In patients total and active patients, eotaxin correlated with DLCO% (0.39 and 0.52). In inactive patients, MIP1b correlated with DLCO% (0.47). In patients total, IP-10 correlated with any HRCT finding and airways disease (0.37 and 0.44); which were also present in active patients (0.34 and 0.61). In patients total, MCP-1 correlated with any HRCT finding and calcinosis (0.34 and 0.40); in active patients MCP-1 and calcinosis correlated (0.34). Eotaxin correlated with ILD in both patients total and active patients (0.37 and 0.38).

The following cytokines correlated with HRCT findings in inactive patients only; IL-17 with any HRCT findings and ILD (0.61 and 0.52), MIP1a with any HRCT findings (0.63) and IL-5 with ILD (-0.42).

Conclusion: In JDM, lung involvement was associated with mainly proinflammatory cytokines in active and profibrotic/proinflammatory cytokines in inactive patients. The association between higher eotaxin and better gas diffusion was interesting, since we have previously demonstrated an association between higher eotaxin and cardiac dysfunction (Ref 2).

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OP0257

CHILDHOOD-ONSET MONOGENIC SYSTEMIC LUPUS ERYTHEMATOSUS NOT DUE TO COMPLEMENT DEFICIENCY: A SYSTEMATIC REVIEW OF 90 CASES

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Background: The paediatric SLE (pSLE) are estimated 10 to 17% of SLE (1). Studies comparing pSLE with adult-onset SLE reported high prevalence of pre-existing organ damage at diagnosis due to the diagnosis delay, more severe disease clinical course with high disease activity resulting in worse prognosis in pSLE (1,2). Recently, whole exome sequencing and next-gene sequencing have facilitated the identification of rare monogenic variants associated with SLE and lupus-like phenotypes with high penetrance (3,4)

Objectives: To reporting the whole spectrum of monogenic pSLE according to pathway abnormalities, using a meta-analysis (MA) approach.

Methods: A systematic review of studies focused on monogenic pSLE were analysed. To be included in the systematic review, studies had to meet simultaneously all the following criteria: (i) reported at least 1 case of SLE (ii) started before the age of 16 (iii) with clearly description of the diagnosis criteria and (iv) provided genetic data of the case.

We pooled the genes into three main groups of signalling pathways: (i) genes that affect adaptive immunity pathways including the lymphocyte apoptosis (FAS, KRAS, PTPN11, SHOC2) and the B and T cell development checkpoints (PRKCD, RAG2, IKZF1, DOCK8, WASP), (ii) genes containing type I interferon (IFN) pathways (ACP5, TREX1, SAMHD1, RNASEH2A, RNASEH2B, RNASEH2C, IFIH1, DNASE1L3, DNASE1, TMEM173, Tetrasomy 9), and (iii) metabolism pathways including both amino acid metabolism (PEPD, SLC7A7, MAN2B1) and oxidative metabolism (NCF2). Clinical and laboratory data were analysed according to the SLICC classification.

Fig 1: The PRISMA 2009 flow diagram of literature searched process for the study.

Results: We included 42 articles encompassing 88 patients, plus 2 additional unpublished cases of one author (AB). The MA process were carried out according to the PRISMA statement.

According to the SLICC classification, the blood disorders were significantly higher in the group of the metabolism pathway as well as the positivity of direct

Coombs test. The positivity of antibodies anti-Sm was also a distinguishing element for this group, while the positivity of ALP antibodies was the lonely manifestation to distinguish the adaptive immunity pathway from the others. Fever, recurrent infections, hepatosplenomegaly, and pulmonary diseases were significantly predominant in the metabolism pathways. In the group of the IFN pathway, the most distinctive criteria were the neurological manifestations and bone changes. There were no statistically significant differences in all groups concerning all other clinical or immunological criteria.

Conclusion: This is the first quantitative analysis of multiples cases reports, comparing the whole spectrum of monogenic pSLE according to three different pathways. Our results suggest that abnormalities in signaling pathway delimitate different SLE juvenile subset. The clinical and biological differing between the three signaling pathway were not specific, but our results may help to detect phenotypes requiring targeted genetic investigations. Subsets of SLE syndrome have to be identified to drive the treatment and propose personalized medicine.

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OP0258

LESSON FROM EUROFEVER REGISTRY AFTER THE FIRST TEN YEARS OF ENROLLMENT

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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 assess the impact of the Eurofever Registry on scientific community with particular interest in the geographical coverage, diagnostic delay, access to treatment and publications.

Methods: The data analyzed in the study were extracted from the Eurofever registry, which is hosted in the PRINTO website.

Results: Up to date 4175 patients have been enrolled from 62 countries (3843 of them with complete demographic data). Most of patients (72%) are resident in Western Europe, 8% in Central-Eastern Europe, 11% in Southern-Eastern Mediterranean, 2% in South America and 7% in other countries. Compared to the first Eurofever report (Toplak et al, 2012) we have observed an increase of enrolled patients from 1388 to 2651 in Western Europe, from 106 to 313 in Central-Eastern Europe, from 294 to 406 in Southern-Eastern Mediterranean. The median onset age is 4 years (range 1 month – 75 years), the median diagnosis age is 8 years (range 1 month – 78 years). The median diagnostic delay observed in 2012 was 7.3 years (range 0.3–76), from patients enrolled after 2012 it was 1.9 years (range 0–57). Comparing the mean diagnostic delay from 1980 to 2018, we have observed an encouraging constant reduction of period between AIDs onset e diagnosis (from a mean diagnostic delay value of 20 years for patients born before 1980, to a mean value of 1 year for patients born after 2011, Figure 1). Complete information on access to treatment were available in 2430 patients. DMARDs were used in 1031 (42%), biologics in 396 (16%) patients. According to the number of enrolled patients, biologics were used in 361/1782 (20%) of Western Europeans, 17/342 (5%) of Central-Eastern Europeans, 6/259 (2%) of Southern-Eastern Mediterranean patients. Regarding Eurofever impact on Scientific Community, during this first 10 years the Registry provided 12 papers with more than 800 citations. Detailed analysis of clinical features collected in Eurofever database allowed to perform studies with large cohort of patients, to purpose new classification criteria (Federici et al, 2015), to validate damage and activity score (Piram et

al, 2014 and N Ter Haar et al, 2017) and to evaluate genotype/phenotype correlation (Papa et al, 2017).

Figure 1. Diagnostic delay (years) according to the year of birth

Conclusion: In the last years we have observed an encouraging increase of involved Countries, with a greater number of patients coming from geographic area poorly represented in the first epidemiologic study of Toplak et al. Eurofever data analysis has confirmed an improvement of diagnostic ability during the last years, with a significant reduction of mean diagnostic delay. Longterm studies will help understand the efficacy and safety of different treatments used in these rare conditions.

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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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OP0259

OVERWEIGHT AND OBESITY IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS ENROLLED IN THE GERMAN NATIONAL PAEDIATRIC RHEUMATOLOGIC DATABASE (NPRD)

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Background: Patients with juvenile idiopathic arthritis (JIA) may have a different body composition associated with reduced muscle mass and increased fat mass [1]. They display decreased physical fitness, perform less strenuous physical activities, and spend more time sleeping than do healthy children. A lower level of physical activity is associated with deconditioning and functional deterioration, favoring an inactive lifestyle. The risk of overweight might be further increased by the glucocorticoid treatment.

Objectives: Since obesity can increase inflammatory processes, cause early atherosclerotic changes and promote metabolic disorders, the objectives were a) to determine the prevalence of overweight and obesity in children and adolescents with JIA, and b) to examine the association between overweight and health-related parameters in this population.

Methods: A cross-sectional analysis of physicians' recorded body weights and heights of patients with JIA enrolled in the NPRD in the year 2016 was performed. Overweight was defined as BMI >90th sex- and age-specific percentile and obesity as BMI >97th percentile. For comparison with data from the general German population [2], patients aged 3 to 17 years were considered. A linear regression model was used to explore the association between overweight and both clinical as well as self-reported outcomes.

Results: In total, data from 6.860 children and adolescents with JIA (age 11.5 ± 4 years, disease duration 4.6 ± 3.6 years, 67% girls, 39% persistent oligoarthritis) were analyzed. Overweight was found in 14% (including 6% obesity) of JIA cases. Comparative data from the German general population report an overweight prevalence of 15% (including 6% obesity). In contrast to the general population, overweight rates in JIA differed between girls and boys (girls 14% vs. boys 16%, $p < 0.05$). Patients with psoriatic arthritis (20%) and systemic JIA (18%) showed the highest overweight rates. In multivariate analyses, age (OR 1.06; 95%CI: 1.04-1.09), male sex (OR 1.21; 95%CI: 1.01-1.44), functional limitations (OR 1.29; 95%CI: 1.04-1.59), as well as therapy with biological DMARDs (OR 1.48; 95%CI: 1.22-1.80) and systemic glucocorticoids (OR 1.40; 95%CI: 1.14-1.71) were significantly associated with overweight.

Conclusion: The prevalence of overweight and obesity in young patients with JIA is similar to that of children and adolescents in the general population. The overweight rate increases with age and is strongly associated with functional restrictions and treatment with glucocorticoids. The role of overweight in the long-term outcome of JIA is an issue that still needs to be addressed.

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