

Table 6 (web only) Multivariate analysis of prognostic factors- Europe only population

Variable	Chi-Squared	Significance	Goodness of fit
Cardiac n=320			
eGFR ($< 60\text{ml}/\text{min}/1.73\text{m}^2$)	39.16	<0.001	0.9135
Hearing impairment ^s	Not included		
LVM index ($\geq 50\text{ g}/\text{m}^2$)	13.26	<0.001	
Proteinuria ($>300\text{mg}/24$ hour)	11.36	<0.001	
Presence of vertigo	15.01	<0.001	
Presence of Abnormal ECG	4.29	<0.05	
Presence of Anhidrosis/Hypohidrosis	4.19	<0.05	
Presence of Angiokeratomas or telangectasias	*	$\ll 0.0001^*$	

Renal n=179			
eGFR ($< 60 \text{ ml/min/1.73m}^2$)	52.13	<0.001	0.9250
Male Gender	18.99	<0.001	
Proteinuria ($>300\text{mg}/24$ hours)	4.75	<0.05	
Presence of Angiokeratomas or telangiectasias		All patients had yes on this variable	
Neuro n=320			
Hearing impairment ^s	12.18	<0.001	0.3388
eGFR ($< 60 \text{ ml/min/1.73m}^2$)	16.18	<0.001	
Presence of Vertigo	Not included		
Presence of Anhidrosis/ Hypohidrosis	6.90	<0.01	
Death n=320			
eGFR ($< 60 \text{ ml/min/1.73m}^2$)	17.05	<0.001	0.5178
Presence of Abnormal ECG	7.65	<0.05	

Composite n=320			0.3048
eGFR (< 60 , ml/min/1.73m ²)	74.51	<0.001	
Hearing impairment [§]	30.85	<0.001	
Microalbuminuria	5.67	<0.01	
LVM index (≥ 50 g/m ²)	7.26	<0.01	
Presence of anhidrosis/ hypohidrosis (Y/N)	Not included		
Proteinuria *(>300 mg/24hour)	13.31	<0.001	

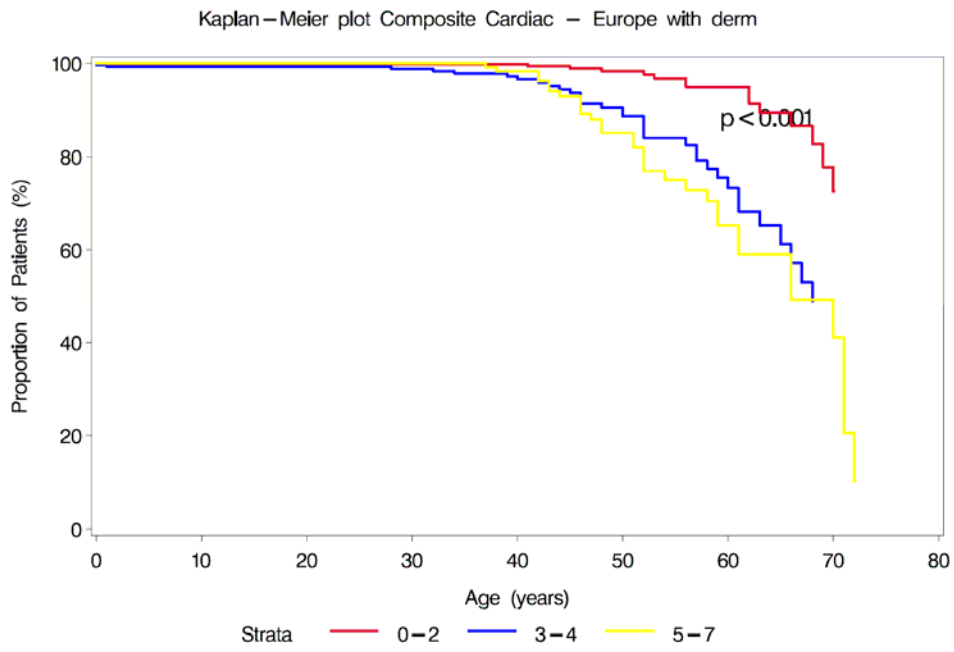
* Added

[§] Hearing impairment is average pure tone audiometric thresholds at 0.5, 1 & 2 kHz in one or both ears > 25 dB ISO

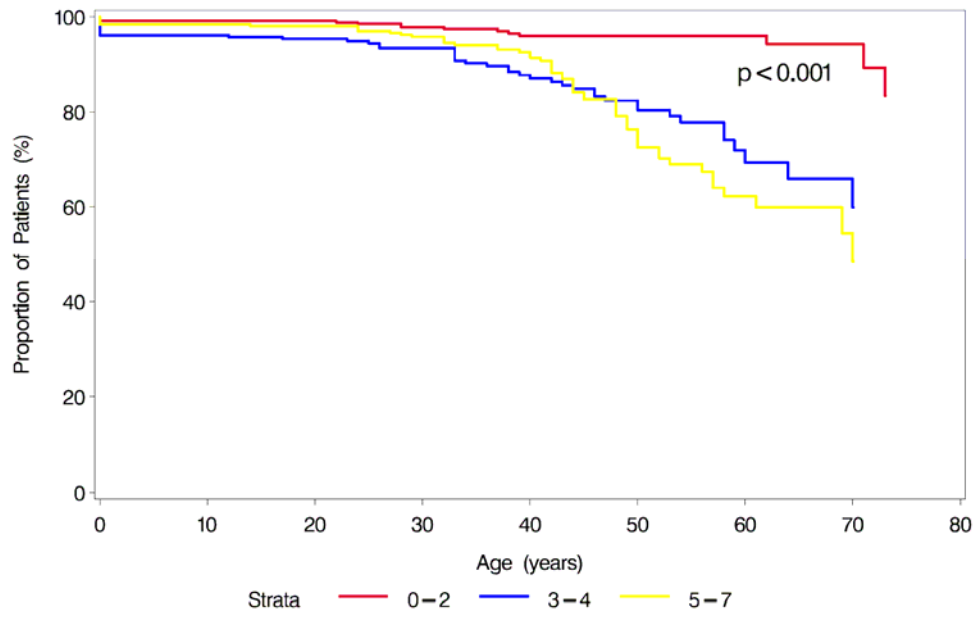
Figure 3 (web only)

Kaplan-Meier Analysis of Predictive Scores - Europe only population

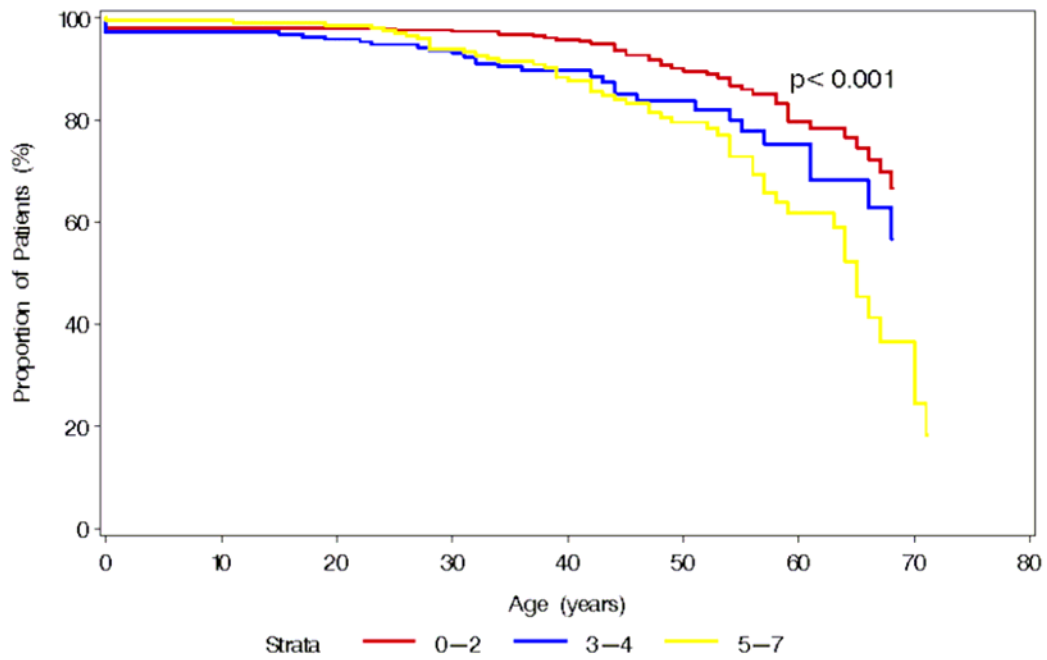
- (a) Cardiac score
- (b) Renal score
- (c) Neurological
- (d) Composite



Kaplan–Meier plot composite Renal - Europe



Kaplan–Meier plot Composite CNS - Europe



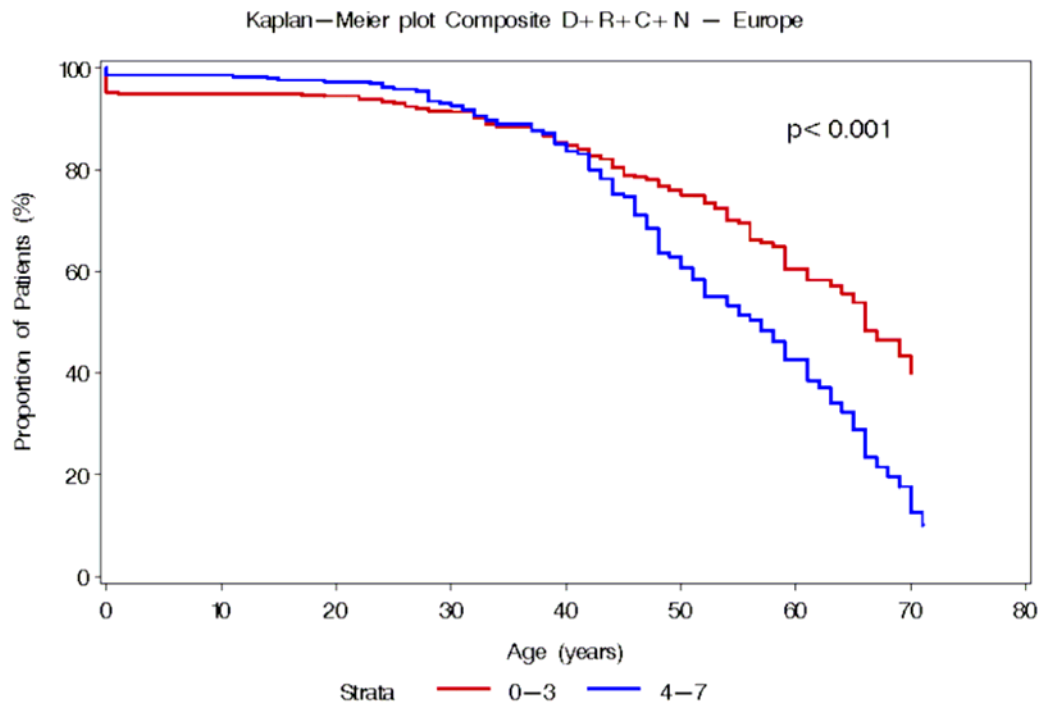


Figure 4 (web only) Kaplan-Meier Analysis of Predictive Scores- Randomly selected population

- (a) Cardiac score
- (b) Renal score
- (c) Neurological
- (d) Composite

Kaplan-Meier plot Cardiac - rand

