Background: Tumour necrosis factor (TNF) receptor-associated periodic syndrome (TRAPS) is one of the best-known monogenic auto-inflammatory disorders resulting from an autosomal dominant variation in the TNF superfamily receptor 1A (TNFRSF1A) gene (1).

Objectives: To define best treatment approach in patients with TRAPS and effect on long-term outcomes.

Methods: We reviewed evidence data on patients with TRAPS enrolled in the Eurofever international registry according the INSADA gene variant classification and the new Eurofever/PRIINTO classification criteria (EPCc).

Results: Data on 226 patients were available. Patients not fulfilling the EPCc carrying likely benign/variant (21 patients, 9%) or VOUS/not classified variants (40 patients, 18%) displayed a milder disease than the patients fulfilling the EPCc with VOUS/not classified variants (38 patients, 17%) or pathogenic/likely pathogenic variants (127 patients, 56%). In particular, in patients not fulfilling the EPCc, less frequent abdominal pain and skin rashes, higher efficacy rate (>85% complete response), while Etanercept was less effectively used and discontinued in 65% of patients.

Conclusion: Anti-IL-1 drugs are the best maintenance treatment in TRAPS with potential to reverse the most serious disease complications of AA amyloidosis and infertility. The diagnosis of TRAPS should be considered very carefully in patients carrying VOUS/not classified variants not fulfilling the EPCc.

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References:

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EFFICACY AND SAFETY OF PCV13 VACCINATION IN JIA PATIENTS WITH SYSTEMIC MANIFESTATIONS ON TOCILIZUMAB AND CANAKINUMAB TREATMENT

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Background: The need for continuous use of immunosuppressive drugs leads to increased risk of developing infectious diseases in children with juvenile idiopathic arthritis with systemic manifestation (sJIA). Questions about choosing the optimal vaccination time and the effect of different classes of therapy on vaccination effectiveness are still open.

Objectives: To study clinical and laboratory effectiveness of PCV13-vaccination in children with sJIA on tocilizumab (TOC) and canakinumab (CAN) treatment depending on disease activity during vaccination.

Methods: Prospective cohort study included 2 groups of sJIA patients: in stable remission (Remission group, n=53) receiving CAN (n=10) or TOC (n=43) treatment, and in acute stage of disease (Acute group, n=25) which started to received CAN (n=7) or TOC (n=18) either before vaccination (Acute Treated before subgroup, n=17) or after vaccination (Acute Treated After subgroup, n=8).

Results: Four weeks after vaccination, protection level of anti-pneumococcal antibodies was achieved by 36 (67.9%) patients in Remission group, 16 (64%) patients in Acute group (intergroup p=0.932), and in 8 (74.06%) patients in Acute Treated Before subgroup and in 8 (100%) patients in Acute Treated After subgroup (intersubgroup p=0.022). PCV13 have shown high clinical effectiveness in both Remission group and Acute group. Reducing of acute respiratory infections, frequency of antibiotics treatment courses, frequency of temporary withdraw of biologics treatment due to severe infections. Frequency of events were counted per patients-years.

Results: Duration of antibiotics treatment reduced from 2.31 to 0.81 weeks per 1 patient-year in Remission group (p<0.001) from 1.97 to 0.74 in Acute group (p<0.001). Among patients who were previously treated with biologics, frequency of therapy withdrawal reduced from 4.34 to 2.42 per patient-year in Remission group (p<0.001) and from 5.33 to 1.18 in Acute Before subgroup (p<0.002). The incidence of reactions to vaccination of PCV13 (loose stools, sensations of hypothermia, pain, subcutaneous temperature) was similar in groups (22 (41.5%) for Remission group and 7 (28%) for Acute group, p= 0.319).

Conclusion: Vaccination with the 13-valent PCV has demonstrated high clinical efficacy and safety in children with sJIA both in the acute stage of the disease and during remission. Vaccination of patients in acute stage of sJIA before treatment has advantages over vaccination during remission or after prolonged immunosuppressive therapy in terms of achieving an adequate vaccine response.

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TOFACITINIB TREATMENT IN RECAGRITANT JDM PATIENTS

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