Rheumatic Manifestations and Connective Tissue Diseases in Autoimmune Hepatitis of the Child and the Adult.

Federico Zazzetti1, Nora C. Fernandez1, Javier Benavidez1, Luis A. Colombato1, Graciela R. Rodriguez2, Graciela Nardi2, Carolina Bru Moro´n3, Oscar L. Rillo4, Nelo A. Quadrini3, Stella M. Garay5, Mariana Fabi5, Teresita Gonzalez5 and Juan C. Barreira1. 1Buenos Aires British Hospital, Buenos Aires, Argentina, 2Hospital Dr. Ignacio Pirovano, Buenos Aires, Argentina, 3Hospital Dr. Enrique Tornu´, Buenos Aires, Argentina, 4Hospital Tornu´, Buenos Aires, Argentina, 5Hospital IAEP Sor Marı´a Ludovica, La Plata, Argentina

Background/Purpose: Autoimmune hepatitis (AIH) is a progressive fibrosing inflammatory disease of the liver of unknown etiology, leading to cirrhosis. Its course is usually fluctuating and its clinical manifestations protean. Forty % of type I AIH and 80% of type II is diagnosed in childhood. Up to 30% of patients with AIH will develop rheumatic manifestations. Moreover, AIH can be associated with various connective tissue diseases (CTD), although there is a lack of studies comparing pediatric and adult populations in this regard. The aim was to analyze the frequency of rheumatic manifestations and CTD in patients with AIH in a pediatric population (16 years old) compared with adults.

Methods: Patients of any age who fulfilled the 1998 modified diagnostic criteria for AIH of the Autoimmune Hepatitis International Group assisted in Hepatology services of the intervening centers. They were clinically evaluated by a hepatologist and referred to a rheumatologist. Demographic, clinical and serological variables, as well as additional studies and biopsies were recorded. Rheumatic manifestations and CTD were registered and defined according to ACR criteria. Means were reported for numeric variables and percentages for categorical. To compare two groups Mann Whitney and 2tests were applied. A 0.05 alpha-error was chosen.

Results: Forty-eight patients with AIH were analyzed: 28 adults, 89% female, mean age at inclusion 50 17 years (range19–75) and age at diagnosis of AIH 41 18 years (range18.6–70), and 20 pediatric, 75% female, mean age at inclusion 13.8 4 years (range5–16) and age at diagnosis of AIH 9.7 4 years (range1.5–16). Sixty-four % (18/28) of adults and 55% (11/20) of pediatric had at least one rheumatic manifestation (pn/s). There were no differences in the type and frequency of rheumatic manifestations analized in the two age groups, including malar rash, oral ulcers, parotitis, photosensitivity, xerophthalmia, xerostomia, telangiectasias, Raynaud's, vasculitis, xeroderma, subcutaneous nodules, arthralgia, arthritis, myositis, interstitial lung disease, serositis, citopenias, glomerulonephritis, proteinuria and nervous system involvement. Nine CTD were found, 5 in adults (17.8%, SLE4, Sjo¨gren1) and 4 in pediatric patients (20%, SLE3, JIA 1). Comparing both groups, the pediatric group had more jaundice (p0.03), hepatomegaly (p0.0001), cirrhosis (p0.03), splenomegaly (p0.003), anemia (p0.003), ESR (p0.04), AST (p0.0006), ALT (p0.006), FAL (p0.01), total bilirubin (p0.003), ANA (p0.04), ASMA (p0.0001) and LKM (p0.03). There were no differences regarding histological findings of liver biopsies. Autoimmune thyroiditis was more frequent in adults 4/28 vs 1/20 (p0.001).

Conclusion: The frequency of rheumatic manifestations and CTD was similar in both groups, but pediatric patients had a more severe hepatic involvement and higher levels of autoantibodies. Regardless of age, 2 of 10 patients with AIH presented an associated CTD, and at least half of them developed rheumatic manifestations. Systemic lupus was the most frequent CTD associated to both groups. The systematic search for defined CTD and/or rheumatic manifestations should be an essential part of the clinical evaluation of AIH.