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 microscopied 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 bacteriuria. "Indeterminate" specimens would be repeated in the clinical setting and were not further analysed in our laboratory based study.

Results 65 samples were microscopied. 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

doi:10.1136/archdischild-2013-304107.079

¹C Platt, ²J Dudley, ³J Hartley. ¹General Paediatrics, Gloucestershire Hospitals NHS Foundation Trust, Gloucester, UK; ²Paediatric Nephrology, United Hospitals Bristol NHS Foundation Trust, Bristol, UK; ³Department of Hepatology, Birmingham Children's Hospital, Birmingham, UK

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

doi:10.1136/archdischild-2013-304107.080

M Gupta, R Wasala. Department of Paediatrics, Sandwell and West Birmingham Hospitals NHS, West Midlands, UK

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/1.73m² 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

doi:10.1136/archdischild-2013-304107.081

¹JH Frost, ²A Bagul, ³IH Mohamed, ³MG Mould, ⁴HK Jahn, ⁵G Koffman. ¹Oxford University Hospitals NHS Trust, Oxford, UK; ²Guys and St Thomas NHS Foundation Trust, London, UK; ³Royal North Shore Hospital, Sydney, Australia; ⁴Leicester Royal Infirmary, Leicester, UK

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 post-axial 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.