

years (0–13). The median SLEDAI of the patients was 2 points (0–19). As a result of psychiatric screening, separation anxiety disorder, generalized anxiety disorder and specific phobia were significantly more common in SLE patients than in healthy controls (respectively  $p=0,054$ ,  $p=0,052$ ,  $p=0,018$ ). The Wechsler Intelligence Scale for Children-IV (WISC-IV) was applied to children and adolescents. In the test results, Perceptual Reasoning Converted Index (PRCI) and Perceptual Reasoning Standard Index (PRSI) were significantly lower in SLE patients compared to the healthy control group (respectively  $p=0,039$ ,  $p=0,046$ ). The ‘comprehension’ subtest, ‘symbol search’ subtest and Verbal Comprehension Standard Index (VCSI) were found significantly lower in SLE patients compared to the healthy control group (respectively  $p=0,046$ ,  $p=0,017$ ,  $p=0,036$ ).

**Conclusion** This study is the first in the literature about this field. It is important to show early neuropsychiatric involvement with neurocognitive and neuropsychological tests in pediatric SLE patients. We also examined the incidence of psychiatric diagnosis in SLE patients. SLE patients should be evaluated with neurocognitive and neuropsychological tests regardless of disease activity, even though they do not have neurological signs and symptoms. Patients should be closely monitored for neuropsychiatric involvement and tests should be repeated if necessary.

**Keywords** Systemic Lupus Erythematosus (SLE), SLEDAI, Neuropsychiatric SLE (NPSLE), neurocognitive disorder, neuropsychological assessment, psychometric test, psychiatric screening, WISC-IV, WISC-IV subtests, clusters of WISC-IV score

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### THE CLINICAL FEATURES AND OUTCOME OF LUPUS NEPHRITIS PATIENTS PRESENTING WITH MYOCARDITIS: A SINGLE CENTER EXPERIENCE

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**Background/Objectives** Lupus Myocarditis (LM) is one of the rare, but life-threatening manifestations of SLE. The objective was to describe the clinical, laboratory and echocardiographic findings, management, and outcome of 10 Lupus Nephritis (LN) patients presented with LM.

**Patients and Methods** A retrospective analysis of data of patients presented to our tertiary care center with LN and new onset myocardial involvement from June 2017 till May 2019, was done. All patients fulfilled the SLICC criteria for the diagnosis of SLE. LM was diagnosed by a combination of new-onset cardiac symptoms and echocardiography showing myocardial involvement in the form of global/segmental hypokinesia and reduced left ventricular ejection fraction (LVEF). **Results** Regarding baseline characteristics, there were 10 patients with 8 females, with a mean age of 22.3 (15–31) years and mean duration of SLE diagnosis of 3.8 (0–12) years. 2 patients had a history of hypertension, while none of them had a previous cardiac history or DM.

Regarding the onset of LM, in 6 patients it coincided with a diagnosis of LN, in the others, LN preceded LM with a 2-year mean interval.

### Abstract P115 Table 1

1- Baseline patient characteristics (n=10)			
Age (years)	22.3 years		
Female Sex	8 (80%)		
SLE diagnosis duration	3.8 years		
History of hypertension	2 (20%)		
History of DM	0 (0%)		
Previous cardiac history	0 (0%)		
2- Clinical data			
Onset of LM	LN and LM	LN preceding LM	
	6 (60%)	4 (40%)	
Clinical presentation	Cardiogenic shock	Acute heart failure	Syncope
	4 (40%)	5 (50%)	1 (10%)
3- Laboratory and renal biopsy data			
CBC	HB level	8.7 gm/dl	
	Wbcs count	6.1 10 <sup>3</sup> /μl	
	Platelets count	199 10 <sup>3</sup> /μl	
Creatinine level	2.6 mg/dl		
Protein: Creatinine ratio	3.6 mg/mg		
Patients with renal biopsy	5 (50%)		
	Class III	Class IV	
	1 (20%)	4 (80%)	
4- Baseline Echocardiography			
LVEF	26.3%		
ESD	46.1 mm		
EDD	56.3 mm		
Global hypokinesia	9 (90%)		
Mitral regurgitation	10 (100%)		
Tricuspid regurgitation	8 (80%)		
Pericardial effusion	7 (70%)		
Pulmonary hypertension	4 (40%)		
5- Management			
Pulse steroids	10 (100%)		
IV cyclophosphamide	7 (70%)		
mycophenolate mofetil	3 (30%)		
Immunoabsorption	1 (10%)		
6- Follow-up echocardiography after 3 months			
LVEF	42.8%		
ESD	38.9 mm		
EDD	52 mm		
7- Outcomes			
Complete recovery	4 (40%)		
Partial improvement	2 (20%)		
Mortality	3 (30%)		

Data is represented as mean or number (%)

Regarding clinical presentation, 4 patients presented with cardiogenic shock, while 5 had symptoms of acute heart failure and 1 had repeated syncope.

Regarding laboratory and renal biopsy data, mean creatinine was 2.6 (0.3–9.6) mg/dl, Protein: Creatinine ratio was 3.6 (1–14) mg/mg. 5 patients had a renal biopsy, 1 had class III LN, 4 had class IV LN, 2 of them progressed to end-stage renal disease (ESRD) and were on maintenance hemodialysis (MHD) at time of presentation with myocarditis.

Baseline Echocardiography showed mean LVEF of 26.3 (15–40)%, LV end-diastolic dimension (LVEDD) of 56.3 (46–

62) mm, and LV end-systolic dimensions (LVESD) of 46.1 (30–55) mm.

Regarding management, all patients received pulse steroids, 7 were treated with IV cyclophosphamide, 3 started mycophenolate mofetil, and 1 patient underwent 3 sessions of immunoadsorption.

Follow-up echocardiography after 3 months showed a mean LVEF of 42.8 (15–66)%, LVEDD of 52 (42–62) mm, and LVESD of 38.9 (22–55) mm.

Regarding outcomes, 4 patients had a complete recovery and normalization of LVEF, all of them were females, 3 presented with cardiogenic shock and 1 with acute heart failure, with prompt initiation of immunosuppressive therapy. 2 patients became asymptomatic but achieved partial LVEF improvement. Mean duration to LVEF improvement in all patients was 2.7 (1–4) weeks. 4 patients failed to respond to treatment, three of them died within 3-months of LM diagnosis, including the 2 males and those on MHD, all 3 had biopsy-proven class IV LN.

**Conclusion** It is crucial to recognize LM in the setting of acute heart failure & cardiogenic shock in SLE patients, as early diagnosis and prompt treatment with pulse steroids and other immunosuppressive drugs may achieve complete remission of myocarditis. Male gender with class IV LN biopsy proven or on MHD may be useful as predictors of poor prognosis.

#### P116 OUTCOMES AND SAFETY OF RITUXIMAB USE IN SYSTEMIC LUPUS ERYTHEMATOSUS – A SINGLE-CENTRE ANALYSIS

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**Background/Purpose** Rituximab (RTX) has been used worldwide in moderate to severe Systemic Lupus Erythematosus (SLE), despite failure in clinical trials. We reviewed our centre's experience in efficacy, tolerability and safety of RTX in SLE patients.

**Methods** Retrospective single-centre (35 year long, 700 SLE patient cohort) review of records of all SLE adults treated with RTX from 2009 until September 2019. Outcomes were based on physician's assessment, SLEDAI variation, drug reactions, infections, neutrophil count, immunoglobulin and B-cell count.

**Results** 45 patients (6,4% of total) were treated with RTX, 38 had sufficient data for analysis. Thirty (93,8%) female, mean diagnosis age 30,5 years, mean disease duration at first RTX of 123,1 months ( $\pm$  119). Five patients had more than one induction and 11 patients had maintenance doses - total 63 administrations of RTX. Induction regimen was mainly 1 g 15 days apart. The main indications for treatment were lupus nephritis (n=12), arthritis (n=7) and skin involvement (n=6). Mean pre-treatment SLEDAI was  $9,86 \pm 6,4$  points. Most patients had a favorable response (84,2%, n=32) with a mean SLEDAI reduction of 7,2 points ( $\pm$  5,2). B-cell depletion at 3 or 6 months

(52,9%) was more frequent in responders (p=0,003), but 8 non-depleters also responded. Non-responders had lower C3 and hemoglobin pre-RTX (p<0,05). Hypersensitivity reactions occurred 3 times (during the first cycle), 1 requiring adrenaline. One patient had a late-onset allergic reaction. Other adverse outcomes included infection requiring hospitalization (7,9%, n=5), non-serious infection (6,4%, n=4), non-severe neutropenia (3,2%, n=2), acute heart failure (1,6%, n=1) and death due to serious infection (1,6%, n=1). There were 2 cases (3,2%) of IgG hypogammaglobulinemia.

**Conclusion** Our centre has a higher use of RTX than that reported in the literature. Success rate for RTX is high in our cohort with very few serious adverse events.

#### P117 RITUXIMAB THERAPY FOR PRIMARY SJÖGREN'S SYNDROME – A RETROSPECTIVE SINGLE-CENTRE STUDY

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**Background** The rationale for B cell depletion therapy with rituximab (RTX) in primary Sjögren's syndrome (pSS) relies upon the well-established role of B cell hyperactivity in immunopathogenesis. We reviewed our centre's experience in efficacy, tolerability and safety of RTX in pSS patients.

**Methods** Retrospective single-centre (35 years long, 115 pSS patients cohort) observational study of RTX use in pSS adults from 2006 until September 2019, based on medical records, with data concerning indication and duration of treatment. Outcomes were assessed by subjective physician's perspective, ESSDAI variation, drug reactions, infections, neutrophil count, immunoglobulin and B-cell count. ESSDAI scores were calculated for pre and post whenever possible.

**Results** Ten female pSS patients were treated with RTX, with an average diagnosis age of 50,7 years and an average follow-up time of 5,6 years. Indications for RTX were: 3 peripheral nervous system (NS) manifestations, 3 central NS manifestations, 1 vasculitis, 1 vasculitis, central NS and macrophagic activation syndrome, 1 disabling musculoskeletal manifestations and 1 interstitial lung disease. Six patients became asymptomatic (4 of them with CD19 depletion), 2 did not experienced any benefit (1 with CD19 depletion) and 2 had symptomatic improvement (1 with CD19 depletion). Two patients had severe adverse reactions to rituximab (anaphylactic reaction and sweet syndrome). Although they needed hospital admission, they were able to recover completely. Three patients developed serum sickness. There were no cases of hypogammaglobulinemia or neutropenia after the treatment.

**Conclusions** Despite of the scarcity of studies validating its use, RTX can be considered for severe or refractory pSS involvement, with a reasonable safety profile.