



Autoimmune Liver Disease Associated With Celiac Disease in Childhood: A Multicenter Study

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Abstract:

BACKGROUND & AIMS: Celiac patients are at risk to develop an autoimmune liver disease. The aim of this study was to describe the clinical features of children and adolescents presenting with an autoimmune liver disease associated with celiac disease.

METHODS: A retrospective multicenter national survey was made for the period 1990-2005. **RESULTS:** Among 140 pediatric patients with autoimmune liver disease in Italy, we identified 23 with celiac disease: 19 with autoimmune hepatitis, 2 with autoimmune cholangitis, and 2 with overlap syndrome. Diagnosis of celiac disease preceded the diagnosis of liver disease in 18 of them, but elevation of aminotransferase activity was present in 16 when celiac disease was diagnosed. Acute hepatitis developed in 2 infants on gluten-free diet, and a hidden celiac disease was discovered in 5 other patients. Nineteen patients had liver-related non-organ-specific autoantibodies. Liver histology showed inflammatory lesions with features of autoimmune damage and different degrees of fibrosis in all of them and cirrhosis in 4. All patients, on gluten-free diet, achieved remission on immunosuppressive therapy, 14 relapsed because of discontinuation of therapy or during spontaneous gluten challenge, 20 are still on immunosuppressive treatment, and 3 could stop therapy.

CONCLUSIONS: Autoimmune liver diseases are frequently associated with celiac disease, but they might remain undiagnosed because of lack of symptoms, because of absence of liver-specific autoantibodies, or because of a misdiagnosis of celiac hepatitis. Acute hepatitis in celiac patients should induce one to suspect an autoimmune origin. Patients with autoimmune liver disease might have a hidden celiac disease, suggesting a rigorous check in any cryptogenic liver disease.

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