G420(P) A RARE CASE OF SYNDROMIC CONGENITAL HYPERINSULINISM IN A PRETERM BABY

R Oba, U Aboushafa, B Olorunsola. *Paediatrics, North Lincolnshire and Goole NHS FT, Scunthorpe, UK*

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Clinical Case Presentation A case of Syndromic Congenital Hyperinsulinism in a preterm baby, caused by Beckwith-Wiedemann Syndrome(BWS) was seen in our unit. Baby was born at 34 week gestational age by emergency lower segment caesarean section, as a result of fetal distress. Baby was noted to be macrosomic at birth with a weight of 4.37 kg (above the 99.6th centile) and transverse earlobe crease with post-auricular pits. However, there was no other clinical features of Beckwith-Wiedemann Syndrome noted.

Baby was born to a 30 yr old primigravida lady with high BMI of 47. Antenatal Gestational Diabetic screening with oral glucose tolerance test was negative on three separate occasions. There was no history of Polyhydramnios. Anomaly scan was normal. Both parents are non-consanguineous. Dad suffers from Watson Syndrome(a variant of Neurofibromatosis type1) and Mum is Healthy, apart from unexplained High BMI.

Baby was born in good condition but developed respiratory distress soon after birth. Baby was therefore admitted to NICU requiring respiratory support via vapotherm. Baby was noted to have developed hypoglycaemia within the first hours of birth.

Management Hypoglycaemia persisted despite initial 10% dextrose mini-boluses, 20% dextrose and 50% dextrose infusion. Baby required up to 20 mg/kg/min of dextrose infusion including oral feeds. Normoglycaemia was eventually achieved with Glucagon infusion 5 microgram/kg/hour and sustained on oral Chlorothiazide at 7 mg/kg/day, as well as Diazoxide at 5 mg/kg/day.

Insulin level was raised at 98 pmol, as well as C-peptide at 1812 pmol at the time of hypoglycaemia. Baby has a normal Growth Hormone, cortisol level. Ammonia, plasma Amino Acids were also within normal limits.

Baby has clinical and radiological signs of hepatomegaly but normal sized kidneys and pancreas. Echocardiography showed septal and ventricular wall hypertrophy.

In view of macrosomia, transverse earlobe crease with post auricular pits,hyperinsulinism and hypoglycaemia genetic test for BWS was sent.

The genetic testing showed Hypomethylation at KCNQ10T1:TSS-DMR(11P15.5) confirming Beckwith-weiderman Syndrome (BWS)

Conclusion This case exemplified a rare case of Beckwith-weidermann diagnosed in a preterm baby, presenting with Congenital Hyperinsulinism. BWS is associated with preterm delivery in 43% of reported cases. Early suspicion and diagnosis improve outcome.

G421(P) INCIDENCE AND MANAGEMENT OF DIABETIC KETOACIDOSIS IN CHILDREN WITH TYPE 1 DIABETES MELLITUS

¹NAEE Hanna, ²RM Williams, ^{1,2}AEJ Hendriks. ¹Department of Paediatrics, University of Cambridge, Cambridge, UK; ²Department of Paediatric and Adolescent Diabetes and Endocrinology, Cambridge University Hospitals NHS FT, Cambridge, UK

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Aims Analyse incidence of diabetic ketoacidosis (DKA) over time in children with type 1 diabetes mellitus (T1DM).

Audit compliance with 2015 NICE guidelines on DKA management in children with T1DM.

Methods Audit of children <16 years of age with T1DM in a single academic centre. DKA admissions were identified from electronic records. Data was collected from 01/01/2011 until 31/12/2012 and from 01/01/2017 until 31/12/18. Pearson Chi-Square test compared DKA incidence between time periods. Local DKA guidelines were updated in July 2016. Audit data was therefore collected between 01/08/2016 and 31/12/2018. DKA management was evaluated by 6 standards: DKA diagnosis, estimation of dehydration, fluid calculation, intravenous fluid type and insulin dose and timing.

Results A total of 612 children were included, 283 in 2011/12 and 329 in 2017/18. There were 81 and 73 hospital admissions in 2011/12 and 2017/18, respectively. There was a significant decrease in admissions for DKA from 25 (31%) admissions in 2011/12 to 11 (15%) admissions in 2017/18 ($p=0.02$). There was no difference between time periods in whether DKA admissions were at diagnosis of T1DM (15 (60%) in 2011/12; 8 (73%) in 2017/18).

Between 01/08/2016 and 31/12/2018, 371 children were audited. Seventeen admissions for DKA out of 91 total hospital admissions (19%) were included. Newly diagnosed T1DM accounted for 33 (36%) admissions, hypo- and hyperglycaemia for another 17 (19%) and 24 (26%) admissions were due to other reasons. DKA diagnosis was appropriate in 17/17 (100%) admissions. Estimation of dehydration was correct in 16/17 (94%) admissions. Fluid requirement calculation was accurate in 16/17 (94%) admissions. Appropriate intravenous fluids were started in 16/17 (94%) admissions. The correct intravenous insulin dose was given in 16/17 (94%) admissions, and this was started >1 hour after starting intravenous fluids in 16/17 (94%) admissions. Overall, only 12/17 (71%) admissions were fully managed according to DKA guidelines on these key standards.

Conclusions DKA admission incidence has significantly decreased in our hospital in 2017/18 compared to 2011/12. Compliance with DKA guidelines on management was suboptimal and further education of healthcare professionals is required.

G422(P) THE USE OF SOCIAL MEDIA TO IMPROVE DIABETES RELATED HEALTH LITERACY IN PARENTS OF A TYPE 1 DIABETES CLINIC COHORT

¹D Greene, ²K Matyka. ¹Paediatrics, Royal Surrey County Hospital, Guildford, UK; ²Post Graduate, University of Warwick, Warwick, UK

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Background Social media use is becoming more common in people with chronic illness.

Objective To determine if posts to a Facebook page can be used to increase the Health Literacy of parents of children with Type 1 Diabetes.

Methods All parents of the 149 children served by a clinic in a district general hospital were invited to join a purpose created Facebook group. Specific health related information was posted regularly and the change in Health Literacy was measured pre and post-intervention using a validated questionnaire.

Results Half of the 10 questions showed improvement in Health Literacy score post-intervention. Parents were more

engaged with the posts relating to the topics that scored lowest in the pre-intervention assessment. The question that scored the lowest pre-intervention showed the most engagement on Facebook and biggest increase in score.

Conclusion Posting health related information to a Facebook group is not a replacement for face to face education but may serve as an adjunct to good diabetes care.

G423(P) UNCOMMON PRESENTATION OF SEPTO-OPTIC DYSPLASIA

A Jeyaraman, C Brew, V Mathew, S Gupta. *Paediatrics, Hull Royal Infirmary, Kingston upon Hull, UK*

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Introduction Neonatal metabolic acidosis is a common presentation of wide range of causes varying from sepsis to rare metabolic and endocrine problems. Metabolic acidosis with normal anion gap help as to narrow down the cause as GI loss, RTA or adrenal insufficiency. We present a rare case of neonatal metabolic acidosis turned out to be due to adrenal insufficiency and subsequently diagnosed as SOD. Septo-optic dysplasia (SOD) is a rare congenital malformation syndrome characterised by a triad of optic nerve hypoplasia, midbrain abnormality and pituitary dysfunction.

Case Report 19 day-old girl baby presented with lethargy, poor feeding and poor weight gain. On examination, she was hypothermic, hypoglycaemic, hyperkalaemia with early shock and metabolic acidosis. She was treated as presumed sepsis with dextrose, fluid bolus and antibiotics. She had persistent metabolic acidosis with normal anion gap despite on full feeds. Her septic screening and initial metabolic screening were negative. She was noted to have adrenal insufficiency (cortisol <50), which was confirmed by synacthen test. Her cranial ultrasound was normal. She was sent home with steroid supplements. She was noticed as not fixing and following with roving eye movements at the age of 4 months. Found to have bilateral optic nerve dysplasia and later registered as blind. Further imaging of MRI head confirmed SOD with underdevelopment of optic nerve and hypoplasia of pituitary gland. She failed the dynamic growth hormone stimulation test. In view of her slow growth and low IGF, she was started on growth hormone treatment. She also has hypothyroidism on thyroxine supplements. She is making good progress in her growth and development with hormone supplements and MDT management.

Conclusion Clinical followed by hormonal evaluation and imaging can guide us to its appropriate diagnosis and treatment of SOD. Early recognition of SOD is paramount in order to reduce morbidity and to optimise management. Without adequate treatment, panhypopituitarism can have devastating effects on growth and neurodevelopment of children.

G424(P) RE-INTERPRETING THE APPROPRIATE GROWTH HORMONE RESPONSE TO HYPOGLYCAEMIA

¹UA Khan, ¹FY Wong, ¹S Azam, ²O Blake, ^{1,3}AM Murphy, ^{1,3}OM Neylon, ^{1,3}CS O'Gorman. ¹Dept of Paediatrics, University Hospital, Limerick, Ireland; ²Dept of Biochemistry, University Hospital, Limerick, Ireland; ³Dept of Paediatrics, Graduate Entry Medical School, University of Limerick, Limerick, Ireland

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Aims 'Critical samples' consist of assessment of an individual's metabolic and endocrine status during a hypoglycaemia episode to evaluate for a pathological contributor towards aetiology of the episode. Growth hormone (GH) level is part of this work up to assess likelihood of GH deficiency. The aim of this study was to identify if the preset cut off value for growth hormone '7 microgram/Litre' during hypoglycaemia is appropriate and how likely values below this are to be indicative of growth hormone deficiency.

Methods The study population consisted of paediatric patients aged up to 16 years old who had critical samples sent during hypoglycaemia over a 12 month period from 1st Jan 2018 until 30th Dec 2018 inclusive at University Hospital Limerick. Retrospective review from a prospectively collected database of glucose and GH levels as well as other growth parameters.

Results Forty blood samples were collected during the study period during hypoglycaemia. One sample was insufficient for analysis of GH level and of the remaining 39, only three (7.6%) patients exhibited a 'normal' GH response to hypoglycaemia of ≥ 7 mcg/L and thirty-six patients (92%) had suboptimal GH levels. Data was skewed to the right with median GH reading of 3.19 mcg/L. The available data on the other parameters i.e. Insulin-like Growth Factor (IGF)-1 levels, Growth Velocity/Height centiles and Growth hormone stimulation test (GHST) was limited but normal.

Conclusion No patient had documented GH deficiency or loss of height centiles, despite only 7.6% (3/39) having 'adequate' GH response during hypoglycaemia. These data suggest that the optimal level for GH during hypoglycaemia should be re-evaluated in larger prospective studies, to reduce unnecessary evaluation for GH deficiency with attendant anxiety and cost generated therein.

G425(P) AVERAGE HbA1c LEVELS IN DIABETIC CHILDREN WITH INSULIN PUMPS AND MULTIPLE DOSE INSULIN

L Horne, Y Galal, R Wilson. *Paediatrics, Blackpool Teaching Hospital NHS Trust, Blackpool, UK*

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Aims To correlate patient's average HbA1c scores to the modality of insulin therapy utilised to identify whether there is a statistically significant difference in the average HbA1c levels of patients who initially received multiple dose insulin (MDI) and then were commenced on continuous subcutaneous insulin infusions (CSCI). This information would then be used to deliver realistic counselling for patients changing from MDI to CSCI based on real data from our trust.

Methods Within this service evaluation, average HbA1c scores were calculated for all paediatric patients who were initially taking multiple dose insulin therapy and then subsequently commenced subcutaneous insulin infusions. The average HbA1c scores were reviewed at 1 year prior to CSCI and then 1 year and 2 years after the change in modality of insulin delivery. The service evaluation was conducted in 2017 and then again in 2019.

Results In 2017, a mean difference of in HbA1c of 2.03 (95% confidence interval 1.17–2.88) was seen with a significant difference of 0.033. This concludes that there was no significant difference between the average HbA1c of patients