



SESSION 1

HEPATOBIILIARY MANIFESTATIONS OF IBD

Gideon Hirschfield, MA MB BChir FRCP PhD

Patients living with IBD are frequently at risk of having a co-existing liver disease. The nature and severity of the liver injury can vary substantially, but may become the major determining factor for patient outcomes. In reaching a diagnosis of liver disease in a patient with IBD, a systematic and logical approach to etiology, severity, and risk of progression is needed.

The leading issues when considering hepatobiliary manifestations of IBD are to first exclude underlying liver disease (such as non-alcoholic fatty liver disease), then to evaluate the possibility that the manifestation is a consequence of drug therapy. Tools such as transient elastography, which can evaluate of disease severity at baseline and over time, can be helpful adjuncts. Having excluded relatively common (but usually less significant) concerns, attention frequently turns to the diagnosis of primary sclerosing cholangitis (PSC), the classic and most clinically significant hepatobiliary manifestation of IBD. In patients younger than 40 years with ulcerative colitis, development of PSC is associated with a 6-fold increase in mortality, and 7-fold increased risk of colorectal cancer, highlighting the profound impact of the diagnosis. Other autoimmune liver diseases can also be present including, autoimmune hepatitis, primary biliary cholangitis or, rarely, IgG4 disease. These are, however, less common than PSC-IBD.

PSC-IBD has a distinct pathophysiology, and clinicians need to be aware of: the heterogeneous presentation across all ages; the diagnostic choices (importantly, MRI as a diagnostic tool, and limited ERCP for specific interventions); and the stratified approach to confirming the diagnosis, liver disease stage and risk of progression. Therapies for patients with PSC-IBD remain very limited and many patients ultimately require liver transplantation, although strategies involving ongoing surveillance of the liver and bowel are also important. Treatment with ursodeoxycholic acid does not, unfortunately, offer significant benefit. Consequently, many new insights from basic pathophysiology studies are driving early phase trials and there is a dedicated PSC-IBD programme at the University of Toronto focused on delivering integrated liver and GI research-driven, clinical-trial focused, whole-patient, life-long care. This focused approach is justified; while it is relatively infrequent, PSC-IBD is a disease with substantial unmet needs related to the heightened risk of events and complications (e.g., liver failure, recurrent cholangitis, recurrent disease in the liver graft, and elevated malignancy risk in the hepatobiliary tree and colon). Additionally, the symptom burden for patients (e.g., pain, pruritus, fatigue) is an added axis of clinical disease that requires recognition and management.

#PSC, November 2018, @AutoImmuneLiver, e:gideon.hirschfield@uhn.ca