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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Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2022-eular.2628

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.
Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2022-eular.2654

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%)	-

Ann Rheum Dis: first published as 10.1136/annrheumdis-2022-eular.2675 on 23 May 2022. Downloaded from http://ard.bmj.com/ on October 30, 2022 at Eskisehir Osmangazi University. Protected by copyright.

whereas 44% were diagnosed at onset of uveitis.The frequency of this population was estimated 6% among all BS patients,. Characteristics of uveitis were as following; unilateral (45/99), bilateral (54/99), anterior (12), posterior (30), panuveitis (30), retinal vasculitis (9) and not defined (9). 25 patients experienced only one attack, whereas more than one attack was seen in 45 patients during follow up. 74 patients whose complications data was available, cataract, maculopathy, glaucoma, and optic atrophy developed in 10, 7, 6 and 3 patients, respectively. Vision loss was seen in 8 patients as partial, in 28 as total (Table 1).

Table 1. Characteristics of uveitis, complications and other components of BS patients

Male/female, (n), (%)	71/28 (%65,1/%34,9)
Age at diagnosis, years (Mean± SD)	28 ±8,48
Age at uveitis, years (Mean± SD)	27 ±8,42
Uveitis-Behcet's diagnosis relationship, (n)	109
-Before, (n), (%)	36 (%33)
-At the time of, (n), (%)	48 (%44)
-After, (n), (%)	25 (%23)
Characteristics of uveitis, (n)	99/109
-Unilateral/Bilateral	45/54
-Anterior	12
-Posterior	30
-Panuveitis	30
-Retinal vasculitis	9
Complications, (n)	26/74
-Maculopathy	7
-Glaucoma	6
-Cataract	10
-Optic atrophy	5
-Vision loss (partial/total)	8/28

BS; Behcet's syndrome

Conclusion: Male gender and onset at young age are important risk factors for uveitis development. Uveitis is seen as a critical component for diagnosis. Complications, despite declining in the era of biologics use, are still a major concern. Retrospective design and missing data are limitations of this study.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2022-eular.2675

AB1293 INTRATHECAL INJECTION OF METHOTREXATE COMBINED WITH DEXAMETHASONE FOR COGAN SYNDROME WITH NEUROLOGICAL INVOLVEMENT: A CASE REPORT

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Background: Cogan syndrome (CS) is a rare autoimmune disease, mainly occurring among young people, with an incidence rate of <0.11/100000 (100000). CS is mainly characterized by interstitial keratitis, visual loss associated with cochlear vestibular involvement and Meniere type vertigo. It usually involves all systems of the body, and about 10% to 13% of cases have nervous system involvement. Fewer than 400 cases have been reported worldwide (*1*). Glucocorticoid is mainly used in acute attack as the basis of treatment, the effect was not satisfactory(*2*). Here, we report a case of CS with nerve injury as the main manifestation, which was successfully treated by intrathecal injection of methotrexate combined with dexamethasone.

Objectives: CS often involves multiple systems throughout the body and has a poor clinical prognosis. From a review of our patient's treatment experience, intrathecal methotrexate combined with dexamethasone may be an effective treatment modality for CNS involvement in patients with refractory recurrent Cogan syndrome.

Methods: The clinical manifestation, laboratory test, treatment, and outcome were described.

Results: A 31-year-old man presented with recurrent bimanual joint pain for 2 years, after which the patient developed interstitial keratitis, hearing loss and was diagnosed with CS. The patient received treatment with glucocorticoid, cyclophosphamide, methotrexate, tacrolimus, leflunomide, mycophenolate mofetil, hydroxychloroquine, adalimumab. The joint pain was improved, but the headache, dizziness, tinnitus and hearing impairment were significantly aggravated, and syncope occurred. Cerebrospinal fluid examination for lumbar puncture on admission of the patient showed CSF pressure > 185 mmH_2O, CSF microprotein 1.10 g/l, and CSF nucleated cell count 120x10^6 /L. At the beginning, we successively treated the patient with methylprednisolone (40 mg, QD), gamma globulin (10 g, QD), methotrexate (20 mg, QW) and cyclophosphamide (100 mg, QOD), but the effect was not satisfactory. After excluding infection, we treated this patient with intrathecal methotrexate 10 mg combined with dexamethasone 10 mg, and the patient's symptoms of dizziness, headache, and tinnitus improved significantly, and his hearing did not decline significantly any more. The patient received a total of seven intrathecal injections (cumulative methotrexate 60 mg + dexamethasone 65 mg) during the subsequent 6 months. His dizziness, headache, tinnitus and other neurological symptoms improved significantly, no further syncope, fever. CSF protein quantification and the number of CSF nucleated cells decreased significantly.

Conclusion: CS patients are often accompanied by multisystem injuries, so it is essential to correctly identify CS and provide appropriate treatment as soon as possible. For patients with neurological involvement who do not respond to treatment with glucocorticoids and immunosuppression, intrathecal methotrexate combined with dexamethasone may be considered as a treatment option for CS in the future.

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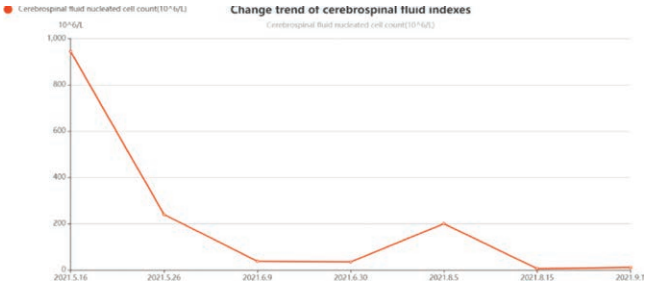


Figure 1. Change trend of nucleated cell count in cerebrospinal fluid during treatment

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2022-eular.2839

AB1294 CO-OCCURRENCE OF IGG4 RELATED DISEASE AND OTHER SYSTEMIC IMMUNO-MEDIATED DISORDERS.

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Background: IgG4-related disease (IgG4RD) is a fibroinflammatory disease with unique clinical and pathophysiological features. According to the recently released Classification Criteria, the presence of other concomitant immunological disorders (OID) represents an exclusion criterion for the classification of IgG4RD (1). Rigorous application of the Classification Criteria, however, can lead to misdiagnosis because our increasing appraisal of IgG4RD indicates that this condition may coexists with OID (2).

Objectives: To describe IgG4RD presentation in the context of OID.

Methods: IgG4RD was diagnosed based on definitive histological findings. OID were diagnosed based on specific classification/diagnostic criteria.

Results: In six out of 234 (3%) patients IgG4RD occurred in accompaniment with OID, including two patients with sarcoidosis (33%), two with granulomatosis with polyangiitis (33%) one with eosinophilic granulomatosis with polyangiitis (16%) and one with Takayasu arteritis (16%). In three cases IgG4RD preceded OID (Table 1).

Conclusion: Our data demonstrate that IgG4RD can occur in overlap with OID, especially granulomatous diseases. Further research is needed to define whether this clinical scenario is associated with different pathological bases and long-term outcomes.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2022-eular.3086

Ann Rheum Dis: first published as 10.1136/annrheumdis-2022-eular.2675 on 23 May 2022. Downloaded from http://ard.bmj.com/ on October 30, 2022 at Eskisehir Osmangazi University. Protected by copyright.