Background Cystic fibrosis related diabetes (CFRD) is a combination between reduced insulin secretion and peripheral insulin resistance that only people with cystic fibrosis can get. CFRD is associated with a decline in lung function, poor nutritional status and high mortality rate.

Aim Study of the clinical course and therapeutic management in adolescents with CFRD.

Methods We present 2 cases of CFRD in a female patient (15 years old) and a male patient (17 years old) hospitalised at the CF Centre. The diagnosis of CF was confirmed by positive sweat test (Macroduct USA), identification of CFTR mutation (F508del/F508del), small amounts of elastase in stool. Confirmation of diabetes was achieved by a blood glucose test, blood glucose profile, glycosylated Hb, C peptide, glucose and ketones in urine.

Results On the background of pulmonary exacerbation, both patients had hyperglycemia (9.8/14.1 mmol/l). Subsequently, it triggered clinical syndromes suggestive for diabetes - polydipsia, polyuria, weight loss. Glycemic profile variations 7.8–15.8 mmol/l in boy and 10.4–21.0 mmol/l in girl. Confirmation of diabetes was achieved by a blood glucose test, blood glucose profile, glycosylated Hb, C peptide, glucose and ketones in urine.

Conclusions Patients with CFRD shows a specific clinical framework and require a strict medical diet control and surveillance of the insulin therapy, case that differs from the other types of diabetes. If early introduced, it significantly improves life expectancy towards these patients.

Background Vitamin D status in children with type 1 diabetes.

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Conclusions Patients with CFRD shows a specific clinical framework and require a strict medical diet control and surveillance of the insulin therapy, case that differs from the other types of diabetes. If early introduced, it significantly improves life expectancy towards these patients.

Background and aims Serum vitamin D (sVD) deficiency may contribute to the development and progression of diabetic nephropathy (DN). Research has shown urinary vitamin D binding protein (uVDBP) excretion is increased after renal injury, and is associated with tubulo-interstitial damage. Animal studies suggested that sVD deficiency may be associated with urine loss due to kidney damage. The aim was to test associations of sVD with levels of uVDBP, urinary vitamin D (uVD).

Methods 42 children aged 6–17 years with type 1 diabetes were examined: 24 normoalbuminuric patients (1st group) and 18 microalbuminuric (2nd group). 15 healthy children were included in controls. We measured serum and urine 25(OH)D levels, uVDBP concentrations and tested their correlations.

Results sVD levels were decreased in the patients of the 1st and 2nd groups, compared with controls (22.03 (17.23; 24.44) and 14.42 (12.02; 19.63), compared with 30.65 (28.45; 35.05) ng/ml, respectively) (p < 0.001)). uVDBP levels were elevated in the patients of the 1st and 2nd groups, compared with controls ((179.5 (174.0; 189.0) and 219.0 (216.0; 222.0), compared with 125.0 (116.5;136.0) ng/mg, respectively) (p < 0.001)). uVD levels were increased in the patients of the 1st and 2nd groups, compared with control group ((3.2 (2.9; 3.3) and 3.9 (3.7; 4.1), respectively).
Compared with 2.2 (2.1;2.6) ng/mg, respectively (p < 0.001). The correlations between the levels of sVD and uVD (r = -0.74, p < 0.01), sVD and uVDBP (r = -0.64, p < 0.01) were determined.

**Conclusions** These data suggest that, theoretically, one of the causes of VD deficiency in patients with DN is a urine loss.

**Abstract PO-0068 Table 1**

<table>
<thead>
<tr>
<th>Mean HbA post diagnosis quarter (95% CI)</th>
<th>HbA &gt;7.5% (Numbers/%)</th>
<th>Reduction of HbA levels achieved (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>G1a 8.67(7.87–9.48)</td>
<td>11(78) 21</td>
<td></td>
</tr>
<tr>
<td>G1b 10.45(9.10–11.81)</td>
<td>13(93) 7</td>
<td></td>
</tr>
<tr>
<td>G2a 7.87(7.29–8.44)</td>
<td>12(60) 40</td>
<td></td>
</tr>
<tr>
<td>G2b 7.81(7.33–8.29)</td>
<td>13(65) 35</td>
<td></td>
</tr>
</tbody>
</table>

Conclusions No significant difference of HbA in two groups post diagnosis; mean HbA1C level (p = 0.082). Comparing the changes over time in the two groups, an increase of HbA1C of 1.78 (Table2) percentage points in G1 (without intensive monitoring) was significantly greater than the decrease of 0.06 percentage points in G2 (p = 0.008).

**Discussion** Intensive initial management of type 1 Diabetes can significantly reduce future HbA1C. We aim to follow G2 over next 5 years to establish that an improved metabolic memory could reduce HbA1C levels over longer periods.

**Abstract PO-0069**

**TERMINAL DELETION OF CHROMOSOME 15Q RESULTING IN HAPLOINSUFFICIENCY OF THE IGF-1 RECEPTOR AND MARKED ELEVATION OF IGF-1**

A O’Riordan, N McGrath, F Shirali, MJ O’Grady. Paediatrics, Midland Regional Hospital Mullingar, Mullingar, Ireland

**Introduction** Ten to fifteen percent of small for gestational age (SGA) infants demonstrate failure of catch-up growth. Haploinsufficiency of the insulin-like growth factor-1 receptor (IGF1R) gene due to monosomy 15q is an extremely rare cause with 16 cases reported in the literature. We describe the phenotype of such a patient including biochemical findings, auxology and management.

**Case** A three-year-old female with global developmental delay and autistic spectrum disorder was evaluated for short stature. Height was 82 cm (-3.7 SDS) and weight was 12.5 kg (-1.3 SDS). She was born at term weighing 2.88 kg and was microcephalic and dysmorphic. Array CGH revealed an unbalanced translocation resulting in trisomy of the terminal portion of 15q.

Design A follow up cohort study of 34 children and adolescents in a large district hospital with diagnosis of Type 1 diabetes between 2005 and 2011.

**Results** In the first group (G1=14), without intensive blood sugar monitoring, mean HbA1C post diagnosis was 8.67% (95% CI 7.87–9.48%), 11 out of 14(79%) of them had HbA1C above target level (7.5%). In the second group (G2=20) with intensive monitoring, mean HbA1C post diagnosis was 7.87% (95% CI 7.29–8.44%). 12 out of 20 (60%) had HbA1C above target level (7.5%)

**G1a/b: Group without intensive initial monitoring** 2005/12
**G2a/b: Group with intensive initial monitoring** 2011/12

**Abstract PO-0068 Table 2**

<table>
<thead>
<tr>
<th>HbA trends (%)</th>
<th>95% CI (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>G1a vs. b</td>
<td>+1.78</td>
</tr>
<tr>
<td>G2a vs. b</td>
<td>-0.06</td>
</tr>
</tbody>
</table>

Cl: Confidence Interval