

m).Of the 29 lupus patients,13 charts were retrieved. The mean age at diagnosis was 13.3 years(SD 3.4) from 6–18 years old. Oedema(53.8%), fever(46.1%), abdominal pain (38.4%) and easy fatigability (30.7%) were the most common features at disease onset while renal involvement (84.6%), malar rash (53.8%) and oral ulceration(46.1%) were common at the time of diagnosis. All of 9 patients with ANA titers were positive. Anti-dsDNA antibodies were high in 3 patients. Low complement values were seen in 83.3%. The follow-up period ranged from 0.2-2y with a mean duration of 1.2 ± 0.6 y.Four went into remission but 3 patients died, 3 patients transitioned to adult section and 3 were lost to follow-up.

Conclusions Oedema and renal involvement were the most common feature at the onset and at the time of the disease, respectively. All male lupus patients had positive ANA and low C3 results. Causes of death were:active disease, sepsis and DIC. Early recognition and diagnosis will lead to prompt institution of treatment that will benefit lupus patients.

140 ATOPY IN CHILDREN WITH JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS IS ASSOCIATED WITH SEVERE DISEASE

¹G Ruru*, ¹L Liangjing, ²C Lanfang, ²C Junjia, ²Z Yangqing. ¹Renji Hospital-School of Medicine-Shanghai Jiao Tong University, Rheumatology, Shanghai, China; ²Renji Hospital-School of Medicine-Shanghai Jiao Tong University, Pediatric, Shanghai, China

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Background and Aims We aimed to assess the influence of co-existing atopy on the prognosis of juvenile systemic lupus erythematosus (JSLE)

Methods Patients diagnosed with JSLE between October 2005 and April 2016 were enrolled in a prospective cohort study and followed for 2 years. Management of patients was evaluated using SLEDAI-2K score. Eighty JSLE patients were enrolled at diagnosis and were divided into those with atopy and those without.

Results Atopic patients had significantly higher SLEDAI-2K at disease onset (16.09 *vs.* 11.18), higher anti-double-stranded DNA (66.58 *vs.* 44.55 IU/ml), higher erythrocyte sedimentation rate (52.89 *vs.* 38.27 mm/h), higher percentage of total B-cells (25.85 *vs.* 19.51%), lower percentage (7.26 *vs.* 9.03%) and activity (9.92 *vs.* 11.32%) of natural killer cells, lower complement C3 (0.51 *vs.* 0.69g/L), and lower complement C4 (0.06 *vs.* 0.12g/L) ($p < 0.05$ for all comparisons). At 1 month, 3, 6, 12, 18 and 24 months, JSLE patients with atopy reached higher SLEDAI-2K and lower Δ SLEDAI-2K improvement rate (at 1 month, 8.34 *vs.* 4.71 and 43.63 *vs.* 57.95%, respectively; at 3 months, 8.57 *vs.* 2.62 and 48.39 *vs.* 75.10%, respectively; at 6 months, 6.91 *vs.* 2.38 and 53.59 *vs.* 77.26%, respectively; at 12 months, 4.71 *vs.* 1.80 and 69.54 *vs.* 84.10%, respectively; at 18 months, 4.66 *vs.* 2.02 and 68.14 *vs.* 82.93%, respectively; at 24 months, 8.57 *vs.* 2.62 and 70.00 *vs.* 81.88%, respectively; all $p < 0.05$).

Conclusions Co-existing atopy in children with JSLE may exert an adverse influence on JSLE, with atopic patients manifesting more severe disease at diagnosis and poorer outcome.

141 SOLUBLE RECEPTOR ACTIVATOR OF NUCLEAR FACTOR K B LIGAND (S RANK-L) LEVELS IN PAEDIATRIC ONSET SLE

S Sandal*, A Rawat, A Gupta, S Singh. PGIMER, Paediatrics, Chandigarh, India

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Background and aims Receptor Activator of Nuclear Factor κ B (RANK), its ligand (RANKL) and osteoprotegerin are the key mediators of bone remodelling and the final effector pathway in osteoclast development and differentiation. The data on RANKL axis in paediatric Systemic Lupus Erythematosus (SLE) is lacking. Thus we proposed to estimate serum sRANKL levels in paediatric SLE and to correlate sRANKL levels with the SLE disease activity

Methods Consecutive children with SLE attending Paediatric Rheumatology Clinic of Advanced Paediatrics Centre, PGIMER, Chandigarh were enrolled. The study group was divided into active (with ongoing disease activity) and inactive (no disease activity) subgroups based on SLE disease activity index (SLEDAI) scores. The sRANKL ligand levels were measured at enrollment using an enzyme-linked immunosorbent assay (sRANKL – ELISA MyBioSource@, USA).

Results Thirty-one children (12 boys) with a mean age of 13.4 ± 3.2 years were included. The median (interquartile range) sRANKL level of the cohort was 52.3 (24.1, 66.4) pg/mL. Serum RANKL levels were not significantly different in active and inactive disease subgroups [median (interquartile range): 55.2 (21.3, 66.4) pg/mL *versus* 53.3 (29.3, 64.9) pg/mL, respectively] ($p = 0.89$). There was no statistically significant correlation between sRANKL levels and SLEDAI scores, Spearman correlation coefficient $\rho = 0.083$, $p = 0.65$,

Conclusions There was no significant difference in sRANKL levels between the inactive and active disease group. Also there appears no correlation between sRANKL level and SLEDAI scores.

142 CLINICAL PROFILE AND LONG TERM OUTCOME OF CHILDHOOD SYSTEMIC LUPUS ERYTHEMATOSUS

S SAWHNEY*, A SHIVPURI, M AGARWAL. Sir Ganga Ram Hospital, Paediatric Rheumatology, NEW DELHI, India

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Background and aims Systematic study of all diseases is essential to understand the spectrum of the disease presentation, the severity of the disease and the outcome. There is paucity of data from India on details of paediatric SLE.

This study aims to define:

- Describe the clinical and immunological profile of SLE within six months of disease onset in three age categories
- To compare the performance of ACR 1997 criteria *vs* SLICC 2012 criteria to classify disease in first 6 months of onset
- To define the mean value of SLEDAI at presentation and over a 5 year follow up

Methods Children attending the paediatric rheumatology clinic from January 2009 to September 2016 were included and details recorded.

Abstract 142 Table 1 Age wise breakup of clinical and immunological feature at disease onset

Age distribution	Up to 6 years N=15	6-11 years N=61	11-18 years N=93
Constitutional features			
Fever	73%	85%	82%
Fatigue	13%	72%	70%
Oral ulcers	47%	60%	35%
Non scarring alopecia	60%	54%	58%
Organ system involvement			
Arthritis	67%	57%	56%
Neurological involvement	0	11%	8%
Renal involvement	27%	44%	37%
Cutaneous involvement	66.6%	72%	57%
Serositis	6.6%	14.7%	
Hematological	40%	51%	17.2%
			56%
Immunological profile			
Direct coombs test	47%	11%	41%
Low complements	60%	80%	86%
APLA	20%	28%	26%

Abstract 142 Table 2 Comparison of ACR 1997 criteria vs SLICC 2012 criteria for disease classification

No. fulfilling both ACR and SLICC criteria at onset of disease	136/169= 80.4%
No. fulfilling no criteria	9/169=5.3%
No. fulfilling SLICC criteria	160/169=94.6%
No. fulfilling SLICC criteria but missed by ACR criteria	24/169=14.2%
No. fulfilling ACR but missed by SLICC	0

Abstract 142 Table 3 SLEDAI

SLEDAI	No. of patients	Median SLEDAI
At onset(first 6 months)	169	14(0-52)
At one year	139	2(0-16)
At 2 years	100	2(0-28)
At 3 years	70	2(0-24)

Results Of 169 children, 139 (82%) females. Median age at disease onset:11.4 years(3.4–18),median age at diagnosis:12 (3.5–19).

20% had history of autoimmune disease in first degree relative.

Therapy at disease onset(first 6 months):

Hydroxychloroquine: 100%,

Glucocorticoids: 98%,

Mycophenolate: 33%,

Methotrexate: 27%,

Azathioprine: 16%,

Cyclophosphamide: 9%,

Rituximab: 2%.

At last follow up: Glucocorticoids: 37%

Median follow up 48 months(1–195 months), Mortality:4%,lost to follow up: 19%, active disease at last follow up:25%

Conclusions Patients seen at our centre had a significant disease burden with a median SLEDAI score of >20 at presentation.

Upto 1/2 of the study population did not have a malar rash. 38% had renal disease. Fever was seen in 82% and often was the cause for seeking medical opinion. This is a small data set from a tertiary level centre and not representative of the community disease.

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RITUXIMAB IN SLE MEMBRANOUS NEPHROPATHY

¹R Sinha*, ²S Akhtar, ²P Giri, ²P Pal. ¹Kolkata, India; ²Institute of Child Health, Paediatric Nephrology, Kolkata, India

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Background and Aims We hereby report three cases of biopsy proven Type 5 SLE membranous nephropathy (T5: SLEMN) which responded to Rituximab after failed first line induction agents.

Methods Retrospective chart review

Results Case 1: Rituximab was used in this 14 year old girl as she failed to standard therapy of cyclophosphamide (750 mg/m²) and high dose corticosteroid along with angiotensin convertase inhibitor (ACEI). 30 days post rituximab 24 hour proteinuria dropped (6790 to 876 mg) and albumin rose (1.8 mg/dl to 3 mg/dl). She is in remission at 14 month on low dose steroid and mycophenolate mofetil (MMF).

Case 2: 10 year old girl whose presentation was similar to Case 1 and failed to show any significant improvement to standard therapy. 45 days post rituximab, 24 hour proteinuria dropped (4900 to 690 mg) and albumin rose (1.9 mg/dl to 3.3 mg/dl). At 12 month she is in remission on low dose steroid and MMF.

Case 3: 12 year old girl presented with features of nephrotic syndrome. At 30 days follow up there were no improvement despite standard therapy and she also started to have neuropsychiatric manifestation. 60 days post rituximab 24 hour proteinuria dropped (3548 to 300 mg) and albumin rose (1.8 mg/dl to 3.7 mg/dl). Her neuro-psychiatric manifestation also improved.

Conclusions In all 3 cases 2 doses of rituximab at 375 mg/m² each achieved CD 19 count of zero (which normalised by 14, 9 and 11 month respectively) along with significant drop in proteinuria.

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ROLE OF TUBULOINTERSTITIAL LESIONS IN PREDICTING RENAL OUTCOME AMONG PAEDIATRIC ONSET LUPUS NEPHRITIS – A RETROSPECTIVE COHORT STUDY

¹CY Wu*, ²HP Chien, ³HY Yang, ⁴MH Tseng, ¹KW Yeh, ¹JL Huang. ¹Chang-Gung Memorial Hospital- Linko branch, Division of Allergy- Asthma- and Rheumatology- Department of Paediatrics, Taoyuan city, Taiwan R.O.C; ²Chang-Gung Memorial Hospital- Linko branch, Department of Pathology, Taoyuan city, Taiwan R.O.C; ³Chang-Gung Memorial Hospital- Linko branch, Department of Nephrology, Taoyuan city, Taiwan R.O.C; ⁴Chang-Gung Memorial Hospital- Linko branch, Division of Paediatric Nephrology- Department of Paediatrics, Taoyuan city, Taiwan R.O.C

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Background and aims Raising evidence supported a prognostic utility of tubulointerstitial lesions in lupus nephritis (LN). The exact prevalence of tubulointerstitial abnormalities and its predictive value among paediatric onset systemic lupus erythematosus (pSLE) cases, however, remained unknown.

Methods Sixty-seven pSLE subjects diagnosed with LN with initial renal samples available were enrolled and followed for an average of 6.43±3.06 years. Renal histology was evaluated according to the International Society of Nephrology/Renal Pathology Society classification, National Institute of Health classification and tubulointerstitial activity index (TIAI).

Results Tubulointerstitial injuries were observed in 38.81% of all LN cases, including 13.33% with non-proliferative lupus nephritis (nPLN) and 46.15% of with proliferative lupus nephritis (PLN). Tubulointerstitial injuries occurred solitarily in cases with nPLN(13.33%), but always associated glomerular changes and significantly impacted renal survival (p=0.032) among those with PLN. TIAI associated glomerular abnormalities (p=0.031) but did not correlate renal performance or subsequent outcome (p=0.445). Among the chronicity index, it was the chronic tubulointerstitial lesions which provided prognostic information (p=0.012). We observed a synergistic effect of all tubulointerstitial abnormalities rather than an individual factor attributed the prognostic utility (p=0.025 vs. p=0.083, 0.055, 0.354). Finally, considering tubulointerstitial injuries in PLN further discriminated subsequent renal outcome (p=0.006).

Conclusions The prevalence and clinical significance of tubulointerstitial abnormalities were similar among the pSLE and the adult population. With its importance in identifying those at risk of renal failure, histologic classification considering tubulointerstitial lesions may potentially assist outcome prediction.

Patient-submitted abstracts

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A PATIENT'S FOUR DECADE JOURNEY TO WELLNESS : A MODEL OF CARE FOR LIVING WELL WITH LUPUS

M Erceg*. ¹Perth, Australia

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Background and aims Mary Erceg is a former teacher and senior public servant who has lived with systemic lupus erythematosus for over 40 years since initial diagnosis.

This presentation explores her personal journey through initial diagnosis; medications; flares; acute relapses; and treatment errors which resulted in 6 days in a coma, four months