

Table S2 – Cardiovascular risk factors in N = 330 vEDS adult patients at study according to the type of *COL3A1* variant.

Characteristics^a	All N=330	Dominant negative N=266	Haplo- insufficiency N=64	<i>P</i>^b
Overweight (na*=110)	47 (14.2%)	38 (14.3%)	9 (14.1%)	0.738
Diabetes (na=125)	10 (3.0%)	9 (3.4%)	1 (1.6%)	0.506
High LDL-c (na=124)	48 (14.5%)	38 (14.3%)	10 (15.6%)	0.627
Hypertension (na=134)	34 (10.3%)	27 (10.2%)	7 (10.9%)	0.846
Smoking (na=111)	43 (13.0%)	33 (12.4%)	10 (15.6%)	0.749

^a Categorical data are presented as number (%).

^b The *P* value for categorical data was calculated through logistic regression.

No difference in the subset of patients with arterial lesions, data not shown.

* na: not available.

Table S3 – Characteristics and comparison of N = 227 patients with MSA lesions only (no aortic lesion) and N = 45 patients with aortic lesion (associated or not with MSA lesions).

Characteristics ^{a,b}	Patients with arterial lesions N=272	No aortic lesion N=227	Aortic lesion N=45	<i>P</i> ^c
Females	157 (57.7%)	136 (59.9%)	21 (46.7%)	0.103
Index cases	185 (68.0%)	155 (68.3%)	30 (66.7%)	0.832
Type of variant				0.027
- Dominant negative	225 (82.7%)	193 (86.3%)	32 (71.1%)	
- Haploinsufficiency	47 (17.3%)	34 (13.7%)	13 (28.9%)	
Age at molecular diagnosis (years)	38.0 (28.0-49.0)	37.0 (27.0-46.0)	49.0 (35.0-61.0)	2.4e-5
Deceased	43 (15.9%)	31 (13.7%)	12 (26.7%)	0.032
- Age at death (years)	38.0 (29.0-49.5)	38.0 (27.5-41.5)	48.5 (30.5-54.0)	0.027
- Time between molecular diagnosis and death (years)	3.0 (0.5-5.0)	4.0 (1.0-5.5)	2.0 (0.0-5.0)	0.221
Age at first arterial lesion (years)	35.0 (28.0-43.0)	34.0 (27.0-41.8)	43.5 (32.8-53.3)	6.9e-6
Non-aortic arterial lesion	263 (96.7%)		36 (80.0%)	
- Limb arteries	140 (51.7%)	113 (50.0%)	27 (60.0%)	0.441 ^d
- Supra-aortic trunk arteries	149 (54.9%)	129 (57.1%)	20 (44.4%)	0.228 ^e
- Renal arteries	109 (40.2%)	91 (40.3%)	18 (40.0%)	0.815 ^f
- Digestive arteries	135 (49.8%)	117 (51.8%)	18 (40.0%)	0.277 ^g
- Coronary arteries	11 (4.1%)	7 (3.1%)	4 (8.9%)	0.240 ^h

^a Categorical data are presented as number (%).

^b Continuous data are presented as median (IQR).

^c The *P* value for continuous variables was calculated using ANOVA. The *P* value for categorical data was calculated through logistic regression.

^d Adjusted age-groups did not reveal a significant difference for limb artery lesions in patients without aorta lesions in comparison with patients with aorta lesions: OR for age 1.02, IQR 1.00-1.04, *P* 0.035.

^e Adjusted age-groups did not reveal a significant difference for SAT lesions in patients without aorta lesions in comparison with patients with aorta lesions: OR for age 0.99, IQR 0.96-1.01, *P* 0.563.

^f Adjusted age-groups did not reveal a significant difference for renal artery lesions in patients without aorta lesions in comparison with patients with aorta lesions: OR for age 1.021 IQR 0.99-1.03, *P* 0.252.

^g Adjusted age-groups did not reveal a significant difference for digestive artery lesions in patients without aorta lesions in comparison with patients with aorta lesions: OR for age 0.99, IQR 0.97-1.01, *P* 0.502.

^h Adjusted age-groups did not reveal a significant difference for coronary artery lesions in patients without aorta lesions in comparison with patients with aorta lesions: OR for age 1.04, IQR 0.99-1.08, *P* 0.129.

Table S4 – Characteristics of the vEDS patients studied or not by carotid echotracking.

Characteristics ^{a,b}	No echotracking data N=197	Echotracking data N=133	P^c
Females	115 (58.4%)	83 (62.4%)	0.464
Index cases	116 (58.9%)	83 (62.4%)	0.521
Type of variant			0.006
- Dominant negative	149 (75.6%)	117 (87.9%)	
- Haploinsufficiency	48 (24.4%)	16 (12.0%)	
Age at molecular diagnosis (years)	37.0 (23.0-49.0)	35.5 (25.0-44)	0.548
Deceased*	33 (16.8%)	10 (7.5%)	0.017
- Age at death (years)	40.0 (29.0-52.0)	36.0 (28.3-38.0)	0.248
- Time between molecular diagnosis and death (years)	2.0 (0.0 – 5.0)	5.5 (4.3-6.8)	0.221
Age at first arterial lesion (years)	35.0 (26.0-43.0)	33.0 (27.3-42.0)	0.355
Arterial lesions	158 (80.2%)	114 (85.7%)	0.389
MSA lesions	151 (76.6%)	112 (84.2%)	0.389
- Limb arteries	81 (41.1%)	59 (44.4%)	0.910
- SAT	76 (38.6%)	73 (54.9%)	0.015
- Renal arteries	58 (29.4%)	51 (38.3%)	0.187
- Digestive arteries	75 (38.1%)	60 (45.1%)	0.494
- Coronary arteries	5 (2.5%)	6 (4.5%)	0.335
Aorta lesions	32 (16.2%)	13 (9.8%)	0.099

^a Categorical data are presented as number (%).

^b Continuous data are presented as median (IQR).

^c The *P* value for continuous variables was calculated using ANOVA. The *P* value for categorical data was calculated through logistic regression.

* Deceased at the time of this retrospective study.

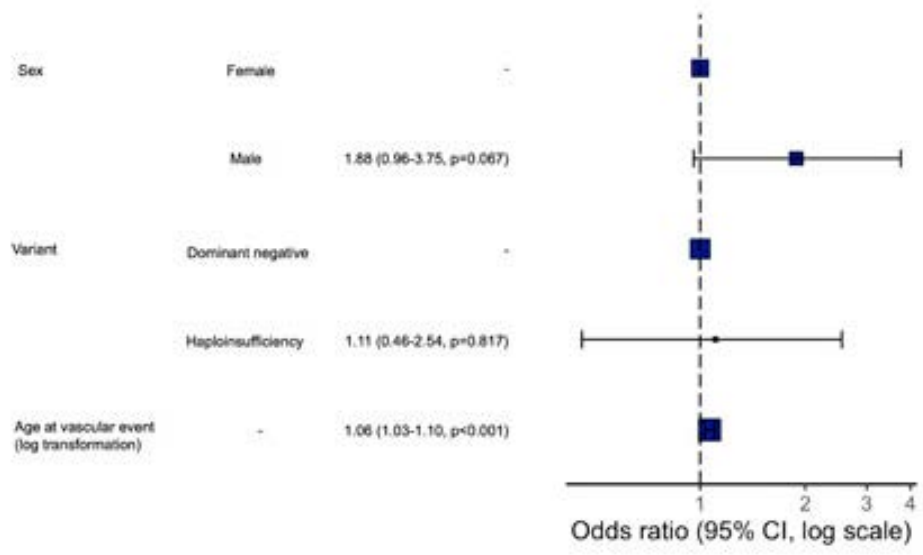


Fig1 Suppl.

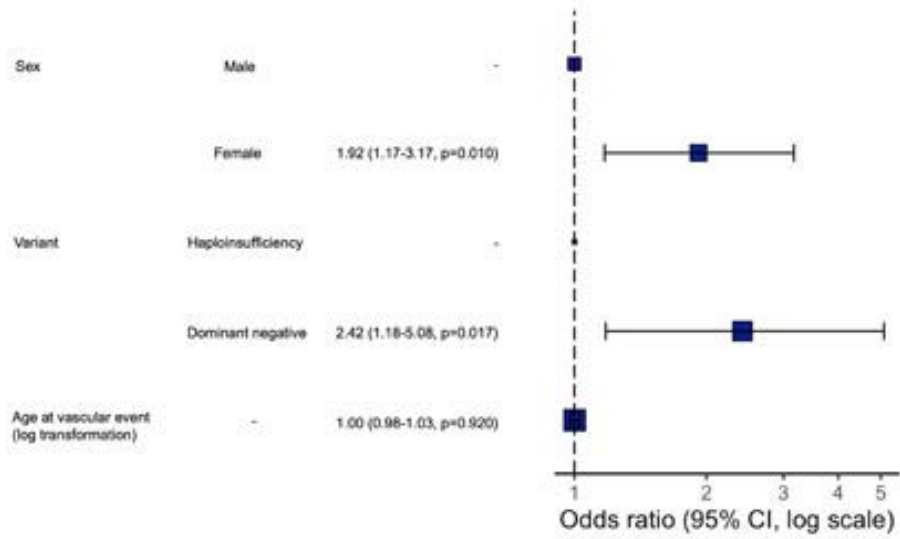


Fig2 Suppl.