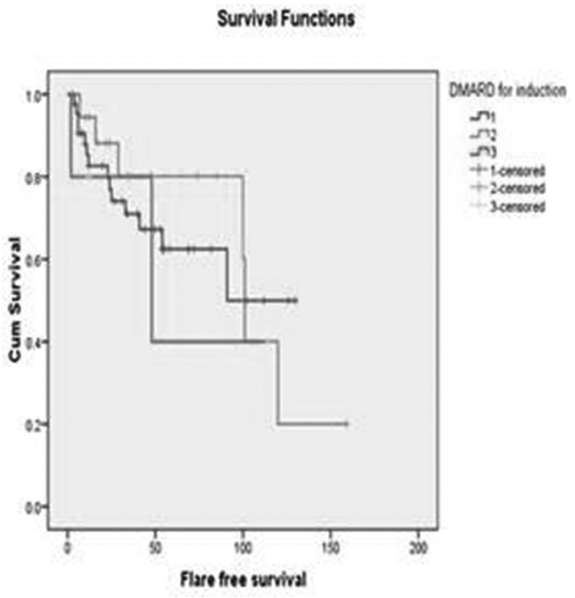

Abstract 124 Figure 1 Kaplan Meir survival curve: Median time to flare with Drug1 (MMf) 91 months as induction agent vs 101 months with drug 2(CYC) and 48 months with drug 3(Aza)

125 OUTCOME OF CHILDHOOD LUPUS NEPHRITIS: A 15 YEARS EXPERIENCE OF A SINGLE CENTRECENTER

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Background and aims To report the long-term renal outcome of a cohort of Saudi children with systemic lupus erythematosus (SLE)

Methods All patients with childhood lupus nephritis (cLN) proved by renal biopsy seen between January 2000 and June 2015 were reviewed The renal outcome was assessed according to serum creatinine level, protein/creatinine ratio at the last follow-up visit, and/or evidence of renal impairment during follow-up period and end stage renal disease (ESRD). Additional outcome measures include accrual damage measured by pSDI and death related to SLE was determined.

Results A total of 84 (72 females) cLN patients with follow-up duration of 9.3 years were included. The mean current age was 19.4 years and mean age at onset was 9.2 years. The most frequent histological class was proliferative glomerulonephritis (64.3%) followed by membranous nephritis (27.4%). The mean activity and chronicity were 6 and 4 respectively. Renal microthrombosis was found in 9 (10.7%) patients. All patients treated with immunosuppressive medications; cyclophosphamide used in 64 followed by mycophenolate mofetil in 42 while rituximab used in 24 patients. At last follow up visit, the mean serum creatinine was 147 and the mean protein/creatinine ratio was 0.8 while the mean total SDI was 1.89. Sixteen (19%) patients had ESRD. However, there was no significant difference in ESRD by histological class. The overall survival rates were 5 years: 94% and 10 years: 87%. Infection was the leading cause of mortality.

Conclusions Our patients had severe cLN and required intensive treatment. However, the survival rate is comparable to other studies.

126 AUTOIMMUNE HEMOLYTIC ANAEMIA EVOLVING TO SYSTEMIC LUPUS ERYTHREMATOSUS

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Background and Aims The first manifestation of systemic lupus erythrematosus (SLE) could vary, atypical, and often confusing. Hemolytic anaemia can be the only manifestation of SLE. With low index of clinical suspicion or inadequate follow up, the diagnosis of SLE could be delayed. We present a case of autoimmune hemolytic anaemia (AIHA) patient to describe the nature of AIHA which could evolve to SLE.

Methods Case report of a 12-year-old Indonesian female was admitted to Dr. Hasan Sadikin General Hospital with complaints of pallor, chest pain, prolonged fever and redness on her cheek. She was diagnosed with idiopathic AIHA but never routinely checked up after being discharged from the hospital 6 months ago. On admission she was alert, febrile, had muffled heart sound, and malar rash. Her laboratory investigation revealed anaemia (Hb: 6,8 g/dl), low C3 and C4, and increased titer of anti ds-DNA. Chest radiograph showed cardiomegaly and echocardiography showed pericardial effusion. A diagnosis of autoimmune hemolytic anaemia, systemic lupus erythrematosus, and pericardial effusion was confirmed. She was administered metilprednisolon 40 mg/day.

Results The patient had good response to metilprdnisolon. Fever, malar rash, chest pain start to resolve at day 4th of steroid administration. She was discharged at day 7th. One month after hospital discharge lupus activity disease remained stable and steroid was tappered off.

Conclusions We conclude that AIHA can be the only first symptoms of SLE. A careful observation of AIHA is important to prevent late diagnosis and treatment of SLE.

127 CARDIOVASCULAR COMPLICATIONS IN PEDIATRIC-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS IN SAUDI ARABIAN PATIENTS

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10.1136/lupus-2017-000215.127

Background and Aims Cardiac involvement among pSLE patients is a known complications. Early diagnosis and treatment of pSLE cardiac complications is crucial, as they may carry poor prognosis.We are reporting the prevalence and different types of cardiovascular complications in Saudi Arabian patients with pSLE.

Methods 46 pSLE patients (6 to 19 years) were following from January 2014 to September 2015 at the rheumatology clinic of King Abdul-Aziz University Hospital, Jeddah. Laboratory data such as CRP, ANA, anti-dsDNA, C3 and C4 complements, were collected. Cardiac evaluation included chest x-ray, ECG, and echocardiography, along with estimation of SLE activity by calculating the SLE Disease Activity Index (SLE-DAI) score according to SELENA Modification

Results Prevalence of cardiac manifestations was 47.8%, occurring at a mean±SD age of 14.0±2.28 years. Valvular heart diseases were detected in 16 (34.8%) cases, followed by pericarditis in 6 (13%), and silent valvular diseases in 8 (17.4%) cases. Of the 16 valvular diseases, tricuspid and pulmonary

valves were involved in 9 and 8 cases, respectively. Cardiac involvement was silent in 36.4% and occurred as an initial presenting symptom of SLE in 9.1% cases. Biologically, patients with cardiac involvement had higher levels of CRP and anti dsDNA, and lower levels of complement C3 compared to patients with no cardiac involvement; while SLE activity was the only significant predictor for cardiac involvement (Beta=0.654; p=0.020)

Conclusions Cardiac complications are common (1 out of 3 times). They are predicted by high SLE activity and anti-dsDNA, CRP and low C3 levels. Reguiar echocardiography is erommended for Patients with high SLE activity.

128 OUTCOMES OF 847 CHILDHOOD-ONSET SYSTEMIC LUPUS ERYTHEMATOUSUS PATIENTS IN THREE AGE GROUPS

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Background and aims To assess outcomes of childhood systemic lupus erythematosus (cSLE) in three different age groups evaluated at last visit: group A early-onset disease(<6 years), group B school age(≥6and<12 years) and group C adolescent (≥12and<18 years).

Methods Observational cohort study of 10 Paediatric Rheumatology centres, including 847 cSLE patients.

Results Group A had 39 (4%), B 395 (47%) and C 413 (49%). Median disease duration was higher in group A compared to groups B and C[8.3 (0.1–23.4) vs. 6.2 (0–17) vs. 3.3 (0–14.6) years, p<0.0001]. The median SLICC/ACR-DI[0 (0–9) vs. 0 (0–6) vs. 0 (0–7), p=0.065] was comparable in all groups. Further analysis of organ/system damage revealed that frequencies of neuropsychiatric(21% vs. 10% vs. 7%, p=0.007), skin (10% vs. 1% vs. 3%, p=0.002) and peripheral vascular involvements(5% vs. 3% vs. 0.3%, p=0.008) were more frequent in group A compared to B and C. Frequencies of severe cumulative lupus manifestations such as nephritis, thrombocytopenia and autoimmune hemolytic anaemia were similar in all groups(p>0.05). Mortality rate was higher in group A compared to groups B and C(15% vs. 10% vs. 6%, p=0.028). Out of 69 deaths, 33/69 (48%) occurred within the first two years after diagnosis. Infections accounted for 54/69 (78%) of the deaths and 38/54 (70%) had concomitant disease activity.

Conclusions: This large multicenter study provided evidence that early-onset cSLE group had distinct outcomes, with higher mortality rate and neuropsychiatric/vascular/skin organ damages in spite of comparable frequencies of severe cumulative lupus manifestations. We also identified that overall death in cSLE patients was an early event mainly attributed to infection associated with disease activity.

129 PANNICULITIS IN CHILDHOOD-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS: A MULTICENTRIC COHORT STUDY

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Background and Aims To evaluate prevalence, clinical manifestations, laboratory abnormalities, treatment and outcome in a multicenter cohort of childhood-onset systemic lupus erythematosus(cSLE) patients with and without panniculitis.

Methods Panniculitis was diagnosed due to painful subcutaneous nodules and/or plaques in deep dermis/subcutaneous tissues and lobular/mixed panniculitis with lymphocytic lobular inflammatory infiltrate in skin biopsy. Statistical analysis was performed using Bonferroni correction(p<0.004).

Results Panniculitis was observed in 6/847 (0.7%) cSLE. Painful subcutaneous erythematosus and indurated nodules were observed in 6/6 panniculitis patients and painful subcutaneous plaques in 4/6. Generalised distribution was evidenced in 3/6 and localised in upper limbs in 2/6 and face in 1/6. Histopathology features showed lobular panniculitis without vasculitis in 5/6(one of them had concomitant obliterative vasculopathy due to antiphospholipid syndrome) and panniculitis with vasculitis in 1/6. Comparison between cSLE with panniculitis and 60 cSLE without panniculitis with same disease duration [2.75 (0–11.4) vs. 2.83 (0–11.8) years, p=0.297], showed higher frequencies of constitutional involvement (67% vs. 10%, p=0.003), leukopenia (67% vs. 7%, p=0.002) and median C-reactive protein (10.5 vs. 0.5 mg/L, p=0.001). Cutaneous atrophy and hyperpigmentation occurred in 83% of patients.

Conclusions Panniculitis is a rare skin manifestation of cSLE occurring in the first three years of disease with considerable sequelae. The majority of patients have concomitant mild lupus manifestations.

130 NORMAL-PRESSURE HYDROCEPHALUS IN A NINE YEAR OLD FILIPINO FEMALE WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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10.1136/lupus-2017-000215.130

Background and Aims Central nervous system involvement in paediatric systemic lupus erythematosus (SLE) is common, however, normal-pressure hydrocephalus is unusual.

Methods The medical records of a nine-year old Filipino female with SLE was reviewed.

Results Patient presented with prolonged fever, weight loss, generalised weakness, lapses in memory, and mood lability. She was poorly nourished, non-ambulatory, pale, febrile, and ill-looking. She was tachycardic, with normal blood pressure,