Supplementary information for

Nusinersen rescues taurine deficiency in patients with type 1 Spinal Muscular Atrophy

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Suppl. Table 1. Age-dependent decrease of taurine levels in the CNS of wild type (WT) and SMA mice from P1 to P12.

		[Taurine] median (IQR) P1	[Taurine] median (IQR) P5	[Taurine] median (IQR) P12	Kruskall Wallis *p*-value	Dunn's multiple comparisons with Bonferroni correction *p*-value
Brainstem	WT	180.3 (158.1-191.6)	112.2 (101.7-136.6)	93.13 (83.75-103.2)	< **0.0001**	P1 vs P12: p = **0.0005**
	SMA	161.3 (157.9-181.9)	114.6 (101.8-144.0)	71.23 (70.46-82.28)	**< 0.0001**	P1 vs P12: p < **0.0001**
Spinal cord	WT	122.6 (96.94-106.5)	94.26 (93.74-109.2)	71.73 (61.64-84.17)	< **0.0001**	P1 vs P12: p = **0.0011**
	SMA	144.2 (106.5-154.7)	96.35 (87.36-108.7)	67.62 (65.46-76.40)	**< 0.0001**	P1 vs P12: p = **0.0002**
Cortex	WT	241.1 (231.9-248.5)	227.4 (206.4-235.7)	160.0 (153.8-170.6)	< **0.0001**	P1 vs P12: p = **0.0004**
	SMA	241.9 (207.7-254.0)	216.5 (200.8-223.9)	158.9 (152.3-165.1)	< **0.0001**	P1 vs P12: p = **0.0009**
Cerebellum	WT	176.5 (155.4-230.3)	156.2 (147.5-169.6)	131.4 (123.4-146.5)	**0.0009**	P1 vs P12: p = **0.0023**
	SMA	181.3 (155.0-199.7)	138.6 (134.1-156.1)	125.3 (122.1-129.0)	**0.0010**	P1 vs P12: p = **0.0023**

Taurine concentration is expressed as nmol/mg of protein and reported as median (IQR).

Suppl. Table 2. Gastrostomy, NIV, or tracheostomy in SMA1 or SMA2 patients does not influence taurine concentration in the CSF.

	Non-invasive ventilation					**Tracheostomy**					**Gastrostomy**				
	No		Yes			No		Yes			No		Yes		
	N	Median (IQR)	N	Median (IQR)	*p-value*	N	Median (IQR)	N	Median (IQR)	*p-value*	N	Median (IQR)	N	Median (IQR)	*p-value*
SMA1 (Taurine (µM))	7	8.60 (5.84-9.17)	6	6.06 (4.94-10.78)	0.836	8	7.67 (5.54-9.98)	5	5.84 (5.69-8.60)	0.524	3	8.92 (6.15-9.17)	10	6.13 (4.94-10.78)	0.573
SMA2 (Taurine (µM))	9	9.57 (8.98-16.31)	5	12.82 (12.82-17.45)	0.518										

Data are expressed as median (IQR). *P*-values refer to two-sided Mann-Whitney test.