The Journal of Pediatrics

Volume 164, Issue 4, April 2014, Pages 714-719.e2

Original Article

Outcome after Discontinuation of Immunosuppression in Children withÂ Autoimmune Hepatitis: A Population-Based Study

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https://doi.org/10.1016/j.jpeds.2013.12.008

Objective

To assess sustained immunosuppression-free remission (SIFR) in children with autoimmune hepatitis (AIH).

Study design

We retrospectively reviewed all children with AIH in the region between 1986 and 2011 using a population-based methodology.

Results

We identified 56 children with AIH (62.5% females; median age, 11.1Â years [IQR, 5.7-14.4Â years], followed for a median of 5.6Â years [IQR, 2.8-8.6Â years]). Liver disease

14.1Â years], followed for a median of 5.8Â years [IQR, 2.8-8.8Â years]). Liver disease was characterized by type II AIH in 8.9%, cirrhosis in 14.0%, and primary sclerosing cholangitis in 21.4%. Coexisting nonhepatic immune-mediated diseases occurred in 37.5%. Biochemical remission on immunosuppressive therapy was achieved in 76.4% of all patients with AIH at a median of 1.2Â years (IQR, 0.4-3.6Â years); 23.1% of these patients experienced a subsequent relapse. Discontinuation of all immunosuppressive medications was attempted in 16 patients and was successful in 14 patients (87.5%) with type 1 AIH (median age at discontinuation, 8.9Â years [IQR, 3.5-17.9Â years], treated for a median of 2.0Â years [IQR, 1.3-3.5Â years] after diagnosis), with SIFR occurring at a median of 3.4Â years (IQR, 2.6-5.8Â years) of follow-up. Excluding patients with inflammatory bowel disease who received immunosuppressive therapy independent of their liver disease, the probability of achieving SIFR within 5Â years of diagnosis of AIH was 41.6% (95% CI, 25.3%-62.9%). Baseline patient characteristics associated with an inability to achieve biochemical remission on immunosuppression or SIFR were elevated international normalized ratio, positive antineutrophil cytoplasmic antibody titer, cirrhosis, and a nonhepatic autoimmune disorder.

Conclusion

We found a high rate of successful discontinuation of all immunosuppressive medications in carefully selected patients with AIH in a population-based cohort. SIFR is an achievable goal for children with AIH, particularly those with type I disease in stable biochemical remission on immunosuppressive therapy.

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AIH, Autoimmune hepatitis; ANCA, Antineutrophil cytoplasmic antibody; ASC, Autoimmune sclerosing cholangitis; IBD, Inflammatory bowel disease; INR, International normalized ratio; PSC, Primary sclerosing cholangitis; SIFR, Sustained immunosuppression-free remission; TSH, Thyroid-stimulating hormone; TTG, Tissue transglutaminase

Supported by the National Center for Research Resources and the National Center for Advancing Translational Sciences, National Institutes of Health (NIH; 8UL1TR000105 [formerly UL1RR025764]). The content is solely the responsibility of the authors and does not necessarily represent the official views of the NIH. The authors declare no conflicts of interest.

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