adults. Males comprise 4-22% of all SLE patients. Gender distribution of pSLE is 4.5-5: 1 (female-to-male) as opposed to 9-10: 1 (female-to-male) in the adult population. However male patients have been known to have a more severe disease with higher morbidity, especially due to renal causes.

Objectives: To evaluate clinical and immunological profile and outcomes in a follow up series of males with pediatric onset SLE

Methods: We analyzed the clinical and immunological profile and outcome of male patients diagnosed with SLE at less than 16 years of age. These children were followed-up in Pediatric Rheumatology Clinic, Advanced Pediatrics Centre, Postgraduate Institute of Medical education and research, Chandigarh, India. Details on demographic data, clinical presentation, laboratory findings, immunological profile, treatment regimens and outcomes of these children were retrieved from clinic files.

Results: Forty-three boys were diagnosed to have SLE between January 1998 to December 2018. Mean age at presentation was 9.7 years (range: 9 months-12 years). Total patient years of follow up was 192 years. The most common clinical presentation was fever in 38 (88%) patients; rash in 27(63%); pallor in 21(49%); edema with urinary abnormalities in 17(40%) and photosensitivity in 16 (37%). A diagnosis of lupus nephritis was made in 25 (58%) patients out of which 17 (39.5%) had nephritis at presentation. Renal biopsies were performed in 18 patients; 11 had class IV disease, 2 had class 5 disease, 2 had class IV/V. Neuropsychiatric manifestations were seen in 11 (26%) patients- 7 of these had symptoms at presentation. Seizures were the predominant manifestation in 9 patients (21%)-6 of these had MRI changes consistent with posterior reversible encephalopathy syndrome while 3 had cerebral infarcts. Other central nervous system abnormalities included psychosis (1 patient) and chorea (1 patient). We also noted a family history of lupus like illness in 4 patients and 3 (6.9%) were found to have early complement deficiencies. Antiphospholipid antibodies (aPLA) were detected in 8 (18.6%) patients - anticardiolipin antibody was positive in 8 and lupus anticoagulant was positive in 6. Dual aPLA positive was seen in 6(13.9%) and triple positive was positive in none. Infections were seen in 16 (37%) patients during follow-up. All patients received steroids in gradually tapering doses along with hydroxychloroquine following a diagnosis of SLE. Cyclophosphamide was given for induction in 13 patients who had severe forms of lupus nephritis. Remission was maintained through azathioprine in 8 patients and 8 required Mycophenolate mofetil. Ten patients (23%) had a relapse on therapy -1 had CNS relapse, 3 had renal relapse, 2 had muco-cutaneous relapse and 4 had hematological relapse. Six fatalities (14%) were recorded during follow-up -1 with severe disease activity and neuropsychiatric manifestations, 1 had disseminated tuberculosis, 1 had CNS flare with status epilepticus, 4 had sepsis.

Conclusion: This is one of the largest series on boys with pediatric onset SLE from a developing country. It appears that while the severity of lupus nephritis in boys is no different from that in girls, neurological disease is more severe in the former. Further, boys appeared to have earlier onset of neuropsychiatric lupus as compared to girls. The incidence of complement deficiency lupus was also more in boys. Mortality in boys with SLE appears to be comparable to our previously published mortality data on pSLE(1).

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AB0950 CORRELATION OF QUANTITATIVE MAGNETIC RESONANCE IMAGING BIOMARKERS AND AXIAL PAIN IN ENTHESITIS-RELATED ARTHRITIS PATIENTS

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Background: Magnetic resonance imaging is increasingly used to diagnose, stratify and monitor inflammation in patients with spondyloarthritis, including young patients with enthesitis-related arthritis (ERA). Recently, there has been interest in the use of quantitative magnetic resonance imaging (qMRI) as an objective method of assessing disease. For example, diffusion-weighted imaging (DWI) and fat fraction (FF) mapping have

emerged as biomarkers of oedema and structural damage, respectively, in inflamed bone¹. However, as yet there are limited data to tie these biomarkers to underlying immunopathology.

Objectives: To analyse the relationships between quantitative imaging biomarkers (QIBs), axial pain and serum biomarkers – including 'inflammatory' biomarkers and those reflecting bone turnover - in a cohort of ERA patients.

Methods: 28 patients with ERA were prospectively recruited. The hips, sacroiliac joints and lumbar spine were assessed with chemical shift-encoded MRI and diffusion weighted imaging. Quantitative MRI parameters, such as the apparent diffusion coefficient (ADC) and fat fraction (FF) were measured in the sacroiliac joints. Visual scores for inflammation and fat metaplasia were also reported. BASDAI questionnaires were completed. Serum cytokines and bone markers were measured via multiplex bead-based immunoassays (Luminex) and results were available for 14 patients with ERA.

Results: The mean visual analogue score for spinal and hip pain (BASDAI Q2) was 3.9 (Table 1). There was a correlation between ADCmean and spinal and hip pain ($r_s=0.393$, $P=0.043$). There was also a negative correlation between FFmean and the above pain score ($r_s = -0.378$, $P=0.047$). There was a positive correlation between matrix metalloproteinase-3 (MMP-3) levels and FFmean ($n=14$, $r_s=0.698$, $P=0.006$). However, the correlation did not reach statistical significance when corrected for multiple testing.

Conclusion: The correlation of ADCmean, which increases with increased tissue water/inflammation, with spinal and hip pain suggests that BASDAI Q2 relates to pain of active inflammation. Similarly the inverse correlation of FFmean, which increases with bone healing and reduced inflammation, and BASDAI Q2, would also support this relationship.

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Abstract AB0950 Table 1

Baseline Characteristics of ERA patients	
Number, n	28
Age (years), mean	17.96 (12-22)
Disease duration (years)	5.64 (0-16)
Sex	
Male, n (%)	19 (67.9%)
Female, n (%)	9 (32.1%)
Biologic monotherapy, n (%)	1 (3.6%), (Etanercept)
csDMARD monotherapy, n (%)	7 (25%), (4 Methotrexate, 3 Sulfasalazine)
Biologic and csDMARD, n (%)	12 (42.9%)
NSAIDs only, n (%)	2 (7.1%)
No treatment, n (%)	6 (21.4%)
BASDAI (mean)	3.41 (0-8)
Spinal and Hip pain (BASDAI Q2, 0-10)	3.89 (0-10)
HLA-B27 positive, n (%)	12 (42%)
CRP (mean, mg/dl), n=23	4.07 (0.5-22)
Sacroiliitis on MRI, n (%)	21 (75%)

Disclosure of Interests: None declared

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AB0951 MULTICENTRIC CARPOTARSAL OSTEOLYSIS SYNDROME – A RARE BUT IMPORTANT DIFFERENTIAL OF JUVENILE IDIOPATHIC ARTHRITIS. CASE REPORT OF TWO BROTHERS WITH AN IDENTICAL MAFB MUTATION

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Background: Carpotarsal osteolysis is observed in a rare group of 'vanishing bone' skeletal dysplasias. Diagnosis requires a high index of suspicion combined with clinical and radiological signs. Inheritance varies amongst subtypes.

Multicentric carpotarsal osteolysis syndrome (MCTO) is characterised by aggressive osteolysis in early childhood predominantly in the carpal and tarsal bones. It is associated with kidney disease in >50% which can progress rapidly to end stage renal failure. Prediction of renal impairment is not currently possible. MCTO shows an autosomal dominant inheritance pattern with heterozygous mutations in the *MAFB* gene.