G434(P) FAMILIAL DIABETES MELLITUS: 2 SIDES OF A COINP Vasista, P Sundaram. *Paediatrics, Leicester Royal Infirmary, UHL NHS Trust, Leicester, UK*

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Aim To understand the difference in presentation of an inherited condition depending on the mode of inheritance.

Case Report *Case A:* 11 years old girl with history of tiredness, normal BMI, had high blood glucose BG (7.1 mmol/L) and HbA1c (45 mmol/mol). There was family history of Type 2 Diabetes mellitus (DM) in mother, diagnosed at age of 23 years.

Oral glucose tolerance test:

	BG (mmol/L)
0 min	6.2
60 min	9.8
120 min	8.0

HbA1c	45 mmol/mol
C-peptide	616 (298 – 2350 pmol/l)
Insulin	10.3 (4.4– 26 mIU/l)
Insulin antibodies	< 2 mg/L
Islet cell antibody	Negative
Anti GAD antibody	< 5 IU/ml

Blood Glucose (BG)	19 mmol/l
Insulin	<2 mIU/L (4.4 –26)
C-peptide	42 pmol/L (298–2350)
IA2 antibody	<10 IU/ml (0–10)
Anti GAD antibody	<5 U/ml (0–10)
Islet cell antibody	Negative
Thyroid function test	Normal

Genetic test confirmed MODY – *Heterozygous* GCK mutation. She did not require any treatment and had stable HbA1c on further regular monitoring. *Case B* - 10 days old term female baby, weighing 1.88 Kg (<0.4th centiles), admitted in hospital for jaundice, was found to have glycosuria and hyperglycaemia (BG =24 mmol/L). Her mother had gestational Diabetes and father had type 2 DM.

Genetic test confirmed MODY – *Homozygous* GCK mutation. She was managed with IV fluids and intravenous insulin infusion and then started on sensor augmented insulin pump. **Conclusion** MODY GCK mutation accounts for ~15% of the familial DM. Glucokinase acts as 'glucose sensor' for secretion of insulin in pancreas. Mutation of GCK gene results in 'resetting' of the glucose sensors, causing delayed secretion of insulin.

The manifestation of symptoms varies with type of mutation in GCK-MODY. The commoner *heterozygous* mutation is asymptomatic with stable mild hyperglycaemia and no treatment is required. The rarer *homozygous* mutation presents as permanent neonatal DM requiring life-long insulin therapy.

G435(P) HYPERTENSION IN CHILDREN WITH DIABETES

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Aim To Monitor the persistence of abnormal blood pressure recordings in young age diabetes patients.

Method The data was collected from a Paediatric diabetes unit of a DGH for national Paediatric diabetes audit for the year 2017 – 18. There were 106 children in the age group of 12 – 18 years. We classified any blood pressure reading above the 95th centile for the age, sex and height as hypertension.

Results There were 27(25.4%) patients out of 106 who had at least one blood pressure reading above the 95th centile. 16 were male and 11 females (table 1, 2 and 3).

Abstract G435(P) Table 1 Ethnic origin

Ethnic origin	Numbers
White British	15
Asian Pakistani	4
Black African	2
Asian Bangladeshi	1
Asian Other	1
Any white	1
Mixed white and black African	1
Mixed white and black Caribbean	1
Not tested	1

Abstract G435(P) Table 2 BMI

Category	BMI centiles	Numbers
Obese	>98th	5
Overweight	91–98	5
Normal weight	2–91	15
Underweight	<2	2

Abstract G435(P) Table 3 Blood pressure readings

Number BP reading	No. of patients	4 abnormal readings	3 abnormal readings	2 abnormal readings	1 abnormal reading
4	7	0	1	0	6
3	8	-	1	0	7
2	7	-	-	2	5
1	5	-	-	-	5

23 out of 27 patients (85%) had only one abnormally high BP, 2 (7.4%) had 2 readings and 2 (7.4%) had 3 abnormally high BP.

Conclusion Majority of children with hypertension had only one abnormal reading, only with 15% of the patients a high blood pressure was recorded more than once.

One abnormal high blood pressure may not be indicative of hypertension in young people with diabetes. If blood pressure recordings are multiple and persistently raised, ambulatory blood pressure monitoring may need to be considered.

G436(P) AUDIT AND QUALITY IMPROVEMENT OF THE NATIONAL HEALTH SERVICE ENGLAND (NHSE) SPECIALISED SERVICE FOR CHILDREN WITH ALSTROM SYNDROME

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Background Alstrom syndrome is a rare (1:1,000,000) multi-system condition associated with infancy-onset obesity and insulin-resistant diabetes during puberty. It is unknown whether deterioration of renal function begins in childhood and whether this relates to glycaemic control.

Primary Aims To audit children attending the national service (n=24) against the standard of annual BMI, renal, liver function, and glucose control and compare to the 2018 audit.

To design BMI-SDS graphs to allow clearer indication of BMI progress.

Methods Case notes and laboratory data were reviewed for 2015–2019. eGFR was calculated using the Counahan-Barratt method.

Results Screening of blood pressure, U&Es and urine albumin creatinine measurements (target level:80%) has improved from 10/24(42%), in the first audit cycle, to 15/22(68%).

11/20(55%) had a reduction in eGFR (median decline 25 ml/min/yr, range:9–58 ml/min/yr) (declining eGFR group) compared to 9/20 (45%) (stable eGFR group). Comparing the declining eGFR to the stable eGFR group, median age was 14.5 yrs (4–17.9 yrs) vs 10.3 yrs (6.5–16.8 yrs); 6/11 (55%) had HbA1c $\geq$ 6.5%(median 7.3% (5.7–7.7)) vs 9/20 (45%); 7/11 (64%) were overweight ($\geq$ 2 BMI-SDS) vs 6/9 (67%); 4/11 (36%) were obese ($>$ 3 BMI-SDS) vs 1/9 (11%); and 8/11(73%) were of South Asian origin vs 3/9 (33%).

The average BMI-SDS remained at $>$ 2.67SDS ($>$ 99.6th centile) in the Asian cohort of Alstrom patients over the 4 years, but decreased from $>$ 2.67SDS to 1.5SDS in the White European cohort.

Conclusions Higher BMI-SDS, South Asian origin, older age and poorer glycaemic control were associated with a reduction in eGFR over time. The co-occurrence of these features at puberty may relate to a 'metabolic syndrome' phenotype associated with high levels of insulin resistance known to occur in mid puberty. The results for the South Asian cohort suggest the tendency for the phenotype to remain obese into adulthood.

Recommendations Prospective studies should investigate the drivers to loss of glycaemic control and reduction in eGFR around puberty and the reasons for the ethnic differences in BMI-SDS. Improved monitoring in childhood and earlier

pharmacologic and lifestyle interventions may be required to prevent eGFR deterioration and achieve better glycaemic control. Bespoke BMI-SDS charts may empower children and families for weight management. Culturally specific dietetic advice may be helpful.

G437 VALIDITY OF THE WASHINGTON GROUP MODULE ON CHILD FUNCTIONING IN 2-YEAR-OLD CHILDREN; DISABILITY OUTCOME OF THE SHINE TRIAL

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Introduction The WHO estimate that 1 billion people live with disability, with 80% living in developing countries. For children, this is associated with increased mortality, risk of becoming a victim of violence and decreased school attendance. Through identifying these children early, actions can be taken to improve their quality of life.

Global childhood disability data collection to date has used various tools and methodologies, affecting comparability. In 2016, the Washington Group and UNICEF developed the child functioning module (WGCFM) to identify functional difficulty (FD) in children aged 2–17, for use in census and surveys. Field testing has highlighted a need for further validation. This study aims to identify prevalence of disability according to the WGCFM in over 1500 2-year-old children from the Sanitation Hygiene Infant Nutrition Study (SHINE) trial in 2 districts of Zimbabwe, and to assess concurrent validity with the Malawi Developmental Assessment Tool (MDAT).

Methods All consenting pregnant females aged 15–49 were followed up throughout pregnancy and infancy. At age 2, their child was assessed for disability by one of 11 local research nurses using both the WGCFM and MDAT (a culturally appropriate tool for developmental assessment in rural Africa – adapted, translated and back translated for use in Zimbabwe). Quality control and reliability tests were performed on all measures between research nurses. Other measures included anthropometry (e.g. height, weight), maternal HIV status and behavioural measures.

Results 1606 (49.9% female) participants from SHINE were eligible for disability follow-up. FD was identified in 4.17%, severe FD in 0.68%. MDAT and WGCFM scores showed a significant negative correlation, $P<0.001$. Those with FD had a significantly lower MDAT score, $P=0.012$; this became more significant when comparing those with severe FD to no FD, $P=0.002$.

Conclusions Following removal of the 'controlling behaviour' domain, prevalence of FD by WGCFM in rural Zimbabwe is comparable to that in field testing in Mexico, Samoa and Serbia. These results demonstrate that the WGCFM has concurrent validity for use within a population of 2-year-old children in Zimbabwe.