



Bone status in relation to ambulatory performance in girls with Rett syndrome: a 10-year longitudinal study

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Table 1- Clinical characteristics, biochemical and ultrasonographic parameters at baseline, after 5 and 10 years in non-ambulatory and ambulatory Rett girls

	NON AMBULATORY (N = 28)			AMBULATORY (N = 30)		
	Baseline	5 years	10 years	Baseline	5 years	10 years
Age (yrs)	5.1 ± 2.1	9.7 ± 3.5	14.6 ± 3.4	5.9 ± 3.2	10.9 ± 3.4	15.8 ± 3.4
Birth Weight (Kg)	2.944 ± 0.350			3.143 ± 0.387*		
Weight (Kg)	19.2 ± 4.2	29.4 ± 5.2	33.9 ± 9.2	22.9 ± 12.1	31.8 ± 13.6	38.8 ± 13.6
Height (cm)	104.9 ± 14.1	129.1 ± 8.5	140.6 ± 10.6	113.5 ± 21.5	131.9 ± 14.4	142.7 ± 8.2
BMI (Kg/m ²)	16.3 ± 0.1	16.7 ± 1.3	16.8 ± 2.9	16.4 ± 3.5	17.4 ± 4.6	18.8 ± 5.2*
Waterlow score [#]	16/28 (57.1%)	20/28 (71.4%)	16/28 (85.7%)	9/30 (30.0%)*	19/30 (63.3%)	23/30 (76.6%)
Age Menarche (yrs)	11.7 ± 2.8			11.9 ± 2.4		
Calcium (mg/dl)	9.5 ± 0.5	9.4 ± 0.6	9.6 ± 0.4	9.5 ± 0.5	9.6 ± 0.6	9.5 ± 0.5
Phosphorous (mg/dl)	5.2 ± 0.8	5.0 ± 0.7	4.9 ± 0.6	5.0 ± 0.7	5.1 ± 0.6	4.8 ± 0.6
25OHD (ng/ml)	15.2 ± 13.2	24.3 ± 14.7	28.1 ± 18.9	16.2 ± 10.8	26.7 ± 16.8	26.5 ± 16.3
AD-SoS (m/s)	1805.5 ± 43.1	1846.9 ± 21.2	1865.3 ± 36.8	1840.5 ± 137.2*	1886.5 ± 88.5*	1906.2 ± 66.6*
AD-SoS Z-score	-2.27 ± 1.06	-2.39 ± 1.14	-3.15 ± 0.30	-1.59 ± 1.67*	-1.96 ± 1.87*	-2.68 ± 1.40*
BTT (µs)	0.35 ± 0.19	0.60 ± 0.18	0.53 ± 0.19	0.51 ± 0.23*	0.73 ± 0.27	0.88 ± 0.22*
BTT Z-score	-1.91 ± 0.30	-2.01 ± 0.15	-3.27 ± 0.28	-1.10 ± 1.78*	-1.52 ± 2.84*	-2.68 ± 1.70*

Values expressed as mean ± SD

[#] Categorical variable (Chi-squared test)

*p<0.05 between non ambulatory versus ambulatory groups

Figure 1- Percent change from baseline of AD-SoS Z-score in non-ambulatory and ambulatory Rett girls

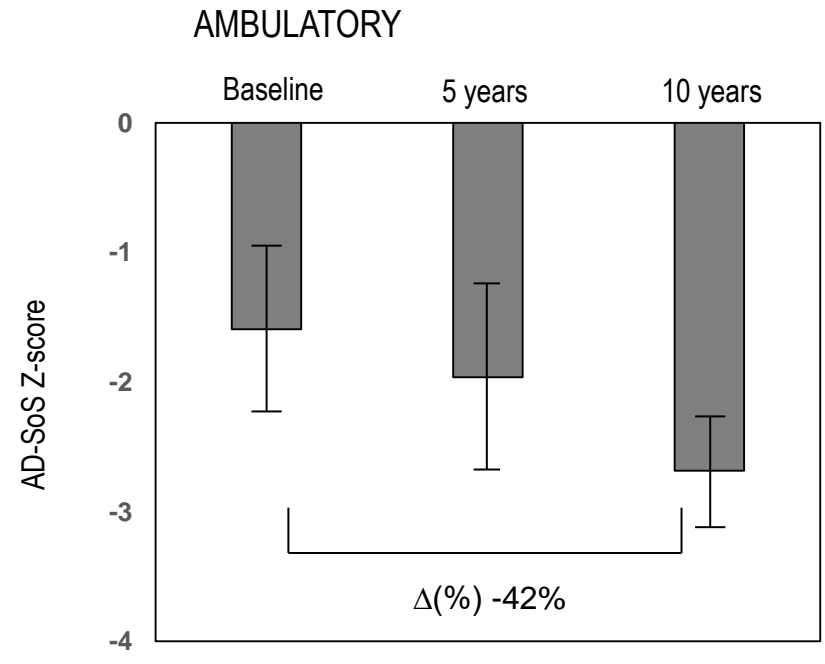
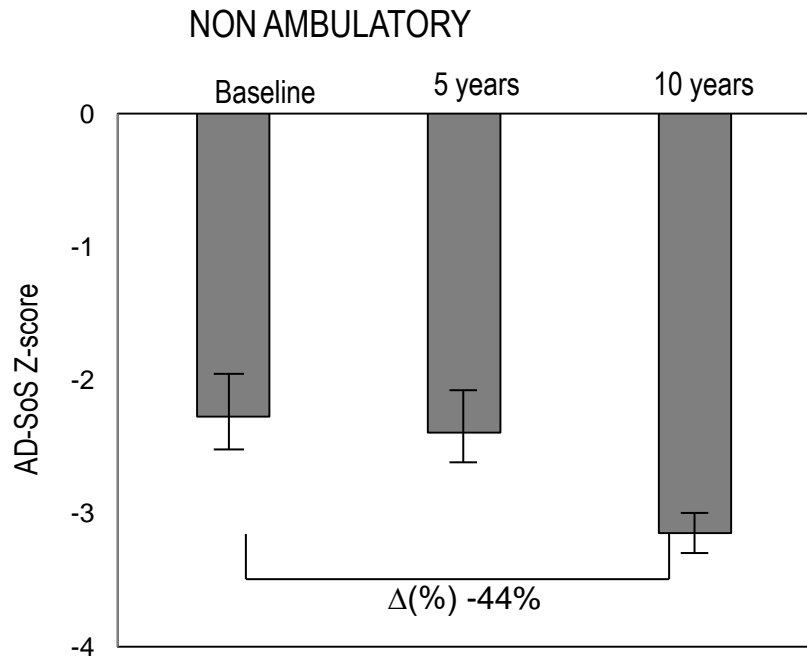


Figure 2 - Percent changes from baseline of BTT Z-score in Non-ambulatory and ambulatory Rett girls

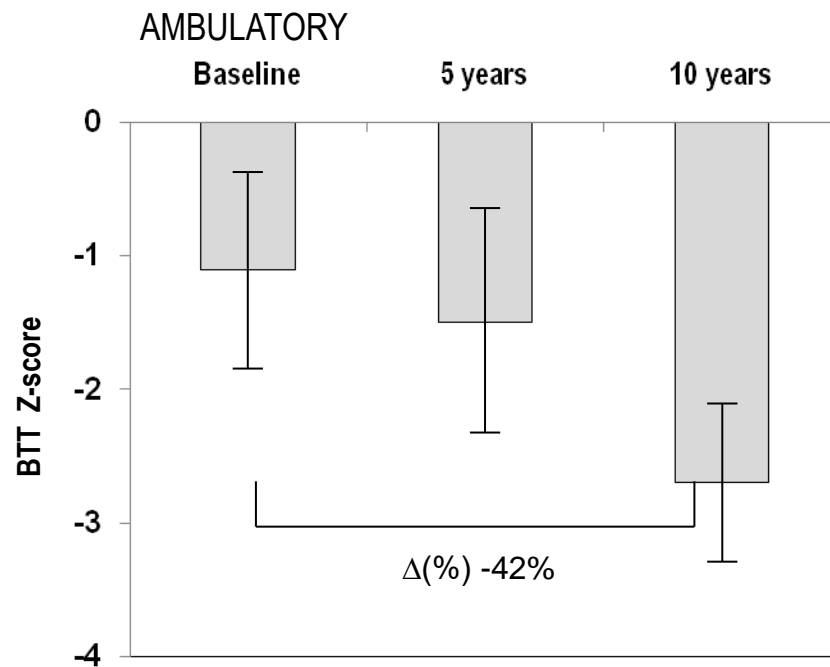
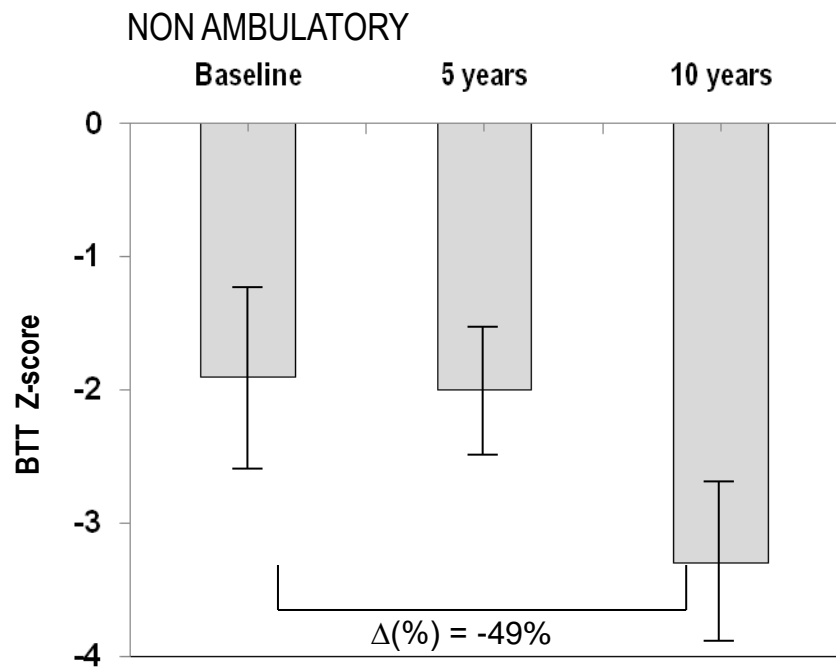


Table 2 . Multiple linear regression analysis of predictors of QUS parameters in Rett girls at baseline and after 5 and 10 years

	Variable	Unstandardized coefficient, b	95%CI	p
Baseline	AD-SoS (m/s)			
	Height (cm)	4,352	2,105 to 6,600	<0,0001
	Weight (Kg)	5,786	0,99 to 10,58	0,019
	BTT (μs)			
	Height (cm)	0,009	0,006 to 0,012	<0,0001
5 years	AD-SoS (m/s)			
	Height (cm)	5,188	2,867 to 7,509	<0,0001
	Weight (Kg)	4,180	1,547 to 6,814	0,003
	Ambulatory	43,874	5,453 to 82,294	0,026
	BTT (μs)			
	Height (cm)	0,011	0,006 to 0,015	<0,0001
10 years	AD-SoS (m/s)			
	Ambulatory	51,298	12,933 to 89,662	0,010
	BTT (μs)			
	Height (cm)	0,029	0,002 to 0,056	0,036

Whole set of variables included into the models: age, weight, height, 25OHVitD, fracture, scoliosis, ambulatory capacity, nutritional status