Objectives
To shed light on the clinical course and outcome of juvenile Behçet disease.

Methods
A single center retrospective descriptive study of 13 children with Behçet disease. The mean follow-up period was for 3 years.

Results
We had 8 boys and 5 girls. Four of our patients were consanguineous. The mean age was 9 years. The mean interval between onset and diagnosis was 3 months. All of our patients had recurrent aphthous stomatitis with a median of 6 episodes per year. The other symptoms at diagnosis were concomitant genital ulcers in 4 cases, pseudofolliculitis in 7 cases, urticarial rush in 4 cases, severe headache in 2 patients, stroke in one patient, prolonged and/or recurrent fever in 9 patients, uveitis in 4 cases, episcleritis in one case, arthritis in 5 cases, pulmonary hemorrhage in one case, and pyoderma gangrenosum in one case. The pathergy test was performed in all the patients, and was positive in 3 of them. Diagnosis was based on the International Criteria for Behçet's Disease. Acute phase reactants (ESR and CRP) were high in all patients. Meanwhile, granulocytosis was found in 10 patients. HLA 51 was positive in 8 patients, from whom three were siblings. All our patients were on steroids, five had colchicine, seven had azathioprine, two had methotrexate, two had cyclophosphamide, three had etanercept and one had tocilizumab. Complete remission was obtained in 8 cases in a mean time of 15 months, while five children had partial remission, and one boy lost sight of his left eye after a severe retinitis.

Conclusion
Behçet's disease can cause severe sequelae if not diagnosed and treated on time. More studies are needed to establish management guidelines for pediatric population.

Abstract citation ID: keac496.057
61 THE NAILFOLD VIDEOCAPILLAROSCOPY IN PEDIATRIC BEHÇET’S DISEASE: A MULTI-CENTER STUDY

Figen Çakmak1, Şeyda Doğantan2, Rana İşgüder3, Müşerref Kasap Cüceoğlu4, Şengül Çağlayan5, Hafize Emine Sönmez6, Özlem Akgün1, Betül Sözeri7, Ayşenur Paç Kısaarslan7, Erbil Ünsal8, Seza Özen9 and Nuray Aktaş Açay1
1Istanbul University, Istanbul Faculty of Medicine, Pediatric Rheumatology, 2Erciyes University, Faculty of Medicine Pediatric Rheumatology, 3Dokuz Eylül Faculty of Medicine, Pediatric Rheumatology, 4Hacettepe University, Faculty of Medicine, Pediatric Rheumatology, 5Ümraniye Health and Science Hospital, Pediatric Rheumatology, 6Kocaeli University, Istanbul Faculty of Medicine, Pediatric Rheumatology

Background
Behçet’s disease (BD) is a chronic inflammatory disease characterized by recurrent oral aphthous and genital ulcers accompanied by eye, joint, skin, gastrointestinal and central nervous system involvement. The vascular involvement may affect both the arterial and venous systems. Nailfold videocapillaroscopy (NVC) is an easy and non-invasive method used in the evaluation of microcirculation. With this study, we aimed to find the characteristics and prevalence of nailfold capillary alterations in pediatric Behçet’s disease.
patients with juvenile BD and to analyze their possible relationship between clinical characteristics and activity of the disease.

Methods

Patients aged 5–21 years with a diagnosis of juvenile BD and followed up for at least six months were included in the study. Demographic and clinical characteristics of the patients were recorded. NVC was performed on 8 fingers of both hands, excluding the thumbs, and four consecutive non-overlapping fields for each of fingers were evaluated (32 fields per patient). Capillary density, capillary width (arterial width, venous width, apical loop), capillary morphology and the presence of meandering capillary, micro hemorrhage, avascular area, neangiogenesis, capillary ramification were evaluated from the images.

Capillary morphology were evaluated by classifying them into four groups as normal, minor abnormalities, major abnormalities and scleroderma pattern. The presence of abnormalities in at least two fingers were recorded as capillary abnormality. The semi-quantitative rating score 1–3 was applied for each capillaroscopic alteration.

Results

37 patients from 6 pediatric rheumatology centers were included in the study. The mean age of patients was 17 years (IQR 13–19) and 20 (54.1%) of them were girls. The patients were evaluated in four clusters according to their clinical presentations. Nineteen patients had mucocutaneous involvement, 9 patients had uveitis, 8 patients had vascular and neurological involvement, and 4 patients had gastrointestinal system involvement. During the follow-up period, genital ulcers developed in 22 patients, erythema nodosum in 9 patients, pseudofolliculitis in 18 patients, uveitis in 10 patients, vascular involvement in 8 patients, and neurological involvement in 5 patients. Anterior uveitis was present in five, posterior uveitis in three, panuveitis in one, and retinal vasculitis in three of the patients with ocular involvement. Four patients had lower extremity venous thrombosis, three patients had central nervous system (CNS) thrombosis, and one patient had both lower extremity and CNS thrombosis. When capillary morphology was evaluated; normal morphology was present in 16 patients, minor abnormality in 13 patients, and major abnormality in 8 patients. Median capillary density was 8, capillary length was 325 μm, arterial width was 12 μm, venous width was 16 μm, apical loop width was 18 μm, capillary width was 39 μm, and intercapillary distance was 107 μm. Neoangiogenesis was seen in 13 patients, enlarged capillaries in 12 patients, capillary meandering in 9 patients, bushy capillaries in 5 patients, bizarre capillaries in 4 patients, and microhemorrhage in 3 patients. Neoangiogenesis was found to be significantly more common in the NVC evaluation of patients with lower haemoglobin values at the time of diagnosis (p = 0.014).

Conclusion

NVC is an in vivo, non-invasive, and inexpensive imaging technique that allows the direct observation of the capillary network in living tissue throughout the skin and it may be preferred in juvenile BD for evaluating microvascular involvement.