and a longer duration of disease (months) (5.7 ± 1.6 vs 3.1 ± 3.2, *p 0.06*). In addition, fewer relapses were found among patients with peroneal involvement [0 (0-0) vs 5 (1-6.7), *p 0.05*], a high white blood cell count at diagnosis (r -0.569, *p 0.02*) and those who were not treated with pamidronate [0 (0-5) vs 5 (1.2-6.7), *p 0.07*]. Finally, the lack of sequelae was related to the administration of pamidronate (100% vs 57.1%, *p 0.07*), younger age at diagnosis (years) (9.08 ± 1.4 vs 11 ± 2, *p 0.07*) and lesser CRP level at diagnosis (mg/L) [2.9 (2.9-2.9) vs 3.8 (2.9-3.8), *p 0.005*].

Conclusion: Advanced cases of NBO, without radiological signs of sclerosis or lysis, were more likely to achieve remission. Elevation of acute phase reactants was related to a better response to treatment and a lower relapse rates, but with a greater probability of sequelae. As described in the literature, bisphosphonates achieve higher rates of clinical and radiological remission. In our study, they are also associated with a lower relapse rates and the lack of sequelae. The last is the main reason to be considered the first treatment choice in NBO.

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SAT0511 CANAKINUMAB IN SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS: CLINICAL INACTIVE DISEASE RATE AND SAFETY IN ITALIAN PATIENTS

Manuela Pardeo[1], Claudia Bracaglia[1], Anna Lucia Piscitelli[1], Arianna De Matteis[2], Jessica Tibaldi[3], Maria Alessio[2], Achille Marino[4], Giovanni Conti[5], Maria Cristina Maggio[6], Clotilde Alizzi[7], Francesco Licciardi[8], Alma Nunzia Olivieri[9], Giovanni Filocamo[10], Francesca Orlando[2], Silvana Martino[8], Rolando Cimaz[4], Angelo Ravelli[3], Fabrizio De Benedetti[1]. *[1]IRCCS Ospedale Pediatrico Bambino Gesù, Division of Rheumatology, Rome, Italy; [2]University Federico II, Department of Pediatrics, Rheumatology Unit, Naples, Italy; [3]University of Genova and Giannina Gaslini Institut, Department of Paediatric Rheumatology, Genoa, Italy; [4]Meyer Children's Hospital, University of Florence, Department of Neurosciences, Psychology, Drug Research and Child Health, Rheumatology Unit, Florence, Italy; [5]University of Messina, Unit of Pediatric Nephrology and Rheumatology, Messina, Italy; [6]University of Palermo, University Department Pro.Sa.M.I. "G. D'Alessandro", Palermo, Italy; [7]University of Palermo, University Department Pro.Sa.M.I. "G. D'Alessandro", Palermo, Italy; [8]University of Turin, Department of Pediatrics and Infectious Diseases, Torino, Italy; [9]Second University of Studies of Naples, Department of Woman, Child, and General and Specialistic Surgery, Naples, Italy; [10]Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Intermediate Pediatric Care Unit, Milano, Italy*

Background: Systemic juvenile idiopathic arthritis (sJIA) is a polygenic autoinflammatory disease. The innate immune mechanisms play a central role with overproduction of inflammatory cytokines. The increased knowledge on the role of these cytokines has provided a change in the natural history of the disease with the introduction of the targeted treatments. Remarkable results has been observed with canakinumab, an anti-interleukin-1β monoclonal antibody, in two clinical trials but little information are available in real life.

Objectives: To evaluate clinical inactive disease rate and safety of canakinumab in Italian patients with sJIA.

Methods: We have collected retrospectively clinical and laboratory data of patients with sJIA treated with canakinumab in 9 Italian Pediatric Rheumatology centers. Clinically inactive disease (CID) at 6 months was defined according to Wallace criteria. We analyzed the effect of canakinumab on fever, rash, number of actives joints, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and physician's global assessment of disease activity score.

Results: Forty seven patients (26 F) were included in the analyses. The median age (range) at the diagnosis and at the beginning of treatment with canakinumab was 7.6 (1-14.7) and 10.2 (1.7-22.2) years, respectively. Twenty seven patients (57.4%) had been previously treated with other biologic agents (18 with anakinra, 1 with tocilizumab, 6 with both and 2 with etanercept), withdrawn for inefficacy in 15/27 (55.5%). Thirty patients (63.8%) were receiving concomitant treatment with glucocorticoids at the median dose (range) of 0.69 (0.02-2.75) mg/kg/die. Thirtynine out of 47 patients had > 6 months of follow-up. Among these 39 patients, 27 (69.2%) achieved CID at 6 months and 5/27 (18.5%) were still on glucocorticoids. Of the 30 patients who received concomitant glucocorticoids at baseline, 24 achieved 6 months of follow-up and 12 (50%) of these were able to withdraw glucocorticoids. Minor adverse events were reported in 5/30 (16.6%) patients: upper respiratory tract infections in 4 and transient injection site reaction in 1. No cases of macrophage activation syndrome was reported.

Conclusion: Our results provide initial real world evidence of the efficacy of treatment with canakinumab in patients with sJIA. In our study the percentage of patients who reached CID at 6 months is slightly higher (69.2%) than reported at the end (from 3 months to one year) of the 2 published randomized trials (60%). No serious adverse events were recorded in our population.

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SAT0512 LONG-TERM FOLLOW-UP OF CARDIOVASCULAR OUTCOMES IN KAWASAKI DISEASE. OBSERVATIONAL STUDY FROM A SINGLE CENTER

D. Prieto-Peña, José Luis Martín-Varillas, Lara Sánchez Bilbao, Eva Peña Sainz-Pardo, Monica Calderón-Goercke, Natalia Palmou-Fontana, María Teresa Viadero, María Jesús Cabero, Miguel A. González-Gay, Ricardo Blanco. *Marqués de Valdecilla University Hospital, Santander, Spain*

Background: Kawasaki disease (KD) is an acute systemic vasculitis of unknown aetiology which can lead to coronary aneurysm (CA) formation in 15-20% of patients. Long-term rates of adverse cardiac events are not well-known.

Objectives: a) to compare demographic, clinical and laboratory features of patients who developed coronary aneurysm with patients who did not. b) to assess long-term cardiovascular outcomes in the subgroup of patients who developed cardiac aneurysms.

Methods: Single center study of 55 patients with KD who were diagnosed between 1994-2009. KD diagnosis was based on the classic classification criteria[1]. We considered two groups: a) patients who presented CA at diagnosis. b) patients who did not present CA. Statistical analysis was performed with SPSS. Student´s t test or Mann-Whitney U test was used to compare continuous variables, and Chi-squared test or Fisher´s exact test for categorical variables as appropriate.

Results: Out of 55 patients with KD, 8 patients (3 men/5 women) presented CA at diagnosis. Differences between the two groups mentioned above are shown in **TABLE 1.** Mean age at diagnosis was lower in patients who presented CA (1.74 vs 3.88 years; p=0.044). The duration of symptoms was longer in patients with CA (14.0 vs 12.0 days; p=0.015), however there were no qualitative differences in the symptomatology. Regarding laboratory markers, C- reactive protein (CRP) levels were significantly higher in patients who developed CA (17.0 vs 10.8 mg/dL; p=0.047). There was also a tendency to higher levels of erythrocyte sedimentation rate (ERS) and higher platelet count. All patients received intravenous immunoglobulin (IVIG) and antiplatelet agents at the time of