Abstract THU0599 – Figure 1 Efficacy of canakinumab with adapted ACR Paediatric criteria in SJIA patients with/without prior use of tocilizumab (TCZ)

Conclusions: CAN 4 mg/kg q4w s.c. provided improvement in disease activity and a reduction of oral steroid dose in Japanese SJIA pts, including those who were not well-controlled with TCZ. No new safety concerns were reported.

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THU0600

HIGH VACCINE COVERAGE RATES ARE NOT ENOUGH: VACCINATION DELAY AND RISK FOR VACCINE-PREVENTABLE DISEASES IN PAEDIATRIC PATIENTS WITH RHEUMATIC DISEASES WITH AND WITHOUT IMMUNOSUPPRESSIVE THERAPY

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Background: Paediatric patients with rheumatic diseases (PedRD) are more susceptible to invasive infectious diseases, due to their underlying disease, high disease activity and immunosuppressive therapy (IT).^{1,2} In Switzerland, specially indicated (SI) vaccinations for PedRD patients against pneumococcus, varicella zoster, hepatitis B, influenza and human papilloma virus should be administered as early as possible. On-time and complete coverage reduces the risk of vaccine-preventable infections³ and its associated morbidity. Information about on-time vaccination is scarce, especially in PedRD patients.

Objectives: To assess the relationship of vaccine coverage and on-time vaccination in PedRD patients in Switzerland with and without IT.

Methods: Multicenter retrospective prevalence study based on the Juvenile Inflammatory Rheumatism (JIR) cohort, an international registry for rheumatic diseases. Patients treated in Swiss paediatric rheumatology centres (Basel, Geneva, Lausanne, Lucerne, Zurich) were included. Vaccine coverage and timeliness of administered vaccines were assessed for routine and for SI vaccinations according to the Swiss Federal Office of Public Health.

Results: 239 patients were included. 93 (39%) received IT during a median observation period of 2.7 years (IQR 1.1–5.4). Median age was 6.7 years (IQR 2.9–10.3) at diagnosis, 10.9 years (IQR 7–14.3) at reference date (12th April 2016). At diagnosis, 185 patients (77%) had complete routine vaccine coverage; 76 (32%) fulfilled Swiss on-time immunisation criteria. At reference date, the routine vaccine coverage rate decreased to 74% with on-time vaccination achieved in 29% of PedRD patients without IT and in 23% with IT, respectively. Completeness of SI vaccinations was documented in 19 PedRD patients (13%) without IT and 6 patients (6.5%) with IT. On-time vaccination for SI vaccination was fulfilled in 7 patients without IT; no patient with IT was vaccinated on-time. Only 23 (9.6%)

patients showed complete and thereof 4 (1.6%) on-time vaccination status for routine and SI vaccinations.

Conclusions: Despite moderate to high routine vaccination coverage in Switzerland, vaccinations particularly recommended for PedRD patients to prevent from complications, are incompletely administered. Moreover, the high frequency of delayed vaccination implicates a potential increase of vaccine-preventable disease. Close surveillance of vaccination status improves standard of care for this susceptible population.

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THU0601

CARRIER RATE OF FAMILIAL MEDITERRANEAN GENE (MEFV) MUTATION IN PARENTS OF CHILDREN WITH FAMILIAL MEDITERRANEAN FEVER

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Background: Familial Mediterranean Fever (FMF) is one of the most prevalent periodic fever syndromes, MEFV, the responsible gene for the disease is in the short arm of chromosome 16. In the considerable count of the FMF patients, only one mutation is found in the MEFV, and parents who was the obligatory carriers for that mutation, were asymptomatic.

Objectives: The aim of this study was to evaluate these asymptomatic parents in regard to mutation in MEFV gene and similarity between parents and offspring patients.

Methods: In this cross sectional study, asymptomatic parents of FMF patients enrolled the study, which were referred to periodic fever clinic or paediatric rheumatology clinic of Tehran University of medical sciences. The patients should have at least one mutation in MEFV gene and none of them had any family history of auto inflammatory disease. Twelve mutations in MEFV gene was assessed in the parents by Vienna Lab FMF Strip Assay kit by MAS PCR/Reveres hybridization.

Results: Forty three patients and their parents participated in the study. Sixty three percent²⁷ of patients were male. Onset of disease symptoms in 31 patients (72%) were before 4 years of old. Nine (21%) of the patients were homozygote, 16 (37%) compound heterozygote and 17 (40%) heterozygote for MEFV mutation, there was a case of complex alleles mutations (2%). M694V/M694V in 4 patients (9%) was the most homozygote genotype, M694V/R761H in 4 (9%) and E148Q in 7 (16%) were the most compound heterozygote and heterozygote genotype, respectively. M694V, M680I and E148Q were the most mutation in the parents. Overall, 41 patients, had mutations similar to their parents' mutation, except in 2, which parents had not mutation, but patient did.

Conclusions: It seems that occurrence of new mutations in offspring is not prevalent among FMF patients, and there are other reasons for different clinical presentation in similar mutation carriers. On the other hand, in ethnicities with high prevalence of FMF, new mutation in descendant may occurs, infrequently.

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