features of sepsis. Diagnosing DKA in this age group is crucial as any delay in initiating right treatment may result in significant morbidity and mortality.

P77 REYE-LIKE SYNDROME AND SEVERE CARNITINE DEFICIENCY IN A BOY WITH AUTISM SPECTRUM DISORDER AND RESTRICTED DIET

Lydia Healy*, Eimear Forbes, Jane Rice, Jane Leonard, Ellen Crushell. Temple Street Children’s University Hospital, Dublin, Ireland

10.1136/archdischild-2019-epa.432

Case Description A 9-year old boy presented to the Emergency Department with an acute history of vomiting, ataxia and reduced consciousness in the setting of a respiratory illness. In the two months prior he had intermittent episodes of vomiting, without headaches or visual disturbance.

On examination, he was encephalopathic (GCS 9/15), with marked hepatomegaly. Height and weight were on the 0.4th-2nd percentile and 9th percentiles respectively.

He had a background of high-functioning autism spectrum disorder (ASD) and a long history (since aged 18 months) of rigid eating behaviour, with a diet restricted to corn-based crisps (‘Rancheros’), rich-tea biscuits (2-3/day), French-fries and Coca-Cola (10% glucose, drinking 1 L/day). He had recently commenced a multivitamin preparation.

Blood-work showed anaemia, elevated hepatic transaminases and creatinine kinase (60000U/l), low albumin, hypoglycaemia (2.7 mmol/l) and deranged electrolytes. CT brain showed moderate, communicating hydrocephalus without pressure effects. An extra-ventricular drain was inserted but later removed as this was deemed to be a coincidental finding of arrested congenital hydrocephalus.

Management and Investigation He required intubation and ventilation for respiratory compromise (RSV infection). Given the Reye-like scenario and abnormal biochemistry, a metabolic disorder was suspected. Acylcarnitine profile showed undetectable free carnitine and amino acid levels were low, as were vitamin levels. He received carnitine supplementation and very cautious nasogastric feeding; biochemistry and neurological status improved. Extensive investigations including carnitine uptake studies did not reveal an inherited metabolic disorder.

Detailed nutritional analysis of the patient’s diet revealed very low daily protein (0.37 g/kg/day) and fat (0.77 g/kg/day) but with normal total daily caloric intake of 1200kCal, indicative of protein-energy-malnutrition (Kwashiorkor). He received intensive inpatient dietetic and psychological input and by discharge from hospital he was consuming an expanded range of foods. Carnitine is synthesised from essential amino acids lysine and methionine, but primarily it is obtained directly from protein in the diet. Cereal-based diets such as above are particularly low in the amino acids methionine and lysine.

Diagnosis and Follow-up On follow-up 2 years later, carnitine levels remain normal without supplementation. Protein intake is normal and diet is varied. Vertical growth has improved significantly with height now on the 91st centile and hepatomegaly has resolved. The final diagnosis was dietary protein-energy-malnutrition with profound secondary carnitine deficiency resulting in Reye-like syndrome.

This case reminds us that severe malnutrition exists in the developed world, and emphasises the need for detailed nutritional assessment of children on self-restricted diets, especially those with ASD.