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THU0570 EFFICACY AND SAFETY OF ANAKINRA IN THE TREATMENT OF AUTOIMMUNE MYOCARDITIS

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Background: Virus-negative or "autoimmune" 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 maintenance 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) in patients with VNM.

Methods: Biopsy-proven VNM patients were enrolled and treated with ANK 100 mg daily subcutaneously. All patients 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, all patients underwent a full evaluation with assessment of 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) was 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(F/M=5/6, mean age 46,2±12,2 years) diagnosed with EBM-proven VNM were enrolled. 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. On EMB, 6 patients were classified as i-DCM, 3 with acute VNM e1 with active and chronic VNM³⁻⁴. Clinical onset was characterized by congestive heart failure in the most cases(72.7%). The majority of patients(72.7%) was in NYHA class III-IV. 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 29,2%±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. 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 not statistical significant(2582,6±5048,1 pg/ml,p=0,06). We did not observe any myocarditis-related death or complications, nor any ANK-related AEs.

Conclusion: Our pilot study supports the efficacy and safety of ANK in the treatment of inflammatory heart failure in VNM and provides the first

clinical evidence to support the therapeutic blockade of IL-1 in myocarditis.

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THU0571 THE IMPACT OF AGING ON FAMILIAL MEDITERRANEAN FEVER PATIENTS

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Background: Familial Mediterranean Fever (FMF) is a monogenic autoinflammatory disorder with innate immune activation with an onset before age 20 in approximately 90% of the patients. There is scarce data on the effect of aging on FMF patients over 40 years of age

Objectives: This study aims to collect data on FMF patients who have survived over 40 years of age. Here we report our preliminary data on disease course and treatment status and comorbidities of our patients with FMF.

Methods: Among the FMF patients who have been followed in our FMF outpatient clinic with a pool of approximately 5000 patients, those who have aged 40 and over are being included to the study. As by today 180 patients are considered for evaluation. The files of patients were reviewed and a standard questionnaire was used to interview the patients. Here we report the results of 100 of these patients (56%) who were contacted for this purpose. These patients were questioned on their demographic characteristics, comorbid conditions, colchicine treatment details, and attack information. In order to see the trend of the change in the parameters assessed, the patients were divided into two groups based on their present age (Group 1: 40-50 years, Group 2: ≥50 years).

Results: A total of 100 (78 F, 22M) patients were evaluated. There were 61(46F, 15M) patients aged between 40-50 years and 39 (32F, 7M) over 50. The demographic characteristics and clinical features of these patients are given in Table 1. Besides 3, all patients were still on colchicine regularly. Ninety-six percent of the patients declared overall benefit from colchicine therapy; however 38% experienced a side effect related to this treatment. Over 88% of the patients reported decrease in severity and frequency of FMF attacks. The mean daily colchicine dose was lower in the age 50 and over group (1.7±0,77 mg versus 1,35 ±0,38 mg). There were no patients with AA amyloidosis in neither age group. The mean duration from the last attack increased from 15.3 ±19.7 months to 35.6±52 months in the older patients. One or more additional disease was present in 75% of this patient group. Among the comorbidities hypertension was the most frequent, diagnosed in 25% of the patients, followed by hypothyroidism (16%), diabetes mellitus (10%) and cardiac disease (5%). Sixty-five of the patients were receiving other medications in addition to colchicine.

Table 1. Clinical course and co-morbidities in two age groups over 40 years

n	Group 1* (n=61)	Group 2** (n=39)	p
Sex (F:M); current age (mean±SD) (yr)	(46 :15);45.5 ± 2.29	(32:7); 57.05 ± 6.81	0,43; <0.001
Mean duration since the last episode, (mean±SD, mo)	15.3 ± 19.7 (1-60)	35.67 ± 52.05 (1-276)	0,012
Number of patients on colchicine therapy, n (%)	59 (96.7)	38 (97.4)	NS
Mean colchicine dose, mg/day (current)	1.7±0.76	1.41±0.45	0.03
Number of patients with decrease in attack severity, n (%)	54(88,5)	35 (89,7)	NS
Number of patients with decrease in attack frequency, n (%)	57(93,4)	37 (94,8)	NS