%) cases. More patients with rheumatoid-factor negative (54 vs 50%; $p=0.04$) and rheumatoid-factor positive pJIA (13 vs 10%, $p=0.04$) received combination with MTX, while more patients in the monotherapy cohort had extended oligoarthritis (40 vs 32%, $p<0.001$). Patients with MTX had a shorter disease duration (4 vs 5.5 years, $p<0.001$) and received concomitant steroid more often (34 vs 24%), $p<0.001$). There was no statistical difference regarding disease activity parameters (active joint count, patient assessment and physician assessment of disease activity, ESR, CRP, CHAQ-DI, JADAS10). Discontinuation due to ineffectiveness was reported for ETA/ADA/GOL/TOC in 20%/18%/14%/28% of patients, respectively in 3.7/4.9/6/10.5 patients/100 treatment years. Thus discontinuation due to inefficacy was reported less frequently with ETA compared to ADA ($p=0.046$) and TOC ($p<0.001$) and with ADA compared to TOC ($p<0.001$).

Patients on ETA and ADA had a slightly, but not statistically significant lower rate of withdrawal for ineffectiveness if on methotrexate (Figure 1). There was no difference regarding baseline disease activity parameters in patients with ETA/ADA monotherapy compared with combination with MTX, apart from patients with ETA+MTX receiving more often systemic steroids at baseline (36 vs 24%, $p<0.001$). For both GOL and TOC treatment, no baseline differences in disease activity between cohorts with monotherapy and MTX combination could be shown. The combination with MTX led to significantly lower rates of discontinuation due to inefficacy ($p<0.05$) with GOL and TOC (Figure 1).

Conclusion: Patients with pJIA mostly were treated with a combination of the biologic and MTX rather than with biologic monotherapy. Treatment was discontinued due to lack of efficacy in 14% to 28%. No statistically significant effect of combination treatment with MTX versus monotherapy could be observed regarding the rate of treatment failures in patients treated with ETA or ADA. However, combination treatment with MTX significantly prolonged the survival of GOL and TOC in patients with polyarticular JIA. The results are limited by low patient numbers in the GOL cohort and possible bias by JIA category.

Figure 1. Kaplan Meier plot of drug survival in patients with monotherapy or with combination with MTX of the indicated biologic

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POS1302 THE MUSCULOSKELETAL SYSTEM MANIFESTATIONS IN CHILDREN WITH FAMILIAL MEDITERRANEAN FEVER

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Background: Familial Mediterranean fever (FMF) is a monogenic inherited periodic fever syndrome presenting with episodes of self-limiting fever and inflammation of serosal membranes. The attacks emerge with arthritis were defined as one of the major diagnostic criteria besides involvement of serosal membranes. Non-specific musculoskeletal findings such as myalgia, arthralgia, transient synovitis, and more rare manifestations like protracted febrile myalgia can also be seen in FMF patients attacks

Objectives: We aim to reveal the frequency and genotype association of musculoskeletal manifestations in children with FMF.

Methods: The patients diagnosed with FMF between January 1, 2017 and June 1, 2019, and followed for at least 6 months in our pediatric rheumatology clinic were included in the study. Musculoskeletal manifestations of patients were

enrolled. The patients were grouped according to the "Mediterranean Fever" (MEFV) gene variants. Musculoskeletal manifestations of the patients were compared between the groups

Results: The study group included 634 children with FMF (336 female and 298 male, F/M: 1.13/1). The clinical manifestations of patients in attack period were as follows: 99% of the patients had fever, 87.3% had abdominal pain, 20.7% had chest pain, 11.3% had vomiting, 10.7% had erysipelas like erythema, and 9.3% had headache. The musculoskeletal symptoms were accompanied by 58.6% (n: 372) of the patients during the attack period. The most common musculoskeletal manifestation was found as arthralgia (32.6%, n: 206). Also, the other musculoskeletal manifestations were found as follows during attacks; arthritis in 23.7% (n: 150), myalgia in 20.5% (n: 130), exertional calf pain in 6.5% (n: 41), and protracted febrile myalgia in 1% (n: 7) of the patients. It was observed that the musculoskeletal manifestations were significantly higher in patients with homozygous M694V variant in exon-10 ($p=0.017$). Also, it was found that the musculoskeletal manifestations are more common in the attack periods of patients carrying the M694V variant in at least one allele ($p = 0.019$).

Conclusion: It was determined that the musculoskeletal manifestations were seen as an attack symptom in more than half of FMF patients. Also, homozygous and compound heterozygous MEFV mutations including M694V variant found as a risk factor for emerge of musculoskeletal manifestations. In children with unexplained and recurrent musculoskeletal symptoms, especially in ethnicities with the high frequency of FMF, analysis of MEFV gene can help reveal the underlying cause.

REFERENCES:

- [1] Brik R, Shinawi M, Kasinetz L, Gershoni-Baruch R. The musculoskeletal manifestations of familial Mediterranean fever in children genetically diagnosed with the disease. *Arthritis Rheum* 2001;44:1416-9.
- [2] Jarjour RA, Dodaki R. Arthritis patterns in familial Mediterranean fever patients and association with M694V mutation. *Mol Biol Rep* 2011;38:2033-6.

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POS1303 EXPERIENCE WITH ADALIMUMAB BIOSIMILAR USE IN CLINICAL PRACTICE: DATA FROM THE GERMAN BIKER-REGISTRY

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Background: In 2017, Adalimumab Biosimilars became approved. Comparative studies to the originator have been performed in adult patients with rheumatoid arthritis, ankylosing spondylitis and psoriasis and extrapolation led to approval for juvenile idiopathic arthritis (JIA).

Objectives: So far there is limited experience with biosimilars in JIA: The large national data base of the BIKER-registry was used to describe experience with Adalimumab biosimilars in clinical practice

Methods: This retrospective analysis used data of the German BIKER-registry. The data basis was screened for patients exposed to Adalimumab. Subcohorts with initiation of treatment after 2017, use of the originator and of biosimilars were built. The course of JADAS10, Physician global assessment VAS 0–100-mm, Parent/patient global assessment VAS 0–100-cm, Active joint count 0–71, truncated at 10, ESR and CHAQ-DI was analyzed. Descriptive statistics was used for demographic, clinical data, drug exposure, adverse events (AEs) and events of special interest (ESI).

Results: Until 31.10.2020, 1173 JIA patients were reported to have received Adalimumab. 352 treatments have been started after January 1, 2017. A biosimilar was used first line in 44 patients. Further 55 patients switched for the originator to a biosimilar. 2 patient switched from a biosimilar to the originator. 3 patients switched to a second biosimilar while 5 patients who switched from the originator to a biosimilar reswitched back to the originator.

After 2017, 33 pediatric rheumatology centres reported initiation of Adalimumab treatment. 17 have used a biosimilar. 15 centres have switched at least 1 patient from the originator to a biosimilar and 14 have used first line a biosimilar in at least 1 patient. In a single centre, initiation of a biosimilar was used more frequently (8 versus 7).

The patients' characteristics and disease activity parameters were brightly comparable. The JIA category rheumatoid factor (RF) negative polyarthritis was less frequent in the biosimilar first cohort while RF positive polyarthritis and psoriatic arthritis was more frequent. In patients with idiopathic uveitis the originator was used more often. In the switching cohort, more patients had RF negative polyarthritis, persistent oligoarthritis but less had psoriatic arthritis and no had RF positive polyarthritis.

No difference in disease activity parameters between patients receiving the originator or biosimilars were noted, neither at baseline, during the course of treatment nor at last observation upon treatment (Figure 1). At the time of switching, 46 (92%) had JADAS minimal disease activity (MDA) and 30 (69%) were in JADAS remission. At last observation, those numbers were comparable with 42 (86%) with JADAS MDA and 28 (57%) with JADAS remission.

In total, 45 adverse events (AE) were reported in 45 patients upon biosimilar treatment. 26 patients had 1, 12 patients had 2 and 6 patients reported 3 and 1 reported 4 events. Adverse event of special interest were Infection associated leukopenia (n=1), COVID 19 infection (n=1), Uveitis flare (n=8), other disease deterioration (arthritis flare) (n=20), injection site reaction n=2. A single serious AE was reported. A 16 year old female adolescent was admitted for unexpected CK elevation. In 10 patients, Adalimumab was discontinued, in 2 it was temporarily paused.

Conclusion: This article is the first attempt to present a large sample of data on JIA patients exposed to Adalimumab biosimilars. Since approval of Adalimumab-Biosimilars, limited experience from clinical practice is available. Biosimilars are used in a minority of patients and by a minority of centers although no difference in efficacy or safety was noted from our analysis.

JADAS 10

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POS1304

JUVENILE SYSTEMIC SCLEROSIS (JSSC) PATIENTS WITH OVERLAP CHARACTERISTICS DO NOT HAVE MILD DISEASE. RESULTS FROM THE JSSC INCEPTION COHORT. WWW.JUVENILE-SCLERODERMA.COM

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Background: Juvenile systemic sclerosis (jSSc) is an orphan disease with a prevalence of around 3 in 1, 000,000 children. It is known that in pediatric jSSc cohorts, there are a significant number of patients with overlap features, such as arthritis and myositis. However, the disease burden between those with and without overlap features in jSSc has not been defined.

Objectives: Compare the clinical phenotype between children with and without overlap features in the juvenile systemic sclerosis inception cohort (jSScC).

Methods: A cross-sectional study was performed using baseline visit data. Demographic, organ system evaluation, autoantibody profile, treatment, and patient and physician reported outcome variables were extracted from jSScC. Comparison between patients with and without overlap features was performed using chi-square test and Mann Whitney U-test.

Results: At the time of data extraction, 175 jSSc patients were enrolled in the cohort, 81% were Caucasian and 81% female. Mean disease duration was 3.1 year (±2.7). Mean age at Raynaud's onset was 10 years (±3.8) and mean age of first non-Raynaud's was 10.2 years (±3.8). Overlap features occurred 17%

(n=30) of the cohort, 12.5% in the diffuse cutaneous (dc) jSSc and in 30% in the limited cutaneous (lc) jSSc. Significant differences in clinical characteristics were found between those patients with compared to without overlap characteristics. Patients with overlap features presented more frequently with Gottron papules (p=0.007), swollen joints (p=0.019), muscle weakness (p=0.003), and lung involvement documented by decreased DLCO < 80% (p=0.06) and/or abnormal high resolution computed tomography (p=0.049). Anti-PM/Scl autoantibodies were also more common in this group (p=0.001). Significantly more patients without overlap features had Raynaud's (p=0.006). Physician Global Assessment of disease activity was significantly higher in patients with overlap features (41 vs 34; p=0.041). (Table 1.)

Table 1. Demographic and clinical characteristics of jSSc patients with and without overlap features.

	Whole Cohort N=175	Patients without overlap N=145	Patients with overlap N=30	P value
Female to Male Ratio	4.3:1 (142/33)	4:1 (116/29)	6.5:1 (26/4)	0.395
Cutaneous subtype				
Diffuse subtype (N)	73% (128)	112	16	
Limited subtype (N)	27% (47)	33	17	
Mean disease duration (years)	3.1 (± 2.7)	3.2 (± 2.8)	3.1 (± 2.2)	0.291
Mean age of onset of Raynaud's (years)	10.0 (± 3.8)	10.0 (± 3.8)	10.0 (± 3.7)	0.931
Mean age of onset of non-Raynaud's (years)	10.2 (± 3.8)	10.2 (± 3.9)	9.8 (± 3.7)	
Disease modifying drugs (N)	88% (154)	89% (129)	83% (25)	0.388
Raynaud's phenomenon	90% (158)	93% (135)	77% (23)	0.006
Anti-PM/Scl	18% (12/68)	9% (5/53)	47% (7/15)	0.001
Gottron Papules (N)	27% (46/171)	23% (33/144)	48% (13/27)	0.007
DLCO <80% (N)	44% (39/88)	39% (28/71)	65% (11/17)	0.06
Abnormal findings in HRCT (N)	44% (59/133)	40% (43/107)	62% (16/26)	0.049
Proportion of patients with swollen joints	18% (32)	14% (21)	37% (11)	0.019
Muscle Weakness (N)	21% (31/149)	16% (20/123)	42% (11/26)	0.003
Physician global disease activity (0-100) min -max	35 (0-90) n=141	34 (0-90) n=114	41 (0-80) n=27	0.041

Conclusion: Results from this large international cohort of jSSc patients demonstrate significant differences between patients with and without overlap features. Patients with overlap have significantly more interstitial lung disease and more physician rated disease activity and should not be considered to have more "mild disease."

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POS1305

TREATMENT RESPONSE TO TUMOR NECROSIS FACTOR INHIBITORS IN ADULTS WITH JUVENILE IDIOPATHIC ARTHRITIS: DATA FROM THE NOR-DMARD STUDY

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Background: Juvenile idiopathic arthritis (JIA) can cause considerable pain and disability in childhood and adulthood. Studies exploring the efficacy of medications in adult JIA patients are limited, although tumor necrosis factor inhibitors (TNFi) have been increasingly used in this patient group.

Objectives: To explore the efficacy of TNFi ± comedication on disease activity in adult JIA patients, compared to a weighted rheumatoid arthritis (RA) cohort.

Methods: Data from NOR-DMARD, a longitudinal observational study including patients > 18 years starting or switching DMARD treatment, was used [1]. Patients with a clinical JIA diagnosis, or patients with other inflammatory joint diseases diagnosed before 16 years were identified from the study population. RA patients were included for comparative purposes.

Disease activity measurements and remission rates among patients starting treatment with TNFi ± comedication were collected at baseline, 3 and 6 months.