Background: Behçet's disease (BD) is an systemic inflammatory vasculitis characterized by recurrent oral aphthae and genital ulcers. In the course of the disease, skin, eye, musculoskeletal, nervous system and gastrointestinal system involvements can be detected.
Objectives: To evaluate the clinical, laboratory and radiological findings of pediatric cases diagnosed with BD.
Methods: Fifty patients (0-20 years old) who were followed up with the diagnosis of BD at the Pediatric Rheumatology outpatient clinic of Uludag University Faculty of Medicine between January 2011 and July 2021 were included in our study. The patients were diagnosed according to the diagnostic criteria of the International BD Study Group.
Results: Twenty-four (48%) of the patients were male and 26 (52%) were female. The mean age at the diagnosis of patients was 9 ±4.55 years (10.75±4.55 years in boys, 12.35±3.65 years in girls). Twenty patients (36.3%) had a family history of Behçet's Disease. Oral aphthae were present in 96% (n=48) patients, while genital ulcers were in 32% (n=16) (Table 1). Of the nine patients with uveitis, 6 had panuveitis, 2 had anterior uveitis and 1 had posterior uveitis. . There was no difference in the distribution of symptoms according to the gender of the patients. HLA-B51 allel was found in 39 (78%) and ANA was positive in 14 patients (28%). Immunological tests showed that serum immunoglobulins were low in 11 (32.3%) of 34 patients. Low IgG levels were detected in 6 patients, low IgM levels were in 3, and low IgA levels were in 2. Thrombus was presented in three cases (thrombus in the right ventricle in one case, in the intracranial transverse sigmoid sinus and left jugular vein in two cases). The most commonly used drug for aphthae was colchicine (n=45, 82%). The use of biological agents according to patient manifestations is shown in Table 2. In the follow-up, clinical findings improved in 35 patients (70%) Complete improvement was detected only with biological agents in 8 patients with uveitis. One patient was operated due to the development of complicated cataracts secondary to uveitis. Three patients were diagnosed with Covid-19, one of them was followed without treatment, while two of them were treated with favipiravir at home. Three patients with Covid-19 infection were using only colchicine treatment.

Table 1.

Findings at the time of diagnosis	n	%
Uveitis	9	18
Skin findings	18	36
Gastrointestinal tract involvement	19	38
Central nervous system involvement	6	12
Vascular involvement	3	6
Arthritis	17	34

Table 2.

Findings Biological Agents	Uveitis (n)	Arthritis (n)	Venous thrombus (n)	Central nervous system involvement (n)
Azathioprine	2	2	3	1
Methotrexate	2	2	-	-
Adalimumab	3	1	-	-
Infliximab	3	1	-	-

Conclusion: Behçet's disease is rare in childhood. Although it is not common, life-threatening complications can be observed. To reduce morbidity and complications, physicians should be aware of manifestations and rare clinical pictures of the BD.
REFERENCES:
[1] Hu YC, Chiang BL, Yang YH. Clinical Manifestations and Management of Pediatric Behçet's Disease. Clin Rev Allergy Immunol. 2021 Oct;61(2):171-180.
[2] Costagliola G, Cappelli S, Consolini R. Behçet's Disease in Children: Diagnostic and Management Challenges. Ther Clin Risk Manag. 2020 Jun 11;16:495-507.
[3] Balt J, Jamyanjav B, Jav S, Dandii Z, Ganbold C, Horie Y, Lennikov A, Uehara O, Ohno S, Kitaichi N. Clinical features of Behcet's disease in Mongolia: a multicenter study. Clin Rheumatol. 2020 Sep;39(9):2697-2706.
[4] Muruganandam M, Rolle NA, Sibbitt WL Jr, Cook GB, Emil NS, Fangtham M, Reiter KJ, Bankhurst AD. Characteristics of Behcet's Disease in the American Southwest. Semin Arthritis Rheum. 2019 Oct;49(2):296-302.
Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2022-eular.481

AB0580 BEHÇET'S DISEASE IN OSLO NORWAY IN PERIOD 1999 – 2021

J. L. Lorentsen[1], K. Lerang[1], T. Garen[1], Ø. Palm[1]. [1]*Rikshospitalet, Rheumatology, Oslo, Norway*

Background: Bechet's disease (BD) is a multisystemic, inflammatory disease, characterized by variety of clinical manifestations, including mucocutaneous, ocular, vascular, neurological (CNS) and gastrointestinal (GI) involvements. The disease can be seen at any age and occurs across gender, ethnic groups, and geographical areas. However, there are only a few studies from northern Europe.
Objectives: To study the demographics and clinical feature of patients with Bechet's disease (BD) in Oslo Norway.
Methods: In this retrospective study we reviewed hospital records from Oslo University Hospital (OUS), a referral hospital for systemic inflammatory vasculitis in Norway. All patients living in Oslo with ICD-10 code M35.2 were identified, and their BD-related data recorded. Those fulfilling either the International Study Group criteria (ISG) or International Criteria for Bechet's disease (ICBD) were included. Patients were defined as non-Norwegian when none of the parents was of Norwegian ancestry. The differences between groups were compared by Chi-square test using the Statistical Package for the Social Sciences (SPSS).
Results: We identified a total of 58 patients fulfilling the inclusion criteria (Table 1). Among them, 30 (52%) were females and 25 (43%) of Norwegian ancestry. Mean age at onset was 26 years, and at diagnosis 32 years.

Table 1. Demographics of Behcet`s patients in Oslo Norway

Counts (%)	Norwegian ancestry	Non-Norwegian ancestry	All (%)
All (%)	25 (43)	33 (57)	58 (100)
Male (%)	8 (29)	20 (71)	28 (48)
Female (%)	17 (57)	13 (43)	30 (52)
ISG Criteria (%)	20 (47)	23 (53)	43 (100)
ICBD Criteria (%)	25 (43)	33 (57)	58 (100)
Age at diagnosis mean (SD) years	33 (11.85)	32 (9.36)	32 (10.41)
Age at onset mean (SD) years	24 (9.34)	27 (9.52)	26 (9.42)

The most prevalent clinical manifestations were oral ulcers (n=58, 100%), genital ulcers (n=49, 84%) and skin lesions (n=40, 69%). Ophthalmic complications (n=21, 36%), arthritis (n=20, 34%), vascular involvement (n=7, 12%), CNS (n=3, 5%) and gastrointestinal (GI) complications (n= 4, 7%) were also present. There was no significant difference of clinical features between patients with Norwegian ancestry and those with non-Norwegian ancestry (data not shown).

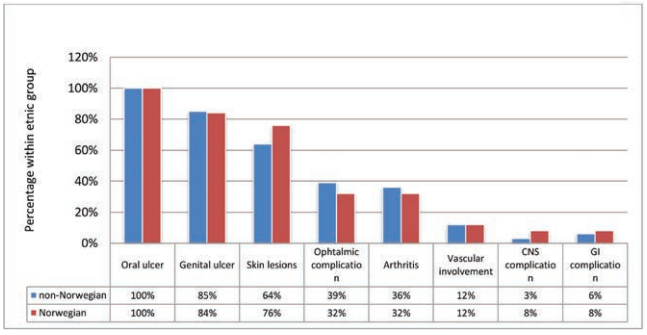


Figure 1.

Conclusion: BD was most prevalent among patients of non-Norwegian ancestry. We found a wide spectrum of clinical manifestations occurring during the disease course. Mucocutaneous lesions were most frequent, but potential serious manifestations were also present. These manifestations were similarly distributed between patients with Norwegian and non-Norwegian ancestry (Figure 1).
REFERENCES:
[1] Davatchi F. et al. Behcet`s disease: epidemiology, clinical manifestations and diagnosis. Expert Review of Clinical Immunology, 13:1, 57-65, DOI: 10.1080/1744666X.2016.1205486
Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2022-eular.635

AB0581 TRACHEAL STENOSIS; AN IMPORTANT INVOLVEMENT IN GRANULOMATOSIS WITH POLYANGIITIS

O. Ozdemir Isik[1], F. Tuncer[1], O. Sadioglu Cagdas[1], A. Yazici[1], A. Cefle[1]. [1]*Kocaeli University, Faculty of Medicine, Rheumatology, Kocaeli, Turkey*

Background: Granulomatosis with polyangiitis (GPA) is a granulomatous systemic vasculitis of unknown etiology that can affect many organs. Approximately 90% of patients with GPA have upper respiratory tract involvement, including the nasal cavity, sinuses, ear and trachea. Subglottic stenosis (SGS) may develop in some patients due to tracheal involvement.
Objectives: It is aimed to evaluate the general characteristics, treatments and disease prognosis of our GPA cases with tracheal stenosis as it is a significant cause of morbidity and mortality.

Ann Rheum Dis: first published as 10.1136/annrheumdis-2022-eular.795 on 23 May 2022. Downloaded from http://ard.bmj.com/ on December 16, 2022 at Kocaeli Universitesi. Protected by copyright.

Methods: The data of 48 patients diagnosed with GPA between 2000-2021 were analyzed retrospectively and the data of 6 patients with tracheal stenosis (TS) were evaluated.
Results: TS was present in 13% of the patients. All patients with TS were female. The mean age of the patients with TS was 46.5±6.5 years, the mean age of disease onset was 35.5 ±13.4 years, and the mean disease duration was 10.8±10.2 years. Constitutional symptoms were in 17% of patients, and 17% had mastoiditis, sinusitis, otitis, bloody nasal discharge and renal involvement. None of the patients had hearing loss, skin, eye and neurological involvement. 83% of patients had limited disease. While all patients had SGS, one patient also had glottic and bronchial stenosis. The most common symptoms in patients were hoarseness and shortness of breath. All patients received systemic treatment according to their organ involvement, local treatment (dilatation, steroid injection) was also applied to two patients. All patients had anti-neutrophil cytoplasmic antibody (ANCA) positivity. When patients with and without tracheal stenosis were compared, there was a difference in terms of gender, lung and kidney involvement, presence of anemia, increased erythrocyte sedimentation rate, pANCA, cANCA and PR3-ANCA positivity, and myalgia. In addition, our patients with tracheal stenosis had a younger age of onset and a longer delay in diagnosis.
Conclusion: The incidence of SGS in large series has been reported as 8-23% of GPA patients.[1,2] Although the number of cases in our study was small, this rate was 13%. Symptoms range from cough and shortness of breath to progressively life-threatening severe stridor. Early diagnosis and treatment are important. For this reason, TS is strongly emphasized.
REFERENCES:
[1] Girard C, Charles P, Terrier B, Bussonne G, Cohen P, Pagnoux C, et al. Tracheobronchial stenoses in granulomatosis with polyangiitis (Wegener's). Medicine. 2015;34:1-6.
[2] Costantino CL, Niles JL, Wright CD, Mathisen DJ, Muniappan A. Subglottic stenosis in granulomatosis with polyangiitis: therole of laryngotracheal resection. Ann Thorac Surg 2018;105:249-53.

Table 1. Comparison of data from GPA patients with and without tracheal stenosis

N(%)	Tracheal Stenosis +GPA(n=6)	Tracheal Stenosis - GPA (n=42)	P
Gender Female	6 (100)	21(51)	***0.031***
Male	0	20(49)	
Mean Age	46.5±6.5	55.6±13.2	0.095
Symptom Onset Age	35.5±13.4	49.3±14.5	***0.041***
Delay in Diagnosis (months)	44.2±6.5	9.98±14.8	***0.023***
Myalgia	2(33)	33(81)	***0.030***
Fever	1(17)	21(51)	0.194
High ESR	1(17)	31(78)	0.007
High CRP	5(50)	32(80)	0.138
Anemia	1(17)	28(70)	***0.020***
Upper Respiratory Involvement	6(100)	24(59)	0.074
Lung Involvement	3(50)	36(90)	***0.037***
GIS involvement	1(17)	6(15)	1
Eye Involvement	-	11(27)	NA
Skin Involvement	-	12(29)	NA
Urogenital Involvement	-	1(2)	NA
Neurological Involvement	-	13(32)	NA
Renal Involvement	1(17)	29(71)	***0.018***
ANCA positivity	6(100)	38(93)	1
MPO-ANCA	2(33)	4(10)	0.162
PR3-ANCA	1(17)	33(81)	***0.004***
p-ANCA	5(83)	6(15)	***0.000***
c-ANCA	1(17)	31(78)	***0.001***

Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2022-eular.795

AB0582 TAKAYASU ARTERITIS PRESENTS AT AN OLDER AGE IN EUROPEAN POPULATION

F. Coath[1], G. Ducker[1], T. Eccleston[1], S. Fordham[1], C. Mukhtyar[1] on behalf of Mukhtyar Vasculitis group. *[1]Norfolk & Norwich University Hospital, Rheumatology, Norwich, United Kingdom*

Background: The definition of Takayasu arteritis includes the age of onset of disease as being <50 years. It has been reported most frequently in young females from Asian populations.
Objectives: We investigated whether the epidemiological features are the same in Norfolk, UK, where the population is predominantly of Northern European ancestry.
Methods: In our centre, cases are classified using validated international criteria. The electronic records of our vasculitis clinic were reviewed to identify all individuals with a new diagnosis of large vessel vasculitis between 2011-2020 who met a modified ACR 1990 criteria for Takayasu arteritis. The original criteria include decreased brachial artery pulse, bruit over subclavian arteries or aorta, and arteriogram abnormality. The respective modifications of these three criteria were to accept decreased radial pulse as well as the brachial pulse, bruit over the axillary artery as well as the subclavian artery, and abnormality of positron emission tomography and/or ultrasonography instead of arteriograms.
Results: 15 individuals met the modified ACR 1990 criteria for Takayasu arteritis. 2 of them also met the criteria for GCA and were excluded. 13 individuals were classified as having Takayasu arteritis. 11 were female. The mean (standard deviation) age at diagnosis was 66.9 (8.9) years. None of individuals were <40 years old. On presentation - 9 had upper limb claudication, 7 had pulse discrepancy, 3 had a blood pressure difference >10 mm Hg systolic, 7 had an audible bruit over either subclavian or axillary artery. The diagnosis was confirmed on positron emission tomography in 8. Ultrasonography of extracranial arteries was abnormal in 7. The mean (standard deviation) ESR was 53.2 (33.6) and CRP was 37.5 (32.8).
Conclusion: Takayasu arteritis can present at an older age in Norfolk, UK, where the population predominantly has Northern European ancestry. This has implications for treatment, identification of immunogenetic factors and disease classification.
REFERENCES:
[1] Arend WP et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum 1990; 33(8), 1129-34.
Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2022-eular.1151

AB0583 REFERRAL PATTERN AND TREATMENT OF POLYMYALGIA RHEUMATICA IN GENERAL PRACTICE: AN INTERNATIONAL QUESTIONNAIRE BASED STUDY

A. Overgaard Donskov[1], S. Mackie[2], E. M. Hauge[1,3], C. Toro Gutiérrez[4], A. Hemmig[5], A. Van der Maas[6], B. Dalsgaard Nielsen[7], I. Hansen[1,3], M. Yates[8], L. Frølund[1], K. Douglas[9], K. Van der Geest[10], E. Rezus[11], S. Monti[12], M. Gromova[13], V. Ocampo[14], S. Appenzeller[15], M. Erraoui[16], A. Ajibade[17], A. Marun Lyrio[18], R. Grainger[19], M. Sandovici[10], T. Helliwell[20], E. Brouwer[10], C. Dejaco[21,22], K. Keller[1,3] on behalf of The International PMR/GCA Study group. *[1]Aarhus University Hospital, Department of Rheumatology, Aarhus, Denmark; [2]Leeds Teaching Hospital NHS trust, Rheumatology, Leeds, United Kingdom; [3]Aarhus University, Clinical Medicine, Aarhus, Denmark; [4]Centro de Referencia en Osteoporosis & Reuatología, Reumatología, Cali, Colombia; [5]University Hospital Basel, Rheumatology, Basel, Switzerland; [6]St Maartenskliniek, Rheumatology, Nijmegen, Netherlands; [7]The Regional Hospital in Horsens, Department of Medicine, Horsens, Denmark; [8]Norfolk and Norwich University Hospital, Rheumatology, Norwich, United Kingdom; [9]Dudley Group NHS, Rheumatology, Dudley, United Kingdom; [10]University of Groningen, University Medical Center Groningen, Department of Rheumatology and Clinical Immunology, Groningen, Netherlands; [11]Clinical Rehabilitation Hospital, Rheumatology, Lassi, Romania; [12]University of Pavia, Rheumatology, Pavia, Italy; [13]Pirogov National Russian Research Medical University, Faculty Therapy, Moscow, Russian Federation; [14]Scarborough Health Network, Reumatology, Toronto, Canada; [15]University of Campinas, Rheumatology, Campinas, Brazil; [16]Ibn Zohr University, Department of Rheuamatology, Agadir, Morocco; [17]Obafemi Awolowo University Teaching Hospitals Complex, Rheumatology Unit., Ile-Ife, Nigeria; [18]PUC Campinas Hospital, General Medicine / Rheumatology, Campinas, Brazil; [19]University of Otago, Department of Medicine, Otago, New Zealand; [20]School of Medicine Keele University, Global Health, Staffordshire, United Kingdom; [21]Hospital of Bruneck, Department of Rheumatology, Bruneck, Italy; [22]Medical University Graz, Department of Rheumatology, Graz, Austria*

Background: In most countries polymyalgia rheumatica (PMR) is diagnosed and managed by both general practitioners (GP) and rheumatologists. However, the referral pattern from GP's to specialist around the world has not been described. The initial prednisolone dose recommended by EULAR/ACR is between 12.5 and 25 mg[1], but little is known about whether these guidelines are followed everywhere by GP's in clinical practice[2].
Objectives: This study aims to describe the refererral pattern and treatment strategy for PMR in general practice in several countries worldwide.
Methods: An English language questionnaire was drafted by a working group of rheumatologists and GP's from 6 different countries. The questionnaire contained questions on: 1: Respondent, 2: Referral pattern and 3: Prednisolone. Questionnaires were distributed to GP's via members of the International PMR/GCA study group. Answers were collected via an online survey tool (Redcap), from 3rd of November 2021 to 27th of January 2022. Countries with more than 15 responders to the questionnaire were included in the analysis.
Results: Data from 11 countries were analysed. Referral patterns differed widely among countries (Table 1). Almost all patients initially seen by rheumatologists were returned to GP's for treatment. In all countries a proportion of the GP's prescribed higher initial prednisolone doses than recommended, with a large variation between countries (Table 1).

Ann Rheum Dis: first published as 10.1136/annrheumdis-2022-eular.795 on 23 May 2022. Downloaded from http://ard.bmj.com/ on December 16, 2022 at Kocaeli Universitesi. Protected by copyright.