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EVOLUTION OF PEDIATRIC AUTOIMMUNE CHOLANGITIS AND PRIMARY SCLEROSING CHOLANGITIS INTO ADULTHOOD

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Background: The natural history of primary sclerosing cholangitis (PSC) in children seems to differ from PSC in adults. However, studies on this matter have been limited by short follow-up periods and inconsistent classification of patients with autoimmune cholangitis (AIC) (or overlap syndrome). Consequently, it remains unclear if long-term outcomes are affected by the clinical phenotype.
Aims: The aims of this is study are to describe the long-term evolution of PSC and AIC in a pediatric cohort with extension of follow-up into adulthood and to evaluate the influence of phenotype on clinical outcomes.
Methods: This is a retrospective study of patients with AIC or PSC followed at CHU-Sainte-Justine, a pediatric referral center in Montreal. All charts between January 1998 and December 2019 were reviewed. Patients were classified as either AIC (duct disease on cholangiography with histological features of autoimmune hepatitis) or PSC (large or small duct disease on cholangiography and/or histology). Extension of follow-up after the age of 18 was done for patients followed at the Centre hospitalier de l'Université de Montréal. Clinical features at diagnosis, response to treatment at one year and liver-related outcomes were compared.
Results: 40 patients (27 PSC and 13 AIC) were followed for a median time of 71 months (range 2 to 347), with 52.5% followed into adulthood. 70% (28/40) had associated inflammatory bowel disease (IBD) (78% PSC vs 54% AIC; p=0.15). A similar proportion of patients had biopsy-proven significant fibrosis at diagnosis (45% PSC vs 67% AIC; p=0.23). Baseline liver tests were similar in both groups.
At diagnosis, all patients were treated with ursodeoxycholic acid. Significantly more patients with AIC (77% AIC vs 30 % PSC; p=0.005) were initially treated with immunosuppressive drugs, without a significant difference in the use of Anti-TNF agents (0% AIC vs 15% PSC; p= 0.12). At one year, 55% (15/27) of patients in the PSC group had normal liver tests versus only 15% (2/13) in the AIC group (p=0.02). During follow-up, more liver-related events (cholangitis, liver transplant and cirrhosis) were reported in the AIC group (HR=3.7 (95% CI: 1.4-10), p=0.01). Abnormal liver tests at one year were a strong predictor of liver-related events during

follow-up (HR=8.9(95% CI: 1.2-67.4), p=0.03), while having IBD was not (HR=0.48 (95% CI: 0.15-1.5), p=0.22). 5 patients required liver transplantation with no difference between both groups (8% CAI vs 15% CSP; p=0.53).
Conclusions: Pediatric patients with AIC and PSC show, at onset, similar stage of liver disease with comparable clinical and biochemical characteristics. However, patients with AIC receive more often immunosuppressive therapy and treatment response is less frequent. AIC is associated with more liver-related events and abnormal liver tests at one year are predictor of bad outcomes.

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