THU0517

THE LONGITUDINAL EUROFEVER PROJECT: AN UPDATE ON ENROLLMENT

Ilaria Gueli, Martina Finetti, Fabrizio De Benedetti, Jordi Anton, Maria Alessio, Joost Frenkel, Luca Cantarini, Romina Gallizzi, Judith Sanchez Manubens, Marco Cattalini, Efimia Papadopoulou-Alataki, Rolando Cimaz, Donato Rigante, Alma Nunzia Olivieri, Pavla Dolezalova, Alberto Martini, Nicolino Ruperto, Marco Gattorno. *IRCCS Istituto Giannina Gaslini, UOSD Centro Malattie Autoinfiammatorie e Immunodeficienze, on the behalf of the Paediatric Rheumatology International Trials Organisation (PRINTO) and the Eurofever Project, Genoa, Italy*

Background: In 2008 the Paediatric Rheumatology European Society (PReS) promoted an International Project for the study of Autoinflammatory Diseases (AIDs) named Eurofever, whose main purpose is to create a web-based registry for the collection of information in AIDs patients.

Objectives: To implement the Registry with the new recently described AIDs and to increase the collection of longitudinal data.

Methods: The data were extracted from the Eurofever registry, which is hosted in the PRINTO website (<http://www.printo.it>). From February 2015 we started the longitudinal collection of follow-up data with particular focus on treatment, modification of the clinical picture, onset of complication/adverse events. We have enrolled patients included in the registry up to 28 September 2018.

Results: Up to date 4175 patients have been enrolled (3843 of them with complete demographic information, 1903 M e 1940 F) from 62 countries. For 3356 (87%) patients also clinical data from onset to diagnosis, collected during the first visit performed at referred pediatric or adult center, are available. For each disease the number of enrolled patients is: FMF 1086 pts (951 with complete clinical data); TRAPS 273 pts (256 complete); CAPS 298 pts (279 complete); MKD 205 pts (190 complete); Blau's disease 49 pts (26 complete); PAPA 42 pts (41 complete); NLRP12 mediated periodic fever 13 pts (11 complete); DADA2 14 pts (9 complete); DIRA 3 pts (all complete); SAVI 3 pts (all complete); CANDLE 1 pt (complete) and Majeed 4 pts (all complete). Among multifactorial autoinflammatory diseases: PFAPA 676 pts (551 complete); CNO 581 pts (540 complete); Behcet 214 pts (186 complete), undefined periodic fever 368 pts (292 complete) and Schnitzler 13 pts (all complete). The median onset age is 4 years (range 1 month – 75 years), the median diagnosis age is 8 years (range 1 month – 78 years). Most of patients (3509, 91%) presented disease onset during pediatric age (<16 years), 334 (9%) during adult age (81 FMF, 31 CAPS, 53 TRAPS, 40 CRMO, 12 Schnitzler syndrome e 90 unknown fever). 405 of 3509 (12%) patients with pediatric onset received diagnosis during adult age. The median diagnostic delay is 5 years; diseases with longer diagnostic delay are: NLRP12 (24 years, range 4-76), CAPS (15 years, range 0-77), PAPA (14 years, range 2-57), TRAPS (12 years, range 0-77). 396 patients have been treated with at least one biologic drug, 1031 with DMARDs, 427 with systemic steroid and 686 with others drugs. The most frequent diseases treated with biologic drugs are: CAPS (38%), multifactorial diseases (22%), TRAPS (14%), MKD (11%), rare monogenic (8%: 1 CANDLE, 2 DIRA, 2 NALP12, 3 Majeed, 8 DADA2 and 14 PAPA), and FMF (7%). Since February 2015, longitudinal visits have been inserted for 477 (12%) patients, with detailed data on treatment and safety.

Conclusion: The enrollment in Eurofever Registry is still ongoing. The analysis of data will improve our knowledge both on the natural history of the single disease and on the efficacy/safety of treatment commonly used in the clinical practice.

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AbbVie, Biogen, Boehringer Ingelheim, Bristol-Myers Squibb, EMD Serono, Janssen, Novartis, Pfizer, R-Pharm.

The money received for these activities are directly transferred to the Gaslini Institute's bank account. Before March 2016, I was the head of the Pediatric Rheumatology Department at the G. Gaslini Hospital, where the PRINTO Coordinating Centre is located. For the coordination activity of the PRINTO network, the Gaslini Hospital received contributions from the industries listed in this section. This money has been reinvested for the research activities of the hospital in fully independent manners besides any commitment with third parties., Nicolino Ruperto Grant/research support from: The Gaslini Hospital, where NR works as full-time public employee, has received contributions (> 10.000 USD each) from the following industries in the last 3 years: BMS, Eli-Lilly, GlaxoSmithKline, F Hoffmann-La Roche, Janssen, Novartis, Pfizer, Sobi. This funding has been reinvested for the research activities of the hospital in a fully independent manner, without any commitment with third parties., Consultant for: Received honoraria for consultancies or speaker bureaus (< 10.000 USD each) from the following pharmaceutical companies in the past 3 years: Ablynx, AbbVie, Astrazeneca-Medimmune, Biogen, Boehringer, Bristol-Myers Squibb, Eli-Lilly, EMD Serono, GlaxoSmithKline, Hoffmann-La Roche, Janssen, Merck, Novartis, Pfizer, R-Pharma, SanofiServier, Sinergie, Sobi and Takeda., Speakers bureau: Received honoraria for consultancies or speaker bureaus (< 10.000 USD each) from the following pharmaceutical companies in the past 3 years: Ablynx, AbbVie, Astrazeneca-Medimmune, Biogen, Boehringer, Bristol-Myers Squibb, Eli-Lilly, EMD Serono, GlaxoSmithKline, Hoffmann-La Roche, Janssen, Merck, Novartis, Pfizer, R-Pharma, SanofiServier, Sinergie, Sobi and Takeda., Marco Gattorno Grant/research support from: MG has received unrestricted grants from Sobi and Novartis

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THU0518

EFFICACY AND SAFETY OF ETANERCEPT IN JUVENILE IDIOPATHIC ARTHRITIS – 18 YEAR EXPERIENCE USING DATA OF THE BIKER REGISTRY

Ariane Klein¹, Gerd Ganzer¹, Michael Rühlmann¹, Frank Dressler¹, Toni Hospach¹, Kirsten Minden¹, Ralf Trauzeddel¹, Boris Huegle¹, Sonja Mrusek¹, Ivan Foeldvari¹, Jasmin Kuemmerle-Deschner¹, Frank Weller-Heinemann¹, Andreas Urban¹, Gerd Horneff². ¹BIKER Registry, Sankt Augustin, Germany; ²Paediatric Rheumatology Centre, General Paediatrics, Sankt Augustin, Germany

Background: Since 2000, Etanercept is approved for polyarticular juvenile idiopathic arthritis (JIA). Subsequently, approval has been extended to additional JIA categories.

Objectives: To describe efficacy and safety of Etanercept in clinical practice in JIA patients in comparison to a biologic-naïve JIA cohort using the German Biologics registry (BiKEr).

Methods: Baseline demographics and disease activity parameters were documented. Efficacy was determined using the JADAS10. Safety assessments were based on adverse events reports (AE) processed according to MedDRA.

Results: Until October 1, 2018, 2645 JIA patients treated with Etanercept were registered, representing 5820 patient-years (PY) of exposure. The total observation time (from date of first dose until last follow-up, censored, if another biologic was started) was calculated as 8080 PY.

In general, the cohort treated with Etanercept had experienced disease duration of 4.2±/3.7 years (mean±/SD). 2303 patients (87%) were pre-treated with methotrexate, 100 with alternative biologics. Concomitantly, 1822 patients (69%) received methotrexate, 2098 (79%) NSAIDs, 949 (36%) systemic corticosteroids as well as further drugs in lower numbers. In the control cohort, 1517 biologic naïve JIA patients started methotrexate.

At last follow-up, 68%/62%/50%/34% of patients reached JIAACR30/50/70/90 criteria. The median (IQR1-3) JADAS10 score decreased from 15.0 (10-20.4) at baseline to 3.5 (0.7-10.1). 60% of patients achieved a JADAS defined minimal disease activity, 40% reached a JADAS remission.

1924 AE were observed (33.1/100PY [95% CI 31.6-34.6]). 221 qualified as SAE (3.8/100PY [3.3-4.3]). These figures were compared to 1292 AE reported during 3714PY in the control cohort (34.8/year [32.9-36.8]) and 52 SAE (1.4/100PY [1.1-1.8]).

The most frequent SAE in the Etanercept cohort were grouped in the MedDRA SOC infections and infestations (n=54) followed by musculoskeletal and connective tissue disorders (n=27), eye disorders (n=21), gastrointestinal disorders (n=17) and nervous system disorders (n=15). There were 5 deaths. Serious infections were of bacterial and viral origin and mostly occurred once only.

Adverse events of special interest were uveitis (n=109), H. zoster (n=24), chronic inflammatory bowel disease (n=19), pregnancies (n=4), depression (n= 11), malignancies (n=8), demyelination (n=2). Adverse Events of Special Interest (AESI) were in the MTX-cohort uveitis (n=52), H. zoster (n=4), chronic inflammatory bowel disease (n=1), depression (n= 2), malignancies (n=4).

A total of 1606 patients (60.7%) discontinued treatment. Of these 638 (39.7%) discontinued due to remission, 565 (35.2%) due to lack of efficacy and 188 (11.7%) because of intolerance.

Conclusion: The current analysis adds to the established safety profile of Etanercept and demonstrates that the rate of SAEs is comparable and consistent with the overall AE profile in paediatric patients. As expected, infections are the most frequent SAE. Uveitis is likely associated with JIA. Of notice, few patients developed a chronic inflammatory bowel disease and some a malignancy. No new safety signals specific to the paediatric population were identified in this large cohort of JIA patients.

REFERENCES:

- Geikowski T, Becker I, Horneff G; German BIKER Registry Collaborative Study Group. Predictors of response to etanercept in polyarticular-course juvenile idiopathic arthritis. *Rheumatology* (Oxford). 2014 Jul;53(7):1245-9

Efficacy measure: JIAACR-Score:

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THU0519 DISEASE COURSE AND OUTCOMES OF PREGNANCY IN JUVENILE IDIOPATHIC ARTHRITIS: ANY DIFFERENCE AMONG DISEASE SUBSETS?

Antía García Fernández¹, Laura Andreoli^{2,3}, Maria Chiara Gerardi^{2,3}, Francesca Crisafulli^{2,3}, Cecilia Nalli², Matteo Filippini², Roberto Gorla², Marco Taglietti², Micaela Fredi^{2,3}, Maria Grazia Lazzaroni^{2,3}, Andrea Lojcono^{3,4}, Sonia Zatti⁴, Franco Franceschini^{2,3}, Angela Tincani^{2,3}. ¹Ramón y Cajal University Hospital, Rheumatology, Madrid, Spain; ²Spedali Civili of Brescia, Rheumatology and Clinical Immunology, Brescia, Italy; ³University of Brescia, Brescia, Italy; ⁴Spedali Civili of Brescia, Obstetrics and Gynecology, Brescia, Italy

Background: Data on the disease activity and pregnancy outcomes taking into account the different types of Juvenile idiopathic arthritis (JIA) patients are limited.

Objectives: To analyze the course of disease during pregnancy and pregnancy outcomes in patients with JIA according to disease subtype, with focus on the medication use.

Methods: A retrospective observational study was conducted on pregnant JIA patients followed in our pregnancy clinic between 2010 and 2018. Disease activity (DAS28-PCR3) was analyzed before conception, at each trimester and postpartum. Flare was defined as an increase in clinical activity leading to intensified therapy with at least one of: new prednisone, increase in dosage ≥ 5 mg/day, treatment with intraarticular glucocorticoids, (re-)treatment with DMARDs/TNFi. Pregnancy outcome were also recorded.

Results: Sixteen patients and 22 pregnancies were included. Two pregnancies were voluntary interrupted (1 for Methotrexate exposure and 1 for fetal encephalocele). Table 1 summarizes disease features and pregnancy outcomes according to disease subsets. 10/22 (45,5%) were pregnancies observed in Oligoarticular Extended (OLA-E), 6 (27,3%) Polyarticular (PLA), 4 (18,2%) Systemic (SYS), 1 (4,5%) Oligoarticular (OLA) and 1 (4,5%) Enthesitis related Arthritis (ERA) JIA patients.

Abstract THU0519 –Table 1.

Table 1	Polyarticular	Oligoarticular	Oligoarticular extended	Systemic	Enthesitis related arthritis
Patients, n(%)	5(31.2)	1(6.25)	6 (37.5)	3 (18.8)	1 (6.25)
Pregnancies, n(%)	6 (27.3)	1 (4.5)	10 (45.5)*	4 (18.2)	1 (4.5)
Age at pregnancy (years) mean+SD, yrs	33 (5.51)	28 (0)	27.8 (5.63)*	24.5 (6.25)	26 (0)
Disease duration, months, mean+SD,	20.5 (2.6)	22 (0)	19.5 (6.2)*	19.5(6.5)	19 (0)
CCS before conception, n (%), daily dose, mg/day, mean+SD	4 (66.7) 6.6 (5.82)	0 (0) 0 (0)	3(33.3)* 4.5 (0.86)*	0 (0) 0 (0)	0 (0) 0 (0)
cDMARDs before conception					
None (%)	5 (83.3)	0 (100)	6 (60)*	0 (100)	0 (100)
Ciclosporin (%)	1 (16.7)	0	0 (0)	0	0
Hydroxychloroquine (%)	0	0	3 (30)*	0	0
Methotrexate (%)	0	0	1 (10)*	0	0
bDMARDs before conception					
None (%)	1 (16.7)	0 (100)	5 (50)	0 (100)	0 (100)
ETANERCEPT (%)	3 (50)	0	5 (50)*	0	0
GOLIMUMAB (%)	1 (16.7)	0	0	0	0
ABATACEPT (%)	1 (16.7)	0	0	0	0
Stop bDMARDs at positive pregnancy test, n (%)	4 (80)	0 (0)	4 (80)*	0 (0)	0 (0)
DAS28PCR, mean+SD					
PC	1.69 (0.42)	1.15 (0)	1.73 (0.45)	1.27 (0.25)	1.15 (0)
1T	2.47 (1.36)	1.64 (0)	1.82 (0.47)	1.53 (0.31)	1.64 (0)
2T	3.11 (1.05)	1.64 (0)	2.29 (0.53)	1.56 (0.30)	2.00 (0)
3T	2.84 (1.61)	1.64 (0)	1.94 (0.29)	1.71 (0.10)	1.99 (0)
PP	2.66 (1.13)	Not available	2.85 (0.71)	1.59 (0.14)	1.60 (0)
Flare, n (%)					
1T	1/7 (14.3)	0/7 (0)	1/7 (14.3)	0/7 (0)	0/7 (0)
2T	3/7 (43)	0/7 (0)	1/7 (14.3)	0/7 (0)	0/7 (0)
3T	1/7 (14.3)	0/7 (0)	1/7 (0)	0/7 (0)	0/7 (0)
PP	2/8 (25)*	Not available	6/8 (75)*	0/8 (0)	0/8 (0)
Pregnancy outcome					
Live-births, n (%)	5 (83.3)		7 (77.8)*	1 (100)	1 (100)
Ongoing pregnancies, n (%)	1(16.7)		0 (0)	0 (0)	0 (0)
Spontaneous pregnancy loss n (%)	0 (0)	No available	0 (0)	0 (0)	0 (0)
VPI, n (%)	0 (0)		2 (22.2)*	0 (0)	0 (0)
P-PROM(%)	1 (16.7)		0 (0)	0 (0)	0 (0)
Delivery week, median min-max	36 (33-41)		38 (35-39)	41 (38-41)	41 (0)
Premature delivery, n (%)	2 (33.3)		3 (37.5)	0 (0)	0 (0)

cDMARDs conventional disease-modifying antirheumatic drug, bDMARDs biological disease-modifying antirheumatic drug. 1T, First trimester, 2T Second trimester, 3T Third trimester, PC preconception, PP postpartum, CCS corticosteroids, VPI voluntary pregnancy interruption, PROM premature rupture of membranes.

* Methotrexate exposure during 1stT at 8W and fetus malformation at 17W.

* Taking into account voluntary pregnancy interruption.

At conception 19(95%) pregnancies were on remission, 1(5%) OLA-E patient in low disease activity. Among the 20 pregnancies ended with a live birth, 1 OLA, 4 SYS and 1 ERA patients maintained sustained remission, without cDMARDs or bDMARDs, throughout pregnancy.

There were 7 flares in 6 (31,8%) pregnancies, 5 (71,4%) of them among PLA patients and 2 (28,6%) among OLA-E patients. No extrarticular flare was recorded. There was no flare among SYS, OLA and ERA patients. One pregnancy in a patient with PLA JIA (4,5%) was characterized by more than one flare, but none had more than two flares. Most of flares (2/29% 1sttrimester, 4/57% 2ndtrimester, 1/14% 3rdtrimester) occurred during 2ndtrimester. Eight flares occurred in postpartum, also in 2 pregnancies after voluntary pregnancy interruption.

Regarding treatment at conception; 8 (40%) pregnancies were on bDMARDs, treatment was stopped at positive pregnancy test in 6 pregnancies, 2 (33,3%) required restart of treatment due to a flare in 1stTrimester (1 CTZ) and 3rdTrimester (1 ETA), and 3 flared in puerperium with restart of bDMARDs. Two (25%) pregnancies continued bDMARDs at positive pregnancy test throughout the pregnancy to prevent a flare, that had been already experienced in previous pregnancies (not followed in our centre).

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

Giovanni Civiari¹, Biagio Castaldi², Giorgia Martini¹, Ornella Milanese², Francesco Zulian¹. ¹Pediatric Rheumatology, Department of Woman's and Child's Health, University of Padova, Padova, Italy; ²Pediatric Cardiology, Department of Woman's and Child's Health, University of Padova, Padova, Italy

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.

REFERENCES:

- [1] Martini G et al. Arthritis Rheum 2006; 54:3971-3978.
- [2] Spethmann S et al. Eur. Heart J Cardiovascular Imaging 2012; 13:863-870.
- [3] Dedeoglu R et al. Pediatric Cardiol 2017; 38:1686-1695.
- [4] La Torre F et al. Arthritis Rheum 2012; 64:4143-4150.
- [5] Geyer H et al. J Am Soc Echocardiogr 2010; 23:351-369.

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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

Claudia Sengler¹, Svea Horn¹, Nadine Grösch¹, Jens Klotsche¹, Martina Niewerth¹, Fabian Speth², Peter Haas³, Claas Hinze⁴, Gerd Horneff⁵, Tilmann Kallinich⁶, Frank Weller-Heinemann⁷, Nadine Unterwalder⁸, Kirsten Minden¹. ¹German Rheumatism Research Center, Berlin, Germany; ²University of Rostock, Rostock, Germany; ³German Center for Pediatric and Adolescent Rheumatology, Garmisch-Partenkirchen, Germany; ⁴University of Münster, Department of Pediatric Rheumatology and Immunology, Münster, Germany; ⁵Center for General Pediatrics and Neonatology, Asklepios Klinik, Sankt Augustin, Germany; ⁶Charité – Universitätsmedizin Berlin, Department of Pediatric, Division of Pneumology and Immunology, Berlin, Germany; ⁷Prof.-Hess-Kinderklinik, Berlin, Germany; ⁸Labor Berlin, Berlin, Germany

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-TIF1 γ 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-TIF1 γ , 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