A rare and unexpected cause of diabetes in childhood

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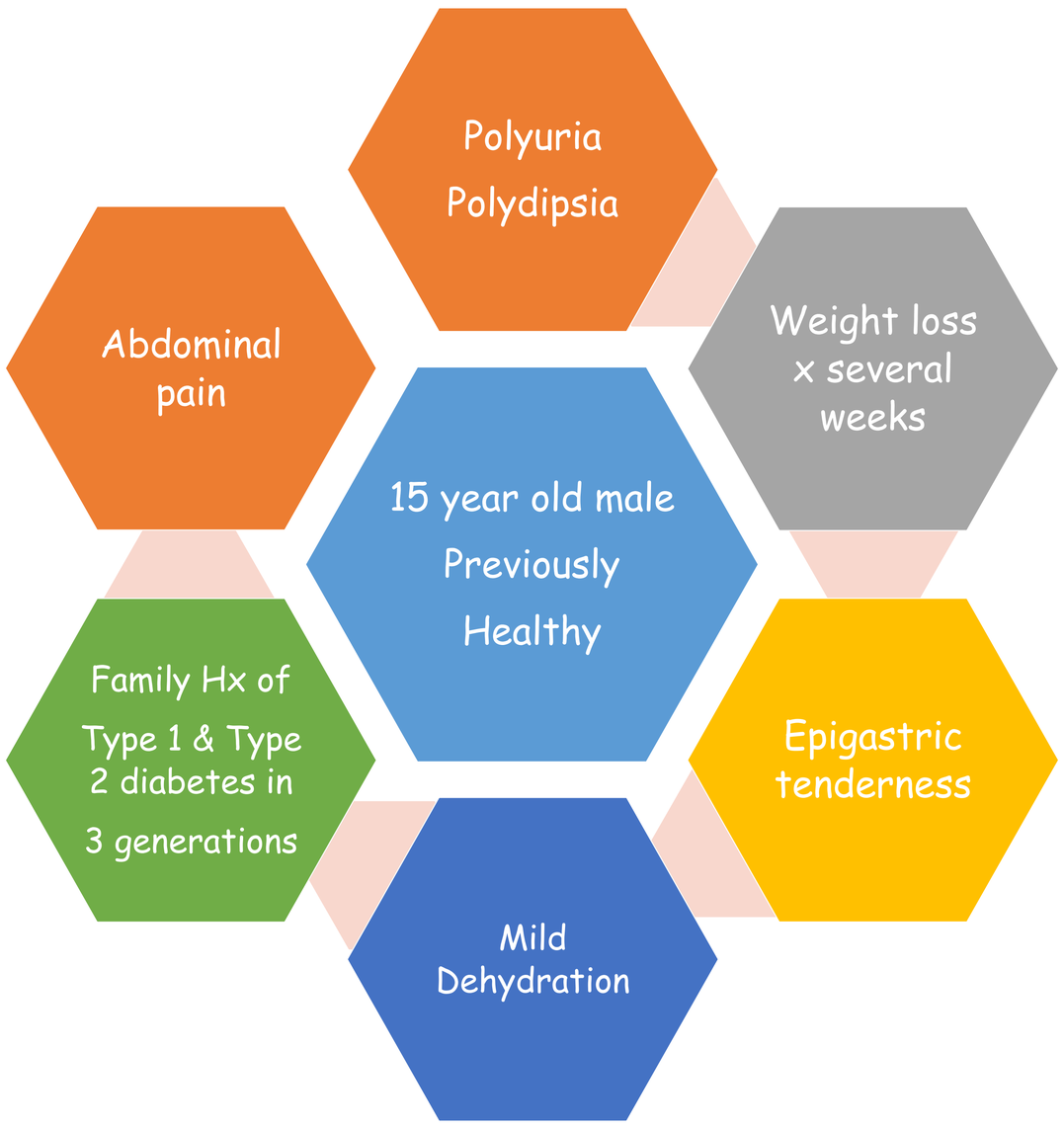
Introduction & Objective:

Pancreatogenic diabetes is rare in children. It is also called **type 3c diabetes mellitus (T3cDM).** The underlying pathophysiology is destruction of islet cells by pancreatic inflammation. The prevalence is 5-10% of all cases of diabetes in the developed world.
We present a case in a teenager, highlighting the challenges in making the correct diagnosis.

At presentation:



Initial blood tests:

Tests	Results	Normal range
Random blood glucose (BG)	18.8 mmol/L	4-7
HbA1c	90 mmol/mol	20-42
Amylase	185 IU/L	25-125
Blood ketones	0.1 mmol/L	<0.3

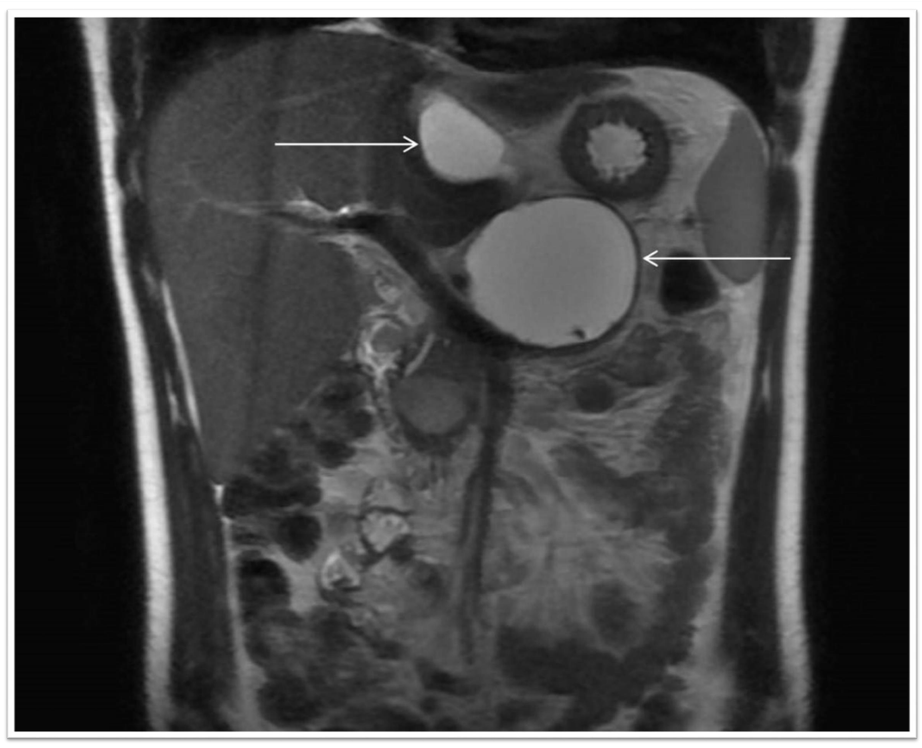
Further tests confirmed diabetes, but the type remained undetermined.

Oral glucose tolerance test	0 hour	2 hours
Blood glucose	7.3mmol/L	18.8mmol/L
C-peptide	274pmol/L	498pmol/L

- Glutamic acid decarboxylase, Islet antigen 2, Islet cell antibodies were negative.
- Started on subcutaneous insulin (Glargine and NovoRapid®).
- Next generation sequencing for all known monogenic diabetes genes were negative.

Following weeks:

On-going central abdominal pain, significant weight loss, early satiety and pain on eating larger portions raising suspicion of gastric outlet obstruction (GOO). Clinical signs with high repeat amylase (415 IU/L) prompted an urgent abdominal MRI.



MRI abdomen showed two pancreatic pseudocysts, the largest one measuring 11cm in diameter. It also revealed pancreatic duct disruption.

Results:

Investigations confirmed diabetes secondary to chronic pancreatitis - also referred to as T3cDM.

Following endoscopic drainage of the pseudocyst his symptoms of GOO quickly resolved. His insulin requirement is slowly weaning. He was commenced on vitamin D as levels of fat soluble vitamins showed vitamin D deficiency.

Further tests looking at genetic causes for idiopathic pancreatitis (SPINK1 and PRSS1 genes) were negative.

Discussions:

T3cDM is a complex condition. It is often complicated by co-morbidities such as malabsorption and malnutrition. It is mis-diagnosed as T2DM in over 87% of patients, but requires insulin therapy more urgently than those with T2DM.

Making the diagnosis of T3cDM is important in order to appropriately manage both the exocrine and endocrine pancreatic insufficiency.

Take home messages:

- Early pancreatic imaging is recommended in suspected cases.
- Avoid alcohol and smoking.
- Consider oral pancreatic enzyme supplementation.

References:

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2. Ewald N, Kaufmann C, Raspe A, Kloer HU, Bretzel RG, Hardt PD. 'Prevalence of diabetes mellitus secondary to pancreatic diseases (type 3c)'. *Diabetes Metab Res Rev.* 2012;28(4):338–342. https://doi.org/10.1002/dmrr.2260
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