

Supplemental figures and tables

Figure legends

Figure I: Survival curves for each mutation. They compare cases from our centres versus cases from the bibliography. No significant differences were found for mutations Arg719Trp (a), Gly716Arg (b) and Ile736Thr (c). For Arg719Gln (d), differences in survival rate were not significant below the age of 50.

Figure II. Survival curves for Mutation Arg723Gly vs all mutations in the converter region. Mutation Arg723Gly showed a better prognosis than the other mutations in the region

Figure III: Survival curves for cases with one vs two mutation. No significant differences were observed.

Figure I a

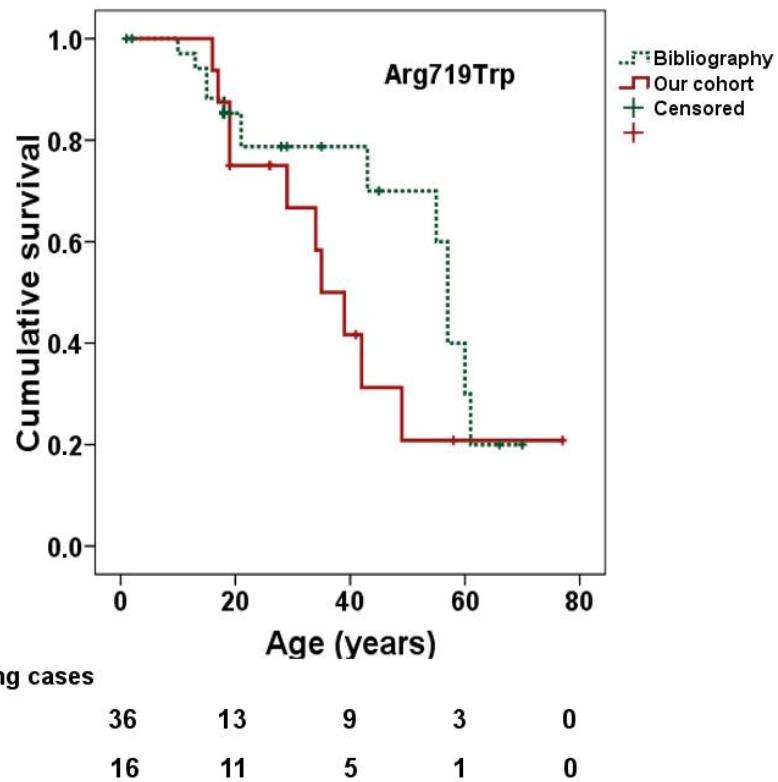


Figure I b

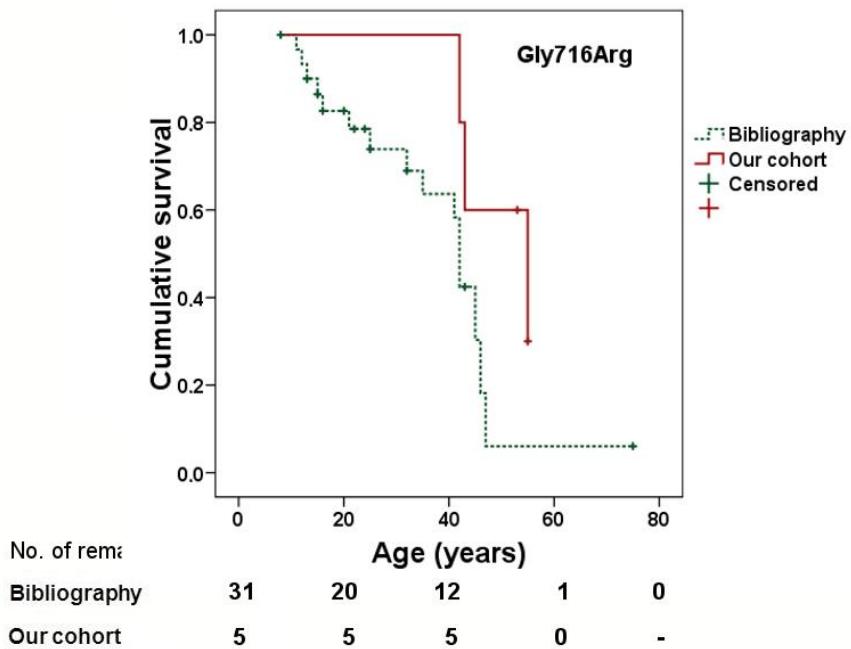


Figure 1c

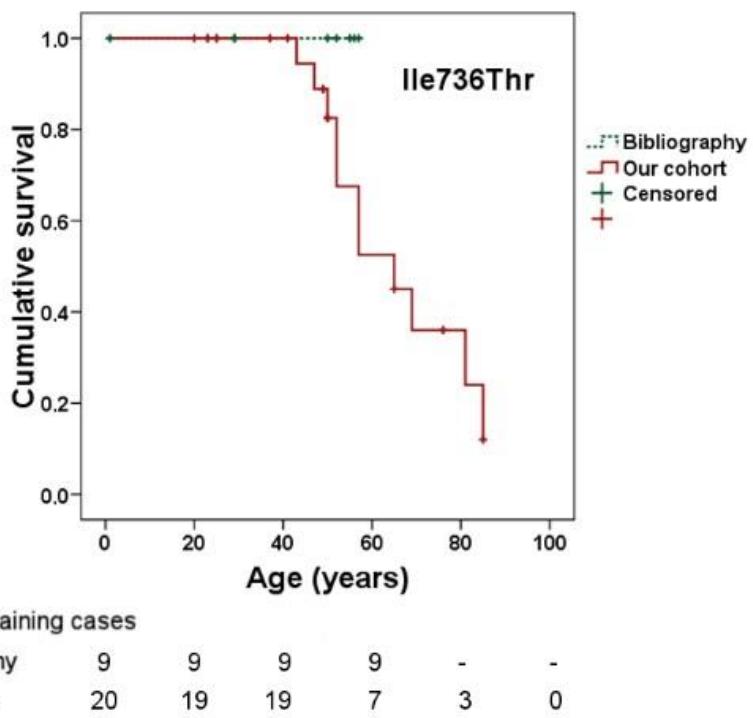


Figure 1d

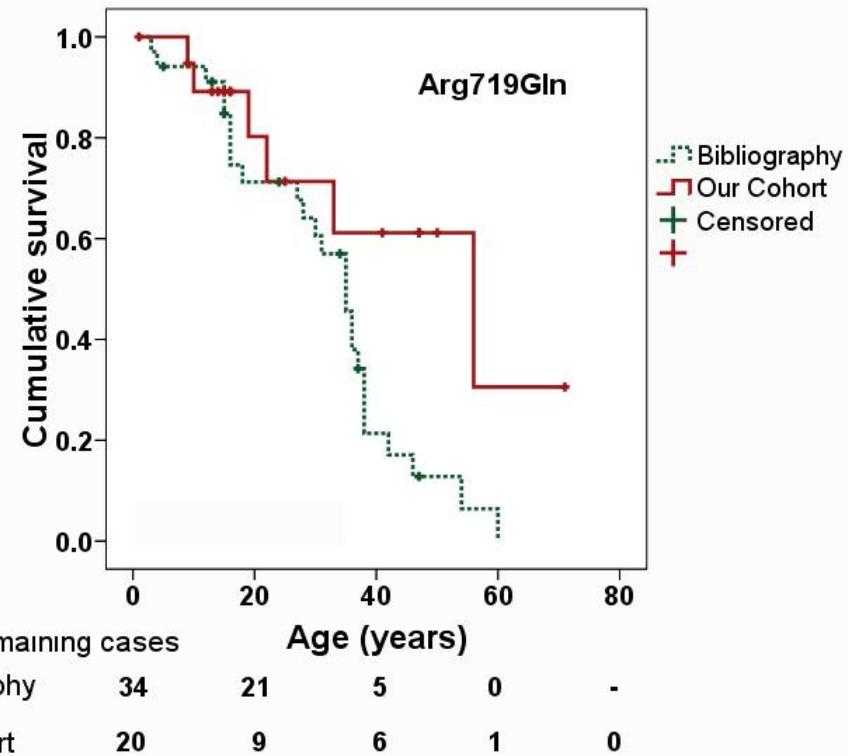


Figure II

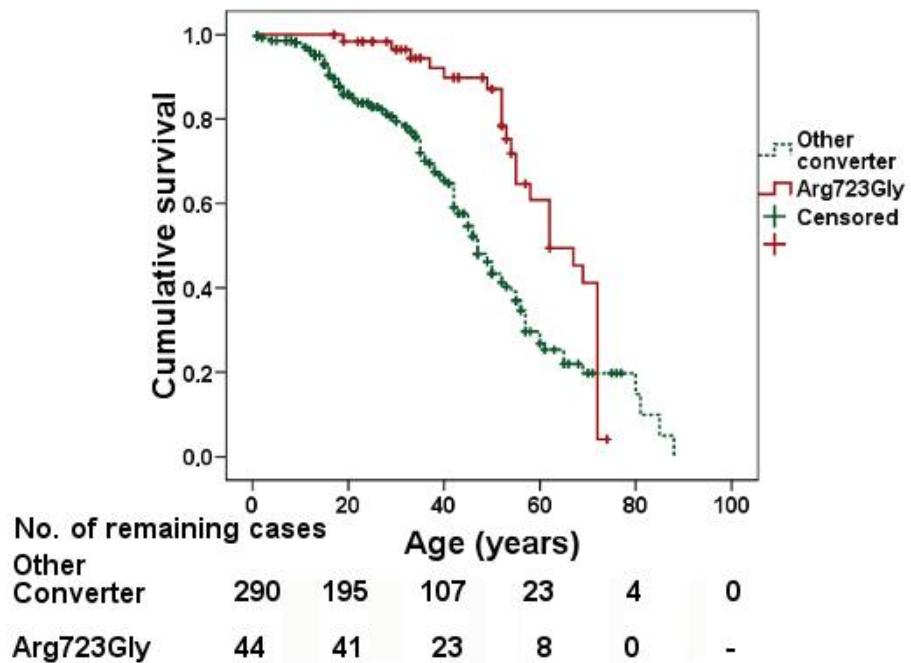


Figure III

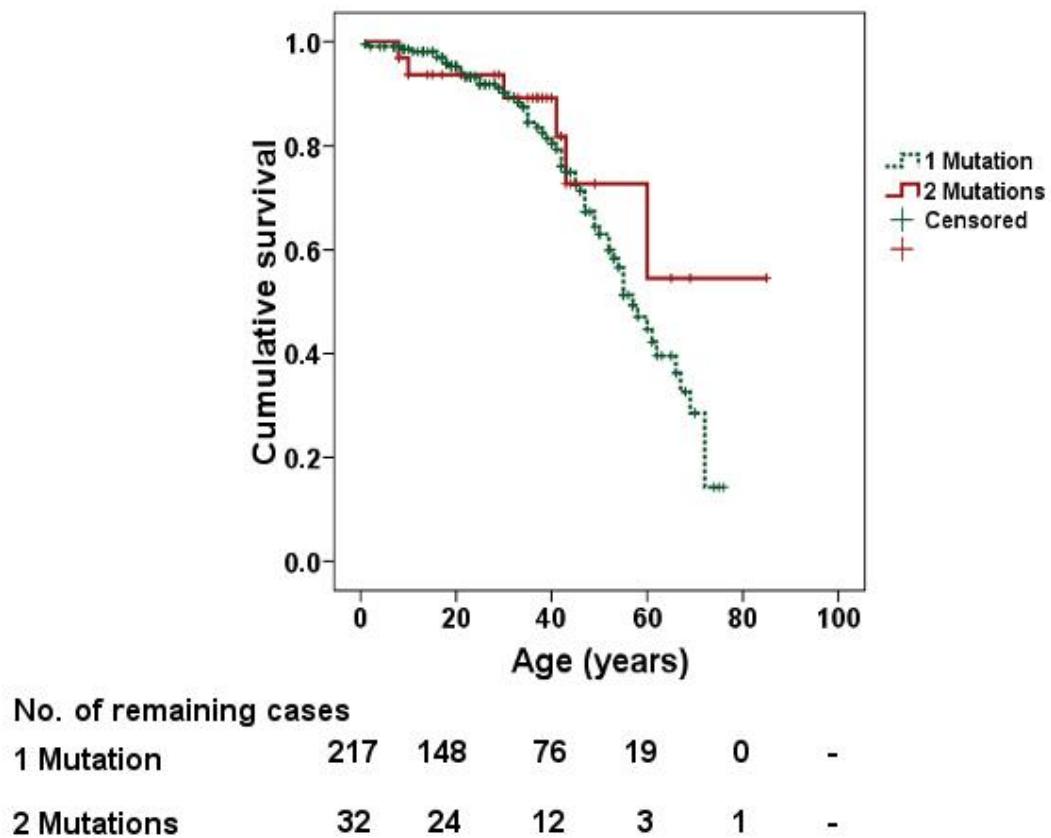


Table A: Hazard ratios and confidence intervals of Frailty Cox-Propotional Model. Comparisons of mutations from the bibliography vs mutations of our cohort, particular mutations vs all others in the converter region, and carriers of 1 mutation vs carriers of more than one mutations (at least one of them located in converter region)

	Mutation	HR (95% CI)	p-value
Cases from bibliography vs cases from our cohort	Arg719Gln	0.36 (0.14, 0.89)*	0.02
	Gly768Arg	0.09 (0.001, 8.85)	0.31
	Arg719Trp	1.31 (0.28, 5.9)	0.73
	Gly716Arg	0.59 (0.12, 2.75)	0.51
	All Mutations	0.66 (0.31, 1.37)	0.27
Mutation Vs All others in converter region	Arg719Gln	3.92 (1.91, 8.01)*	<0.01
	Ile736Thr	0.26 (0.091, 0.77)*	0.01
	Gly716Arg	1.75 (0.63, 4.86)	0.28
	Arg723Gly	0.44 (2.23, 1.11)	0.08
	Asp717Gly	2.66 (0.38, 6.7)	0.053
1 mutation carriers vs 2 mutation carriers		0.39 (2.5, 1.14)	0.08

* Statistically significant at p-value ≤ 0.05.

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