

Supplementary Table 1. The relationship of baseline clinical characteristics and occurrence of any arrhythmia requiring treatment during follow-up (Cox regression analysis)

Predictor variable		Multivariable analysis		
		HR	95% CI	<i>p-value</i>
Age (years)	41-50	0.44	0.10-2.03	0.292
	51-60	0.77	0.23-2.55	0.663
	61-70	0.69	0.18-2.67	0.591
	>70	1.73	0.43-6.93	0.438
Male sex		0.76	0.30-1.92	0.568
QRSd (ms)*		0.93	0.83-1.06	0.270
LGE		2.12	0.56-8.05	0.270

*HR for QRSd is defined using 10ms intervals.

Table legend: CI – confidence interval, HR – hazard ratio, LGE – late gadolinium enhancement, QRSd – QRS duration.

Supplementary table 2. Implantable Loop Recorder (ILR) usage in Fabry disease for the duration of the study period.

	Male	Female	<i>p-value</i>
Sample size (n, %)	10	9	<i>N/A</i>
Age (years)	46±5	58±13	0.023
Follow-up (years)	2.2 (1.5-4.6)	1.7 (0.8-3.5)	0.497
Indication:			
Palpitations & dizziness (n, %)	6	2	0.139*
Previous stroke ?AF (n, %)	2	1	
Suspected VA (n, %)	2	3	
Inconclusive Holter (n, %)	0	3	
Arrhythmia:			
AF requiring anticoagulation (n, %)	2	1	0.498*
High VE burden (n, %)	1	0	
Bradycardia/pauses – did not fulfil criteria for PPM (n, %)	1	3	

P-values are for male-female comparisons. *statistical significance investigated using Chi-squared testing.

Table legend: AF – atrial fibrillation, PPM – permanent pacemaker, VE – ventricular arrhythmia

Supplementary table 3. Baseline demographic data in the FD-CIED cohort split by device type

	PPM	ICD	CRT	p-value*
Sample size (n)	38	43	9	<i>N/A</i>
Age (yrs)	53±13	57±12	63±9	<i>0.052</i>
Male gender (n, %)	27 (71)	33 (77)	9 (100)	<i>0.182</i>
On ERT (n, %)	27 (71)	31 (72)	4 (44)	<i>0.247</i>
Classical mutation (n, %)	17 (45)	17 (40)	5 (56)	<i>0.660</i>
BMI (kg/m ²)	26±5	29±6	21±7	<i>0.014</i>
SBP (mmHg)	126±15	122±20	136±31	<i>0.499</i>
DBP (mmHg)	75±8	70±12	86±21	<i>0.077</i>
MSSI	14.5±10.7	14.8±8.6	18.4±10	<i>0.541</i>
Comorbidities				
IHD (n, %)	3 (8)	2 (5)	1 (11)	<i>0.719</i>
CKD stage 3-5 (n, %)	10 (26)	5 (12)	3 (33)	<i>0.147</i>
HTN (n, %)	8 (21)	8 (19)	2 (22)	<i>0.948</i>
DM (n, %)	3 (8)	5 (12)	0 (0)	<i>0.516</i>
Stroke/TIA (n, %)	14 (37)	5 (12)	3 (33)	<i>0.025</i>

*p-values are calculated using one-way ANOVA with post-hoc Tukey correction for continuous variables and Chi-squared testing for categorical variables.

Table legend: BMI – body mass index, CKD – chronic kidney disease, CRT – cardiac resynchronization therapy, DBP – diastolic blood pressure, DM – diabetes mellitus, ERT – enzyme replacement therapy, HTN – hypertension, ICD – implantable cardioverter defibrillator, IHD – ischemic heart disease, MSSI – Mainz severity score index, PPM – permanent pacemaker, SBP – systolic blood pressure, TIA – transient ischemic attack.

Supplementary figure 1

The incidence of arrhythmic events in the FD-CIED group on ERT vs. no therapy. It includes the following: (i) total documented arrhythmia, (ii) treated arrhythmia (including initiation of or a change in medication) and (iii) individual arrhythmia (AF and sustained VT requiring therapy). * denotes $p=0.008$. All other comparisons were non-significant.

Figure legend: AF – atrial fibrillation, ATP – anti-tachycardia pacing, VT – ventricular tachycardia.