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

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

Methods: We reviewed data on patients with TRAPS enrolled in the Eurofever international registry according the INSAD gene variant classification and the new Eurofever/PRINTO classification criteria (EPCC).

Results: Data on 226 patients were available. Patients not fulfilling the EPPC carrying likely benign/variant variants (21 patients, 9%) or VOUS/not classified variants (40 patients, 18%) carried a milder disease than the patients fulfilling the EPCC with VOUS/not classified variants (38 patients, 17%) or pathogenic/likely pathogenic variants (172 patients, 76%). In particular, in patients not fulfilling the EPPC, less frequent abdominal pain and skin rashes, higher efficacy rate of colchicine and no development of AA amyloidosis have been reported. Almost 90% of patients fulfilling the EPPC required maintenance therapy and anti-inferleukin (IL)-1 drugs were the most frequently used, with the highest 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.

References:


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

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Objectives: 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.

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

Results: 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). 0.5 ml of the 13-valent PCV was administered once subcutaneously. Efficacy was evaluated by achieving of protection level of anti-pneumococcal antibodies after 4 weeks and by clinical indicators after 6 month follow-up: frequency of acute respiratory infections, frequency of antibiotics treatment courses, frequency of temporary withdrawal of biologics treatment due to severe infections. Frequency of events was counted per patient-years.

Conclusion: Four weeks after vaccination, protection level of anti-pneumococcal antibodies was achieved for 36 (67.9%) patients in Remission group, 16 (64%) patients in Acute group (intergroup p=0.932), and in 8 (47.06%) 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 was as follows: from 4.57 to 2.15 episodes per patient-year in Remission group (p=0.001) and from 4.32 to 1.28 per patient-year in Acute group (p=0.001). 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 3.53 to 1.18 in Acute Treated Before subgroup (p=0.002). The incidence of reactions to vaccination of PCV13 (local pain, redness, swelling) 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 RECALCITRANT JDM PATIENTS

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Background: Juvenile dermatomyositis (JDM) is a systemic, autoimmune inflammatory muscle disorder and vasculopathy that affects children younger than 18 years. Although the cause of JDM remains unknown it is clear that genetic and environmental influences play a role in the aetiology. New treatments are becoming available and being tested through international multicentre trials. Increasing evidence suggests a role for types I and II IFN in juvenile and adult dermatomyositis, including elevated IFN-response gene signatures in the muscle, skin and blood. It has recently been reported that patients with refractory JDM responded well to treatment with tofacitinib, a JAK inhibitor, with corresponding downregulation in selected IFN-response genes.

Objectives: In this study, we evaluated our cases with resistant JDM who received tofacitinib treatment.

Methods: Six patients who received tofacitinib because of severe skin involvement of JDM were included in the study. The data were obtained retrospectively from the hospital records.

Results: The age ranges of the cases were between 7-17 years and the ratio of girls and boys was 1 (3/3). The age of diagnosis was between 2-13 years, and the follow-up period was between 3-9 years. Calcinosis cutis in 5 cases, decreased muscle strength in 3 cases, joint involvement in 4 cases were detected. Systemic steroids, methotrexate, and non-steroid anti-inflammatory drugs were given in all cases before tofacitinib treatment. Pamidronate was used in 4 cases because of severe skin calcinosis, high dose intravenous immunoglobulin in 4 cases, mycophenolate mofetil in 3 cases, rituximab in 3 cases and cyclophosphamide in 1 case previously. Tofacitinib treatment (10mg/qd) was started in six cases with treatment-resistant JDM. Five cases had been treated with tofacitinib for 6-24 month intervals. The treatment was discontinued in one case because of severe allergic reaction. Variable level of improvement were detected in the skin findings of all cases during the therapy period. The treatment was interrupted for 1 month in only one case due to neutropenia.

Conclusion: Tofacitinib seems to be an effective and safe treatment option in patients with JDM who are resistant to conventional treatments. More studies are needed on this subject.

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