Outcomes in Children with Primary Sclerosing Cholangitis or Autoimmune Hepatitis-Overlap and Associated IBD

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Introduction:

- Primary Sclerosing Cholangitis (PSC) and Autoimmune liver disease (autoimmune hepatitis & overlap syndrome (AIHO)) are rare entities.
- PSC and AIHO have been reported to be associated with IBD.

Aims/Objective:

- To study outcomes of children diagnosed with AIHO and PSC who also have a diagnosis of IBD.
- Outcomes included portal hypertension, biochemical remission, survival of native liver and mortality

Methods:

- Retrospective study (2000-2020) of 193 patients diagnosed with AIH
- 23 patients had AIHO+IBD (n=9) or PSC+IBD (n=14)
- Casenotes were examined at intervals of 1, 3, 5, 7 and 10 years
- Biochemical remission was defined as:
  - ALT <50 iu/ L, or GGT <40 iu/ L
- Portal hypertension was defined as having any of below:
  - Platelet count <120, splenomegaly or gastroesophageal varices

Results:

- Of the PSC group: 35% (n=5) had gastric, duodenal and colonic disease; 1 patient underwent pan-proctocolectomy
- Of the AIHO group: 66% (n=6) of patients had colonic disease
- Biochemical remission:
  - AIH: 75% at 1 year (n=8); 100% by year 3 (n=3)
  - PSC: 71% at 1 year (n=8); 75% by year 3 (n=6)
- Portal hypertension: 13% (n =2) AIHO & UC
- No patient deaths or Liver transplant (median follow up 4 years)

Conclusion:

- Patients with PSC were more likely to have a diagnosis of IBD-U
- Patients with AIHO were more often diagnosed with UC
- In both groups, normalisation of GGT or ALT did not correlate with resolution of endothelial thickening of the CBD on ultrasound.