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PAEDIATRIC NEPHROLOGY

MP823 **MANAGEMENT OF CHILDREN WITH CONGENITAL NEPHROTIC SYNDROME - LESSONS FROM AN ESPN SURVEY**

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INTRODUCTION AND AIMS: Congenital nephrotic syndrome (CNS) is one of the most challenging conditions within the field of paediatric nephrology. Patients present in infancy with severe proteinuria, hypalbuminaemia and oedema and may suffer from life-threatening complications secondary to profound hypoalbuminemia. The early management remains crucial, both for initial survival, but also to minimize long-term morbidity. Due to rarity of the disease it is extremely difficult to establish clinical trials or develop evidence based guidelines.
METHODS: We conducted a 6-year survey across members of the European Society for Paediatric Nephrology Dialysis Working Group to evaluate early management strategies as well as dialysis management of children with CNS across Europe.
RESULTS: 82 children (52% male) were included in the 6-year survey across 19 tertiary nephrology units in 11 EU countries. Median age at presentation was 9.5 (inter quartile range 0-164) days with serum albumin 11 (4-29) g/L and serum creatinine 27 (2-480) μmol/L. Albumin infusions were given in 73 patients (89%) at a median age of 10 (2-46) days with a dose of 1.5 (1-3.3) g/kg/day and 7 (3.6-7) sessions per week. Antiproteinuric treatment was given in 47 (57%) with an overall response rate (defined as an increase in serum albumin 4 weeks after start) of 70%. In those, delta increase of serum albumin was 7 (1-14) g/L ($p<0.001$) and the frequency of albumin infusion could be reduced significantly ($p=0.006$). Response rate to antiproteinuric treatment for NPHS1 was 70%, for NPHS2 100% and for WT1 50%. Antithrombotic medication was given prophylactically in 47 (57%) children and after first thrombotic event in 5 (6%) starting at a median age of 28 (16-81) days. Long-term dialysis was initiated in 52 (63%) at a median age of 8.5 (6-16.5) months. Of those, 17 (33%) were on dialysis by the age of 6 months, 36 (70%) by the age of 1 year and 47 (92%) by the age of 2 years (see figure 1). PD was the modality of choice, however 34% needed to switch to HD. There was no difference in complication rate and growth between PD and HD patients. At final follow-up at a median age of 31 (0-73) months, 27 (33%) were transplanted, 19 (24%) on dialysis and 21 (26%) did not require renal replacement therapy. 15 children (18%) died at a median age of 7 (0 - 53) months. Causes of death were sepsis in 7, hyperkalaemia in 1, respiratory failure in 1, severe prematurity in 1, sudden cardiac arrest in 2 and withdrawal of active treatment in 3.
CONCLUSIONS: 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.

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