Dysfunctional Mitochondria in cardiac fibres of a Williams-Beuren Syndrome Mouse Model

Noura Abdalla , Gloria Garrabou, Alejandro Gonzalez, Ester Tobías-Baraja, Gustavo Egea and Victoria Campuzano

Supplemental Tables

Supplemental Table S1: Statistical data of Oxygen consumption and ATP production

Unpaired t-test- Multiple comparisons Holm-Sidak method				
	t-ratio	**df**	**P value**	**Adjusted P value**
LEAK	1.88927	10	0.088168	0.088168
OXPHOS CI	2.66538	10	0.023680	0.046799
OXPHOS CI+CII	3.63116	10	0.004604	0.013748
OXPHOS CIV	8.10377	9	0.000020	0.000100
ETC CII	4.37771	10	0.001382	0.005518
Unpaired t-test				
	t-ratio	**df**	**P value**	
RCR	3.470	10	0.0060	
MEFs-ATP	4.315	4	0.0125	

Supplemental Table S2: Statistical data of genomic copy number and OXPHOS complex

Unpaired t-test: genomic copy number				
	t-ratio	**df**	**P value**	
Cardíac fibers	2.658	9	0.0261	
MEFs	4.066	8	0.0036	
Unpaired t-test- Multiple comparisons Holm-Sidak method				
	t-ratio	**df**	**P value**	**Adjusted P value**
CI-NDUFB8	5.038	7	0.0015	0.0075
CII-SDHB	4.021	7	0.0050	0.0125
CIII-UQCRC2	4.425	7	0.0031	0.0122
CIV-MTCO1	3.051	6	0.0225	0.0225
CV-ATP5A	4.169	7	0.0042	0.0125

Supplemental Table S3: Statistical data of mitochondrial morphology

Unpaired t-test			
	t-ratio	**df**	**P value**
Density	2.626	6	0.0393
Average Size	3.321	6	0.0160
Circularity	2.865	6	0.0286

Supplemental Table S4: Statistical data of mitochondrial dynamics

Unpaired t-test- Multiple comparisons Holm-Sidak method				
	t-ratio	**df**	**P value**	**Adjusted P value**
L-OPA1	3.387	12	0.005400	0.010632
S-OPA1	3.394	12	0.005330	0.010632
Unpaired t-test				
	t-ratio	**df**	**P value**	
FIS1	4.887	7	0.0018	
MTF1	3.859	8	0.0048	
MTF2	0.205	7	0.8431	

Supplemental Table S5: Primer sequences for genotyping and genomic copy number

Gene	**Sequence**	**Tm ºC**
HPRT	5'-CTCTGAGGCTTCAAAGGTTC-3'	56.7
	5'-AATCCAGCTTGTTTGGGCTA-3'	59.7
TRDC	5'-CAAATGTTGCTTGTCTGGTG-3'	57.7
	5'-GTCAGTCGAGTGCACAGTTT-3'	56.4
GADPH	5′-ATGACTCCACTCACGGCAAAT-3′	61.9
	5′- GGGTCTCGCTCCTGGAAGAT-3′	63.0
mt-ND1	5'-GGATCCGAGCATCTTATCCA-3'	60.0
	5'- GGTGGTACTCCCGCTGTAAA-3'	60.0