

Bilateral Nephrectomy In Children With Congenital Nephrotic Syndrome – Is It Still The Way To Go?

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Background

Management of children with congenital nephrotic syndrome (CNS) is controversial and includes different approaches ranging from conservative management to unilateral or bilateral nephrectomies to dialysis and transplantation.

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

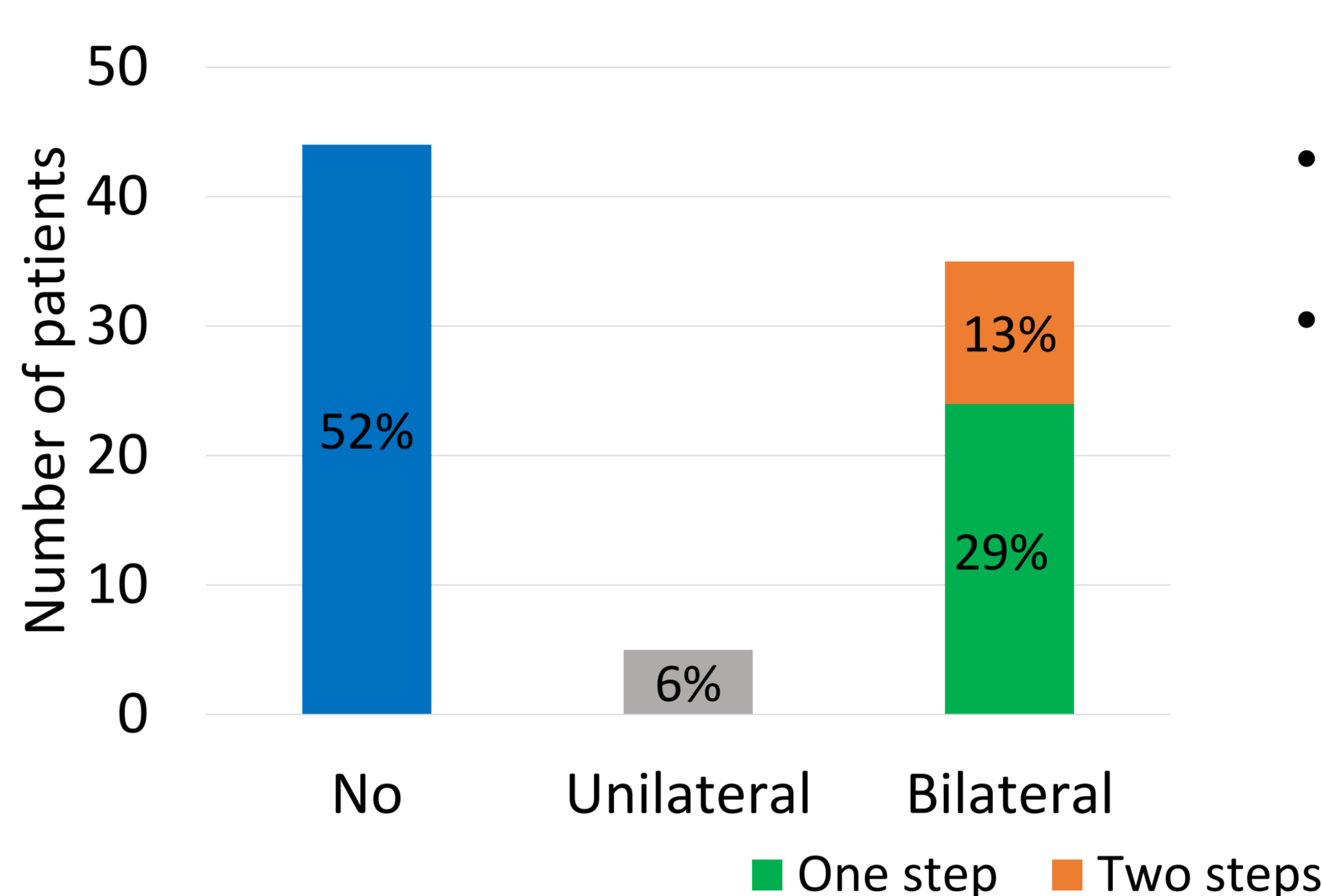
At presentation in tertiary center

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

IQR: Inter quartile range

Nephrectomy

Total of 40 children (48%) underwent nephrectomy



- Age for unilateral nephrectomy: median 4 (IQR 3 - 6) months
- Age for bilateral nephrectomies: first kidney median 6 (IQR 4 - 11) months, second kidney median 8 (IQR 6 - 22) months

Bilateral Nephrectomy vs Conservative Management?

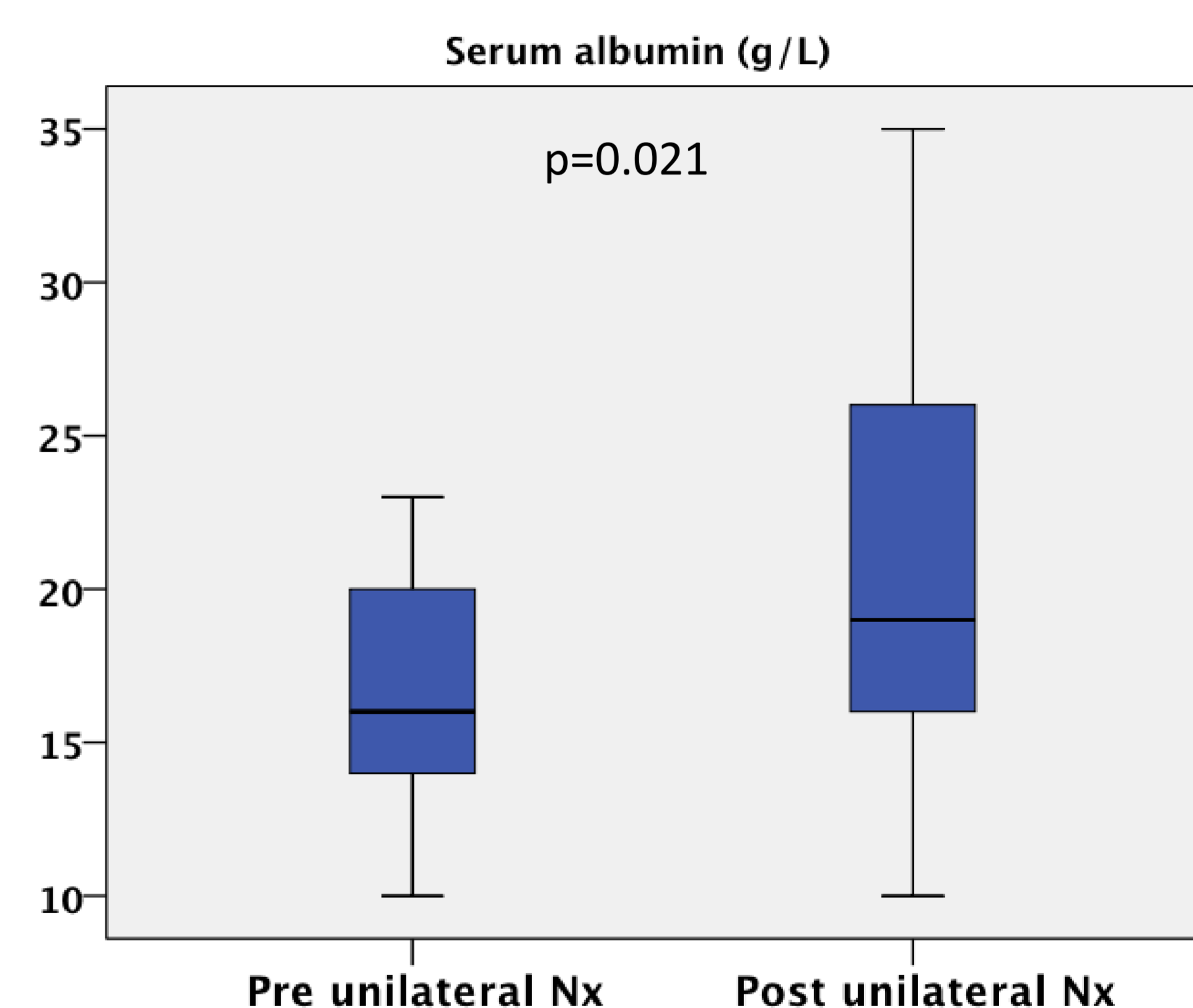
To compare outcomes of nephrectomy, only children with NPHS1 mutations and >12 months follow-up (n=43) were studied:

	Bilateral nephrectomy n = 26	Conservative management n = 17	P value
Gestational Age (weeks)	37 (IQR 34 - 38)	36 (IQR 35-38)	0.70
Birth Weight (g)	2480 (2125-2847)	2510 (2468-2852)	0.52
Parameters at presentation			
Age (days)	3 (0-15)	29 (7-50)	0.01
Creatinine (µmol/l)	20 (16-51)	20 (9-26)	0.19
Albumin (g/l)	8 (7-12)	10 (7-11)	0.21
ACE Inhibitors	8 (31%)	16 (94%)	< 0.01
Antithrombotic medication	21 (81%)	11 (65%)	0.24
Long-term Dialysis			
Age at start (months)	26 (100%)	6 (35%)	< 0.01
	8.5 (7-14)	25 (20-31)	< 0.01
Complications			
Peritonitis	8 (31%)	2 (13%)	0.18
Central line infections	12 (46%)	8 (47%)	0.95
Septic episodes	13 (52%)	9 (53%)	0.95
Thrombus formation	4 (15%)	2 (12%)	0.74
Transplantation			
Living related donor	21 (81%)	4 (24%)	< 0.01
	13	0	
Age at Tx (months)	17 (12-24)	33 (27-45)	< 0.01
Time on dialysis before Tx	6 (4-11)	11 (4-25)	0.34
Survival			
Time of follow-up (months)	25 (96%)	15 (88%)	0.32
	40 (22-49)	33 (22-54)	0.96

Values are represented as median and inter quartile range

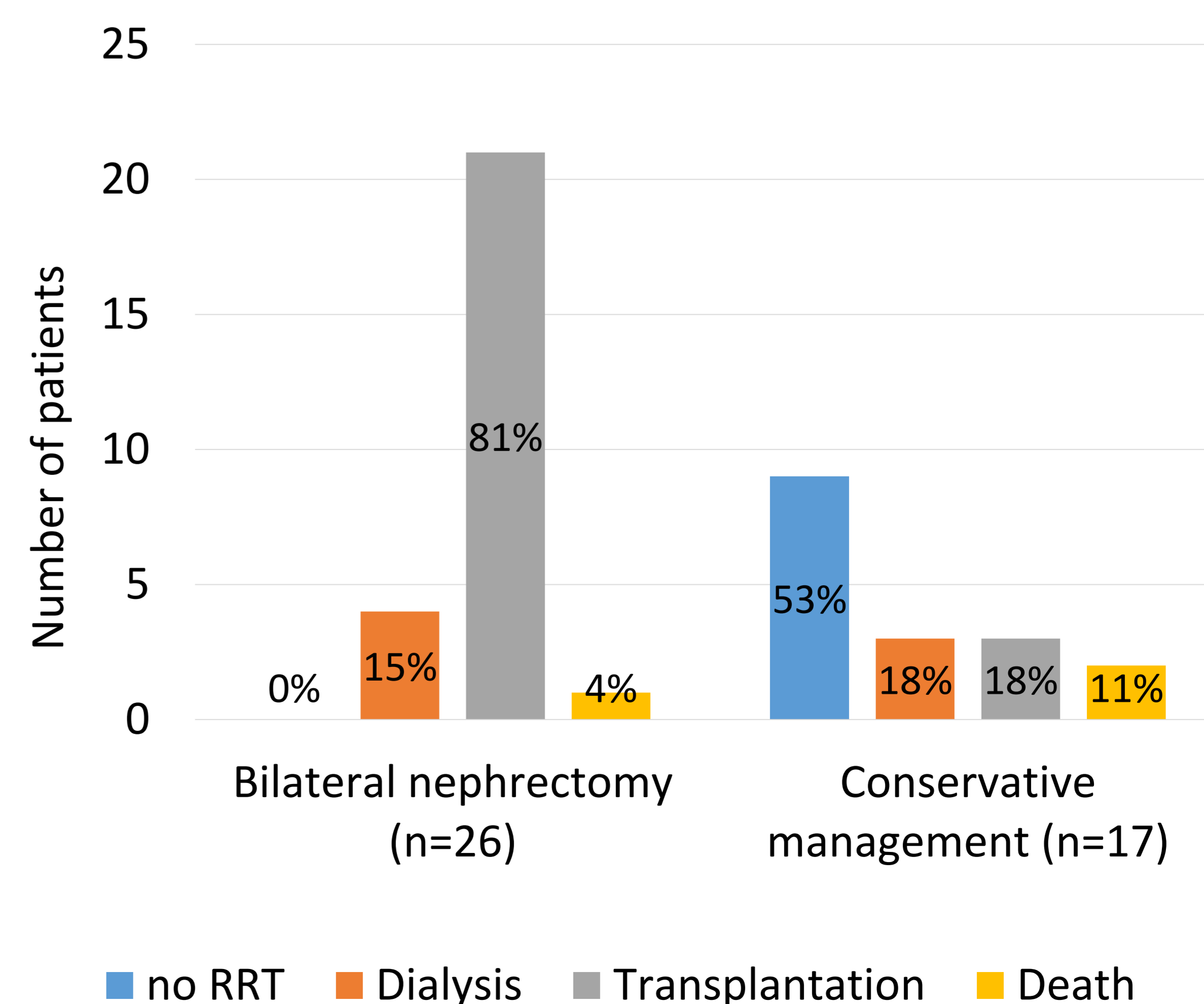
Unilateral Nephrectomy?

Boxplot for change in serum albumin with unilateral nephrectomy for children not on dialysis (n=9)



Reduction of amount of albumin given from 7.5 to 3.6 g/kg/week (p=0.068)

Outcome



Conclusion

An individualised, stepwise approach, with prolonged conservative management, followed by unilateral nephrectomy may be a reasonable alternative to early bilateral nephrectomies in children with CNS. This approach requires further study in large prospective cohorts.

