

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

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

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

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