Supplementary Table 1. Ranking of pathogenic variants in mtDNA associated to HON in Italian, French and Global cohorts. The arrow ↓ indicate the lower ranking in the national cohort as compared to the global cohort and the arrow ↑ indicate the higher ranking in the national cohort as compared to the global cohort.

Patogenic variants in mtDNA									
	Italian cohort (n=125)			France cohort (n=199)			Total (n=324)		
nt position	N	%	Ranking	N	%	Ranking	N	%	Ranking
m.11778 G>A/*MT-ND4*	74	59.2%	1 (=)	133	66.8%	1 (=)	207	63.9%	1
m.3460 G>A/*MT-ND1*	20	16.0%	2 (=)	30	15.1%	2 (=)	50	15.4%	2
m.14484 T>C/*MT-ND6*	18	14.4%	3 (=)	21	10.6%	3 (=)	39	12.0%	3
m.14482 C>A/*MT-ND6*	0	0.0%	-	4	2.0%	4 (=)	4	1.2%	4
m.13051 G>A/*MT-ND5*	1	0.8%	6 (↓)	2	1.0%	5 (=)	3	0.9%	5
m.4171 C>A/*MT-ND1*	1	0.8%	6	1	0.5%	7 (↑)	2	0.6%	6
m.10254 G>A/*MT-ND3*	2	1.6%	4 (↑)	0	0.0%	-	2	0.6%	6
m.14487 T>C/*MT-ND6*	2	1.6%	4 (↑)	0	0.0%	-	2	0.6%	6
m.13042 G>A/*MT-ND5*	0	0.0%	-	2	1.0%	5 (↑)	2	0.6%	6
m.3733G>A/*MT-ND1*	1	0.8%	6 (↑)	0	0.0%	-	1	0.3%	10
m.3890 G>A/*MT-ND1*	1	0.8%	6 (↑)	0	0.0%	-	1	0.3%	10
m.14477 A>G/*MT-ND6*	1	0.8%	6 (↑)	0	0.0%	-	1	0.3%	10
m.10663 T>C/*MT-ND4L* m.12033 A>G/*MT-ND4* m.14258 G>A/*MT-ND6*	1	0.8%	6 (↑)	0	0.0%	-	1	0.3%	10
m.14258 G>A/*MT-ND6* m.10680 G>A/*MT-ND4L*	1	0.8%	6 (↑)	0	0.0%	-	1	0.3%	10
m.14258 G>A/*MT-ND6* m.14582 A>G/*MT-ND6*	1	0.8%	6 (↑)	0	0.0%	-	1	0.3%	10
m.3460 G>A/*MT-ND1*	1	0.8%	6 (↑)	0	0.0%	-	1	0.3%	10

m.14484 T>C/*MT-ND6*									
m.3395 A>G/*MT-ND1*	0	0.0%	-	1	0.5%	7 (↑)	1	0.3%	10
m.3700 G>A/*MT-ND1*	0	0.0%	-	1	0.5%	7 (↑)	1	0.3%	10
m.9957 T>C/*MT-CO3*	0	0.0%	-	1	0.5%	7 (↑)	1	0.3%	10
m.13513 G>A/*MT-ND5*	0	0.0%	-	1	0.5%	7 (↑)	1	0.3%	10
m.13679 C>T/*MT-ND5*	0	0.0%	-	1	0.5%	7 (↑)	1	0.3%	10
m.14459 G>A/*MT-ND6*	0	0.0%	-	1	0.5%	7 (↑)	1	0.3%	10