post-prandial vomiting. Three patients received conservative treatment including proton pumping inhibitor successfully. Only one patient received operation with Billroth I +vagotomy due to pyloric complete obstruction. No recurrent gastric outlet obstruction was noted in all the patients

Conclusions: NSAIDs ingestion was an important cause of peptic ulcer in children. Conservative treatment including PPI should be first line therapy for the gastric outlet obstruction due to peptic ulcer in children. Surgical treatment should be reserved for the patients with failed medical treatment.

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COMORBIDITY OF AUTOIMMUNE DISEASES IN CHILDREN WITH DIAGNOSED COEALIC DISEASE

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Introduction: Coeliac disease is an inflammatory enteropathy with an immunological background, characterised by a permanent gluten intolerance in genetically predisposed individuals.

Aim of the study: The aim of the study was to analyse the comorbidity of autoimmune diseases in children with diagnosed coeliac disease, hospitalized in the Department of Paediatrics , in the period 2003- 2008.

Patients and methods: The analysis included 88 childred (55% girls and 45% boys), aged from 11 months to 18 years, in whom coealic disease was diagnosed on the basis of the clinical manifestation, the presence of p/IgAEmA antibodies and/or tTG antibodies, and abnormalities in the histopathological examination of the small intestine. The course of disease and comorbidity of other autoimmune diseases were evaluated in the examined children. The patients were analysed in relation to gender, age and time of disease manifestation.

Results: In 9/88 children (10.2%) with diagnosed coeliac disease, the coexistence of type1 diabetes was observed; in 4/88 children with coeliac disease inflammatory bowel disease was found; in 4/88 children - lymphocytic colitis was also present.

In children with diagnosed coeliac disease, thyroid gland diseases (2/88), psoriasis and vitiligo were less common.

In 11/88 patients (12.5%) with coeliac disease food allergy was diagnised, most often to cow´s milk protein and egg.

Autoimmune diseases were significantly more frequent in the subgroup of older children (>7 years of age).

Summary: In this study we would like to emphasize the frequent coexistence of autoimmune diseases in patients with diagnosed coeliac disease (24%), especially in older children.

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RISK FACTORS OF GASTROINTESTINAL DYSFUNCTION IN PATIENTS WITH SPINAL MUSCULAR ATROPHY

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Spinal muscular atrophy (SMA) is a neurodegenerative disease affecting motor functions, which may compromise feeding and swallowing ability, gastrointestinal motility, and nutritional status. We conduct a questionnaire survey for addressing the prevalence of and predictive factors for above problems in patients with SMA.

112 genetically confirmed SMA patients participated. The questionnaire recorded demographic data, neuromuscular status such as respiratory support and current ambulatory functions, feeding and swallowing difficulties, and gastrointestinal motility problems. Body weight was measured and insufficient weight gain was defined as Z score of weight-for-age less than -2.

Four patients with type I SMA were excluded, thus 108 type II & III SMA patients met the inclusion criteria (median age: 13.0 ±10.68 years, type II 60 cases and type III 48 cases). The prevalence rate of swallowing difficulties in type II SMA was higher than type III patients (41.7% vs. 6.3% in pre-oral phase; 38.3% vs. 10.4% in oral phase; 46.7% vs. 10.4% in pharyngeal phase; and 25% vs. 6.3% in esophageal phase, respectively). Multivariate analysis showed SMA type, neuromuscular status are independent predictive factors. The prevalence

of GERD diagnosis was 11.6% in type II, and 6.3% in type III. Pulmonary complication such as recurrent pneumonia occurred in a half of patients with both GERD and insufficient weight gain.

Swallowing difficulties are very common in type II SMA. GERD and esophageal complaints also occurs in a significant portion of type II and III SMA. Early recognition and intervention of such gastrointestinal dysfunction is mandatory.

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CRIGLER-NAJJAR SYNDROME TYPE II CAUSED BY HOMOZYGOUS DOUBLE MUTATION [T1456G, G211A] OF *UGT1A1* GENE IN A TAIWANESE SIBLING

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Objective: This study aimed to analyze the mutations of UDP-glucuronosyltransferase 1A1 (*UGT1A1*) gene in a Taiwanese family.

Patients and methods: Two brothers aged 24 and 2 months, respectively were born to a Chinese mother who married to a Taiwanese man. They had profound unconjugated hyperbilirubinemia in the newborn period (30.7 and 25.2 mg/dL, respectively). The liver function, hemoglobin and glucose-6-phosphate dehydrogenase levels were within normal limit. They both received intensive phototherapy and the serum bilirubin level gradually decreased to 6-10 mg/dL. The blood samples of the brothers and their parents were sent for DNA analysis including the promoter area of the UGT1A1 gene, exons 1-4, the coding region of exon 5, and their flanking intronic regions. The polymerase chain reaction-restriction fragment length polymorphism (PCR-RFLP) and direct DNA sequencing were used.

Results: The PCR results for the *UGT1A1* gene showed homozygous G > A substitution at nucleotide 211 in exon 1 and homozygous T > G substitution at nucleotide 1456 in exon 5 ($Gly^{71}Arg$ plus $Tyr^{486}Asp$) in both patients. No other variation was detected in the *UGT1A1* gene. Analyzing the *UGT1A1* gene of the parents showed a homozygous 211G > A plus a heterozygous 1456 T > G variations in their father and a heterozygous 211G > A plus a heterozygous 1456 T > G variations in their mother. All of the tests were confirmed by the direct gene sequence.

Conclusion: Double homozygous mutation [T1456G, G211A] of *UGT1A1* gene is rare in CN-2 and is a novel mutation in Taiwanese children.

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PROLONGED JAUNDICE SCREEN AUDIT-RATIONAL APPROACH TO INVESTIGATIONS

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Background: Extensive laboratory investigations are routinely performed to exclude rare but potentially catastrophic causes of prolonged jaundice.

Aims: This study aims to review the investigations and outcomes of Prolonged Jaundice Screening (PJS) and considers whether it is possible to safely reduce the number of unnecessary blood tests.

Methods: Retrospective analysis of all babies for PJS, between June 2008 and March 2010. Data from the Prolonged Jaundice Proforma was collated in Excel and was analysed using simple statistical methods and percentage calculations.

Results: 100 babies reviewed, age of presentation were between 2 to >8weeks. 97% were fully or partially breast fed and 97.3% showed adequate growth. 90.4% term and 9.6% pre-term. Stool colour and clinical examination was normal in all cases.

8% had conjugated bilirubin above 20% of the total, 19% were noted to have >200mmol/L unconjugated bilirubin levels. All were subsequently found to have normal second line investigations. 55% of babies had abnormal blood results requiring repeat testing. All results normalised on subsequent testing (up to five repeats).

Urine was positive in 25 cases with possible contamination in 20 cases and E.coli in five cases. 2 babies had positive reducing substances, however, Galactosaemia was ruled out in both cases. No cases of cholestasis were identified.

Conclusions: Good clinical history and examination along with split bilirubin is sufficient as first line investigation for PJ screen. Second line investigations are indicated only if there are any concerns on initial screen.