

Supplementary table 3: Clinical characteristics and survival rates by era of presentation.

		1980-1989 (n=11)	1990-1999 (n=117)	2000-2009 (n=254)	2010-2017 (n=305)	P value
Gender	Male	6 (55%)	67 (57%)	161 (63%)	200 (66%)	+ 0.412
Syndrome	Idiopathic/ non- syndromic	5 (45%)	88 (75%)	157 (62%)	183 (60%)	+ 0.025
	RASopathy	4 (36%)	19 (16%)	48 (19%)	55 (18%)	
	Friedreich ataxia	1 (10%)	9 (8%)	26 (10%)	23 (8%)	
	Inborn error of metabolism	1 (10%)	1 (1%)	21 (8%)	41 (13%)	
Length of follow up in years (median, IQR)		15 (12.6-24)	11.8 (6.5 - 18)	8.5 (5.25- 11.2)	2.6 (1.3 - 4.1)	
Mortality rate (per 100 person years at risk)		2.27 (0.852 - 6.05)	1.38 (0.890- 2.14)	1.71 (0.123- 2.373)	2.54 (1.672 - 3.858)	* 0.735

	1980-1989 (n=11)	1990-1999 (n=117)	2000-2009 (n=254)	2010-2017 (n=305)	P value
SCD rate (per 100 person years at risk)	1.13 (0.284 - 4.537)	1.17 (0.729 - 1.886)	1.14 (0.765- 1.703)	1.73 (1.044 - 2.872)	* 0.876

Data expressed as number (%). Total number of patients is 687 unless otherwise stated. + Indicates comparisons were made using Chi square test. * Indicates comparison made using Log Rank Test.

SCD = sudden cardiac death