been associated with nephronophthisis (NPHP) and Jeune asphyxiating thoracic dystrophy, both of which are ciliopathies, but also with focal segmental glomerulosclerosis (FSGS), a glomerular kidney disease. During this search, there was a notable incidence (57%) of hypertension out of the 56 total cases, 84% of which aligned with a p.(Pro209Leu) variant. Searching the Genomics England 100,000 Genomes Project revealed one additional case of arterial hypertension associated with biallelic TTC21B variants.

Conclusion

In conclusion, biallelic variants in TTC21B have been shown to produce a wide spectrum of kidney phenotypes, resembling both NPHP and FSGS. This mixed tubulointerstitial and glomerular disease can often present with early-onset hypertension, and the diagnosis may be overlooked due to the lack of typical ciliopathy features. The addition of TTC21B to the early-onset hypertension gene panels can ensure a timelier genetic diagnosis for this rare tubuloglomerular kidney disease (figure 1).

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Conclusion Higher grade VUR result in high chance of renal scarring. However, both higher grade and lower grade VUR can cause renal damage, microalbuminuria, and small contracted kidneys, hence need to follow up until adolescence or later life.

REFERENCES
HEALTH-RELATED QUALITY OF LIFE, PERSONALITY AND PSYCHOLOGICAL MALADJUSTMENT, EMOTIONAL AND BEHAVIOURAL PROBLEMS, AND FAMILY IMPACT IN CHILDREN WITH NEPHROTIC SYNDROME

1Randula Ranawaka, 1Prasad Chathurangana, 1Haitha Lijanaarachchi, 1Prasanjith Dissanayake, 1Danush Mhiran, 1Supun Maranithunga, 1Niranga Devanarayana, 1Shaman Rajindrajith, 1University of Colombo; 2The National Hospital of Sri Lanka, Colombo; 3Faculty of Medicine, University of Kelaniya

Aims Nephrotic Syndrome (NS) is the commonest chronic glomerular disease of childhood. A majority (70–80%) have relapsing disease persisting throughout childhood causing a significant psychosocial impact on both children and their families. Objectives of this study were to assess the quality of life (QOL), personality and psychological maladjustment, emotional and behavioural problems, and family impact in children with NS compared to a matched healthy control group.

Methods A case-control study was conducted in the nephrology clinic, Lady Ridgeway Hospital for Children, Colombo, Sri Lanka. Personality and psychological maladjustments were assessed using the Personality Assessment Questionnaire. The Child Behavior Checklist and Strengths and Difficulties Questionnaire were used to assess the emotional and behavioural problems and psychological symptoms, respectively. The family impact was quantified using the Impact on Family questionnaire. The centre for epidemiological studies depression (CESD) scale was used for the assessment of depressive symptoms of mothers. The quality of life of children was assessed using the Health-Related Quality of Life questionnaire. Data were analysed using the R language for statistical computing.

Results The nephrotic syndrome (NS) group and the control group contained 151 (age 6.7 ±4.03 years) and 125 (age 7.95 ±4.31 years) subjects respectively.

A statistically significant difference of the means was found between the control and NS groups for personality assessment (p<0.05), psychological problems score (p<0.001), family impact (p<0.001), depressive symptoms of mothers (p<0.001) and health-related quality of life (p<0.001).

The score for the strengths and difficulties significantly correlated with all the other 5 test scores. Strong positive correlations were observed between the family impact and the depressive symptoms scores of mothers (r = 0.66, p-value < 0.001), family impact and child behavioral checklist scores (r = 0.58, p-value < 0.001) and strengths and difficulties and child behavior checklist (r = 0.55, p-value < 0.001).

Conclusion Nephrotic syndrome had a significant psychosocial and behavioural impact on the child as well as the family. Care should be taken to address these issues when treating nephrotic syndrome in children.

CONGENITAL CHYLOTHORAX FLUID MANAGEMENT STRATEGIES IN NEONATES: A 6-YEAR RETROSPECTIVE REVIEW

Efthychia Thodi, Alessandra Glover Williams, Jan Dudley, University Hospitals Bristol and Weston NHS Foundation Trust

Aims To describe a single, tertiary-NICU’s experience of neonatal chylothorax over a 6-year period (2016-2021) examining fluid management strategies and associated survival.

Methods Retrospective single-centre review of neonates with congenital chylothorax as defined by a diagnosis made on or before day 30 of life without an operant cause.

Results 10 neonates were diagnosed with congenital chylothorax. Average gestational age was 33±3 weeks (29-36±4 weeks) with a mean birthweight of 2515grams (1230-4100g).

Analysis showed an overall survival rate of 60% and a mean length of stay of 52.3 days (19-106 days). 90% (9) of the neonates were diagnosed antenatally; 88.8% (8) of these underwent pleuroamnionic shunting. Infants who received in utero intervention had a 62.5% survival rate; the infant who did not receive antenatal intervention did not survive.

Survival rate of neonates diagnosed with both congenital chylothorax and hydrops during their admission was 33.3% (2/6) while survival of those with chylothorax alone was 100% (4/4).

There was no correlation between maximum fluid input per day (ml/kg/day) and mortality noted. The maximum amount of fluid given to any infant in the cohort was 263ml/kg/day; this infant survived.

Human albumin solution (5%, 20%) was administered in 90% of the neonates included in the cohort. 70% received mostly 5% HAS with a survival rate of 57%. 20% received a 50/50 combination of 5% and 20% HAS, survival was 50%. Of those who did not receive HAS, survival rate was 100%.

Diuretics were given as stat doses or as a continuous infusion, with neither method being associated with improved survival.

Peritoneal Dialysis was commenced in 2 patients; neither patient survived.

Of note, mortality was strongly associated with other serious accompanying conditions (e.g. Noonan’s syndrome, Twin-To-Twin-Transfusion Syndrome), as previously recognised in the literature.

Conclusion Although limited by small numbers and single-centre cohort, this retrospective analysis adds new fluid management strategy detail to the literature on the challenging management of chylothorax. Approach continues to be perplexing and treatment always requiring of individualisation dependent on associated diagnoses. Continuing larger, detailed cohort studies are needed to accumulate data on a group of patients that holds potential for systematic review and meta-analysis in what is a rare diagnosis.

HYPER EOSINOPHILIA: A MULTISPECIALITY DIAGNOSTIC APPROACH

1Noman Choudhery, 2Matt Harmer. 1Bristol Royal Hospital for Children; 2Southampton Children’s Hospital, Southampton, UK

Aims Severely increased blood eosinophils (ie, > 5 x 10^9/L), whether discovered incidentally on a full blood count or found with signs and symptoms of associated organ involvement, warrant diagnostic workup and often therapeutic interventions. Our aim was to systematically evaluate Hyper-eosinophilia and its secondary effects in a patient with Chronic kidney disease.