Objectives: In this study, we aimed to evaluate the clinical features and course regarding cluster analysis of BD in Kurdish patients in Van provience, Eastern Turkey at the intersection of the silk road.
Methods: This retrospective observational study was carried out in a tertiarry reference center in Van province, East of Turkey. Patients data was obtained from their charts. All patients were asked about their ethnicity and patients with other ethnicities rather than Kurdish were also excluded from the study. Seven disease manifestations were included to the cluster analysis as follows; mucocutaneus findings (recurrent oral ulcer [ROU], genital ulcer [GU], pseudofolliculitis, erythema nodosum (EN)-like lesions, musculoskeletal (MSK) involvement (arthritis and spondylitis), gastrointestinal (GIS) involvement, vascular involvement (venous and/or artery), parenchymal central nervous system (CNS) involvement.
Results: We included 444 patients (of 59.2 % male) to the study. Mean±standard deviation (SD) of the patients was 35.8±10.2 years and median (Interquartile range [IQR]) disease duration was 12 [12]. Mean±SD age at initial symptom and age of diagnosis were 19.4±7 and 26.4±8.2, respectively. Fulfillment of ISG and ICBD criteria were 91.6 % and 96 %, respectively. Family history of Behçet disease and ROU were 35.4 % and 70.2 %, respectively. In cluster analysis, four clusters were observed as follow; 132 patients (31.2 %) in vascular (C1), 66 patients (15.6 %) in eye (C2), 60 patients (14.2 %) in musculoskelethal (C3) and 165 patients (39 %) in mucocutaneus (C4) clusters. Male gender (OR: 6.5, 95 % Confidence interval [CI]: 2-21.4), superficial thrombophlebitis (OR: 4.7, 95 % CI: 1.9-11.4) and uveitis (OR: 3.6, 95 % CI: 1.3-9.9) were associated with vascular involvement in multivariate analysis.
Conclusion: Among Kurdish patients with BD four disease clusters were observed. Parenchymal lung and CNS involvement were associated with vascular cluster. One third of patients had vascular involvement among Kurdish patients with BD which was quite higher compared to patients with Turkish ethnicity [1].
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[1] Tascilar K et al. Vascular involvement in Behçet's syndrome. PMID: 24907156.

Table 1. Cluster analysis of the study population

Disease manifestations	Vascular	Eye	Musculoskelethal	Mucocutaneus
	C1, n=132 (31.2 %)	C2, n=66 (15.6 %)	C3, n=60 (14.2 %)	C4, n=165 (39 %)
Mucocutaneus	Yes	Yes	Yes	Yes
Musculaskelethal	Yes	No	Yes	No
Gastrointestinal	No	No	No	No
Uveitis	No	Yes	No	No
Thrombophlebitis	Yes	No	No	No
Vascular	Yes	No	No	No
CNS paranchimal	No	No	No	No

Table 2. Univariate analysis of associated factors in disease clusters

Variables	Disease clusters				
	Vascular	Eye	Musculoskeletal	Mucocutaneus	
	C1, n=132 (31.2 %)	C2, n=66 (15.6 %)	C3, n=60 (14.2 %)	C4, n=165 (39 %)	p value (Odds ratio)
Gender, male, n (%)	111 (81)	33 (50)	34 (56.7)	69 (41.8)	***<0.001 (56.5)***
Disease manifestations, n (%)					
Recurrent oral ulcer (ROU)	130 (98.5)	65 (98.5)	59/59 (100)	164 (99.4)	0.7
Genital ulcer	112 (84.8)	46 (69.7)	52 (86.7)	132 (80)	***0.045 (8)***
Thrombophlebitis	79 (59.8)	0	0	0	***<0.001 (214)***
Pseudofolliculitis	114 (86.4)	46 (69.7)	47 (78.3)	123 (74.5)	***0.027 (9.2)***
Erythema nodosum	90 (68.2)	34 (51.5)	29 (48.3)	62 (37.6)	***<0.001 (27.7)***
Pathergy positivity	78 (75)	37 (69.8)	31 (79.5)	74 (58.3)	***0.016 (10.3)***
Arthritis	72 (54.5)	19 (28.8)	58 (96.7)	0	***<0.001 (212)***
Spondylitis	2 (1.5)	1 (1.5)	5 (8.8)	0	***0.001 (17.7)***
Gastrointestinal	6 (4.5)	2 (3)	2 (3.3)	0	0.07
CNS parenchymal	12 (9)	1 (1.5)	1 (1.7)	0	***<0.001 (20.6)***
Lung parenchymal	4 (3.1)	0	0	0	**0.03 (9)**
ISG fulfillment, n (%)	117/124 (94.4)	63/64 (98.4)	49/53 (92.5)	134/156 (86)	***0.009 (11.5)***
ICBD fulfillment, n (%)	130 (98.5)	65 (98.5)	55/57 (96.5)	147/159 (92.5)	***0.042 (8.2)***
HLAB51 positivity, n (%)	8/9 (89)	3/4 (75)	1/3 (33.3)	16/18 (89)	0.1

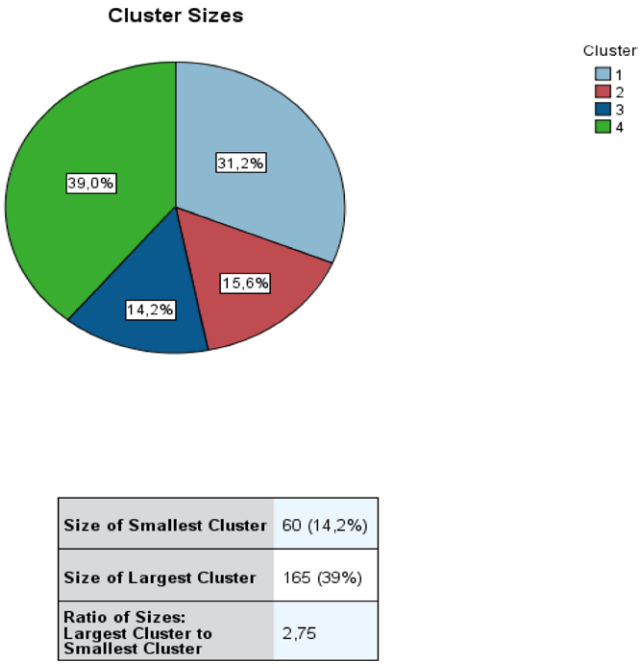


Figure 1.

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AB1512

CHARACTERISTICS OF PATIENTS WHO RECEIVED BIOLOGICAL TREATMENT AFTER KIDNEY TRANSPLANTATION AND THE INCIDENCE OF SERIOUS INFECTIONS: A MULTICENTER STUDY

Keywords: bDMARD, Kidneys

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Background: Kidney transplant patients due to both primary kidney involvement of chronic/autoimmune inflammatory diseases and end-stage kidney disease related to amyloidosis are followed up in rheumatology clinics. Biological agents one of the treatment options in kidney transplant recipients with chronic/autoimmune inflammatory disease.
Objectives: However, there is insufficient data on the development of infection in kidney transplant recipients who received biological treatment. Herein, we aimed to determine the incidence of serious infections in patients with kidney transplant recipients who are received biological therapy.
Methods: Kidney transplant recipients who are received biological agents due to rheumatologic disease were included in the study. Patients' demographic features, transplantation data, biological treatment, development of infection and severity of infection were screened retrospectively. Infections that requiring hospitalization were defined as severe infections.
Results: A total of 31 patients were included in the study, 14 (45%) of whom were female and mean age was 41 ±9 years. Twenty-five patients (80%) of them were non-preemptive kidney transplant and mean duration of hemodialysis before the transplantation was 40 ±40 months. Twenty-three patients (74%) had end stage kidney failure due to FMF-amyloidosis(Figure-1-). Seventeen patients (54%) received anakinra, 11 patients (35%) received canakinumab and 3 patients (10%) received etanercept with other immunosuppressive treatment. Mean treatment duration of biological agents was 4.2±2.6 years. Two patients developed solid organ malignancy and one patient developed hematological malignancy after transplantation. Sixteen of the patients (52%) were hospitalized at least once due to infection and 4 patients (13%) died due to infection. The cause of decease in two patients was COVID-19.
Conclusion: Rheumatic diseases are an important cause of end-stage renal disease and definitive treatment is kidney transplantation. Kidney transplant recipients due to rheumatological disease also use biological agents in the post-transplantation period.

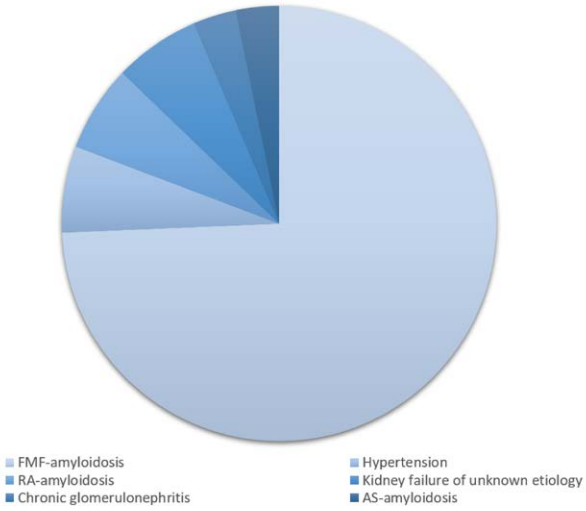
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Kidney transplant recipients have higher risk for the development of infection since they receive immunosuppressive therapy and use of biologic agents may further increase the risk for development infection. Meyer et al reported that infection developed in 54 of 187 solid organ transplant recipients using biological agents.[1] Mean treatment duration of biological agents was 12 months in this study. The incidence of infection was 54% in our study. Mean treatment duration of biological agent was 4.2 year was considered main reason for higher incidence of infection in our study.

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[1] Meyer F, Weil-Verhoeven D, Prati C, Wendling D, Verhoeven F. Safety of biologic treatments in solid organ transplant recipients: A systematic review. Semin Arthritis Rheum. 2021 Dec;51(6):1263-1273. doi: 10.1016/j.semarthrit.2021.08.013. Epub 2021 Aug 26. Erratum in: Semin Arthritis Rheum. 2022 Aug;55:152015. PMID: 34507811.

Figure-1-Causes of end stage kidney failure in this study



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AB1513 MANAGEMENT OF RETROPERITONEAL FIBROSIS PATIENTS: A SCOTTISH COHORT OBSERVATIONAL STUDY

Keywords: Rare/orphan diseases, Descriptive Studies, Outcome measures

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Background: Retroperitoneal fibrosis (RPF) is a rare condition characterised by the inflammation and fibrosis of tissues in the retroperitoneum. Often presenting non-specifically with a variable picture of abdominal pain, constipation, and systemic symptoms of malaise, fever, and weight loss. RPF may also present with severe obstructive uropathy. Initial response to steroids is usually good, but a significant proportion of patients relapse. Evidence for second line treatment is scarce, and a variety of immunosuppressive therapies, such as rituximab, have been used. Surgical intervention and interventional radiology may be useful in treating acute ureteric obstruction and preserving renal function.
Objectives: As an orphan disease without, traditionally, a clear home speciality, retroperitoneal disease is likely underdiagnosed and undertreated especially in those who do not have ureteric obstruction. We characterised patients with retroperitoneal fibrosis across West and South-East Scotland to better understand these patients and their outcomes.
Methods: Patients were previously identified in East Scotland by contacting rheumatologists and by searching the urological stent registry from 2019 to 2022. The method was replicated in the West of Scotland though was less successful as the urological stent register did not record diagnoses, and clinicians did not have an accessible database for this condition. A further avenue of patient identification was then pursued. All available radiological reports and their attached clinical data on the request from 2006 to September 2022 were searched for the term "RPF" and "retroperitoneal fibrosis". Patients were included if they were given a clinical diagnosis of retroperitoneal fibrosis. Patients were excluded if RPF was concurrent with metastatic malignancy, associated with infection or other secondary cause. Patient data was acquired from their electronic records.
Results: 78 patients were identified: 56 in the West of Scotland and 22 in the South-East. Patient demographics are shown in Table 1. The diagnosis was most often made by CT scan. The estimated annual incidence in the last 10 years 2.2 is per million population in the West and 1.6 per million population in the East, for catchment populations of 1.2 million and 0.9 million, respectively. Treatment was heterogenous in both groups. Renal outcomes for these patients were interrogated. Patients were classified as having lost renal function if there was either biochemical or radiological evidence.

Table 1. Summary of findings in the two cohorts across Scotland compared with literature.

	South-East Scotland	West Scotland	Literature
Number	22	56	
Age at diagnosis	60y (range 43-81y)	61.5y (range 45-83y)	54-64y[1]
Female: male ratio	1:2.7	1:1.9	1:1.2-3.4[1]
Lost renal function	73% (16)	75% (42)	
eGFR <60ml/min/1.73m^2	73% (16)	61% (34)	32%[2]
eGFR <30 ml/min/1.73m^2	27% (6)	18% (10)	

Conclusion: The literature suggests renal impairment is variable in retroperitoneal fibrosis with only 32% of patients with abnormal creatinine reported in one study which is much lower than our results. The current literature is sparce for alternative therapies. More work is required in determining efficacy for more recent therapies such as rituximab to further the treatment of this orphan disease and improve patient outcomes.

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Disclosure of Interests: None Declared.
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AB1514 EPIDEMIOLOGY OF BEHÇET'S DISEASE IN NORTHERN SPAIN

Keywords: Epidemiology, Geographical differences, Behcet's disease

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Background: The prevalence of Behçet's disease (BD) has a considerable geographical and temporal variability. Data regarding epidemiology in Spain are limited. (1,2)
Objectives: To assess the epidemiology and clinical domains of BD in a population-based cohort from northern Spain and to compare the results with other geographical areas.
Methods: We conducted a cross-sectional study of a well-defined population in Northern Spain. Cases of suspected BD between January 1980 and December 2018 were identified. The diagnosis of BD was established according to the International Study Group (ISG) for Behçet's Disease. The incidence of BD between 1999 and 2018 was estimated by sex, age, and year of diagnosis.
Results: Of 120 patients with probable BD, 59 patients met ISG criteria and were finally included in the study, with a male/female ratio of 0.97; mean age 49.7±14.7 years. Incidence during the period of study was 0.492 per 100,000 people, observing an increase from January 1999 to December 2018. Prevalence was 10.14 per 100,000 in 2018. Figure 1. Clinical manifestations were relapsing aphthous stomatitis (100%), genital ulcers (78%), skin involvement (84.7%), joint involvement (64.4%), uveitis (55.9%), central nervous system (16.9%), vascular (10.2%), and gastrointestinal manifestations (6.8%). Table 1.
Conclusion: The prevalence of BD in Cantabria is higher than in other Southern-European countries. This difference may reflect a combination of geographic, genetic, or methodological variations, as well as the free accessibility to the Spanish Public Health System. Clinical phenotypes observed are similar to those described in other world regions.

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