AB1290 SOMETIMES IT IS ALL IN THE HEART; CARDIAC INVOLVEMENT IN BEHCET'S SYNDROME

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Background: Cardiac involvement is an uncommonly seen but might be a fatal clinical manifestation of Behcet's syndrome (BS), if not recognized early and treated aggressively.
Objectives: Our aim was to retrospectively evaluate the characteristics of cardiac involvement in Behcet's syndrome and investigate possible risk factors.
Methods: 800 patients with BS diagnosis according to International Study Group for Behcet's Disease (ISG) criterias in our center were screened. 14 patients with cardiac involvement were included in this study.
Results: All the patients were male with positive smoking history. The age at the onset cardiac involvement was 32.5 ± 7.8 years. In 3 patients, diagnosis of BS was made simultaneously with cardiac involvement. Types of cardiac involvement were as following; intracardiac thrombosis (ICT) in 8, right coronary artery aneurysm in 2, and myocardial infarction in 4 patients. The majority of ICT was seen in right ventricle followed by right atrium (75% vs 25%). Deep vein thrombosis (DVT) was the most common accompanying other vascular involvement (57%).Thrombophilic factors were studied in 8 patients, whom only 2 cases were found positive for heterozygous factor V Leiden mutation. Cyclophosphamide was the most common used immunosuppressive agent and anticoagulation was used in 9 patients (Table 1).

Table 1. Characteristics of cardiac involvement in BS patients

Male/female, (n), (%)	14 (100%)
Diagnosis age, years (Mean ± SD)	30 ± 7
Age of cardiac involvement, years, (Mean ± SD)	32.5 ±7.8
Smoking history, (n), (%)	14 (100%)
Other vascular involvement, (n), (%)	12 (85%)
-Deep vein thrombosis, (n)	8
-Pulmonary involvement, (n)	4
-Peripheral artery aneurysm, (n)	1
Cardiac involvement patterns	
-ICT, (n), (%)	
• RV	6
• RA	2
-CAA, (n), (%)	2(14.3%)
-AMI, (n), (%)	4(28.6%)
Immunosuppression, (n)	
-Steroid	14
-CYC	9
-AZA	3
-IFN	1
Anticoagulation, (n)	9

ICT; intracardiac thrombosis, RV; right ventricle, RA; right atrium, CAA; coronary artery aneurysm, AMI; acute myocardial infarction, CYC; cyclophopshamide, AZA; azathioprine, IFN; interferon

Conclusion: Most common type of cardiac involvement is pericarditis, whereas intracardiac thrombosis has its unique characteristics (1). Male gender and smoking seems to be major significant risk factors.
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AB1291 AGE OF ONSET AND DIAGNOSTIC DELAY IN PATIENTS WITH MONOGENIC AUTOINFLAMMATORY DISEASES AND SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS - DATA FROM THE FEDERAL CENTER IN RUSSIAN FEDERATION

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Background: Autoinflammatory diseases (AIDS) are a group of rare monogenic and multifactorial diseases manifested by systemic inflammation, organ disorders, the development of complications, primarily amyloidosis, and possible death without timely targeted therapy. Rapid diagnosis contributes to the early appointment of treatment, prevention of the development of organ damage and improvement of prognosis in such patients.
Objectives: To establish the age of the onset and the duration of the delay of diagnosis in patients (pts) with monogenic AIDS (m AIDS) and systemic juvenile idiopathic arthritis (sJIA) according to the Federal Rheumatology Center.
Methods: 273 pts were included in the study for the period from 2007 to 2021, 158 of them with mAIDS: FMF - 75, CAPS - 50, TRAPS - 28, HIDS/MKD - 5; 115 pts with sJIA.
Results: The age of pts with mAIDS ranged from 6 months to 65 y.o. (120 children and 38 adults), among pts with sJIA there were only children aged 12 months to 17 y.o. Most of the pts with CAPS, TRAPS, HIDS/MKD were female (62.4%), as with sJIA (57.4%). FMF was more often affected by male persons (56.6%). The age of the onset of mAIDS ranged from 0 to 53 y.o., sJIA - from 4 months to 15 y.o., was the smallest in pts with CAPS and HIDS/MKD. 33.5% pts with mAIDS had the onset of the disease from birth or within the first year of life, more often with CAPS and HIDS /MKD, in the first decade of life in half of the pts (52.5%), more often in FMF and TRAPS, from 10 to 18 years - 8.2%, in adulthood - 5.7%. In pts with sJIA, the onset of the disease was mainly in the first 5 years of life – 69 (60%), from 5-10 years – 38 (33%), rarely in the first year of life and after 10 years of age (6.95%). The duration of the disease at the time of diagnosis ranged from 1 month to 48 years in FMF pts, from 1 month to 46 years with CAPS, from 3 months to 59 years with TRAPS, from 3.5 to 16.5 years with HIDS/MKD, from 3 weeks to 7.8 years with sJIA. Within 1 year the diagnosis was established in 15.2% of children with mAIDS, in 12.2% with sJIA, in the first 5 years of the disease in 35.4% and 15.6% respectively. The delay in diagnosis for more than 5 and 10 years was observed in 24.1% and 25.9% of pts with mAIDS, practically did not occur in sJIA. The least delay in diagnosis was noted in pts with FMF.
Conclusion: Most of the pts with mAIDS had the onset of the disease in the first 5-10 years of life, an earlier age of onset was noted in pts with CAPS and HIDS/MKD. To date, the delay in diagnosis in pts with mAIDS remains very long and reaches decades, especially with CAPS, TRAPS and HIDS/MKD. In pts with sJIA, the diagnosis was established faster, according our opinion a later diagnosis is associated with a polycyclic course of the disease. Earlier diagnosis and the appointment of targeted therapy are needed, which will help to avoid complications, improve the prognosis and quality of life of pts.
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AB1292 CHARACTERISTICS OF UVEITIS IN BEHCET'S SYNDROME; A RETROSPECTIVE ANALYSIS OF 109 PATIENTS

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Background: Uveitis is still an important cause of morbidity leading to vision loss in Behcet's syndrome (BS).
Objectives: Our aim in this study was to retrospectively evaluate characteristics of uveitis, long term complications, and its association with the other components of BS.
Methods: 800 patients with BS diagnosis according to International Study Group for Behcet's Disease (ISG) criterias in our center were screened and among those, data of 109 patients with BS-related uveitis were included in this study.
Results: The overall prevalence of uveitis was found 13% among all BS population. Majority of BS-related uveitis were male (65,1%/34,9%). Mean age for BS diagnosis and onset of uveitis were found 28 ± 8,48 and 27 ± 8,42 years. In this group, uveitis developed before BS diagnosis in 33% of patients,

Table 1.

	FMF	CAPS	TRAPS	HIDS/MKD	sJIA
Age (me, IQR) y	10,5[6,0;15,4]	13[7,0;31,5]	9,0[7,9;14,5]	9,0[5,0;17]	4,5[2,8;7,6]
Age of onset	3[1,5;5]	1,0[0,15;7,0]	4,0[1,0;7,1]	0,5[0,4;1,5]	4,2[2,1;6,75]
(me, IQR), y	18 (24%)	24(48%)	8 (28,6%)	3 (60%)	8 (6,95%)
≤1y	46 (61,3%)	18 (36%)	17 (60,7%)	2 (40%)	99 (86,1%)
>1-10y	7 (9,3%)	5 (10%)	1 (3,6%)	-	8 (6,95%)
>10-18y	4 (5,3%)	3 (6%)	2 (7,1%)	-	-
>18y					
Delayed diagnosis (me, IQR), y	10,5[6,0;15,4]	13[7,0;31,5]	9,0[7,9;14,5]	9,0[5,0;17]	0,25[0,1;0,5]
≤1y	11 (14,7%)	9 (18%)	4 (14,3%)	-	14 (12,2%)
>1-5y	30 (40%)	12 (24%)	12 (42,9%)	2 (40%)	18 (15,6%)
>5-10y	20 (26,7%)	11 (22%)	6 (21,4%)	1 (20%)	1 (0,9%)
>10y	13 (17,3%)	18 (36%)	8 (28,6%)	2 (40%)	-

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