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Objective Food protein-induced enterocolitis syndrome (FPIES) is a non-IgE-mediated gastrointestinal food hypersensitivity manifesting as profuse repetitive vomiting, often with diarrhea. FPIES consists of two types, acute and chronic. In the acute type, ingestion of triggering foods induces vomiting, diarrhea, lethargy, and hypotension in up to 4 hours. Episodes can last 24 hours, and both vomiting and systemic symptoms benefit from ondansetron. In the chronic type, there is vomiting and diarrhea, but the time between ingestion and symptoms is longer and the presentation milder. Mild but persistent vomiting and diarrhea cause malnutrition and hypoalbuminemia. Diagnosis is based on clinical symptoms and confirmed via positive food challenge. Symptoms resolve after elimination of triggering foods and recur with re-exposure. This report aims to review the presentation of FPIES using a case-driven approach.

CASE Female 1-year-old admitted with repetitive, projectile emesis, and diarrhea. Symptoms subside after 24 hours with iv fluid resuscitation, and vomiting began with complementary food. Initially diagnosed with a cow milk allergy, but after ingesting bread, she had another attack. This attack led to a diagnosis of Celiac disease, and she was prescribed a gluten-free diet.

After ingesting gluten free pasta, she had another attack with loss of consciousness. After fluid resuscitation, laboratory tests including anti-endomysium IgA, cow-milk IgE, wheat IgE, egg yolk IgE, Immunoglobulins, and ANCA were normal. Esophagogastroduodenoscopy and colonoscopy were normal, with a pathology report of mucosal fragments including lenoid follicles. An oral food challenge with possible triggering foods was then performed. The patient was admitted, and a peripheral intravenous line was placed. After 30 minutes of feeding with rice, vomiting with diarrhea presented with tachycardia and hypotension. Furthermore, her neutrophil count was increased. IV saline fluid bolus, IV ondansetron, and IV methylprednisolone were administered. Emesis stopped, and abdominal pain subsided within 30 minutes. After 24 hours, all symptoms subsided. With a diagnosis of acute FPIES, she was discharged with strict dietary restrictions as well as future food challenge plans for other possible triggering foods.

Conclusion FPIES is often misdiagnosed as an acute viral gastrointestinal illness, sepsis, or anaphylaxis, and a diagnosis of FPIES can often be delayed for months. As in this case, delay in diagnosis lead to unnecessary diagnostic evaluations and erroneous treatments. After diagnosis is approved with an oral food challenge, prognosis is good. However, avoiding offending foods and appropriate episode management requires hard work from both the families and the health personnel.

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The diagnosis of acute pancreatitis (AP) in children has been improving worldwide probably due to better awareness of physicians evaluating children with gastrointestinal symptoms. The common causes include anatomic abnormalities, drug toxicity, trauma, and systemic diseases, however, a significant proportion of cases are still being labeled as idiopathic. The aim of our study was to highlight the etiopathogenesis of acute pancreatitis in children diagnosed in a University Hospital Centre during a 7 year period.

The charts of pediatric patients, from 2013.-2020. with at least one documented episode of acute pancreatitis requiring hospitalization, were reviewed retrospectively. Data included a clinical picture, laboratory findings (serum and urine amylase and lipase), abdominal ultrasound and MRCP findings as well as results of genetic analysis in a group of patients with previously unresolved etiology.

We identified 33 patients with AP, 18 boys and 15 girls. The mean age at diagnosis was 12, 42 (range 3-18, median 14). The most frequent was idiopathic AP (n=19), followed by anatomical abnormalities (n=7).

Drug-induced AP was seen in four patients due to valproate, enalapril and in two of azathioprine toxicity. Post-traumatic pancreatitis was diagnosed in one of our patients. Two of our youngest patients showed positive results in gene testing with both of them being heterozygous for serine protease inhibitor Kazal type 1 (SPINK1) mutation. Both of them developed acute recurrent pancreatitis later.

In this retrospective analysis, the etiology of acute pancreatitis has similar distribution to previously reported causes in the paediatric literature. Genetic testing is a valuable tool in identifying the nature of previously designated idiopathic pancreatitis, especially in those with a more severe and recurrent presentation.

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Alagille syndrome (AGS) is a rare genetic disorder inherited in autosomal dominant pattern that affects the liver, heart, and other organ systems. Major feature of AGS is liver damage caused by paucity of the bile ducts which leads to chronic cholestasis marked by jaundice, pruritus, xanthomas and sometimes cirrhosis. Common associated anomalies include cardiac anomalies (peripheral pulmonary stenosis), butterfly vertebrae, posterior embryotoxon, dysmorphic facies and renal dysplasia.

We present a long-term follow up of two siblings aged 13 and 7 year diagnosed with AGS during infancy.

The older sibling, female, presented at 7 weeks with jaundice and history of acholic stools and dark urine. Laboratory tests confirmed cholestasis with elevated liver enzymes, bile acids, alpha-feto-protein and 5'-nucleotidase.

Hepatic scintigraphy confirmed severe intrahepatic cholestasis.

Ursodeoxicollic acid (UDCA) and phenobarbitone were introduced. Due to dysmorphic facial features (prominent forehead, broad nasal bridge, triangular facies and deep set eyes) AGS was suspected and confirmed by further evaluation

(peripheral pulmonary stenosis and posterior embryotoxon). At 3 years old liver biopsy revealed paucity of bile ducts, changes typical to AGS, which definitely confirmed the diagnosis.

When the younger, male, sibling presented with typical phenotype and intrahepatic cholestasis in the neonatal period, AGS was immediately suspected. Of associated anomalies, peripheral pulmonary stenosis, posterior embryotoxon and butterfly vertebrae were all present. Stomatology exam revealed hypodontia in both siblings. Brain MRI conducted at age 3 revealed signs of hepatic encephalopathy with no associated vascular anomalies.

Both siblings had severe cholestatic liver disease in pre-school period which had stabilized by school age with conservative treatment for pruritus and malabsorption. Both had severe hypercholesterolemia, in the younger sibling it was extreme with LDL cholesterol levels reaching up to 30,1 mmol/L. The levels of bile acids were also extremely high, reaching a maximum of 634,4 $\mu\text{mol/L}$ in the younger sibling. Therapy included chronic use of UDCA, liposoluble vitamins and MCT with a hypercaloric diet.

The natural evolution of the disease is favorable if liver transplantation is avoided until reaching school age. In both our patients, despite severe liver disease, psychosomatic development and quality of life remained intact.

249

UNUSUAL INCIDENCE OF CRIGLER-NAJJAR SYNDROME TYPE 1 IN CROATIA

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To present patients with Crigler-Najjar syndrome type 1 (CN1). It is a rare autosomal recessive disorder with an incidence of 1: 1 000 000 live births, characterised by severe unconjugated hyperbilirubinemia which arises as a consequence of the absence of hepatic bilirubin uridine diphosphate glucuronosyl transferase (UGT1A1) activity.

In the last 30 years we treated seven children with this syndrome at the Department of Pediatrics, University Hospital Center Zagreb.

They were from five families- two pairs of siblings (brother and sister) and three unrelated patients (two boys and a girl). Genetic testing of UGT1A1 gene was performed in six patients (two pairs of siblings and two unrelated boys). Unfortunately, one patient's result was lost.

Three patients had frameshift mutations in exon 1: Patient 1 (c.722_723delAG p.Glu241Glyfs*16), Patients 2 and 3 were siblings and had identical mutation (c.717_718delAG p.Q239fsX256). Two patients (4 and 5, also siblings) had identical nonsense mutation in exon 3 (c.1021C>T p.Arg341*).

Genetic testing, as it was not widely available at the time, was not performed in one girl whose diagnosis was made by the chromatographic analysis of bilirubin glucuronides in the bile.

Four patients underwent a liver transplant from living related donors. In two auxiliary procedure was performed (siblings at the age of 7 and 9 years) and in two segmental liver transplant (at the age of 6 and 10 years). Prior to

surgery, there was also an unsuccessful attempt of hepatocyte transplantation in one patient.

Three liver transplant procedures were successful, and one patient died in the early post-operative course due to primary graft dysfunction.

Three patients who have not yet undergone liver transplant (a 3-year-old boy and two siblings 1.5-year-old girl and her 6-month-old brother) are currently treated with phototherapy. At least 10-14 hours long treatment is necessary to keep their bilirubin at an acceptable level (around 250 $\mu\text{mol/L}$). Their psychomotor development is appropriate and they have no neurologic impairment.

Considering the number of births per year in Croatia we noticed a remarkably high incidence of CN1, more than five times as expected (5,4: 1 000 000). We don't have explanation for this finding, at least not by mutations observed. Nevertheless, three of our patients are offspring of two families originating in small Croatian enclave in Kosovo where they were isolated for several centuries. Perhaps there are epigenetic factors we are unaware of that may play a role and contribute to this unexpectedly high incidence.

250

TO BOLUS OR NOT TO BOLUS: A RECURRING SITUATION ENCOUNTERED IN PAEDIATRIC ED

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To analyse the adherence to guidelines for assessment and management of dehydration in children presenting with acute gastroenteritis/gastritis in a Pediatric ED.

40 charts were reviewed retrospectively over three months for children who had symptoms of vomiting and/or diarrhea. HSE clinical guidelines for assessment and management of Gastroenteritis were used as a standard².

Specific emphasis was given to the appropriate prescription of normal saline and dextrose boluses³.

Charts were reviewed again after giving appropriate education sessions.

100% documentation was noticed for vital signs and capillary refill time.

While making an assessment of dehydration status, degree of dehydration was documented in 17.5% of cases, this improved to 86% with massive correction in individual components in degree of dehydration.

Children who required IV fluid, boluses improved from 21.5% to 95% for appropriately prescribed saline and dextrose boluses.

Acute Gastroenteritis is a common childhood illness and its severity is linked to etiology, though rotavirus is the most severe infectious agent. Dehydration is a frequent association and its severity must be monitored by established score system¹.

As proven by this audit, assessment and management of Gastroenteritis, although common, can still prove tricky in the acute emergency setting.

However, through education sessions optimal results were achieved.