Accounts of disease weakening, and the advent of newer medications have prompted efforts to enhance conservative management. However, treatment continues to be challenged by the condition's heterogenous presentation and varied response to drugs, both of which appear to be genetically influenced. Thus, this study aimed to review the correlation between genotype and phenotype of patients with CHI.

Methods Retrospective review of the phenotype of patients with genetic forms of CHI managed at a specialised centre between 2010 and 2018.

Results The study identified 71 children with genetic forms of CHI. K_{ATP} channel defects (*ABCC8/KCNJ11*) made up the majority of cases ($n=55$). The remaining mutations included *HNF4A*, *GLUD1*, *GCK* and *PMM2*. The median age at first hypoglycaemia for most mutations varied between day 0–2; *GLUD1* probands were an exception with a median presentation of 260 days. Normal birth weights were recorded in *HNF4A*, *GLUD1* and paternally-inherited K_{ATP} defects; remaining subgroups were large for gestational age. Compound heterozygous, homozygous and paternally-inherited K_{ATP} defects made up the bulk of Diazoxide-unresponsive cases but interestingly some showed partial response. All probands with non- K_{ATP} channel mutations expressed either full or partial response to Diazoxide. Three *PMM2* mutation-holders displayed positive response to Diazoxide with adjunctive Nifedipine. Sirolimus and lanreotide were used in up to 12 patients with medically managed K_{ATP} defects. Disease weakening was observed across most genetic subgroups. Natural remission was seen in 1 compound K_{ATP} (1.56 years), 5 paternal K_{ATP} (median age 2.19 years), 5 (50%) maternal K_{ATP} (median age 0.33 years) and 1 *HNF4A* (0.66 years) mutation. Twenty-one (29.6%) patients underwent pancreatectomy. Neurodevelopmental delay was noted in 20 (28.2%) patients.

Conclusion The study reinforces the heterogenous nature of CHI and the potential for genetic understanding to enhance management. Experimental therapies, sirolimus and lanreotide, appear to be promising alternatives to invasive treatment in severe cases. The weakening severity observed across genetic subgroups and the morbidity associated with surgery, appear to support a shift towards conservative therapy.

G414 EVALUATING THE IMPACT OF A 'HIGH HbA1c PATHWAY' IN A PAEDIATRIC DIABETES CLINIC

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Aims HbA1c is an established marker of glycaemic control in diabetes mellitus. A high HbA1c is associated with greater risk of diabetic ketoacidosis, avoidable hospital admissions and long-term complications. In line with national recommendation our paediatric diabetes clinic has a 'high HbA1c pathway' involving intensive support to improve glycaemic control in patients whose HbA1c is ≥ 70 mmol/mol. We evaluated the impact of our 'High HbA1c pathway' on our patient outcomes.

Methods Medical notes of 109 young people on the High HbA1c programme from June 2015 to June 2017 were reviewed retrospectively. Data was analysed using Microsoft Excel.

Results In total 109 patients (53% females), mean age 13 years (range 2–19 years) were commenced on the pathway

during the 2 years period. 54% (59) of patients continued on the pathway for over 12 weeks. 46% (50) of patients were on an insulin multi daily injection regime opposed to pump therapy. 8% (9) had their insulin delivery method changed whilst on pathway.

Mean HbA1c on initiation was 76 mmol/mol, with subsequent measurement at 6 months being 73 mmol/mol (where available). The mean decrease after 12 weeks on the pathway was 7 mmol/mol.

Three patients had social care involvement. 20 patients had psychological input.

No statistical difference was found in the change in HbA1c whilst on the pathway between patients of different sex or ages. 65% of those who had psychological input successfully came off the pathway (HbA1c < 70 mmol/mol). 78% of those in whom attempts at regular contact were successful within 6 weeks of starting on the pathway, subsequently came off the pathway.

Conclusion The High HbA1c pathway has had a mixed impact on our paediatric diabetes population. 46% of patients were able to improve their HbA1c sufficiently to come off the pathway within 12 weeks. Factors such as early contact with patients and access to psychological services seem to lead to better patient outcomes. Some patients' HbA1c didn't improve despite multiple regular contacts. Future work should aim to qualitatively explore what deems a contact successful. New innovative ways of engaging these patients need to be developed in order to reach optimal glycaemic control.

G415 LITERATURE REVIEW OF DIAGNOSTIC TESTS USED FOR THE ASSESSMENT OF GLYCAEMIC ABNORMALITIES OR CFRD IN CHILDREN WITH CYSTIC FIBROSIS (CF)

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Introduction CFRD is the most common co-morbidity in CF.¹ Long-term survival depends on the nutritional status, which influences pulmonary exacerbation and lung function.² Hence need for a reliable and reproducible diagnostic test, which identifies glucose intolerances early for improved survival in CF.

Aim To review recent evidences on various diagnostic modalities used for assessment of abnormal glucose tolerance (AGT) or CFRD by literature review.

Methods Systematic search of Medline and Embase database (up to March 2018) to identify relevant studies using appropriate inclusion & exclusion criteria along with various limits. A comparative analysis was carried out to assess the current evidence base.

Results Five relevant studies were identified for this review. Four studies examined oral glucose tolerance test (OGTT) of which two compared OGTT with CGMS. The prevalence of CFRD was 32–38%. OGTT identified glycaemic abnormalities in 50–60% while CGMS showed abnormalities in 79–93%. The sensitivity of HbA1c was low at 36% (range 11–69%). Self-monitoring of blood glucose (SMBG) with 1 hr postprandial levels offers valuable diagnostic information.

Conclusions Diagnosis of AGT/CFRD continues to be a huge challenge. HbA1c is unreliable as a diagnostic marker. OGTT is still favoured by international societies in spite of high inpatient variability. SMBG requires a huge commitment from patients. CGMS is promising but lack of clinical guidance