

Background

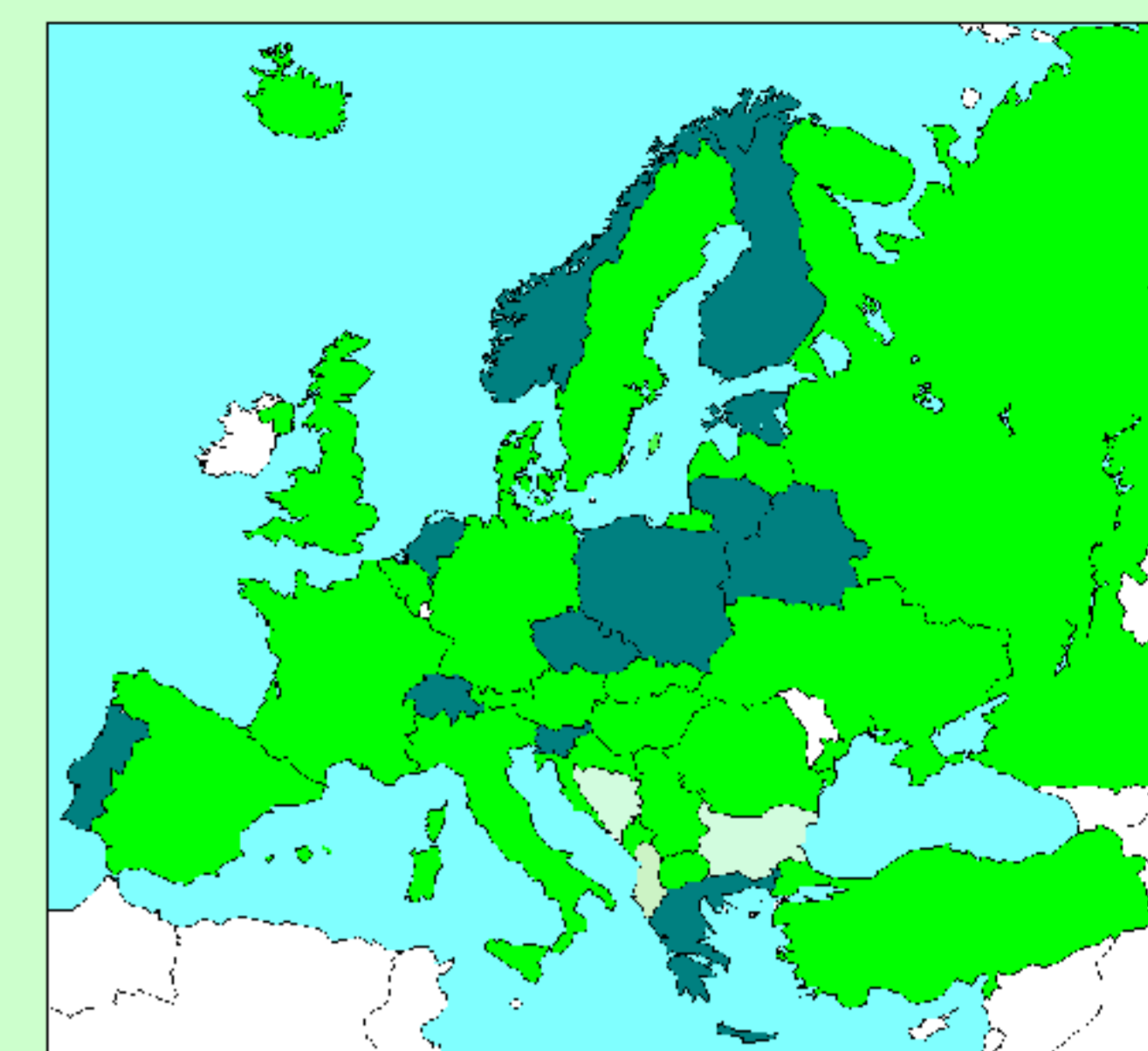
Congenital nephrotic syndrome of the Finnish type (CNF) is a rare congenital renal disease presenting with heavy proteinuria starting within 3 months after birth. Data on outcome of renal replacement therapy (RRT) for children with CNF are scarce. Here we present outcomes for 209 children with CNF at start of RRT (dialysis or pre-emptive renal transplantation, RTX). CNF is much more common in Finland and we compared treatment strategies in Finland (Finnish CNF) with those in other countries. As for not all types it was sure whether it was CNS of the Finnish type we also had a group of CNS - unknown

Methods

Countries in the ESPN/ERA-EDTA Registry for which data on CNF over the period January 1, 1991 and December 31, 2011 were available are shown in Figure 1, as marked with the dark green colors. Countries in lighter green are those participating in the Registry.

Statistical tests were two-tailed and differences were considered statistically significant when $p < 0.05$. Data were analysed using SPSS statistical program (version 20, SPSS, Chicago, Ill., USA)

Fig. 1 Participating countries



Results

	Finnish CNF patients	Non-Finnish CNF patients	Non-Finnish CNS patients
Patient number	66	70	73
Age at start RRT (years)	0.66	1.96	1.78
Height Z-score at start of RRT	-1.44	-2.59	-2.58
at age of 5 years	-1.58	-1.90	-2.34
eGFR at start (ml/min/1.73m ²)	172	7.6	8.4
RRT at start:			
PD/HD	100 / 0 %	68.6 / 18.6 %	69.9 / 19.2 %
pre-emptive RTX	0%	12.9%	5.5%
Dialysis time (years)	0.87	1.12	1.82
Age at RTX (years)	1.63	3.37	4.00

Figure 2. Patient survival

25 patients died, and 5 year patient survival was 90%. There was no significant difference between patients from Finland and from outside Finland both in time since birth and time since start of RRT

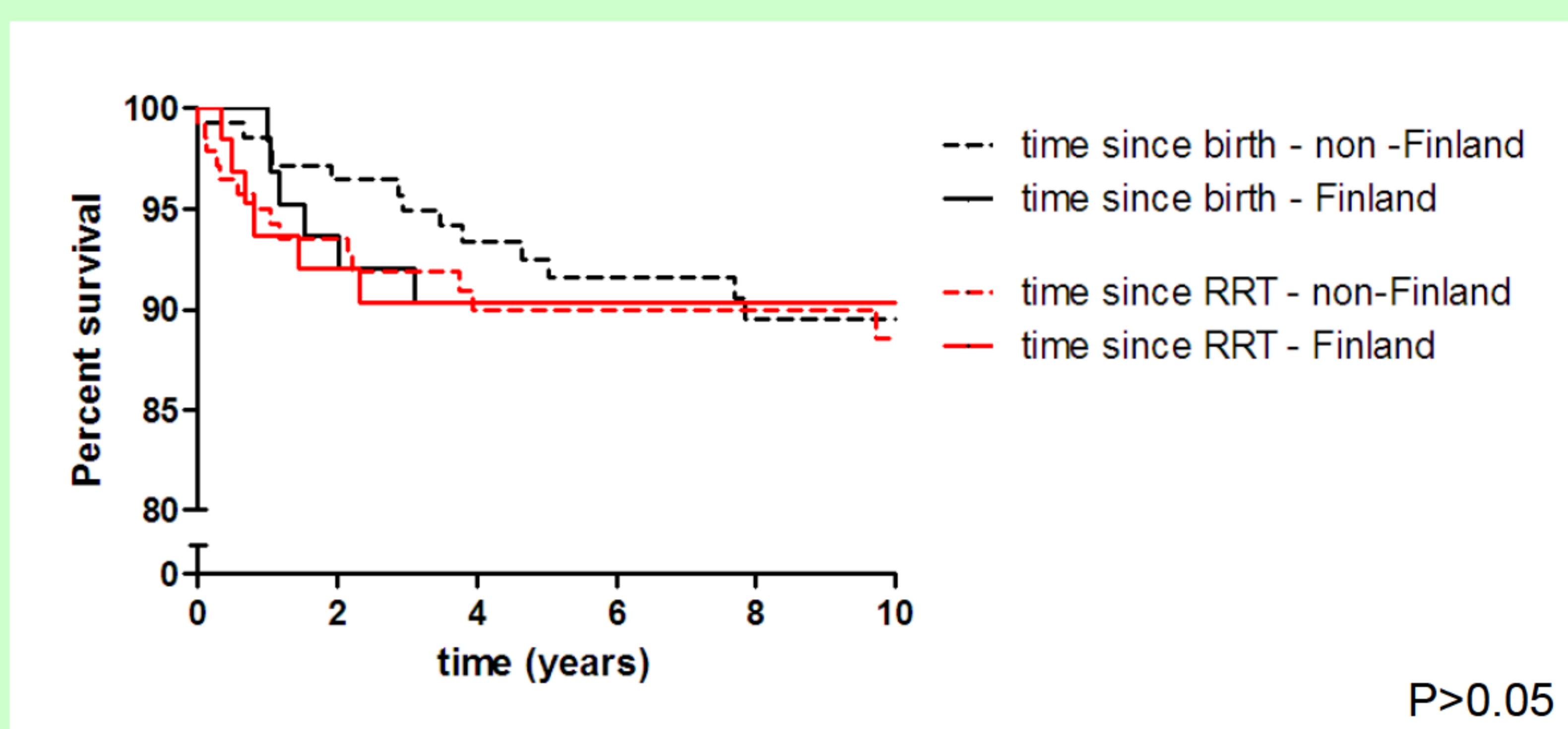
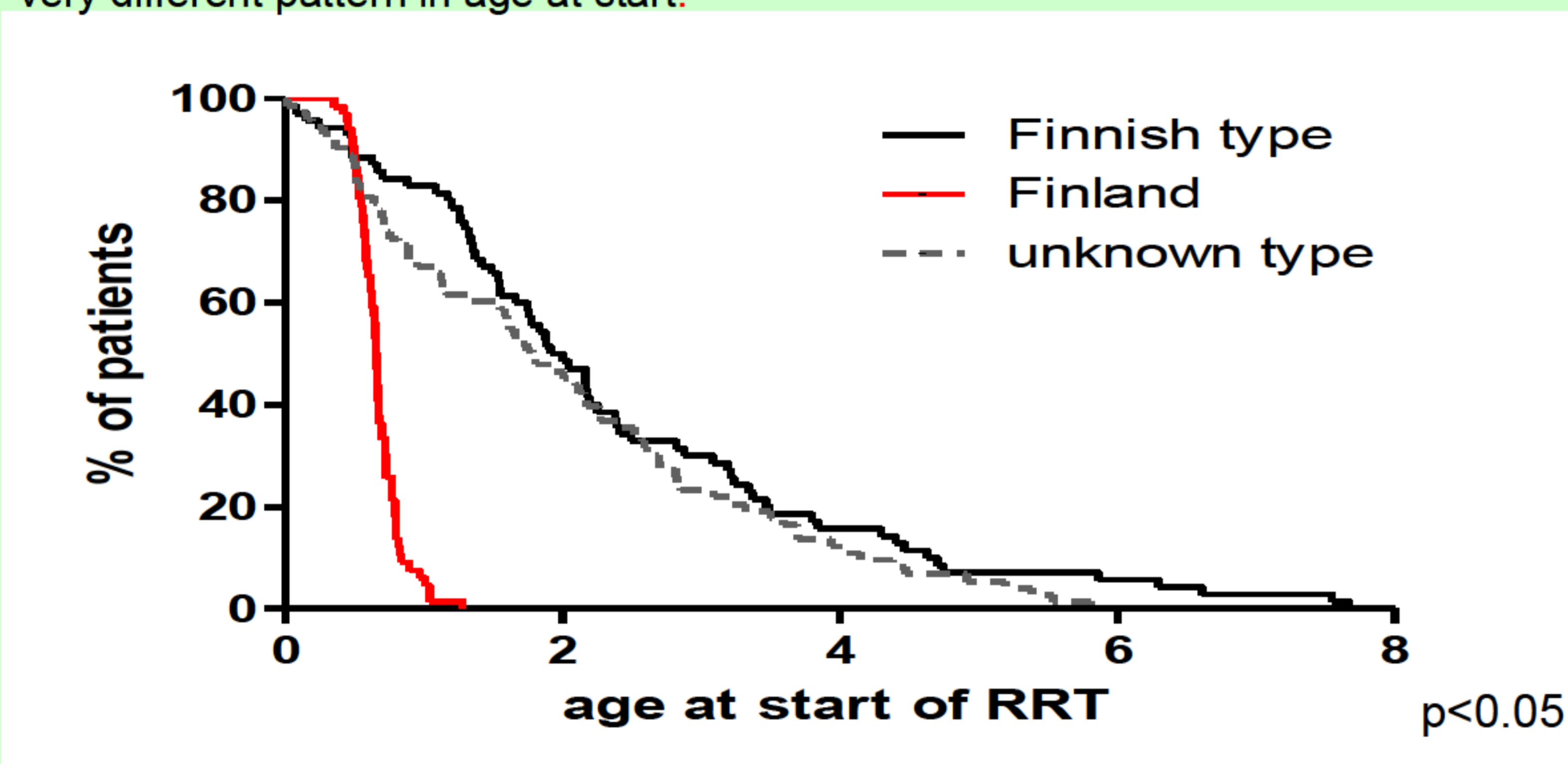


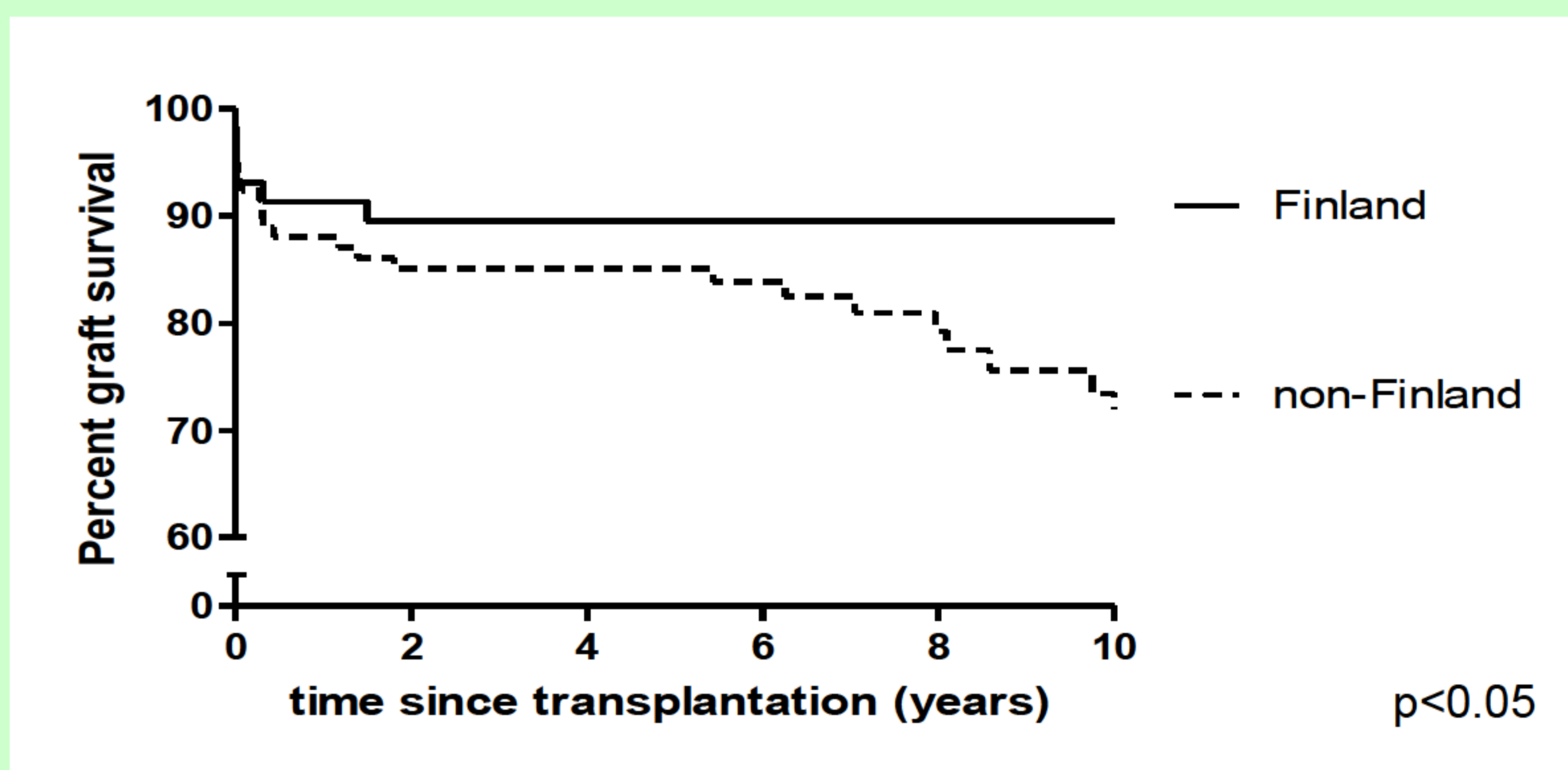
Figure 3. Mean age at start of RRT

Median age at RRT start was very different across the 3 groups, and there was a very different pattern in age at start.



Graft survival censored for patient death

5 year graft survival was 85% outside Finland and 90% in Finland



Discussion

- Patients with CNF from Finland started RRT at a much younger age, as compared to patients from outside Finland. All patients in Finland followed a very similar pattern, all being nephrectomised at 1 year, but this strategy was very different from that in the other countries.
- As a consequence eGFR at start was very different between the groups
- Height at start and height at 5 years were significantly higher in patients from Finland as compared to other patients.

- Patient survival was not different between the groups
- Graft survival was significantly better in Finland as compared to the other countries
- Unfortunately, no information was available on the exact genetic cause and pre-dialysis treatment. Part of the differences might be explained by the occurrence of the Fin-Minor and -Major mutations, explaining almost all of the cases in Finland leading to uniform treatment protocol. In other countries the mutations are variable requiring different treatment protocols.

Conclusions

Growth and graft survival were better in Finnish patients compared with those from other countries. These differences require further study, but may suggest that starting RRT before the decline in eGFR with a bilarectal nephrectomy might be beneficial in this group.

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