

USEFULNESS OF MYOCARDIAL STRAIN IMAGING IN SPINAL MUSCULAR ATROPHY

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Introduction

- Patients with Spinal Muscular Atrophy (SMA) may have cardiac impairment. Main cardiac dysfunction in these patients has been related to **autonomic dysfunction**.
- Cardiac dysfunction in this group of patients could be related to increased survival associated with the best ventilatory control and better management of the deformities of the rib cage.
- This study assesses the myocardial function with strain of pediatric patients with SMA

Material and method

- Prospective study of cases and controls. Patients diagnosed with SMA, period 2013-2015, with controls matched for age and sex are included.
- Variables systolic / diastolic function echocardiographic analysis are collected as well as Strain pattern.
- Statistical analysis: T Student or ANOVA

Results

- We include 31 case. Median age 7,2 years (0 – 12 years), 87% SMA type II. 29 controls.

Variables	SMA (n=31)	Controls (n=29)	p
HR (bpm)	96 ± 12	73 ± 11.2	0.014
Systolic PA (mmHg)	118 ± 11	115 ± 11	0.3
Diastolic PA(mmHg)	71 ± 11	68 ± 13	0.089
Body surf (m ²)	1.8 ± 0.13	1.5 ± 0.16	0.068
BMI (kg/m ²)	22.7 ± 2.1	21.1 ± 1.9	0.18
LVEF %	61.4 ± 2.8	64.8 ± 4.2	0.04
LVTd mm	44.8 ± 2.4	41.2 ± 3.8	0.51
LVTsd mm	26.4 ± 2.5	25.8 ± 3.1	0.47
IVSs mm	9.2 ± 1.3	9.0 ± 0.9	0.26
Max volumen LA ml	17.5 ± 2.5	21.5 ± 3.1	0.04

Variables	SMA	Controls	P
GLS VI %	16.2 ± 3.3	19.8 ± 2.1	0.08
Gtime to peak (ms)	504 ± 58	463 ± 44	0.005
A m/s	0.98 ± 0.18	0.7 ± 0.19	0.044
E m/s	1.1 ± 0.2	1.2 ± 0.18	0.012

Fig 2. Mean values of variables in cases and controls (GLS: Global longitudinal strain; Gtime: global time)

Fig 1. Mean values of variables in cases and controls

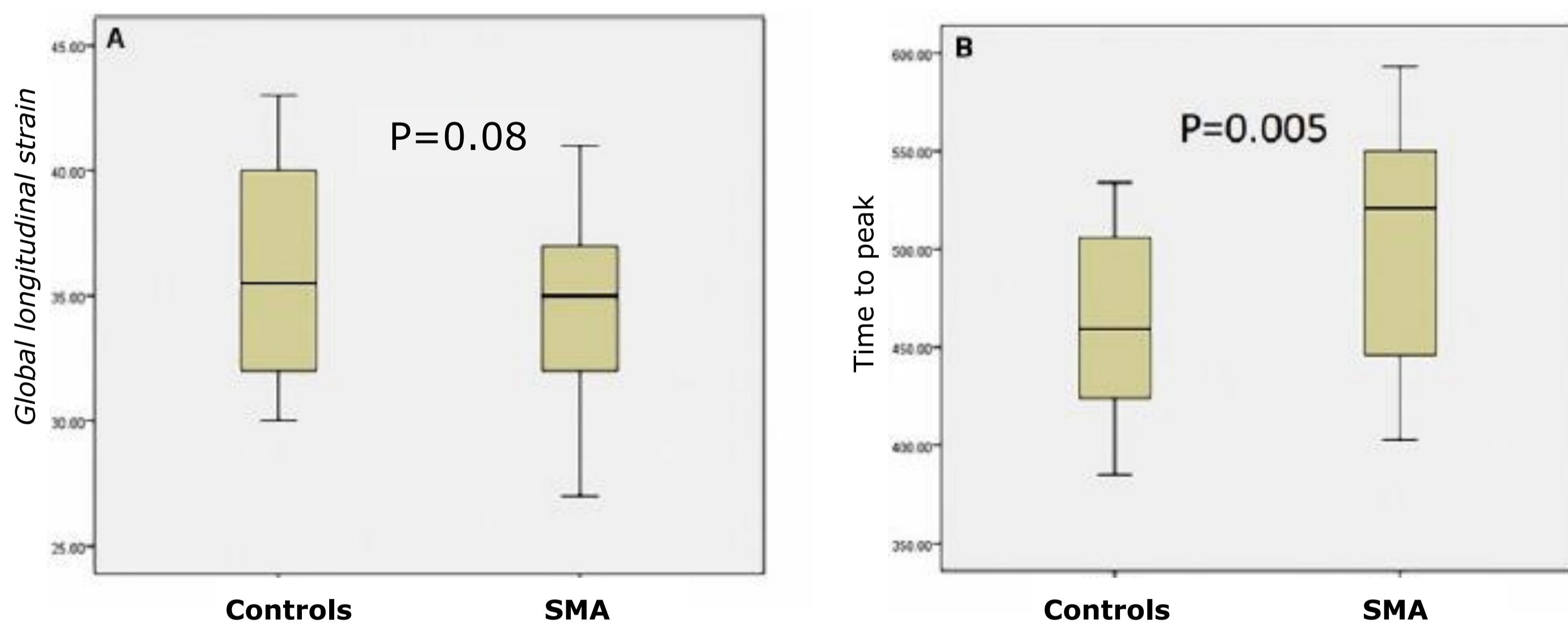


Fig 3. Comparison longitudinal strain values and time to peak in SMA patients vs controls

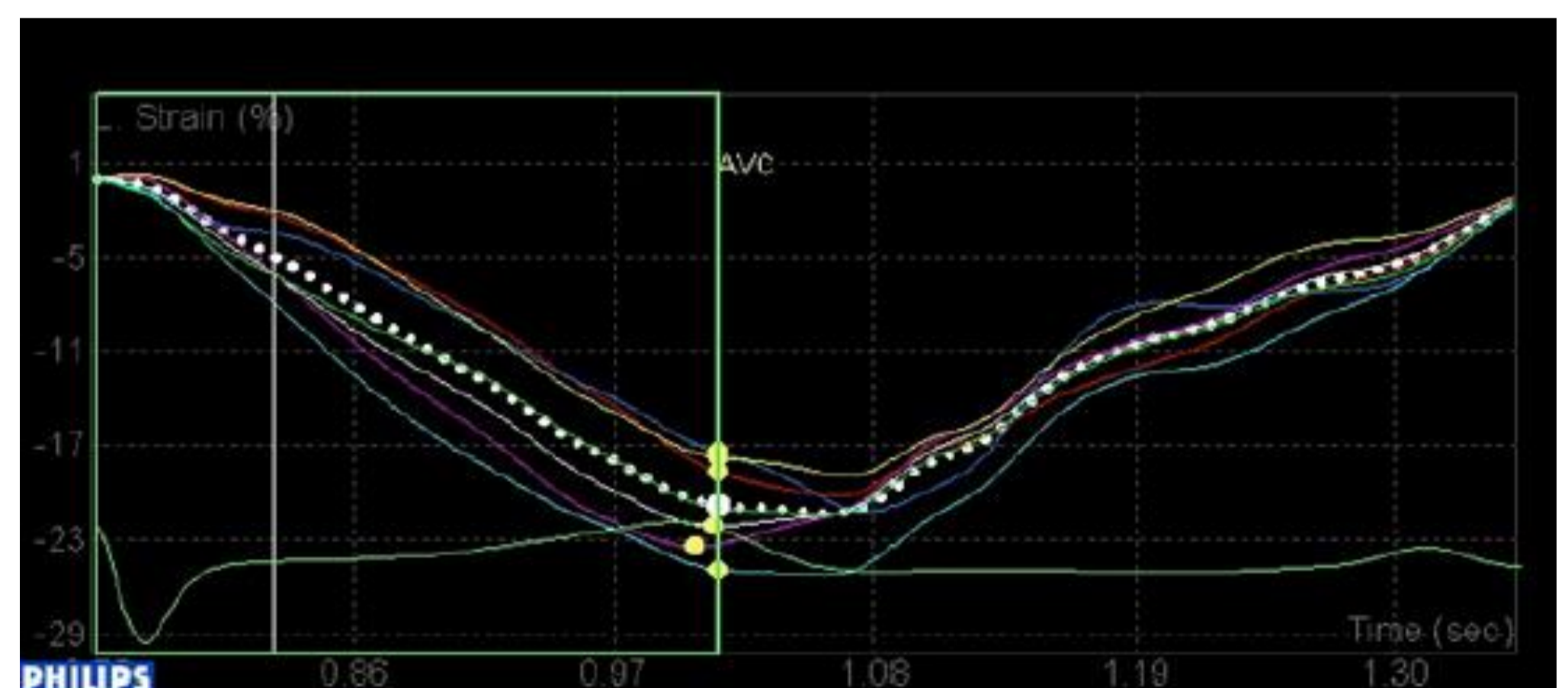
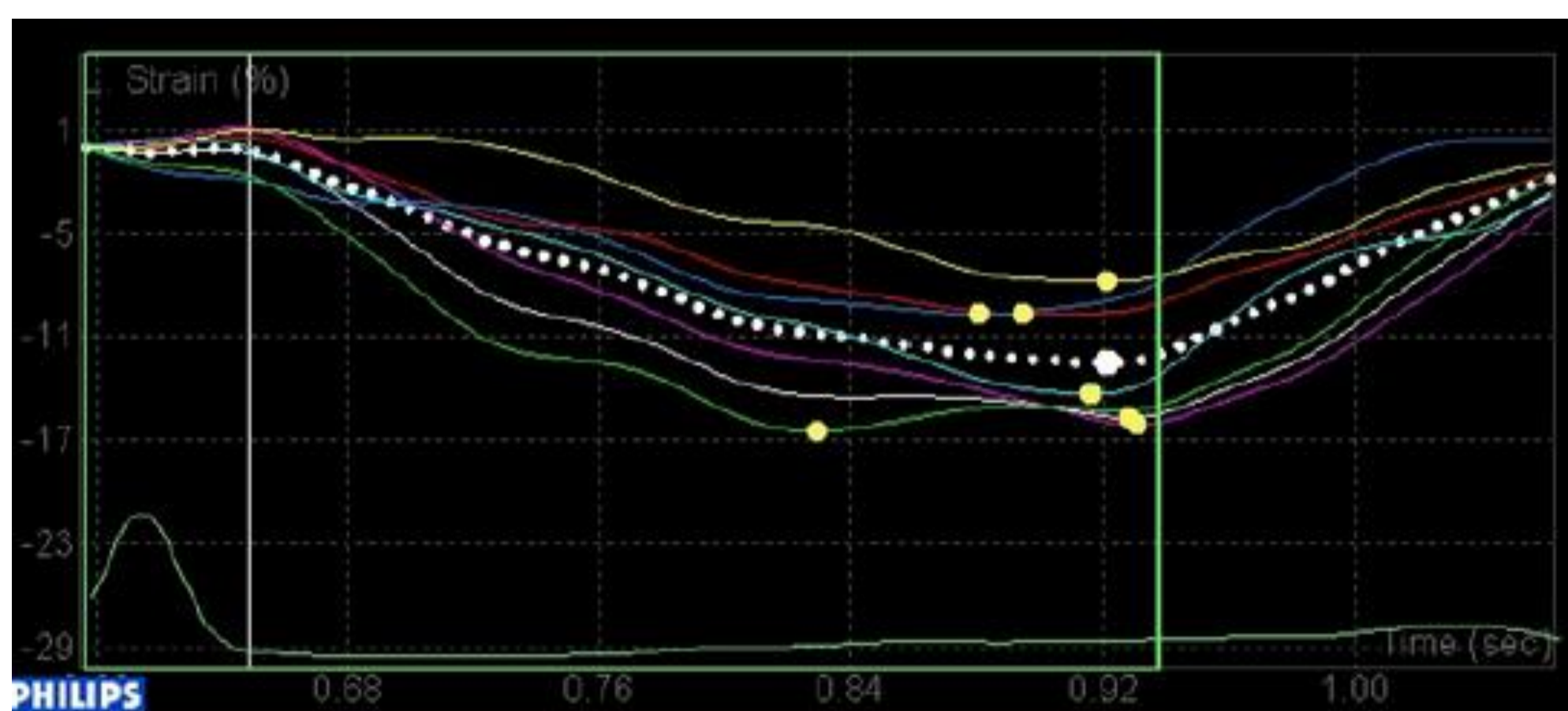


Fig 4. Comparison strain and dispersion pattern between SMA (left) and controls

Discussion

- Children with SMA have significant reductions in parameters of deformation (strain) despite maintaining a global systolic function.
- The pattern of strain could be considered in the diagnosis and monitoring of disease.
- The prognostic value of these findings needs further time tracking of this patient's group