The majority are female (85%). The average age is 15±2.5 years. This cohort includes Hispanic (36%), Asian (29%), African-American (21%) and Caucasian (11%) children. LN class III or IV was diagnosed in 67% of patients. Nearly all were treated with hydroxychloroquine and steroids (95%). Other drugs used include mycophenolate mofetil (85%), cyclophosphamide (43%), rituximab (27%) and tacrolimus (2%).

A significant increase in urinary HER2, TWEAK and VCAM1 levels was found in LN patients (p=0.005; p=0.006; p=0.01, respectively) when compared to controls. HER2 levels reflected disease activity, increasing during flares.

In an adult cohort of LN patients (N=126) composed mainly of females (80%) with an average age of 46±13 years, we also found a significant increase of urinary HER2 when compared to controls (p=0.002).

A strong correlation between the urinary levels of HER2, TWEAK and VCAM1 was not found.

Conclusions: Urinary HER2, TWEAK and VCAM1 were significantly increased in a paediatric cohort of LN patients. In addition, significantly higher urinary HER2 levels were also found in an adult LN cohort. This on-going study will further evaluate if these urinary markers, alone or in combination, can reflect disease activity and predict renal flares, analysing their potential value in clinical practice.

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Disclosure of Interest: None declared

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THU0491 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 the clinical and serological profile of pediatric and adult patients with neuropsychiatric lupus (NPSLE) treated in a Spanish tertiary center.

Methods: A retrospective study of patients with jSLE (age of onset: 0–18y) and aSLE (age of onset: >18y) seen in our center during the period 1988–2016 was performed. Case definitions of the American College of Rheumatology were used to identify NPSLE manifestations. Demographics, clinical and serological data were obtained through a review of their medical records.

Results: A total of 69 patients with NPSLE were included, aSLE 41 (59%) and jSLE 28 (41%), the comparison of groups is presented in the table. Most of them were Caucasian (92%), mean age at diagnosis in adults was 36.4 years (range: 19–68) and 13.9 years (range: 8–18) in children. The proportion of males was higher in the latter group. The mean duration of the disease was significantly greater in adults, as well as the time from SLE diagnosis to NP manifestation onset, although without significant difference. Central NP manifestations were the most frequent in both groups (aSLE 93%, jSLE 96%) regarding to the peripheral manifestations (aSLE 12%, jSLE 11%). The most frequent manifestations in aSLE were headache (29%), cerebrovascular disease (27%), seizures (17%) and myelopathy (15%), whereas in jSLE were seizures (46%), headache (29%), mood disorder/depression (25%), psychosis (18%) and autonomic disorders (18%). A significant group of patients presented ≥2 central manifestation during their evolution (aSLE 32%, jSLE 41%), with the mean number of manifestations in adults being 1.36 (range: 1–3) and in children 1.44 Range: 1–4). Patients with jSLE developed lupus nephritis (LN) with a significantly higher frequency, as well as higher titres of anti-DNA antibodies, erythrocyte sedimentation rate (ESR) and hypocomplementemia. During the study period there was mortality in 2 cases of aSLE and 2 jSLE (5% and 7%, respectively).

	Juvenile NPSLE	Adult NPSLE	p-value
No. 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-ß2 glycopotein 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*

Conclusions: Our results corroborate that juvenile patients with NPSLE present higher disease activity compared to adults. There was no significant diference in the 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 manifestation. Mortality continues to be important in NPSLE in both age groups.

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THU0492 MACROPHAGE ACTIVATION SYNDROME AS THE INITIAL MANIFESTATION OF JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Macrophage activation syndrome (MAS) is a severe, potentially life-threatening complication of autoimmune diseases in children. Little is known about the association between MAS and the onset of juvenile systemic lupus erythematosus (jSLE).

Objectives: The aim of this study was to determine the frequency and clinical features of MAS as the initial complication of jSLE.

Methods: During 2004 and 2016, we retrospectively reviewed the clinical and laboratory features of 46 jSLE patients diagnosed at the Saitama Children's Medical Center. Patients who were complicated with MAS at the same time as jSLE. These patients were compared with a control group composed of jSLE patients without MAS. The MAS was diagnosed according to preliminary guidelines.

Results: Fifteen patients (32.6%) developed MAS during the initial stage of jSLE. Fever, leukopenia, thrombocytopenia, hyperferritinemia hypofibrinogenemia, increased aspartate aminotransferase (AST), and increased lactate dehydrogenase (LDH) were more frequently observed in patients having jSLE with MAS than in those having jSLE without MAS. No differences were observed in serum C3 and C4 levels, or erythrocyte sedimentation rate (ESR) ($P<0.05$). Especially, Seven patients (46.7%) had *neurologic* symptoms that were significantly higher in those with MAS ($P<0.01$). All patients received corticosteroids when jSLE with MAS diagnosis was established, among whom seven received pulse methylprednisolone therapy. Two patients were treated with IVIG. Nine patients with MAS were treated with immunosuppressants, including cyclophosphamide and mycophenolate mofetil, *azathioprine*. No patient involved in this study died.

Conclusions: MAS can be the initial manifestation of jSLE. MAS may be an underrecognized complication of jSLE. MAS with jSLE should be suspected in patients with high fever, cytopenia, and a liver disorder. In addition, we found that in jSLE with MAS patients, they had more neurologic symptoms compared to jSLE without MAS. Early diagnosis and intensive therapy is essential to improve the clinical outcome.

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THU0493 STUDY OF LONG-TERM OUTCOME OF CHILDREN WITH JUVENILE DERMATOMYOSITIS FROM A SINGLE-CENTRE IN NORTH INDIA

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Background: Juvenile dermatomyositis (JDM) is a rare inflammatory myopathy seen in children. There have been few studies on long-term outcome in children with JDM

Objectives: To assess long-term outcome of JDM using validated measures of outcome

Methods: All children diagnosed to have JDM for more than 2 years and registered in Pediatric Rheumatology Clinic at PGIMER, Chandigarh, India, were deemed eligible for recruitment. Study period was from January 1, 2015 to June 30, 2016. Those who were not on regular follow-up were called for assessment which was done by a single observer using Childhood Myositis Assessment Scale (CMAS), Manual Muscle Testing 8 (MMT8), Myositis Disease Activity Assessment Tool (MDAAT), abbreviated cutaneous assessment tool (aCAT), Myositis Damage Index (MDI) and Childhood Health Assessment Questionnaire (CHAQ).

Results: Thirty-five children were enrolled in this study, 22 (62.9%) were on regular follow-up. Mean age was 13.9yrs (range 4–29yrs). Mean age at diagnosis was 7.51yrs with median interval between onset of symptoms and diagnosis

being 5 months. Mean duration of disease at the time of enrolment was 7.18yrs. Disease course was monocyclic in 24 (68.6%). Muscle strength was normal in 71.4%. Severe involvement defined as MMT8 score below 64 was seen in 8.6%. Cutaneous activity was determined by aCAT with 40% children having some form of cutaneous activity. Based on MYOACT, 31.4% children had evidence of disease activity at the time of cross-sectional assessment with skin being the commonest organ system involved in 28.6% followed by muscles in 22.9%. Twenty-one (60%) children had some form of cutaneous damage. Calcinosis in 12 (34.3%) and lipodystrophy in 8 (22.9%). Twenty four subjects had an MDI score of $\geq$1 suggesting damage in at least one organ system. Most commonly affected organs were skin, endocrine and muscles in 20, 12 and 9 subjects respectively. Nine (25.7%) subjects in our study had some form of a physical dysfunction suggested by a CHAQ score above 0. Previous studies on long-term outcomes in children with JDM have either not used validated outcome measures or have used fewer measures [1–3].

Conclusions: Highlight of our study is the use of validated outcome measures for evaluation of long-term outcomes. After mean disease duration of 7.18 yrs, 1/3rd subjects had evidence of disease activity with almost 1/10th having moderate to severe activity. About 2/3rd had damage in at least one organ system. Skin was the most common organ affected by activity as well as damage. About 1/4th had reduced physical functioning. Thus, JDM is not a disease where one time treatment would suffice and regular long-term follow-up is required. Counselling of the caregivers is also critical for them to adhere to follow-up. Larger long-term studies using validated outcome measures are required to confirm these findings.

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THU0494 CLINICAL FEATURES OF CHILDREN WITH KAWASAKI DISEASE IN DIFFERENT AGE GROUPS IN SOUTHWEST CHINA

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Background: Kawasaki disease (KD) is a systemic vasculitis characterized by acute and prolonged fever. The prevalence of coronary artery abnormality (CAA) is as high as 11%. The young infants usually have the increased risk of CAA, but do not have the typical clinical manefestations of KD.

Objectives: To explore clinical features of children with KD in different age groups to improve the prognosis of KD.

Methods: A total of 218 children with kawasaki disease were divided into the infants group, the toddlers' group, the pre-school age group and the school age group. Retrospective analysis of clinical data were performed among the groups. Categorical data were compared with each other statistically by Chi-square analysis. Statistical significant was defined as $P < 0.05$. Due to the insufficient cases of school age group and five cases of patients with entire clinical data, the analysis was focused on the other three groups and excluded the five cases in the following statistical analysis.

Results: (1) Among the 218 KD patients, the male to female ratio was 1.5:1 and the recurrence rate was 1.8%. Seven cases (3.2%) were diagnosed as atypical KD, and 84 (38.5%) patients accepted intravenous gamma globulin (IVIG) treatment after the sixth day of KD onset. The incidence of IVIG-resistent KD was 8.7% and the rate of coronary dilation was 11.5%. (2) Fever was the most common clinical feature (100%). The bilateral bulbar conjunctiva injection and the change in mucosa of oropharynx were 85.4% and 81.2% respectively. Moreover, cough (40.5%), diarrhea (16.9%) and vomiting (8.5%) were also very common in the present KD patients. (3) Patients from the toddlers' group were more common to develop lymphadenopathy and skin rash (χ^2=7.784, P=0.02; χ^2=10.794, P=0.005), but were less frequently to be documented with cough and diarrhea (χ^2=7.334, P=0.026; χ^2=18.447, P=0.000). (4) The incidence of increased platelets was more common in the infants group (χ^2=7.552, P=0.023). Comparing with the urine test among three groups, the toddlers' group had a higher incidence of sterile pyuria (χ^2=10.653, P=0.005), and infants younger than 12 months old had a lower incidence of proteinuria and positive urine ketone (χ^2=15.507, P=0.000; χ^2=40.336, P=0.000).

Conclusions: The respiratory tract, the digestive and urinary systems are involved commonly in Kawasaki disease, and patients from different age groups showed different clinical features, which should be pay more attention to promote the prognosis.

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THU0495 EFFECTIVENESS OF CHILDHOOD VACCINATIONS IN CAPS PATIENTS TREATED WITH CANAKINUMAB: RESULTS FROM AN OPEN-LABEL PHASE 3 EXTENSION STUDY

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Background: Canakinumab (CAN) has been shown not to impair antibody production following vaccination in children in an open-label phase 3 study (NCT01302860).[1] Here we present the results from the extension of this study.

Objectives: To evaluate the presence of protective antibody levels following immunisation with inactivated vaccines in CAPS patients during extension study.

Methods: Patients who completed the core study were allowed to continue into the extension study on the standard dosing regimen of 2 mg/kg subcutaneous CAN every 8 weeks or on last dose/dosing regimen received in the core study. Vaccination response was evaluated using post-vaccination antibody titres at 4 and 8 weeks after immunisation. Patients were considered assessable for an antibody response to a specific vaccination if they had a measurement of antibody titre 0–14 days post-vaccination (pre-vaccination assessment) and at least 1 subsequent measurement of antibody titre at 4 weeks and/or 8 weeks post-vaccination. However, for patients with adequate pre-dose antibody titres and maintained during the trial, the specific patient vaccination was deemed non-assessable.

Results: During the extension phase, of 17 patients ($\leq$6 years), 4 received 8 types of vaccinations against *Corynebacterium diphtheriae*, *Bordetella pertussis*, *Neisseria meningitidis*, *Clostridium tetani*, influenza type A and type B, *Haemophilus influenzae* B, *Streptococcus pneumoniae*, or hepatitis B. Of 20 unique patient-vaccination cases, 17 were assessable for a vaccination response, whereas for the remaining 3, pre-dose antibody titre was not available. For 16 (94.1%) assessable cases, post-vaccination antibody titres increased above protective levels. For one patient who received Tetravec formulation (diphtheria, tetanus and acellular pertussis combination), the response observed for 1 (vaccination against *Clostridium tetani*) of the 3 vaccines included in Tetravac represented optical density rather than antibody concentrations and hence considered non-evaluable. For 19/20 patient-vaccinations, including those without pre-dose antibody titres, protective levels were observed during the study, which were maintained throughout the extension study.

Conclusions: Canakinumab appeared to have no effect on post-vaccination antibody production following the administration of non-live vaccines in CAPS patients.

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THU0496 PULMONARY SYMPTOMS AS THE FIRST PRESENTATION OF KAWASAKI DISEASE IN CHILDREN

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Background: Kawasaki disease (KD) is a medium vessel vasculitis which predominantly affects children less than 5 years of age. Though principal clinical features are mucocutaneous, KD in children may have multiple systemic manifestations, including pulmonary, which may create diagnostic difficulties for the treating physician.

Objectives: We describe our experience of managing children with uncommon pulmonary presentation of KD.