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Case Report

Type 2 Diabetes with Hyperglycaemic Hyperosmolar

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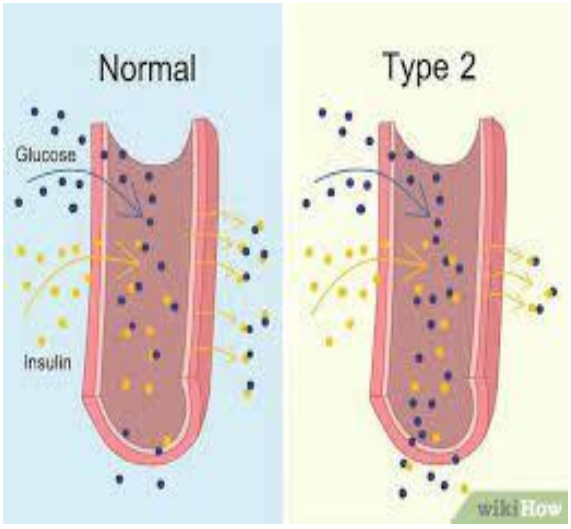
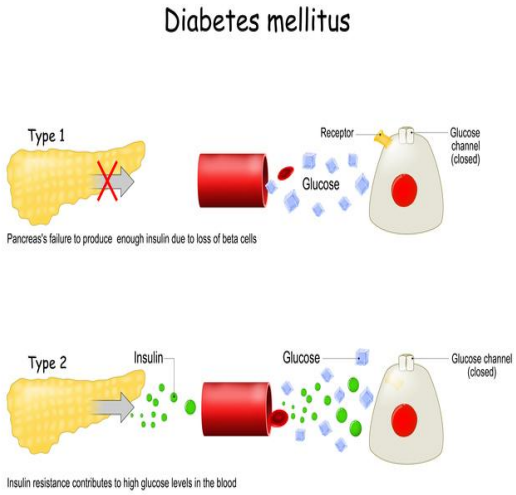
Hyperglycaemic hyperosmolar state (HHS) is a life-threatening condition rarely seen in paediatrics. It is becoming increasingly recognised with the growing incidence of childhood type 2 diabetes mellitus (T2DM). We present a 16-year-old boy with Bardet-Biedl syndrome, with comorbidities including chronic renal impairment requiring renal transplant, isolated growth hormone (GH) deficiency and obesity, who presented on routine follow-up with new onset T2DM and in HHS. Investigations revealed hyperglycaemia (45.7 mmol/L), ketones of 0.1 mmol/L, pH 7.38 and osmolarity 311 mOsmol/kg. After acute management with fluid resuscitation and intravenous insulin, he is now stable on metformin. He has lost weight, renal function is stable and he has stopped GH therapy. We discuss the dilemmas encountered in his long-term management due to his renal transplant and comorbidities, and whether or not, given his significant T2DM risk, this case was preventable or predictable.



Hyperglycaemic hyperosmolar state (HHS) was previously thought to be a rare presentation of type 2 diabetes

mellitus (T2DM) in children, but reported cases are on the rise. This is likely due to the increased prevalence of both T2DM and obesity in the paediatric population. Between 2001 and 2008, 65 cases of paediatric HHS were reported,[1] but the true incidence of this condition is still not known. Bardet-Biedl syndrome (BBS) is a rare heterogeneous autosomal recessive disorder affecting ciliary function. It has an estimated incidence of 1:160 000 in European populations and the abnormal ciliary function has wide reaching effects on multiple organ systems. There is a wide range of clinical variability, but common features include retinitis pigmentosa, renal tract abnormalities (10% develop end-stage renal failure), developmental delay, polydactyly and hypogonadotrophic hypogonadism.



It is also associated with obesity (72–92% of cases), hypercholesterolaemia and many children develop T2DM (6–48%).[2] We describe a rare case of a child with BBS, presenting with HHS, and the issues that were encountered in his management due to his multiple comorbidities and renal replacement therapy. In addition, this patient had multiple risk factors for T2DM—a predisposing syndrome, obesity, family history of T2DM, a high-risk ethnic background and treatment with steroids, tacrolimus and growth hormone (GH). In complex cases such as this, especially when diabetogenic medications are prescribed, significant T2DM risk accumulates and the risk-benefit balance for prescribing such medication must be re-evaluated. Likewise, screening such patients for diabetes is essential to

ensure early treatment and hence prevent unnecessary morbidity and mortality due to damaging a precious transplanted kidney, or the onset of HHS.

Case presentation

A 16-year-old boy with BBS presented with an incidental finding of high blood glucose level on routine screening at a renal outpatient clinic. for his full case history. Of note, he was on immunosuppression therapies, and had been started on GH treatment from the age of 9 years.

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