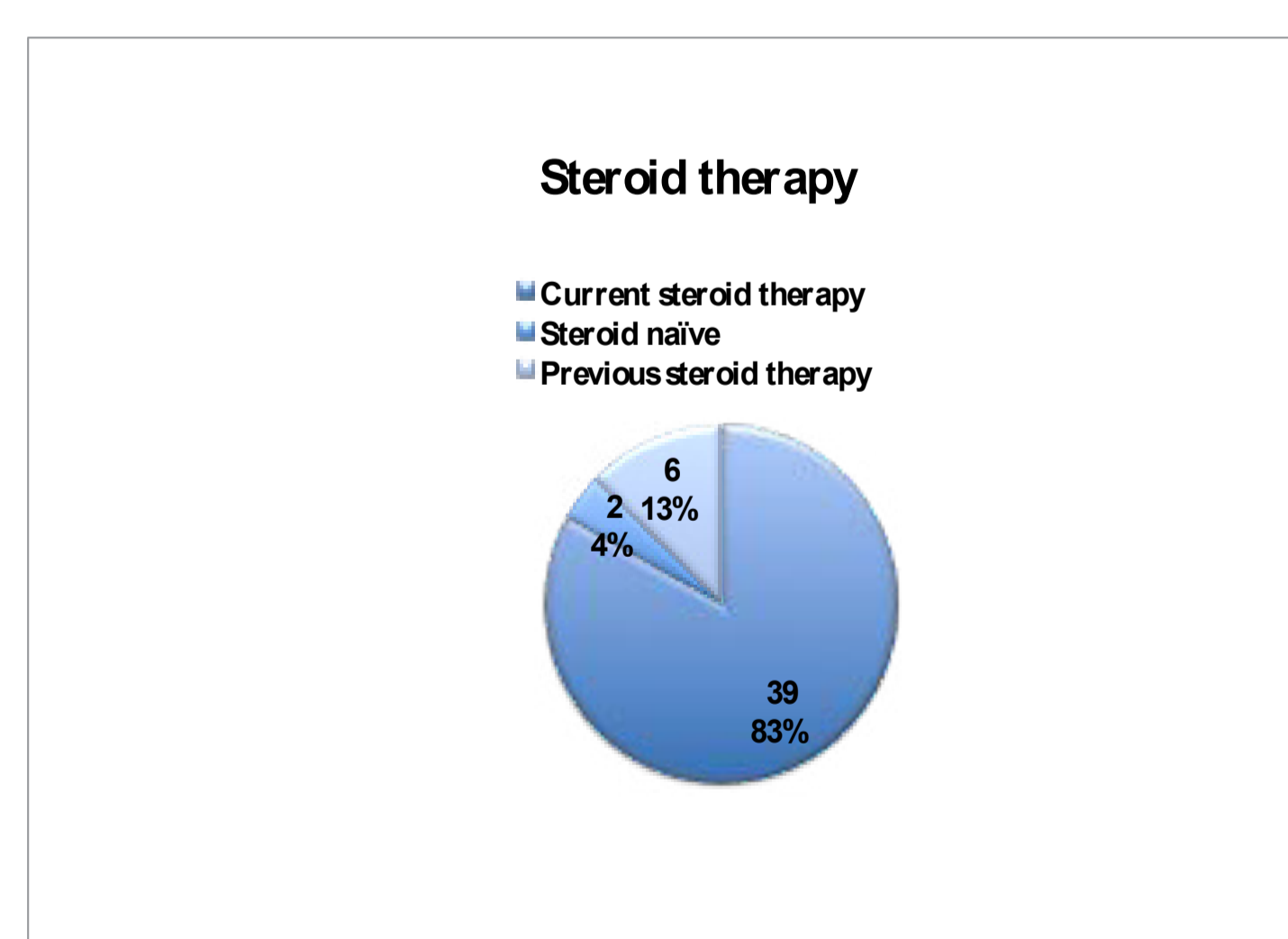


Table 1: Stages of disease and mobility

Stages	Functional ability/mobility
1 Presymptomatic	No gait disturbance
2 Early ambulatory	Waddling gait, Gower's positive, able to climb stairs
3 Late ambulatory	Increasingly laboured gait, loss of ability to rise from floor and climb stairs
4 Early non-ambulatory	Able to self propel and maintain posture on wheel chair
5 Late non-ambulatory	Loss of upper limb function and ability to maintain posture

Adapted from Diagnosis and management of DMD part1: Lancet Neurol 2010;9:77-93

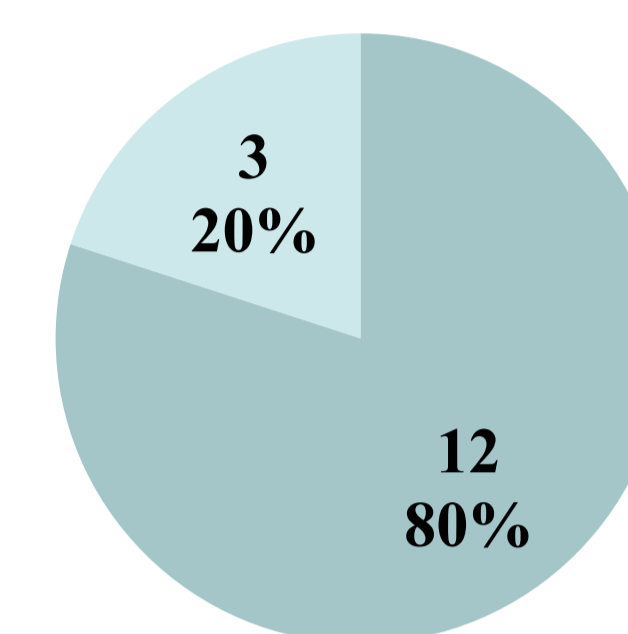
Delayed puberty

5/10(50%) of those over 14 had delayed puberty and all of these boys were treated with testosterone therapy.

Results continued

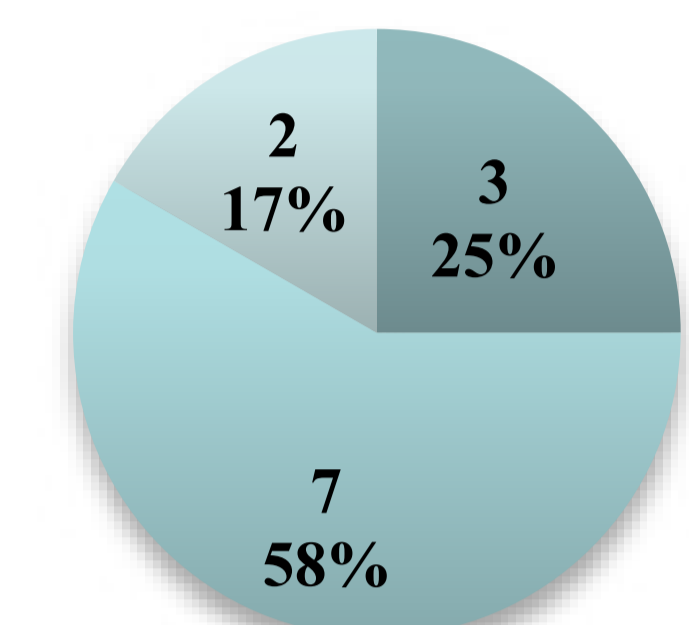
12/47 (26%) boys had sustained a total of 15 symptomatic fracture events.

Proportion of Appendicular vs. Vertebral Fractures



Appendicular fractures

- Femur
- Tibia/Fibula
- Humerus/Radius/Ulnar



Appendicular fractures

Patient	Age at fracture (years)	Length of steroid (years)	Mechanism of Injury	Fracture site	Mobility status	DXA TB BMC (SDS)	DXA LS BMC (SDS)	Vitamin D (nmol/L)	PTH (pg/mL)
1	2	0	Fall	Femur	Ambulant	N/A	N/A	N/A	2
2	2	0	Fall	Tibia	Ambulant	N/A	N/A	N/A	N/A
2	3	0	Fall	Tibia	Ambulant	N/A	-1.2	38	N/A
3	5	1	Fall	Humerus	Ambulant	-0.8	-0.1	21	6.3
4	6	1	Fall	Radius/ulnar	Ambulant	N/A	N/A	35	3.9
5	6	1	Fall	Tibia	Ambulant	-0.3	-0.2	34	4.3
6	8	4	Fall	Radius	Ambulant	1.0	-0.5	51	3.3
7	11	8	Fall	Tibia	Late ambulant	-0.1	1	28	3.6
8	12	7	Fall	Tibia	Non-ambulant	0.1	-0.8	<20	3
8	12	9	Fall	Fibula	Non-ambulant	0.7	0.4	41	2.7
9	13	8	Being lifted	Femur	Non-ambulant	0.3	-0.1	56	5.3
10	14	7	Fall	Femur	Ambulant	-0.5	-1.0	57	4.1

Vertebral fractures

Patient	Age at vertebral fracture (years)	Length of steroid (years)	Mechanism of injury	Mobility status	X-ray report	DXA TB BMC (SDS)	DXA LS BMC (SDS)	Vitamin D (nmol/L)	PTH (pg/mL)
1	9	5	Atraumatic	Ambulant	Multiple compression fracture at T9, T10 thoracic & all lumbar vertebrae	1.8	-1.3	59	2.9
2	11	6	Atraumatic	Late ambulant	Multiple compression fracture at lower thoracic & upper lumbar vertebrae	0	1.9	29	1.1
3	13	8	Atraumatic	Non-ambulant	Multiple compression fracture at lower thoracic & lumbar vertebrae	-0.5	-0.3	54	2.9

Conclusion

In boys with DMD, symptomatic vertebral fractures occur in older children, with longer duration of steroid therapy.

Appendicular fractures occur in younger boys and can also present in very young, ambulant, steroid naïve boys.

Coincidental severe Vitamin D deficiency or reduced BMC were not common findings at a fracture event.

