Disclosure of Interests: Riccardo Papa: None declared, Marta Rusmini: None declared, Stefano Volpi: None declared, Roberta Caorsi: None declared, Paolo Picco: None declared, Alice Grossi: None declared, Francesco Caroli: None declared, Francesca Bovis: None declared, Valeria Musso: None declared, Laura Obici: None declared, Cinzia Castana: None declared, Angelo Ravelli Grant/research support from: Angelini, AbbVie, Bristol-Myers Squibb, Johnson & Johnson, Novartis, Pfizer, Reckitt Benkiser, and Roche, Consultant for: Angelini, AbbVie, Bristol-Myers Squibb, Johnson & Johnson, Novartis, Pfizer, Reckitt Benkiser, and Roche, Speakers bureau: Angelini, AbbVie, Bristol-Myers Squibb, Johnson & Johnson, Novartis, Pfizer, Reckitt Benkiser, and Roche, Marielle E van Gijn: None declared, Isabella Ceccherini: None declared, Marco Gattorno Grant/research support from: MG has received unrestricted grants from Sobi and Novartis

DOI: 10.1136/annrheumdis-2019-eular.3562

FRI0569

WHAT DOES IT MEAN TO BECOME PREGNANT WITH JUVENILE IDIOPATHIC ARTHRITIS? A MONOCENTRIC EXPERIENCE IN A TERTIARY CENTRE OF MILAN DEDICATED TO YOUNG ADULTS AFFECTED BY JIA

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Background: During the last seventeen years, biological and non-biological therapies have been used in our center in an open prospective study for the treatment of refractory JIA not only in paediatric age but also in young adults. The availability of more effective drugs for JIA has dramatically increased the number of patients willing to become pregnant. Data regarding the effect of JIA on pregnancy outcome are scant.

Objectives: The study aims to evaluate the capability of JIA patients to become pregnant, the effects of drugs on pregnancy outcome and the impact of gestation on the disease course.

Methods: JIA patients regularly followed up in our Transition Clinic of the Division of Rheumatology, Gaetano Pini Institute of Milan, were enrolled in the study at positive pregnancy test. During the whole pregnancy patients underwent monthly clinical examination and obstetric ultrasound. Data regarding disease activity and pregnancy course (fetal morphometric parameters, fetal heart rate, fetal growth, Doppler velocimetry) were collected. Gestational age, birth weight and APGAR score were also recorded.

Results: 29 pregnancies in 23 women affected by refractory JIA became pregnant during a 17 years follow up. All patients had a long lasting polyarticular disease (median duration of 23 years) not responsive to DMARDs, and became pregnant during biologic therapy. Patients were treated with Etanercept (11 patients), Golimumab (4 patients), Rituximab (3 patients), Adalimumab and Certolizumab (2 patients respectively) as monotherapy, and in most of the cases after multiple switches. One woman was treated with Etanercept during the first pregnancy and Adalimumab during her second pregnancy. Three patients decided for an elective termination and 3 experienced an early miscarriage; among 23 pregnancies resulting in live born infants, only 3 had premature births and 1 cleft palate. A pregnancy was complicated by gestosis, two by placental detachment. All the babies were followed up during the 17 years of observation and did not experience any major late complication. Eight patients were subjected to intra-articular infiltrations during pregnancy to switch off the disease, 18 patients resumed therapy shortly after childbirth and only 7 patients decided to breastfeed.

Conclusion: despite a large amount of studies demonstrating the safety of anti-TNF during pregnancy, data regarding the effects of biologics on pregnancy outcome in JIA are still lacking. The very low number of patients treated with traditional DMARDs achieving low disease activity underline the pivotal role of new biologic drugs in the management of aggressive long standing JIA, to improve the quality of life of these patients, including family planning. In our experience, no greater number of unexpected complications or side effects were observed in JIA patients during pregnancy compared to the other autoimmune diseases. Discontinuation of therapy has increased the risk of flare, requiring local therapy, confirming that EULAR recommendations for the use of biologics during pregnancy can be applied also in JIA.

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Disclosure of Interests: Irene Pontikaki: None declared, Maria Gerosa: None declared, Lorenza Maria Argolini: None declared, Carolina Artusi: None declared, Martina Cornalba: None declared, Laura Trespidi: None declared, Pier Luigi Meroni Consultant for: Inova, Thermofisher, Teofarma

DOI: 10.1136/annrheumdis-2019-eular.7718

FRI0570

CLINICAL CHARACTERISTICS IN PATIENTS WITH PEDIATRIC SYSTEMIC LUPUS ERYTHEMATOSUSDIAGNOSED BEFORE 6 -YEARS OLD IN A THIRD-LEVEL HOSPITAL IN MEXICO

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Background: Pediatric-onset Systemic Erythematosus Lupus (pSLE) is known to be more aggressive than adult-onset SLE. The disease is extremely rare under the age of 5 years and with severe course.

Another big difference is the association of pSLE with monogenic primary immune deficiencies (PID). Some PIDs have been associated with SLE or lupus-like manifestations: deficiencies of the classical complement pathway, female carriers of X-linked chronic granulomatous disease, IgA deficiency (present in 5% of pSLE).

It is important to consider PIDs in patients with severe manifestations in pSLE, in our country there are no studies of this association.

Objectives: To describe the clinical manifestations, laboratory features, characteristics and therapeutics of patients with diagnosis of SLE before 6 years of age at National Institute of Pediatrics in Mexico City

Methods: This is a retrospective study with the review of the patient's records with diagnosis of SLE before 6 years old, from the Immunology department between January 2005 and December 2016 at National Institute of Pediatrics in Mexico City.

Results: Between 2005-2016 there were 295 newly patients diagnosed with pediatric Systemic Lupus Erythematosus (pSLE) at our hospital, from these patients 2.7% (8) were diagnosed with pSLE before 6 years of age. According to this, 7 (87.5%) were female. The youngest patient was 26 months at moment of diagnosis.

Every patient had at least 4 criteria of the ACR classification for SLE. Only 2 (25%) had malar rash at diagnosis, and 1 (12.5%) presented discoid rash. Seventy-five percent of patients presented glomerulonephritis at diagnosis and also 75% had an Hematological disorder.

In terms of serology, 37.5% had positive native DNA antibody, 37.5% antibody to Sm protein and 37.5% antiphospholipid antibodies. One hundred percent of patients had presence of antinuclear antibody (ANA) at moment of diagnosis.

Conclusion: During 2005-2016 we found 295 newly patients with diagnosis of pediatric Systemic Erythematosus Lupus (pSLE), from these, eight patients (2.7%) were diagnosed before 6 years of age. Seventy-five percent presented recurrent infections and 50% with severe infections, and one patient died due to septic shock.

Systemic erythematosus lupus with an early onset has a more severe phenotype and has been associated with monogenic primary immune deficiency, presenting with severe infections and higher mortality.

In our country, we do not have genetic testing to help us determine the endless monogenic defects associated with early-onset SLE, being necessary to determine etiology and specific treatment as needed.

We recommended that in every patient with severe pSLE who develops life-threatening, infections or due to unusual germs, associated with hypocomplementemia and/or consanguinity, to perform an approach to rule out any primary immune deficiency.

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Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2019-eular.8237

FRI0571

MEASUREMENT PERFORMANCE OF REDUCED VERSIONS OF MUSCLE STRENGTH TOOLS IN JUVENILE DERMATOMYOSITIS

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Background: Assessment of muscle strength is a fundamental component of the clinical evaluation of children with juvenile dermatomyositis (JDM). Regular measurement of muscle strength in daily care requires the availability of simple and quick muscle assessment tools.

Objectives: To investigate whether reduced versions of the MMT8 and CMAS are equally reliable as the original tools.

Methods: The following 4 reduced instruments were devised: 1) MMT4 (score 0-40), including 4 items of MMT8 (neck flexors, deltoid middle, gluteus maximus, and gluteus medius); 2) MMT6 (score 0-60), composed of the same items of MMT4 plus biceps brachii and quadriceps; 3) head lift time of CMAS (0-120 seconds or 0-5 points); 4) sum of CMAS head lift time in points and the 6 sit-ups maneuvers of CMAS (score 0-11). Validation was conducted according to OMERACT filter on 213 patients followed in standard clinical care at 13 international pediatric rheumatology centers and evaluated at baseline and after a median of 5.9 months.

Results: All reduced instruments revealed strong correlations ($r > 0.7$) with muscle activity VAS and total DAS, moderate correlations ($r = 0.4-0.7$) with physician's global VAS, muscle DAS, skin activity VAS, pain VAS, parent's overall wellbeing VAS, and CK. Correlations with skin DAS and fatigue VAS were low ($r < 0.4$). Cronbach's alpha was excellent (0.92-0.95) for all reduced tools for which this property could be assessed. SRM was good-to-moderate (0.60-0.91) for all reduced instruments in patients judged as improved by the physician. All reduced tools discriminated strongly between patients classified in different disease activity states by the physician ($p < 0.0001$), and between patients whose parents were satisfied or not satisfied with their children's disease status ($p < 0.0001$). Overall, the metrologic performance of the reduced instruments was comparable to that of MMT8 and CMAS.

Conclusion: We found that reduced versions of the MMT8 and CMAS have good metrologic properties and perform similarly to the original tools in a population of patients followed in standard clinical care. Our results suggest that these simplified and shortened instruments could serve as surrogate for the complete measures in routine practice, particularly in a busy clinical setting.

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Disclosure of Interests: Silvia Rosina: None declared, Giulia Camilla Varnier: None declared, Angela Pistorio: None declared, Alessandro Consolaro Grant/research support from: AbbVie, Pfizer, Clarissa Pilkington: None declared, Susan Maillard: None declared, Adele Civino: None declared, Elena Tsitsami: None declared, Jaime de Inocencio: None declared, Marija Jelusic: None declared, Jelena Vojinovic: None declared, Graciela Espada: None declared, Balahan Makay Speakers bureau: Enzyvant, Novartis, Roche, Abbvie, Maria Katsikas: None declared, Polixeni Pratsidou-Gertsis: None declared, Dragana Lazarevic: None declared, Anand Prahalad Rao: None declared, 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., Angelo Ravelli Grant/research support from: Angelini, AbbVie, Bristol-Myers Squibb, Johnson & Johnson, Novartis, Pfizer, Reckitt Benkiser, and Roche, Consultant for: Angelini, AbbVie, Bristol-Myers Squibb, Johnson & Johnson, Novartis, Pfizer, Reckitt Benkiser, and Roche, Speakers bureau: Angelini, AbbVie, Bristol-Myers Squibb, Johnson & Johnson, Novartis, Pfizer, Reckitt Benkiser, and Roche

DOI: 10.1136/annrheumdis-2019-eular.2228

FRI0572

DISABILITY AND HEALTH-RELATED QUALITY OF LIFE OUTCOMES IN PATIENTS WITH SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS TREATED WITH TOCILIZUMAB IN A PHASE 3 RANDOMIZED CONTROLLED TRIAL

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Background: Tocilizumab (TCZ) intravenous (IV) formulation was approved for the treatment of patients with systemic juvenile idiopathic arthritis (sJIA) based on the results of a large phase 3 clinical trial. Physical function, measured by the Childhood Health Assessment Questionnaire-Disability Index (CHAQ-DI), and health-related quality of life (HRQOL), measured by the Child Health Questionnaire (CHQ), were evaluated.

Objectives: To examine measures of disability and HRQOL in patients with sJIA treated with TCZ IV for up to 2 years in post hoc analysis of data from the phase 3 trial of TCZ.

Methods: Changes within 3 months of treatment initiation with TCZ (baseline) were compared between TCZ- and placebo (PBO)-treated patients using CHAQ-DI, pain global assessment (Pain-GA), physician global assessment, and patient global assessment (Pt-GA) using analysis of variance adjusted for treatment group. Changes in CHAQ-DI overall and domain scores and changes in CHQ domain and summary scores from baseline to 2 years were compared for patients treated with TCZ using the unpaired *t* test.

Results: Patients with sJIA experienced clinically relevant improvement in physical function (CHAQ-DI) and reduction of pain (Pain-GA). Mean (SD) CHAQ-DI scores for patients treated with PBO and TCZ were 1.7 (0.8) for both groups at baseline and 1.2 (1.0) and 0.9 (0.8), respectively, at week 12 (week 12 mean difference, -0.2; median [range] change from baseline to week 12 difference, -0.3 [-0.6 to 0.0]). These patients also had significantly improved CHQ socialization, behavior, mental health, and psychosocial summary scores after 3 months compared with those receiving PBO (Figure 1). Improvement in all CHAQ-DI domains over 2 years was observed with TCZ treatment (Figure 2); improvement rates in patient well-being (Pt-GA) were 87.7%. Similar improvements in physical function were observed in patients with polyarticular JIA in the phase 3 trial that led to approval of TCZ IV in these patients (not shown). There was also significant improvement ($p < 0.05$) in most domains of HRQOL (CHQ domain scores) in patients with sJIA; of note, patients experienced marked improvement in mean scores from baseline to week 104 for pain/discomfort (31.7 to 75.3), self-esteem (61.0 to 76.0), mental health (62.1 to 76.8), and social limitation-emotional (52.6 to 86.2).

Conclusion: Two years of TCZ treatment resulted in statistically significant and clinically relevant improvements in function and HRQOL in patients with sJIA.