

Table S1. Correlation and comparison between ultrasound measurements in the entire cohort.

Total cohort (<i>n</i> = 36)	Correlation	<i>p</i> -value
CSA tibial nerve left – CSA tibial nerve right	0.912 (<i>< 0.001</i>)**	<i>Z</i> = -1.440; <i>p</i> = 0.150
Microvascular blood flow left – Microvascular blood flow right	0.540 (<i>< 0.001</i>)**	<i>Z</i> = -1.645; <i>p</i> = 0.100
CSA tibial nerves – Microvascular blood flow	0.375 (0.024)**	---

Data presented as Spearman's rank correlation coefficient (*rho*) (*p*-value), ***p*-value considered significant after Bonferroni adjustment (ie, *p* ≤ 0.05 / 2 = 0.025). CSA, Cross sectional area.

Table S2. Correlation and comparison between nerve conduction study – tibial nerve.

	CIDP (<i>n</i> = 5)		CMT (<i>n</i> = 5)	
	Correlation	<i>p</i> -value	Correlation	<i>p</i> -value
CMAP ankle left– CMAP ankle right	1.000 (<i>< 0.001</i>)**	<i>Z</i> = -1.604; <i>p</i> = 0.109	0.900 (0.037)	<i>Z</i> = -0.944; <i>p</i> = 0.354
CMAP popliteal fossa left – CMAP popliteal fossa right	1.000 (<i>< 0.001</i>)**	<i>Z</i> = -1.633; <i>p</i> = 0.102	-0.700 (0.188)	<i>Z</i> = -0.135; <i>p</i> = 0.893
NCV left – NCV right	0.959* (0.010)**	<i>T</i> (4) = 0.482; <i>p</i> = 0.655	-0.981* (0.003)**	<i>T</i> (4) = -0.110; <i>p</i> = 0.917
CMAP ankle – CMAP popliteal fossa	0.973 (0.005)**	<i>Z</i> = -1.604; <i>p</i> = 0.109	1.000 (<i>< 0.001</i>)**	<i>Z</i> = -2.023; <i>p</i> = 0.043

Data presented as Spearman's rank correlation coefficient (*rho*) (*p*-value), *Pearson correlation coefficient (*r*) (*p*-value), ***p*-value considered significant after Bonferroni adjustment (ie, *p* ≤ 0.05 / 4 = 0.0125); CIDP, Chronic inflammatory demyelinating polyradiculopathy; CMT, Charcot-Marie-Tooth disease; CMAP, compound muscle action potential; NCV, nerve conduction velocity.

Table S3. Subtype ALS comparison ultrasound data.

US data ALS (<i>n</i> = 11)	ALS bulbar (<i>n</i> = 3)	ALS limbs (<i>n</i> = 8)	<i>p</i> -value
CSA (mm ²)	14.06±0.82 (13.39-14.97)	10.94±3.07 (5.82-15.47)	<i>T</i> (9)=1.681; <i>p</i> =0.127
Microvascular blood flow	0.0138±0.0131 (0.00-0.03)	0.0091±0.0092 (0.00-0.02)	<i>T</i> (9)=0.675; <i>p</i> =0.516
US data ALS limbs (<i>n</i> = 8)	ALS lower limb (<i>n</i> = 2)	ALS upper limbs (<i>n</i> = 6)	<i>p</i> -value
CSA (mm ²)	12.32±4.46 (9.16-15.47)	10.49±2.67 (5.82-13.21)	<i>T</i> (6)=0.703; <i>p</i> =0.508
Microvascular blood flow	0.0172±0.0042 (0.01-0.02)	0.064±0.0090 (0.00-0.02)	<i>U</i> =2.000; <i>Z</i> = -1.341;

p=0.286

Data presented as mean \pm standard deviation, (range); US, ultrasound; ALS, Amyotrophic lateral sclerosis; CSA, Cross-sectional area. T-test and Mann-Whitney-U-Test were used. Significance was set at $p \leq 0.05$.

Table S4. Group comparison ultrasound data in CMT 1A.

US data		ALS (<i>n</i> = 11)	CIDP (<i>n</i> = 5)		CMT 1A (<i>n</i> = 4)	CON (<i>n</i> = 15)	
CSA (mm ²)		11.79 ± 2.97 (5.82-15.47)	20.97 ± 8.31 (9.69-33.05)		25.00 ± 2.83 (20.99-27.52)	9.15 ± 2.15 (5.61-13.80)	
Microvascular blood flow		0.0104 ± 0.0099 (0.00-0.03)	0.0318 ± 0.0246 (0.00-0.06)		0.0100 ± 0.0093 (0.00-0.02)	0.0044 ± 0.0037 (0.00-0.01)	
US data	<i>p</i> -value (ANOVA)	ALS vs. CIDP	ALS vs. CMT 1A	ALS vs. CON	CIDP vs. CMT 1A	CIDP vs. CON	CMT 1A vs. CON
CSA (mm ²)	F(3) = 25.59; <i>p</i> < 0.001	< 0.001	< 0.001	0.554	0.759	< 0.001	< 0.001
Microvascular blood flow	F(3) = 7.592; <i>p</i> < 0.001	0.007	1.000	1.000	0.034	< 0.001	1.000

Data presented as mean \pm standard deviation, (range); US, ultrasound; ALS, Amyotrophic lateral sclerosis; CIDP, Chronic inflammatory demyelinating polyradiculopathy; CMT, Charcot-Marie-Tooth disease, CON, Controls; CSA, Cross-sectional area. One-way ANOVA and post-hoc Bonferroni were used. Significance was set at $p \leq 0.05$. Significant values are shown in bold.

Table S5. Correlation ultrasound data vs. clinical scores in CMT 1A.

Correlation	CMT 1A (n = 4)			
	CMTNS total	CMTNS leg score	MRC-sum-score	MRC-lower limbs
CSA	-0.395 (0.605)	-0.111 (0.889)	-0.135 (0.865)	-0.512 (0.488)
Microvascular blood flow	0.233 (0.767)	-0.219 (0.781)	-0.250 (0.750)	-0.216 (0.784)

Data presented as Pearson correlation coefficient (r) (p-value), **p-value considered significant after Bonferroni adjustment ($p \leq 0.05 / 8 = 0.00625$). CMT, Charcot-Marie-Tooth disease; CSA, Cross-sectional area; MRC-sum-score, Medical research council sum score; CMTNS, CMT-neuropathy score.

Table S6. Correlation ultrasound data vs. nerve conduction study – tibial nerve in CMT 1A.

Correlation	CMT 1A (n = 4)		
	CMAP ankle	CMAP popliteal fossa	NCV
CSA	0.204* (0.796)	0.400 (0.600)	0.693* (0.307)
Microvascular blood flow	-0.242* (0.758)	0.200 (0.800)	-0.669* (0.331)

Data presented as Spearman's rank correlation coefficient (rho) (*p*-value), *Pearson correlation coefficient (r) (*p*-value), *p*-value considered significant after Bonferroni adjustment ($p \leq 0.05 / 6 = 0.008$). CMT, Charcot-Marie-Tooth disease; CSA, Cross-sectional area; CMAP, Compound muscle action potential; NCV, Nerve conduction velocity.

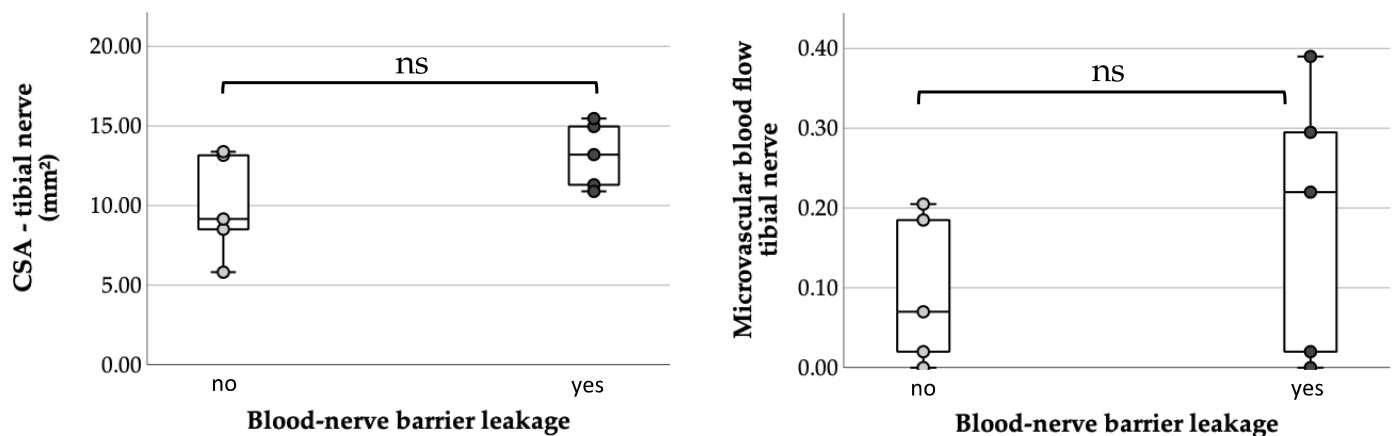


Figure S1. Box plots ALS – Blood-nerve barrier leakage.

A. Comparison of two ALS subgroups after division into present blood-nerve barrier leakage (yes) and intact blood-nerve barrier (no). The subgroup with blood-nerve barrier leakage shows not significant slightly larger CSA values. **B.** Illustration of microvascular blood flow without significant difference of ALS subgroups with blood-nerve barrier leakage (yes) and intact blood-nerve barrier (no). Median (horizontal bars), 25th to 75th quartile (box), range (whiskers) and single values (dots) are shown, CSA, Cross-sectional area; CSF, cerebral spinal fluid; ns, not significant.

Table S7. Subgroup comparison clinical scores – ALS.

ALS (n = 11)	ALSFRS/R total			ALSFRS/R gross-motor		
	severe (n = 8)	mild (n = 3)	<i>p</i> -value	severe (n = 6)	mild (n = 5)	<i>p</i> -value
CSA (mm ²)	11.26 ± 2.82 12.24 (5.82-13.81)	13.20 ± 3.51 1.97 (9.16-15.47)	T(9) = -0.958; <i>p</i> = 0.363	11.11 ± 3.11 12.24 (5.82-13.81)	12.61 ± 2.91 13.21 (8.52-15.47)	T(9) = -0.821; <i>p</i> = 0.433
Microvascular blood flow	0.0067 ± 0.0087 0.0021 (0.00-0.02)	0.0202 ± 0.0059 0.0202 (0.01-0.03)	U = 3.000; Z = -1.854; <i>p</i> = 0.085	0.0082 ± 0.0090 0.069 (0.00-0.02)	0.0130 ± 0.0116 0.0142 (0.00-0.03)	T(9) = -0.777; <i>p</i> = 0.457

Data presented as mean \pm standard deviation, median (range). *p*-value considered significant after Bonferroni adjustment (ie, $p \leq 0.05 / 4 = 0.0125$). ALS, Amyotrophic lateral sclerosis; CSA, Cross sectional area; ALSFRS/R, ALS-Functional Rating Scale/revised.

Table S8. Subgroup comparison clinical scores – CIDP.

CIDP (<i>n</i> = 5)	ONLS total			ONLS leg grade scale		
	severe (<i>n</i> = 2)	mild (<i>n</i> = 3)	<i>p</i> -value	severe (<i>n</i> = 2)	mild (<i>n</i> = 3)	<i>p</i> -value
CSA (mm ²)	27.50 \pm 7.85 27.50 (21.95-33.05)	16.62 \pm 6.01 19.67 (9.69-20.49)	T(3) = -1.785; <i>p</i> = 0.172	27.50 \pm 7.85 27.50 (21.95-33.05)	16.62 \pm 6.01 19.67 (9.69-20.49)	T(3) = -1.785; <i>p</i> = 0.172
Microvasc ular blood flow	0.0241 \pm 0.0342 0.0241 (0.00-0.05)	0.0370 \pm 0.0231 0.0257 (0.02-0.06)	T(3) = 0.515; <i>p</i> = 0.642	0.0241 \pm 0.0342 0.0241 (0.00-0.05)	0.0370 \pm 0.0231 0.0257 (0.02-0.06)	T(3) = 0.515; <i>p</i> = 0.642
CIDP (<i>n</i> = 5)	MRC-sum-score			MRC leg		
	severe (<i>n</i> = 2)	mild (<i>n</i> = 3)	<i>p</i> -value	severe (<i>n</i> = 2)	mild (<i>n</i> = 3)	<i>p</i> -value
CSA (mm ²)	27.50 \pm 7.85 27.50 (21.95-33.05)	16.62 \pm 6.01 19.67 (9.69-20.49)	T(3) = 1.785; <i>p</i> = 0.172	27.50 \pm 7.85 27.50 (21.95-33.05)	16.62 \pm 6.01 19.67 (9.69-20.49)	T(3) = 1.785; <i>p</i> = 0.172
Microvasc ular blood flow	0.0241 \pm 0.0342 0.0241 (0.00-0.05)	0.0370 \pm 0.0231 0.0257 (0.02-0.06)	T(3) = -0.515; <i>p</i> = 0.642	0.0241 \pm 0.0342 0.0241 (0.00-0.05)	0.0370 \pm 0.0231 0.0257 (0.02-0.06)	T(3) = -0.515; <i>p</i> = 0.642

Data presented as mean \pm standard deviation, median (range). *p*-value considered significant after Bonferroni adjustment (ie, $p \leq 0.05 / 8 = 0.00625$). CIDP, Chronic inflammatory demyelinating polyradiculopathy; CSA, Cross sectional area; ONLS, Overall neuropathy limitation scale; MRC, Medical research council.

Table S9. Subgroup comparison clinical scores – CMT.

CMT (<i>n</i> = 5)	CMTNS total			CMTNS leg		
	severe (<i>n</i> = 2)	mild (<i>n</i> = 3)	<i>p</i> -value	severe (<i>n</i> = 2)	mild (<i>n</i> = 3)	<i>p</i> -value
CSA (mm ²)	24.26 \pm 4.62 24.26 (20.99-27.52)	23.07 \pm 4.65 25.23 (17.73-26.26)	T(3) = -0.279; <i>p</i> = 0.798	22.63 \pm 6.92 22.63 (17.73-27.52)	24.16 \pm 2.79 25.23 (20.99-26.26)	T(3) = 0.365; <i>p</i> = 0.739
Microvascular blood flow	0.0081 \pm 0.0037 0.0081 (0.00-0.01)	0.0083 \pm 0.0119 0.0031 (0.00-0.02)	T(3) = 0.028; <i>p</i> = 0.980	0.0043 \pm 0.0017 0.0043 (0.00-0.01)	0.0190 \pm 0.0110 0.0107 (0.00-0.02)	T(3) = 0.804; <i>p</i> = 0.480

CMT (<i>n</i> = 5)	MRC-sum-score			MRC leg		
	severe (<i>n</i> = 3)	mild (<i>n</i> = 2)	<i>p</i> -value	severe (<i>n</i> = 3)	mild (<i>n</i> = 2)	<i>p</i> -value
CSA (mm ²)	22.08 ± 4.99 20.99 (17.73-27.52)	25.74 ± 0.72 25.74 (25.23-26.26)	T(3) = -0.981; <i>p</i> = 0.399	23.84 ± 5.32 26.26 (17.73-27.52)	23.11 ± 3.00 23.11 (20.99-25.23)	T(3) = 0.170; <i>p</i> = 0.876
Microvascular blood flow	0.0064 ± 0.0039 0.0055 (0.00-0.01)	0.0110 ± 0.0155 0.0110 (0.00-0.02)	T(3) = -0.522; <i>p</i> = 0.638	0.0102 ± 0.0102 0.0055 (0.00-0.02)	0.0054 ± 0.0076 0.0054 (0.00-0.01)	T(3) = 0.556; <i>p</i> = 0.617

Data presented as mean ± standard deviation, median (range). *p*-value considered significant after Bonferroni adjustment (ie, *p* ≤ 0.05 / 8 = 0.00625). CMT, Charcot-Marie-Tooth disease; CSA, Cross sectional area; CMTNS, CMT-neuropathy score; MRC, Medical research council.

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