

Background

Delayed puberty is common in Duchenne muscular dystrophy (DMD) due to long-term glucocorticoid (GC) therapy. However, there has been no prospective study of testicular development in this cohort.

Objectives

To evaluate testicular development and function in DMD over a 12 month follow-up period from a prospective, longitudinal study.

Methods

23 boys with DMD with median age 12.4 (10.0, 16.8) years had pubertal assessment carried out by a single endocrinologist. Testes volume was converted to Z scores adjusted for bone age (TVBA-Z score). Plasma luteinising hormone (LH), follicle stimulating hormone (FSH) and testosterone levels were measured. Inhibin B levels were converted to Z scores adjusted for bone age.

For statistical analysis, cohort was split into:

Group A (pre-pubertal; G1 and testes <4ml),

Group B (virilised by testosterone therapy; G2+ but testes <4ml), and

Group C (spontaneous puberty; G2+ and testes 4+ml). Results expressed as median (range).

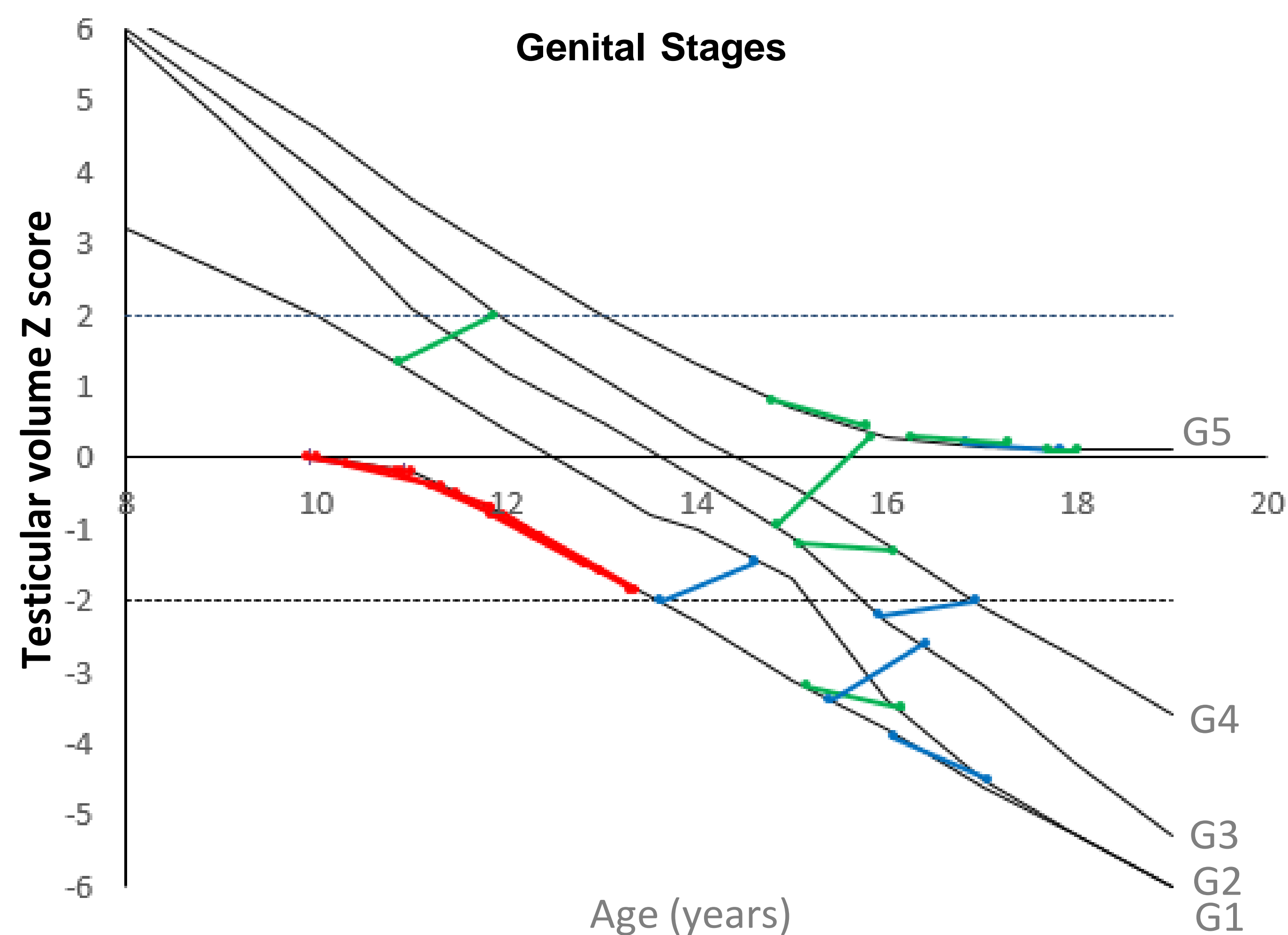
Results

	Group A	Group B	Group C
Number	11/23 (48%)	5/23 (22%)	7/23 (30%)
Age (years)	11.2 (10.0, 12.4)	15.9 (13.6, 16.8)	15.1 (10.9, 16.6)
Bone age delay (years)	0.3 (-1.3, 5.1)	4.6 (2.3, 7.1)	-0.2 (-2.4, 4.2)
Height velocity (cm/year)	3.2 (0.9, 6.4)	2.8 (0.3, 3.5)	1.0 (0.1, 7.9)

Cohort characteristics (n=23).

Group	Current GC Therapy		GC Regimen		Daily GC	
	Baseline	12 Months	Baseline	12 Months	Baseline	12 Months
Group A (n=11)	10/11 (91%)	11/11 (100%)	DFZ: 7/11 (64%)	DFZ: 7/11 (64%)	10/11 (91%)	10/11 (91%)
Group B (n=5)	5/5 (100%)	5/5 (100%)	DFZ: 5/5 (100%)	DFZ: 5/5 (100%)	5/5 (100%)	5/5 (100%)
Group C (n=7)	4/7 (57%)	4/7 (57%)	DFZ: 1/4 (25%)	DFZ: 1/4 (25%)	3/7 (43%)	4/7 (57%)

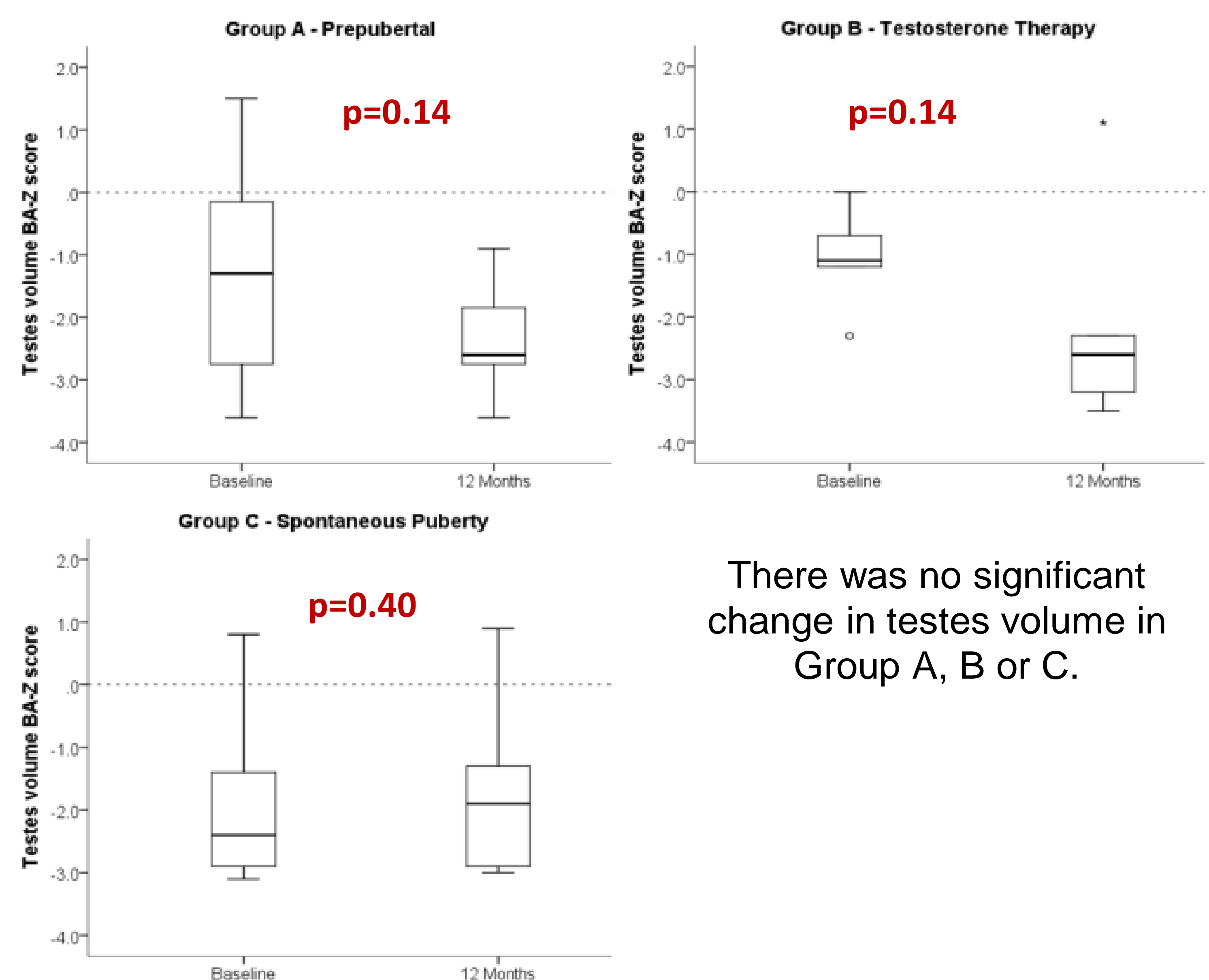
GC therapy at baseline and 12 months.
DFZ: Deflazacort.



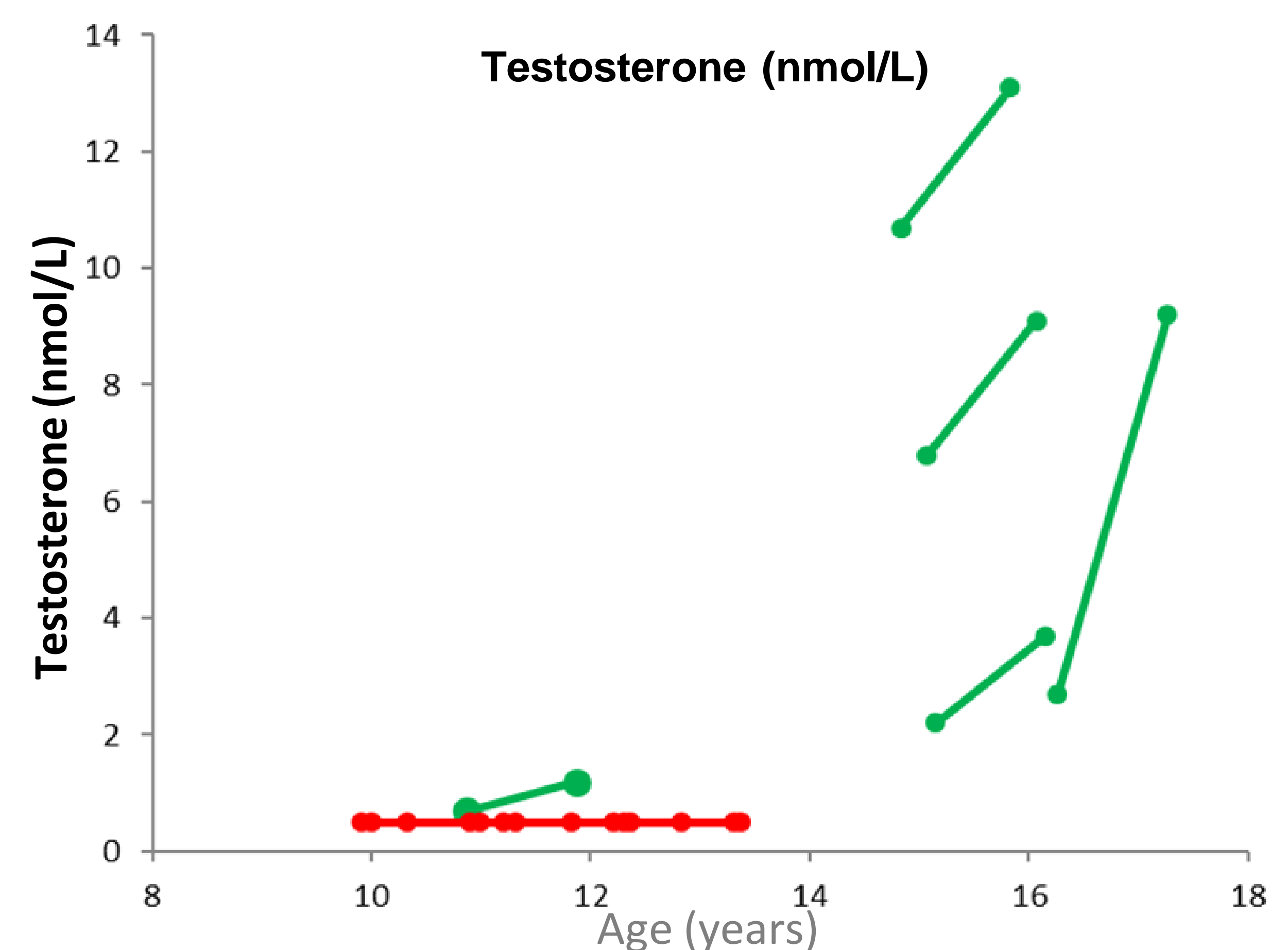
Genital stage change from baseline to 12 months.
Red: Group A. **Blue:** Group B. **Green:** Group C.

Results continued

Testes Volume Z Scores

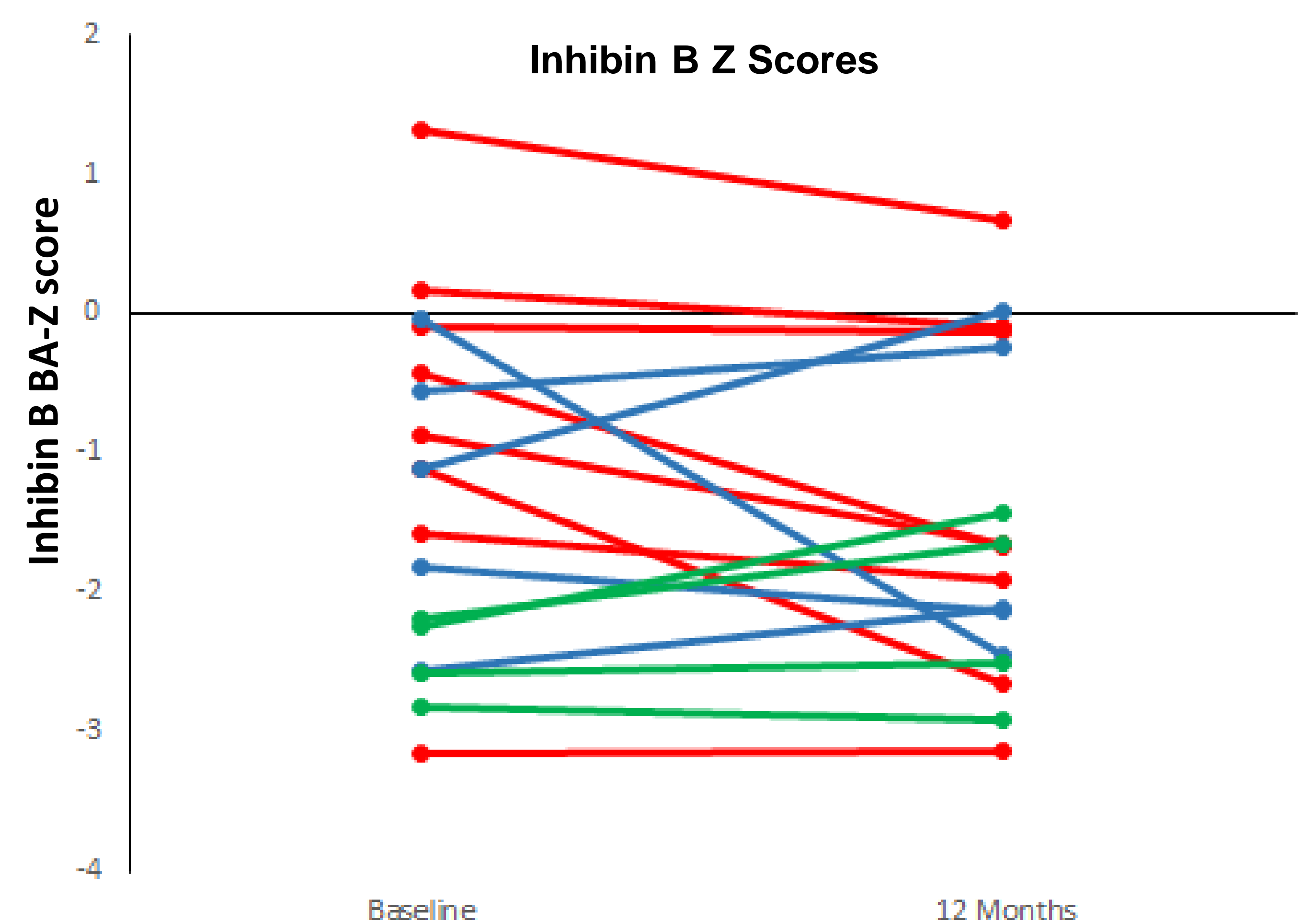


There was no significant change in testes volume in Group A, B or C.



Red: Group A: 9/11 (82%) had undetectable testosterone at baseline and 12 months. 8/11 (73%) & 9/11 (82%) had undetectable LH at baseline and 12 months

Green: Group C: 0/7 (0%) had undetectable testosterone at baseline and 12 months. None had undetectable LH at baseline and 12 months.



Inhibin B Z scores adjusted for bone age.

Red: Group A (p=0.03). **Blue:** Group B (p=0.89). **Green:** Group C (p=0.27).

Conclusion

- 1- DMD boys have relatively small testes when compared with healthy boys and adjusted for bone age.
- 2- Longer period of follow-up of puberty and further details of gonadal function in these boys are needed and underway.