WHAT'S HOT WHAT'S NEW - AN UPDATE FROM THE IPNA 2016 MEETING

CKD-MBD

1. This international survey describes the high prevalence (~35%), increased morbidity (longer hospital stays) and >4 fold higher mortality in neonates who develop AKI. Preventable causes of AKI need to be identified followed by prompt recognition and management of AKI.

FP-S13-1 - Assessment of Worldwide AcuteKidney Injury Epidemiology in Neonates (AWAKEN): Incidence and Outcomes from an International Multi-Center Cohort Study

C. Mammen[1], J. Jetton[2], L. Boohaker[3], R. Griffin[3], D. Askenazi[3], .. On Behalf of the neonatal kidney collaborative[4]

[1]University of BC and Children's and Women's Health Center, Vancouver, Canada
[2]University of Iowa, Iowa City, Iowa, United States
[3]University of Alabama at Birmingham, Birmingham, United States
[4]Canada, India, Australia, United States

Objectives: Single center studies report neonatal acute kidney injury (AKI) incidence of 12-70%; neonates with AKI appear to have worse outcomes. These findings prompted the formation of the Neonatal Kidney Collaborative (NKC) and the development and implementation of the AWAKEN study. The objective of this study is to assess the incidence of neonatal AKI and its association with outcomes in a large, multi-national cohort

Methods: NKC includes 24 institutions from 4 countries (USA, Canada, Australia and India). Neonatal intensive care unit (NICU) admissions from Jan 1 to Mar 31, 2014 were screened. Inclusion criteria need for intravenous (IV) fluids ≥48 hrs. Exclusion criteria: 1) admission at > 2 weeks of age, 2) congenital heart disease requiring surgical repair at < 7 days of age, 3) lethal chromosomal anomaly, 4) death within 48 hours of admission, 5) severe congenital kidney disease. AKI was defined as either urine output (UOP) < 1 mL/ kg/hr during any 24 hour period fromdays 2-7 of life, a rise in serumcreatinine (SCr) of 0.3 mg/dL from previous lowest value and/or receipt of renal replacement therapy.

Results: Of 4,274 screened neonates, 2163 met study criteria in whom 143 (6.5%) had insufficient SCr and UOP data, 765 (35.4%) had AKI, and 1257 (58.1%) did not have AKI. Subjects diagnosed with AKI had higher mortality than those without AKI (58/765 (8.4%) vs. 21/1257 (1.7%) (OR = 4.8; 95% CI = 2.9 -8.0); P< 0.001) and prolonged length of stay (LOS) (45.8 + 45.1 vs. 23.2 + 22.1 days, P < 0.0001). (Table)

Table: Clinical Outcomes by AKI status

	ANY AKI			AKI MAX Stage				
	NO (n=1257)	YES (n= 765)	p-value	0 (n=1257)	1 (n= 223)	2 (n=210)	3 (n=332)	P-value
Survived			<0.0001					<0.0001
Yes	1236 (98.3)	707 (92.4)		1236 (98.3)	204 (91.5)	197 (93.8)	306 (92.2)	
No	21 (1.7)	58 (7.6)		21 (1.7)	19 (8.5)	13 (6.2)	26 (7.8)	
Length of Stay (Days)	23.2 ± 22.1	45.8 ± 45.1	<0.0001	23.2 ± 22.1	21.0 ± 21.2	47.5 ± 38.3	61.5 ± 53.0	<0.0001

*141 Enrolled patients removed from analysis as they had less than 2 SCr and no UOP data collected.
**Among the subjects who did not die, 306 were transferred for convalescence or escalation of care

Conclusions: Our study demonstrated that at least 35.4% of infants admitted to NICU who required IV fluids develop AKI. Subjects with AKI have worse increased risk of death and longer LOS. Our data contribute to the growing evidence that neonatal AKI is common, with significant impact on clinical outcomes. Further analysis of this robust database is underway.

FP-S10-1 - Congenital nephrotic syndrome of the Finnish type, ESPN/ERA-EDTA Registry data

T. Holtta[1], M. Bonthuis[2], K. Jager[2], J. Groothoff[3], K. Van Stralen[2], J. Harambat[4], F. Schaefer[5]

[1]Children's Hospital, University of Helsinki, HUS, Helsinki, Finland
[2]ESPN/ERA-EDTA Registry, Deparment of Medical Informatics, AMC, University of Amsterdam, Amsterdam, Netherlands
[3]Emma Children's Hospital AMC, University of Amsterdam, Amsterdam, Netherlands
[4]Department of pediatrics, Bordeaux University Hospital, Bordeaux, France
[5]Center for Pediatrics and Adolescent Medicine, University of Heidelberg, Heidelberg, Germany

Objectives: Congenital nephrotic syndrome of the Finnish type (CNF) is a rare congenital renal disease with heavy proteinuria, hypoproteinemia ja edema caused by defect in the nephrin gene. There are scarce data on outcome of renal replacement therapy (RRT) for chidlren with CNF. Here we present registry data for 170 children with genetically proven CNF at the time of starting RRT (dialysis or pre-emptive renal transplantation, RTX).

Methods: ESPN/ERA-EDTA registry data were collected between January 1, 1991 and December 31, 2011. Patients were identified from 11 European countries contributing the registry. Since CNF is most prevalent in Finland and therapeutic approach differs from most other countries, outcomes between Finnish and non-Finnish patients were compared. Age-matched patients form the same registry with congenital anomalies of the kidney and urinary tract (CAKUT) served as controls.

Results: Median (IQR) age at RRT start was 0.9 years (0.6-1.9). The Finnish CNF patients (n=66) were significantly younger than non-Finnish patients (n=104), both at start of RRT (0.7 vs. 1.7 years) and time of RTX (1.6 vs. 3.0 years). Six percent of patients received a pre-emptive RTX, 80.8%initiated RRT on peritoneal dilaysis (PD) and 13.2% on hemodialysis. After a median dialysis time of 0.9 years (0.6-1.7) 88.8% of the CNF patients had received a RTX (32.4%living donors). The overall 5-year patient survival on RRT (91%) and graft survival (89%), were similar to controls and in Finnish and non- Finnish patients. At the start of RRT height SDS was higher in Finnish compared to non-Finnish patients (median (IQR): -1.41 (-1.93 to -0.68) and -2.4 (-3.24 to -1.67), $p<0.01$). However, at 5 years of age height and BMI SDS were similar to those in controls.

Conclusions: Overall 5-year patient and graft survival of CNF patients was excellent and similar to CAKUT controls with equally early RRT onset and were not affected by timing of entering RRT and RTX.

3. This study follows from the registry data above and challenges the current dogma of managing children with congenital nephrotic syndrome by performing routine (protocol driven) nephrectomies followed by dialysis. Unilateral nephrectomy may reduce the need for albumin infusions and allow a longer dialysis-free period. There is no difference in the risk of infections or thrombotic episodes and growth is comparable with both approaches.

FP-S25-06 - Dialysis management of children with congenital nephrotic syndrome

S. Dufek[1], T. Holtta[2], E. Ylinen[2], A. Trautmann[3], C.P. Schmitt[3], E. Vidal[4], A. Edefonti[5], R. Shroff[1]

[1]Great Ormond Street Hospital for Children NHS Foundation Trust, London, United Kingdom
[2]Children's Hospital, University of Helsinki and Helsinki University Hospital, Helsinki, Finland
[3]Center for Pediatric & Adolescent Medicine, Heidelberg, Germany
[4]University-Hospital of Padova, Padua, Italy
[5]Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy

Objectives: Dialysis management of children with congenital nephrotic syndrome (CNS) is challenging.

Methods: We conducted a 5-year survey across the ESPN Dialysis Working Group members to review the dialysis approach in children with CNS.

Results: Data on 61 children (32 male) from 12 centres were analysed. Shortterm dialysis (<4 weeks) was required in 5 (8%) children (PD in 2, CVVH in 2 and HD in 1) at a median age of 11 (5-16) weeks for a median duration of 16 (8-18) days secondary to sepsis (n=2), acute fluid overload (n=2) or AKI following ACEI (n=1). Thirteen (21%) patients required long-term dialysis by the age of 6 months, 30 (50%) by 1 year, 38 (62%) by 2 years, 41 (67%) by final follow up at a median of 34 months. Of those, 9 (22%) patients did not have nephrectomies, 29 (71%) had nephrectomies before and 3 (7%) after commencingdialysis. In 37 (90%) PD was the modality of choice. Amongst those on PD, CCPD with a day exchange was the commonest prescription (43%). Eleven (30%) patients needed to switch to HD due to PD catheter dysfunction (n=5), peritonitis (n=3), inadequate UF (n=2) or development of pleuro-peritoneal fistula (n=1). Peritonitis rate was 0.95/12 patient months. None developed thrombosis on PD. All patients on HD received 3 sessions of 4 hours per week.No HD line infections and no development of thrombosis were reported. There was no difference in growth between PD and HD patients. Twenty-five patients (61%) received a transplant at a median of 6.5 (0-47) months after start of dialysis of which 15 with living donor. One patient died whilst on PD (palliative treatment) and 2 patients died whilst on HD (hyperkalemia and candida peritonitis).

Conclusions: The need for long-term dialysis in children with CNS is very likely by the age of 1 year. PD is the modality of choice, however the peritonitis ratewas higher than recommended, butmay reflect the high percentage of infants in this study. A significant number switched between dialysis modalities.