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. RET/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.

Disclosure of Interests: None declared

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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 antibodies 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%. This condition is associated with a high rate of fetal/neonatal mortality and renal and thrombotic disease. 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 (IHCHB) were reported (1). 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 Ro/SSA positive.

Methods: Data on retrospective registry, treatment, maternal, neonatal and childhood outcome, and follow-up were collected through an online electronic database. 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 (76%). Child mortality was observed in 27 (27.5%) cases, during the perinatal 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<0.03). 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. Interestingly, one 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: Lupus involvement 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: To assess the 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 Sjögren’s syndrome. Arthritis and skin involvement were documented in 60% for both clinical domains. 40% had SLEDAI 8.

At 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%). Analysis, 67% had elevated levels of CRP and hypovitaminosis D; 40% elevated DNA titers, 50% hyocomplementemia 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 a frequent manifestation in SLE, it usually associates respiratory symptoms and pulmonary functional repercussions with the diagnosis. The most characteristic radiological pattern is the NINE.
In our study, ILD was more frequently associated with women and joint or skin symptoms, as well as with data on immunological activity, without statistical significance.

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SUBSPECIALTY LUPUS CLINIC CARE IS ASSOCIATED WITH HIGHER QUALITY CARE THAN SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Healthcare quality for systemic lupus erythematosus (SLE) is a modifiable target for improving patient outcomes. Disease-specific subspecialty clinics offer experienced healthcare professionals, collaborative multidisciplinary teams and streamlined care processes. A single centre study in the USA has suggested superior performance of the subspecialty lupus clinic in the provision of quality care (1), but this has not been examined outside the USA where access to care may be influential.

Objectives: To assess the quality of SLE care provided in a subspecialty lupus clinic compared with hospital general rheumatology and private rheumatology clinics in a non-US, universal healthcare setting.

Methods: Lupus patients (n = 258) were recruited in 2016 from various clinic settings in Australia, including a subspecialty lupus clinic (n = 147), two hospital general rheumatology clinics (n = 56) and two private clinics (n = 55). Quality of care was assessed using 31 validated SLE quality indicators (QI) encompassing diagnostic work-up, disease and comorbidities assessment, drug monitoring, preventative care and reproductive health. Data were collected from medical records and patient questionnaires. Overall and individual QI performance was calculated and compared between the three clinic settings, and multivariable regression was performed to adjust for sociodemographic, disease and healthcare factors.

Results: Median [IQR] overall performance on eligible QIs was higher in the lupus clinic (66.7% [16.9]) than the hospital general rheumatology (52.7% [10.6]) and private rheumatology (50.0% [18.0]) clinics (p <0.01), and remained significant with multivariable adjustment. This trend was still observed when the overall performance was reassessed to include patient self-report (73.1% [14.8] vs 68.1%, 11.5% [63.2%] vs 0.0%), this difference may be due to consistent formal assessments of disease activity (100% vs 0%, p <0.01) and disease damage (95.9% vs 0%, p <0.01). Performance was high across all clinic settings for disease activity (100% vs 0% vs 0%, p <0.01) and disease damage (95.9% vs 0% vs 0%, p <0.01). This difference may be due to consistent formal assessments of disease activity (100% vs 0% vs 0%, p <0.01) and disease damage (95.9% vs 0% vs 0%, p <0.01) across all clinic settings for disease activity (100% vs 0% vs 0%, p <0.01) and disease damage (95.9% vs 0% vs 0%, p <0.01). This difference may be due to consistent formal assessments of disease activity (100% vs 0% vs 0%, p <0.01) and disease damage (95.9% vs 0% vs 0%, p <0.01). This difference may be due to consistent formal assessments of disease activity (100% vs 0% vs 0%, p <0.01) and disease damage (95.9% vs 0% vs 0%, p <0.01). This difference may be due to consistent formal assessments of disease activity (100% vs 0% vs 0%, p <0.01) and disease damage (95.9% vs 0% vs 0%, p <0.01).

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DEVELOPMENT OF A RISK PREDICTION MODEL FOR VENOUS THROMBOEMBOLISM IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS: THE SLE-VTE SCORE

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Background: Patients with systemic lupus erythematosus (SLE) have a substantially increased risk of venous thromboembolism (VTE). An individual VTE risk assessment is important to ensure that all patients are assessed and given adequate thromboprophylaxis.

Objectives: We conducted this study to develop a risk score for VTE in patients with SLE.

Methods: Patients with SLE who participated in the Chinese SLE Treatment and Research group were enrolled in this study. Patient baseline information and clinical laboratory indicators were obtained, and VTE events were recorded every 3-6 months during follow-up visits. The risk prediction model was created and internally validated using the bootstrap methods, and a scoring system was established (Figure 1).

RESULTS: Median [IQR] overall performance on eligible QIs was higher in the lupus clinic (66.7% [16.9]) than the hospital general rheumatology (52.7% [10.6]) and private rheumatology (50.0% [18.0]) clinics (p <0.01), and remained significant with multivariable adjustment. This trend was still observed when the overall performance was reassessed to include patient self-report (73.1% [14.8] vs 68.1%, 11.5% [63.2%] [13.4], p <0.01). This difference may be due to consistent formal assessments of disease activity (100% vs 0% vs 0%, p <0.01) and disease damage (95.9% vs 0% vs 0%, p <0.01) at the lupus clinic. Performance was high across all clinic settings for disease activity (100% vs 0% vs 0%, p <0.01) and disease damage (95.9% vs 0% vs 0%, p <0.01). This difference may be due to consistent formal assessments of disease activity (100% vs 0% vs 0%, p <0.01) and disease damage (95.9% vs 0% vs 0%, p <0.01). This difference may be due to consistent formal assessments of disease activity (100% vs 0% vs 0%, p <0.01) and disease damage (95.9% vs 0% vs 0%, p <0.01).

Figure 1. Flow chart of study design.