The mean duration of tocilizumab therapy was 14.75 months. 2 patients received s.c. accoding the poly JIA dosing and all other i.v. There were diferent i.v doses applied, 5 of them 8mg/kg every 4 weeks, one of them 8 mg /kg every three weeks, 1 every two weeks and 1 pateints received 10 mg /kg every 3 weeks. 3/11 received TOC as monotherapy. 8/11 as combination therapy, 6 of them with Methotrexate and one of each with Mycophenolate or Tacrolimus. Therapy success was refelected by a decreased mLoSSI in 8/11 patients and in 6 patients by a decrease in the Localosed Scleroderma Skin Damage Index [1] (LoSDI). No new lesion occured during the treatment and in the patients with Parry Romerg subtype (n=2) no increase in the facial atrophy occured. In 8/8 patients physician global (VAS 0–100) decreased and in 8/8 the patients global disease activity (VAS 0–100) decreased. In 3/3 patients, were it was applicable, the number of acitve joints decreased, in one patients the limb discrepancy decreased. The mean modified Rodnan skin score assessed in 8 patients decreased from the mean value of 9.6 to 5.5.

Conclusions: In this small cohort of patients TOC seems to be a promising rescue medication in methotrexate/mycophenolate nonresponsive patients. A prospective controlled study would be important to prove the seen effect in a controlled way.

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THU0512 FLUORESCENCE OPTICAL IMAGING IN JUVENILE PATIENTS WITH AND WITHOUT INFLAMMATORY PEDIATRIC RHEUMATIC JOINT DISEASES

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Background: Imaging techniques play an important role in making a diagnosis and in the evaluation of treatment effectiveness as well as in the outcome assessment of juvenile idiopathic arthritis (JIA). Fluorescence optical imaging (FOI) has been shown to visualize inflammation in arthritis of wrist and finger joints. FOI is a simple and cost-effectively imaging technique that is well tolerated by the patients.

Objectives: Firstly, to determine the association and agreement of FOI with ultrasonography (US) and physician's assessment of swollen and active joints. Secondly, to estimate the predictive power of FOI to distinguish between patients with and without inflammatory pediatric rheumatic joint diseases.

Methods: A total of 95 patients were enrolled in three pediatric rheumatology centers in Berlin, Germany. FOI and US (in greyscale (GS) and power Doppler (PD)) were performed in each patient. The FOI software automatically generated the PrimaVista mode (PVM). Furthermore, three phases (P1, P2, P3) were defined dependent on the signal intensity in the fingertips. Each joint was scored semiquantitatively (0=no signal up to 3=strong signal, more than 50% of affected joint area) in each of the three phases and PVM. US was additionally graded by a semiquantitative score of each joint for synovitis (synovial thickening and joint effusions) in GS and hyperperfusion in PD mode. The joints were defined as active if the FOI or US reached a score of at least 1, respectively. We report the results on 27 patients in this interim analysis.

Results: The mean disease duration was 3.5 years (SD=3.2), the mean cJADAS-10 was 11.0 (SD=12.3), the mean number of active joints in the hand was 3.4 (SD=5.8). Half of the patients had polyarthritis (51.8%) and one third had a non-inflammatory rheumatic disease. A total of 810 joints in 27 patients could be analyzed. Among those, 140 (17.3%) had a positive US synovitis score, 87 (10.7%) a positive US power Doppler signal, 93 (11.5%) a clinically active joint and 133 (16.4%) a positive FOI PVM. Taking the US synovitis score as reference, the FOI PVM had a sensitivity of 40%, a specificity of 86% and an overall agreement of 79%. Taking the active joint count as reference, the FOI PVM had a sensitivity of 46%, a specificity of 88% and an overall agreement of 83%. The area under curve was 0.91 for US power Doppler, 0.84 for US GS synovitis, 0.76 and 0.93 for FOI PVM and P2 for the ability to distinguish between patients with and without inflammatory rheumatic diseases. FOI and US scores correlated highly with the cJADAS-10 and the physicians global. In contrast, the patient-reported outcomes pain and fatigue did not show any correlation with FOI and US scores.

Conclusions: FOI and US had a comparable predictive power to distinguish between patients with and without inflammatory rheumatic diseases in pediatric/juvenile patients. The agreement between active joint count, US and FOI was high. FOI may provide a cost-effective method to evaluate inflammation in finger and hand joints.

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Disclosure of Interest: None declared

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THU0513 NEONATAL MANIFESTATIONS OF IMMUNE-MEDIATED RHEUMATIC DISEASES: A RETROSPECTIVE LONGITUDINAL STUDY IN A TERTIARY HOSPITAL

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Background: Autoimmune rheumatic diseases such as systemic lupus erythematosus (SLE), antiphospholipid antibody syndrome (APS) and Sjögren's syndrome (SS) are part of a clinical spectrum eligible to affect women in child-bearing ages, affecting neonatal outcomes. Cardiac, cutaneous, haematological, hepatic complications and, more rarely, pulmonary complications have been described.

Objectives: This project aims to describe the occurrence of neonatal lupus manifestations and possible associated clinical factors among women with immune-mediated rheumatic diseases.

Methods: A retrospective longitudinal study was performed including pregnant women with immune-mediated rheumatic diseases seen in a multidisciplinary group for autoimmune diseases during pregnancy between January 2010 and December 2015. Clinical and demographic data as well as and pregnancy outcomes and neonatal manifestations were collected through consultation of clinical files. Patients with and without neonatal lupus were compared using Mann-Whitney, qui-square and fisher tests (SPSS 24.0). Significance level was set as <0.05.

Results: We included 151 gestations from a total of 140 women with a mean age of 32,5±4,4 years; 4 gestations were twin pregnancies. Within these 151 gestations, 54 (35,8%) women had SLE, 17 (11,3%) had Sjögren's syndrome, 17 (11,3%) had rheumatoid arthritis, 41 had APS (27,2%), 11 (7,3%) had Behçet's disease, 4 (2,6%) had systemic sclerosis, 8 (5,3%) had mixed connective tissue disease and 16 (10,6%) had other immune-mediated diseases. 35 (23,2%) had anti-SSA/La antibodies, 18 (11,9%) had anti-SSB antibodies, 6 (4,0%) had anti-URNP antibodies and 43 (28,5%) had anti-nuclear antibodies. During follow-up, 142 (94,0%) babies were born and 7 (4,6%) abortions and 2 (1,3%) foetal losses occurred. 6 (4,2%) neonates were born with neonatal lupus and 1 (0,7%) died *in uterus* with a complete heart block. Out of the 6 babies with manifestations, 4 (66,7%) were cardiac, 2 (33,3%) were cutaneous, 1 (16,7%) was hepatic, 2 (33,3%) were haematological and 1 (16,7%) was pulmonary. Neonatal lupus manifestations occurred more frequently in mothers with SS (23.5% vs 2.2%; p=0.003), anti-SSa/Ro (20% vs 0%; $p<0.001$), and anti-SSb/La (27.7% vs 1.5%; $p<0.001$).

Conclusions: Our study proved a link between immune-mediated rheumatic diseases and specific neonatal outcomes.

Disclosure of Interest: None declared

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THU0514 PREDICTIVE VALUE OF SUBCLINICAL SYNOVITIS DETECTED BY DOPPLER ULTRASOUND IN RELATION TO FLARE IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS TREATED WITH BIOLOGIC THERAPY AFTER TAPERING BIOLOGIC THERAPY

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Background: Anti-TNF therapy is effective and safe in JIA. Changes in anti-TNF doses are common when remission is achieved[1]. Subclinical synovitis on Doppler mode (PD) detected by ultrasound can predict flares in adult RA, but it is not yet clear in JIA[2]

Objectives: The aim of this study is to evaluate the predictive value of subclinical synovitis detected by PD-US in relation to flares in patients with JIA on remission under anti-TNF when therapy is tapered. The preliminary results were presented at the EULAR congress 2015 in Rome (FRI0520).

Methods: Observational, prospective and multicenter study. We included JIA patients on remission at least 6 months with anti-TNF, ETN and ADA, in whom anti-TNF was tapered due to clinical decision. ETN was tapered by increasing the injection 3 days and ADA by increasing a week. Patients were clinically assessed every 3 months and also with PD-US at baseline. Bilateral US assessment included joints and tendons. Adult synovitis definitions and semiquantitative scoring system were used, no synovitis definitions are available for JIA. We collected demographics (date of birth, JIA subcategory, previous and current treatments. Flare was defined as clinical signs and/or symptoms of arthritis that required increase of systemic therapy

Results: We included 57 patients, with 19 patients (33.33%) having a flare

during the 12 months follow-up. 38 patients (66.67%) were receiving ETN and 19 (33.3%) ADA, of which 11 patients (28.95%) had a flare with ETN and 8 patients (42.11%) with ADA. Table 1 shows demographics. Median time to flare was 5.73 months (IR 2–93–8.9). Concomitant methotrexate was lower in patients with flare (26.32% vs 71.05%). In 18 patients (31.58%), a previous tapering was done and median time of remission before being included was 22 months (IR 15.5–28.5). US does not predict flare in our cohort. Global synovitis score at baseline was 4 (IQR 1.3–10.8) and 0 in BM and PD respectively, and tenosynovitis was 0 both BM and PD

	All patients (n: 57)	Patients with flare (n: 19)
Gender; women (%)	39 (68.42)	10 (52.63)
Time of remission previous to tapering median months (IR)	22 (15.5–28.5)	35 (35)
Biologic therapy (ETN/ADA)	38/19	11/8
Concomitant DMARD, n (%)	32 (56.14)	5 (26.32)
Previous tapering, n (%)	18 (31.58)	5 (26.32)
JIA subcategories: n (%)		
Persistent oligoarticular JIA	15 (26.32)	5 (33.33)
Extended oligoarticular JIA	14 (24.56)	4 (28.57)
RF– polyarticular JIA	15 (26.32)	5 (33.33)
RF+ polyarticular JIA	2 (3.51)	1 (50)
Enthesitis related JIA	3 (5.26)	2 (66.6)
Psoriatic JIA	8 (14.04)	2 (25)

Conclusions: Anti-TNF tapering was safe in our JIA patients in more than half of patients after 1 year follow-up. US did not predict flares in our patients. Concomitant treatment with methotrexate was more frequent in patients who remained on remission

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THU0515 DISABILITY AND LOWER QUALITY OF LIFE IS ASSOCIATED WITH SOCIOECONOMIC PASSIVITY IN YOUNG ADULTS WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background: Health related quality of life of adult patients with juvenile idiopathic arthritis (JIA) is shown to be significantly lower compared to the general population.[1,2]

Objectives: We aimed to recognize young adults with JIA who are socioeconomically passivated and to assess which areas of self-rated health are associated with the emergence of passivity symptoms.

Methods: We studied 195 young adults with JIA using questionnaires addressing demographics, health behavior, and physical activity. The HAQ questionnaire was used to assess functional ability, quality of life was assessed with RAND-36, depressive symptoms were assessed with BDI-II, and self-esteem was evaluated by using the Rosenberg scale. Patients were classified as active if they were engaged in studying, working, maternal leave or military service; and as passive, if they were unemployed or on disability pension.

Results: 80% of the patients were female, mean age was 23 years, and disease duration was approximately 15 years. Patients in the passive group participated less in leisure time non-physical activities (p =0.033), they felt more disturbed during their leisure time (p =0.010). Leisure time physical activity did not reveal statistically significant differences between the groups. The majority of the patients in the passive group (58%) had only basic education (p<0.001), they visited their doctor more frequently (p=0.019) and they used oral prednisolone more often (p<0.018). Approximately 70% of the patients received disease modifying antirheumatic drugs, and nearly half of the patients were treated with biologicals in both of the groups. Mean disability scores on HAQ were higher in the passive group (p=0.012). Depressive symptoms did not differ between the groups. Self-esteem was lower in the passive group (p=0.002). Results in health related quality of life revealed statistically significant differences between the groups: physical functioning (p=0.049), social functioning (p =0.020), and emotional well-being (p=0.047) were significantly lower in the passive group.

Conclusions: Patients being socioeconomically more passive showed higher degrees of disability, reporting lower physical functioning, self-esteem, emotional well-being, and social functioning. Those patients should be recognized earlier and activating interventions should be provided.

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THU0516 LONGER TERM OUTCOMES OF CRMO IN A TERTIARY ADOLESCENT AND YOUNG ADULT RHEUMATOLOGY CENTRE IN THE UK

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Background: Chronic relapsing multifocal osteomyelitis (CRMO) is a rare autoinflammatory bone condition presenting primarily in children and adolescents. It characteristically affects the epiphysis and metaphysis of long bones, and presents with bony pain, local swelling and warmth.

Objectives: The aim of this study was to collate our tertiary adolescent rheumatology centre's experience of managing patients with CRMO, and establish their longer term outcomes, possible by the fact we have a cohort of patients with CRMO under long-term follow-up.

Methods: We carried out a retrospective case note review of all patients who are known to our service with a diagnosis of CRMO, presenting between age 13–20.

Results: In total 17 patients were identified as having CRMO, presenting between 1999 and 2015. 10 patients were female, and 7 patients male. The median age of initial symptoms, and age of presentation was 12 years (range1–16 years).

Median duration of follow up is 4.75 years (range 1–16.5 years). Since initial diagnosis, 3 patients evolved into a SAPHO (synovitis, acne, pustolosis, hyperostosis, osteitis) phenotype, 3 an ERA (enthesitis related arthritis) phenotype and 2 patients developed oligoarticular juvenile idiopathic arthritis (JIA).

35% of patients have a purely unifocal disease, and 65% have multifocal disease as confirmed by whole body MRI. 70.5% patients had recurrent episodes of inflammation, while 29.5% of patients had only one flare and then reached remission (either clinical or confirmed with MRI). 15 patients had their diagnosis confirmed with biopsy, while 2 did not due to site of disease. Their diagnosis is assumed based on clinic impression, and typical radiographic findings.

Sites of disease confirmed in our patients include lower limbs (70% patients), upper limbs (35% patients), clavicle (29.4%), mandible (17.6%) and spine/pelvis (23.5%).

All patients were treated with NSAIDs. In terms of treatments used since diagnosis, 76% patients have been on methotrexate (MTX), 47% had one infusion of pamidronate, and 23% required more than one infusion of pamidronate. Other medications include sulfasalazine (SSZ), azathioprine, risedronate and anti-TNFs (adalimumab, etanercept and infliximab).

On last clinic review, with or without imaging, 35% of patients continue to have active disease. Currently 29% patients are on MTX alone, 23% patients are on adalimumab and MTX, and 35% are only maintained on NSAIDS. Of those without active disease 5 patients (45%) are not on any DMARD or biologic therapy.

Conclusions: We present here our experience of managing adolescent patients with CRMO.

The perceived wisdom is that CRMO is a self-limiting disease which eventually goes into remission. However our centre's experience is that nearly 50% of our patients have a disease which evolves into another systemic autoimmune disease, mainly SAPHO, polyarticlar or enthesitis related JIA. Previous case series have suggested only 0–30% of patients' disease evolves. This may be a reflection of our older cohort of patients, who are only referred to our service with ongoing disease.

The majority of patients have a recurrent and multifocal course of disease. The most common site of disease was in the lower limbs. All patients were treated with NSAIDS, and a combination of DMARDS, bisphosphonates and biologic agents have been used, which has resulted in remission of disease in the majority of patients.

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THU0517 LARGE VESSEL VASCULITIS IN INFANTS - A CASE SERIES

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Background: Large vessel involvement following recrudescent or recalcitrant Kawasaki Disease or other vasculitidies in young children have been limited to few case reports and outcomes are still unclear.

Objectives: To describe and compare the characteristics and outcomes of 6 patients with large vessel vasculitis diagnosed between 2013 to 2015 in KK Hospital, Singapore

Methods:

Demographic and disease characteristic information were collected and median, interquartile range (IQR) & percentiles were used to describe the data.

Results: 6 patients were included in the analysis. Median age was 3.75 months