82; 45%), anhidrosis/hypohidrosis (44/82; 53.6%), and cornea verticillata (40/62; 64.5%). However, there was a significant delay to diagnosis of up to 51 years (median 18⁹; 27 years). Moreover, diagnosis was established by nationwide screening in dialysis units in 22/82 (26.8%) patients or by family screening in 34/82 (41.4%) patients. At the time of diagnosis, patients usually presented with a clinical picture of systemic disease with mild to moderate proteinuria with or without impairment of kidney function (70/82; 85.4%), left ventricular hypertrophy (56/82; 68.3%), white matter lesions on brain MRI (38/72; 52.8%), and/or a history of stroke (15/82; 18.3%). Twenty six of 82 patients (31.7%) previously had at least one diagnosis of rheumatic disease (Table 1). The common causes for referral to rheumatologist were skin rash and neuropathic pain. In 6 of 26 patients (23.1%), the latter was initially misdiagnosed as joint pain. The possible causes of diagnostic errors included also 'genuine' arthralgia (6/26; 23.1%), episodes of unexplained fever (13/26; 50%), Raynaud phenomenon (2/26; 7.7%), and the laboratory markers of inflammation (11/26; 42.3%).

Abstract SAT0627 – Table 1. 'Rheumatic' diagnoses in Fabry patients

Misdiagnoses	Number of patients
Vasculitis*	6 (7.3%)
Arthritis**	5 (6.1%)
Osler-Weber-Rendu disease	4 (4.9%)
Rheumatic fever	4 (4.9%)
Systemic lupus erythematosus	3 (3.7%)
Familial Mediterranean fever	1 (1.2%)

Note: *Vasculitides included IgA-vasculitis, Behcet disease, etc. **Arthritides included rheumatoid arthritis, juvenile rheumatoid arthritis and osteoarthritis.

Conclusions: Practicing rheumatologists should be aware of FD, given a high occurrence of diagnostic errors. The clues to correct diagnosis include a history of typical symptoms (i.e. neuropathic pain, angiokeratoma, hypohidrosis) from childhood or adolescence and/or the presence of typical manifestations in family members. Notably, FD can initially present as an autoinflammatory disorder with episodes of joint pain and unexplained fever associated with the laboratory markers of inflammation.

Disclosure of Interest: None declared

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SAT0628

A RETROSPECTIVE OBSERVATIONAL STUDY OF AZATHIOPRINE MAINTENANCE THERAPY ON BEHCET'S DISEASE WITH VASCULAR INVOLVEMENT

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Background: Vascular disease which has potential to involve both arteries and veins of all size is one of the major causes of mortality and morbidity in Behçet's disease (BD). There is no controlled studies for the immunosuppressive (IS) therapy in vascular involvement of BD. For the induction and/or maintenance therapy, azathioprine (AZA) is widely used as a corticosteroid tapering IS agent in vascular BD although there are many different clinical presentation.

Objectives: The purpose of this study to investigate relapse rate and the clinical factors affecting it during AZA maintenance therapy.

Methods: Consecutive BD patients with a documentation for arterial or venous involvement were evaluated from two rheumatology centre between January-September 2017. Patients who have been treated for at least 3 months after the complete or partial remission were included to study. The baseline clinical and laboratory findings, treatment protocol, first vascular relapse time and adverse events were obtained medical records. Long-term outcome and factor associated with vascular relapse were assessed.

Results: Totally 78 patients were included to the study and majority of them [59 (%75.6)] was male. The mean age $\pm$ SD of the patients was 37.5 $\pm$ 9 years. Clinical characteristics of patients are seen in table 1. The median duration of maintenance therapy with AZA was 25 (min 3- max 144) months and the mean dose $\pm$ SD of AZA was found as 1.7 $\pm$ 0.3 mg/kg/day. AZA was withdrawn in 4 (% 5.1) patients because of adverse events. Twenty patients (%25.6) were receiving anti-coagulant therapy. Vascular relapse was observed in 14 (%17.9) patients. In relapsing group, arterial involvement and uveitis was higher statistically (p=0.014 and 0.045 respectively). In regression analysis, arterial involvement was independent risk factor for relapse (p=0.016). The percentage of relapses

was calculated as %39 (7/18) in patient subgroup with arterial involvement. Relapse free survival rates according to involving vessel were seen in figure 1.

Abstract SAT0628 – Table 1. Clinical characteristics of vascular Behçet's Disease (n=78)

The mean age $\pm$ SD	37.5 $\pm$ 9.2
Male gender n (%)	59 (76)
Genital ulcer n (%)	49 (63)
Skin lesions n (%)	44 (57)
Patherji positivity n (%)	29 (38)
Uveitis n (%)	17 (22)
Musculoskeletal involvement n (%)	15 (20)
CNS involvement n (%)	1 (1.2)
GIS involvement n (%)	1 (1.2)
Venous involvement n (%)	67 (86)
Arterial involvement n (%)	18 (24)

SD: Standart deviation, CNS: Central nervous system, GIS: Gastrointestinal system

Abstract SAT0628 – Figure 1. Relapse free survival rates according to involving vessels in patients with Behçet's disease (BD) by using Kaplan-Meier method (BDVI: Behçet's disease with venous involvement, BDAl: Behçet's disease with arterial involvement, p=0.001 by log-rank test).

Conclusions: According to our study, AZA is seen as safe in maintenance therapy of BD with vascular involvement, however it can be said that the relapse rate is not favourable especially in patients with arterial involvement. Vascular involvement is considered as related to endotel inflammation resulting BD activity and the effect of anti-coagulant therapy is still controversial. TNF inhibitors or other immunosuppressant may be alternative to AZA for maintenance therapy with the support of randomized-controlled studies.

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SAT0629

IGG4 RELATED CASTLEMAN'S DISEASE OR "SECONDARY" IGG4 RELATED DISEASE OF MULTICENTER CASTLEMAN'S DISEASE?

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Background: It's widely accepted that a diagnosis of IgG4-related disease (IgG4-RD) can be made under a premise of rule out a series of disease including Castleman's disease (CD). Clinically, the two diseases may share similar clinical manifestations, such as multiple organ involvement. Pathologically, in some cases with CD, IgG4 positive plasma cell infiltrations are also evident. Otherwise, IgG4-RD may pathologically exhibits CD-like feature.¹⁻³