cause of gastric outlet obstruction in the 2- to 12-week-old age group. Pyloric stenosis leads to progressive and projectile vomiting.

It affects 2–3 per 1000 infants in Northern Europe but less common elsewhere in the world. Males (especially first borns) are affected approximately 4–6 times as often as females. The offspring of a mother, and, to a lesser extent the father who had pyloric stenosis are at higher risk for pyloric stenosis (Nelson Textbook of Paediatrics, 20th ed, Elsevier Saunders, 2016).

Impetus

- To stimulate discussion and further research on extreme electrolyte
- imbalances babies can tolerate.
- To demonstrate importance of slow correction of electrolyte imbalance over
- 72 hours.
- To consider Pyloric Stenosis as one of the differential diagnosis for
- constipation in early infancy.

Case Report We present a 9-week baby boy presenting with constipation and worsening vomiting for a week. He had very severe metabolic alkalosis (Ph-7.67), hypokalaemia (2.5 mmol/L) and hypochloraeaemia (60 mmol/L). He was amazingly still breathing spontaneously with no apnoea. His electrolyte imbalance was slowly reversed over 72 hrs and later he was transferred to a tertiary surgical care unit for corrective pyloromyotomy. He recovery was uneventful post surgery. We would like to share baby's severe electrolyte imbalances and stimulate research on how much extremes of metabolic alkalosis infants can tolerate.

Abdominal ultrasound Pylorus appears moderately thickened measuring to 2.1 cm in length (normally up to 1.5–1.7 cm). Pyloric transverse diameter measures to 1.7 cm (normally up to 1.3 cm) and pyloric muscle thickness was 8.9 mm

Learning Points

- Slow correction of electrolyte imbalance is critical in treatment of pyloric stenosis.
- Observe very closely for apnoes in babies with severe metabolic alkalosis
- Always think of Pyloric Stenosis in infants presenting with constipation. Pyloromyotomy is performed only after baby's metabolic alkalosis is normalized.

REFERENCES

1. Dalton BG, Gonzalez KW, Boda SR et al, Optimizing fluid resuscitation in hypertrophic pyloric stenosis. *Journal of Pediatric surgery* 2016 Aug;51(8):1279–82. doi: 10.1016/j.jpedsurg.2016.01.013. Epub 2016 Feb 3.

P105 FLOPPY INFANT: FORGOTTEN OR NEGLECTED SYMPTOM OF CELIAC DISEASE

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Introduction Celiac disease (CD) or gluten-sensitive enteropathy is an autoimmune disease of the small intestine precipitated by the ingestion of gluten in genetically susceptible individuals. The prevalence is 1 of every 120 to 300 persons. The association between neurological symptoms and celiac

disease was first described by Cooke and Smith in 1966. Initiation of a gluten-free diet reverses the mucosal lesion and corrects malabsorption with symptomatic improvement.

Case report A 16-months old girl presented to the University children hospital with acute hypotonia and suspected Guillain-Barre syndrome. Two months before admission, she developed vomiting and watery diarrhea, which lasted for few days. Two weeks afterwards, acute symmetrical weakness of upper and lower limbs was occurred, so the patient was admitted to regional hospital. Her past medical history was irrelevant. She had normal growth and normal milestones development. She was able to walk before her first birthday. There was no history of honey and canned food use. She had negative family history of a similar condition. Her physical examination showed a floppy, irritable and afebrile child with body weight of 9900 g (25–50th percentile). Both upper and lower limbs were very weak with no reflexes. After her laboratory tests, abdominal ultrasound and brain CT, the diagnosis of GB was established and the treatment with IVIG was initiated. Her neurological symptoms improved slightly, but after five doses her neurological condition deteriorated with appearance of pretibial and dorsal pedal edema, and then she was sent to our hospital. On admission, she was 16-month old girl, body stature 82 cm (90th percentile), body weight 8450 g (10th percentile), BMI 13.7 kg/m². Subcutaneous fatty tissue was significantly reduced, she also had severe generalised edema. Her abdomen was distended without signs of hepatosplenomegaly. Neurological examination revealed severe general hypotonia of limbs with reduced reflexes, without axial weakness. Routine laboratory revealed normal CBC, hypoproteinemia, hypoalbuminemia with normal kidney and liver function. Inflammatory parameters, the biochemistry and cytology of cerebrospinal fluid, and MR of lumbosacral region were normal. The electro-neuromyography was not performed. Clinical aspect and laboratory results indicated celiac disease. Very high anti-endomysial and anti-transglutaminase antibodies and positive genetic test (HLA DQ2) confirmed clinical suspicion. No duodenal biopsy was performed. During investigation, the therapy with corticosteroids, albumin and furosemide were started. After celiac disease was diagnosed, gluten free diet (GFD), vitamin B complex and folic acid were started with gradually decreasing of corticosteroid therapy. Neurological improvement appeared after five days. After 3 weeks, she improved in her nutritional and psychological aspects.

Conclusion We should think about CD, since many patients have atypical disease.

P106

DIAGNOSIS OF ACUTE MYOSITIS VS DUCHENNE MUSCULAR DYSTROPHY BY TIMING OF BLOOD'S ENZYME PROFILE

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Introduction Elevated Alanine Aminotransferase (ALT) and Aspartate Aminotransferase (AST) enzymes would sway a doctor toward thinking there's a liver problem; a high CK with high ALT and AST levels suggests that something's going on in the muscle. A correct diagnosis may prevent extensive investigations or procedures. In boys one of the worst case scenarios is Duchenne Muscular Dystrophy (DMD) a severe

type of muscular dystrophy, with an incidence of 1 in 3500 male births. The initial feature in most boys with DMD is a gait disturbance, with an onset often before age 3 years. Although there is not curable, it is treatable with an importance of an early diagnosis

Case presentation A 26 months old boy presented to our Paediatric Assessment Unit with a history of fever up to 38°C, vomiting, and poor oral intake. His urine was red in colour, but dipstick and microscopy negative for blood. He had no significant past medical history. Vaccinations were up to date. No history of foreign travel. His development was normal.

On examination he had signs of an upper respiratory tract infection. His inflammatory markers were elevated and he was commenced on IV Augmentin. He received IV fluids antipyretics and anti-emetics.

The following day he refused to weight bear. On repeat blood tests, his ALT value was elevated and remained high on repeat testing. Ancillary test for assessment of liver disease were requested. A Creatine Kinase (CK) was requested and showed a value of 20248 IU/L.

He started to weight bear after 4 days and this continued to improve. CK values decreased quickly in the following two weeks. His neurological exam was normal at clinic follow up 2 months post discharge and his LFTs have now returned to normal.

Discussions In the absence of liver pathology, raised transaminase may be an early sign of occult myopathy and such patients should have CK levels checked to look for evidence of muscular involvement as happens in polymyositis and muscular dystrophy as in Duchenne's (DMD).

In DMD, CK levels typically rise above 10,000 IU/L, but they do not return to normal in a few weeks time. It helps to make a clinical differential diagnosis without performing complex genetic tests for Duchenne's. Laboratory investigations together with the coryzal symptoms from the onset were consistent with our diagnosis of acute viral myositis.

P107

THUMB AUTO-AMPUTATION FOLLOWING TOURNIQUET SYNDROME

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Case Report A 12 month old boy presented to the Paediatric Emergency Department (PED) with ischaemic necrosis of the distal phalanx of the left thumb as a result of Tourniquet Syndrome.

Six days prior to presentation an adhesive bandage had been applied to the digit to discourage thumb sucking. His mother described crying and irritability, and when the dressing was removed forty-eight hours after application, the infant's mother noted the digit to be shrivelled and mottled. The following day the area began to blister and he was brought to his General Practitioner for review, where hospital attendance was advised.

Examination of the left thumb revealed dry necrosis of the nailbed and distal phalanx extending 1 cm proximal to the nailfold, with an obvious tourniquet mark present at the

interphalangeal joint. The infant was admitted under the joint care of the Plastic Surgery and General Paediatric Teams. The injury was initially managed conservatively. The distal phalanx subsequently auto-amputated and formal surgical debridement and terminalisation at the level of the interphalangeal joint was undertaken. At follow-up twelve months later the injury had healed well with a satisfactory functional outcome.

The severity of the injury coupled with the delay in presentation raised suspicion of neglect or non-accidental injury, and the case was referred to the Medical Social Work Department. The history given by both parents was consistent with the presenting injury. No other concerning injuries or previous hospital presentations were noted and appropriate child-parent interaction was noted throughout. A child protection case conference found that the injury was unintentional and that the infant was not at ongoing risk of significant harm through neglect. A family support plan was put in place on discharge to the community.

Discussion Tourniquet Syndrome is a surgical emergency as circumferential constriction impedes lymphatic drainage with resultant venous outflow obstruction and oedema. Raised interstitial pressures then restrict arterial supply, causing ischaemia and associated pain. If it is not possible to remove the tourniquet in the PED, general anaesthetic and removal in theatre may be required. In cases of significant vascular compromise necrosis and auto amputation may occur as illustrated.

Due to its insidious nature, a high index of suspicion for this condition is required for timely detection. Added vigilance is required in the case of preverbal children and those with speech or learning difficulties who may not be able to express discomfort easily.

P108

A RARE CASE REPORT WITH ALOPECIA, ONYCODYSTROPHY AND HYPOGAMMAGLOBULINAEMIA- COMMON VARIABLE IMMUNODEFICIENCY (CVID)

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Introduction Common Variable Immunodeficiency (CVID) is a condition characterised by primary hypogammaglobulinaemia and in consequence increased susceptibility to infections and less frequently ectodermal abnormalities. Additional endocrinopathies have been identified with hypoadrenalism, growth hormone deficiencies etc in the previously reported patients.

Case presentation A 2 and half year old boy presented to paediatric outpatients of Wexford General Hospital with a history of alopecia, onychodystrophy and recurrent infections. Alopecia gradually got worse within one year. He was treated for chest and ear infections with several courses of antibiotics.

On examination we found that his growth was satisfactory. Apart from alopecia and nail dystrophy there were no other abnormalities noted.

Investigations revealed normal full blood count and electrolytes. Low immunoglobulins in all domains were the striking abnormality. Lymphocyte subset and thyroid screening were normal. He was referred to the immunology team.