Aims To determine the accuracy of phase contrast microscopy in predicting urine culture results.

Design and methods A prospective study comparing the results of phase contrast microscopy interpretation of urine samples with that of urine culture. Samples were microscoped at the time culture was being performed.

The sample size though ongoing was based on the availability of the clinician to get to the laboratory at the time of urine culture. Uncentrifuged, unstained urine samples were examined with an Olympus BH2 microscope enabled with phase contrast at 400x magnification mounted on a Hawksley Nebauler counting chamber. Samples were interpreted as being either "Positive," "Negative" or "Indeterminate" based on their level of bacteriurea. "Indeterminate" specimens would be repeated in the clinical setting and were not further analysed in our laboratory based study.

Results 65 samples were microscoped. Immediate determination was made for 62 samples (95.4%). 3 samples were deemed as "Indeterminate."

Abstract G66 Table 1

	Culture Positive	Culture Negative	Total
Microscopy Positive	15	3	18
Microscopy Negative	2	42	44
Total	17	45	62

Of the 62 microscopy interpretations 57 (91.9%) showed concordance with the microbiology culture results. Sensitivity of 15/17 (88.2%), specificity 42/45 (93.3%), positive predictive value 15/18 (83.3%) and negative predictive values of 42/44 (95.5%) were obtained. Video evidence is available showing bacteria in 2 of the samples deemed "Microscopy Positive" but "Culture Negative." If these samples, as they ought to, returned as "Culture Positive" then microscopy would have attained concordance 59 (95.2%), sensitivity 17/19 (89.5%), specificity 42/43 (97.7%), positive predictive value 17/18 (94.4%), negative predictive value 42/44 (95.5%).

Conclusions Phase contrast microscopy afforded immediate interpretation in 62/65 (95.4%) of samples studied. Microscopy interpretation showed high concordance rate, sensitivity, specificity, positive and negative predictive values when compared to the accepted gold standard i.e., microbiological culture even with the apparent error in the culture process.

G67(P) DENT'S DISEASE COMPLICATED BY AN ACUTE BUDD-CHIARI SYNDROME: CASE REPORT

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We present the case of a young boy with Dent's disease, identified as having a mutation in the kidney-specific chloride-proton antiporter CLCN5 during investigation for nephrotic range proteinuria. He went on to develop a growth hormone deficiency requiring treatment with recombinant growth hormone followed by an acute presentation with hepato-renal failure and thrombotic occlusion of both middle and right hepatic veins consistent with a diagnosis of Budd Chiari syndrome, which required a prolonged period of intensive care. We have identified 3 reports in the literature in which growth hormone therapy has been used to treat short stature associated with Dent's disease. This report confirms that growth hormone deficiency is a recognised finding in this rare disease. There are no previous reports on thrombotic complications associated with either Dent's disease or the use of recombinant growth hormone *per se*. The cause of the Budd-Chiari syndrome in this case has yet to be fully elucidated, but potentially widens the Dent's phenotype and gives support to the previous observation that this disease is multifaceted with a possible role for as yet unidentified environmental and/or genetic modifying factors.

G68(P) VESICOURETERIC REFLUX – AN UNUSUAL GENETIC ASSOCIATION OF COMMON CONDITION

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Background Vesicoureteric reflux is the most common urological anomaly in children and is a common cause of end stage renal failure and hypertension in children. It usually occurs in isolation and in vast majority of cases it improves on its own. However rarely it can be associated with coloboma and renal hypoplasia, consequent to PAX2 gene mutation.

Case Report We report a case of 14 year old boy, who was under regular paediatric follow-up since the age of 5 month when he had first episode of urinary tract infection. He was put on trimethoprim prophylaxis and micturating cystourethrogram (MCUG) revealed bilateral grade 3 reflux without obstruction at the age of 8 months. He was lost to follow up for 3 years, delaying investigation. Dimercaptosuccinic acid (DMSA) scan at 4 years of age showed small right kidney with no scarring and differential function of 33%. On routine eye check at age of 5 years he was noted to have bilateral optic disc coloboma, however his vision was not affected. He was noted to have mild impairment in renal function at age of 6 yrs with eGFR of 63ml/min/1.73m^2 which remained stable for next 6 year. During this period he also became significantly obese (body mass index 33) with significant disproportion. He remained normotensive during this period. His renal function started deteriorating for past couple of years with the development of proteinuria. His diagnosis was revisited and was thought to be due to PAX2 gene mutation.

Discussion Paired box (*PAX*) genes play a critical role in human development and disease. The *PAX2* gene is expressed in primitive cells of the kidney, ureter, eye, ear and central nervous system and is required for normal kidney and eye development. Mutation in PAX2 gene has been described in the several families with optic nerve coloboma, renal hypoplasia, mild proteinuria and vesicoureteric reflux.

Conclusion We report this case to raise awareness amongst paediatricians about this uncommon genetic association of vesicoureteric reflux with renal hypoplasia and coloboma which can lead to progressive renal failure.

G69(P) PAEDIATRIC RENAL TRANSPLANTATION WITH BARDET-BIEDL SYNDROME (BBS) AND SITUS INVERSUS TOTALIS

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Introduction Bardet-Biedl syndrome (BBS) is a rare autosomal recessive disorder characterised by a genetic dysfunction that causes cystic malformation of the kidneys alongside features such as postaxial polydactyly, central obesity and a spectrum of learning difficulties. A rare cause of renal failure in children that ultimately requires transplantation at a very young age. We report the first case of successful renal transplantation in a 3 and a half year old child with both BBS and situs inversus totalis .

Patient and Methods The patient had commenced peritoneal dialysis 14 months prior to transplant due to end-stage renal failure. Following transplant workup her father was deemed a suitable donor and a right sided hand assisted laparoscopic donor nephrectomy was performed. The recipient then underwent the transplant with a midline incision to allow access to the aorta and inferior vena cava to which the anastomoses were made. Anatomical reversal of the usual techniques used in paediatric transplantation were needed due to situs inversus. The kidney was then placed in the left iliac fossa. Native kidneys not removed and the abdomen was closed with peritoneal dialysis catheter removed at time of transplant.

Results The patient recovered in the paediatric intensive care unit as per standard transplantation at the unit. Ultrasound imaging was performed immediately post operatively and showed good global perfusion of the kidney with no hydronephrosis or perinephric collection. The patient developed immediate graft function and was discharged with standardised outpatient follow up.

Discussion Six cases of BBS and renal transplantation have been reported in literature and this is the first case report of a successful renal transplantation in a child with both BBS and situs inversus totalis. We therefore conclude that such anatomical malformations should not be considered a contra-indication for renal transplantation in children.

Paediatric Mental Health Association

G70 FOREIGN ACCENT SYNDROME (FAS) IN ASSOCIATION WITH AUTISTIC SPECTRUM DISORDER (ASD). A NEW SYNDROME?

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Background FAS has been described in adults following acute injury to the brain. Children commonly develop an accent closer to that of their peers than their parents' native accent. Children with ASD often have a speech and language disorder. There have been no recorded cases of children speaking with a foreign accent that they have not been exposed to.

We present 3 British children with Asperger's syndrome who speak with an American sounding accent with no prior exposure to it. We will show DVD clips of their speech.

Case 1:
White English girl, 8, with unilateral hearing loss following congenital CMV infection. Attending main stream school with a statement of special educational needs (SEN), but with significant social and communication difficulties; diagnosed as Asperger's syndrome. Fluent speech with a strong American accent.

Case 2:
White English boy, 9, with CHARGE syndrome, attending main stream school with a statement of SEN; good speech, but with poor social interaction skills and significant obsessional behaviour; was diagnosed as Asperger's syndrome. Speaks with a clear American accent.

Case 3:
Black British boy, 6 with behavioural difficulties, in main stream school with significant social and communication difficulties; diagnosed with Asperger's syndrome. Has clear speech with an American accent.

Discussion There are over 90 cases reported from many parts of the world in adults who have suddenly developed a foreign accent following trauma to the brain such as stroke, waking up after anaesthesia, migraine, brain tumour. It is thought to be due to involvement of speech area of brain.

There has been no report of a child with FAS.

None of our children have had any documented acute brain trauma but 2 of them have some co- morbidities. Their unexplained foreign accents have baffled both parents and professionals alike.

Conclusions We hypothesise some children with ASD could have had a brain injury of an obscure nature leading to both the behavioural difficulties and pronunciation disorder that sounds like a foreign accent. We suggest that the combination of ASD and FAS is a new syndrome.

Paediatricians with Expertise in Cardiology/ British Inherited Metabolic Disease Group

G71 A REVIEW OF 24-HOUR AMBULATORY HOLTER ELECTROCARDIOGRAPHY

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Background The 24-hour ECG is being widely used by paediatricians and paediatric cardiologists to assess children who present with palpitations or other cardiac symptoms. It is also used for children who are known to have an arrhythmia or a cardiac condition, have a family history of serious arrhythmia or suffer from a metabolic disease which can affect their heart.

Aims To review the effectiveness of 24-hour ECG in capturing arrhythmias in children.

Methods This was a retrospective review of the 24-hour ECG reports between January 2009 and January 2012. The patients were identified from the cardiology database and the reports were accessed via the electronic medical record system of our hospital.

Results Over this 3 year period, 178 Holter ECGs were performed. That included 72 female and 106 male patients. The age range was from 4 days to 16 years, with most Holters performed between 1 month to 2 years of age (27%). Most patients (90%) had an ambulatory ECG which lasted for 24 hours, while on 9 occasions it lasted for 48 hours and on 8 occasions for 72 hours. The indications for requesting this investigation were divided into 5 categories: known arrhythmia, known cardiac disease, non-specific palpitations (including chest pain, shortness of breath and 'funny turns'), family history of prolonged QTc and SUDS and screening due to metabolic disease (Fabry's). 34 patients (19%) were on medication at the time of the Holter ECG, one had an implanted defibrillator and another one had a pacemaker. In total, significant abnormalities were found in 29 (16%) patients, including SVT, second degree heart block and frequent sinus pauses. Quite importantly, abnormalities were not detected amongst the patients who were being investigated for non-specific palpitations. Of note, 10 such patients experienced symptoms during their 24-hour ECG study; in all cases the recorded rhythm was sinus rhythm.

Conclusion In this study, the 24 hour ECG did not reveal any rhythm abnormalities in the patients being investigated for non-specific palpitations and other associated symptoms. However, it demonstrated important abnormalities in patients under follow-up for known arrhythmia, heart disease or relevant family history.

G72 THE EVOLVING ROLE OF A PAEDIATRICIAN WITH EXPERTISE IN CARDIOLOGY IN A NON-CARDIAC CENTRE

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Aim The Safe and Sustainable Review of Paediatric Cardiac Surgery will change the way that paediatric cardiology is delivered in the United Kingdom[1]. Paediatricians working with cardiologists in