

Clinical and diagnostic characteristics of arterial involvement in paediatric Behçet's disease

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Abstract

Objective

To explore the clinical features, diagnosis, treatment and prognosis of arterial involvement in paediatric Behçet's disease (BD) for clinical reference.

Methods

A retrospective cohort study analysed 76 paediatric BD patients (January 2013 to May 2024). Nineteen with arterial involvement were the experimental group, and 57 without vascular involvement were the control group.

Results

The experimental group mainly involved medium-sized (15/19, 78.95%), large-sized (13/19, 68.42%) or both (9/19, 47.37%) arteries, most commonly abdominal aorta (8/19, 42.11%), pulmonary artery (7/19, 36.84%) and femoral artery (7/19, 36.84%). Notably, we found a high prevalence of coronary artery involvement, exclusively manifesting as left main coronary artery dilation. Lesions were mainly wall thickening (9/19, 47.37%), lumen dilation (8/19, 42.11%) and stenosis (7/19, 36.84%). Compared with the control group, it had later onset age (11.0 vs. 7.0 years), shorter disease duration (6.0 vs. 24.0 months), and higher incidences of fever, multi-organ involvement (neurological, renal, cardiac), and elevated inflammatory markers including C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR). It also had more intensive treatment and surgery but had significantly higher rates of cerebrovascular accidents (15.79% vs. 0%), cardiac complications (31.58% vs. 0%), and mortality (10.53% vs. 0%).

Conclusion

Paediatric BD with arterial involvement is a severe phenotype with a poor prognosis, characterised by intense inflammation and multi-organ damage. This underscores the critical need for early identification, aggressive treatment, and close monitoring to improve long-term outcomes.

Key words

arterial involvement, Behçet's disease, paediatric, vasculitis

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Introduction

Behçet's disease (BD) is a chronic and multisystemic vasculitis of unknown aetiology (1) that can involve multiple organ systems throughout the body. Typical clinical manifestations include recurrent oral and genital aphthous ulcers, skin lesions and ocular uveitis (2). The peak incidence occurs between 25–30 years of age, whereas paediatric BD is relatively rare, with only 4–26% of patients experiencing onset before the age of 16 (3, 4). In recent years, paediatric BD has garnered increasing attention. Its clinical manifestations, distribution of involved organs, and disease severity differ from those in adults.

Vascular involvement is one of the most severe and life-threatening complications of BD, occurring in approximately 15–50% of patients and serving as a major predictor of morbidity and mortality (5, 6). Vascular lesions are diverse, predominantly affecting the venous system, with deep vein thrombosis (especially in the lower extremities) and superficial thrombophlebitis being the most common. Arterial involvement is less frequent (approximately 3–5%) but can manifest as aneurysms (*e.g.* pulmonary artery, aortic aneurysms) or arterial occlusion, presenting a more critical condition (7). Current research on arterial involvement in paediatric BD is limited, largely consisting of case reports or small series, and lacks comprehensive data on its clinical spectrum, diagnostic challenges, and outcomes compared to paediatric BD without vascular involvement. Therefore, this study aims to: (1) describe the clinical and imaging characteristics of arterial involvement in a cohort of paediatric BD patients; (2) compare these features with paediatric BD patients without vascular involvement to identify distinguishing factors; and (3) analyse treatment strategies and prognosis associated with arterial involvement, thereby providing evidence to guide early recognition and management.

Data and methods

Study design and participants

This was a single-centre, retrospective cohort study. We reviewed the medi-

cal records of all paediatric patients diagnosed with BD who were admitted to the Department of Rheumatology and Immunology, Capital Centre for Children's Health, Capital Medical University, between January 2013 and May 2024. The study protocol was approved by the Ethics Committee of Capital Centre for Children's Hospital, Capital Medical University (SHER-LLM2023039), which waived the requirement for informed consent due to the retrospective nature of the study, and in accordance with the Helsinki Declaration of 1975/83.

Inclusion and exclusion criteria

Inclusion criteria were: 1) Age below 18 years at diagnosis; 2) Diagnosis meeting the 2015 Paediatric Behçet's Disease (PEDBD) classification consensus (8) or the 2014 International Criteria for Behçet's Disease (ICBD) (9, 23). For the experimental group, an additional criterion was the presence of arteritis confirmed by ultrasonography, computed tomography (CT), or magnetic resonance imaging (MRI).

Exclusion criteria were: 1) Arteritis caused by other systemic diseases (*e.g.* Takayasu's arteritis, Systemic lupus erythematosus); 2) Vascular lesions due to trauma or infection; 3) Congenital vascular malformations.

Data collection

Patients were divided into two groups: the experimental group (BD with arterial involvement) and the control group (BD without any vascular involvement). Collected data included demographics and clinical features: age at onset, disease duration, sex, family history, and systemic manifestations (fever, oral/genital ulcers, skin, ocular, gastrointestinal, neurological, renal, cardiac involvement). Laboratory parameters: complete blood count (white blood cell count (WBC), haemoglobin, platelet count (PLT)), acute phase reactants (C-reactive protein [CRP], erythrocyte sedimentation rate [ESR]), serum ferritin, immunoglobulin G (IgG), tumour necrosis factor-alpha (TNF- α), interleukin-6 (IL-6), antinuclear antibodies (ANA), and anti-neutrophil cytoplasmic antibodies (ANCA).

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Competing interests: none declared.

Table I. Anatomical distribution and imaging characteristics of arterial lesions in 19 paediatric Behçet's disease patients with arterial involvement.

Category of involved arteries	Specific involved arteries	Number of cases (n, %)	Main lesion types (number of cases)
Large arteries	Abdominal aorta	8 (42.11%)	Wall thickening (6), Lumen stenosis (4), Dissection (1)
	Pulmonary artery	7 (36.84%)	Lumen dilation (3), Thrombosis (2), Wall thickening (2), Aneurysm (1)
	Thoracic aorta	5 (26.32%)	Wall thickening (4), Lumen stenosis (2), Uneven lumen widening (1)
	Ascending aorta/aortic arch	4 (21.05%)	Wall thickening (2), Lumen stenosis (2), Aneurysm (1),
Coronary artery	Left main coronary artery	5 (26.32%)	Lumen dilation (5) (maximum Z-score 4.38)
Cervicothoracic arteries	Internal carotid artery	5 (26.32%)	Wall thickening (4), Lumen stenosis (3), Localised dilation (1)
	Subclavian artery	4 (21.05%)	Wall thickening (4), Lumen stenosis (2)
Visceral arteries	Superior mesenteric artery	5 (26.32%)	Wall thickening (3), Lumen stenosis (2)
	Renal artery	4 (21.05%)	Lumen stenosis (4), Wall thickening (1)
Upper extremity arteries	Axillary artery	4 (21.05%)	Wall thickening (4), Lumen stenosis (3)
	Brachial artery	1 (5.26%)	Occlusion (1)
Lower extremity arteries	Femoral artery	7 (36.84%)	Wall thickening (6), Lumen stenosis (3)
	Popliteal artery	4 (21.05%)	Wall thickening (3), Lumen stenosis (2)
	Iliac artery	1 (5.26%)	Occlusion (1)
	Dorsalis pedis artery	1 (5.26%)	Occlusion (1)
	Posterior tibial artery	1 (5.26%)	Occlusion (1)

- Imaging characteristics

For the experimental group, the number, location (large/medium-sized), and type of arterial lesions (wall thickening, dilation, stenosis, thrombosis, occlusion, aneurysm) were recorded based on imaging reports.

- Treatment and outcomes

Medications used (glucocorticoids, conventional and biologic disease-modifying anti-rheumatic drugs), surgical interventions, and prognostic outcomes (cerebrovascular accidents, cardiac complications, mortality, recurrence, disease stability).

Statistical analysis

Data were analysed using SPSS software (v. 26.0; IBM Corp., Armonk, NY, USA). The normality of distribution for continuous variables was assessed using the Shapiro-Wilk test. Categorical data are presented as counts and percentages (%), and comparisons between groups were performed using the Chi-square test or Fisher's exact test, as appropriate. Normally distributed continuous data are expressed as mean $\pm$ standard deviation and compared using the independent samples t-test. Non-normally distributed continuous data are expressed as median with interquartile range [IQR] and

Table II. Comparison of demographic and clinical characteristics between paediatric Behçet's disease patients with and without arterial involvement.

	Total (n=76)	BD with arteries involvement (n=19)	BD without vascular involvement (n=57)	p-value
General information				
Age at onset (years) (IQR)	8.16 (4.23, 11.0)	11.0 (8.33, 13.0)	7.0 (3.88, 11.0)	0.015
Disease duration (months) (IQR)	12.00 (2.00, 36.00)	6.0 (2.0, 12.0)	24.0 (2.5, 48)	0.024
Male (n, %)	30, 39.47%	6, 31.58%	23, 40.35%	0.416
Family history of BD (n, %)	3, 3.95%	2, 10.53%	1, 1.75%	0.089
Clinical manifestations (n, %)				
Fever (n, %)	41, 53.95%	15, 78.95%	26, 45.61%	0.012
Oral ulcers (n, %)	70, 92.11%	15, 78.95%	55, 96.49%	0.014
Genital ulcers (n, %)	38, 50.0%	8, 42.11%	30, 52.63%	0.427
Skin manifestations (n, %)	40, 52.63%	12, 63.16%	28, 49.12%	0.289
Eye involvement (n, %)	18, 23.68%	6, 31.58%	12, 21.05%	0.169
Nervous system involvement (n, %)	10, 13.16%	6, 31.58%	4, 7.02%	0.001
Pathergy reaction (n, %)	11, 14.47%	2, 10.53%	9, 15.79%	0.572
Gastrointestinal ulcers (n, %)	55, 72.37%	9, 47.37%	46, 80.70%	0.005
Gastrointestinal perforation (n, %)	2, 2.63%	1, 5.26%	1, 1.82%	0.408
Renal involvement (n, %)	17, 22.37%	9, 47.37%	8, 14.04%	0.003
Cardiac involvement (n, %)	6, 7.89%	6, 31.58%	0, 0	0.000

BD: Behçet's disease; IQR: interquartile range; N: number of patients in the group; n: number of cases.

compared using the Mann-Whitney U-test. A two-tailed p -value <0.05 was considered statistically significant.

A *post-hoc* power analysis was performed using G*Power software (version 3.1.9.7). The analysis was based on the observed difference in the primary outcome of all-cause mortality between the arterial involvement group (10.53%, 2/19) and the control group (0%, 0/57). With an alpha level set at

0.05, a sample size of 19 in group 1 and 57 in group 2, the achieved statistical power ($1-\beta$) for detecting this difference was calculated to be 0.72.

Results

Baseline characteristics and arterial lesion profile

A total of 76 paediatric BD patients were included, with 19 (25%) in the arterial involvement group and 57 (75%)

Table III. Comparison of laboratory parameters and inflammatory markers between paediatric Behçet's disease patients with and without arterial involvement.

Variables	Total (n=76)	BD with arteries involvement (n=19)	BD without vascular involvement (n=57)	p-value
WBC (10 ⁹ /L) (IQR)	10.36 (7.48, 16.30)	14.43 (9.82, 24.81)	9.84 (6.91, 15.98)	0.018
Hgb (g/L) (IQR)	118.0 (105.0, 129.25)	105.00 (89.00, 122.00)	121.00 (109.00, 129.50)	0.006
PLT (10 ⁹ /L) (IQR)	366.50 (298.50, 589.60)	458.00 (310.00, 545.00)	357.00 (290.50, 467.50)	0.126
CRP (mg/L) (IQR)	28.05 (2.56, 85.32)	74.70 (18.01, 136.00)	24.80 (1.85, 65.90)	0.004
ESR (mm/h) (IQR)	39.0 (16.00, 60.00)	66.00 (38.00, 103.00)	29.00 (14.50, 51.00)	0.001
SF (ng/ml) (IQR)	90.10 (50.02, 138.72)	105.00 (48.88, 220.22)	83.23 (49.64, 133.84)	0.471
IgG (g/L) (IQR)	12.40 (10.0, 15.70)	14.55 (11.49, 17.58)	11.80 (9.83, 15.20)	0.115
TNF α (IQR)	11.20 (6.51, 18.60)	9.89 (4.62, 21.15)	11.20 (6.63, 16.58)	0.889
IL6 (IQR)	7.99 (2.84, 20.00)	9.42 (5.42, 26.30)	8.11 (2.29, 19.80)	0.325
ANA positive (n, %)	7, 9.21%	2, 10.53%	5, 8.77%	0.823
ANCA positive (n, %)	4, 5.26%	2, 10.53%	2, 3.51%	0.289

ANA: antinuclear antibodies; ANCA: anti-neutrophil cytoplasmic antibodies; CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; Hgb: haemoglobin; IgG: immunoglobulin G; IL-6: interleukin-6; IQR: interquartile range; PLT: platelet count; SF: serum ferritin; TNF- α : tumour necrosis factor-alpha; WBC: white blood cell count.

in the control group. The characteristics of arterial lesions are summarised in Table I. The median disease duration at the time of vascular lesion discovery was 7.0 (IQR 4.0, 12.0) months. Medium-sized arteries were involved in 15/19 cases (78.95%), large arteries (aorta or pulmonary artery) in 13/19 cases (68.42%), and both in 9/19 cases (47.37%). The most common imaging findings were arterial wall thickening (9/19, 47.37%), lumen dilation (8/19, 42.11%) and lumen stenosis (7/19, 36.84%).

Comparison of demographics and clinical manifestations

As shown in Table II, patients with arterial involvement had a significantly later median age at onset (11.0 years vs. 7.0 years, $p=0.015$), and a shorter median disease duration (6.0 months vs. 24.0 months, $p=0.024$) compared to controls. The frequencies of fever (78.95% vs. 45.61%, $p=0.012$), neurological involvement (31.58% vs. 7.02%, $p=0.001$), renal involvement (47.37% vs. 14.04%, $p=0.003$), and cardiac involvement (31.58% vs. 0%, $p<0.001$) were significantly higher in the arterial involvement group. The prevalence of oral ulcers was paradoxically lower in the arterial group (78.95% vs. 96.49%, $p=0.014$).

Comparison of laboratory results

Laboratory results are detailed in Table III. The arterial involvement group exhibited significantly higher levels of

Table IV. Treatment strategies and clinical outcomes in paediatric Behçet's disease patients with and without arterial involvement.

Variables	BD with artery involvement	BD without vascular involvement	p-value
Follow-up duration (months) (IQR)	16.0 (6.0, 50.0)	12.0 (5.75, 21.25)	0.34
Medication usage			
High-dose steroid pulse, n (%)	9/19 (47.37%)	7/57, 12.28%	0.001
Cyclophosphamide, n (%)	14/19 (73.68%)	15/57, 26.32%	0.000
Methotrexate, n (%)	4/19, 21.05%	6/57, 10.53%	0.24
Colchicine, n (%)	1/19, 5.26%	2/57, 3.51%	0.734
Mycophenolate mofetil, n (%)	6/19, 31.58%	3/57, 5.26%	0.002
Tofacitinib, n (%)	5/19, 26.32%	1/57, 1.75%	0.000
Thalidomide, n (%)	10/19, 52.63%	46/57, 80.70%	0.016
TNF α inhibitors, n (%)	12/19, 63.16%	15/57, 26.32%	0.004
Tocilizumab, n (%)	2/19, 10.53%	1/57, 1.75%	0.089
Surgery, n (%)	4/19, 21.05%	1/57, 1.75%	0.003
Prognosis			
Recurrence or disease activity, n (%)	5/19, 26.32%	12/50, 24.00%	0.842
Cerebrovascular accident, n (%)	3/19, 15.79%	0	0.001
Cardiac complications, n (%)	6/19, 31.58%	0	<0.001
Ocular complications (steroid-induced cataracts/glaucoma), n (%)	2/19, 10.53%	6/50, 12.00%	0.864
Death, n (%)	2/19, 10.53%	0	0.02
Stable condition, n (%)	12/19, 41.38%	38/50, 76.00%	0.286
Discontinuation of steroids, n (%)	2/19, 10.53%	8/50, 16.00%	0.564

BD: Behçet's disease; IQR: interquartile range; TNF- α : tumour necrosis factor-alpha.

Denominator for prognosis indicators was adjusted due to lost to follow-up (19→19, 57→50).

systemic inflammation, evidenced by elevated median WBC (14.43 vs. 9.84 $\times 10^9$ /L, $p=0.018$), CRP (74.70 vs. 24.80 mg/L, $p=0.004$), and ESR (66.00 vs. 29.00 mm/h, $p=0.001$). The prevalence of anaemia (haemoglobin <110 g/L) was also higher in this group (68.42% vs. 42.11%, $p=0.006$).

Comparison of treatment and prognosis

Treatment strategies and outcomes are compared in Table IV. The arterial involvement group received more

intensive immunosuppressive therapy, including higher usage rates of high-dose steroid pulse (47.37% vs. 12.28%, $p=0.001$), cyclophosphamide (73.68% vs. 26.32%, $p<0.001$), mycophenolate mofetil (31.58% vs. 5.26%, $p=0.002$), and TNF- α inhibitors (63.16% vs. 26.32%, $p=0.004$). Surgical intervention was also more frequent (21.05% vs. 1.75%, $p=0.003$). Despite aggressive treatment, this group had significantly worse outcomes, including higher rates of cerebrovascular accidents (15.79%

vs. 0%, $p=0.001$), cardiac complications (31.58% vs. 0%, $p<0.001$), and mortality (10.53% vs. 0%, $p=0.020$).

Discussion

BD is a systemic vasculitis characterised by full-thickness inflammation of the blood vessel wall, which can involve arteries, veins, and capillaries of all sizes (10). Its pathogenesis involves both autoinflammatory and autoimmune features, primarily mediated by Th1 and Th17 cell responses, neutrophil activation and perivascular inflammation (11). Activated neutrophils release superoxide radicals and lysosomal enzymes, damaging the vascular wall, leading to endothelial dysfunction and further platelet activation, which promotes thrombosis (12). Venous involvement occurs in approximately 20–40% of BD patients (13), while arterial involvement, although affecting only 3–5% of patients, represents a unique feature of BD (14), that can affect peripheral, visceral and cardiopulmonary regions. Its progression is rapid, with numerous complications and a poor prognosis (15). Our finding that 25% of our paediatric cohort had arterial involvement suggests this complication may be more common in children than adults, warranting heightened clinical vigilance.

In this study, children with BD and arterial involvement had a later age of onset (median 11.0 years) and a shorter disease duration (median 6.0 months), suggesting they may be in a phase of more intense immune activation and more rapid disease progression. The clinical profile was dominated by constitutional symptoms (fever) and severe organ involvement (neurological, renal, cardiac), contrasting with the more classic mucocutaneous presentation seen in the control group. The laboratory hallmark was a pronounced acute-phase response, with markedly elevated WBC, CRP, and ESR. This reflects a more severe systemic inflammatory response and a higher risk of multi-system involvement in children with arterial involvement.

This study found that paediatric BD arterial involvement primarily affected medium-sized (15/19, 78.95%) and large arteries (13/19, 68.42%). The

most commonly affected arteries were the abdominal aorta (42.11%), pulmonary artery (7/19, 36.84%), and femoral artery (7/19, 36.84%). Notably, the involvement rate of lower extremity arteries (*e.g.* femoral, popliteal) was higher than that of the upper extremities, suggesting that clinical assessment should pay special attention to the lower limb vasculature. In adult BD, the most commonly involved arteries are the abdominal aorta, followed by the pulmonary artery, femoral artery, and iliac artery (16), with pulmonary artery involvement reported in 1–18% of cases, predominantly in males. In contrast, this paediatric cohort showed no gender difference in arterial involvement.

Imaging findings in paediatric BD arterial lesions were primarily wall thickening (9/19, 47.37%), lumen dilation (8/19, 42.11%), and lumen stenosis (7/19, 36.84%). The proportions of aneurysms (2/19, 10.53%) and thrombosis (3/19, 15.79%) were lower than in adults (6, 17), which may be related to the absence of traditional cardiovascular risk factors (*e.g.* smoking, diabetes, hypertension, hyperlipidaemia) in children, as well as a shorter disease duration. A study with a mean follow-up of 3.65 years reported that aneurysms were the most common arterial lesion in adult BD with arterial involvement, though occlusions and stenosis could also occur (17, 18); one-third of arterial involvement was multiple, and 50% of patients had thrombosis (19).

It is noteworthy that coronary artery involvement is a major feature in paediatric BD. In our series, all cases presented with dilation of the left main coronary artery, with the widest having a Z-score of 4.38. As the primary blood supply to the heart, any progression to stenosis or infarction of the left main coronary artery directly impacts prognosis. In contrast, coronary artery involvement in adult BD is very rare (overall incidence <0.5%) (7), often involves the left anterior descending artery, and commonly presents as stenosis, occlusion, or pseudoaneurysm (20, 21). This indicates significant differences in the manifestations of coronary artery involvement between paediatric and adult BD.

Recommendations for managing vascular BD are scarce (22). This study found that the proportions of patients receiving high-dose steroid pulse therapy (9/19, 47.37%), cyclophosphamide (14/19, 73.68%), TNF- α inhibitors (12/19, 63.16%), and surgical intervention (4/19, 21.05%) were all significantly higher in the experimental group. Once paediatric BD is complicated by arterial involvement, the treatment strategy must be escalated from routine symptom control to a 'potent, rapid, multi-target' crisis management mode. High-dose steroid pulse therapy is the cornerstone for rapidly suppressing acute vascular inflammation. Cyclophosphamide, as a potent conventional immunosuppressant, had the highest usage rate in this group, reflecting the urgent need to induce remission in severe vasculitis (23). The high usage rate of TNF- α inhibitors represents a recent therapeutic advance. TNF- α plays a key role in vascular wall inflammation; its targeted inhibition helps control vascular inflammation more rapidly, promote vascular wall repair, and may reduce the risk of aneurysm formation and rupture (24–26). The high surgical rate indicates that for structural complications (*e.g.* aneurysms, stenosis, perforation) poorly controlled by medication, surgical or interventional therapy is a life-saving measure. Despite intensive treatment, the experimental group still had high rates of complications and mortality. This suggests that current treatments may have a 'lag effect'. Once structural vascular damage occurs, it is often irreversible. Therefore, the key to future management lies in earlier identification of high-risk children, more proactive vascular imaging screening, and intervention at the early stages of vascular damage, rather than waiting for aneurysms or severe stenosis to appear.

Conclusion

In summary, paediatric BD with arterial involvement is characterised by later-onset but rapidly progressive disease course, intense inflammations, a high burden of multi-system involvement, and a poor prognosis. Clinicians should maintain a high index of suspi-

cion for vascular disease in paediatric BD patients, particularly adolescents presenting with fever and elevated inflammatory markers. Early and systematic vascular imaging evaluation, focusing on the aorta, pulmonary, coronary and lower extremity arteries, is recommended. Treatment should adopt an early, intensive strategy of combined immunosuppressive and biologic targeted therapy, with a proactive approach to surgical intervention. Close follow-up should focus on vascular complications and the protection of end-organ function, aiming to ultimately improve the long-term prognosis of these critically ill children.

Limitations

Several aspects of this study should be considered when interpreting the results. First, the retrospective nature and single-centre setting, while enabling the collection of this relatively large paediatric cohort, may limit the generalisability of the findings. Second, the sample size, though substantial for a focused study on paediatric arterial BD, remains modest. A post-hoc power analysis on the mortality outcome yielded a power of 0.72. While this indicates a reasonable probability of detecting the observed significant difference, it also suggests that future studies with larger sample sizes would be beneficial to confirm these findings and explore potential risk factors with greater precision.

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