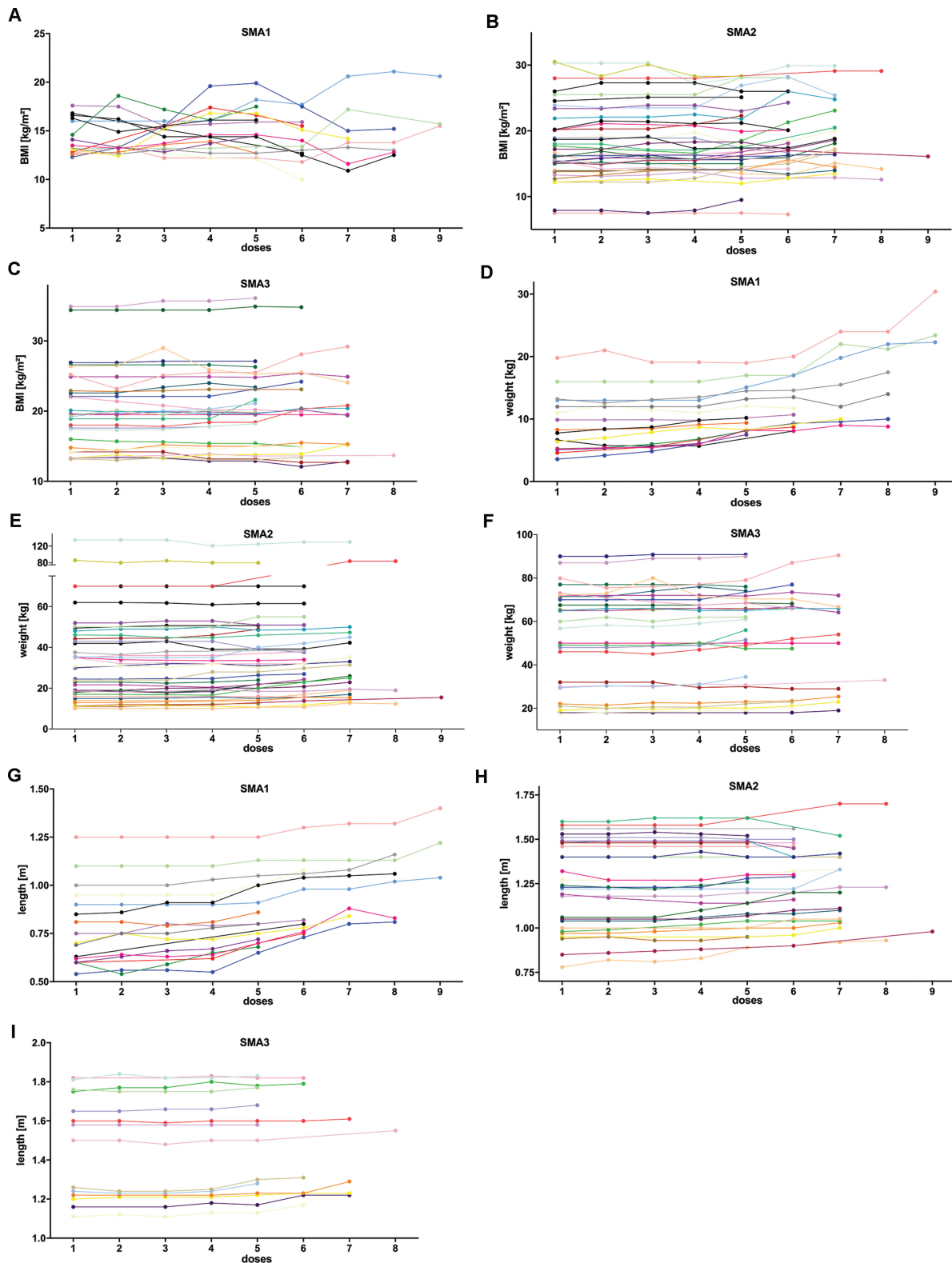
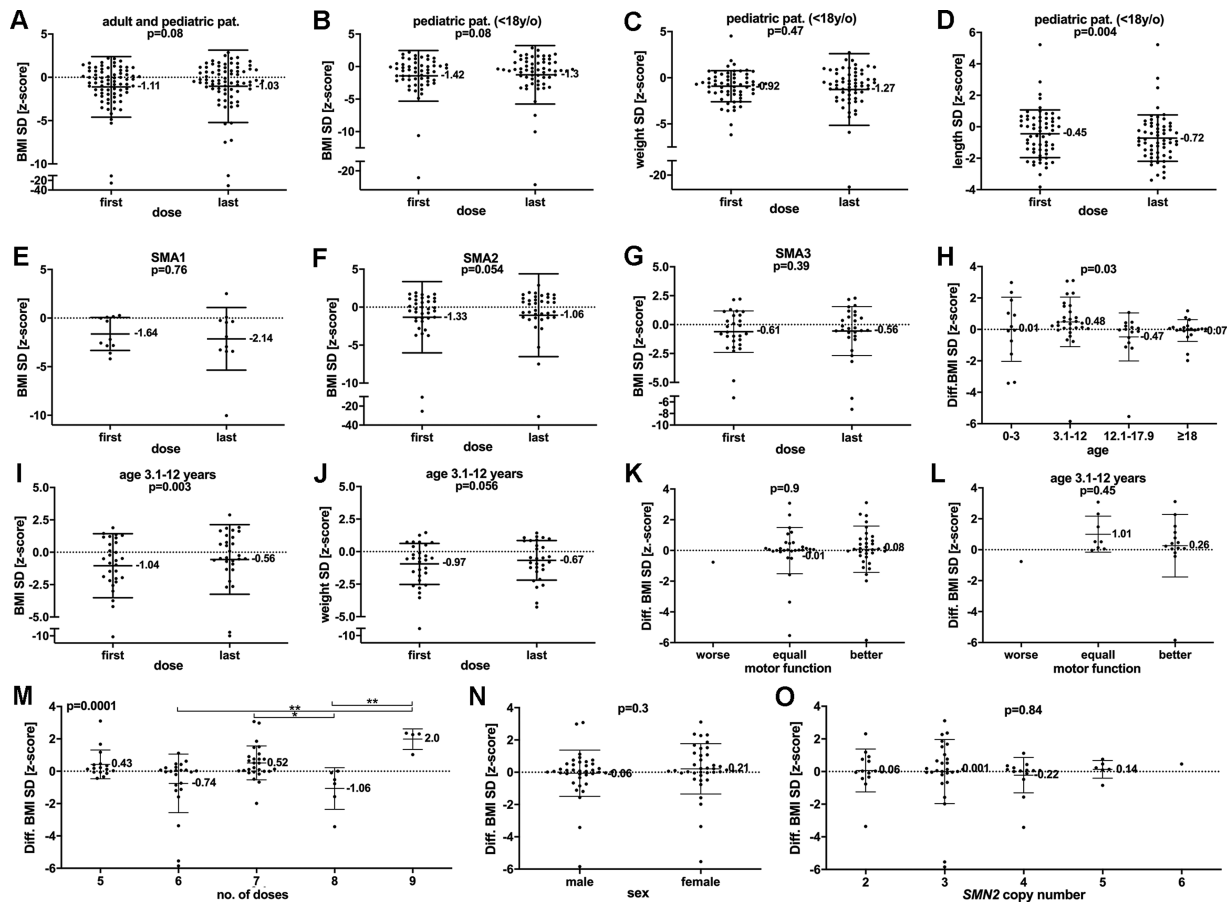


Supplementary Material



Supplementary Fig. S1 BMI/weight/length development of SMA1 to 3. (A) BMI development of patients with SMA1, (B) SMA2, and (C) SMA3. (D) Weight development of patients with SMA1, (E) SMA2, and (F) SMA3. (G) Length development of patients with SMA1, (H) SMA2, and (I) SMA3. BMI, body mass index; SMA, spinal muscular atrophy.



Supplementary Fig. S2 Standard deviations. (A) The BMI SD of adult and pediatric patients increased under treatment with nusinersen ($n = 76$; Wilcoxon’s test; $p = 0.08$), and also in (B) patients <18 years increased under treatment with nusinersen ($n = 56$; Wilcoxon’s test; $p = 0.08$). (C) Weight SD of patients <18 years were not significantly increased when comparing first and last injection point (Wilcoxon’s test; $p = 0.47$). (D) Length SD of patients <18 years were significantly decreased when comparing first and last injection time point (Wilcoxon’s test; $p = 0.004$). (E) BMI SD were not higher in SMA1 (Wilcoxon’s test; $p = 0.76$), (F) SMA2 (Wilcoxon’s test; $p = 0.054$) or (G) SMA3 (Wilcoxon’s test; $p = 0.39$) when comparing first and last injection point. (H) Diff. BMI SD trend did correlate with age of patients at first dose (Kruskal–Wallis test; $p = 0.03$). (I) Body mass index SD increased significantly in the subgroup of patients between 3.1 and 12 years (Wilcoxon’s test; $p = 0.003$), but (J) weight SD merely showed a trend toward an increase (Wilcoxon’s test; $p = 0.056$). (K) Diff. BMI SD trend did not correlate with motor function changes ($n = 64$; Wilcoxon’s test; $p = 0.9$) of the whole cohort or (L) in children between 3.1 and 12 years of age ($n = 22$; Wilcoxon’s test; $p = 0.45$). (M) Diff. BMI SD trend did correlate with the number of doses given (Kruskal–Wallis test, $p = 0.0001$; * $p < 0.05$; ** $p < 0.01$), but not with (N) sex (Wilcoxon’s test; $p = 0.3$), or (O) SMN2 copy number ($n = 61$; Kruskal–Wallis test; $p = 0.84$). BMI, body mass index; Diff. BMI SD, Difference between BMI SD prior to and under treatment; SD, standard deviation; SMA, spinal muscular atrophy; SMN, survival motor neuron gene.

Supplementary Table S1 Overview about body mass index/weight/length z-scores of SMA1 to 3 of our cohort and natural cohorts

		This study			Natural cohorts		
		Prior treatment		Last contact		Bertoli et al (2017) ²²	Baranello et al (2020) ¹⁴
		Mean (SD)	P25/P50/P75	Mean (SD)	P25/P50/P75	Mean (SD)	P25/P50/P75
BMI (z-score)	SMA1	-1.6 (1.7)	-2.9/- 2.2/0.08	-2.1 (3.2)	-3.4/- 1.9/- 0.3	-6.3 (4.4) -2.1 (2.3)	-0.57/- 1.99/- 0.86 -2.52/- 1.72/- 0.52
	SMA2	-1.3 (4.7)	-1.7/- 0.2/1.0	-1.1 (5.4)	-1.4/0.4/1.2		
	SMA3	-0.6 (1.8)	-1.7/- 0.7/0.8	-0.56 (2.1)	-1.1/- 0.4/0.8		
Weight (z-score)	SMA1	-0.8 (1.1)	-1.5/- 0.6/0.07	-0.9 (1.7)	-2.1/- 0.7/0.4	-2.7 (1.6) -1.4 (1.3)	-1.96/- 1.21/- 0.24 -2.11/- 0.90/- 0.22
	SMA2	-1.04 (1.95)	-2.6/- 0.96/0.5	-1.6 (4.8)	-2.4/- 0.9/0.6		
	SMA3	-0.7 (1.97)	-1.5/- 0.5/0.69	-0.6 (2.2)	-0.99/- 0.5/0.7		
Length (z-score)	SMA1	0.3 (1.2)	-0.2/0.5/0.8	0.06 (1.4)	-1.1/0.03/0.6		-0.12/0.76/1.57 -1.41/- 0.13/1.04
	SMA2	-0.9 (1.7)	-2.09/- 1.4/0.1	-1.3 (1.6)	-2.2/- 1.5/- 0.6		
	SMA3	-0.4 (0.95)	-1.1/- 0.2/0.5	-0.4 (0.77)	-1.0/- 0.3/0.4		

Abbreviations: BMI, body mass index; P, centile; SD, standard deviation; SMA, spinal muscular atrophy.