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HEREDITARY TYROSINEMIA TYPE I: LIVER TRANSPLANTATION (LT) WITH LIVING DONOR FOR HEPATOCARCINOMA AFTER 7 YEARS OF NTBC

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Background and aim: Hereditary tyrosinemia type I (HTI) is a severe metabolic disease due to deficiency of fumaryl aceto acetate hydrolase (FAAH). The main clinical consequence of this defect includes hepatic involvement, high risk of liver cancer, renal tubular dysfunction and neurological crisis. Until 1992 transplantation is the only treatment of HTI. Since 1992, the NTBC (2 - (2 - nitro-4 trifluoromethyl benzoyl) - 1,3 - cyclohexane dione) offers a new promising tool for the treatment of HTI. We report the results of this treatment in one Tunisian patients who developed hepatocarcinoma after 7.5 years of treatment.

Observation: The disease was revealed by subacute form at age 9 months by hepatic dysfunction. The diagnosis was confirmed by urinary abnormal excretion of succinyl acetone (SA) and Δ amino levulinate. The treatment by NTBC was started at age of 10 months. The hepatic function was normalized at one month. After 7 years, he developed an hepatocarcinoma with resection of alpha foetoproteine. In the first time the child have a resection of tumor than a liver transplantation with living donor and now he is at 4 years and 8 months of the LT. The rate of the succinyl acetone was raised at 21 μ mol/l.

Conclusion: The NTBC is a promising alternative treatment of HTI to the transplantation and the results reported here highlight the efficiency of the NTBC in early outcome however it doesn't prevent at the long term from the liver cancer.

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ETIOLOGY OF UPPER GASTROINTESTINAL BLEEDING IN CHILDREN

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Background: Upper gastrointestinal bleeding (UGIB) is a relatively common and occasionally

fatal emergency in children. Upper digestive endoscopy is the method of choice when establish the etiology.

Objective: to determine the etiology of UGIB, indications for upper digestive endoscopy, the safety of the procedure in children.

Material and method: 129 children admitted in Pediatric Department, Clinical Hospital Constanta, Romania, with UGIB, undergone a number of 178 upper digestive endoscopies in a prospective study during 5 years. Endoscopy was performed under local/iv general anesthesia/iv sedation, depending on age and agreement of children and parents, after the procedure was explained.

Results: 66 boys and 63 girls aged between 9 months and 16 years (mean age 9.7 years) undergone upper digestive endoscopy for hematemesis (95 cases), melena (88 cases) and inexplicable anemia (4 cases). Endoscopy detected the site of bleeding in 118 cases (91.47%). Association of hematemesis and melena was best correlated with endoscopical findings (96.55%). The etiology revealed most frequently erosive gastritis (59 cases), esophageal varices (24 cases), duodenal ulcers (10 cases), erosive esophagitis (10 cases), erosive duodenitis (8 cases). We found rare etiology in children, such as gastric ulcer (2 cases), duodenal angiodysplasia (2 cases), gastric polyp (1 case), Mallory Weiss syndrome (1 case), gastric adenocarcinoma (1 case). We used local anesthesia (46 cases), iv anesthesia (62 cases) and iv sedation (70 cases). No complications were encountered.

Conclusion: Upper digestive endoscopy was a safe procedure in children, visualizing recent stigmata of hemorrhage in 91,47% of cases of UGIB.

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ANTIBODIES OF HEPATITIS A IN CHILDREN IN SYRIA

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Aim of the study: Studying serological inversion of hepatitis A virus in children and knowing the percentage of asymptomatic hepatitis A.

Material and methods: This study was done in children's hospital in years 2007-2008. The number of children included in the study was 168 (88 male