are under 18-year-old. Each patient was evaluated if he or she satisfied the 1997, 2012, or 2017 criteria.

Results: Among the 100 cases, 9 were male and 91 were female, with wide range of age;^{18–88} mean 44.5-year-old). Asian accounted for 98%, with Hispanic 1%, and Caucasian 1%. The sensitivity of 1997, 2012, 2017 criteria are, 97%, 99%, 87% respectively. The positivity rate of each domain in the 2017 criteria was shown in table 1. The total score that the patients got in the 2017 criteria ranged from 12 to 44 (mean: 27.3). All the cases who were classified as non-SLE in the 2017 criteria had anti-nuclear antibody (ANA) <80; all of them scored more than 10 points.

Abstract SAT0447 – Table 1. Validation of the 2017 SLE classification criteria using clinically diagnosed 100 SLE cases.

Item (point)	Positivity (%)	
Entry Criteria		
ANA ≥80 (required: no point)	87.0	
Constitutional Domain		
Unexplained fever >38.3° C (2)	47.0	
Muco-Cutaneous Domain		
Non-scarring alopecia (2)	48.0	
Oral ulcers (2)	27.0	
SCLE or DLE (4)	42.0	
ACLE (6)	70.0	
Arthritis Domain		
Synovitis or tenderness in ≥2 joints or tenderness and ≥30 minutes of morning stiffness (6)	79.0	
Neurologic Domain		
Delirium (2)	0	
Psychosis (3)	4.0	
Seizure (5)	7.0	
Serositis Domain		
Pleural or pericardial effusion (5)	29.0	
Acute pericarditis (6)	0	
Hematological Domain		
Leukopenia (3)	77.0	
Thrombocytopenia (4)	26.0	
Autoimmune hemolysis (4)	14.0	
Renal Domain		
Proteinuria >0.5g / 24 hours (4)	64.0	
Renal biopsy with Class II or V lupus nephritis (8)	12.0	
Renal biopsy with Class III or IV lupus nephritis (10)	17.0	
Antiphospholipid Domain		
aCL (IgG) >40 GPL units, anti-β2-GPI >40 units, or lupus anticoagulant positive (2) *	17.0	
Complement Proteins Domain		
Low C3 or Low C4 (3)	40.0	
Low C3 and Low C4 (4)	40.0	
Highly Specific Antibodies Domain		
Anti-dsDNA antibody (6)	74.0	
Anti-Smith antibody (6) *	28.0	
Rate of the patients classified as SLE (ANA≥80 and got ≥10 points overall)		
	87.0	

*N/A in 3, *N/A in 1.

Abbreviations: ACLE, acute cutaneous lupus erythematosus; ANA, anti-nuclear antibody; anti-CL, anti-cardiolipin antibody; DLE, discoid lupus erythematosus; N/A, not available; SCLE, Sub-acute cutaneous lupus erythematosus; SLE, systemic lupus erythematosus.

*N/A in 3, *N/A in 1.

Abbreviations: ACLE, acute cutaneous lupus erythematosus; ANA, anti-nuclear antibody; anti-CL, anti-cardiolipin antibody; DLE, discoid lupus erythematosus; N/A, not available; SCLE, Sub-acute cutaneous lupus erythematosus; SLE, systemic lupus erythematosus.

Conclusions: 2017 criteria for SLE accomplished modestly high sensitivity in the real-world practice, but not as high as 1997 and 2012 criteria. They possibly miss-classify the real SLE cases as non-SLE, especially if patients have low titer (<80) of ANA.

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SAT0448

REPORT OF 86 CASES OF CONGENITAL HEART BLOCK: DATA FROM THE ITALIAN REGISTRY (LU.NE REGISTRY)

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Background: Cardiac neonatal lupus is due to placental transfer of maternal anti-Ro/SSA and anti-La/SSB autoantibodies to the fetus and mainly includes congenital heart block (CHB) and dilated cardiomyopathy. 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 in the majority of cases requires pacemaker pacing.^{2–6}

Objectives: The rarity of this condition requires the establishment of collaborative registries in order to improve our knowledge. Here we report the data of the ongoing Italian Registry of the autoimmune congenital heart block (Lu.Ne), which was created in 2016

Methods: The aim was to collect retrospective and prospective pregnancies complicated with CHB in patients with antibodies. Data regarding demography, treatment, maternal and neonatal outcome and follow-up were collected through an online electronic datasheet prepared in a Research Electronic Data Capture platform.

Maternal demography	n=82
Maternal Diagnosis at CHB detection	
Undifferentiated Connective Tissue Disease	26 (31.7)
Sjogren Syndrome	16 (19.5)
Systemic Lupus Erythematosus	4 (4.8)
Carriers Ro/SSA	18 (21.9)
Carriers Ro/SSA+La/SSB	15 (18.4)
Other	3 (3.7)
Autoantibodies profile	
Anti-Ro/SSA	78 (95)
Anti-La/SSB	47 (53)
Pregnancy Outcome	
Live birth	71 (82.5)
Intrauterine fetal death/termination of pregnancy	15 (17.5)
CHB detection	
In utero	81 (94.2)
Neonatal	5 (5.8)
Mean week of detection of CHB (±SD)	22.4±8.5
CHB degree	
II degree	17 (19.7)
III degree	59 (68.7)
II-III intermittent	4 (4.6)
Other/unknown	5 (5.8)
Therapy after in utero CHB detection	
Fluorinated steroids	45 (55)
Intravenous Immunoglobulins	19 (23)
Plasma-exchange	18 (22)
Postnatal outcome of liveborns with CHB	
At birth/Neonatal Pacemaker implantation	27 (38)
Neonatal death	3 (4.4)
Later Pacemaker implantation	23 (32.4)
Infant/childhood death	2 (3.2)

Results: Eighty-six cases of CHB were collected in 82 women with 85 pregnancies that occurred between 1969–2016. CHB was mostly detected in utero (81

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 postnatal. 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 $\pm$ 0.9 weeks vs 24.8 $\pm$ 5.4 weeks; $p=0.007$), hydrops ($p=0.002$; OR=11.3; CI95% 1.84–69.2) and pericardial effusion ($p=0.025$; OR >100; CI95% 2.88–>100).

Conclusions: The Lu.Ne registry is an ongoing project aiming at collecting all Italian CHB. 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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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 were 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 50.9%) and 77.3% immunosuppressant drugs. 105 (99%) 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 manifestations were found in 6.6%. Digestive involvement was only present in 6% of cases. According to the revised SELENA FLARE INDEX (SFI), the first flare was severe for half of the patients (n=55).

Disease course during adulthood had two patterns: 82 patients (77.3%) had at least one SLE flare and 24 (22.6%) a sustained remission. Mean follow up was however significantly higher in the relapsing group (15 years vs 9.8 years, $p=0.0014$). No difference was found between these 2 groups for first flare severity and clinical manifestations during childhood.

Significantly more cutaneous (61.3 vs 42.4%, $p=0.003$), musculoskeletal (75.5 vs 59.4%, $p=0.007$), neuropsychiatric (10.4 vs 3.8%, $p=0.035$), or hematologic manifestations like AIHA (9.4 vs 2.8%, $p=0.039$) or ITP (26.4 vs 10.4%, $p=0.001$), and fever (32.1 vs 3.7%, $p<0.001$) were observed during childhood than during adulthood. Nephritis occurred at similar frequencies in childhood and adulthood (34.9% and 30.2% respectively). Half of adulthood nephritis were relapses of j-SLE nephritis.

At the end of the survey mean global SLICC damage index (SDI) was 0.64. Mean childhood SDI was lower than mean adulthood SDI (0.21 vs 0.45, $p=0.016$). However mean adult follow up was significantly longer. Mean SDI increase per year was similar during childhood and adulthood (0.053 vs 0.049 respectively, $p=0.563$).

13 patients (12.3%), had musculoskeletal damage occurring more frequently during adulthood than childhood (11 vs 2, $p=0.022$), specially avascular necrosis (8 vs 0, $p=0.008$). 5 (4.7%) patients had a renal damage that occurred mostly during adulthood (n=4). Ocular damage was present for 9 (8.5%) patients. Premature gonadal failure occurred for 5 women (5.7%). Among the 88 women 19 had 32 pregnancies, leading to 22 births.

Conclusions: Damage accrual seems to increase at the same pace during childhood and adulthood. SDI was low at the end of the survey. This could reflect a protective role for HCQ, more immunosuppressant use with lower dose of steroids. Juvenile SLE nephritis are at high risk of relapsing during adulthood raising the issue of duration of immunosuppressive treatment.

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SAT0450 ESTIMATING DURATION OF RESPONSE IN SYSTEMIC LUPUS ERYTHEMATOSUS TRIALS

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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 patients was 16.4–24.8 weeks. After adjusting for baseline SLEDAI score, patients with lower anti-dsDNA titers 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.

Responder Index	Landmark response rate at 52 weeks	Probability in response at 52 weeks from MSM (95% CI)	Mean sojourn time in response (95% CI)	Mean total time in response (95% CI)
SRI-4	45.9% (95% CI: 42.3%–49.5%)	61.0% (95% CI: 58.3%–63.5%)	31.5 weeks (95% CI: 28.5–34.8)	24.8 weeks (95% CI: 23.7–25.9)
SRI-5	33.8% (95% CI: 30.2%–37.1%)	43.8% (95% CI: 40.8%–46.3%)	25.7 weeks (95% CI: 23.1–28.7)	17.1 weeks (95% CI: 16.0–18.1)
SRI-6	32.5% (95% CI: 29.2%–36.0%)	41.7% (95% CI: 38.8%–44.7%)	24.3 weeks (95% CI: 21.9–27.1)	16.4 weeks (95% CI: 15.3–17.5)
BICLA	36.0% (95% CI: 33.4%–42.7%)	50.2% (95% CI: 47.1%–53.5%)	20.4 weeks (95% CI: 18.1–23.0)	21.2 weeks (95% CI: 19.7–22.5)

^aDrop-outs and missing data imputed as non-response. Among patients with 14 or >28 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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