

Management Of Children With Congenital Nephrotic Syndrome – Lessons From An ESPN Survey

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Background

Congenital nephrotic syndrome (CNS) is one of the most challenging conditions within the field of pediatric nephrology. The early management remains crucial, both for initial survival, but also to minimize long-term morbidity.

Methods

6-year survey across 17 centers in 11 countries from members of the ESPN Dialysis Working Group

Inclusion criteria:

All patients in a pediatric dialysis unit who are diagnosed with CNS since 01/01/2010 - including those in whom withdrawal of care was performed

Exclusion criteria:

Presentation after the first 3 months of life

Results

Total of 84 children (43 (51%) male)

At birth

	Median	IQR
Gestational age (weeks)	37	35 - 38
Birth weight (g)	2700	2315 - 2060

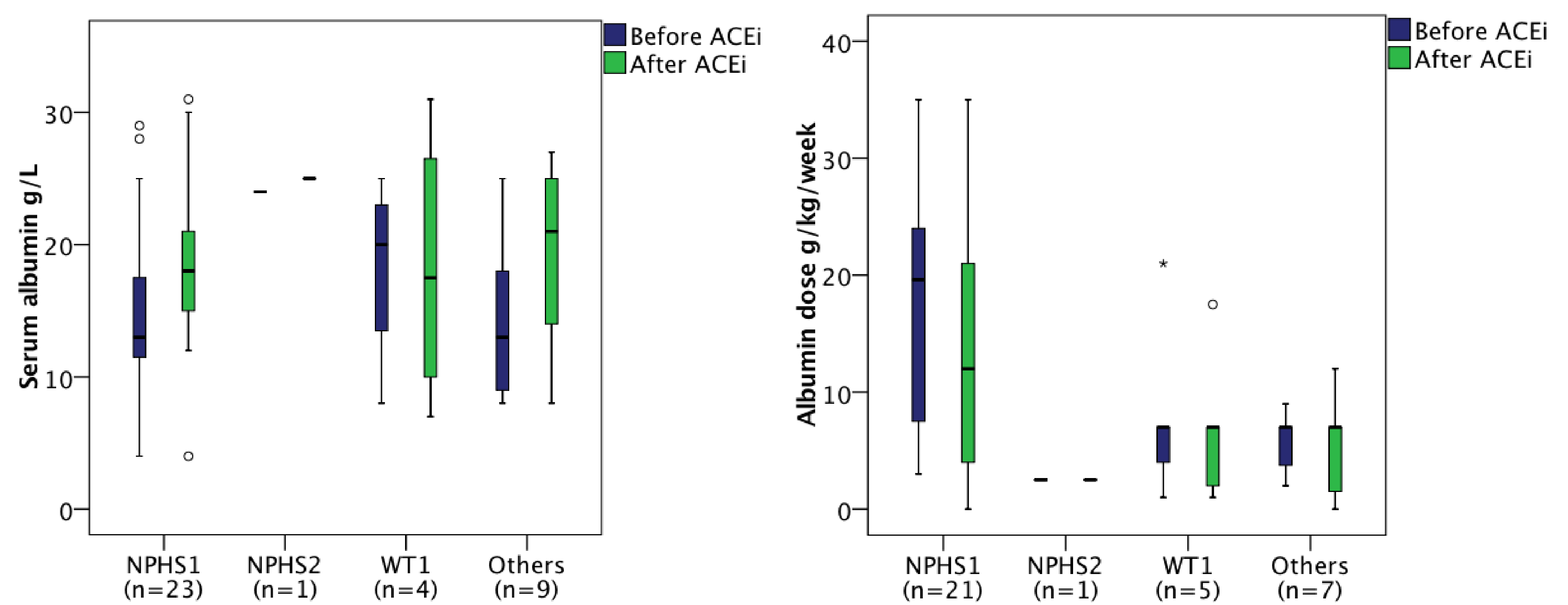
IQR: Inter quartile range

At presentation in tertiary center

	Median	IQR
Age (days)	11	2 - 51
Albumin (g/L)	11	8 - 16
Creatinine (μmol/L)	27	16 - 56

IQR: Inter quartile range

ACE Inhibitor



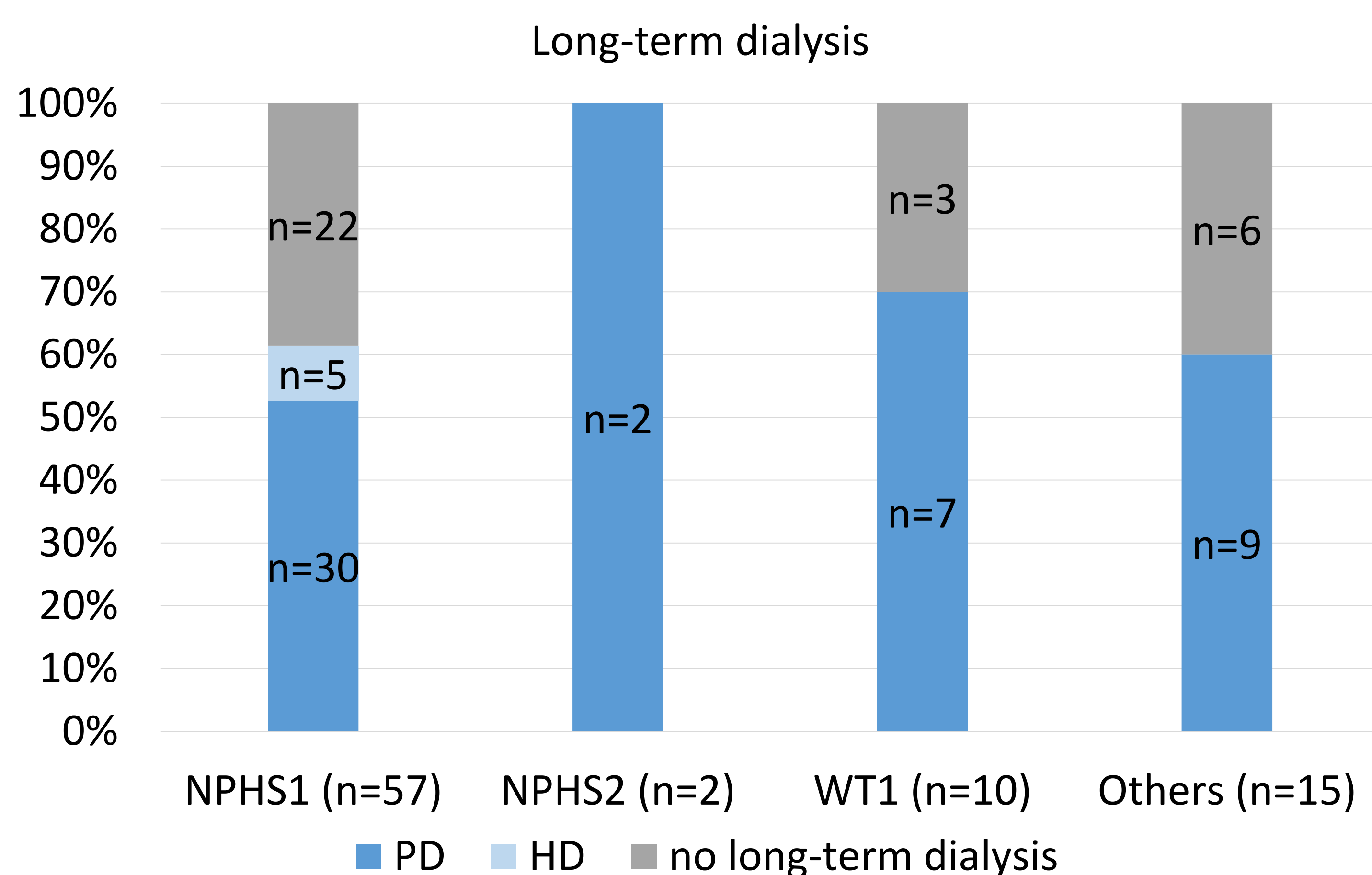
Dialysis

Long-term dialysis was initiated in 53 (63%)

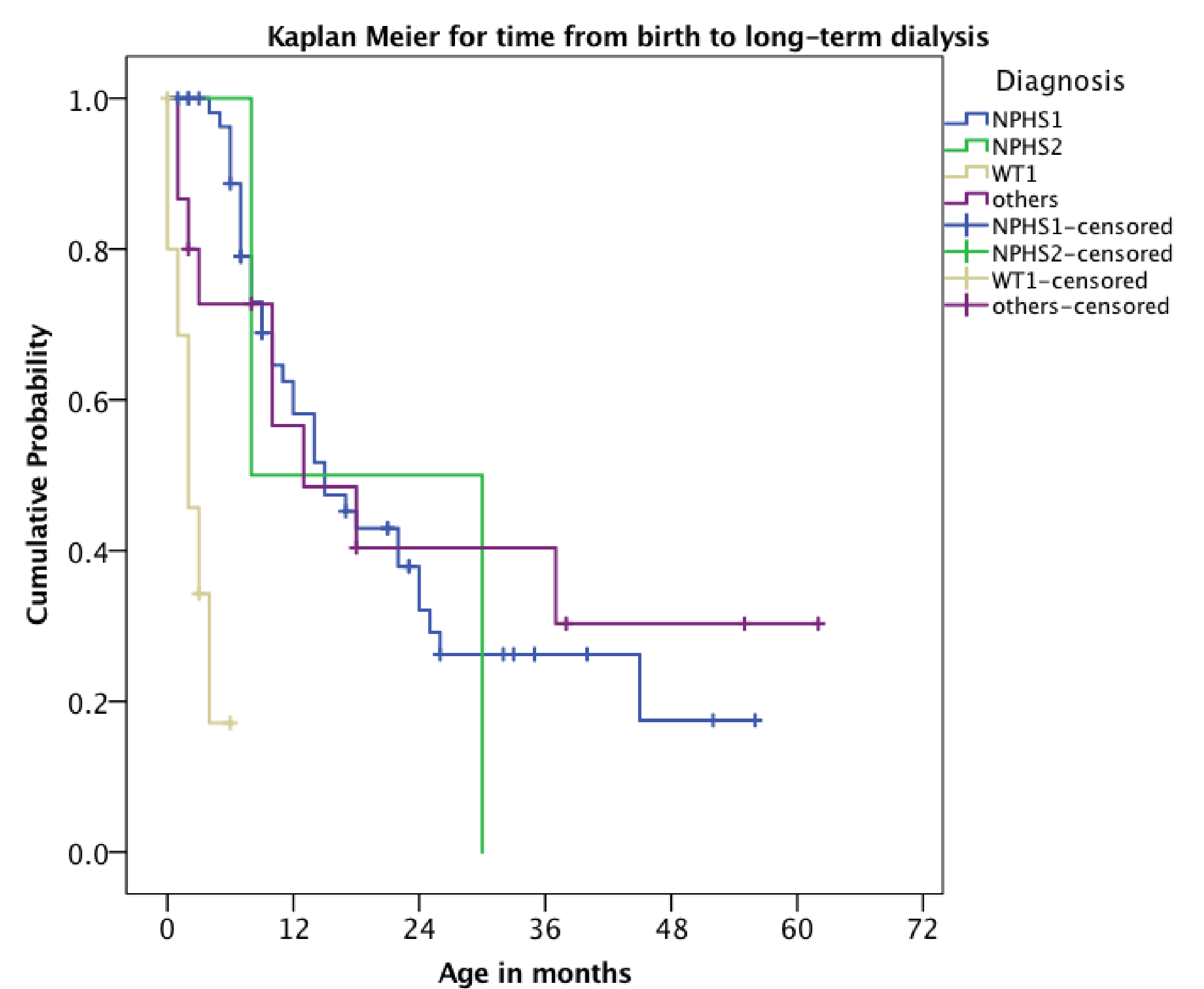
At start of dialysis

	Median	IQR
Age (months)	9	5.5 - 15
Weight (kg)	8.4	6.4-9.6
Albumin (g/L)	22	17 - 27
Creatinine (μmol/L)	246	188 - 294

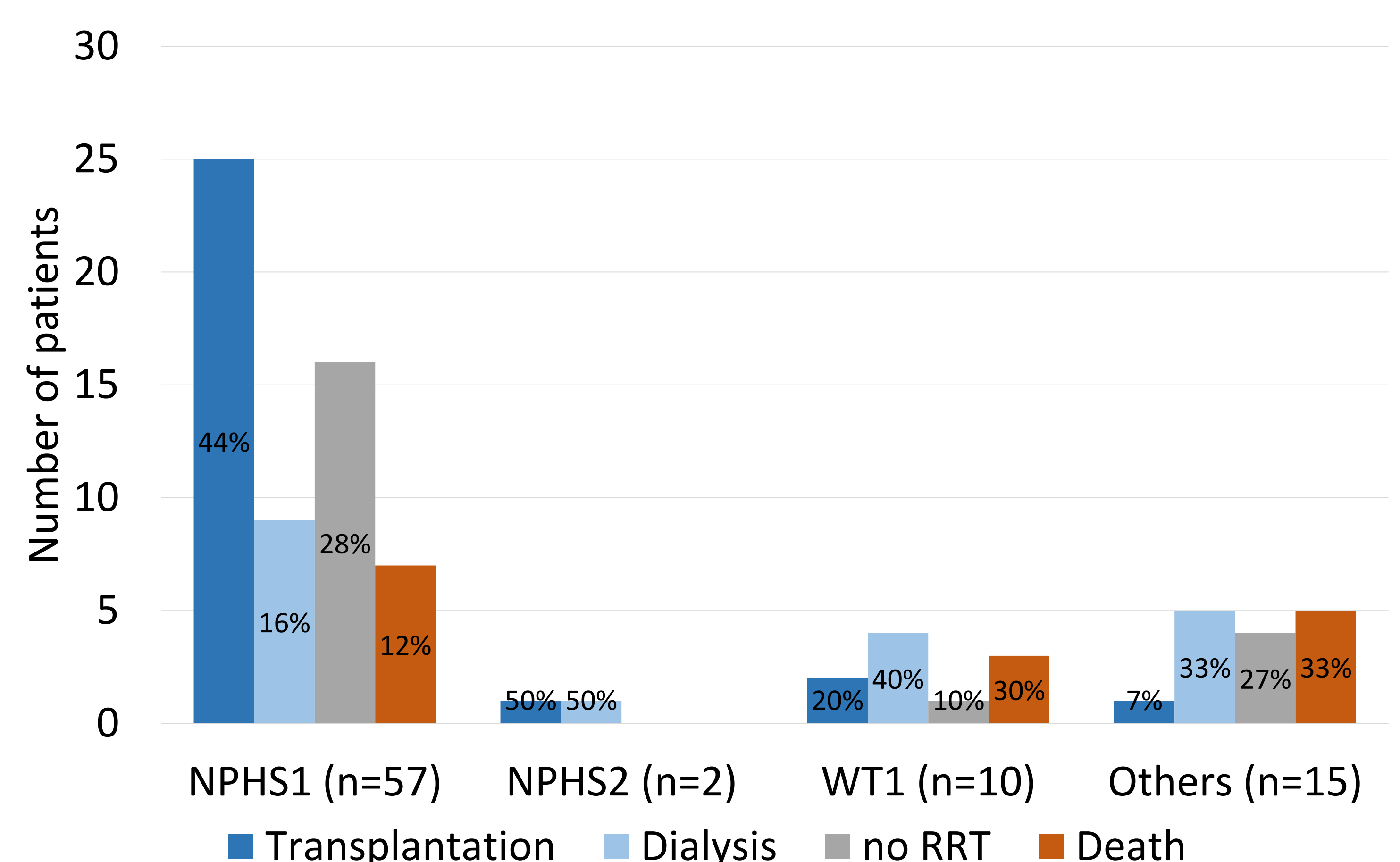
IQR: Inter quartile range



Survival curve for requirement of long term dialysis (n=84)



Outcome



Conclusion

Early management in children with CNS may include a trial of antiproteinuric medication independent of the underlying genetic cause for CNS. 50% of children with CNS start dialysis by the age of one year.

