

# Thromboembolism In Children With Congenital Nephrotic Syndrome – Lessons From An ESPN Survey

Stephanie Dufek<sup>1</sup>, Elisa Ylinen<sup>2</sup>, Agnes Trautmann<sup>3</sup>, Andrea Pasini<sup>4</sup>, Gema Ariceta<sup>5</sup>, Tuula Holtta<sup>2</sup> and Rukshana Shroff<sup>1</sup>

<sup>1</sup> Great Ormond Street Hospital for Children NHS Foundation Trust, London, UK, <sup>2</sup> University of Helsinki and Helsinki University Hospital, Helsinki, Finland, <sup>3</sup> Center for Paediatric & Adolescent Medicine, Heidelberg, Germany, <sup>4</sup> Azienda Ospedaliera Universitaria Sant'Orsola-Malpighi, Bologna, Italy, <sup>5</sup> Hospital Materno Infantil de la Vall d'Hebron, Barcelona, Spain

## Background

Thromboembolism is a serious and potentially fatal complication in children with congenital nephrotic syndrome (CNS). Due to rarity of the disease there are no established guidelines for prophylaxis or management of thromboembolism and outcome remains uncertain.

## 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 and developed thromboembolism

## 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

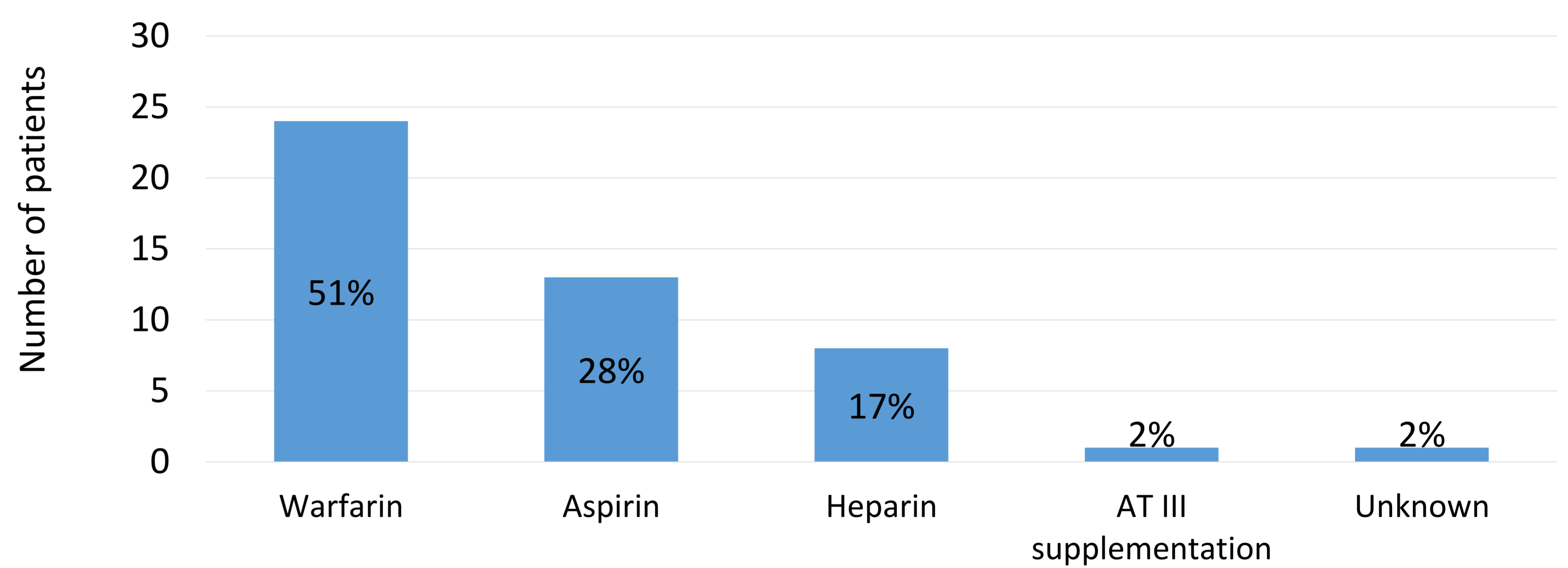
#### At presentation in tertiary center

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

IQR: Inter quartile range

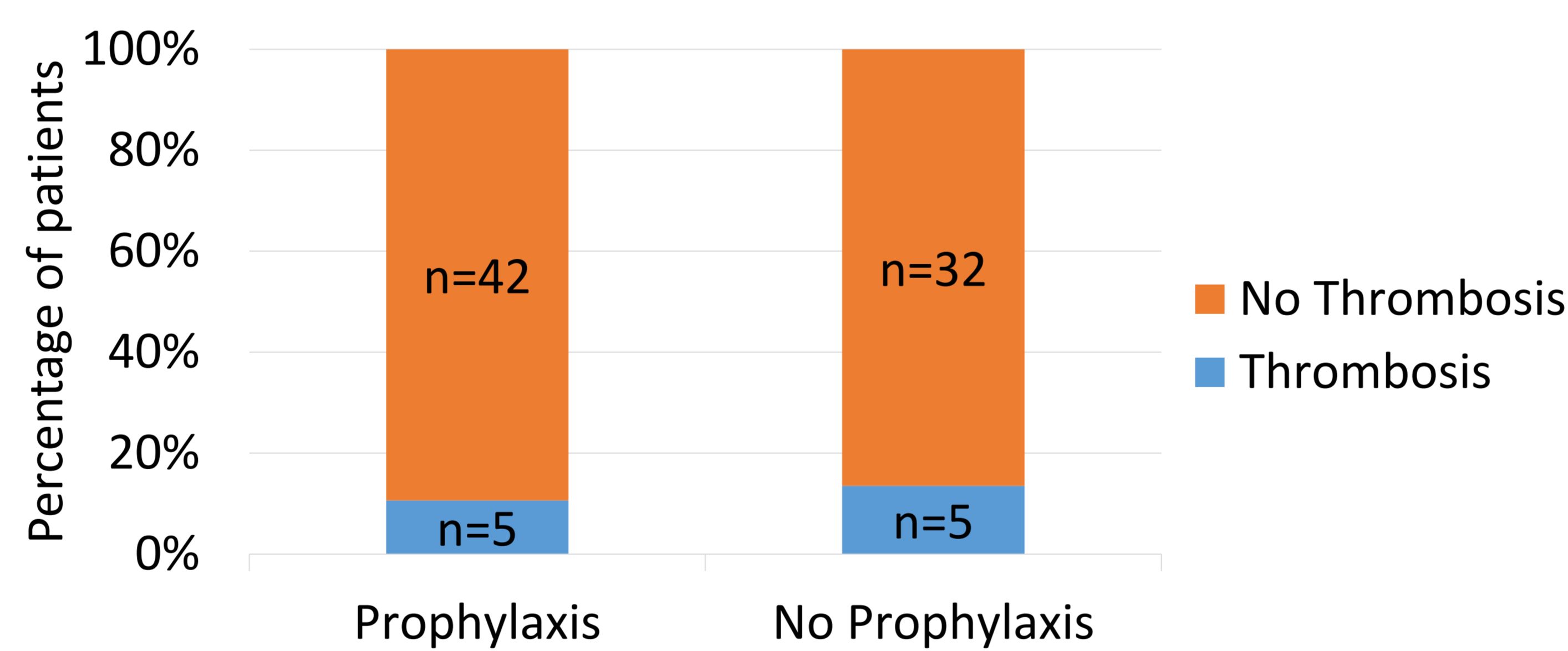
### Antithrombotic Prophylaxis

- Routine antithrombotic prophylaxis was used in 10 (59%) centres as part of their policy for CNS management.
- 47 (56%) children received antithrombotic prophylaxis



### Thromboembolism

- A total of 10 (12%) patients developed thrombosis
- 5 of 47 on prophylaxis versus 5 of 37 not on prophylaxis (p=0.686)



#### Symptoms of thrombosis

- Catheter malfunction: n = 3 (30%)
- Asymmetric extremity: n = 2 (20%)
- Convulsion: n = 1 (10%)
- Fever with malaise: n = 1 (10%)
- Asymptomatic: n = 3 (30%)

### Details on patients with thromboembolism (n=10)

ID	Genetics	Age (months)	Prophylaxis	ACE inhibitor	Diuretic treatment	Dialysis	Dehydration	Line related	Location	Treatment	Outcome associated with thrombosis
1	WT1	2	No	Yes	Yes	No	No	Yes	Right common femoral vein, distal iliac vein	Heparin	Withdrawal of treatment
2	NPHS1	<1	No	No	Yes	No	No	No	Sinus sagittalis	Enoxaparin	Convulsion, hydrocephalus
3	NPHS1	47	No	Yes	No	Yes, HD	No	Yes	Right jugular vein	Heparin	na
4	NPHS1	1.5	No	Yes	Yes	No	Yes	Yes	Bilateral internal jugular veins, superior and inferior vena cava, femoral and iliac veins	Heparin, Enoxaparin	Ongoing thrombosis with collateral formation
5	NPHS1	<1	No	No	Yes	No	No	Yes	Internal jugular vein	Warfarin	Complete resolution
6	NPHS1	12	ASA	Yes	Yes	No	No	Yes	Vena cava superior	Heparin	Complete resolution
7	NPHS1	1.5	Warfarin	No	Yes	No	No	Unclear	Right Atrium	Warfarin + Thrombolysis	Complete resolution
8	NPHS1	3	Warfarin	No	Yes	No	No	No	Vena cava inferior + right renal vein	Enoxaparin	Partial resolution
9	NPHS2	2	Heparin	Yes	Yes	No	No	Yes	Right popliteal vein until right external iliac vein	Heparin, Enoxaparin	Complete resolution
10	NPHS1	1	Warfarin	No	na	No	na	na	Vena cava superior	na	na

na = not available; ASA = Aspirin

## Conclusion

In infants with CNS the risk of thrombotic episodes is high despite prophylactic treatment leading to significant morbidity and mortality. Prospective studies to determine an optimal anticoagulation regime in children with CNS are required.

