Regarding pregnancy complications and outcomes, no cases of Preeclampsia, eclampsia, small for gestational age babies, intrauterine death, were observed. One pregnancy was complicated with pre-term premature rupture of membranes at 35 gestational week, in a patient not taking prednisone. There were 5 (25%) pre-term deliveries.

Conclusion: Nearly one third of pregnant patients with JIA had a disease flare during pregnancy. Flares were observed only in 2 disease subsets (PLA; OLA-E) and associated with discontinuation of bDMARDs at positive pregnancy index. The preconception counseling of patients with JIA should include the disease subset in the risk stratification and consequently the continuation of bDMARDs during pregnancy. Maternal disease control is necessary to minimize the risk of adverse pregnancy outcomes (especially pre-term birth).

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THU0520

EARLY IDENTIFICATION OF VENTRICULAR DYSFUNCTION IN JUVENILE SYSTEMIC SCLEROSIS BY SPECKLE TRACKING ECHOCARDIOGRAPHY

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Background: Juvenile Systemic Sclerosis (JSSc) is a rare multisystemic connective tissue disease, with onset before the age of 16. Cardiac involvement, recognized in 8-24% of the patients [1], begins in early stages of JSSc and has a poor prognosis. The traditional cardiac US imaging, including the left ventricular ejection fraction (EF), evaluates the global function of the heart, thus being inappropriate to assess the sub-clinical course of the disease. A new echocardiographic technique, the speckle tracking echocardiography (STE), has been shown to be able to identify regional ventricular dysfunctions also in early stages of adult-onset SSc[2,3].

Objectives: Aim of our study was to assess the longitudinal strain of right and left ventricle in JSSc patients, in order to identify ventricular dysfunctions earlier and more effectively than with traditional echocardiography. Furthermore, we investigated the evolution of cardiac involvement during the follow-up and possible correlations with the overall disease severity, measured by the Juvenile Systemic Sclerosis Severity Score (J4S)[4].

Methods: Consecutive patients with JSSc underwent clinical and cardiological evaluation. This included traditional echocardiography (such as M-Mode, EF, Pulsed- and Tissue-Doppler), 3D-Echocardiography and STE, measuring the global longitudinal strain of left ventricle (GLS) and the longitudinal strain of right ventricle free-wall (RVLS). Each patient was assessed several times by pediatric rheumatologists for J4S and by cardiologists with STE and standard echo.

Results: 18 JSSc patients (12 F, 6 M), mean age 12.3 years, disease duration 4.5 years, entered the study. At baseline evaluation, EF was abnormal in 1 patient, whereas GLS and RVLS were abnormal in 5. The diagnostic sensitivity of cardiac involvement of STE increased, with a prevalence rising from 22.2% to 38.8%. During the follow-up, lasted mean 30 months (range 17–43), the mean GLS values gradually worsened (-21.2; -20.1; -19.4%) while there was no significant variations of EF. The strong correlation between GLS and J4S, found at baseline, vanished during the follow-up.

Conclusion: Speckle tracking echocardiography is a useful technique to evaluate the cardiac involvement in patients with JSSc. In comparison with traditional EKG or echocardiography, it allows to increase the diagnostic sensitivity of cardiac involvement. Over time, we observed a gradual worsening of GLS, sign of a progressive left ventricular dysfunction, that was not identified by EF. It is possible that the coronary microvascular damage compromises the subendocardial fibers function which are more sensitive to ischemia and whose contractility is well assessed by GLS [5]. Finally, the initial correlation between strain and J4S disappeared during the follow-up, maybe because of the pharmacological

therapy, which was effective on several aspects of the disease but had low impact on the ventricular function.

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THU0521

JUVENILE DERMATOMYOSITIS IN GERMANY – DATA OF THE NATIONAL PEDIATRIC RHEUMATOLOGY DATABASE WITH SPECIAL REGARD TO MYOSITIS-SPECIFIC ANTIBODIES AND ASSOCIATED CLINICAL PHENOTYPES

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Background: Next to weakness of the proximal muscles and typical skin lesions as leading symptoms in juvenile dermatomyositis (JDM) there can be involvement of other organ systems and tissues. Myositis-specific antibodies (MSA) may help to distinguish clinically distinct phenotypes

Objectives: 1) to analyse the clinical presentation of JDM and how diagnosis was verified and 2) to determine the spectrum of MSA and the associated clinical phenotypes in a German cohort of JDM patients.

Methods: We analyzed data of the national pediatric rheumatology database (NPRD), where children and adolescents with chronic rheumatic diseases are documented yearly by means of disease-specific questionnaires. Cross-sectional data of patients with JDM documented between 2014 and 2016 were analyzed. MSA were determined by a commercial multiplex array. To further specify the phenotype and patient's outcome, an additional retrospective chart review was conducted.

Results: We identified 186 patients with a diagnosis of JDM (69% female). Mean age at disease onset was 6.8 ± 3.5 years, mean time between first symptoms and first contact to a pediatric rheumatologist was 6.1 ± 8.9 months, mean age at documentation was 11.5 ± 4.4 years. The following diagnostic procedures were pathologic/positive for JDM: Histology 44/147 (30%), electromyography 28/144 (19%), magnetic resonance imaging 103/148 (70%), Creatine kinase (CK) elevation at diagnosis 126/148 patients (85%). At last consultation (mean disease duration 5.0 ± 3.9 years), the mean physician's global assessment of disease activity (PGA, NRS 0 – 10) was $1.6 \pm SD 2.3$, the mean manual muscle test (MMT, range 0 – 80, best 80) was $69.7 \pm SD 19$, the mean disease activity score (DAS, range 0 – 20, best 0) was $4.4 \pm D 4.7$, the mean CHAQ was $0.5 \pm SD 0.8$. MSA testing was performed on 88 patients (73% female); 42% tested positive: anti-NXP2 16%, anti-TIF1γ 14%, anti-MDA5 6%, anti-Mi2 3%, anti-synthetase-antibodies 3%. The most common clinical feature was dysphagia in patients with anti-NXP2, calcinosis in patients with anti-TIF1γ, lung involvement in patients with anti-synthetase antibodies, and pronounced muscle weakness in patients with anti-Mi2. Patients with anti-MDA5 were characterized by frequent lung involvement, mucosal ulcers, fever and polyarticular arthritis of small joints. Muscle weakness tested by Childhood Myositis Assessment Score (CMAS) was significantly associated with an increased CK level. At last consultation, 32% and 14% received oral glucocorticoids (GC) < 0.2 mg/kg or ≥ 0.2 mg/kg body weight, respectively, 52% were treated with methotrexate (MTX) and 27% with intravenous immunoglobulins (last 12 months). PGA was ≤ 1 in 65% of patients at last consultation, 22% of those were off therapy.

Conclusion: Demographic and clinical parameters of patients with JDM in Germany are comparable to JDM-cohorts in other countries. MRI has gained diagnostic importance and is used more than twice as often as biopsy. MSA could be found in almost half of the patients. Clinical