She had altered behaviour with agitation, disorientation, fluctuating consciousness, hallucinations, and altered sleep. On examination, she had malar rash, icterus, and hepatosplenomegaly. She also had catatonia, mutism, would stare intermittently, had low speech output and psychomotor retardation with rigidity. There was no focal deficit. Investigations revealed pancytopenia, transaminitis, conjugated hyperbilirubinemia, normal renal functions, antinuclear antibody (ANA) - positive (homogenous pattern), high anti-dsDNA with hypocomplementemia.Liver biopsy revealed steatosis with hepatitis. Screen for infections was negative, except CMV. Very high levels of CMV DNA in blood were noted on PCR. It was a clinical dilemma as to whether CMV was causative, co-infection or a re-activation due to immunosuppression. Magnetic resonance imaging (MRI) brain showed cortical atrophy. There was no evidence of any vascular involvement.

She was treated with intravenous (IV) methylprednisolone, IV cyclophosphamide pulses and oral valganciclovir.

Results A repeat CMV viral load done after six weeks of oral valganciclovir therapy was undetectable. She has been followed up for a period of 6 months. She has shown marked improvement in her neurological status and transaminases have normalised.

Conclusions CMV is an important pathogen in patients with SLE; however its exact pathogenesis needs more research.

137 A CASE OF CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME WAS CONTROLLED WITH RITUXIMAB

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Background and Aims CAPLS is a multiple thrombi of small vessels affecting viscera. Patients had previous evidence of APLS. CAPLS was precipitated by infection, invasive procedures. The clinical presentation was involving the cardiopulmonary, CNS, abdomem. Thrombocytopenia and hemolytic anaemia were frequent. Anticardiolipin antibodies were present in almost. The outcome can be disastrous.

Methods Case: 10-year-girl had been developed petechiae on Aug 10, 2009. Patelet 2000, Hb 12.0. The diagnosis was ITP and she received the IVIG. But anaemia was developed on Aug 31, 2009. Platelet 48,000. Hb 8.9. The diagnosis was Acquired hemolytic anaemia. On Sept 21, 2009 the Hb 10.0, reticulocyte 11.0% and Cooms' (+), FANA (++), anti-dsDNA (-). The diagnosis was Evans' syndrome. The following lab were anti-cardiolipin IgG (+), IgM (+), IgA (-), Lupus anticoagulant (+), anti-beta2 GPI IgG (+), IgM (+) on Oct 14, 2009. The final diagnosis was SLE with antiphosolipid syndrome.

Gangrene affecting the right 5th toe on Jan 2014 and gangrened toe was autoamputated on Dec 2014. CT angiogram was performed on Apr 2014. There was superficial occusion on right femoral vein. On July 2014 the disease was flared. Hemolytic anaemia (Hb 7.7) and jaundice developed. Chest CT revealed SLE myocarditis and abdomen CT was bowel SLE.

Results On Sept 2014, catastrophic APS was developed. Platelet 5,000. aPTT 99.3 s. Plasma pheresis was done for 7 times, but the platelet count did not change. Rituximab was injected 4 times. Platelet was 4000 thru 99 000 on Nov 2014. After then platelet was well controlled.

Conclusions This rituximab response to therapy.

138 CALCIUM AND VITAMIN D STATUS IN LUPUS CHILDREN IN JAKARTA, INDONESIA

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Background and Aims Osteopenia and subsequent osteoporosis are complications faced by children living with lupus on steroid therapy. Calcium and vitamin D have been embedded in the management protocol early in therapy.

Aim To evaluate calcium and vitamin D level in children with lupus.

Methods Lupus patient recruited if their age 5–18 years old, on steroid therapy at least 5 month. Lupus diagnosis, follow up record and SLEDAI score were retrieved from medical record. Subjects have DXA-scanning to determine bone density and plasma level for 25-hydroxyvitamin D and calcium at the latest visit. Daily intake of calcium and vitamin D was determined from 3 day food diary.

Results There were 16 patient included, 14 were girls and average age 13.3 years. Duration of steroid use was 27.75 +17.4 month with the last SLEDAI score of mild and moderate flare on 9 subjects. Subjects received 400 mg calcium and vitamin D3 200 IU (5 mcg) daily. Average Calcium intake was 587.58+213.29 mg (RDA 1300 mg/day) and vitamin D 2.9 mcg daily(RDA 15 mcg/day). Plasma level of 25-OHD was 19.3+5.4 ng/ml (normal level 36–108) and calcium was 8.69 +0.5 mg/dl (normal level 8.5–10.2). DXA results showed 7 subjects with Z-score lumbal <−2.0 and mean Z-score −1.75 +1.24

Conclusions Our paediatric Lupus patient showed low plasma level of calcium and vitamin D. Current supplementation should be increased 2 times to achieve relevant RDA and maintaining normal bone density

139 CLINICAL PRESENTATIONS AND OUTCOMES OF FILIPINO MALE LUPUS PATIENTS

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Background and Aims Systemic lupus erythematosus(SLE)is a multisystemic disease affecting almost all of the organ systems. Children frequently manifests with non specific signs and symptoms.Renal involvement accounts for 40%–70% of SLE patients.Male lupus represent a small percentage but should not be underestimated in terms of most health-related issues.

Methods Hospital charts seen at the Philippine General Hospital(PGH) over a 10 year period(2004–2013) were retrieved and reviewed from the Medical Records.Demographic,clinical presenting features and manifestation during the illness and laboratory findings during the course of the disease for each patient were collected.

Results Two hundred fifteen lupus patients were seen at PGH from 2004–2013. The female to male ratio was 6:1(186 f,29

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 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

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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 ΔSLEDAI-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

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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

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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.