S1 Table Patient characteristics stratified for sex, phenotype and ERT type

S1 Table A Patient characteristics at start of ERT Agalsidase alfa (0.2 mg/kg)

	Men		Women	
	Classical	Non-classical	Classical	Non-classical
Patients	69	47	95	37
Age at start ERT (years)	35 (±12)	52 (±16)	46 (±16)	51 (±15)
Follow up time (years)	6.7 (0.8-14.1)	3 (0.8-14.4)	5.3 (1.0-13.3)	4.6 (1.0-13.5)
Events before initiation of ERT				
▪ Dialysis/renal transplant	5 (9%)	2 (4%)	1 (1%)	0
▪ PM/ICD	1 (1%)	14 (30%)	6 (6%)	0
▪ Stroke	7 (10%)	1 (2%)	13 (14%)	1 (3%)
▪ Any of the above	11 (16%)	15 (32%)	16 (17%)	1 (3%)
LysoGb3 (nmol/l)	104 (62-146)	8.0 (4.1-36.2)	10.3 (2.7-22.6)	4.7 (0.7-19.5)
eGFR (ml/min/1.73m^2)	106 (10-139)	80 (10-136)	89 (10-160)	86 (32-126)
CKD category A3	12/52 (23%)	9/29 (31%)	21/82 (26%)	2/32 (6%)
LVMI (gram /m$^{2.7}$)	52 (24-112)	59 (16-117)	44 (23-97)	51 (16-96)
Use of ACEi/ARBs	15/69 (22%)	21/47 (45%)	37/95 (39%)	16/37 (43%)
Hypertension	14/65 (22%)	21/41 (49%)	38/93 (59%)	18/37 (51%)
BMI (kg/m^2)	22 (16-35)	27 (20-41)	25 (18-45)	25 (18-42)
HDL cholesterol (mmol/l)	1.2 (0.6-2.6)	1.3 (0.7-2.4)	1.5 (0.8-2.8)	1.6 (0.7-2.9)
LDL cholesterol (mmol/l)	2.2 (1.1-4.8)	2.6 (0.7-4.8)	2.6 (1.4-5.1)	2.8 (1.2-5.3)
Total cholesterol (mmol/l)	4.2 (2.4-6.2)	4.8 (2.4-7.4)	4.9 (2.8-8.1)	5.1 (3.5-7.4)
Triglycerides (mmol/l)	1.1 (0.5-3.5)	1.4 (0.6-5.6)	1.1 (0.3-4.4)	1.3 (0.5-5.9)

Continuous variables are presented as mean (±SD) or median (range).
ERT: enzyme replacement therapy, PM: pacemaker, ICD: implantable cardiac device, lysoGb3: globotriaosylsphingosine, eGFR: estimated glomerular filtration rate, CKDA: chronic kidney disease albuminuria categories, CKD category A3 is defined as AER >300 gram/day or equivalent, LVMI: left ventricular mass index measured by echocardiography, ACEi: angiotensin-converting-enzyme inhibitors, ARB: angiotensin receptor blocker, hypertension is defined as a diagnosis of increased blood pressure by the treating physician, BMI: body mass index, HDL: high density lipoprotein cholesterol, LDL: low density lipoprotein cholesterol.

S1 Table B Patient characteristics at start of ERT Agalsidase beta (1.0 mg/kg)

	Men		Women	
	Classical	Non-classical	Classical	Non-classical
Patients	71	7	43	18
Age at start ERT (years)	38 (±10)	55 (±13)	51 (±10)	53 (±16)
Follow up time (years)	3.8 (1.0-12.1)	3.7 (1.4-7.3)	4.0 (0.8-7.8)	4.2 (0.8-7.2)
Events before initiation of ERT				
▪ Dialysis/renal transplant	12 (16%)	0 (0%)	0 (%)	0 (0%)
▪ PM/ICD	4 (6%)	1 (14%)	3 (7%)	1 (6%)
▪ Stroke	8 (11%)	1 (14%)	7 (16%)	1 (6%)
▪ Any of the above	19 (27%)	1 (14%)	9 (21%)	2 (11%)
LysoGb3 (nmol/l)	116 (38-178)	9.5 (2.0-39.7)	9.9 (5.1-23.5)	8.3 (3.2-20.0)
eGFR (ml/min/1.73m^2)	85 (10-137)	45 (10-95)	87 (42-140)	90 (40-118)
CKD category A3	25/54 (46%)	5/7 (71%)	9/40 (23%)	3/13 (23%)
LVMI (gram /m$^{2.7}$)	51 (20-149)	75 (42-100)	58 (25-120)	48 (25-70)
Use of ACEi/ARBs	23/71 (32%)	5/7 (71%)	18/43 (42%)	6/18 (33%)
Hypertension	32/70 (46%)	5/7 (71%)	18/42 (43%)	7/18 (39%)
BMI (kg/m^2)	23 (15-34)	26 (23-38)	27 (19-45)	22 (18-38)
HDL cholesterol (mmol/l)	1.3 (0.8-1.9)	1.3 (1.0-1.7)	1.5 (0.8-2.8)	1.6 (1.1-2.3)
LDL cholesterol (mmol/l)	2.6 (1.3-4.2)	3.5 (1.1-4.3)	2.7 (1.4-4.5)	3.6 (1.2-1.6)
Total cholesterol (mmol/l)	4.4 (2.8-6.7)	5.4 (3.7-6.6)	4.9 (3.1-7.2)	5.8 (3.5-7.1)
Triglycerides (mmol/l)	1.3 (0.4-3.4)	2.1 (1.2-2.4)	1.1 (0.4-3.4)	1.4 (0.6-3.6)

Continuous variables are presented as mean (±SD) or median (range).
ERT: enzyme replacement therapy, PM: pacemaker, ICD: implantable cardiac device, lysoGb3: globotriaosylsphingosine, eGFR: estimated glomerular filtration rate, CKDA: chronic kidney disease albuminuria categories, CKD category A3 is defined as AER >300 gram/day or equivalent, LVMI: left ventricular mass index measured by echocardiography, ACEi: angiotensin-converting-enzyme inhibitors, ARB: angiotensin receptor blocker, hypertension is defined as a diagnosis of increased blood pressure by the treating physician, BMI: body mass index, HDL: high density lipoprotein cholesterol, LDL: low density lipoprotein cholesterol.