work in collaborative teams to ensure participation and sustainability of processes. Next steps would be to add additional quality and disease outcome measures.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.6188

AB0947

TREATMENT WITH SIMULTANEOUS BIOLOGICAL AGENTS IN JUVENILE IDIOPATHIC ARTHRITIS: SINGLE-CENTER CASE SERIES AND REVIEW OF LITERATURE

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Background: Several therapeutic choices are currently available for juvenile idiopathic arthritis (JIA) patients. The classical first line therapies are NSAIDs and corticosteroids (CTCs) and the second line encompasses syntheical and biological DMARDs. The clinical efficacy of DMARDs (alone or in combination) has been well demonstrated in RCTs and the treatment is often tailored on an individual basis. Combination of bDMARDs has not been formally evaluated in rheumatology and few publications are available concerning the combination of bDMARDs in patients with severe active forms of JIA, mainly systemic [SoJIA] and polyarticular [PA] categories. (1)

Objectives: To describe efficacy and safety of JIA patients resistant to MTX and bDMARDs requiring a simultaneous bDMARDs treatment.

Methods: Retrospective analysis of 7 (4.7 ± 2.1 years, male sex 75%) patients with JIA of a single tertiary center. A systematic review of literature was made to search observational studies from MEDLINE database (January 1946 to January 2019).

Results: All patients in the Paediatric Rheumatology Department (Lyon University Hospital) receiving combination therapy with bDMARDs were eligible. Seven patients with JIA diagnosis (4 SoJIA and 3 PA) were included. Genetic studies were performed in three patients, with positive LACC1 mutation in one. The exposure to the bDMARDs of the whole cohort was 79 patient-years (PY), including 16.5 PY of combination with simultaneous bDMARDs. The delay between the date of diagnosis and the first prescription of a combination of simultaneous bDMARDs treatment was 8.3 ± 4.8 years. Nine bDMARDs drugs were prescribed: anti-TNFs, tocilizumab and rituximab (nine times each). Ten co-prescriptions of the bDMARDs were administrated with 5 possible types of combinations between them, the most frequent associating tocilizumab/rituximab. Rituximab was used most frequently in combination with another bDMARD. The rituximab schedule was different between patients. Disease activity decreased for all patients, primarily at the biological level without reaching complete clinical remission.

In general, the clinical tolerance to the co-prescription of bDMARDs was acceptable. One patient (P2) experienced a severe hepatitis event in adulthood attributed to the treatment (tocilizumab), dropping the bDMARDs combination. During the exposure time, the most frequent adverse events were infections: repeated otitis media (P1), persisting vaginitis and fungal urinary infection (P3) not requiring hospitalization. One patient (P6) had 2 episodes of purpuric rash on the days of the canakinumab injection requiring oral CTC treatment. No severe acute reactions were found and no patients died.

The systematic review found a publication: retrospective series of 4 patients (SoJIA) combining abatacept and canakinumab (2).

Conclusion: Severe forms of JIA that are refractory to the current therapeutic options available could benefit from the combination of bDMARDs. The combination of bDMARDs may be an alternative therapy to explore in this group of patients.

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Acknowledgement: Sincere thanks to Dr. Maria-Luisa Cabañas (INCAN, Ministry of Health of Paraguay)

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.5712

AB0948

PARADOXICAL TINEA AMIANTACEA IN A PATIENT WITH JUVENILE IDIOPATHIC ARTHRITIS RECEIVING ADALIMUMAB

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Background: Tinea amiantacea is a papulo-squamous condition of the scalp that can lead to scalp fibrosis and subsequent permanent hair loss. It is thought to represent a reaction pattern to inflammatory skin disease as psoriasis or seborrheic dermatitis (1).

Objectives: To highlight an adverse reaction which involved the skin in the disease course of a young JIA (juvenile idiopathic arthritis) patient, during treatment with adalimumab.

Methods: A 16-month-old female patient presented to our clinic with a 4-week history of knee swelling, associated with functional limitation and morning stiffness. Family history was unremarkable, while past medical history revealed atopic dermatitis in the first year of life. The baby was initially treated with NSAIDs, but one month later, due the persistence of arthritis and the appearance of uveitis, subcutaneous methotrexate was started (15 mg/m²/weekly). However 5 months later, given the persistence of uveitis and the onset of a severe hypertransaminasemia, methotrexate was interrupted and adalimumab (24 mg/m² every 2 weeks) was introduced with a prompt and stable control of ocular and articular disease and a gradual normalization of transaminases. One year later the patient developed dry, itchy, red and cracked skin behind her ears, with fissuring in the lower attachment of the ear lobe, and presented right parietal yellowish scalp lesions which were pruriginous, thick, and scaly, attached both to the scalp and to the proximal hair shafts. A first diagnosis of pityriasis amiantacea secondary to atopic dermatitis was made. A paradoxical cutaneous reaction to the anti-TNF therapy was later hypothesized (2), and 7 months later adalimumab was interrupted with quick resolution of the dermatologic lesions. However, both arthritis and uveitis rapidly recurred, showing an inadequate response to a six month cycle of abatacept treatment (10 mg/kg/month). Adalimumab was than reintroduced with a rapid improvement.

Results: Currently, after 16 months of adalimumab treatment, the patient still shows complete disease control, without any new dermatologic lesions up to now.

Conclusion: TNF antagonist-induced tinea amiantacea is a rare adverse reaction that may require the drug discontinuation. Although the exact pathogenetic mechanism is unclear, an imbalance in the cytokine milieu with a selective overexpression of type I interferon has been hypothesized.

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Disclosure of Interests: Maria Costanza Caparello: None declared, Francesca Tirelli: None declared, Gabriele Simonini Grant/research support from: Abbvie, Speakers bureau: Abbvie, Rolando Cimaz: None declared, Teresa Giani: None declared

DOI: 10.1136/annrheumdis-2019-eular.4311

AB0949

IS PEDIATRIC ONSET LUPUS MORE SEVERE IN BOYS? OUR EXPERIENCE AT A TERTIARY CARE CENTER IN NORTH-WEST INDIA

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Background: There is diversity in clinical presentation of pediatric onset SLE (pSLE) and the manifestations are more severe as compared to

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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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.7687

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

DOI: 10.1136/annrheumdis-2019-eular.7742

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.