

that in both groups of patients IL-1 $\beta$ , IL-6 and TNF- $\alpha$  levels were significantly higher than those observed in HS. In addition, we found that LPS-stimulated whole blood cells from non-responder inactive sJIA patients released significantly higher levels of IL-1 $\beta$  and TNF- $\alpha$  compared to responder inactive sJIA patients.

**Conclusion:** Our preliminary results show a dysregulated production of inflammatory cytokines by whole blood cells from sJIA patients in remission disease, when stimulated with a TLR-4 agonist.

#### Disclosure of Interest

None Declared

#### P2065

##### Anakinra drug retention rate and predictive factors of drug survival in systemic juvenile idiopathic arthritis and adult onset Still's disease

Jurgen Sota<sup>1</sup>, Donato Rigante<sup>2</sup>, Antonella Insalaco<sup>3</sup>, Paolo Sfriso<sup>4</sup>, Salvatore de Vita<sup>5</sup>, Rolando Cimaz<sup>6</sup>, Giuseppe Lopalco<sup>7</sup>, Giacomo Emmi<sup>8</sup>, Francesco La Torre<sup>9</sup>, Claudia Fabiani<sup>10</sup>, Alma N. Olivieri<sup>11</sup>, Marco Cattalini<sup>12</sup>, Daniele Cammelli<sup>13</sup>, Romina Gallizzi<sup>14</sup>, Maria Alessio<sup>15</sup>, Raffaele Manna<sup>16</sup>, Ombretta Viapiana<sup>17</sup>, Micol Frassi<sup>18</sup>, Armin Maier<sup>19</sup>, Carlo Salvarani<sup>20</sup>, Rosaria Talarico<sup>21</sup>, Roberta Priori<sup>22</sup>, Maria C. Maggio<sup>23</sup>, Manuela Pardeo<sup>3</sup>, Carla Gaggiano<sup>24</sup>, Salvatore Grosso<sup>24</sup>, Fabrizio de Benedetti<sup>25</sup>, Antonio Vitale<sup>26</sup>, Luca Cantarini<sup>26</sup>

<sup>1</sup>Research Center of Systemic Autoinflammatory Diseases and Behçet's Disease Clinic, Department of Medical Sciences, Surgery and Neurosciences, University of Siena, Siena; <sup>2</sup>Institute of Pediatrics, Fondazione Policlinico A. Gemelli IRCCS; <sup>3</sup>Division of Rheumatology, Department of Pediatric Medicine, Bambino Gesù Children's Hospital IRCCS, Rome; <sup>4</sup>Rheumatology Unit, Department of Medicine, University of Padua, Padua; <sup>5</sup>Department of Medical and Biological Sciences, Rheumatology Clinic, University of Udine, Udine; <sup>6</sup>Rheumatology Unit, Meyer Children's Hospital, University of Florence, Florence; <sup>7</sup>Department of Emergency and Organ Transplantation-Rheumatology Unit, University of Bari, Bari; <sup>8</sup>Department of Experimental and Clinical Medicine, University of Florence, Florence; <sup>9</sup>Pediatric Rheumatology Section, Pediatric Oncology Unit, Vito Fazzi Hospital, Lecce; <sup>10</sup>Ophthalmology Unit, Department of Medicine, Surgery and Neuroscience, University of Siena, Siena; <sup>11</sup>Dipartimento della Donna, del Bambino e di Chirurgia Generale e Specialistica, Seconda Università degli Studi di Naples, Naples; <sup>12</sup>Pediatric Clinic, University of Brescia and Spedali Civili di Brescia, Brescia; <sup>13</sup>Experimental and Clinical Medicine Department, University of Florence, Florence; <sup>14</sup>Department of Pediatrics, Azienda G. Martino, University of Messina, Messina; <sup>15</sup>Department of Pediatrics, University Federico II, Naples; <sup>16</sup>Periodic Fever Research Center, Università Cattolica Sacro Cuore, Rome; <sup>17</sup>Rheumatology Section, Department of Medicine, University of Verona, Verona; <sup>18</sup>Rheumatology and Clinical Immunology Unit, Department of Clinical and Experimental Sciences, University of Brescia and Spedali Civili di Brescia, Brescia; <sup>19</sup>Struttura Semplice di Reumatologia, Ospedale di Bolzano, Bolzano; <sup>20</sup>Rheumatology Unit, Department of Internal Medicine, Azienda Ospedaliera ASMN IRCCS, Reggio Emilia; <sup>21</sup>Rheumatology Unit, Department of Clinical and Experimental Medicine, University of Pisa, Pisa; <sup>22</sup>Department of Internal Medicine and Medical Specialties, Rheumatology Unit, Sapienza University of Rome, Rome; <sup>23</sup>University Department "Pro.S.A.M.I.", University of Palermo, Palermo; <sup>24</sup>Clinical Pediatrics, Department of Molecular Medicine and Development, University of Siena, Siena; <sup>25</sup>Division of Rheumatology, Department of Pediatric Medicine, Bambino Gesù Children's Hospital, IRCCS, Rome; <sup>26</sup>Research Center of Systemic Autoinflammatory Diseases and Behçet's Disease Clinic, Department of Medical Sciences, Surgery and Neurosciences, University of Siena, Siena, Italy

**Correspondence:** Jurgen Sota

Pediatric Rheumatology 2019, 17(Suppl 1):P2065

**Introduction:** Only a few studies have reported the long-term efficacy of interleukin (IL)-1 inhibition in systemic juvenile idiopathic arthritis (sJIA) and adult onset Still's disease (AOSD). We herein describe Anakinra (ANA) effectiveness expressed in terms of drug

retention rate (DRR) and evaluate predictive factors of drug survival in sJIA and AOSD patients.

**Objectives:** Examine the overall DRR of ANA in sJIA and AOSD patients. Explore the influence of biologic line of treatment, and the concomitant use of disease modifying anti-rheumatic drugs (cDMARDs) on DRR in the whole sample and stratified according to the disease thereafter; find eventual predictive factors associated with events leading to drug discontinuation. The corticosteroid (CS)- and cDMARDs-sparing effect, the impact of treatment delay on survival and the record of safety profile constituted ancillary aims.

**Methods:** Medical records from 61 sJIA and 76 AOSD patients treated with ANA in 24 Italian tertiary referral centers were retrospectively reviewed.

**Results:** The cumulative retention rate of ANA at 12-, 24-, 48- and 60-months of follow-up was 74.3%, 62.9%, 49.4% and 49.4% respectively, without any significant differences between sJIA and AOSD patients ( $p=0.164$ ), and between patients treated in monotherapy compared to the subgroup co-administered with conventional cDMARDs ( $p=0.473$ ). On the other hand, a significant difference in DRR was found between biologic-naïve patients and those previously treated with biologic drugs ( $p=0.009$ ), which persisted even after adjusting for pathology ( $p=0.013$ ). In regression analysis, patients experiencing adverse events (AEs) (HR=3.029 [C.I. 1.750-5.242],  $p<0.0001$ ) and those previously treated with other biotechnologic agents (HR=1.818 [C.I. 1.007-3.282],  $p=0.047$ ) were associated with a higher hazard ratio of ANA discontinuation. The median treatment delay was significantly higher among patients discontinuing ANA ( $p<0.0001$ ). A significant CS- ( $p=0.033$ ) and cDMARDs-sparing effect ( $p<0.0001$ ) was also recorded. Less than one third of our cohort developed AEs and 85% were deemed mild in nature, with 70% involving the skin.

**Conclusion:** Our findings display an overall excellent DRR of ANA on the long run for both sJIA and AOSD that may be further optimized by closely monitoring patient's safety issues and employing this IL-1 inhibitor as a first-line biologic as early as possible. Moreover, ANA allowed a significant drug-sparing effect while showing a good safety profile.

#### Disclosure of Interest

None Declared

#### P2066

##### Predictors of effectiveness of anakinra in systemic juvenile idiopathic arthritis

Jessica Tibaldi<sup>1</sup>, Bendetta Saccomanno<sup>1</sup>, Francesca Minoia<sup>2</sup>, Francesca Bagnasco<sup>3</sup>, Angela Pistorio<sup>3</sup>, Andreassa Guariento<sup>4</sup>, Roberta Caorsi<sup>3</sup>, Alessandro Consolaro<sup>1</sup>, Marco Gattorno<sup>3</sup>, Angelo Ravelli<sup>1</sup>  
<sup>1</sup>Istituto G. Gaslini/Università degli Studi di Genova, Genoa; <sup>2</sup>Fondazione IRCCS Ca' Granda, Ospedale, Milan; <sup>3</sup>Istituto G. Gaslini, Genoa, Italy; <sup>4</sup>Instituto de Criança – FMUSP, Rio de Janeiro, Brazil

**Correspondence:** Jessica Tibaldi

Pediatric Rheumatology 2019, 17(Suppl 1):P2066

**Introduction:** Systemic juvenile idiopathic arthritis (sJIA) is the most severe and a rather distinct subtype of JIA. It is common view that sJIA is the most severe form of childhood arthritis and the most difficult to treat. Recently the use of interleukin (IL)-1 antagonists has led to a significant improvement of the disease's long-term evolution and has confirmed this cytokine's key-role in the pathogenesis of sJIA. A number of potential predictors of the therapeutic effectiveness of IL-1 inhibitors have been reported, which include less severe joint disease and increased white blood cell count, shorter disease duration, older age at disease onset and use of IL-1 blockade as first-line therapy. However, because the experience gained so far is still limited, there is a need of further data to better characterize the profile of sJIA patients who are more susceptible to respond to IL-1 blockade.

**Objectives:** To seek predictors of therapeutic response to the interleukin (IL)-1 inhibitor anakinra in children with systemic-onset juvenile idiopathic arthritis (sJIA).