

Supplementary Material

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Supplemental Table 1. Comparison of clinical and Fabry specific renal structural data in Fabry patients with known classic phenotype and those with unknown phenotype.

	Known Classic Phenotype (n=49)	Unknown Phenotype (n=7)	p-value
Age (years)	27 ± 13	22 ± 9	0.29
UPER (g/g)	0.30 ± 0.59	0.32 ± 0.44	0.93
GFR (ml/min/1.73 m ²)	117 ± 37	123 ± 30	0.75
Vv(Inc/PC)	0.41 ± 0.07	0.41 ± 0.04	0.98
Vv(Inc/Endo)	0.12 ± 0.04	0.17 ± 0.08	0.01
Vv(Inc/Mes)	0.06 ± 0.03	0.08 ± 0.04	0.25

Abbreviations: GFR=glomerular filtration rate; UPER=urine protein excretion rate;

Vv(Inc/PC)=fractional volume of GL3 inclusions in; Vv(Inc/Endo)=fractional volume of GL3

inclusions in endothelial cells; Vv(Inc/Mes)=fractional volume of GL3 inclusions in mesangial

cells.

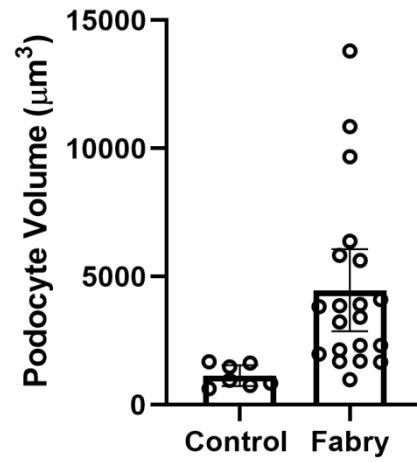
Supplemental Table 2. Relationships between fractional volume of GL3 inclusions in podocytes and podocyte profile area in individual biopsies.

Case	Age (year)	Number of Podocytes	R between podocyte profile area and Vv(Inc/PC)	p-value
1	7	25	0.76	<0.00001
2	18	18	0.67	0.002
3	16	29	0.53	0.003
4	12	27	0.73	<0.00001
5	13	38	0.60	<0.00001
6	34	16	0.88	<0.00001
7	11	40	0.73	<0.00001

Abbreviations: Vv(Inc/PC) = fractional volume of GL3 inclusions in podocytes

Supplemental Figure 1. Comparison of mean podocyte volume in male Fabry patients and healthy controls showing that podocyte volume was ~4 fold greater in Fabry patients.

*p=0.0006. Whiskers represent mean \pm 0.95 confidence interval. Circles represent individual values.



Supplemental Figure 2. Comparison of foot process width in Fabry and healthy control subjects. * $p=0.004$. Whiskers represent mean \pm 0.95 confidence interval. Circles represent individual values.

