it was easily misdiagnosed. The delay in establishing the diagnosis of RPC was common. A survey from the United Kingdom found a median length of 1.9 years from the first disease attack to diagnosis². One recent research reported 64% of RPC patients had a diagnostic delay with more than five years³.

Objectives: We made a retrospective study to explore distinct characteristics of relapsing polychondritis with arthritis as the first attack. By comprehending the nature of disease fully, misdiagnosis at the early phase of illness could be avoided.

Methods: The clinical features and prognosis of 7 RPC patients in Peking Union Medical College Hospital between October 2012 and October 2018, presenting as arthritis at onset, were retrospectively analyzed.

Results: There were five female patients. The female to male ratio of the case series was 5:2. The age of 7 patients was 43.43 ± 9.86 years. The number of affected joints was 20.14 ± 10.92 . The joint involvement was most common in the bilateral proximal interphalangeal joints (PIPs) and interphalangeal joints of the thumb. Recurrent arthritis occurred in 5 patients. There was no accompanying enthesitis. The specific serum indexes of RA, such as rheumatoid factor (RF), anti-cyclic citrullinated peptide (CCP) antibody, were negative. There were three patients administered with conventional synthetic disease modifying anti-rheumatic drugs firstly, and they did not respond well. The median delay time from onset to diagnosis of RPC was ten month (Figure 1). The deformity and destruction of the joint were not observed in all patients, by clinical and radiological assessment. The average duration from arthritis to the occurrence of other involved system was 8.3 months. Four patients were misdiagnosed as rheumatoid arthritis at first. One patient was considered as ankylosing spondylitis at first. Two patients were diagnosed as arthritis at the beginning. As regards to treatment, glucocorticoids (GCs), cyclophosphamide and mycophenolate mofetil were used. Two patient was complicated with palmoplantar pustulosis and Kikuchi-Fujimoto disease. During follow up, myelodysplastic syndrome occurred in one patient. After following up for 35.43 ± 30.92 months, all patients survived and were not with the recurrence of arthritis.

Figure 1. The demonstration of multiple system involvement and the interval from onset to diagnosis in seven patients with relapsing polychondritis.

Conclusion: RPC patients with arthritis onset were characteristic of middle-aged women and symmetric multiple joint involvements. The RF and anti-CCP antibody were negative. The arthritis was recurrent and not erosive. The interphalangeal joints of the thumb and PIPs were the most frequently involved joints. In clinical practice, physicians should be alert to the possibility of RPC, when encountered the patients with abovementioned arthritis. The inquiry about manifestations of other system involvement in RPC and a regular follow-up was warranted. After the combined therapy of GCs and immunosuppressants, all patients had a favorable prognosis.

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THU0569

MANAGEMENT OF ADULT-ONSET STILL'S DISEASE (AOSD) WITH IL-1 INHIBITORS: EVIDENCE- AND CONSENSUS-BASED STATEMENTS BY A PANEL OF ITALIAN EXPERTS

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Background: Still's disease is a rare autoinflammatory disease, presenting in both pediatric [systemic juvenile idiopathic arthritis (SJIA)] and adult patients [adult-onset Still's disease (AOSD)]. Due to the rarity of the disease, clinical trials are limited and treatment guidelines are not available. In patients refractory to the classical therapy with NSAIDs, corticosteroids and DMARDs, the introduction of drugs targeting IL-1 has greatly expanded treatment options. Among these, canakinumab, a human monoclonal anti-IL-1 β antibody, and anakinra, a human recombinant IL-1RA, have been recently approved for the treatment of refractory patients.

Objectives: To produce recommendations, based on evidence and expert consensus, that can help clinicians in choosing the most appropriate treatment of AOSD, with particular attention to anti-IL-1 therapies, in order to achieve disease remission before the development of complications.

Methods: The recommendations development process took place from April to October 2018 and consisted of three steps. The first step was dedicated to a comprehensive literature review and development of statements. Two separate literature searches were performed: a) similarities and differences between SJIA and AOSD; b) efficacy and safety of IL-1 blockade in AOSD. The issue related to the treatment of AOSD with anti-IL1 therapies was specified into 4 questions: 1) efficacy and safety; 2) comparison among IL-1 inhibitors; 3) early versus late treatment; 4) systemic versus chronic articular pattern of the disease. In the second step, the statements were submitted in a Delphi process to a panel of 67 rheumatologists. Consensus threshold was set at 66%: positive, > 66% of voters selected scores 3 to 5; negative, > 66% of voters selected scores 1 or 2. At the third step of the consensus process, the voting results were analyzed, and the statements were finalized.

Results: In the two literature searches, 332 and 358 publications were identified; 30 and 25 publications, respectively, were selected according to the inclusion criteria. Based on the review of the literature and personal clinical experience, 11 statements were developed. 48/67 rheumatologists (72%) participated to the Delphi process. Positive consensus was reached after the first round of voting and was full (> 95%) on the majority of statements. A large consensus was achieved in considering AOSD and SJIA as the same disease. The use of anti-IL-1 therapies in refractory patients was considered quite safe and effective both as first and as subsequent line of biologic treatment, especially in systemic patients. Because of the lack of head-to-head comparisons, a different profile of efficacy among IL-1 inhibitors could not be established. There was a large consensus that failure of the first IL-1 inhibitor does not preclude a therapeutic response with another one. The lack of studies comparing early versus late treatment in AOSD patients did not allow to draw conclusions, however data from SJIA suggest a better response in early treated patients.

Conclusion: The Delphi method was used to develop recommendations that we hope will help clinicians in the management of patients with AOSD refractory to conventional therapies.

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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