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.

REFERENCE

1. Hanas, R. Type 1 Diabetes in Children, adolescents and young adults. Class Publishing, London 2007.

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

doi:10.1136/archdischild-2013-304107.114

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

doi:10.1136/archdischild-2013-304107.115

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.

REFERENCES

1. Waguespack SG, Rich T, Grubbs E, *et al.* A current review of the etiology, diagnosis, and treatment of pediatric pheochromocytoma and paraganglioma. *J Clin Endocrinol Metab* 2010;95:2023–37.
2. Hammond PJ, Murphy D, Carachi R, *et al.* Childhood phaeochromocytoma and paraganglioma: 100% incidence of genetic mutations and 100% survival. *J Pediatr Surg* 2010;45:383–6.
3. Van Hulsteijn LT, Dekkers OM, Hes FJ, *et al.* Risk of malignant paraganglioma in SDHB-mutation and SDHD-mutation carriers: a systematic review and meta-analysis. *J Med Genet* 2012;49:768–76.
4. Pasini B, McWhinney SR, Bei T, *et al.* Clinical and molecular genetics of patients with the Carney-Stratakis syndrome and germline mutations of the genes coding for succinate dehydrogenase subunits SDHB, SDHC, and SDHD. *Eur J Hum Genet* 2008;16:79–88.
5. Armstrong R, Shridar M, Greenhalgh KL, *et al.* Phaeochromocytoma in children. *Archives of Disease in Childhood* 2008;93:899–904.

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

doi:10.1136/archdischild-2013-304107.116

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

fourteen year period. Data was collected on tumour site, histology, treatments used & endocrine complications.

70% of children underwent surgery, 87% received chemotherapy, 40% received cranial radiotherapy, 23% received craniospinal radiotherapy and 16% children received both cranial and craniospinal radiotherapy.

36% of survivors were diagnosed with growth hormone deficiency (all of these children had received radiotherapy). Impaired spinal growth was seen in all children who had received craniospinal radiotherapy, exacerbating short stature. 23% of children were found to have a suboptimal cortisol response; necessitating emergency hydrocortisone treatment. 20% of survivors developed hypothyroidism. Onset of hypothyroidism ranged from 1 to 5 years following treatment. 11% of survivors were diagnosed with precocious puberty; which in 1 case had masked a growth hormone deficiency.

In conclusion, this audit confirms the high prevalence of endocrine late effects in survivors of childhood brain tumours. Growth hormone deficiency was the most common, however there was a high percentage of multiple hormone deficiencies. Data support the establishment of a joint oncology and endocrinology late effects clinic; to ensure early identification and treatment of these serious complications.

G105(P) INTERPRETATION OF CORTISOL LEVELS IN INFANCY IS DEPENDENT ON CLINICAL HISTORY

doi:10.1136/archdischild-2013-304107.117

¹DS Cordiner, ¹HL Miles, ²MS Crane, ¹LE Bath. ¹Endocrinology and Diabetes, Royal Hospital for Sick Children, Edinburgh, UK; ²Biochemistry, Royal Hospital for Sick Children, Edinburgh, UK

Aims To assess cortisol levels in infancy and to determine whether low cortisol levels are indicative of pathology.

Methods A retrospective study of cortisol requests in patients aged up to 64 days for 20 months until August 2012 was undertaken. Data was collected on indications for testing, subsequent results and final outcome. Cortisol was measured using the Abbott Architect Analyser.

Results 47 patients had cortisol measured. The clinical indications were assessment of hypoglycaemia (n = 16), adrenal insufficiency (n = 13), jaundice (n = 11) and hypopituitarism (n = 7).

Cortisol was <100nmol/l in 19 patients: 7 had no further investigation; 3 proceeded directly to standard short synacthen test [SSST]

and passed; 9 had repeat cortisol levels tested: 3 were above 100nmol/l and 5 out of the remaining 6 had further investigation (SSST or Corticotrophin Releasing Test [CRH]) which 3 passed.

For hypoglycaemia the median cortisol was 260nmol/l (range 42–793nmol/l). 1 patient with a random cortisol of 42nmol/l passed a SSST.

For investigation of adrenal insufficiency, the median cortisol was 182nmol/l (range 46–503nmol/l). 2 patients with random cortisols of 82 and 85nmol/l passed a SSST while a third with a level of 93nmol/l had a borderline SSST result.

For jaundice screen, the median cortisol was 132nmol/l (range <40–407nmol/l). One patient with a cortisol of 47nmol/l went on to pass a SSST.

For hypopituitarism, the median cortisol was 40nmol/l (range <40–146nmol/l). Four children in this group with baseline cortisol levels <40nmol/l proceeded to a SSST which 3 passed. A child with suspected septo-optic dysplasia and a baseline cortisol of 87nmol/l failed a CRH test. One infant with baseline of 69nmol/l underwent no further testing.

Results are illustrated in graph 1.

Conclusion Reviewing this cohort of 47 patients, 3 are now known to have cortisol deficiency. In 2, the random cortisol was less than 100nmol/l and they had additional clinical features. A 3rd patient has congenital adrenal hyperplasia, and the cortisol at presentation was 130nmol/l. Interpretation of a cortisol result must be undertaken with the clinical history and additional biochemical results and unless there are features indicating an underlying problem, a random low cortisol is not diagnostic.

Ethics and Law Forum/British Academy of Childhood Disability

G106 NON-THERAPEUTIC CIRCUMCISION IN 'HIGH-RISK' CHILDREN IN A CHILDREN'S HOSPITAL: CONSENT AND SAFETY

doi:10.1136/archdischild-2013-304107.118

¹S Collinson, ²J Brierley. ¹Medical School, St Georges, London, UK; ²Clinical Ethics Service, Great Ormond St Hospital, London, UK

Aims Non-therapeutic circumcision (NTC) is controversial. Religious freedoms and suggested health benefits are balanced against a non-medically indicated surgery in children who cannot consent.

Abstract G105(P) Graph 1