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%)

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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.

Objectives: Describe two brothers affected by MCTO highlighting the importance of a genetic diagnosis.

Methods: B1 (20 years) presented age 3 years with swollen painful feet and restricted movement in his fingers and wrists and was diagnosed with polyarticular juvenile idiopathic arthritis (JIA). After no response to steroid treatment he developed rapid onset fixed flexion deformity of both elbows. He had mild learning difficulties. Based on clinical and radiological findings he was diagnosed with carpotarsal osteolysis age 4 years. Bisphosphonates and vitamin D replacement did not halt progressive bony destruction. Consistent with the literature osteolysis stabilised in the teenage years. Renal function remains normal.

B2 (5 years) is a dizygotic twin and younger sibling of B1. All other family members: parents (non-consanguineous), twin sister, sister, brother and half-brother are healthy. B2 presented aged 21 months with bilateral painful swollen wrists and reduced range of movement. His development was normal and comparable to his twin. Clinical presentation was similar to B1 who was yet to receive a genetic diagnosis. Plain films were not diagnostic as carpi unossified. Regular monitoring of renal function was advised although it was noted the nephropathy associated with MCTO does not normally manifest until the second decade.

At genetic review an autosomal recessive form of carpotarsal osteolysis was considered most likely. The elbow contractures in B1 were consistent with this phenotype although subcutaneous nodules were absent. Genetic testing for *MAFB* gene alterations identified an identical mutation in B1 and B2 (*MAFB* – c176c>Tp(Pro59Leu) confirming the autosomal dominant MCTO. B2 developed hypertension, heavy proteinuria and hypoalbuminaemia at 3 years, responding to tacrolimus.

Results: Two brothers with clinical features of MCTO and identical mutations in *MAFB* gene are described. The phenotype varied significantly with early renal disease in one and mild learning difficulties in the other. Inheritance of this autosomal dominant condition in these two brothers with unaffected parents could result from either gonadal mosaicism or incomplete penetrance. There is a case of incomplete penetrance in MCTO described in the literature. [2]

Conclusion: Individually skeletal dysplasias are rare yet as a group they represent an important differential to JIA requiring different management. Pursuing a genetic diagnosis in carpotarsal osteolysis is important not only for prediction of recurrence risk. It allows a definitive diagnosis to be made earlier than is possible clinically/radiologically. Significantly different phenotypes yet the same genetic mutation lends weight to the possibility of modifier genes or other epigenetic mechanisms/environmental factors affecting disease penetrance as proposed by Dworschak et al [2]. A greater understanding of these may enable prediction of renal involvement and aid development and stratification of new therapies.

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AB0952

RETROSPECTIVE STUDY OF PROCEDURAL PAIN ANALYSIS DURING JOINT INFILTRATION IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background: Juvenile Idiopathic Arthritis (JIA) is the rheumatic disease more common in childhood. Intra-articular corticosteroid injections (IAC) are a mainstay in the treatment of this disease, unfortunately it is often associated with pain and anxiety, for this reason is extremely important to pay attention both to procedural analgesia and to the reduction of anticipatory anxiety.

Objectives: The primary aim was evaluate the impact of the our analgesic procedure on the perception of infiltration pain; the secondary aims

were stratify the results according to the demographic characteristics, number of joints infiltrated for each patient.

Methods: 40 patients (pt) studied between august 2016 and august 2017 (1 year) received a IAC [F/M=25/15, age 8.63 ± 3.41 yrs, weight 32.1 ± 13.9 kg; monoarticular JIA 7 pt (17.5%), oligoarticular 19 (47.5%), polyarticular 14 (35%). 27/40 (67.5%) pt infiltrated in one joint, 13/40 (22.5%) in two or more joints].

One hour earlier of the procedure for each pt was applied 1% prilocaïne cream and 30 minuts before administered Midazolam orally at a dose of 0.5 mg/kg (maximum 15 mg), 50% nitrogen protoxide mixture dispensed with a mask, in spontaneous breathing and continuous flow. In 25/40 (62.5%) pt distraction techniques were performed during IAC and in 24/40 (60%) was also applied ice spray before IAC. To monitor pain, before and after IAC, appropriate scales have been used, stratified by age: CHIPPS scale (pt ≤ 5 yrs), Wong-Baker scale for ages 5 to 8, numerical scale analog visual VAS (pt ≥ 8 yrs). CHIPPS and VAS scales <4 : indicative of mild pain. For each pt pain assessment was performed before and after IAC.

Results: VAS before IAC = 0.90 ± 1.67 /Median (M) = 0, after IAC = 1.07 ± 1.56 /M = 0; CHIPPS before IAC = 0.82 ± 1.88 /M = 0, during IAC = 2.75 ± 3.01 /M = 2. No significant correlation between VAS scale scores and after IAC and of CHIPPS both before and during IAC and respectively of age, weight, sex, application of ice spray, application of distraction techniques before the execution of the procedure, number of infiltrated joints. Significant correlation between VAS before and after IAC (RHO Spearman 0.397, $p = 0.018$), CHIPPS before and during IAC (RHO Spearman 0.599, $p \leq 0.0001$). Highly significant correlation coefficient between VAS and CHIPPS before IAC (RHO Spearman 0.6, $p = 0.0001$) and between CHIPPS during IAC and VAS after IAC (RHO Spearman 0.403, $p < 0.05$). Significant difference between CHIPPS before and after IAC (Wilcoxon Ranks Signed Test, $R = 8.5$, $p < 0.0001$), not between VAS before and after IAC ($R = 83$, $p = 0.62$).

Conclusion: In our study the intensity of pain would seem exclusively subjective, in fact it does not depend: on age, number of IAC, on the application of distraction techniques and application of ice spray. Anyway the protocol that we used seems to be effective on pain control (VAS and CHIPPS scale respectively after and during IAC: $m \pm sd$ and M. <4 points) and it can be performed on outpatients safely without an anesthetic procedure. Our results encourage to expand the series with a future prospective trial.

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AB0953

EAR INVOLVEMENT IN PATIENTS AFFECTED BY JUVENILE IDIOPATHIC ARTHRITIS

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Background: Few clinical studies in pediatric patients have shown a possible link between hearing loss and Juvenile idiopathic arthritis (JIA). It could be related to the involvement of the joints of the ossicular chain as a result of the autoimmune inflammatory process in the middle and inner ear

Objectives: The aim of this study was to assess the frequency of hearing impairment in patients affected by JIA versus a healthy control group.

Methods: We studied 64 ears of 32 JIA patients (25 girls and 7 boys, mean age 15 years, age range 8-19 years) from June to December 2018. The mean disease duration was 7 years (± 4 DS). Polyarticular JIA was diagnosed in 6 patients, oligoarthritis JIA in 19; 5 patients had psoriatic arthritis and 2 spondyloarthritis. The control group consisted of 60 ears of 30 healthy children, sex-and age-matched. Patients with previous otitis media at least 3 months and/or ear surgery were excluded. The study was approved by the local ethics committee and parents and/or patients signed their informed consent. All subjects underwent