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IL-1 blockade in paediatric recurrent pericarditis: a multicentric retrospective study of the Italian cohort

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Introduction: Acute pericarditis is an inflammatory condition causing the occurrence of pericardial effusion. In a third of patients, the disease is recurrent. First line treatment of idiopathic pericarditis consists in non-steroidal anti-inflammatory drugs (NSAIDs) and colchicine; glucocorticoids represent the second line treatment in resistant or intolerant cases. A recent clinical trial has enlightened the effectiveness of anakinra in adults and paediatric patients with colchicine-resistant recurrent pericarditis.
Objectives: To describe the clinical characteristics and response to treatment in a cohort of paediatric patients with recurrent pericarditis treated with IL inhibitors.
Methods: paediatric patients with recurrent pericarditis followed at 19 Italian centers of paediatric rheumatology or cardiology and treated with IL1 inhibitors were included in the study. Demographic, clinical and response to treatment data were retrospectively collected.
Results: 55 patients were included in the study. The mean age at onset of the first episode of pericarditis was 12.53 years. The mean number of relapses of pericarditis before the beginning of treatment with IL1 inhibitors was of 3.4 (range 1 -10). 53 out of 55 patients had previously received treatment with NSAIDS and 44 with colchicine. 44 patients received steroidal treatment: 2 of them displayed a steroidal-resistance and 39 steroidal-dependence with reoccurrence of the symptoms following any attempt to reduce or withdraw this treatment. Anakinra (mean dosage of 1.67 mg/kg/day) was used as first Il-1 inhibitor in 54 of the 55 patients, Canakinumab (150 mg every 4 weeks) in one. 53 out of 54 patients treated with anakinra displayed a complete clinical response to treatment within a mean of 2 days (range 1-7): NSAIDs, glucocorticoids and colchicine were withdraw in 25 out of 26, 31 out of 35 and 15 out of 32 patients respectively. 50 of 54 patients displayed a complete response; among these, three were switched to Canakinumab, 17 patients continued treatment at the same dosage while in 30 patients a reduction of treatment was attempted. 12 patients presented, during anakinra tapering, a disease flare, promptly resolved after an increasing of the dosage. The remaining 18 patients did not present any flare despite the reduction of the drug. Anakinra was withdrawn in 16 patients, with recurrence of the symptoms in 11. 5 patients were treated with Canakinumab: 1 as first anti-IL1 drug, 4 were switched from anakinra (two for poor compliance, one for side effects and one for incomplete control of the disease). 2 out of five patients had a complete control of the diseases, 2 patients discontinued the treatment because of inefficacy and 1 patient required low dose of glucocorticoids to control the disease. At last follow-up 34 patients were on anakinra, 7 on anakinra and colchicine, 2 on canakinumab, 1 oncanakinumab plus colchicine and NSAIDs. In 9 patients all treatments were withdrawn for complete control of the disease.
Conclusion: This study confirm the effectiveness of IL-1 blockade in paediatric patients with recurrent pericarditis. However most of the patients require prolonged treatment to maintain clinical remission. Moreover, in our cohort of patients the rate of response was higher for anakinra then for canakinumab, suggesting a possible role of IL1α in the pathogenesis of this condition.

Disclosure of Interest
None Declared

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Efficacy and safety of anakinra in the treatment of inflammatory heart failure in myocarditis

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Introduction: Virus-negative myocarditis (VNM) is a severe, inflammatory heart disease with a poor prognosis, and is a leading cause of inflammatory dilated cardiomyopathy(i-DCM). Therapies are limited. Preliminary data indicate that interleukin-1 (IL-1) plays a key role in the initiation and maintainance of the inflammatory heart response, sustaining an auto-inflammatory cycle [1,2].
Objectives: to evaluate the efficacy and safety of anakinra (ANK) in improving Left Ventricular Ejection Fraction (LVEF) on transthoracic echocardiography (TTE) and other cardiovascular parameters in patients with VNM.
Methods: Biopsy-proven VNM patients were enrolled and treated with ANK 100 mg daily subcutaneously. All patients also received treatment with the maximum tolerated dose of any beta blockers and ACE-inhibitors, according to current guidelines. At baseline and 8±4 weeks after ANK therapy initiation, all patients underwent a full evaluation with assessment of their functional status (New York Heart Association [NYHA]), measurement of high-sensitive troponin T (hs-TnT) and NT-proBNP serum levels, electrocardiography (ECG), 24h-ECG Holter, TTE and cardiac magnetic resonance (CMR). Any myocarditis-related complication, cardiovascular deaths and adverse events (AEs) were recorded during follow-up. Continuous variables were assessed with the Wilcoxon signed-rank test for non-parametric tests and a p value<0.05 was considered statistically significant.
Results: eleven patients with EBM-proven myocarditis were enrolled and treated with ANK. Nine patients received ANK as first line therapy, and in 5 cases ANK was used as monotherapy; ANK was combined with prednisone (mean dose 31,7±16,7 mg daily) in 6 patients, 5 of them were concomitantly treated with azathioprine. Demographic and baseline clinical characteristics of our cohort are summarized in table 1. Mean LV-EF on TTE at baseline was 38,7%±19,6, with comparable findings on CMR (36,45%±18,0), and 8 patients (72.7%) had a depressed LV-EF(<55%). At baseline, mean levels of hs-TnT and NT-proBNP were 150,0 ± 153,9 ng/L and 6968,8 ± 10788,4 pg/ml, respectively. Hs-TnT and NT-proBNP levels were elevated in 10 (90.9%) and 9 patients (81.8% respectively. At 8 weeks, LV-EF improved in 10 patients (90.9%). The LV-EF increase was >10% in 5 patients (45.5%) and between 5-10% in 5 cases (45.5%); only 1 patient showed a <10% LV-EF decrease. Mean LV-EF at the end of follow-up improved to 49,4%±10,8 (p=0.059). When evaluating the 8 patients with baseline reduced LV-EF, the LV-EF improvement was statistically significant (baseline 29,25% ±12,9; after ANK 45.2% ±9.2, p=0.025). The LV-EF amelioration was paralleled by clinical improvements in all patients, since the majority of them (90.9%) were in NYHA class I-II at the end of follow-up(vs 27.3% at baseline). Consistently, hs-TnT declined after 8 weeks (64,6±100,7 ng/L,p=0,028), and a similar trend was observed for NT-proBNP, even though this not reach