

The mean duration of tocilizumab therapy was 14.75 months. 2 patients received s.c. according to the poly JIA dosing and all other i.v. There were different 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 patient 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 reflected by a decreased mLoSSi in 8/11 patients and in 6 patients by a decrease in the Localized Scleroderma Skin Damage Index [1] (LoSDI). No new lesion occurred during the treatment and in the patients with Parry Romberg subtype (n=2) no increase in the facial atrophy occurred. 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, where it was applicable, the number of active joints decreased, in one patient 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.

References:

- [1] Arkachaisri T, Vilaiyuk S, Torok KS, Medsger TA, Jr.: Development and initial validation of the localized scleroderma skin damage index and physician global assessment of disease damage: a proof-of-concept study. *Rheumatology (Oxford)* 2010, 49(2):373–381.

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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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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 achieved¹. Subclinical synovitis on Doppler mode (PD) detected by ultrasound can predict flares in adult RA, but it is not yet clear in JIA².

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