

Conclusion This data demonstrates that the proposed formula is safe, reliable and effective for the management of hypo- and hyperglycaemia in children with type 1 diabetes mellitus and should be implemented widely to improve safe practise. As it is patient specific and user-friendly, it allows patients/parents to feel in control over their diabetes and contributes to overall patient safety.

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G102(P) CASE REPORT: ATYPICAL PRESENTATION OF HYPERGLYCAEMIC HYPEROSMOLAR STATE IN A YOUNG TYPE 1 DIABETIC

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AEJ Abohweyere, J Wasserman. *Paediatrics (Endocrinology Division), Hospital for Sick Children, University of Toronto, Toronto, Canada*

Aim To highlight the presentation and treatment of Hyperglycaemic Hyperosmolar Syndrome (HHS) presenting atypically in a paediatric diabetic patient.

Case history A previously well 5-year-old boy presented to his local hospital with loss of consciousness. He had a 10 day history of weight loss, polydipsia, polyuria and secondary nocturnal enuresis. He had been drinking large volumes of high sugar content juice. He also had 3 days of abdominal pain and multiple bouts of emesis on the day of presentation. He was found to be lethargic and was immediately transported to hospital. At the emergency department, he was minimally responsive (GCS = 3) and hypopnoeic. He was intubated and transferred to the ICU.

Initial blood glucose was 111 mmol/L. Venous Gas: PH 7.19, pCO₂ 43 mmHg, HCO₃ 15mmol/L, Lactate 3.8mmol/L. Serum Osmolality 381mosm/L. Na 135mmol/L, K 3.8mmol/L, Cl 95mmol/L, Creatinine 446mmol/L, Urea 22.5mmol/L, CPK 7351mmol/L. Urine ketones and serum toxicology screens were negative.

ICU course was notable for severe dehydration requiring copious fluid replacement. Hypotension required multiple inotropes. Hyperthermia requiring ice cooling developed; dantrolene was considered but not used. He had rhabdomyolysis and seizures. MRI showed abnormal FLAIR signal in the sub-cortical white matter of both occipital lobes. His hospital stay was complicated by left leg occlusive thrombus due to a femoral venous catheter. He needed physiotherapy to regain independent mobility.

Islet cell, anti-GAD and Insulin antibodies were positive, confirming Type 1 diabetes. C-peptide levels are pending.

Conclusion We present this case to highlight HHS as an unusual presentation and complication of Type 1 diabetes. The typical patient with HHS is an adult with established diabetes or an obese teenager of African descent. Our patient though black, did not fit this typical profile. HHS occurs less commonly than diabetic ketoacidosis in childhood diabetes although presenting features often overlap. Its management requires large fluid volumes to correct dehydration compared to fluid restriction in DKA. HHS is associated with greater mortality than DKA therefore, paediatricians need a high index of suspicion to diagnose and manage it effectively.

G103(P) AN INTERESTING CASE OF TWO SIBLINGS WITH SDHB MUTATIONS

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RB Mitting, P Kapila. *Paediatrics, North Middlesex University Hospital NHS Trust, London, UK*

Phaeochromocytoma presenting in childhood is rare.¹ It commonly presents with the effects of catecholamine excess, the most consistent of which is hypertension.^{2,3} Patients with phaeochromocytoma

presenting in childhood often have a predisposing genetic mutation.^{1,2} Carriers of succinate dehydrogenase complex subunit B (SDHB) mutations are at risk of developing paraganglioma/phaeochromocytoma, and these tumours have a high rate of malignancy.³ SDHB mutations are also associated with gastro-intestinal stromal tumours.⁴ They are inherited in an autosomal dominant fashion.⁵

We present a case of a 15 year old girl (AB) and her 4 year old sister (MB), who both presented in 2011. AB was diagnosed with an extra-adrenal phaeochromocytoma, (paraganglioma), and MB with intestinal pseudo-obstruction. Both patients have been found to have SDHB mutations.

AB presented with seizures and decreased level of consciousness, requiring intubation and ventilation. It was initially difficult to obtain non-invasive blood pressure (BP) measurement, and once an arterial line was inserted, BP was 255/177. This settled without intervention at the time. Once transferred back to DGH, BP remained 120–70 systolic, so an ultrasound scan was performed, which revealed a mass adjacent to the aortic bifurcation, and urinary catecholamines measured, which revealed raised noradrenaline, and urinary VMA. The mass was removed, and histology confirmed extra-adrenal phaeochromocytoma.

MB presented with bilious vomiting and abdominal distension, so was transferred to a tertiary hospital under the care of the surgical team. She required 3 weeks of TPN and was given a diagnosis of intestinal pseudo-obstruction. Following these episodes, both patients received genetic counselling, and were found to have SDHB mutations, which they inherited from their father.

This case highlights the importance of accurate blood pressure measurement in paediatrics, especially in Accident and Emergency. Hypertension in children is rare,⁵ and therefore when it is discovered, it should be investigated thoroughly as a cause is likely to be found. We are reminded of the need for genetic counselling for children with phaeochromocytoma/paraganglioma and their siblings, as there is a high risk of genetic mutations and, once genetic mutations are identified, there is a risk of synchronous and metachronous cancers.

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G104(P) AUDIT OF ENDOCRINE LATE EFFECTS IN SURVIVORS OF CHILDHOOD BRAIN TUMOURS

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¹E Heffernan, ²A McCarthy, ²R Johnston, ¹S Koppikar, ¹N Abid. ¹Paediatric Endocrinology & Diabetes, Royal Belfast Hospital for Sick Children, Belfast, N Ireland; ²Paediatric Oncology, Royal Belfast Hospital for Sick Children, Belfast, N.Ireland

Brain tumours are the most common solid tumour in children, affecting 400 children in the UK each year. The aggressive treatment required for cure, may have serious consequences, of which endocrine late effects are the most prevalent.

This aim of this audit is to ascertain the frequency and nature of endocrine late effects; in a cohort of 30 survivors, diagnosed over a