Review

Cardiac involvement in Behçet's disease: a comprehensive state-of-the-art review

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ABSTRACT

Behçet's disease (BD) is a chronic multisystemic inflammatory vasculitis which can cause cardiac involvement in less than 6%. It can affect all cardiac wall giving rise to pericarditis, myocarditis, endocardial lesions with valvular involvement (especially aortic insufficiency), intracardiac thrombosis, coronary arteritis, myocardial infarcts, arrythmia, and aortitis. Clinical presentation can be confusing, silent, or overshadowed, so diagnosis is frequently delayed. Early diagnosis is mandatory and different imaging techniques play a pivotal role to screen the heart. Biological agents, including tumour necrosis factor inhibitors, show promising results. Cardiac surgery must be accompanied by immunosuppressive therapy to prevent iatrogenic or trauma-triggered pseudo aneurysm secondary to arterial hypersensitivity. A multidisciplinary approach and tailored monitoring are essential to improve patient outcome. Strong data are still lacking to define evidence-based diagnostic algorithms, risk-stratification tools, and standardised management strategies for this severe cardiac manifestation. Besides, further studies are needed to determine specific biomarkers to refine early diagnosis, assess therapeutic efficiency, and prognosis. Through a comprehensive literature review, we aim to synthesise current data on the epidemiology, clinical presentation, contemporary imaging modalities, prognosis, and evidence-based management strategies of cardiac involvement in BD, and to highlight emerging perspectives.

Introduction

Behçet's disease (BD) is a chronic, relapsing multisystemic, inflammatory vasculitis affecting veins and arteries with multiorgan involvement and heterogeneous clinical manifestations. Its etiopathogenesis remains partially elucidated but is thought to be a compilation of genetic predisposition combined with environmental triggers that lead to an imbalance in the innate and adaptive immune system actors (1). Among the systemic manifestations, cardiovascular involvement represents one of the most severe and prognostically significant features, despite its relative low prevalence (2). Cardiac involvement may affect all cardiac and vascular tunics, including the endocardium, myocardium, pericardium and great vessels, and may present either as isolated manifestation or in association with other vascular or visceral involvement. Clinical presentation is highly heterogenous, ranging from asymptomatic disease detected incidentally on imaging to fulminant heart failure, myocardial infarction, massive haemoptysis or sudden death (3). Confusing clinical presentation, combined with the absence of specific biomarkers and the frequent incompleteness of systemic features, explains why cardiac BD remains underdiagnosed. Another major challenge lies in disease monitoring. Inflammatory markers are unreliable in BD and may remain normal despite active cardiac or vascular inflammation. Moreover, there is no standardised diagnostic or therapeutic strategy, and management remains largely experience-based. Evidence is mainly derived from observational cohorts, case series, and expert opinion (4). In this context, we provide an extended state-of-the-art review of cardiac involvement in BD with a particular focus on epidemiological, clinical features, multimodality imaging, and contemporary management strategies.

Methods

We conducted a comprehensive narrative review of the literature. PubMed, MEDLINE, Embase, ScienceDirect, Google Scholar and the Cochrane Library were searched from database inception to December 2025 using combinations of the following terms: "Behçet's disease", "cardiac", "aortitis", "valvular disease", "aortic insufficiency", "myocarditis", "pericarditis", "intracardiac thrombus", "coronary arteritis", "pulmonary artery aneurysm", "echocardiography", "cardiac magnetic resonance", "computed tomography", and "positron emission tomography". Given the rarity and the heterogeneity of cardiac involvement in BD, observational studies, cross-sectional cohorts, case series, selected case reports, and relevant narrative or systemic reviews were included to provide comprehensive overview of this condition. Studies were selected based on clinical relevance and methodological quality. Editorials without original data and reports lacking sufficient clinical detail were excluded. Data were extracted on epidemiology, clinical presentation, imaging findings, histopathology, treatment strategies and outcome. Evidence was synthetised narratively, with an emphasis on clinically meaningful patterns, areas of consensus and unresolved controversies.

Clinical presentation

The epidemiological, clinical features, management approaches, and outcome of BD patients with cardiac involvement according to the different published case series are summarised in Table I. Figure 1 schematises the main BD-related cardiac involvement.

Endocardium involvement

- Aortic valve insufficiency

Aortic valve insufficiency (Fig. 2) is the most frequent valve involvement in more than two third of Chinese BD patients and seems to be secondary to root aortic dilatation (19). This pattern of cardiovascular involvement also illustrates the geographic variation in disease expression in BD, as aortic valvular lesions are reported far more frequently in Far Eastern countries such as Korea, Japan and China, whereas they are rare or even absent in countries such as Turkey (6). Clinical presentations are non-specific, such as chest pain, shortness of breath, palpitations, or heart failure. Diagnosis can be challenging due to overlap with infective endocarditis. In preoperative echocardiographic study from China enrolling 26 BD patients, moderate to severe aortic regurgitation was found in 22 cases (84.6%) with some distinctive echocardiographic features including prolapse of aortic cusps (61%), vegetation-like mobile lesions (18%), and annular echo-free spaces within the annulus extending into the aortic root or interventricular septum (27%) (10). Besides, aortic cusps thinning and redundancy of aortic leaflets, with or without mobile masses, may be particularly characteristic of BD as compared to other causes of aortic insufficiency (9). Zhang *et al*. studied pathological characteristics of the aortic valve in BD patients. In active subgroup, diffuse inflammatory cell infiltration with predominant neutrophils compared to remission cases (60% *vs*. 7.7%; p=0.044). Mucoid degeneration, neovascularisation, and necrosis were more prevalent in active aortitis. Immunohistochemistry analysis showed diffuse $CD4^+$ T cells (60% *vs*. 20%), $CD8^+$ T cells (75% *vs*. 25%), $CD68^+$ macrophages (75% *vs*. 0%) in active cases (20). Surgical management is particularly challenging in this setting, as postoperative complications such as prosthetic valve detachment, suture dehiscence, and peri-anastomotic leakage are frequent, often requiring technical modifications to standard surgical procedures.

- Mitral valve insufficiency

A cross-sectional descriptive-analytic study from Iran comparing BD patients to controls, concluded that a significant severe mitral insufficiency with consequent higher Systolic Pulmonary artery pressure was significantly higher in BD patients even in asymptomatic patients (21). Echocardiography may reveal mitral valve prolapse, leaflet thickening, and regurgitation jets; tissue Doppler can detect early contractile dysfunction even when conventional systolic function appears preserved (22).

- Tricuspid valve involvement

Tricuspid valve involvement in BD is rare. it is often pauci-symptomatic making systematic echocardiography essential for diagnosis (21). Literature review found reports of either tricuspid regurgitation or stenosis presenting as aseptic endocarditis. Tricuspid insufficiency has been reported to be associated with endomyocardial fibrosis or intracardiac thrombosis. In fact, valvulitis may extent to adjacent structure leading to fibrosis and clot genesis (23).

- Pulmonary valve involvement

To our best knowledge pulmonary valve involvement was reported in only three cases in the English literature and exclusively in patients from Asia. Little is known about this feature, but available data reported low echogenic mass attached to the pulmonary valve with neither associated stenosis (normal jet velocity) nor pulmonary embolism. Right chambers were normal as well (6, 10).

Intracardiac thrombosis (ICT)

ICT occurs in patients originally from the ancient Silk Road areas with a predominance around the Mediterranean countries. It occurs predominantly in males with an age of onset in the third decade (range 12 to 51 years) and is initial in half of cases. In the other cases, it happens with a mean delay of 4 years after BD diagnosis (24). The thrombogenesis in BD is multifactorial. BD is a model of thrombo-inflammation where sustained inflammation causes endothelial damage which increase expression of tissue factor, superoxide-dependent nitric oxide inactivation leading to the loss of physiological anticoagulant function. Activated platelets enhance tissue factor expression and thrombin generation resulting in prothrombotic state (1, 25). Clinical presentation is unspecific and the main reported symptoms were fever, haemoptysis, dyspnoea, chest pain and cough (26, 27). In about one third of cases, patients do not show the classic extra-cardiac symptoms of BD (28). A meticulous work up is mandatory to rule out differential

Table I. Literature review on main studies on BD-related cardiac involvement.

Author	Country	n	Age	M/F	I CI (%)	Symptomatic CI (%)	Silent CI (%)	Pericarditis (%)	AVI (%)	MVI (%)	TVI (%)	PVI (%)	ICT (%)	MI (%)	EMF (%)	Myocarditis (%)	PAA (%)	CAI (%)	GC (%)	IS (%)	Anticoagulation (%)	Surgery (%)	PCI (%)	SC (%)	CR (%)	Relapse (%)	Death (%)
Geri *et al.* (5)	France	52	29.7	0.8	17 (32.7)	18 (34.6)	34 (65.3)	20 (38,5)	9 (17.3)	3 (5.7)	2 (3.8)	0	10 (19.2)	9 (17,3)	4 (7.7)	0	0	12 (23)	39 (75)	31 (59.6)	14 (63.6)	8 (15.4)	4 (7.6)	0	24 (46.1)	8 (15.4)	8 (15.4)
Lee *et al.* (6)	Korea	12	38	2	10 (83.3)	12 (10)	0	0	9 (75)	NS	2 (16.6)	1 (8.3)	4 (33.3)	1 (8.3)	NS	NS	NS	NS	10 (83.3)	8 (66.6)	1 (8.3)	12 (100)	0	4 (33.3)	5 (41.6)	1 (8.3)	1 (8.3)
Chadli *et al.* (7)	Morocco	32	30	3	NS	9 (29)	23 (71)	11 (34)	NS	NS	NS	NS	25 (79)	NS	1 (3)	2 (6)	8 (25)	4 (12.5)	31 (96)	32 (100)	17 (54)	2 (6)	0	0	NS	1 (3)	5 (15.6)
Yildirim *et al.* (8)	Turkey	14	32	∞	3 (21.4)	14 (100)	0	0	NS	NS	NS	NS	8 (57.1)	4 (28.6)	0	0	4 (28.6)	2 (14.3)	14 (100)	13 (92.8)	9 (64.2)	NS	NS	NS	NS	NS	NS
Pu *et al.* (9)	China	63	39	2.9	39 61.9)	38 (60.3)	24 (38)	1 (1,6)	41 (65)	3 (4.8)	NS	NS	3 (4.8)	5 (7.9)	0	0	1 (1.6)	1 (1.6)	NS	NS	NS	43 (66.1)	NS	16 (25.3)	NS	16 (25)	5 (7.9)
Li *et al.* (10)	China	26	37	5.5	20 76.9)	20 (76,9)	6 (23)	NS	22 (84.6)	8 (30.7)	4 (15.3)	1 (3.8)	NS	NS	NS	NS	1 (3.8)	NS	20 (76.9)	20 (76.9)	NS	26 (100)	NS	8 (30.7)	18 (69.2)	NS	NS
Kechida *et al.* (11)	Tunisia	5	31.5	4.3	5 (100)	5 (100)	0	1 (20)	NS	NS	NS	NS	2 (40)	1 (20)	0	1 (20)	NS	NS	5 (100)	5 (100)	1 (20)	0	0	0	5 (100)	0	0
Zhu *et al.* (12)	China	20	35	5.6	NS	NS	NS	NS	13 (65)	0	0	0	7 (35)	NS	NS	NS	NS	NS	20 (100)	20 100)	NS	8 (40)	0	0 NS	NS 0		
Emmungil *et al.* (13)	Turkey	22	29	20	9 (40.9)	18 (81.8)	2 (9)	NS	NS	NS	NS	NS	NS	NS	NS	NS	16 (72)	NS	22 (100)	21 95.4)	8 (36.3)	0	0	0	13 (59)	1 (4.5)	1 (4.5)
Pu *et al.* (9)	China	63	37	2.9	38 (60.3)	36 (57)	27 (42.8)	1 (1,6)	41 (65)	3 (4,8)	0	0	3 (4.8)	1 (1.6)	NS	NS	1 (1.6)	1 (1.6)	NS	NS	NS	43 (68.3)	NS	NS	NS	NS	5 (7.9)
Ahmed *et al.* (14)	Egypt	20	30	3	NS	16 (80)	4 (20)	3 (10)	2 (6,7)	9 (30)	9 (3)	NS	1 (3,3)	0	0	6 (20)	4 (13.3)	2 (6.7)	NS	NS	NS	NS	NS	NS	NS	NS	NS
Hammami *et al.* (15)	Tunisia	10	37.3	4	3 (30)	8 (80)	2 (20)	1 (10)	0	0	0	0	4 (40)	0	1 (10)	1 (10)	1 (10)	5 (50)	10 (100)	10 (100)	7 (70)	1 (10)	3 (30)	0	7 (70)	3 (30)	1 (10)
Yao *et al.* (16)	China	14	NS	NS	14 (100)	14	0	0	1 (7.1)	1 (7.1)	0	0	10 (71.4)	1 (7.1)	0	0	2 (14.2)	5 (35.7)	14 (100)	14 (100)	NS	2 (14.2)	0	0	14 (100)	0	0
Shadmanfar *et al.* (17)	Iran	47	29	1,04	NS	20 (42.5)	27 (57.4)	10 (21.2)	11 (23)	0	0	0	1 (2)	9 (19)	NS	2 (4.3)	2 (4.3)	1 (2)	NS	NS	NS	NS	1	NS	NS	NS	NS
Beyazel *et al.* (18)	Turkey	14	NS	NS	2 (14.2)	0	14 (100)	0	1 (7.1)	9 (64.2)	NS	NS	4 (28.5)	0	0	0	0	0	4 (28.5)	3 (21.4)	4 (28.5)	0	0	0	NS	NS	NS

n: number of patients; M/F: sex-ratio male/female; CI: cardiac involvement; I CI: inaugural cardiac involvement: AVI: aortic valve insufficiency; MVI: mitral valve insufficiency; TVI: tricuspid valve insufficiency; PVI: pulmonary valve insufficiency; ICT: intracardiac thrombosis; MI: myocardial infraction; EMF: endomyocardial fibrosis; PAA: pulmonary artery aneurysm; CAI: coronary artery involvement; GC: glucocorticoids; IS: immunosuppresant; PCI: percutaneous coronary intervention; SC: surgery complications; CR: clinical remission; NS: non specified.

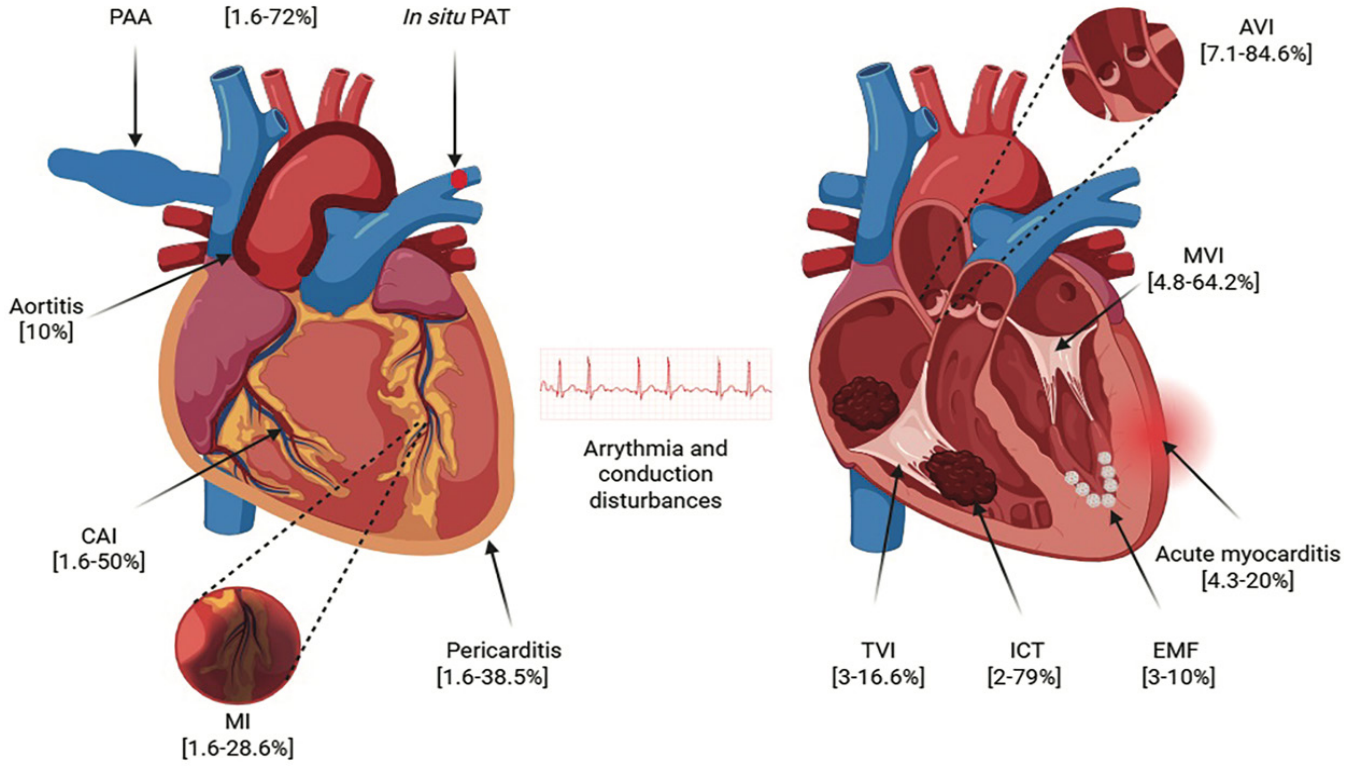


Fig. 1. Cardiac involvement in Behçet's disease.
PAA: pulmonary artery aneurysm; PAT: pulmonary artery thrombosis; CAI: coronary artery involvement; MI: myocardial infraction; TVI: tricuspid valve insufficiency; ICT: intracardiac thrombosis; EMF: endomyocardial fibrosis; MVI: mitral valve insufficiency; AVI: aortic valve insufficiency.

diagnosis such as tumours and infective endocarditis (29). ICT is closely associated with pulmonary artery involvement and other venous thromboses, particularly vena cava thrombosis (13). ICT was mostly reported in the right cavities (Fig. 3) and attributed to extending of thrombi in vena cava and lower pressure of the right heart. ICT in the left heart or in both left and right cavities is also possible (30).

Endomyocardial fibrosis (EMF)
EMF in BD was first reported in a *post-mortem* study, with only 16 cases reported to date (31). Fibrotic genesis is due to endocardium and/or myocardium arteriolar vasculitis with focal fibrinoid deposition. Sustained and repeated endocardial inflammation episodes pave the way of EMF (32). EMF in BD predominately involves the right ventricle but may be also located in the left ventricle and in the atrium. EMF is often subclinical and may be incidentally discovered during routine echocardiography or advanced cardiac imaging. Symptoms occur only when the extent or location of fibrosis leads to significant restrictive cardiomyopathy, heart failure, valvular dysfunction such as tricuspid or pulmonary regurgitation associated with ICT or arrythmias (33). Early recognition is essential, as the prognosis is poor if it is not managed appropriately.

Myocardial involvement
- Acute myocarditis
Myocarditis related to BD is very rare, with diagnosis in only 1.2% of the autopsies performed in a cohort of Japanese patients with BD (34). Nevertheless, subclinical myocardial involvement has been documented in one study, suggesting that myocardial inflammation may be underestimated in BD (14). Recent literature review revealed 8 cases of acute myocarditis, all males aged between 26 and 67 years old originally from Middle East, North Africa and Western Europe. The main clinical presentation was heart failure in the context of new-onset dilated cardiomyopathy (n=4), acute chest pain (n=3), and stroke (n=1) (7, 35-39).
Histopathological findings mirror those observed in other cardiac structures, with inflammatory infiltrates, myocyte necrosis and interstitial fibrosis.

- Myocardial infarct (MI)
In recent systematic review, MI was identified in 62 cases mostly males with a mean age of 37 years old. Clinical presentation, time-dependent troponin level elevation, echocardiographic findings were similar to classic MI. Compared to general population, patients were younger and the prevalence of traditional risk factors were lower: 6% had arterial hypertension, 4.8% hyperlipemia, and 21% history of smoking. Coronarography was performed in 58 patients and was normal in 8 cases (40). In a case control study from Turkey, silent myocardial ischemia was found in 8 (19.5%) out of 41 BD patients. Only one positivity (2.9%) was recorded in the healthy control group on exercise testing and myocardial perfusion SPECT. Coronary angiography was considered normal in all BD patients (41). BD related MI is related to inflammatory coronary arteritis (Fig. 4), *in situ* thrombosis or aneurysmal disease rather than atherosclerosis (42).

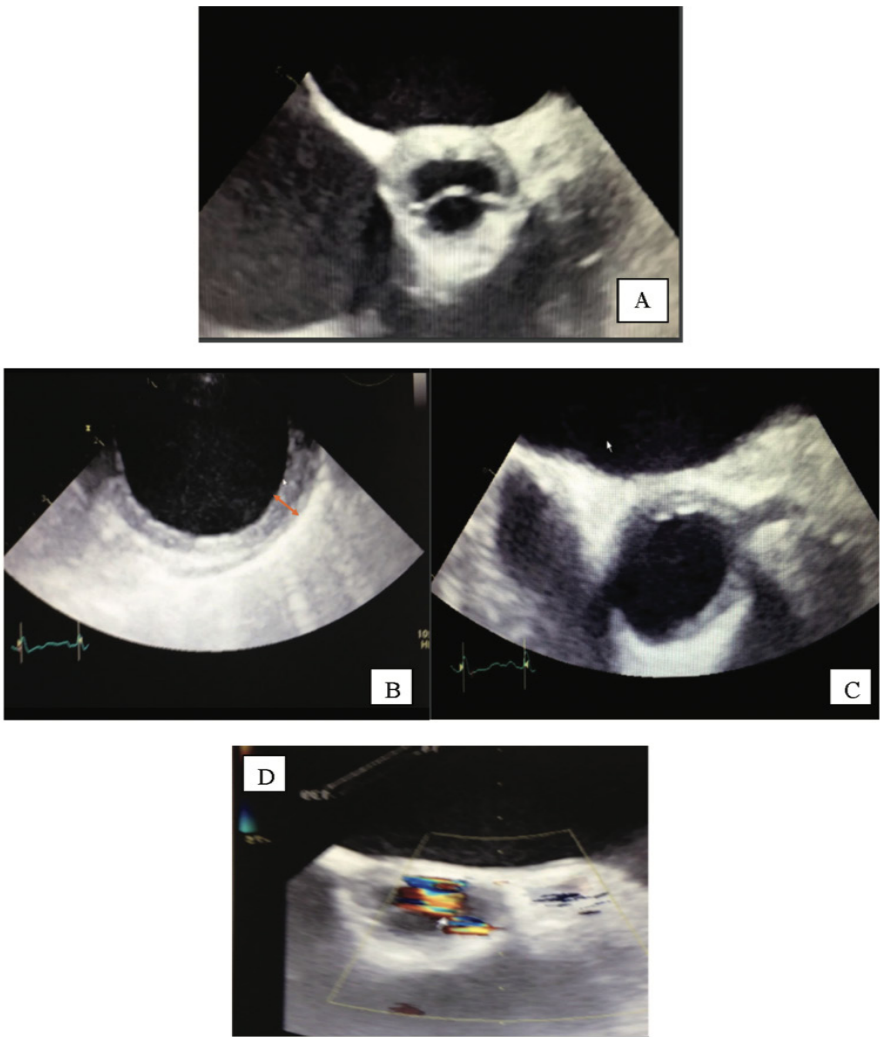


Fig. 2. Echocardiography demonstrating limited mobility of the posterior and left anterior cusp leading to an aortic insufficiency (**A**). Circumferential and hypoechoic thickening of the aortic wall demonstrating BD-related aortitis (**B**) with an extension toward the left main trunk causing stenosis (**C**). Ensheathment of the two aortic cusps resulted in valvular restriction with aortic regurgitation (**D**). (Original data).

- *Septal aneurysm*

In a case control study evaluating echocardiographic features in 54 BD patients free of cardiovascular symptoms and 36 age-matched controls from Turkey, atrial septal aneurysm was more frequent in BD patients than controls (43). Most atrial septal aneurysms in BD are asymptomatic and detected incidentally with cardiac imaging. Otherwise, arrhythmia, heart failure symptoms, or thrombus genesis with embolic events have been reported. Aggressive and invasive therapeutic approach is recommended independently of aneurysm size (44). A handful of reports documented isolated interventricular septal aneurysm in terms of potential clinical feature of cardiac BD. It involves extension from the right sinus of Valsalva into the interventricular septum causing dissection or bulging. Progressive dissection is reported with communication with the aortic cavity through a perforation at the aortic base or with the left ventricular cavity (45).

Pericarditis

According to case series pericarditis is among the most common cardiac manifestations of BD, with prevalence ranging from 26% to 38.5% in patients with cardiac involvement. Most affected patients are young male from Mediterranean regions (Morocco, Egypt, France) (5, 7, 46). Pericarditis in BD is often benign and typically resolves rapidly; however, other presentations including recurrent episodes, cardiac tamponade, or constrictive pericarditis are also possible (Fig. 5) (47). Moreover, there have been reports in which the effusion was attributed to vasculitis of the pleura and the pericardium. Pericardial fluid analysis revealed to be lympho-dominant exudate and pathologic finding showed mild endothelitis with micro-thrombi formation in the lumen (48).

Coronary artery involvement (CAI)

CAI is reported approximately in 0.5% to 2% in some large series and affects young to middle-aged males who often lack traditional cardiovascular risk factors. A time interval of 3 to 16 years has been observed between BD onset and arterial complication. But in one quarter of reports, acute coronary syndrome is the first symptom that leads to the diagnosis (49). The main clinical presentations are ST-elevation myocardial infarction, chest pain, and sometimes heart failure, arrythmia, or sudden death (40). Angiographic findings are highly distinct from typical atherosclerosis coronary artery disease and shows aneurysms, stenosis, pseudoaneurysms, occlusions, and arteritis (50, 51). Coronary artery aneurysm is the hallmark and most common form of coronary vasculitis in BD. These aneurysms may be saccular or fusiform in shape and frequently involve the left anterior descending artery. Unlike atherosclerotic aneurysms, Behçet-related aneurysms can be multiple and often contain intraluminal thrombi. Vessel occlusion typically results from in situ thrombosis, while stenosis arises from diffuse or localised inflammatory thickening of the arterial wall (52). In this context, coronary stent implantation may be complicated by occlusion and/or aneurysm formation in BD (53,54). Pathological findings showed transmural inflammation initially concentrated in the adventitia, often involving the vasa vasorum, before extending into the media and intima. This widespread, destructive inflammation causes two primary lesions: aneurysm formation due to medial and elastic layer destruction, and luminal stenosis/occlusion due to secondary intimal thickening (intimal hyperplasia) (55).

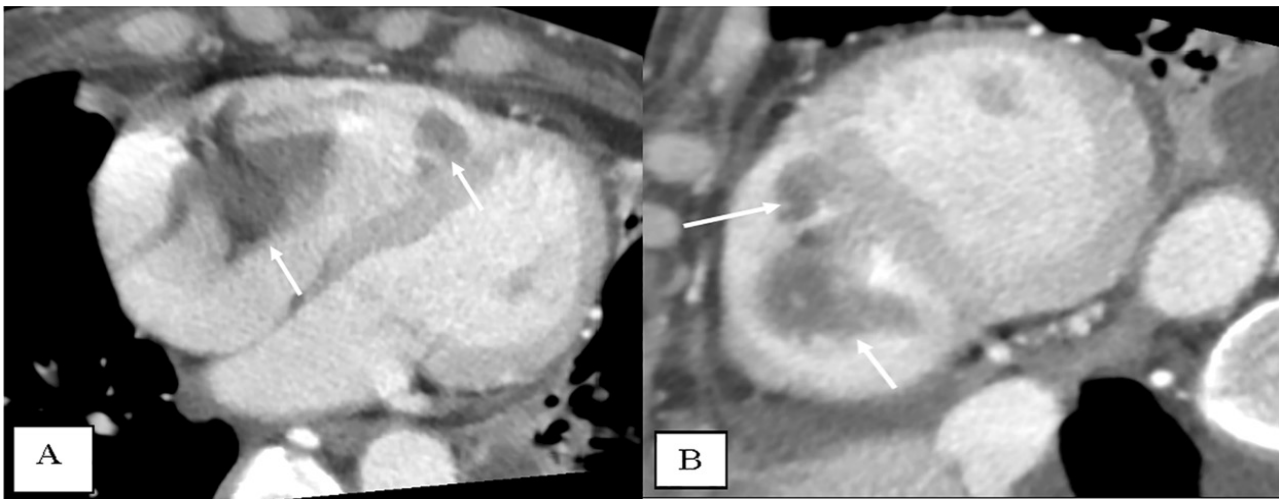


Fig. 3. Four chamber cardiac view with hypodense mass in the right atrium and right ventricle consistent with intracardiac thrombosis. (**A**). Short-axis ventricular view showing endoluminal thrombus in the right ventricle (**B**). (Original data).

Aortitis and pulmonary artery involvement

Aortitis is reported in approximately 10% of BD patients (Fig. 2) and primarily characterised by aortic aneurysms in the aortic root or the abdominal aorta which contribute to disease's severity. Diffuse aortitis is rare in BD (<5%), distinguishing it from other large-vessel vasculitis. Involvement of the abdominal aorta is a late complication, and the diagnosis is usually delayed because of subtle clinical symptoms. Sustained transmural inflammation can destroy the elastic and muscle layers of the media. The destruction of the inner layers leads to pseudoaneurysm genesis. Clinically, patients may be asymptomatic or present with abdominal, thoracic, or lumbar pain pulsatile mass, aortic valve regurgitation, or spontaneous aortic rupture as the initial manifestation. Besides, systemic signs are common, including fever and elevated acute phase reactants (19, 56). Pulmonary artery aneurysms are often multiple, bilateral, and affecting the lower lobes with high risk of rupture in the bronchial tree causing life-threatening haemoptysis (8, 57). PAI frequently coexists with other types of vascular involvement in BD, particularly venous thrombosis and ICT. Pulmonary in-situ thrombosis can occur and may complicate or coexist with aneurysms. Furthermore, small-sized pulmonary vasculitis is also reported with a presentation of alveolar haemorrhage and/or ground-glass appearance (8). Besides, in a cross-sectional study, pulmonary arterial wall thickness evaluated by echocardiography was significantly associated with BD related major organ involvement (58).

Arrhythmias and conduction disturbances

Arrhythmias and conduction disturbances in BD are likely underestimated, yet they signify active cardiac involvement and are a negative prognostic factor. A large population-based study by Lee et al. found that BD patients, particularly younger males (under 40), had a two-fold increased risk of atrial fibrillation (AF) as compared to controls. Sustained inflammation and the upregulation of various multiple inflammatory signalling pathways have been linked to arrhythmogenic mechanisms underlying AF development (59). Tissue Doppler echocardiography analysis of atrial electromechanical delay (inter atrial and intra atrial) revealed a prolonged delay specifically in patients with active BD, confirming this delay as a crucial arrhythmogenic substrate for AF (60). A recent Tunisian case-control study by Ben Mrad et al. evaluated the risk of ventricular arrhythmias in BD patients using heart rate variability (HRV) analysis concluded to parasympathetic system impairment and an increased susceptibility to ventricular arrythmia and sudden death (61). Moreover, electrocardiographic analysis revealed a prolonged QT interval and an increased Tp-e/QT ratio in patients with BD, indicating an elevated risk of cardiac arrhythmia (62).

Cardiac involvements in paediatric BD

In paediatric BD, cardiac involvement predominantly arises within severe vasculo-inflammatory phenotypes. To date, 34 rigorously documented cases of cardiac complications have been reported. Pulmonary artery aneurysms, intracardiac thrombi, and myocarditis occur more frequently than isolated valvular disease (18). Coronary artery lesions typically demonstrate a slowly progressive pattern of arterial dilatation. Early recognition remains challenging, as systemic manifestations may be subtle or evolve insidiously over time.

Cardiac imaging

There is no single gold-standard imaging technique for cardiac assessment. Multimodality imaging plays a central role in the diagnosis, phenotyping and longitudinal assessment of cardiac BD.

Echocardiography

Transthoracic echocardiography is the first-line imaging modality for cardiac screening both at baseline and in symptomatic patients. It allows evaluation of valvular disease such aortic insufficiency, cusp prolapse, vegetation-like mobile lesions, aortic root pseudo-abscesses, and aneurysm formation (10).

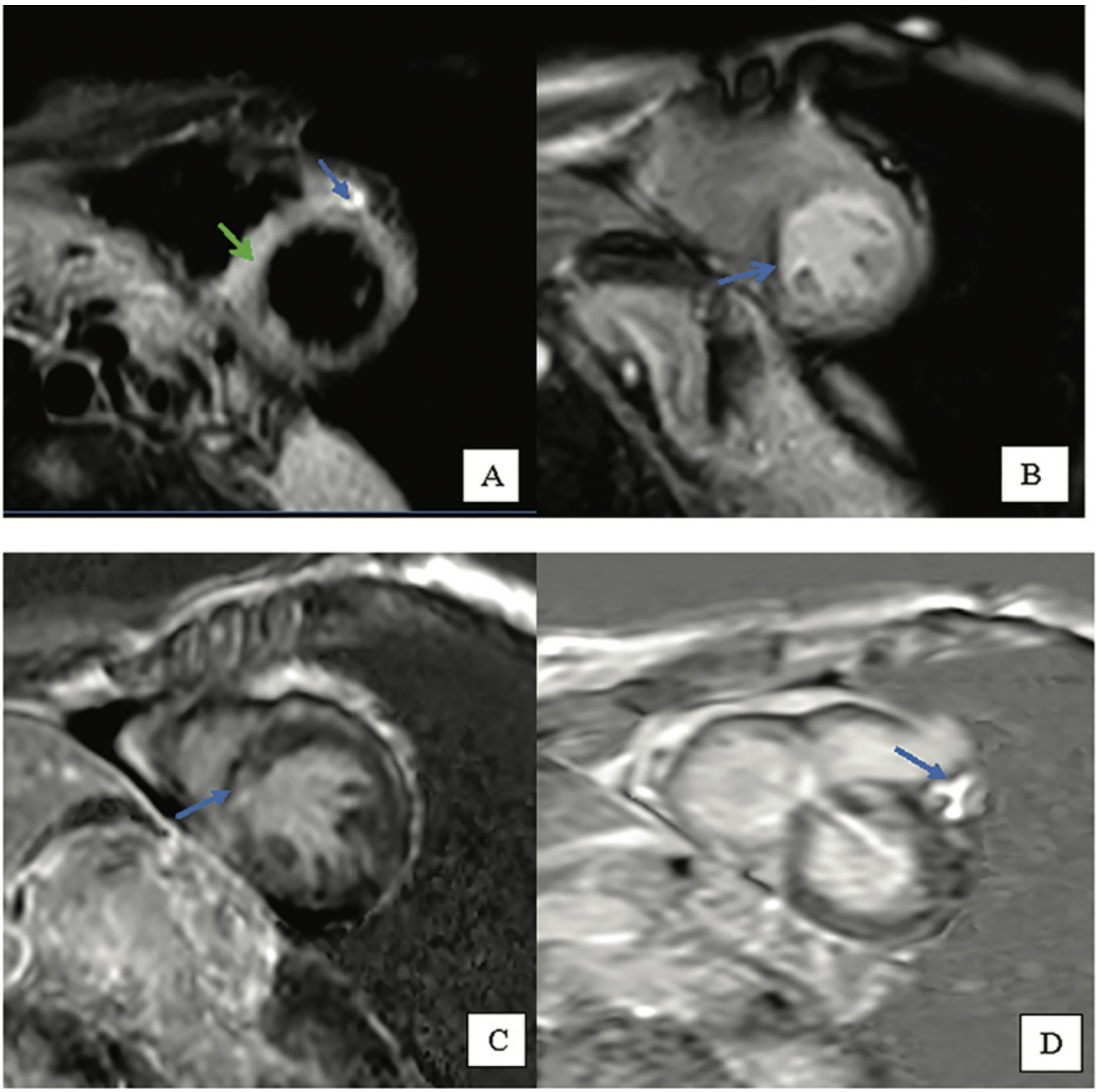


Fig. 4. Coronary arteritis with acute septal Infarction. (**A**). T2 FATSAT short-axis sequence: transmural T2 hypersignal in the mid-infero-septal segment, consistent with myocardial oedema (green arrow), and an oedematous T2 hypersignal in the mid-left anterior descending artery (blue arrow). (**B**). First-pass perfusion: subendocardial hypoperfusion in the mid-inferoseptal wall (blue arrow). (**C**). PSIR Late gadolinium enhancement sequence: mid-infero-septal subendocardial late gadolinium enhancement (blue arrow). (**D**). Late gadolinium enhancement sequence: late contrast enhancement of the left anterior descending artery walls (arteritis). (Original data).

Enlargement of the sinus of Valsalva and proximal ascending aorta has been identified as an independent predictor of moderate-to-severe aortic regurgitation, while left ventricular dilation and reduced ejection fraction correlate with significant mitral regurgitation (33). Advanced techniques, such as speckle-tracking echocardiography including speckle-tracking and three-dimensional imaging, have revealed subclinical right ventricular dysfunction and increased pulmonary arterial stiffness, suggesting early pulmonary vascular remodelling. Diastolic dysfunction, characterised by impaired relaxation and elevated filling pressures, may occur even in asymptomatic cases (22, 63).Transoesophageal echocardiography is reserved for complex valvular disease, intracardiac masses, or prosthetic valve complications (64).

Cardiac magnetic resonance imaging

Cardiac magnetic resonance imaging (CMR) is a non-invasive imaging modality that provides unparalleled tissue characterisation. In a cross-sectional study of 30 BD patients without known cardiac involvement, CMR proved more sensitive than echocardiography, revealing abnormalities in 83% of cases and showing a significant cor-

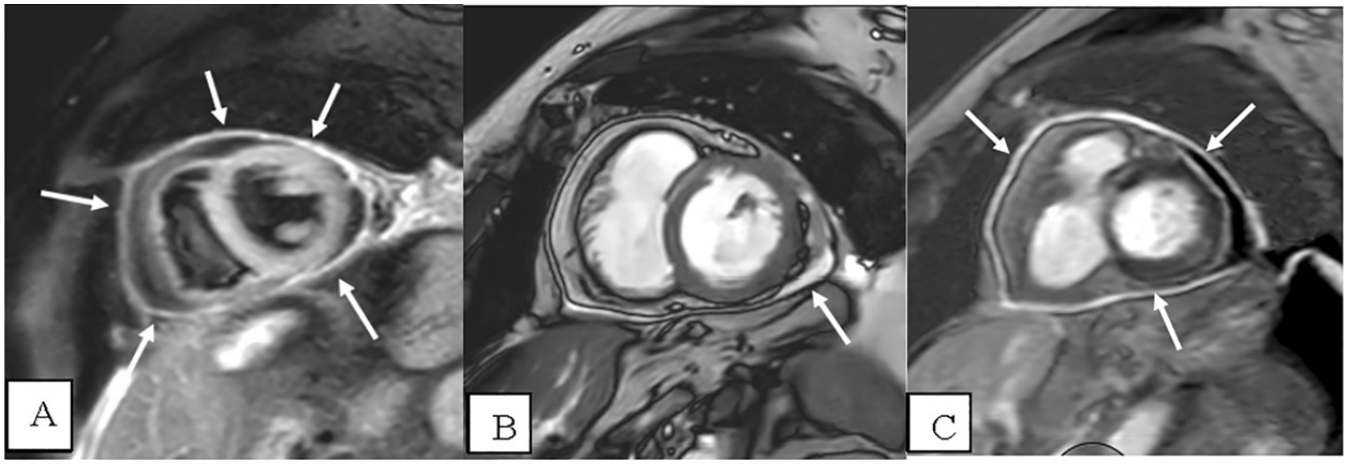


Fig. 5. Pericarditis in BD patients. T2 Fat Sat sequence showing thickening and T2 hyperintensity of the pericardium (**A**). Cine SSFP sequences demonstrating pericardial effusion (**B**). PSIR late gadolinium enhancement sequence with enhanced pericardial thickening. (original data).

relation between cardiac abnormalities and disease activity (14) . CMR can detect and characterise myocardial and intracardiac lesions often missed or incompletely assessed by echocardiography. Late gadolinium enhancement and mapping techniques enable differentiation between with active myocarditis (Fig. 6) or chronic scar which may guide therapeutic decision (14, 65). CMR distinguishes intracardiac thrombi from tumours or vegetations and identifies fibrosis through characteristic enhancement patterns (66). Although its exact role in the diagnostic algorithm remains undefined, CMR is a valuable tool for early detection and risk assessment in cardiac BD. CMR increasingly complements computed tomography (CT) angiography for assessing aortic wall inflammation and periaortic extension particularly when planning surgical interventions for vascular complications (14).

Computed tomography angiography

Computed tomography (CT) angiography is essential for evaluating coronary artery involvement, aortic and pulmonary artery offering excellent spatial resolution and comprehensive assessment of the entire vascular tree. It reliably distinguishes inflammatory lesions, such as circumferential wall thickening, aneurysms, or pseudoaneurysms, from atherosclerotic disease and aids preoperative planning (13). It provides excellent spatial resolution and allows comprehensive evaluation of the entire vascular tree. CT angiography is regarded as the reference imaging modality for confirming pulmonary arterial disease, as it can demonstrate single or multiple aneurysmal 'ballooning' and/or *in situ* thrombosis. CT is often preferred over invasive catheter-based angiography because it avoids arterial puncture and the associated risk of triggering a pathergy phenomenon (40).

Positron emission tomography

Data on ^{18}F-FDG PET/CT accuracy for diagnosing and monitoring BD vascular lesions remain limited, with only small retrospective case series available. These studies report a median vascular FDG uptake index of 1.46 (range 0.58–2.61) in patients undergoing cardiovascular assessment. Increased uptake occurred at multiple sites, including pseudoaneurysms, aortitis with aortic regurgitation, sinus of Valsalva aneurysms, ascending aorta atherosclerosis with regurgitation, and pulmonary artery aneurysms. FDG uptake positively correlated with systemic inflammation. Larger prospective studies are needed to validate its diagnostic utility (67). Emerging tracers such as fibroblast activation protein inhibitors (FAPI) shows promise for diagnosing and monitoring systemic vasculitis, including BD, by enhancing specificity for vascular inflammation and fibrosis compared to ^{18}F-FDG PET/CT. Prospective trials are needed to establish its precise role in BD vascular assessment (67).

Management

Although research data on the management of BD are steadily increasing, current treatment recommendations still rely largely on observational, uncontrolled studies and expert opinion. The marked heterogeneity of cardiac involvement, the absence of standardised outcome measures, and selection bias in published cohorts make it difficult to compare the efficacy of different therapeutic agents. Consequently, the management of cardiac manifestations in BD remains largely empirical and requires a multidisciplinary approach. The core principle is intensive immunosuppressive therapy to control the underlying vasculitis, irrespective of the specific cardiac presentation, combined with carefully planned surgical or interventional procedures and meticulous perioperative care when indicated.

Pharmacological therapy

In line with EULAR guidance (2018), the pharmacological management of BD with cardiovascular involvement is primarily based on high-dose glucocorticoids (GC) combined with potent immunosuppressive therapy to rapidly control severe, acutely active inflammation (68). Recently, treatment strategies for severe BD have evolved, with an increasing tendency to use biologic agents as first line therapy. In a phase 2 Bayesian multicentre randomised controlled trial, patients with major cardiovascular involvement were allocated to receive either intravenous infliximab

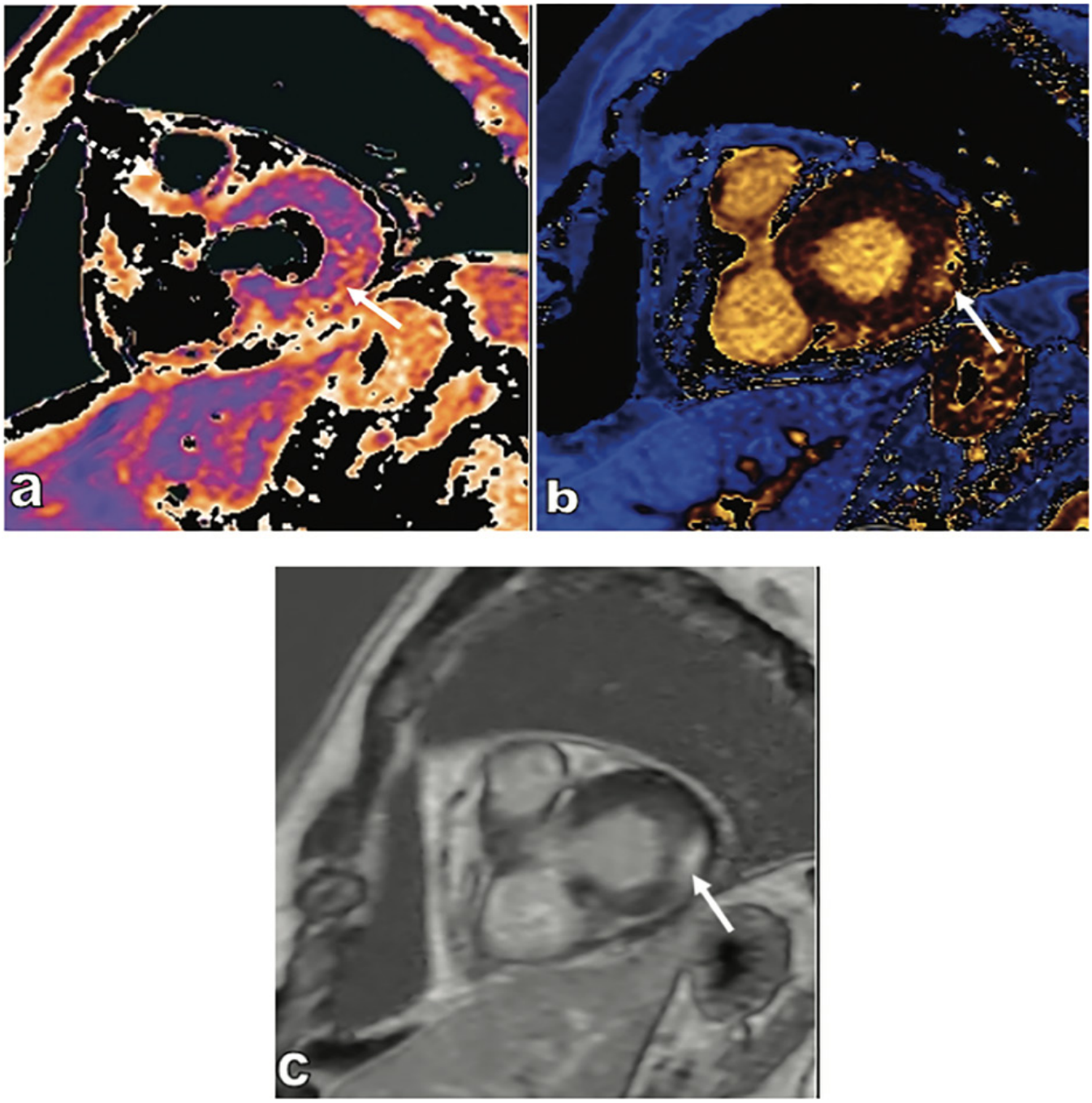


Fig. 6. T2 mapping sequences showing hyper T2 in the subepicardial layer of the mid-inferolateral segment consisting with myocardial oedema (**a**). T1 mapping sequence showing elevated native T1 values in the basal inferolateral wall (**b**). PSIR late gadolinium enhancement sequence showing basal and mid-inferolateral subepicardial late gadolinium enhancement (**c**). (original data).

or cyclophosphamide. Induction with tumour necrosis factor (TNF)-α inhibitors achieved a higher rate of complete response and was associated with fewer adverse events compared with cyclophosphamide-based induction (69). According to last guidance from the British Association of Dermatologists and the British Society for Rheumatology (2024), patients with arterial involvement should receive high-dose oral GC in combination with either cyclophosphamide or an anti-TNF agent for induction, followed by at least two years of maintenance therapy with tapering GC plus azathioprine or ongoing anti-TNF treatment (70, 71). However, their use is contraindicated in patients with moderate to severe heart failure (NYHA class III–IV) due to evidence of worsening outcomes in non-BD populations (72). In patients with pulmonary arterial involvement who respond inadequately to cyclophosphamide or anti-TNF therapy, experts suggest considering tocilizumab or interferon-α2a (70, 71). For ICT, first-line therapy combines glucocorticoids and anti-TNF agents, followed by maintenance with the same biologic or

azathioprine for ≥2 years. Experts recommend against routine anticoagulation alongside immunosuppression for thrombosis, as bleeding risks typically outweigh benefits (70, 71). BD-related pericarditis treatment targets underlying inflammation, adapting 2025 ESC guidelines: first-line colchicine plus NSAIDs; low-moderate dose glucocorticoids if ineffective/contraindicated; IL-1 inhibitors (anakinra, rilonacept) for recurrent/refractory cases (73). ESC recommends aggressive immunosuppression with high-dose glucocorticoids plus azathioprine, cyclophosphamide, or anti-TNF agents for biopsy-proven autoimmune myocarditis (73).

Surgery and interventional procedures

Standard surgical treatment for aortic insufficiency in BD involves aortic root replacement (Bentall/Cabrol procedures) with the button technique for coronary reimplantation, rather than isolated valve replacement. Peri- and postoperative high-dose glucocorticoids plus immunosuppression are essential to prevent complications such as pseudoaneurysms, prosthetic dehiscence, or new vascular lesions (74). Transcatheter aortic valve replacement (TAVR) may be considered for inoperable patients despite technical challenges related to annular dilatation and impaired healing, though evidence remains limited to case reports (75). Arterial aneurysm treatment has shifted toward endovascular/hybrid approaches combined with intensive immunosuppression. Endovascular stent-grafts are preferred for aortic/peripheral aneurysms due to lower perioperative morbidity. Hybrid repair with debranching plus endovascular exclusion is increasingly utilised. Open resection with prosthetic/autologous grafts is reserved for non-endovascular candidates (75). PCI with drug-eluting/covered stents is indicated for focal lesions, urgent haemodynamic instability, or high-surgical-risk cases, including prior CABG restoration. CABG is reserved for multisite lesions or giant aneurysms but carries high early postoperative mortality; on-pump CABG appears feasible in carefully selected patients >40 years requiring multiple distal grafts. A multidisciplinary approach with strict periprocedural inflammation control is mandatory given the arterial pathergy risk (76).

Prognosis and perspectives

Recent cohort studies suggest that cardiac involvement in BD is more frequent in males, current smokers, patients with papulopustular lesions, and those with prolonged activated partial thromboplastin time (APTT >33.15 s), while longer disease duration and hypertension have been less consistently associated. These factors may reflect a vasculitic and prothrombotic phenotype predisposing to pericarditis, intracardiac thrombosis, myocarditis, and valvular disease. Such findings support systematic cardiac screening, particularly by echocardiography, even in asymptomatic patients, to allow earlier initiation of corticosteroids, immunosuppressive therapy, or anticoagulation. However, these associations should be interpreted cautiously, as current evidence is based mainly on small retrospective single-centre cohorts, with limited statistical power and no prospective validation (77, 78). Cardiac involvement and pulmonary artery aneurysm or dilatation are independent adverse prognostic factors in BD with arterial involvement. In a recent long-term Chinese cohort, 5- and 10-year survival rates among patients with cardiovascular involvement were respectively 87.4% and 84.1%, respectively (79). Failure to recognise BD before urgent cardiovascular surgery and to institute appropriate immunosuppressive therapy markedly increases postoperative complications and mortality. Male sex, active smoking, and papulopustular skin lesions have also been significantly associated with a higher cardiac complication (40). Furthermore, several studies have reported that patients in the cardiovascular Behçet subgroup exhibit significantly higher levels of activated partial thromboplastin time, troponin, and CRP, supporting a link between biomarker profiles and cardiac events in this population (77). There is little data on the progression from subclinical to symptomatic cardiac disease. One of the hypotheses of silent vascular damage is altered oxysterol profiles in BD patients from Tunisia. This new biomarker could be an interesting tool to distinguish different forms and its outcome and/or identify efficient treatments (80). Mortality studies on BD are scarce. In a study population from Korea, authors reported a hazard ratio of 1.28 among the population with BD. Interestingly, the risk of mortality was highest in the first 12 months post diagnosis and the most frequent causes of death were cardiovascular events, infections, and gastro intestinal bleeding (81). Additionally, data on long-time survival of BD with cardiac involvement are lacking and the prognostic factors of long-term survival are unknown. There is a real need for tools to assess damage, along with BD activity, and treatment efficiency and/or potential need for surgery. Altogether, larger prospective studies are mandatory to establish diagnostic algorithms, incorporating biomarkers and imaging, as well as personalised treatment. Disease biology is increasingly being explored through microRNA profiling, with miR-155 emerging as a potential independent molecular marker of susceptibility to BD via modulation of pro-inflammatory IL-6/TNF-α pathways (82). A deeper understanding of these mechanisms is crucial for refining risk stratification and optimising management of patients with cardiac involvement.

Conclusion

Cardiac involvement in BD represents a rare but devastating manifestation of a complex systemic vasculitis. It is driven by intense inflammation, thrombo-inflammation and profound tissue fragility, leading to high rates of complications and surgical failure. Early recognition, multimodality imaging and inflammation-adapted medical and surgical strategies are essential to improve outcomes. Close collaboration between internists, cardiologists, cardiac surgeons and rheumatologists remains the cornerstone of optimal care. Future research must prioritise prospective cohorts to delineate diagnostic algorithms, prognostic indices, and personalised treatment paradigms.

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