

cases. Infection and neurological and cardiovascular diseases were the most frequent causes of death.

Conclusion LN seems to be severe in our study, with a predominance of proliferative forms, severe renal manifestations, and poor renal and overall survival.

PS8:164 **COMPARISON OF CLINICAL AND SEROLOGICAL FEATURES OF JUVENILE AND ADULT-ONSET NEUROPSYCHIATRIC LUPUS IN SPANISH PATIENTS**

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Background Neuropsychiatric (NP) manifestations are a main cause of morbidity and mortality in juvenile-onset systemic lupus erythematosus (jSLE). Some studies suggest that they are more frequent and severe in jSLE than in adult-onset SLE (aSLE).

Objectives To compare clinical and serological profiles of paediatric and adult patients with neuropsychiatric lupus (NPSLE) treated in a Spanish tertiary centre.

Methods We performed a retrospective study of patients with jSLE (onset age: 0–18 y) and aSLE (onset age: >18 y) seen in our centre during the period 1988–2016. ACR's case definitions were used to identify NPSLE manifestations. Demographics, clinical and serological data were obtained from their medical records.

Results 69 patients with NPSLE were included (aSLE 41, jSLE 28), most of them Caucasians (92%). Mean age at diagnosis was 36.4 y (range: 19–68) in adults and 13.9 y (range: 8–18) in children. The proportion of males was higher in the latter group. Mean disease duration was significantly greater in adults, as well as time from SLE diagnosis to NP manifestation onset, although without significant difference (comparison of groups is presented in the table). Central NP manifestations were more frequent (aSLE 93%, jSLE 96%) than peripheral manifestations (aSLE 12%, jSLE 11%). The most common manifestations in aSLE were headache (29%), cerebrovascular

disease (27%) and seizures (17%), whereas in jSLE were seizures (46%), headache (29%) and mood disorder/depression (25%). A significant group of patients presented 2 or more central manifestations during follow-up (aSLE 32%, jSLE 41%); mean number of manifestations was 1.36 (range: 1–3) in adults and 1.44 (range: 1–4) in children. jSLE patients with developed lupus nephritis more frequently, as well as higher anti-DNA antibodies titres, increased erythrocytation rate (ESR) and hypocomplementemia. Mortality occurred in 2 cases of aSLE and 2 jSLE.

Conclusions Our results corroborate that juvenile patients with NPSLE present higher disease activity compared to adults. There was no significant difference in time from SLE diagnosis to NP manifestation onset, but tended to be shorter in jSLE. The spectrum of NPSLE was varied both groups and an important proportion developed 2 or more manifestations. Mortality continues to be important in NPSLE in both age groups.

PS8:165 **NEUROPSYCHIATRIC DAMAGE IN DECEASED PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS**

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Purpose Neuropsychiatric damage (ND) is a major determinant of morbidity in SLE. We analysed ND in a group of deceased SLE patients and identified features associated with ND.

Methods We retrospectively analysed 90 patients (68 females) deceased during 2002–2011. All patients fulfilled at least 4 classification criteria of the ACR. We identified patients with ND, as defined by the SLICC/ACR damage index, and its components: cognitive impairment/major psychosis (CIMP), seizures, cerebrovascular accident (CA), cranial/peripheral neuropathy and transverse myelitis (TM). Following variables were compared between patients with and without ND (ND + and ND, respectively): demographics, ACR criteria at diagnosis and cumulatively at death (available at diagnosis for

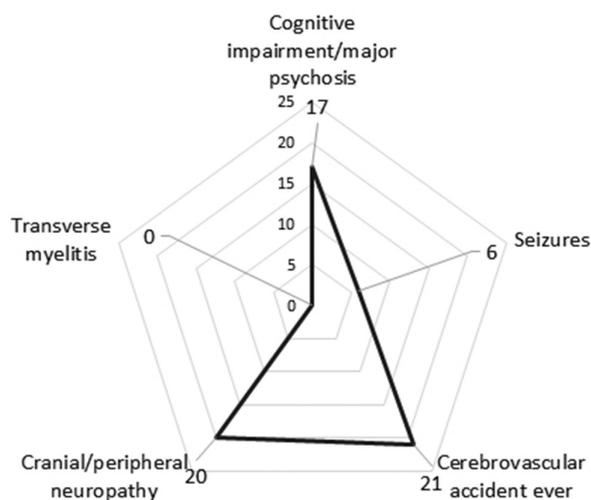
Abstract PS8:164 Table 1

	Juvenile NPSLE	Adult NPSLE	p-value
N° of patients	28 (41%)	41 (59%)	-
Women:men	20:8	39:2	0.0060*
Time of disease (months)	19.8	232.5	0.0001*
NP manifestations at onset	7 (25%)	11 (27%)	0.8651
Time from diagnosis to NP manifestation (months)	42.4	87.1	0.1268
Lupus nephritis	16 (57%)	9 (22%)	0.0028*
Antiphospholipid syndrome	5 (18%)	10 (24%)	0.5182
ANA ≥ 1/1280	8 (29%)	11 (27%)	0.8736
Anti-DNA ab (IU/ml)	178.9	39.4	0.0005*
Anti-Ro/SSA ab	10 (36%)	17 (41%)	0.6308
Anticardiolipin ab	4 (14%)	10 (24%)	0.3054
Anti-β2 glycoprotein I ab	5 (18%)	7 (17%)	0.9328
Lupus anticoagulant	8 (29%)	10 (24%)	0.6977
Cryoglobulins	6 (21%)	3 (7%)	0.0874
ESR (mm/h)	53.8	35.7	0.0199*
CRP mg/l	4.6	4.7	0.9687
C3 low (< 80 mg/dl)	22 (79%)	16 (39%)	0.0012*
C4 low (< 16 mg/dl)	22 (79%)	13 (32%)	0.0001*

73 patients), damage and its components one year following diagnosis and at death, disease activity (ECLAM) at diagnosis, components of the metabolic syndrome, smoking and immunosuppressive treatment. Frequencies were compared using the chi-square and Fisher's exact test, and continuous variables using the t-test and Mann-Whitney U-test. Variables associated with ND were included in a multivariate logistic regression model.

Results We identified 44/90 ND +patients: 17/44 with CIMP, 6/44 with seizures, 21/44 with CA, 20/44 with neuropathy, none with TM. ND +patients had a higher cumulative count of ACR criteria compared to their ND- counterparts (6.02 ± 1.23 vs 5.39 ± 1.26). They had a higher proportion of neurologic disorder (NRL-D) (6/37 vs 0/36) and a lower proportion of serositis (4/37 vs 11/36) at diagnosis, as well as higher cumulative proportions of NRL-D (9/44 vs 2/46), hematologic disorder (41/44 vs 34/46) and lymphopenia (34/44 vs 24/46) ($p < 0.05$). ND +patients also had higher cumulative damage (6.43 ± 3.13 vs 3.43 ± 2.54), higher cumulative proportions of pulmonary (14/44 vs 6/46) and musculoskeletal damage (MSKD) (32/44 vs 21/46), and higher proportions of arterial hypertension (40/44 vs 34/46), secondary sicca (17/44 vs 9/46) and Hughes syndrome (11/44 vs 4/46) ($p < 0.05$). Serositis at diagnosis and cumulative MSKD were associated with ND in the multivariate model (OR 0.17 (95% CI: 0.03 to 0.89) and 6.00 (95% CI: 1.64 to 21.91), respectively).

Conclusions Serositis may be associated with a lower likelihood of ND, while NRL-D was present at diagnosis only in patients that accrued ND. The association between MSKD and ND requires further elucidation.



Abstract PS8:165 Figure 1

PS8:166 COMPARISON OF CLINICAL AND SEROLOGICAL CHARACTERISTICS BETWEEN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS WITH AND WITHOUT ASSOCIATED JACCOUD'S ARTHROPATHY

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Objectives To compare the profile of clinical and serological manifestations of patients with SLE with and without JA followed in a tertiary care hospital of Madrid.

Methods We performed a retrospective observational study of a cohort of patients diagnosed with SLE (4 or more ACR criteria) between June 1977 and December 2015. The variables evaluated included demographics, clinical, analytical and radiological manifestations. The definition of JA was based on the presence of clinical criteria (reversible joint deformities) and absence of radiographic erosions typical of rheumatoid arthritis.

Results We included 108 patients representing a sample of 24% of the total number of patients with SLE treated at our centre during that period. The majority of patients were women (89.8%), mean age at diagnosis was 30 ± 12.29 years (range: 7–75) and duration of disease was 127 months (range: 2–411). Thirteen patients (12.03%) had findings compatible with JA. There were no significant differences in age, sex or race, but the duration of disease was higher in JA patients (190 vs 118.2 months, $p = 0.0299$). There were significant differences in the presence of malar rash ($p = 0.0009$), photosensitivity ($p = 0.0050$), oral ulcers ($p = 0.0032$) and pericarditis ($p = 0.000001$), which were more frequent in patients without JA, but arthritis, nephritis, pleuritis, seizures, psychosis, Raynaud's phenomenon and antiphospholipid syndrome had a similar distribution between both groups. Among the immunological features, no significant difference was found in relation to hemolytic anaemia, lymphopenia, thrombocytopenia, ANA, anti-ENA, anti-DNA, anticardiolipin, anti- $\beta 2$ glycoprotein I and lupus anticoagulant, but leukopenia was also more frequent in patients without JA ($p = 0.0041$).

Conclusions In the analysed sample of patients in our centre JA was a relatively frequent finding and was associated with a longer duration of the disease. It was not possible to corroborate other JA associations suggested in previous studies such as a lower frequency of lupus nephritis or major secondary antiphospholipid syndrome, probably due to limited sample size, but there are also other studies that do not demonstrate significant differences in relation to clinical and serological findings in patients with SLE with JA with respect to those who do not present it.

PS8:167 THERAPEUTIC EFFICACY OF BELIMUMAB IN ADDITION TO STANDARD THERAPY FOR LUPUS NEPHRITIS AND NEUROPSYCHIATRIC LUPUS – CASE SERIES OF ROUTINELY COLLECTED DATA AT A SINGLE CENTRE

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Purpose Belimumab (BEL) is approved for Systemic Lupus erythematosus in addition to standard immunosuppressive therapies. Clinical studies have excluded patients with particular organ manifestations from participation in most clinical trials. Most importantly, Lupus nephritis (LN) and neuropsychiatric Systemic Lupus erythematosus (NPSLE) were exclusion criteria in the relevant clinical studies. We aim to report our experience of BEL's effect on SLE manifestations which have not been formally addressed in clinical trials.

Methods We performed an observational study of routinely collected clinical data of all patients receiving BEL with or without other immunosuppressive therapy at our institution.