colchicine and the group that needed to renew therapy in demographic, genetic and most clinical parameters, including the age (13.4 ± 3.9 vs 11.9 ± 3.7 p=0.26), level of SAA at enrolment (4 ± 3.6 vs 3.3 ± 2.4 p=0.7) and time of last attack prior to enrolment (12.6 ± 9.6 vs 8.6 ± 8.2 months p=0.08). Myalgia and arthritis were more common among children that required to renew therapy compared to the group that didn't (31% vs 6.7% p=0.058 and 31% vs 3% p=0.024 respectively).

Conclusion: Cessation of colchicine therapy following prolonged remission in selected group of patients who are not homozygous for MEFV mutation could be considered. Patients with arthritis or arthralgia are more likely to have an attack after ceasing colchicine therapy.

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THU0529 WHOLE BODY MAGNETIC RESONANCE IMAGING IN JUVENILE DERMATOMYOSITIS. A LONGITUDINAL STUDY

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Background: Juvenile dermatomyositis (JDM) is a rare multisystem connective tissues disorder of unknown aetiology. Assessment of disease activity is a challenge in clinical practice.

Objectives: To compare whole-body MRI (WB-MRI) with clinical examination in the assessment of disease activity in JDM.

Methods: We included consecutive JDM patients followed in the rheumatology unit. All patients were submitted to clinical and laboratory evaluation. WB-MRI images were obtained using a 1.5 T MRI scanner and short T inversion recovery sequences (STIR). Muscle, peripheral inflammation and subcutaneous inflammations signal abnormalities were scored in 42 muscular groups. Muscle inflammation was classified as: 0 = absent; 1 = Mild to moderate/involvement less than 50% of muscle extension and 2 = Accentuated/greater than 50%. Peripheral and subcutaneous inflammations were classified as: 0 = absent; 1 = present; and on proximal and distal extremities. WB-MRI and clinical assessments were performed concurrently and results compared. Evaluation was repeated after 12 months. Statistics was performed according to the nature of the variable.

Results: WB-MRI revealed muscle inflammation in 6 (31.6%) at study entry. We observed grade 2 muscle inflammation of the right and left scapular girdle (1/19 patients), right and left pelvic girdle (2/19 patients) and right and left thigh (1/19 patients). Grade 1 inflammation was observed in peripheral right and left arm (2/19 patients), peripheral right and left thigh (1/19 patients). Grade 1 subcutaneous inflammation was observed in right and left thigh (1/19 patients) and left leg (1/19 patients). Additionally we observed sacroiliitis (1/19 patients), spinal cord infarction (21%) and osteonecrosis (5.2%). All patients were treated with standardized treatment. After 12 months 13/19 (68.4%) patients repeated the WB-MRI. Five (38.4%) patients had new/worsening of muscle and subcutaneous inflammation, one (7.7%) patient had tibial medullary infarction. Correlations between WB-MRI muscle score and disease activity measures were excellent (Manual Muscle Test: $r = -0.88$, Childhood Myositis Assessment Scale: $r = -0.81$). Patients with subcutaneous inflammation developed clinically evident subcutaneous calcifications during follow-up.

Conclusion: WB-MRI provides additional information to clinical evaluation and represents a promising tool to determine the grade of muscle inflammation to additional peripheral and subcutaneous tissue inflammation.

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THU0530 IDIOPATHIC RECURRENT PERICARDITIS: CLINICAL FINDINGS AND TREATMENT APPROACH

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Background: Recurrent pericarditis affects 15-30% of patients with acute pericarditis. The etiology is poorly understood, with about 80% being idiopathic. Several treatment options are available for recurrences, including NSAIDs, colchicine, glucocorticoids and IL-1 inhibitors (i.e. Anakinra). Standardized guidelines for the management of these patients are still lacking

Objectives: To analyze clinical findings and treatment approach in a cohort of pediatric patients with recurrent pericarditis

Methods: Patients with at least two episodes of idiopathic pericarditis, followed at two Pediatric Rheumatology centers between 2006 and 2018, were included

Results: A total of 42 patients (18 males) were included. Mean age at disease onset was 11.8 years (range 4-17). Chest pain and fever were the presenting symptoms in all patients. In 47% pleural effusion was detected. Laboratory tests showed increased white blood cell count (mean $14.509/\text{mm}^3$), C-reactive protein (mean 18.01 mg/dl) and erythrocyte sedimentation rate (mean 39 mm/h) in all patients. The first episode was variably treated: 18/42 (43%) received NSAIDs alone, 5/42 (11.9%), colchicine alone or associated to NSAIDs and 3/42 patients (7%) received antibiotics alone. 16/42 (38%), not responsive to NSAIDs or colchicine, received glucocorticoids. Patients who received glucocorticoids at the first episode relapsed earlier (median time of 2.1 months range 10 days-5 months), than patients treated with NSAIDs (6.6 months range 10 days -24 months) or with colchicine (5 months range 10 days-5 months) (p<0.05). In our study, initial treatment of the first episode did not affect the number of subsequent flares. To evaluate treatment strategy at relapses, we divided our study population in two groups: Group 1 (20 pts) in which recurrence was treated with NSAIDs, colchicine or glucocorticoid (alone or combined); group 2 (22 patients) in which anakinra was started. Among patients belonging group 2, 9 received anakinra at first relapse, 7 at the second, 2 at the third and 2 at the fourth. Anakinra treatment was followed by a prompt resolution of symptoms and inflammatory signs within 2 days. During daily treatment with full dose anakinra, no relapses were reported over a median of 13.3 months (range 5-24 months). In 13 out of 22 patients, anakinra was gradually tapered reducing the days of administration during the week. Four of these patients relapsed. The mean time from the start of anakinra to tapering was 17 ± 4 months (range 14-23 months) in the 4 patients who experienced a relapse versus 14 ± 4 months (range 7-21 months) in patients who did not flare, with no statistical difference. Among the 22 patients belonging to group 2 anakinra was finally discontinued in 11 after a mean time of 23.4 months (range 12-36). Among these, 8 relapsed after anakinra withdrawal including 2 of the 4 patients already relapsed during tapering). Only 3 patients didn't present any relapse (up to 20.3 months of follow-up). All patients who relapsed responded quickly to the reintroduction of anakinra

Conclusion: Our study confirms the lack of a standardized treatment approach in patients with recurrent pericarditis. Patients treated with glucocorticoid at first episode relapse before than those treated with other drugs. Anakinra is an effective treatment; however, tapering/discontinuation of the drug lead to relapses in several cases. Further experience on larger population is needed to define the best treatment duration and approach to withdrawal of IL-1 inhibitor

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