Conclusion: We showed that SLE patients who had constitutional findings and LAP could be the presenting feature and may confer a significant diagnostic challenge in which the patients might require invasive procedures and advanced imaging modalities. PET/CT revealed reactive changes in lymph nodes in all patients but lymph node biopsy revealed atypical changes in some patients suggestive of viral infections. On the other hand, LAP patients with constitutional symptoms represents a severe phenotype of the SLE as there were significant renal and thrombotic disease.

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POS0751

COMORBIDITY AND LONG-TERM OUTCOME IN PATIENTS WITH CONGENITAL HEART BLOCK: PRELIMINARY DATA OF THE ITALIAN REGISTRY ON THE IMMUNE-MEDIATED CONGENITAL HEART BLOCK

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Background: Congenital heart block (CHB) is due to placental transfer of maternal anti-Ro/SSA autoantibodies to the fetus. The prevalence of CHB has been estimated as 1-2% in anti-Ro/SSA women while the recurrence rate is 16-19% (1). This condition is associated with a high rate of fetal/neonatal mortality and most of the cases requires pacemaker (PM) pacing. Given the rarity of CHB, limited data are available regarding the long-term follow-up of the offspring other than the cardiovascular complications.

Objectives: The results of the Italian Registry of the autoimmune congenital heart block were recently described (2). A peculiarity of this cohort was that most of the mothers had an established diagnosis of systemic autoimmune disease at CHB detection, in contrast with other registries where CHB was mostly incidentally detected in healthy women. Here we report an update, with the preliminary data regarding the long-term outcome of patients with CHB, their unaffected siblings and health controls born from mothers positive for Ro/SSA.

Methods: Data regarding demography, treatment, maternal, neonatal outcome, and follow-up were collected through an online electronic datasheet. A dedicated questionnaire was created with the aim to investigate general health, cardiovascular follow-up, and frequency of autoimmune diseases.

Results: One-hundred and five cases of CHB in 99 patients were included from 1969 to December 2020. CHB was mostly detected in utero (97 cases, 92.3%) with 8 neonatal cases. Third degree CHB occurred in 71 cases (67.6%). Child mortality was observed in 29 (27.6%) cases: 20 in utero, 7 during neonatal period and 2 during childhood. Overall, a PM was implanted in 54 out of the 85 live births (63.5%). Then, our cohort was divided into 2 subgroups: pregnancy that occurred

before (N=61) and after 2010 (N=44) with the aim to evaluate possible differences among the subgroups. Whereas mortality, PM, CHB degree were similar, CHB more frequently occurred in the last 10 years among Ro/SSA asymptomatic carriers than in the group of pregnancies before 2010 (53.6% vs 32.8%, p=0.038). Questionnaires from 14 surviving CHB cases, 8 unaffected siblings 12 controls born from mothers Ro/SSA positive were collected. Among CHB cases, 6 were males and 8 females, median age 12 years (range 6-28). All presented a third degree CHB, 10 required a neonatal PM pacing and one had an implantable ECG recorder. PM was substituted at least once in 9 patients, the oldest patient had to change it four times. No dilated cardiomyopathy occurred and most of the patients maintain an annual follow-up. Two cases of autoimmune diseases were registered among CHB cases, one idiopathic juvenile arthritis and one Cogan's vasculitis, both born from mothers with Sjogren Syndrome. Four cases of neurodevelopmental disorders occurred: three cases of learning disabilities (one in each group) and one case of speech disorder in the sibling group. In addition, a CHB case presented a stress disorder linked to frequent hospitalizations.

Conclusion: This registry is an ongoing project aiming at collecting all Italian CHB. Moreover, here we reported the preliminary data concerning the evaluation of long-term follow-up of CHB patients. Our data, even if need to be confirmed in larger cohort, seems reassuring: no differences were reported comparing CHB patients with unaffected siblings or controls.

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POS0752

INTERSTITIAL LUNG INVOLVEMENT IN SYSTEMIC LUPUS ERYTHEMATOSUS: RELEVANT CLINICAL CHARACTERISTICS. OUR EXPERIENCE IN A THIRD LEVEL HOSPITAL

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Background: Lung involvement¹ in Systemic Lupus Erythematosus (SLE) is heterogeneous, with the pleura being more frequently affected. Associated diffuse interstitial lung disease (ILD) is rare (3-13%) with difficult diagnosis and therapeutic management.

Objectives: Assess the main clinical-epidemiological characteristics of patients diagnosed with SLE with ILD in a third level hospital and analyze the possible relationship between them.

Methods: Descriptive observational study of patients diagnosed with SLE (SLICC criteria) with ILD in our hospital between 1973 and 2020. The following clinical-epidemiological characteristics were evaluated such as sex, mean age at diagnosis, presence of cardiovascular risk factors (CVRF), smoking, baseline cardiac and respiratory comorbidities, laboratory markers (autoimmunity, CRP and vitamin D), baseline SLEDAI, manifestations of SLE, interstitial pattern on HRCT, correlation with conventional radiology (Rx), respiratory function tests, clinical and physical examination at diagnosis.

Results: 455 patients diagnosed with SLE were included, of whom 20 had ILD (4.4%). 65% were women with a mean age at diagnosis of 63 ± 16.23 years. 30% presented ILD as the first clinical data for the diagnosis of SLE. 30% had jobs with exposure to chemicals. 25% were ex-smokers and 10% were active smokers. 10% had associated CVRF, highlighting HT (35%). 30% had valve disease on echocardiography such as mitral regurgitation (30%) and tricuspid regurgitation (20%). Only 10% had associated bronchopathy. 35% and 25% associated, respectively, pleural serositis and secondary Sjogren's syndrome. Arthritis and skin involvement were documented in 60% for both clinical domains. 40% had SLEDAI > 6.

At the diagnosis of ILD, 90% had symptoms such as dyspnea (75%) and cough (60%). 80% had crackles on examination. 85% had alterations in the X-ray and 100% in the CT scan with the NINE patterns as the main one (65%) with progression at 2 years despite treatment in 25%. Spirometry differentiated an obstructive pattern (25%), restrictive (20%) and normal (55%) with a tendency to stability at one year (40%) with the prescribed treatment. 90% had a decrease in diffusion, the majority being mild (67%). Analytically, 67% had elevated levels of CRP and hypovitaminosis D; 40% elevated DNA titers, 50% hypocomplementemia and 60% ENAS positive, highlighting Ro (45%); La (15%) and RNP (15%). Leukopenia and thrombopenia (30% and 10% respectively) were also observed.

In our study, we did not find any statistical significance between the variables analyzed, due to the small sample size.

Conclusion: 1)ILD is an infrequent manifestation in SLE, it usually associates respiratory symptoms and pulmonary functional repercussions with the diagnosis. The most characteristic radiological pattern is the NINE.