Paediatric rheumatology

Silent but significant: subclinical cardiovascular changes in paediatric Behçet's disease

İ. Bingöl¹, H.E. Sönmez², S. Doğan³, H. Bornaun⁴

¹Department of Paediatrics, University of Health Science, Kanuni Sultan Süleyman Education and Research Hospital, Istanbul; ²Department of Paediatric Rheumatology, Kocaeli University, Kocaeli; ³Department of Paediatric Radiology, University of Health Science, Kanuni Sultan Süleyman Education and Research Hospital, Istanbul; ⁴Department of Paediatric Cardiology, University of Health Science, Kanuni Sultan Süleyman Education and Research Hospital, Istanbul, Turkey.

Abstract Objectives

Behçet's disease (BD) is a systemic, autoinflammatory vasculitis with recurrent mucocutaneous ulcerations, ocular involvement, and multisystemic manifestations, including cardiovascular complications. While extensively studied in adults, paediatric BD, particularly regarding cardiovascular involvement, remains underexplored. This study assesses cardiovascular manifestations in paediatric BD through echocardiographic, vascular, and microvascular evaluations.

Methods

A total of 58 subjects were enrolled, including 29 paediatric BD patients and 29 healthy children. Exclusion criteria included comorbidities or prior cardiovascular diseases. Capillaroscopy assessed nailfold microvascular changes, and carotid artery intima-media thickness (CIMT) was measured via ultrasonography. Transthoracic echocardiography evaluated cardiac parameters, including ventricular function and indices of aortic stiffness and pulmonary vascular resistance.

Results

The mean age of BD patients was 15.8 years, with disease onset at 12.4 years. Elevated left ventricular myocardial performance index (LV MPI), reduced right ventricular systolic function, and increased pulmonary arterial pressure. Aortic stiffness was also significantly higher, indicating early vascular remodelling. CIMT showed no significant differences. Mild capillary tortuosity was observed in some BD patients.

Conclusion

The findings suggest early cardiovascular changes in paediatric BD, including diastolic dysfunction, increased pulmonary arterial pressure, and impaired aortic elasticity. These results highlight the need for routine cardiovascular screening, even without overt cardiac symptoms, to mitigate long-term cardiovascular risks. Further studies are needed to understand these abnormalities' progression and optimise interventions.

Key words

Behçet's disease, cardiovascular involvement, CIMT

İlyas Bingöl, MD Hafize Emine Sönmez, MD Sümeyra Doğan, MD Helen Bornaun, MD Please address correspondence to: İlyas Bingöl, Department of Paediatrics, Faculty of Health Sciences, Kanuni Sultan Süleyman Education and Research Hospital. 34303 Istanbul, Turkey. E-mail: drilyasbingol@hotmail.com Received on April 13, 2025; accepted in revised form on September 11, 2025. © Copyright CLINICAL AND EXPERIMENTAL RHEUMATOLOGY 2025.

Introduction

Behçet's disease (BD) is classified as a primary vasculitis of variable vessels, encompassing both arterial and venous involvement of varying calibres. However, the hallmark clinical features of BD were initially recognised as recurrent oral ulcers, cutaneous abnormalities, and uveitis. Over time BD has been recognised as a disease with a broad and diverse spectrum of clinical manifestations, affecting multiple organ systems and presenting with varying degrees of severity (1, 2). The prevalence of BD has been reported as 10.3 per 100,000 population (2). However, there is no definitive data on the prevalence of paediatric-onset BD. Studies suggest that 4-26% of BD patients experience the onset of the disease in childhood (3). Paediatric and adult BD exhibit distinct differences not only in prevalence but also in clinical presentations and disease courses. For instance, cardiac involvement in paediatric patients is less prevalent than in adults, with its occurrence ranging from 5% to 20%. Major cardiovascular manifestations of BD include pericarditis, myocarditis, endocarditis, endomyocardial fibrosis, valve lesions, intracardiac and venous thrombosis, coronary vasculitis, ventricular arrhythmias, and dilated cardiomyopathy. Intracardiac thrombosis is frequently associated with pulmonary involvement and often correlates with right ventricular dysfunction (4-6). While cardiac manifestations may often be asymptomatic, when present, they represent a significant contributor to both morbidity and mortality.

This study aimed to evaluate the clinical and laboratory features of paediatric patients with BD referred to the paediatric cardiology outpatient clinic and to determine the presence of cardiovascular abnormalities associated with the disease. To comprehensively assess both macrovascular and microvascular involvement in paediatric BD, a combination of diagnostic tools was used. Echocardiography was employed to evaluate cardiac structure and function, with a focus on diastolic function and aortic elasticity. Carotid intimamedia thickness (CIMT) measurement served as a non-invasive marker of early atherosclerotic changes. Capillaroscopy was utilised to investigate microvascular involvement.

Materials and methods

Study population

This study was conducted between February 2020 and September 2022 at the Department of Paediatrics, Ministry of Health, University of Health Sciences Kanuni Sultan Süleyman Training and Research Hospital. Patients diagnosed with paediatric BD based on classification criteria for paediatric BD and without any comorbidities were included (4). The study participants ranged in age from 8 to 18 years.

Patients diagnosed with BD who were over 18 years of age were excluded from the study. A total of 29 paediatric patients diagnosed with BD, without any cardiological or comorbid conditions, were included in the study. For the control group, 29 healthy children who visited the Paediatric Cardiology outpatient clinic of our hospital for pre-sports and general medical examinations, and who had no pathological findings based on examinations and imaging studies, were selected. The control group was matched to patients on a 1:1 basis by age (±1 year) and sex to minimise confounding due to developmental cardiovascular changes.

Capillaroscopy

The nail bed of each case was assessed using a 'Dino-lite' brand capilloscope at a magnification of x200, following a 15-20-minute rest period for the 2nd-5th fingers of both hands at room temperature, after the application of cedar oil in a seated position. A paediatric rheumatologist then performed capillaroscopy to examine the nailfold capillaries of both hands, recording normal and abnormal images. The evaluation focused on assessing capillary morphology, including parameters such as capillary tortuosity, capillary crossing, dilated capillaries, giant capillaries, and branched capillaries. A capillary count of fewer than 9 per 1mm in each nail bed was recorded as capillary loss. Focal punctate haemorrhagic foci within the capillary bed were noted as microhaemorrhages. Homogeneously

Competing interests: none declared.

dilated or locally expanded capillaries were classified as capillary dilatation. Capillaries exceeding 50 micrometres in width were categorized as giant capillaries. Curved, bush-like, branched, spiral, and elongated capillaries were interpreted as indicative of neoangiogenesis (7).

Measurement of carotid artery intima media

Measurements of CIMT were performed and recorded by the same paediatric cardiologist using a Toshiba Aplio 300 ultrasonography device with an 11L+ probe. Measurements were taken while the patients and control group were in a supine position with the neck gently hyperextended. Three manual measurements were made on each carotid artery, 1-2 cm proximal to the bifurcation, and the average of these values was calculated for each case (8).

Cardiac evaluation

All patients and controls underwent a thorough transthoracic echocardiographic examination using a GE Vivid 7 Pro ultrasound system with a 3 MHz probe, following established guidelines for cardiovascular imaging. For each parameter, three consecutive measurements were taken, and the results were averaged.

Two-dimensional, M-mode, pulsed wave (PW) Doppler, and tissue Doppler echocardiographic images from both the patient and control groups were analysed. Interventricular septum thickness (IVSD), left ventricular posterior wall thickness (LVPWD), ejection fraction (EF) and aortic root width (Ao) were evaluated in both the patient and control groups. Left atrial width (LA), LA/Ao ratio, end-systolic aortic diameter (As), end-diastolic aortic diameter (Ad), aortic stiffness ratio, stiffness index, distensibility, aortic elastic modulus (Ep), right ventricular tricuspid annular plane systolic excursion (RV TAPSE), left ventricular mitral annular plane systolic excursion (LV MAPSE), and pulmonary vascular resistance (PVR) values were also measured. To evaluate ventricular diastolic dysfunction, Doppler echo-

Table I. Comparison of left ventricular evaluations with conventional PW Doppler and tissue Doppler echocardiography in the patient and control groups.

9 (8-9) 45.2 ± 4.5	9 (7.5-9)	0.823
45.2 ± 4.5	` ,	0.823
		0.020
7 (5 5 0)	44.6 ± 5.2	0.609
/ (3.3-9)	6 (6-7)	0.124
25 (23-28)	25 (23-27)	0.888
30 (28-32)	30 (28-31.5)	0.622
1.19 ± 0.15	1.23 ± 0.16	0.366
67.7 ± 5.1	70.4 ± 5.9	0.067
1,40 (1,25-1,45)	1,40 (1,35-1,50)	0.222
0.99 ± 0.18	0.97 ± 0.12	0.681
0.64 ± 0.11	0.58 ± 0.08	0.023
1.60 (1.39-1.74)	1.64 (1.55-1.78)	0.189
0.17 ± 0.03	0.17 ± 0.02	0.359
0.09 ± 0.02	0.08 ± 0.02	0.279
1.99 ± 0.34	2.14 ± 0.29	0.076
52 (48-52)	48 (44-52)	0.181
60.2 ± 6.1	54.7 ± 7.1	0.002
288 (266-297.5)	288 (275.5-305)	0.314
0.39 ± 0.05	0.36 ± 0.05	0.007
	30 $(28-32)$ 1.19 ± 0.15 67.7 ± 5.1 1,40 (1,25-1,45) 0.99 ± 0.18 0.64 ± 0.11 1.60 (1.39-1.74) 0.17 ± 0.03 0.09 ± 0.02 1.99 ± 0.34 52 (48-52) 60.2 ± 6.1 288 (266-297.5)	$\begin{array}{cccccccccccccccccccccccccccccccccccc$

PW-Doppler: pulsed-wave Dopple; IVSd: interventricular septal thickness in diastole; LVDD: left ventricular diastolic diameter; LPWD: left posterior wall diameter; Ao: aortic root diameter; LA: left atrium diameter; MAPSE: mitral annular plane systolic excursion; EF: ejection fraction; IVCT: isovolumetric contraction time; IVRT: isovolumetric relaxation time; ET: ejection time; MPI: myocardial performance index (Tei index).

*Mean ± standard deviation and others are shown as median (25-75th quartile).

cardiographic measurements of mitral and tricuspid inflow velocities are commonly utilized. These include the early diastolic peak velocity (E wave), the late diastolic peak velocity associated with atrial contraction (A wave), and the E/A ratio for both ventricles. Tissue Doppler imaging (TDI) was also employed to assess myocardial velocities during early and late diastole (denoted as E' and A', respectively) at the lateral annulus of the mitral and tricuspid valves. In normal diastolic function, the E wave typically exceeds the A wave; however, in the presence of impaired relaxation, the E/A ratio decreases due to the elevated contribution of atrial filling. Isovolumetric relaxation time (IVRT), isovolumetric contraction time (IVCT), and myocardial performance index (MPI) were also recorded. An MPI value of 0.45 was considered within the normal range (9, 10). For assessment of aortic elasticity, the ascending aorta was visualized from the parasternal long-axis window, approximately 3 cm above the aortic valve. Using M-mode echocardiography, systolic aortic diameter was recorded at the peak anterior motion of the aortic wall (systolic phase), while diastolic diameter was measured at the Q wave on the ECG (end-diastolic phase). The mean values from three consecutive cardiac cycles were used for analysis. Four parameters reflecting aortic stiffness were computed:

- Aortic strain = (systolic diameter diastolic diameter) / diastolic diameter.
- Aortic distensibility = aortic strain / brachial pulse pressure.
- Stiffness index = ln (systolic BP / diastolic BP) / aortic strain.
- Elastic modulus = (systolic BP diastolic BP) / aortic strain.

Statistical analysis

Results were analysed using Microsoft Excel and SPSS V.21 programs. The normality of continuous data was assessed using histograms, Q-Q plots, and the Kolmogorov-Smirnov and Shapiro-Wilk tests. The homogeneity of the data was analysed with Levene's test. Continuous data are expressed

as mean ± standard deviation (SD) if normally distributed, and as median (25th-75th percentile) if not normally distributed. Categorical data are expressed as frequency and percentage. The Chi-square test was used to compare categorical data. For comparisons of continuous data between two groups, the independent t-test was used if the groups were normally distributed, and the Mann-Whitney U test was used if the groups were not normally distributed. All tests were two-tailed, and *p*<0.05 was considered statistically significant.

Results

A total of 58 individuals participated in the study, including 29 patients with BD and 29 healthy children. The mean age of the patients was 15.8±2.8 years, while the mean age at diagnosis of BD was 12.4±3 years. Among the patients with BD, 41.4% (n=12) were female, and 58.6% (n=17) were male. A family history of BD was identified in 34.5% (n=10) of the patients for first-degree relatives and in 17.2% (n=5) for second-degree relatives.

Oral aphthae were present in 96.6% (n=28) of the patients, genital aphthae in 44.8% (n=13), and erythema nodosum in 20.7% (n=6). Papulopustular skin lesions were observed in 37.9% (n=11) of the patients. The pathergy test results were positive in 44.8% (n=13) and negative in 55.2% (n=16) of the patients. Arthritis was observed in 13.8% (n=4) of the patients, while arthralgia was present in 51.7% (n=15). Ocular findings were noted in 10.3% (n=3) of the patients. Thrombosis was observed in 10.3% (n=3) of the patients, including deep vein thrombosis (DVT) in one patient, sinus vein thrombosis in one, and left arm thrombosis in one. None of the patients exhibited pulmonary or gastrointestinal involvement, while 3.4% (n=1) had neurological findings. Among the patients, 65.5% (n=19) were on medication: colchicine was used by 34.5% (n=10), colchicine and azathioprine by 27.6% (n=8), and methotrexate by 3.4% (n=1).

The mean pulse rate was 81.8±14.6 in the patient group and 74.8±10.3 in the control group. The pulse rate in

Table II. Comparison of right ventricular evaluations with conventional PW Doppler and tissue Doppler echocardiography in the patient and control groups.

Parameters	Patient group (n=29)	Control group (n=29)	<i>p</i> -value
Right ventricular systolic function			
RVOT VTI (cm)	22.1 (18.8-24.6)	19.5 (18-20.9)	0.016
RV TAPSE (cm)	2.2 (1.9-2.3)	2.3 (2.2-2.4)	0.005
Pulmonary pressure and resistance	` '	` '	
P max (mmHg)	4.6 (3.9-5.7)	3.5 (2.8-4.6)	0.004
PVR (WOOD Unit)	1.19 (1.11-1.41)	1.22 (1.02-1.35)	0.316
TRV (m/s)	2.26 (2.15-2.39)	2.01 (1.75-2.16)	< 0.001
Tricuspid inflow (PW-Doppler)			
Tricuspid E (m/s)	0.69 ± 0.12	0.73 ± 0.13	0.289
Tricuspid A (m/s)	0.54 ± 0.09	0.54 ± 0.09	0.934
Tricuspid E/A	1.28 ± 0.13	1.34 ± 0.17	0.104
Tricuspid annular velocities (Tissue	Doppler)		
Tricuspid E' (m/s)*	0.15 ± 0.03	0.15 ± 0.02	0.733
Tricuspid A' (m/s)	0.13 (0.1-0.16)	0.11 (0.1-0.14)	0.112
Tricuspid E'/A' *	1.23 ± 0.19	1.33 ± 0.25	0.090
Time intervals			
Tricuspid IVCT (m/s)	55 (52-63)	59 (52-63)	0.627
Tricuspid ET (ms)*	268.1 ± 19.4	277.4 ± 30.6	0.173
Tricuspid IVRT (ms)	63 (57-70)	52 (48-63)	< 0.001
Right ventricular index			
TA*	387.9 ± 21.8	389.2 ± 34	0.862
RV MPI	0.44 (0.41-0.50)	0.40 (0.36-0.43)	0.008

PW-Doppler: pulsed-wave Doppler; RVOT VTI: right ventricular outflow tract velocity-time integral; TAPSE: tricuspid annular plane systolic excursion; Pmax: maximum pulmonary pressure; PVR: pulmonary vascular resistance; TRV: tricuspid regurgitation velocity; IVCT: isovolumetric contraction time; IVRT: isovolumetric relaxation time; ET: ejection time; TA: tricuspid acceleration time; MPI: myocardial performance index (Tei index).

*Mean ±s tandard deviation and others are shown as median (25-75th quartile).

the patient group was significantly higher than that in the control group (p=0.039). The median systolic blood pressure in the patient group was 115 (110-120), and the median diastolic blood pressure was 70 (70-80), whereas the control group had a systolic blood pressure of 100 (100-110) and a diastolic of 70 (70-75). While there was no significant difference between the diastolic blood pressures of the patient and control groups (p=0.306), the systolic blood pressure in the patient group was significantly higher than in the control group (p=0.002), although it remained within normal limits. When comparing the electrocardiogram (ECG) measurements between the patient and control groups, no significant differences were found in PR interval, QRS duration, and QTc interval.

Nailfold capillaroscopy examination of the patients revealed a mild tortuous appearance in 3 patients and mild dilation in one patient.

The comparison of two-dimensional, M-mode, pulsed-wave (PW) Doppler, and tissue Doppler echocardiographic images between the patient and control groups is presented in Tables I and

II. When comparing the left ventricular tissue Doppler values between the patient and control groups, no significant differences were observed in TD Mitral E', TD Mitral A', TD Mitral E'/A', Mitral IVCT, Mitral ET, MYS, or Total Time values. However, the Mitral IVRT and LV MPI values in the patient group were significantly higher than those in the control group. Additionally, the RVOT VTI, P max, and TRV values in the patient group were higher than in the control group, while the RV TAPSE value was statistically significantly lower. The tricuspid IVRT and RV MPI values in the patient group were also higher than those in the control group.

When comparing aortic and carotid artery measurements between the patient and control groups, no significant difference was found between the right and left CIMT (p=0.554, p=0.876, respectively). The aortic systolic and diastolic diameters in the patient group were significantly higher than those in the control group. The Aortic Artery Stiffness Ratio and Aortic Strain values were significantly lower in the patient group compared to the control group.

Table III. Comparison of aortic and carotid artery measurements between patient and control groups.

Parameters	Patient group (n=29)	Control group (n=29)	<i>p</i> -value
Right CIMT (mm)	0.33 (0.30-0.39)	0.35 (0.29-0.41)	0.554
Left CIMT (mm)	0.35 (0.30-0.40)	0.36 (0.31-0.40)	0.876
Systolic diameter of aorta (cm)	2.5 (2.45-2.7)	2.4 (2.3-2.6)	0.017
Diastolic diameter of aorta (cm)	2.3 (2.3-2.4)	2.1 (2-2.3)	0.007
Aortic Artery Stiffness Ratio*	1.12 ± 0.03	1.13 ± 0.03	0.025
Aortic Strain (%)	0.13 (0.09-0.014)	0.14 (0.11-0.15)	0.018
Aortic Stiffness index	4.0 (3.2-4.6)	2.9 (2.4-3.8)	0.005
Aortic Distensibility (10 ⁻⁶ cm ² /dyne)	0.006 (0.005-0.007)	0.008 (0.006-0.010)	0.003
EP (Pressure Strain Elastic Modulus)*	364.3 ± 121.3	273.2 ± 91.8	0.002

CIMT: carotid intima-media thickness.

*Mean ± standard deviation and others are shown as median (25-75th quartile).

The Aortic Stiffness Index was significantly higher in the patient group than in the control group. A comparison of aortic and carotid artery measurements between the patient and control groups is presented in Table III.

Discussion

Data on BD in childhood remains limited, and there is a lack of comprehensive studies specifically focusing on the cardiovascular evaluation of paediatric patients with BD. In the present study, the cardiovascular assessment of a paediatric patient with BD demonstrated diastolic dysfunction in both the left and right sides of the heart, suggesting an increased risk for the development of atherosclerosis.

In a retrospective study conducted by Atmaca et al. (11) in 2011, which evaluated 3,382 patients diagnosed with BD, the prevalence of paediatric patients was determined to be 3.3% (n=110). Among these paediatric cases, 62.7% were female, and the mean age at diagnosis was 14.1±2.1 years. The study also revealed that 100% of the paediatric patients presented with oral ulcers, while 82.7% exhibited genital ulcers. In an international prospective cohort study conducted by Koné-Paut et al. (4) in 2016, involving 156 patients, the male to female ratio was found to be 1:1. The mean age at diagnosis was 13.87±3.82 years. In the present study, the mean age at diagnosis of BD was 12.4±3 years, with a male to female ratio of 1.4:1, and, consistent with other studies, recurrent oral aphthae and genital lesions were found to be the most frequent manifestations.

In the meta-analysis conducted by Aslam et al. (12), a decrease in the E/A ratio and an increase in IVRT were observed in adult BD patients. In contrast, the study by Yağmur et al. (13) found no statistically significant differences in the A wave, E/A ratio, or IVRT when comparing adult BD patients to healthy controls. The elevated A wave suggests that increased atrial contraction is required to facilitate complete ventricular filling due to impaired ventricular relaxation during the diastolic phase. In the presence of clinically significant diastolic dysfunction, the E wave, which reflects ventricular relaxation, diminishes, while the A wave increases. Consequently, the E/A ratio, a marker of diastolic dysfunction, decreases below the normal threshold of 1. In the present study, conventional Doppler and tissue Doppler examination of the left ventricle revealed that the mitral A wave was slightly higher in the patient group compared to the control group (p=0.023). Furthermore, IVRT, a key indicator of diastolic dysfunction, was found to be significantly higher in the patient group (p=0.002). Tavil et al. (14) found that although systolic and diastolic function parameters were comparable between BD patients and controls, tissue Doppler-derived myocardial left ventricular relaxation time and MPI were significantly impaired in the patient group. Correspondingly, in present study, the MPI was also significantly elevated in the patient group (p=0.007). The increase in the MPI value suggests that both diastolic and systolic dysfunction should be considered. Several cases of pulmonary artery aneurysm have been reported in adult BD patients, along with right ventricular involvement due to myocardial and vascular changes, like the involvement seen in the left ventricle. However, no echocardiographic studies have specifically investigated the right ventricular involvement in paediatric BD cases. In the present study, values for IVRT and right ventricular MPI, analysed using tissue Doppler in right ventricular examination results, were found to be statistically significantly higher in the patient group (p=0.001-0.008). Additionally, TAPSE values were significantly elevated in this group (p=0.005). Indicators of pulmonary artery pressure, namely P max and TRV, were also found to be elevated in the patient group (p=0.004, p=0.001).

An increase in the aortic stiffness index, along with an increase in the aortic elastic modulus (Ep) and a decrease in aortic distensibility and strain, are significant markers for atherosclerosis, particularly in the pre-atherosclerotic phase, prior to the onset of clinical symptoms. These parameters may be valuable for early detection of the condition. Aortic Ep, a reduction in aortic strain and distensibility, and an increase in elastic modulus are indicative of heightened aortic stiffness. Furthermore, an increase in both aortic systolic and end-diastolic diameters is commonly interpreted as supportive of atherosclerosis. Notably, an increase in aortic stiffness is one of the earliest changes observed in atherosclerosis and may serve as an early warning sign for subclinical atherosclerosis. Several studies have reported a decrease in aortic distensibility and aortic strain in adult populations. For instance, in a study by Aslam et al. (12), aortic strain and aortic distensibility were found to be statistically significantly lower in adult BD patients compared to the control group (p<0.001 for both parameters). Similarly, a meta-analysis conducted by Upala et al. (15), which included 303 adult BD patients, found that the risk of atherosclerosis was higher in the patient group, as evidenced by carotid-femoral pulse wave velocity (PWV) measurements. A paediatric study conducted by Demir et al.

(16) showed abnormal ABPM (Ambulatory Blood Pressure Monitoring) in 34.4% of paediatric BD patients. Furthermore, BD patients with vascular involvement exhibited significantly higher velocity and velocity-time integral of the left ventricular outflow tract, which may indicate increased aortic stiffness. In the present study, the systolic and diastolic diameters of the aorta were significantly larger in the patient group compared to controls (p=0.017 and p=0.007, respectively).In addition, the patient group demonstrated higher aortic stiffness index (p=0.005) and pressure strain elastic modulus (p=0.002), along with lower aortic strain (p=0.018) and aortic distensibility (p=0.003). The aortic stiffness ratio was also slightly lower in patients (p=0.025). Previous studies have not shown an increase in CIMT (16, 17); similarly, no difference was found in our study. While CIMT is a common surrogate for atherosclerosis, the paediatric BD population may demonstrate vascular changes primarily at the functional (elasticity/microvascular) rather than structural level, especially in early disease. Consistent with previous studies in childhood rheumatic diseases, aortic stiffness has been recognised as a reliable marker for the detection of early atherosclerosis (18, 19).

This study has several limitations. First, it was conducted at a single centre, which may limit the generalisability of the findings. Second, due to the rarity of the disease in childhood, the sample size was relatively small, potentially reducing the statistical power of our analyses. Additionally, pulse wave velocity, an important parameter for vascular assessment, could not be evaluated. We acknowledge this as a limitation and recommend that future prospective studies incorporate this measurement to provide a more comprehensive evaluation. Finally, in the absence of standardized Z-scores for all parameters, certain measurements could not be fully adjusted for paediatric reference ranges, which may limit the precision of interpretation.

In conclusion, echocardiographic examination of the right and left ventricles revealed early diastolic dysfunction, suggesting the need for careful followup of paediatric patients with BD. Additionally, aortic stiffness ratio, aortic strain and aortic distensibility, which indicate the ability of arteries to relax, were significantly lower in the patient group. In contrast, aortic end-systolic and end-diastolic diameters, Aortic Stiffness Index, and EP, which reflect arterial stiffness, were significantly higher in the patient group. These findings suggest that paediatric patients with BD may be at increased risk for atherosclerosis, highlighting the importance of regular monitoring in these patients.

References

- KARINCAOGLU Y, BORLU M, TOKER SC et al.: Demographic and clinical properties of juvenile-onset Behçet's disease: A controlled multicenter study. J Am Acad Dermatol 2008; 58(4): 579-84.
- https://doi.org/10.1016/j.jaad.2007.10.452
 2. KONE-PAUT I, YURDAKUL S, BAHABRI SA, SHAFAE N, OZEN S, OZDOGAN H, BERNARD JL: Clinical features of Behçet's disease in children: an international collaborative study of 86 cases. *J Pediatr* 1998; 132(4): 721-5. https://doi.org/10.1016/s0022-3476(98)70368-3
- YILDIZ M, KOKER O, ADROVIC A, SAHIN S, BARUT K, KASAPCOPUR O: Pediatric Behçet's disease - clinical aspects and current concepts. Eur J Rheumatol 2020; 7: 38-47. https://
- doi.org/10.1016/s0022-3476(98)70368-3
 4. KONE-PAUT I, SHAHRAM F, DARCE-BELLO M *et al.*: Consensus classification criteria for paediatric Behçet's disease from a prospective observational cohort: PEDBD. *Ann Rheum Dis* 2016; 75(6): 958-64. https://doi.org/10.1136/annrheumdis-2015-208491
- DEMIRELLI S, DEGIRMENCI H, INCI S, ARI-SOY A: Cardiac manifestations in Behçet's disease. *Intractable Rare Dis Res* 2015; 4(2): 70-5.
 - https://doi.org/ 10.5582/irdr.2015.01007
- 6. MENDES LA, MAGRAW LL, ALDEA GS, DA-VIDOFF R: Right ventricular thrombus: an unusual manifestation of Behçet's disease. *J Am Soc Echocardiogr* 1994; 7(4): 438-40. https://
 - doi.org/10.1016/s0894-7317(14)80208-3
- CUTOLO M, HERRICK AL, DISTLER O et al.: Nailfold Videocapillaroscopic Features and Other Clinical Risk Factors for Digital Ulcers in Systemic Sclerosis: A Multicenter, Prospective Cohort Study. Arthritis Rheumatol 2016; 68(10): 2527-39.
 - https://doi.org/10.1002/art.39718
- 8. STEIN JH, KORCARZ CE, HURST RT et al.: Use of carotid ultrasound to identify subclinical vascular disease and evaluate cardiovascular disease risk: a consensus statement from the American Society of Echocardiography Carotid Intima-Media Thickness Task Force. Endorsed by the Society for Vascular

- Medicine. J Am Soc Echocardiogr 2008; 21(2): 93-111.
- https://doi.org/10.1016/j.echo.2007.11.011
- 9. QUINONES MA, OTTO CM, STODDARD M, WAGGONER A, ZOGHBI WA: Doppler Quantification Task Force of the N, Standards Committee of the American Society of E. Recommendations for quantification of Doppler echocardiography: a report from the Doppler Quantification Task Force of the Nomenclature and Standards Committee of the American Society of Echocardiography. *J Am Soc Echocardiogr* 2002; 15(2): 167-84. https://doi.org/10.1067/mje.2002.120202
- 10. LANG RM, BADANO LP, MOR-AVI V et al.: Recommendations for cardiac chamber quantification by echocardiography in adults: an update from the American Society of Echocardiography and the European Association of Cardiovascular Imaging. J Am Soc Echocardiogr 2015; 28(1): 1-39.
- https://doi.org/ 10.1016/j.echo.2014.10.003 11. ATMACA L, BOYVAT A, YALCINDAG FN, ATMACA-SONMEZ P, GURLER A: Behçet disease in children. *Ocul Immunol Inflamm* 2011; 19(2): 103-7. https://doi.org/ 10.3109/09273948.2011.555592
- ASLAM F, BANDEALI SJ, CROWSON C, ALAM M: Cardiac Function and Diastolic Dysfunction in Behçet's disease: A Systematic Review and Meta-Analysis. *Int J Rheumatol* 2016; 2016: 9837184. https://doi.org/10.1155/2016/9837184
- 13. YAGMUR J, SENER S, ACIKGOZ NY et al.: Subclinical left ventricular dysfunction in Behçet's disease assessed by two-dimensional speckle tracking echocardiography. Eur J Echocardiogr 2011;12(7):536-41. https://doi.org/ 10.1093/ejechocard/jer088
- 14. TAVIL Y, OZTURK MA, SEN N et al.: The assessment of cardiac functions by tissue Doppler-derived myocardial performance index in patients with Behçet's disease. Clin Rheumatol 2008; 27(3): 309-14. https://doi.org/10.1007/s10067-007-0707-x
- 15. UPALA S, YONG WC, SANGUANKEO A: Increased Arterial Stiffness in Behçet's disease: a Systematic Review and Meta-Analysis. *Korean Circ J* 2017; 47(4): 477-82. https://doi.org/10.4070/kcj.2017.0004
- 16. DEMIR S, DUZOVA A, KARAGOZ T et al.: The risk of cardiovascular comorbidity in children with Behçet's disease. Rheumatology (Oxford) 2024; 63(SI2): SI188-SI94. https:// doi.org/10.1093/rheumatology/kead505
- 17. AKGUN G, SOZERI B, BASAR EZ et al.: Evaluation of epicardial adipose tissue thickness and carotid intima-media thickness in children with Behçet's disease. Clin Exp Rheumatol 2024; 42(10): 2086-91. https://doi.org/10.55563/clinexprheumatol/ghk2ya
- DEDEOGLU R, YILDIZ M, KARAGOZLU F et al.: Unexpected increase of aortic stiffness in juvenile Spondyloarthropathies. Cardiol Young 2020; 30(12): 1806-14. https://doi.org/10.1017/S1047951120002796
- DEDEOGLU R, ULUG MURT N, GUNALP A et al.: Exploring cardiovascular implications of juvenile dermatomyositis: Insights from aortic stiffness analysis and 3D echocardiography. Echocardiography 2024; 41(2): e15776. https://doi.org/10.1111/echo.15776