consecutive sampling technique and SPSS version 16 was used to analyze data.

Results One hundred patients of T1DM were enrolled, 82 were male and 18 were females. Mean age of the patients were 11.70±2.38 years (10–17 years) and 59% were <15 years. Mean duration diagnosis of T1DM was 7.05±0.7 years. Mild non proliferative Diabetic retinopathy (NPDR) was found in 17% patients and none had proliferative diabetic retinopathy.

Conclusion(s) Mild Non Proliferative Retinopathy is quite high in our study population which could later on Progresses Proliferative Retinopathy. Screening for all the children should be mandatory for early diagnosis, management and future of eye complications. The screening for eye complications for all children Type I diabetes mellitus should be initiated annually after 5 years of diagnosis and in the children who are 10 years are older.

64 A CASE SERIES OF TWO ADOLESCENT DIABETIC SIBLINGS DUE TO NOVEL MUTATION IN CFAP126

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Background CFAP126 gene play a role in the regulation of pancreatic beta cell.

Bader et al in 2016 showed Fltp knocked out mice does exhibit a minor defect in insulin secretion.CFAP126 was significantly downregulated in human islet cells of pre-diabetic as compared to non- diabetic and further downregulated in type 2 diabetics.

To date CFAP126 has not been reported to cause clinical diabetes.

We are reporting 2 siblings with diabetes carrying a heterozygous variant of CFAP126 gene.

Case Report(s) Two siblings (both females) from non-consanguineous parents presented with incidental finding of hyperglycaemia at the age of 16 years (sibling 1) and 13 years (sibling 2). Sibling 1 is asymptomatic and sibling 2 had history of polyuria and polyphagia for last 2 months. There was a strong family history of type 2 diabetes in maternal side including mother, 2 maternal uncle, 4 maternal aunt, and maternal grandmother.

Siblings 1 and 2 had BMI of 16 kg/m2 and 23.07 kg/m2 respectively without any clinical evidence of insulin resistance. Investigation revealed that sibling 1 had HbA1C of 12.3%, insulin level (6.6 IU/L) and C peptide (1.8 ng/ml). Sibling 2 had HbA1C of 12.6%, insulin level (7.3 IU/L) and C peptide (2.02 ng/ml). Both were negative for anti GAD65 and anti-insulin antibodies and had normal celiac screen and thyroid profile. They were started on basal bolus insulin with total dose of 1 unit/kg/day (sibling 1) and 0.7 unit/kg/day (sibling 2) and genetic were sent for MODY.

On 3 month follow up both siblings 1 and 2 were having good control of diabetes with HbA1C of 6.6% and 6% respectively. However, their whole exome sequencing turned out to be negative for all variant of MODY gene, but a heterozygous mutation was identified in CFAP126 gene with variant c.310A>T p. (Lys104*) in both siblings and mother.

After informed consent and counselling, Glibenclamide was started in both sibling with written plan of gradual tapering of insulin if needed. Sibling 2 is on glibenclamide for last 3 months with obvious reduction of insulin requirement from 0.7 unit/kg/day to 0.2 unit/kg/day, recent HBA1C 6.6%, and C peptide 4.82 ng/ml. Sibling 1 was on glibenclamide for last one month with reduction of insulin requirement from 1 unit/kg/day to 0.5 unit/kg/day.

Conclusion(s) Mutation in CFAP126 seems to be associated to cause diabetes which do respond to oral sulfonylurea. Further studies are needed to confirm the affect of this gene on insulin secretion at molecular level.

65 RABSON-MENDENHALL SYNDROME WITH SEVERE INSULIN RESISTANCE TYPE A WE NEED TO ACT FASTER THAN THE DISEASE

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Background Rabson-Mendenhall syndrome (RMS) is an autosomal disorder where severe insulin resistance is observed. Insulin levels decrease over time and suppress gluconeogenesis in the liver. Fatty acid oxidation is affected leading to frequent episodes of ketoacidosis. The changes in RMS with type 2 diabetes are much faster than in patients with type 2 diabetes without RMS. RMS patients have a significantly reduced life expectancy and may die during adolescence or early adulthood.

Case Report(s) A 15-year-old girl with poorly-controlled diabetes. She was diagnosed with RMS at the age of 50 days and her genetic study showed a homozygous mutation for R141W in the INSR gene. Her insulin levels were high at 737 µIU/mL, IA-2, her GAD antibodies were negative and her C-peptide was > 18 ng/ml. There is a strong family history of RMS on her mother's side. For the first six years, her hyperglycaemia was treated with an insulin pump (requiring up to 300 units of insulin/day) and oral Rosiglitazone, after which Rosiglitazone was replaced by oral Insulin-like-growth factor (IGF1). Over the last three years, she had four further episodes of DKA triggered by infections and severe lipodystrophy. A trial of leptin and subcutaneous IGF1 has failed. Currently, the patient is with a closed-loop insulin pump MiniMed 780G with a total daily dose of 261 units (4.6U/kg/day).

Conclusion(s) To act faster than the disease progression, we need to know the whole list of issues our patient could face as this will help us look at the entire picture rather than treating different pieces separately. Effective communication and cooperation between the teams is the key point and needs to be organised through a family physician or by the team most involved in patient care. Although technology has some limitations, it still helps when used appropriately.