COLECTOMY RATE IN PAEDIATRIC-ONSET ULCERATIVE COLITIS – ONE CENTRE EXPERIENCE

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Paediatric-onset ulcerative colitis (UC) is often more extensive than in adults, and as disease severity is associated with disease extent, children are more prone to refractory severe episodes, sometimes requiring colectomy.

Previous population-based studies in patients with UC revealed variable colectomy rates. However, a decrease in colectomy rates was observed during the last two decades. The aim of our study was to assess the colectomy rate in paediatric patients with UC and to compare the clinical features of children who had to those who did not have colectomy.

In our hospital, data on children diagnosed with inflammatory bowel disease have been prospectively collected since January 2004. Retrospectively we analysed data (disease history, age at diagnosis, sex, baseline characteristics, and course of disease) on all children diagnosed with UC (n = 170) from 2004 to January 2018. Four children were lost to follow-up and were not included into analysis.

Of 166 children diagnosed with UC, 12 had colectomy (7.2%). Patients with colectomy, compared with UC patients who did not have colectomy, did not significantly differ in gender (girls 58% vs. 48%), age at diagnosis (12.27 vs. 12.62 years), body mass index at the time of diagnosis (median –1.2 vs. –0.22), Paediatric Ulcerative Colitis Activity Index (PUCAI) at the diagnosis (median 32.5 vs. 40), proportion of patients with extensive disease (E4) (75% vs. 57%) nor in extraintestinal manifestations (for all p was >0.05). There was a significant difference in family history positive to IBD (25% vs. 7%, p < 0.05), in the highest PUCAI each patient had during the observation period (65 vs. 40, p < 0.05), number of patients treated with azathioprine (92% vs. 28%, p < 0.01) and anti-TNF therapy (58% vs. 3.9%, p < 0.01). We found a significant decrease of colectomy rate (from 2004 to 2010 rate was 13.5% (7/52) vs. 4.4% (5/114) in the period from 2011 to 2018, p < 0.05). During the same time, the proportion of children with UC treated with anti-TNF therapy increased, although not significantly (p > 0.05): from 2004 to 2010, two children (3.8%) were treated with anti-TNF therapy, and both required colectomy, while in later period, 11 children (9.7%) received biologics, and 5 of them underwent colectomy.

Our paediatric patients presented with extensive form of the ulcerative colitis in nearly 60% of patients, requiring colectomy in overall 7.2% of patients. However, the colectomy rate decreased significantly during the observed period.

Wilson’s disease (WD) is an autosomal recessive genetic disorder that leads to the impairment of cellular copper metabolism.

Clinical presentation is heterogeneous, with predominantly hepatic, neurological and psychiatric manifestations. Acute compensated WD presenting as fulminant liver failure is a life-threatening condition for which liver transplantation is the ultimate treatment.

14-year-old girl presented with acute abdominal pain and peripheral oedema lasting two weeks before onset of abdominal pain. On initial examination, patient was febrile, complaining of periumbilical pain, dyspnea, cough, presenting with anasarca, extensive limb oedema and ascites (gained 17 kg), without encephalopathy. Laboratory evaluation revealed Coombs-negative haemolytic anaemia (Hb 101 g/L, Rtc 125×10^9/L), thrombocytopenia (79×10^9/L), leukocytosis (19.27×10^9/L), mildly elevated inflammatory markers, hypergamma globulinemia (IgG 21.7 g/L) with reduced complement components (C3 0.24 g/L, C4 <0.08 g/L), coagulopathy (INR >2.5), marked hypoalbuminemia (>15 g/L), without encephalopathy. Laboratory evaluation revealed Wilson's disease was practically confirmed after demonstrating pronounced cupriuria (17.8 μmol/24h); serum ceruloplasmin was low (0.13 g/L) and serum copper levels slightly reduced (10.1 μmol/L). Initial treatment was supportive (albumin infusions, vitamin K, fresh frozen plasma, diuretics, lactulose, antibiotics). Liver transplant