cases, 90.6%) with 5 neonatal CHB. Demographic description of the mothers, pregnancy outcomes and treatment are reported in table 1. Child mortality was observed in 22 (25.5%) cases: 12 fetal, 5 termination of pregnancy and 5 neonatal. Maternal and fetal risk factors for fetal mortality were analysed and, at univariate analysis, factors associated with death were an earlier detection of CHB (20.9 ±0.9 weeks vs 24.8±5.4 weeks; p=0.007), hydrops (p=0.002;OR=11.3;95%CI 1.84–69.2) and pericardial effusion (p=0.025;OR=100;95%CI 98.8–100).

Conclusions: The Lu.Ne registry is an ongoing project aiming at collecting all Italian cases. Our data showed similar rate of fetal/neonatal death and of PM implantation previously reported. We confirmed that hydrops and pericardial effusion are risk factors for fetal death. A peculiarity of our cohorts is that the majority of the mothers (59%) had an established diagnosis of systemic autoimmune disease at CHB detection. This is in contrast with other registries showing that usually CHB was incidentally detected in healthy women and related to the recruiting Centres all belonging to Rheumatology Society. The collection of cases from Gynaecological and Paediatric Centres, planned in the next months, will complete our analysis.

REFERENCES:

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SAT0449 JUVENILE ONSET SYSTEMIC LUPUS ERYTHEMATOSUS OUTCOME IN ADULTHOOD: A MONOCENTRIC RETROSPECTIVE COHORT

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Background: Outcome of juvenile-onset SLE (j-SLE) during adulthood is poorly described.

Objectives: To report adult outcome of j-SLE and compare SLE course during childhood and adulthood.

Methods: j-SLE was defined as a SLE fulfilling ACR criteria and diagnosed before the age of 16 years. Mac Nemar test for paired nominal data and Wilcoxon signed rank test for paired data were used.

Results: One hundred and six patients j-SLE (88 women and 18 men; female to male ratio: 4.9): mean age at diagnosis: 12.3 years followed during a mean duration of 13.8 years, from childhood (mean: 4 years) to adulthood (mean: 10.3 years). 97.2% patients received corticosteroids (with intravenous pulses for 18.7%). Other DMARDs were used: 75.2% patients received methotrexate, 60.7% hydroxychloroquine, 35.9% azathioprine, 35.9% mycophenolate mofetil, 21.4% cyclosporine, 17.7% cyclophosphamide. 10.3% patients received anti-malarial drugs.

Clinical manifestations of the first flare were: arthritis (67.9%), cutaneous (57.5%), nephritis (23.6%), fever (17.9%), hematologic: ITP, AIHA (15%). Neuropsychiatric 5.2% and clinical manifestations during childhood. Disease course during adulthood had two patterns: 82 patients (77.3%) had at least one relapse (24.8±5.4 weeks; p=0.007), hydrops (p=0.002;OR=11.3;95%CI 1.84–69.2) and pericardial effusion (p=0.025;OR=100;95%CI 98.8–100).

Conclusions: The Lu.Ne registry is an ongoing project aiming at collecting all Italian cases. Our data showed similar rate of fetal/neonatal death and of PM implantation previously reported. We confirmed that hydrops and pericardial effusion are risk factors for fetal death. A peculiarity of our cohorts is that the majority of the mothers (59%) had an established diagnosis of systemic autoimmune disease at CHB detection. This is in contrast with other registries showing that usually CHB was incidentally detected in healthy women and related to the recruiting Centres all belonging to Rheumatology Society. The collection of cases from Gynaecological and Paediatric Centres, planned in the next months, will complete our analysis.

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SAT0450 ESTIMATING DURATION OF RESPONSE IN SYSTEMIC LUPUS ERYTHEMATOSUS TREATMENTS

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Background: The primary endpoint in SLE trials is usually response to therapy at a landmark visit. However, during a trial, patients may alternate between response and non-response states. Duration of response would therefore be important to assess, but the optimal approach for estimating response duration has not been determined. Analysing response duration only among responders at a landmark visit can result in selection bias. Drop-outs and missed visits further complicate estimation of response duration.

Objectives: To quantify response duration and assess baseline predictors of transitions into and out of response in SLE patients receiving standard of care (SoC) by fitting a multi-state Markov (MSM) model.

Methods: Data on 759 SLE patients with active disease (SLEDAI >6 at entry) randomised to SoC in 52 week trials was obtained from the Collective Data Analysis Initiative (CDAI) database of the Lupus Foundation of America. The following monthly response endpoints (without medication stipulations) were analysed: SRI-4, SRI-5, SRI-6, and BICLA. A MSM model allowing for bi-directional transitions between response and non-response states was fit to estimate the probability of being in response at 52 weeks, average duration of response (sojourn time) and mean total time in response. Predictors of attainment and loss of SRI-5 response were also identified.

Results: Based on the MSM model, the probability of being in response at 52 weeks ranged from 42% (SRI-6) to 61% (SRI-4), higher than conventional 52 week landmark response rates that assume non-response for missing data. The estimated mean duration of response ranged from 20.4 weeks (BICLA) to 31.5 weeks (SRI-4). Mean total time in response over 52 weeks based on all transitions was 16.4–24.8 weeks. After adjusting for baseline SLEDAI score, patients with lower anti-dsDNA titer were more likely to achieve and maintain SRI-5 response (p<0.001). Younger age (p<0.001) and higher protein/creatinine ratio (p<0.001) were associated with higher frequency of SRI-5 response but also shorter response duration. Response duration was also shorter in patients who were non-White (p<0.001), had longer history of disease (p<0.03), and lower lymphocyte count (p=0.001) at baseline.

Conclusions: Factors associated with greater disease severity were consistently associated with shorter response duration on SoC, despite exhibiting variable effects on the probability of achieving response at a given time. Response duration might therefore provide a more discriminating measure to distinguish effective investigational treatments from background SoC, although this remains to be tested. Multi-state models make better use of complex longitudinal clinical trial data and provide a more comprehensive view of the response profile and the role of patient characteristics in different aspects of response.

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