

Abstract Category:

- HEPATOLOGY

Abstract Title:

Prevalence and nature of the associated autoimmune disorders in children with autoimmune liver disease

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Body of Abstract:

Objectives: Timely detection of concurrent autoimmune disorders (AID) present in autoimmune liver disease (AILD) may influence the subsequent patient outcome. This prospective study determined the prevalence of autoimmune disorders of type-1 diabetes mellitus (T1DM), celiac disease (CD), hypothyroidism, autoimmune hemolytic anemia (AIHA) in children with AILD. Methods: From June 2014 to June 2015, 55 consecutive AILD children (<18 years) diagnosed according to simplified scoring system were enrolled¹. After detailed history and examination, blood investigations were done (fasting plasma sugar, post glucose

challenge plasma sugar after 2 hours of 1.75g/kg glucose (max 75g), glycosylated hemoglobin, thyroxine, thyrotropin stimulating hormone, IgA tissue transglutaminase, IgA and direct coombs test (DCT). Duodenal biopsy was done in children with tTG >3times upper limit of normal. Results: 55 children (31 girls) with AILD (31 probable, 24 definite) were enrolled. The median age at diagnosis was 9 (range 2-17) years and median duration of symptoms was 7 (1-108) months. 71% (39/55) patients had type 1 AILD, 23% (13/55) had type 2 and 6% (3/55) were seronegative. 18% (10/55) children had a family history of AID (hypothyroidism, T1DM, vitiligo, CD). 20% (11/55) children had vitiligo. Concurrent autoimmune disorders were detected in 20(36%) cases : 7 CD (3 potential CD), 7 DM (5 required insulin), 4 AIHA (DCT positive in all, 2 required blood transfusion), 2 hypothyroidism and 2 hypoparathyroidism (1 had tetany and other asymptomatic, PTH low in both). AID were equally common in type 1 and 2 AILD (13/39vs 6/13;p=0.51) . 55%(11/20) children were symptomatic for the concurrent AID. 5/20 patients had multiple AID. One had autoimmune polyendocrinopathy syndrome-1 (APS-1) with symptoms of hypoparathyroidism (confirmed by anti- interferon autoantibodies), second had APS type-3b with symptoms of autoimmune hypothyroidism (anti-thyroid peroxidase antibody positive) along with pernicious anemia (anti-parietal cell antibody positive, atrophic gastritis and megaloblastic anemia), third had hypoparathyroidism and idiopathic thrombocytopenia, fourth had potential CD with DM and fifth child had multiple autoimmune disorders [DM, AIHA, renal tubular acidosis with hypokalemia, sjogren syndrome (dry mouth and anti SS-A(Ro), anti SS-B (La) positive)]. Conclusions: A third of children with AILD have concurrent autoimmune disorders, equally common in type 1 and 2 disease. 45% are asymptomatic and would be missed unless screened. All AILD children should undergo screening tests to detect these disorders. Reference Mileti E et al. Validation and modification of simplified diagnostic criteria for autoimmune hepatitis in children. Clin Gastroenterol Hepatol 2012;10:417-21.