Caution-the failed pancreas transplant

Choudhury M, Boregowda K, Bolsusani H, Roberts A, Dayan C.

Department of Diabetes and Endocrinology at University Hospital Wales Cardiff and Cardiff University School of Medicine

Case History

- 38 Year old male with Alstrom syndrome-
- Referred to Diabetes clinic with raised blood glucose levels following a pancreas transplant.

PMH-

- Diagnosed with "type 1 diabetes" since 11 years old
- HbA1c 5-6% very rarely experienced hypoglycaemia or hyperglycaemia
- Registered blind since 1985
- Hypertension

Family history

- One brother died at 5 months of age-fibro-elastosis
- 2 brothers had diabetes
- Mother was hearing impaired

- **Case history cont'd**
- Patient developed progressive renal failure with a creatinine rising to 400 in 1997
- Renal failure believed to be secondary to diabetic nephropathy; no histological diagnosis
- Underwent pre-emptive cadaveric renal transplant in 2005
- Progressive hearing impairment

- Referred for pancreas transplant by transplant team in 2006
- Underwent pancreas transplant 19/9/2006

- Blood glucose levels rising 9-13mmol/l checked at home in 2012
- HbA1c increased from 5.6% in 2007 to 8.3% in 2011 **(fig 1)**
- BMI 38.8 kg/m^2
- Referred to Diabetologist in 2013

Alstrom syndrome

- Alstrom syndrome is rare condition
- Prevalence is 1/1000000
- It has an autosomal recessive inheritance
- A mutation in the ALMS protein is the underlying defect in Alstrom syndrome
- This impairs ciliary function
- Childhood blindness; renal impairment; and insulin resistance are features characteristic of the condition, (**fig 2**)

Figure 2 Features of Alstrom syndrome

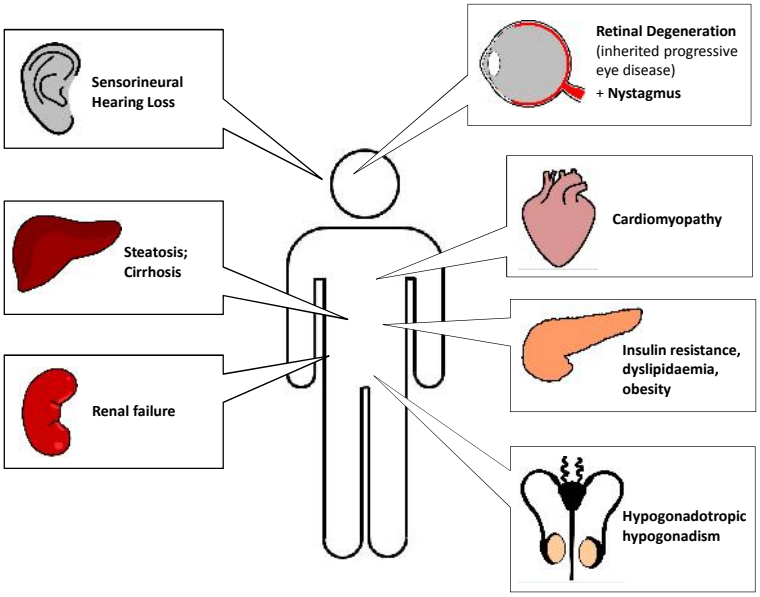

Figure 1 Glycaemic control

	BS	HbA1c
2005		5.0%
2006 – pre- tplt	6.6	
2006 – post (off insulin)	6.7-9.4	
2007	5.7	5.6%
2008	9.2 post food	
2009 OGTT	5.9 to 9.5	
2010		6.4% c-peptide 3969
2011		8.3% c-peptide 3840 (Creat 98)
2012	8.7, post meal 14.3	94 = 11.8% Start Glicalzide 160 bd
2013		70 C-peptide 3065

Outcome

- Patient commenced on metformin therapy
- Inco-oporated more exercise into his lifestyle
- HbA1c decreased from: 70mmol/mol to 42mmol/mol
- His weight remained stable

Alstrom syndrome and diabetes

- Childhood onset diabetes is a feature of Alstrom syndrome
- Insulin resistance and obesity are a key features

- Studies have revealed exercise can lead to significant improvements in glycaemic control (1,2)

Learning points

- Alstrom syndrome is a condition with multi-organ involvement
- Renal, retinal and cardiac disease are a result of the gene defect and not subsequent to diabetes
- Insulin resistance is a predominant component of Alstrom syndrome
- Lifestyle changes and metformin may prevent the early need for insulin treatment in diabetes related to Alstrom
- Pancreatic failure post transplantation may not always necessitate immediate insulin therapy in this condition

References

- *Paisey RB Modification of severe insulin resistant diabetes in response to lifestyle changes in Alstrom syndrome*
- *Mokashi A, Cummings EA; Presentation and course of diabetes in children and adolescents with Alstrom syndrome; Pediatric Diabetes; 2011: 270-275*