a follow up of one year (n=21), 2 years (n=11), and 3 years (n=5). A reduction in the daily median dose of prednisone from 10 mg [0–15 mg] to 0 [0–0 mg] in 3 years, ($p<0.05$) was observed. After a median follow-up of 20.5±11.7 months in 4 patients, the interval between TCZ doses was increased to 5 weeks (n=2), 6 weeks (1) and 7 weeks (1) because of remission. TCZ had to be withdrawn due to articular inefficiency (1) or articular and ocular inefficiency (1). The main adverse effects were severe autoimmune thrombocytopenia, autoimmune anemia and thrombocytopenia, pneumonia, viral conjunctivitis and bullous impetigo in 1 patient each.

Conclusions: TCZ is useful at short and long term follow-up for severe Juvenile Idiopathic Arthritis-associated uveitis. It is possible to optimize the TCZ dose.

Disclosure of Interest: None declared

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THU0527 PEDIATRICIAN AND ADULT RHEUMATOLOGIST COLLABORATING IN A MULTIDISCIPLINARY REUMA-PED CLINIC. IS THIS TRANSITIONAL CARE MODEL EFFECTIVE?

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Background: Transitional care should be a planned movement of adolescents with chronic diseases from child-centred to adult-oriented health care system. Recently a EULAR/PReS taskforce has developed the first international set of recommendations and standards for transitional care[1].

Objectives: To describe the results from a specific transitional care programme.

Methods: The current transitional care programme includes a multidisciplinary PED-REUMA clinic (MPRC) weekly and a non-defined period of rheumatologic follow-ups by the same rheumatologist of MPRC. The transitional care team is composed of two pediatricians, one adult rheumatologist -as transition coordinator-, a clinical nurse specialist and administrative support, as well as a psychologist and physiotherapists. Clinical information and therapies were collected throughout the disease course, and the HEADSS method of psychosocial interviewing has been included recently.

Population of the present study included young patients (YP) from that programme who are going to transfer to adult-oriented health care system. Descriptive study of socio-demographics and clinic features was included, as well as patients' adherence. YP confidence to be transferred and satisfaction with the current transitional process were measured using on a scale of 0 to 10. In patients suffering from juvenile idiopathic arthritis (JIA), clinical status of disease activity and clinical remission and CHAQ were tested before transfer.

Results: Twenty-seven YP with female predominance (63%) were included. The average age was 21±3 yo at time of a planned transfer and 16±3.4 yo at inclusion to the MPRC. JIA was the commonest condition whereas dermatomyositis was uncommon. Up to 63% patients required some DMARDs during the MPRC follow-up, but only a 37% needs maintained immunosuppressive therapies and three (11%) patients required changing the therapeutic target before transfer. YP adherence to rheumatologist appointments was high. Regarding HEADSS data: most YP were students and living at family home, around 50% gave up sports or other activities due to homework or exams, 29% of YP occasionally drank some alcohol but none used tobacco, and 47% of YP felt sad or down once in a while. Patient's confidence to be transferred was 7.7±2.1 (mean±SD; min-max: 2–10). YP showed high satisfaction with the current transitional process, 9.7±0.4 (min-max: 9–10).

Before transfer, 17 patients with JIA showed a mean±SD value of JADAS10 of 2±5 (min-max, 0–18), clinical remission on/off medication was 23% and 53% respectively. Mostly functional status reported by patient was low, YP-rated CHAQ (mean 0.06; min-max, 0.0–0.75).

Conclusions: To the best of our knowledge, this is the first study evaluating a Spanish transitional care programme. The study reports a positive impact across adolescence of our transitional care model in a real life situation. Implementation of recommendations depended on the local available resources.

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THU0528 STING-ASSOCIATED VASCULOPATHY WITH ONSET IN INFANCY (SAVI): A DIFFERENTIAL DIAGNOSIS OF INFLAMMATORY INTERSTITIAL LUNG DISEASE

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Background: STING-Associated Vasculopathy with onset in Infancy (SAVI) is an auto-inflammatory monogenic disease. SAVI is caused by an upregulation of type I interferon signaling due to sporadic or inherited gain-of-function mutations in the STimulator of INterferon Gene (STING)/Transmembrane Protein 173 (*TMEM173).* The first description of this phenotype, and the identification of the mutated gene, was in 2014. SAVI is characterized mainly by cutaneous vasculopathy leading to necrotic lesions, and progressive interstitial lung disease with secondary fibrosis.

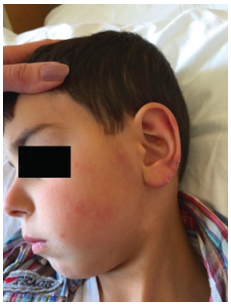
Objectives: SAVI is a rare disease with unknown prevalence. To the best of our knowledge, 25 cases have been reported so-far. These patients variably manifest cutaneous lesions and inflammatory lung disease. Here we present a case of SAVI with onset of features at the age of 3 years.

Methods: A now 8 year old boy born to non-consanguineous parents was described to have experienced recurrent fevers, polyarthralgia and polyarthritis, livedo of the limbs, facial telangiectasia, necrotic lesions of auricles and digits and failure to thrive since the age of 3 years. He has normal cognitive development. His familial history is notable for epilepsy in 2 of his siblings and Behcet disease in a paternal cousin.

Laboratory tests at age 6.5 years revealed increased ESR (60mm/h) while CRP varied from 3 to 14 mg/L. ANA titres were positive (1/640), with normal complement level and negative anti-DNA. c-ANCA and p-ANCA were negative. ECA and lysozymes were in the normal range. Although the child had no respiratory symptoms, chest X-ray revealed diffuse interstitial parenchymal infiltrates. Chest angio-CT showed ground-glass lesions with fibrotic bands and mediastinal and para-hilar adenopathies. FVC and DLCO were reduced on pulmonary functional testing. Lung biopsy was not performed.

Empirical treatment with pulse and oral corticosteroids along with azathioprine was started. Over a period of one year, systemic inflammation and skin involvement regressed dramatically, but his lung disease showed no improvement.

Results: Genetic testing identified a previously reported V155M mutation in *TMEM173*.



Conclusions: SAVI is associated with significant morbidity and mortality. The diagnosis should be considered in children with interstitial lung disease after more common causes have been ruled out. Steroids and immunosuppressive therapies apparently show no efficacy in avoiding progression to irreversible lung damage. Promising results with treatment by Janus Kinase inhibitors as a means of blocking signaling downstream of the type I interferon receptor were recently published.

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THU0529 METHOTREXATE TREATMENT RESPONSE IN NON-SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS

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Background: Methotrexate is approved and recommend as first line disease modifying antirheumatic drug (DMARD) in polyarticular juvenile idiopathic arthritis (JIA). In can be used orally or via s.c. injection.

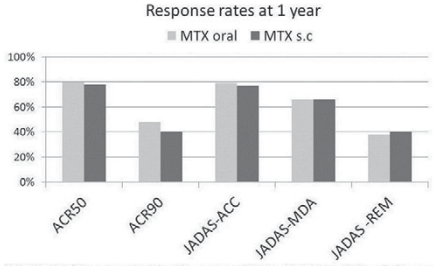
Objectives: S.c MTX is thought to be more efficious or act more rapidly than oral MTX. Thus we want to analyse the kinetic of response in JIA patients treated with oral versus s.c. MTX.

Methods: In the German BIKER registry a cohort of biologics naïve JIA patients starting treatment with MTX was built. The data bank was screened for patients treated with MTX orally vs. s.c for the first time. The JIA-ACR 90 and the JADAS10 definition of remission were used as outcome parameters.

Results: 410 JIA patients received treatment with oral MTX and 384 received s.c. MTX. RF negative polyarthritis was the most common JIA category (50%/51%)

followed by by extended oligoarthritis (27%/26%), polyarticular psoriatic arthritis (18%/16%) and RF positive polyarthritis (5%/8%). Disease duration (2.3+/-3.0 vs 1.9+/-2.7 was statistically higher in the oral cohort (p=0.04) but age at onset and baseline were similar. The baseline disease activity was higher in the s.c. cohort (JADAS10 16.5+/-7.2 compared to 14.7+/-8.2; $p<0.001$ and active joint count 9.0+/-10.1 vs. 7.4+/-7.7; p=0,011). The weekly MTX dosages were comparable with 13.6+/-5.4mg and 13.3+/-4.5 mg. Concomitant treatment with NSAIDS (95%/89%), oral steroids (24%/25%) or intraarticular steroids (6%/8%) were comparable.

After 12 months of treatment, 150 (38.3%) reached a JIA ACR90 with oral MTX and 131 (35%) with s.c MTX while 86 (21. 8%) and 72 (19.5%) reached JADAS-remission (JADAS10$\leq$1). By Kaplan-Meyer- analysis no difference in the early kinetic of response was found. Upon total observation for up to 7.5 years in the intention to treat population (patients discontinuing MTX due to inefficacy or intolerance or starting a biologic were calculated as non-responders) more patients in the oral cohort reached a JADAS-remission (162; 41%) than with s.c MTX (126; 34%) which was stastically borderline significant (p=0,05; odd's ratio 1.2 [95CI 1.0–1.8]).



JADAS-ACC= acceptable disease activity, MDA=minimal disease activity, Rem=remission

Conclusions: Data from the BIKER registry out of the clinical practice do show a high rate of JIA patients reaching a significant JIA-ACR response as well as JADAS-remission upon MTX as a sole DMARD. However, on the long term more patients with oral MTX reached JADAS remission. By Kaplan Meyer analyses we did not observe a superiority of s.c. MTX in the kinetic of response. The limitations of our analysis lie the character of a registry study, the lack of randomisation and study protocol leaving all decisions to start or to stop MTX by the responsible rheumatologist. Thus such data are preliminary and should be confirmed by randomized studies.

Disclosure of Interest: None declared

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THU0530 CHARACTERIZATION OF A COHORT OF PSORIATIC JUVENILE IDIOPATHIC ARTHRITIS PATIENTS FROM A PAEDIATRIC UNIVERSITY HOSPITAL IN SPAIN

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Background: Juvenile Psoriatic Arthritis (JPsoA) is a subtype of Juvenile Idiopathic Arthritis (JIA) present in 7% of JIA patients (1). Psoriasis is present in 0.5–1% of children. Diagnosis is often difficult, with the articular manifestations often preceding skin disease by years. Data is scarce in the Spanish population.

Objectives: Describe demographical and clinical caracteristics of our cohort of JPsoA.

Methods: Descriptive, transversal study of patients attended from 1/2012–12/2016. Included were all complyers with ILAR Criteria (2) for JPsoA (Edmonton 2001). We also included the Wallace Criteria for clinical inactive disease (3) as a variable and endpoint. Data were included and analyzed using SPSS MAC 20.

Results: 31 patients were included: 18 (59%) girls, 13 (41%) boys. All Caucasian. They comprised 5% of all our JIA patients in that period.

Mean age at diagnosis was 7.4 years. All were RF-; 9 (29%) ANA+; 4 (12.9%) HLA-B27+; articular onset 17 (55%) and cutaneous onset 14 (45%). 9 (29%) had Temporomandibular Joint (TMJ) symptoms: 5 (16%) had pain and 4 (12.9%) had a positive MRI for TMJ synovitis.

Plaque psoriasis 14 (45%), guttata 3 (9.6%) and 3 (9.6%) had both. Dactylitis 8 (26.6%); enthesitis 6 (19.35%). Joint disease was mainly oligoarticular 15 (48%), monoarticular 14 (45%) and polyarticular 1 (3.2%). Axial disease 4 (12.9%) at follow-up. 7 (22.5%) uveitis; 5 (77%) were ANA +. 3 (9.6%) onicodistrophia and 6 (19.3%) enthesitis.

All patients received NSAIDs; 30 (96%) methotrexate; 6 (19.3%) switched to leflunomide. 16 (51.6%) received biologic treatment and 9 (29%) more than one. Articular debutants 10 (32.2%) received biologic treatment more than the those with cutaneous onset 6 (19.35%). We report 3 (9.6%) anti-TNF/paradoxal psoriasis events.

Wallace Inactivity Criteria were achieved in 25 (80.6%), with no differences between the biologic and DMARDS groups in time up to Achieving Wallace Criteria (TimeWall). TMJ positive MRI did have a negative effect on TimeWall with 2 (66.6%) $\geq$8 yr (8, 11y) to TimeWall.

Conclusions: We describe the clinical features and demographics of a series of spanish JPsoA patients. We found more oligoarticular and monoarticular involvement and an important presence of enthesitis and dactylitis; higher frecuency of uveitis than published data (22.5% vs. 10–15%). Some were ANA-, reinforcing the need for screening. More than half required biologic treatment, and several cases we needed to switch drugs. Almost 60% of the patients were girls. Articular onset was associated with more active, harder to treat disease. TMJ positive RMI was associated with longer TimeWall. However, Wallace Criteria were not achieved globally.

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Disclosure of Interest: None declared

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THU0531 USE OF RITUXIMAB IN PAEDIATRIC RHEUMATOLOGY - EXPERIENCES FROM A SINGLE TERTIARY CENTRE

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Background: Rituximab is an anti-CD20 monoclonal antibody therapy used widely in the management of paediatric rheumatological conditions. Studies suggest that Rituximab is safe and effective in rheumatic autoimmune diseases, but data on paediatric use remains limited. Although Rituximab spares plasma cells, hypogammaglobulinaemia can still develop, leading to recurrent infections. Frequency of hypogammaglobulinaemia in children receiving Rituximab for rheumatological conditions is unknown.

Objectives: To analyse the use of Rituximab in a tertiary Paediatric Rheumatology centre over the last 15 years (2001–2015). The primary aims were to identify the number of patients who received Rituximab, the underlying diagnoses and the response to treatment. Our secondary aims were to identify the incidence of hypogammaglobulinemia associated with Rituximab use and the frequency and severity of infections. Frequency of monitoring of immunoglobulin levels, lymphocyte subsets and functional antibodies to pneumococcus were noted.

Methods: Retrospective analysis of case notes, electronic records and laboratory data of patients who received Rituximab in the paediatric rheumatology department from 2001–2015.

Results: A total of 22 patients received Rituximab (total of 1500mg/m^2 per cycle over 2 – 4 divided doses) during the study period. 3 were excluded due to insufficient data. Median time of commencement of Rituximab from diagnosis was 2 years 8 months. Of these, 12 patients achieved remission within 6 to 12 months. Rituximab was discontinued in the non-responders at 12 months.

Diagnosis	No of patients		Median No of cycles	Remission achieved
Polyarticular JIA	RF +	2	3.5	2 (100%)
	RF -	4		0
JDM	6		3	5 (83%)
SLE	5		2	4 (80%)
Vasculitis (GPA)	2		2.5	1 (50%)

8 patients (42%) were noted to have hypogammaglobulinaemia at some point. The role of cyclophosphamide contributing to hypogammaglobulinaemia could not be excluded in 2 and a further patient is currently being investigated for an underlying primary immune deficiency. In the remaining 5 (26%) patients, we believe the low IgG levels are secondary to Rituximab, of which two needed long term Ig replacement. Overall 12 patients reported recurrent/severe infections of which 6 had low immunoglobulin levels.

Conclusions: RF+ JIA patients appear to have responded the best to Rituximab and RF- JIA patients the least (0/4), with good results in JDM and SLE subgroups (80–83%). The incidence of hypogammaglobulinaemia secondary to Rituximab in our cohort was 26%, which can be prolonged and worsen with increased number of cycles. Prior treatment with cyclophosphamide may be contributory. We suggest regular monitoring of immunoglobulin levels and lymphocyte markers on all patients prior to commencement of Rituximab and regular intervals subsequently, including further cycles.

Disclosure of Interest: None declared

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