Review the diagnosis of IBD in children with AILD (Auto immune liver disease) – 8 years’ experience in a tertiary Centre

Maria Misiou, Huey Miin Lee, Ben Hope, Marianne Samyn, Babu Vadamalayan
Department of Paediatric Gastroenterology, Hepatology and Nutrition, King’s College Hospital, London, United Kingdom

Objectives
The association of autoimmune liver disease (AILD) and inflammatory bowel disease (IBD) is well documented. IBD affects about 45% of children with autoimmune sclerosing cholangitis (AISC) and about 20% of those with autoimmune hepatitis (AIH).

The aim of this study was to describe the clinical features of AILD associated with IBD, to evaluate the role of fecal calprotectin and the time to look for IBD in patients with primary diagnosis of AILD.

Methods
• Retrospective review of paediatric patients with primary diagnosis of AILD and IBD between 2010 and 2018.
• Patients who were referred or diagnosed with IBD first, were excluded.
• Diagnosis of IBD was based on clinical history, endoscopic appearance and histology findings.
• AILD patients were classified to AIH or AISC according to histology, radiology results and circulating antibodies.
• Patients demographics, symptoms, FC, blood tests, timing before IBD diagnosis and treatment were collected.

Conclusions
20% of patients with primary diagnosis of AILD had IBD. 40% had simultaneous diagnosis; all had raised FC since AILD was identified.

We recommend FC routinely in children with AILD for the early diagnosis of IBD. Colonoscopy should be considered in patients with symptoms of IBD and the ones with clearly elevated FC. The timing of the assessment is of paramount as immunosuppressive treatment can mask symptoms and change the disease activity.

Results
114 patients with AILD were identified

74 (64%) had screening with FC
48/74 (64%) abnormal
26 (35%) normal

67% of FC performed at the diagnosis of liver disease.

25/114 (21%) patients were diagnosed with IBD
✓ 14/25 (56%) had AISC
✓ 11/25 had AIH

• 14 were males and the mean age at diagnosis was 10 years.
• 25% had family history of autoimmunity.

Mean FC at diagnosis of IBD was 646 (range 60-4004)
✓ 18/25 (72%) had ulcerative colitis(UC)
✓ 2/25 (0.08%) Crohn’s disease
✓ 5/25 (0.2%) indeterminate colitis(IBDU)

Endoscopic features: pancolitis 60%, ileitis 20%
Histology: mild to moderate UC (78%)

All patients were started on steroids
✓ 9/25 were already on AZA before the endoscopic assessment for IBD

10/25 (40%) of patients had simultaneous diagnosis of AILD and IBD, all presented with symptoms of bowel disease (6/10 had PR bleeding).

15/25 (60%) were diagnosed with AILD and concomitant IBD after 19 months (mean time)
✓ 7/15 gut symptoms improved since immunomodulators started but FC was raised,
✓ 3/15 had no gut symptoms but raised FC on screening
✓ 5/15 developed bowel symptoms after liver diagnosis, (in 3 of them FC was raised since liver diagnosis)

48 patients had at least one endoscopy
25(52%) were diagnosed with IBD, 8 patients had normal endoscopy but positive FC of mean value 162, (range 60-332).
15 had no endoscopy despite abnormal FC.
6/26 patients with normal FC underwent endoscopy which was normal. Reason was persistently elevated liver enzymes, relapse of AILD or ongoing bowel symptoms.

Conflict of Interest: Nothing to declare