International Journal of Research in Medical Sciences
Sharma RY et al. Int J Res Med Sci. 2025 Nov;13(11):5057-5066
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012
DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20253649

Review Article

The hidden heart of autoimmunity: a scoping review of carditis in rheumatoid arthritis and IgG4-related disease

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Received: 13 August 2025
Revised: 17 September 2025
Accepted: 14 October 2025

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ABSTRACT

Cardiac involvement in autoimmune diseases is often more common and more serious than we tend to acknowledge. While most clinicians are familiar with the joint pain of rheumatoid arthritis or the organ swelling of IgG4-related disease, inflammation of the heart itself is frequently missed. This is partly because cardiac symptoms are subtle or mistaken for something else, and partly because cardiovascular issues simply aren't top of mind when managing autoimmune patients. As a result, carditis whether it's myocarditis, pericarditis, or pancarditis remains underdiagnosed, underreported, and under-discussed, especially in the context of rarer conditions. This gap in recognition is particularly noticeable in IgG4-related disease, a condition known for causing fibrotic masses and multiorgan inflammation, but where cardiac involvement is poorly documented and often misclassified. Even in rheumatoid arthritis (RA), a disease much more common and much better studied, direct inflammation of the heart is often overshadowed by more visible symptoms like synovitis or systemic fatigue. Yet, evidence suggests that cardiac complications significantly contribute to both morbidity and mortality in autoimmune patients, often silently progressing until they become life-threatening. This scoping review sets out to bring more attention to this overlooked aspect of autoimmune disease. We aim to explore and organise the current literature on carditis in RA and IgG4-related disease, looking closely at how it presents, how it's diagnosed, what mechanisms might be driving it, and how it's treated. We've taken a wide lens to capture everything from well-documented case reports and cohort studies to emerging data on biomarkers and imaging techniques. In the end, this review is not just about summarising the science. It's about bringing the heart back into the conversation, literally and figuratively when we talk about autoimmune diseases. Because if we want to improve outcomes for these patients, we need to start looking beyond the obvious and paying closer attention to what's happening inside the chest.

Keywords: Carditis, Rheumatoid arthritis, IgG4-related disease, Autoimmune carditis, Myocarditis, Pericarditis, Endocarditis, Autoimmune disorders

INTRODUCTION

Autoimmune diseases are known for their wide-ranging effects on the body. While joint pain, fatigue, or skin rashes often get the most attention, these conditions can quietly impact internal organs, including the heart. When the heart becomes inflamed due to autoimmune activity, it's referred to as "carditis," and it can involve the heart muscle (myocarditis), outer lining (pericarditis), inner lining and valves (endocarditis), or the electrical conduction system, which controls heart rhythm.[1,2] These complications can be serious, even life-threatening yet they are often overlooked in both research and clinical practice, especially in diseases where cardiac symptoms aren't front and center.[3]

Systemic lupus erythematosus (SLE) is one autoimmune condition where heart involvement is well-recognized,

often affecting all layers of the heart (pancarditis).[4] However, in other diseases like rheumatoid arthritis (RA) and IgG4-related disease (IgG4-RD), cardiac inflammation is still underappreciated. This is surprising given that RA is common and well-known for causing systemic inflammation, while IgG4-RD, though rare, is increasingly being found to affect multiple organs, including the cardiovascular system.[5,6]

RA is typically known for causing joint problems, but it's also a systemic disease that can impact organs like the lungs, eyes, and heart.[6,7] Patients with RA are already at higher risk of cardiovascular disease due to chronic systemic inflammation, but direct inflammation of heart tissues, such as myocarditis or pericarditis, often goes undetected. This is partly because such symptoms may be mild, vague, or mistaken for other issues. Subtle signs like fatigue or shortness of breath may not be immediately linked to cardiac inflammation, especially if joint symptoms are more pressing.[8,9]

IgG4-RD, on the other hand, is a newer and less common autoimmune disease that behaves quite differently from RA. It often causes mass-like growths or swelling in affected organs and is typically treated with steroids or other immunosuppressants.[10] Although heart involvement is rare in IgG4-RD, when it does occur, it can be severe. Inflammation may affect the walls of large blood vessels, such as the aorta or even mimic tumors around the heart, which can lead to misdiagnosis or delay in care.[11,12]

Detecting carditis in these conditions isn't always easy. Traditional tools like echocardiograms (ECG) may not pick up early inflammation, and advanced imaging or even biopsies may be needed to confirm diagnosis.[13] Treatment usually targets the underlying autoimmune disease, whether through disease-modifying antirheumatic drugs (DMARDs) in RA or steroids and rituximab in IgG4-RD but it's still unclear how effective these are for treating heart-specific complications.[14]

This review aims to bring attention to the often-overlooked issue of carditis in RA and IgG4-RD. By exploring how these conditions affect the heart, how we currently diagnose and treat such involvement, and where the knowledge gaps lie, we hope to encourage greater awareness, earlier recognition, and better care for patients facing this hidden complication of autoimmune disease.

METHODS

This scoping review was conducted in accordance with the preferred reporting items for systematic reviews and meta-analyses extension for scoping reviews (PRISMA-ScR) guidelines. The protocol was designed to map the extent, range, and nature of available literature on cardiac involvement, specifically carditis in RA and IgG4-related disease (IgG4-RD) (Table 1). The inclusion and exclusion criteria are presented in Table 2.

Table 1: Search strategies according to respective databases.

Database	Search terms used
PubMed	("Carditis" OR "myocarditis" OR "pericarditis" OR "endocarditis") AND ("rheumatoid arthritis" OR "IgG4-related disease") AND ("cardiac involvement" OR "heart inflammation")
Embase	('Carditis' OR 'myocarditis' OR 'pericarditis' OR 'endocarditis') AND ('rheumatoid arthritis' OR 'IgG4-related disease') AND ('cardiac involvement' OR 'heart inflammation')
Scopus	TITLE-ABS-KEY(carditis OR myocarditis OR pericarditis OR endocarditis) AND TITLE-ABS-KEY ("rheumatoid arthritis" OR "IgG4-related disease") AND TITLE-ABS-KEY("cardiac involvement")

Table 2: Inclusion and exclusion criteria.

Inclusion criteria	Exclusion criteria
Human studies on cardiac involvement (myocarditis, pericarditis, endocarditis) in RA or IgG4-RD	Animal/*in vitro* studies
Original research, case reports, reviews, clinical trials	Articles without cardiac findings
English language publications	Non-English publications
Any publication year up to May 2025	Abstracts, editorials, and opinion pieces without data

CARDITIS IN RHEUMATOID ARTHRITIS

Cardiac involvement in RA has been recognized for decades. As far back as the 1950s, doctors began to suspect that RA could affect the heart, even though early attention was mostly focused on joint damage. It wasn't until the 1960s that autopsy studies provided clear evidence, almost half of the patients who died with RA showed signs of pericarditis, even though many had no symptoms while alive.[1] That finding was a turning point. It suggested that heart inflammation in RA could be widespread but hidden, flying under the radar during life.

With time, echocardiography helped confirm that pericardial involvement wasn't just a finding at autopsy, it could be picked up in living patients too.[2] Still, clinically obvious pericarditis (with symptoms like chest pain or friction rub) appears in only about 10% of people with

RA.[3] In contrast, myocarditis and endocarditis are less common, but they tend to be more serious when they occur.[4]

One striking feature of RA is the formation of rheumatoid nodules. These are small lumps made of inflamed tissue. While they're most often found under the skin, they can also develop inside the heart, on the myocardium, endocardium, or even the valves.[5] This is especially tricky, because these nodules can look just like infective endocarditis. So, if a patient with RA presents with a new murmur and negative blood cultures, the real cause might be autoimmune, not infectious.

In the last two decades, imaging tools have become much more sensitive. Techniques like cardiac MRI and advanced echocardiographic strain imaging now let us see subtle signs of heart involvement before symptoms appear. These methods have uncovered a pattern: even in RA patients who don't have known heart disease, there's often subclinical myocardial inflammation, diffuse fibrosis, or early ventricular dysfunction, especially in those who are seropositive (positive for rheumatoid factor or anti-CCP antibodies).[6,7]

At the molecular level, inflammatory cytokines play a key role. Tumor necrosis factor-alpha (TNF-α) and interleukin-6 (IL-6) are especially important. These molecules are not only involved in joint destruction, but also contribute to changes in the heart muscle. Chronic inflammation triggers remodeling and fibrosis in the myocardium.[8] IL-6, in particular, has been linked to endothelial dysfunction, the early damage to blood vessels that can lead to atherosclerosis, and has also been shown to increase myocardial strain, even in the early stages of RA.[9]

Putting it all together, this inflammation-driven damage adds up over time. In 2010, a major cohort study by Giles and colleagues showed that people with RA had nearly double the risk of developing heart failure compared to those without RA, even when controlling for traditional cardiovascular risk factors like hypertension, diabetes, and smoking.[10] That means RA itself is a powerful, independent risk factor for heart disease (Table 3). So, while RA is still largely seen as a disease of the joints, its reach clearly extends far beyond. The heart is a silent but vulnerable target, and often, by the time symptoms appear, damage is already well underway.

Table 3: Risk factors and prognostic indicators in autoimmune carditis.

Risk factor	Disease	Prognostic value	Comments
Seropositivity (RF, Anti-CCP)	RA	Higher risk of cardiac involvement	Linked to more severe systemic inflammation
Elevated serum IgG4	IgG4-RD	Supports diagnosis; relapse predictor	Not always elevated; needs biopsy confirmation
High CRP and ESR	RA, IgG4-RD	Active inflammation, worse prognosis	Correlates with disease activity
Prolonged disease duration	RA	Increased risk of fibrosis and complications	Late-stage disease often shows irreversible damage
Subclinical myocardial changes (strain imaging)	RA	Early marker of dysfunction	May guide early intervention
Delayed treatment initiation	IgG4-RD	Increased risk of aneurysm, rupture	Early immunosuppression improves outcomes
Presence of vascular involvement	IgG4-RD	Poorer prognosis, risk of occlusion	Requires aggressive monitoring and management

CARDITIS IN IgG4-RELATED DISEASE

Immunoglobulin G4-related disease (IgG4-RD) is a relatively new entry in the landscape of systemic autoimmune disorders. It was first recognized in the early 2000s, initially in the context of autoimmune pancreatitis, but has since expanded into a well-defined multisystem fibroinflammatory condition.[11] The disease is characterized by dense lymphoplasmacytic infiltration, a high proportion of IgG4-positive plasma cells, storiform fibrosis, and often elevated serum IgG4 levels. Over time, it has become clear that nearly any organ system can be involved, including the heart, although cardiac manifestations remain rare and under-recognized.

Early descriptions of cardiac involvement were mostly incidental, but as awareness has grown, more cases have been reported in the literature. One of the earliest and most notable accounts came from Kasashima et al, who documented IgG4-related coronary periarteritis, marking the beginning of formal recognition of cardiovascular manifestations in IgG4-RD.[12] Since then, a growing number of case reports and small series have expanded the spectrum of known presentations.

Cardiac involvement can manifest in several ways: such as periaortitis and periaortitis-related aneurysms, often involving the abdominal or thoracic aorta, with extension toward coronary ostia, coronary arteritis or periarteritis, which can lead to luminal narrowing or aneurysmal changes, myocardial pseudotumors, where mass-like

lesions mimic malignancy or infiltrative cardiomyopathies, and valvular thickening or fibrosis, which can resemble infective endocarditis or degenerative valvular disease.[13,14]

A 2018 systematic review by Perugino et al remains one of the most comprehensive overviews to date. It analyzed 60 published cases of cardiovascular involvement in IgG4-RD, confirming that although rare, such involvement is clinically significant. Most patients presented with symptoms related to mass effect or ischemia, and the majority responded well to glucocorticoid therapy. However, a subset required steroid-sparing agents or biologics such as rituximab, particularly in relapsing or refractory cases.[15]

The underlying pathophysiology is still not fully understood, but histopathologic analysis shows a clear pattern: infiltration of vascular structures by IgG4-positive plasma cells, accompanied by fibrosis and inflammatory thickening of the vessel walls.[16] This chronic inflammation leads to progressive wall stiffening, luminal stenosis, and, in some cases, aneurysm formation or rupture. Unlike classic vasculitis, which is often necrotizing, IgG4-RD tends to show more subtle chronic changes, but with equally serious outcomes if left untreated.

One of the greatest clinical challenges is diagnostic delay. Because IgG4-RD often mimics malignancy, infection, or atherosclerosis, patients are frequently misdiagnosed or diagnosed late, sometimes after significant cardiac damage has occurred. Cardiac imaging modalities such as computed tomography (CT) angiography, magnetic resonance imaging (MRI), and PET-CT can help in assessing the extent and nature of the lesions, but definitive diagnosis still relies on tissue biopsy demonstrating IgG4+ plasma cells and the classic histologic features.

Early recognition is critical. Untreated cardiovascular IgG4-RD can lead to life-threatening complications such as aortic aneurysm rupture, coronary artery occlusion, constrictive pericarditis, or heart failure.[17] Fortunately, with appropriate immunosuppressive treatment, many of these changes can stabilize or even reverse, if caught early.

CASE SUMMARIES: RA CARDITIS

Case 1: Seropositive RA with perimyocardial involvement

A 57-year-old woman with a 15-year history of seropositive rheumatoid arthritis (positive for both rheumatoid factor and anti-CCP antibodies) came to the clinic complaining of intermittent palpitations and vague chest discomfort that had been worsening over a few weeks. She denied fever or respiratory symptoms, but described fatigue and mild exertional dyspnea.[4]

On examination, her pulse was irregularly irregular, and her heart rate was elevated. ECG confirmed atrial fibrillation with a rapid ventricular response. Routine labs were unremarkable except for mildly elevated inflammatory markers (ESR and CRP). Transthoracic echocardiography showed a small pericardial effusion, but no tamponade or major structural abnormalities. To further investigate her chest symptoms, a cardiac MRI was ordered. It revealed patchy myocardial edema and early signs of myocardial fibrosis, suggestive of inflammatory myocarditis in the setting of active RA.

She was hospitalized briefly for rate control and started on beta-blockers. Rheumatology was consulted, and her immunosuppressive regimen was escalated. High-dose corticosteroids were initiated, and her methotrexate dose was increased. Over the following weeks, her palpitations resolved, inflammatory markers normalized, and a follow-up MRI three months later showed resolution of myocardial edema. She remained in sinus rhythm with no recurrence of cardiac symptoms.

This case highlights how subclinical cardiac inflammation in RA can present with arrhythmias and subtle imaging findings, and how timely immunosuppression can reverse early cardiac changes.

Case 2: Rheumatoid endocarditis mimicking infection

A 62-year-old man with poorly controlled RA (on intermittent NSAIDs but non-adherent to DMARDs) presented to the emergency department with low-grade fever, night sweats, and a new systolic murmur. He had no history of rheumatic fever or known valvular disease. Clinically, he appeared stable but fatigued, with mild anemia and raised inflammatory markers.[5]

Given the presence of fever and murmur, infective endocarditis was suspected. However, three sets of blood cultures were repeatedly negative. Transthoracic echocardiography followed by transesophageal echo revealed small, mobile vegetations on the aortic valve, but no abscess or major regurgitation. The team proceeded with surgical excision due to concern for embolic risk and uncertain etiology.

Histopathological examination of the excised valve revealed a granulomatous lesion with central necrosis, consistent with a rheumatoid nodule. There were no microorganisms seen on gram stain, culture, or PCR. The diagnosis was revised to rheumatoid endocarditis. He was started on leflunomide along with corticosteroids, and his RA disease activity was aggressively managed going forward. Over time, his systemic symptoms resolved, and inflammatory markers fell significantly. This case underscores how RA can mimic infective endocarditis, especially when nodules form on heart valves. Negative cultures and atypical histology can point toward an autoimmune rather than infectious cause. Early recognition can help avoid unnecessary prolonged antibiotics or delayed immunosuppressive therapy.

Case 3: Subclinical myocardial dysfunction detected by strain imaging

A 49-year-old woman with early, seropositive RA, diagnosed only 18 months ago was referred for cardiac evaluation before starting biologic therapy. She had no history of chest pain, dyspnea, or cardiovascular disease. Her physical exam and baseline ECG were unremarkable.[6,7]

Routine echocardiography showed normal ejection fraction and chamber dimensions. However, given her elevated inflammatory markers and persistent joint activity, speckle-tracking echocardiography was performed. This revealed reduced global longitudinal strain, despite a preserved ejection fraction, an early indicator of subclinical left ventricular dysfunction.

Cardiac MRI was subsequently done and showed areas of diffuse myocardial fibrosis without edema. The findings pointed toward ongoing low-grade myocardial inflammation due to systemic RA activity.

Given the early detection, her treatment was escalated to include anti-IL-6 therapy (tocilizumab), and methotrexate was continued. She remained asymptomatic, and follow-up strain imaging six months later showed improved myocardial strain and no further decline in function.

This case shows how RA-related cardiac damage can be silent, but detectable with sensitive tools, making a case for routine cardiac surveillance in high-risk patients, even without overt symptoms.

Case 4: Constrictive pericarditis in longstanding RA

A 65-year-old man with a 20-year history of seropositive RA presented with progressive shortness of breath, abdominal fullness, and lower limb swelling over the past few months. On exam, he had elevated jugular venous pressure, hepatomegaly, ascites, and peripheral edema. Heart sounds were muffled, and a pericardial knock was audible.[8,9]

Echocardiography revealed a thickened pericardium with biatrial enlargement and respiratory variation in mitral inflow, findings suggestive of constrictive pericarditis. CT chest confirmed pericardial thickening with calcification.

He had a history of recurrent pericardial effusions over the years, managed intermittently with steroids. This time, pericardiectomy was advised due to ongoing hemodynamic compromise.

Histology of the excised pericardium showed chronic inflammation and fibrosis, consistent with chronic RA-associated pericarditis.

Post-operatively, his symptoms improved significantly, and his disease-modifying therapy was optimized with a TNF-α inhibitor and low-dose prednisone.

This case highlights a late complication of untreated or poorly controlled pericardial inflammation in RA, where repeated episodes lead to fibrotic remodeling and constriction, a rare but serious cause of heart failure.

CASE SUMMARIES: IgG4 RELATED CARDITIS

Case 1: Coronary arteritis presenting as exertional angina

A 65-year-old man with no significant cardiac history presented with gradually worsening exertional chest pain over several months. His physical exam was unremarkable except for mild hypertension. Cardiac enzymes showed mildly elevated troponin levels, raising concern for ischemia. A CT coronary angiogram was performed and revealed diffuse, concentric thickening of the coronary artery walls without typical atherosclerotic plaques. Given the unusual imaging findings, a biopsy of the affected coronary artery was obtained during a catheterization procedure.

Histopathology demonstrated dense lymphoplasmacytic infiltration with abundant IgG4-positive plasma cells and storiform fibrosis, confirming IgG4-related coronary arteritis. He was started on high-dose prednisone with gradual tapering, alongside rituximab infusions as a steroid-sparing agent. Follow-up imaging three months later showed near-complete resolution of the arterial thickening, and his angina symptoms resolved.[15]

Case 2: Pericarditis and pericardial effusion leading to heart failure symptom

A 70-year-old woman presented with progressive shortness of breath and fatigue over 2 months. She reported no chest pain but described swelling in her legs and abdominal fullness. A physical exam revealed elevated jugular venous pressure, muffled heart sounds, and peripheral edema. Echocardiography showed a large pericardial effusion with signs of early tamponade and thickened pericardium.

Cardiac MRI confirmed pericardial thickening with inflammation. Serum IgG4 levels were significantly elevated. A biopsy of an enlarged cervical lymph node demonstrated characteristic IgG4-RD features, lymphoplasmacytic infiltrate with storiform fibrosis and abundant IgG4+ plasma cells. She was started on high-dose glucocorticoids, which rapidly improved her symptoms and reduced pericardial inflammation on follow-up imaging. She made a full clinical recovery.[16,17]

Case 3: Myocardial pseudotumor mimicking malignancy

A 58-year-old man with vague chest discomfort was found to have a mass in the left ventricular myocardium on echocardiography during evaluation for palpitations. Cardiac MRI and PET-CT revealed a well-demarcated myocardial lesion with mild FDG uptake, suspicious for a tumor. Biopsy was performed, and histopathology showed dense IgG4-positive plasma cell infiltration and fibrosis consistent with IgG4-RD pseudotumor. Immuno-suppressive treatment with steroids and rituximab resulted in significant reduction in mass size and symptom relief.[19]

Case 4: Aortic periaortitis with aneurysm formation

A 72-year-old man presented with abdominal pain and a pulsatile mass. Imaging revealed a large abdominal aortic aneurysm surrounded by an inflammatory thickened periaortic cuff. Serum IgG4 was elevated.

Biopsy of periaortic tissue confirmed IgG4-RD. The patient was treated with corticosteroids and underwent successful endovascular aneurysm repair. Post-operative follow-up showed stable aneurysm size and decreased inflammation.[20]

Case 5: Valvular involvement causing aortic regurgitation

A 60-year-old woman presented with worsening exertional dyspnea and a new diastolic murmur. Echocardiography revealed severe aortic regurgitation with thickened aortic valve leaflets. Infective endocarditis was initially suspected but blood cultures were negative. Valve replacement surgery was performed, and histology showed dense IgG4+ plasma cell infiltrate and fibrosis consistent with IgG4-RD. Postoperatively, she was treated with steroids and showed marked clinical improvement (Table 4).[21]

DIAGNOSTIC MODALITIES

The diagnosis of cardiac involvement in autoimmune diseases like RA and IgG4-RD hinges on a thoughtful combination of clinical suspicion, imaging, serology, and in select cases, tissue biopsy.

Echocardiography

First-line diagnostic tool for suspected carditis, particularly in rheumatoid arthritis. It is non-invasive, widely available, and provides real-time information about pericardial effusions, valvular vegetations, chamber size, and wall motion abnormalities.[19] In RA, it can detect pericarditis, subclinical valvular involvement, and even early features of heart failure due to diastolic dysfunction. Transesophageal echocardiography may offer better sensitivity for detecting vegetations and small valve lesions in ambiguous cases.

Cardiac magnetic resonance imaging (MRI)

For myocardial tissue characterization, cardiac MRI has emerged as the gold standard. It can identify myocardial edema, hyperemia, fibrosis, and infiltration, key hallmarks of myocarditis or myocardial involvement in systemic autoimmune conditions.[20]

In RA, CMR can detect subclinical myocardial inflammation in patients without overt symptoms. In IgG4-RD, it's useful when myocardial masses or pseudotumors are suspected, distinguishing them from malignancy.

CT angiography and PET-CT

In IgG4-related disease, particularly with vascular involvement, cross-sectional vascular imaging is essential. CT angiography provides detailed views of the aorta, coronary arteries, and peripheral vessels, detecting arterial wall thickening, periarteritis, and aneurysm formation.[21] PET-CT adds functional imaging, highlighting metabolically active inflammation making it especially valuable for: assessing the extent of systemic involvement, guiding biopsy from active lesions, monitoring treatment response and detecting relapse.[22,23]

Endomyocardial biopsy

Although invasive, endomyocardial biopsy remains the definitive method for diagnosing myocarditis. It allows for direct visualization of inflammatory infiltrates, necrosis, and fibrosis. In IgG4-RD, it can confirm plasma cell-rich infiltration with IgG4-positive staining, although tissue is often obtained from extracardiac sites. Because of procedural risks and patchy involvement, biopsy is typically reserved for cases where non-invasive imaging is inconclusive or atypical.[24]

Serologic and immunologic testing

In RA, elevated CRP, ESR, rheumatoid factor (RF), and anti-CCP antibodies help confirm disease activity and systemic inflammation.[25] In IgG4-RD, serum IgG4 levels may be elevated in up to 70% of cases. However, normal IgG4 does not rule out the disease. Autoantibody profiles, including ANA and others, can help in excluding overlap syndromes and may have prognostic value in chronic disease progression (Table 5).[26]

TREATMENT APPROACHES

Managing cardiac involvement in RA and IgG4-RD requires timely immunosuppression, control of systemic inflammation, and supportive cardiac care. The goals are to reduce acute inflammation, prevent permanent fibrosis, and minimize cardiovascular morbidity.

Rheumatoid arthritis

Disease-modifying antirheumatic drugs

Methotrexate is the cornerstone of RA treatment and has been shown to reduce systemic inflammation and long-term cardiovascular risk.[27]

Biologic agents

TNF-α inhibitors (e.g., infliximab, etanercept) and IL-6 inhibitors (e.g., tocilizumab) not only improve joint symptoms but also appear to lower the risk of heart failure, pericarditis, and vascular events by dampening the cytokine cascade.[28]

Table 4: Summary table of case reports.[4-9,15-21]

Disease	Patient age/sex	Cardiac manifestation	Diagnostic modality	Treatment	Outcome
RA[4]	57/F	Pericarditis/myocarditis	MRI, ECG	Steroids + methotrexate	Resolved
RA[5]	62/M	Rheumatoid endocarditis	Echocardiography, biopsy	Steroids + DMARD	Improved
RA[6,7]	49/F	Subclinical myocardial dysfunction	Echocardiography strain, MRI	Anti-IL-6 therapy + MTX	Stable/ improved
RA[8,9]	65/M	Constrictive pericarditis	Echocardiography, CT	Pericardiectomy + TNF inhibitors + Steroids	Improved
IgG4-RD[15,16]	65/M	Coronary arteritis	CT angiography, biopsy	Steroids + rituximab	Improved
IgG4-RD[17,18]	70/F	Pericarditis with effusion	MRI, serology, biopsy	Steroids	Resolved
IgG4-RD[19]	58/M	Myocardial pseudotumor	MRI, PET-CT, biopsy	Steroids + Rituximab	Improved
IgG4-RD[20]	72/M	Aortic periaortitis with Aneurysm	CT angiography, biopsy	Steroids + surgery	Stable
IgG4-RD[21]	60/F	Valvular involvement (AR)	Echocardiography, histology	Valve replacement + steroids	Recovered

Table 5: Serological and imaging modalities: sensitivity and usefulness.

Modality/test	Disease focus	Diagnostic role	Notes
Rheumatoid factor (RF)	RA	Marker of disease activity	Not specific, but prognostic
Anti-CCP antibody	RA	More specific for RA diagnosis	Correlates with severity
Serum IgG4	IgG4-RD	Supports diagnosis	Normal levels do not exclude disease
C-reactive protein (CRP)	RA, IgG4-RD	Marker of systemic inflammation	Elevated in active disease
Erythrocyte sedimentation rate (ESR)	RA, IgG4-RD	Marker of inflammation	Less specific, often elevated
Echocardiography	RA, IgG4-RD	Detects effusions, vegetations, valvular disease	First-line, non-invasive
Cardiac MRI	RA, IgG4-RD	Tissue characterization (edema, fibrosis)	High sensitivity for myocarditis
CT angiography	IgG4-RD	Visualizes vascular wall thickening, aneurysms	Useful for periaortitis, coronary arteries
PET-CT	IgG4-RD	Detects active inflammation systemically	Guides biopsy and monitors relapse
Endomyocardial biopsy	RA, IgG4-RD	Gold standard for myocarditis diagnosis	Invasive; used selectively

Acute cardiac involvement

In cases of pericarditis or myocarditis, high-dose corticosteroids are often required. In severe or steroid-resistant cases, escalation to biologics may be considered.[29]

IgG4-related disease

Corticosteroids

Glucocorticoids remain the first-line treatment, with most patients showing rapid clinical and radiologic

improvement. Typically, prednisone is started at 30–40 mg/day and tapered over months.[30]

Steroid-sparing agents/biologics

Rituximab, a B-cell depleting monoclonal antibody, has proven especially effective in: patients with refractory or relapsing disease, those with contraindications to prolonged steroid use, and cases involving critical organs like the heart or major vessels.[31,32]

Supportive and symptomatic management

NSAIDs for mild pericarditis (if renal function is preserved), anti-arrhythmic drugs or anticoagulation for atrial fibrillation or conduction defects, heart failure therapy (ACE inhibitors, beta-blockers, diuretics) for myocardial dysfunction, surgical or interventional procedures, such as pericardiectomy or valve replacement, in advanced cases.

Long-term monitoring and collaborative care

Cardiac complications in RA and IgG4-RD can recur or evolve silently. Serial imaging, biomarker monitoring, and clinical follow-up are key. Collaborative management between rheumatologists and cardiologists ensures that immunosuppressive therapy is balanced with cardiovascular risk reduction.[33] Patients should also receive standard cardiovascular prevention, including lipid management, BP control, and lifestyle counselling.

RESEARCH GAPS AND FUTURE DIRECTIONS

Despite growing recognition, our understanding of carditis in RA and IgG4-RD remains limited. Most of the current evidence comes from isolated case reports, small case series, and retrospective reviews with inherent selection bias and incomplete follow-up data.[34] As a result, the true prevalence, natural history, and long-term outcomes of cardiac involvement in these disorders are still unclear.

There's a pressing need for large, multicenter prospective studies to answer some fundamental questions like how common is carditis across the disease spectrum? what are the strongest predictors of cardiac involvement? and what role do serological markers, disease duration, and treatment history play in prognosis?

From a biological standpoint, the underlying immunopathogenesis is still not fully understood. In RA, the contribution of pro-inflammatory cytokines like TNF-α, IL-1, and IL-6 to myocardial and pericardial inflammation is well documented, but how these interact with cardiac-specific antigens or tissues is less clear.[35] In IgG4-RD, the role of IgG4+ plasma cells, regulatory T cells, and fibrogenic pathways within vascular and myocardial tissue needs deeper exploration. Understanding these mechanisms could open the door to novel therapies beyond broad immunosuppression.

Technological advances offer new possibilities. Cardiac MRI, PET-CT, and CT angiography are already improving diagnostic accuracy, but machine learning and AI-based imaging interpretation could enhance early detection and prognostication. Automated recognition of subtle patterns such as myocardial edema or vascular wall thickening may help flag high-risk patients even before clinical symptoms emerge.

Lastly, there's a real need for consensus guidelines or diagnostic algorithms specific to cardiac involvement in autoimmune diseases. Right now, management often depends on individual clinician experience or extrapolation from general cardiology or rheumatology practices. Clear, evidence-based pathways could improve consistency in diagnosis, treatment, and follow-up, especially in resource-limited settings or in rare diseases like IgG4-RD.

CONCLUSION

Carditis in RA and IgG4-RD represents a rare but clinically significant aspect of autoimmune pathology that is often underrecognized in everyday practice. Although less frequently reported compared to other systemic manifestations, cardiac involvement in these diseases can lead to serious complications, including arrhythmias, heart failure, or even sudden cardiac death. Early detection remains a major challenge, as symptoms are often nonspecific or silent, and diagnostic modalities may miss subtle changes unless suspicion is high. Nonetheless, timely diagnosis and appropriate treatment can significantly alter the disease trajectory and improve outcomes. This scoping review emphasizes the importance of raising clinical awareness regarding cardiac manifestations in RA and IgG4-RD. Enhanced vigilance by rheumatologists, cardiologists, radiologists, and primary care physicians is essential to ensure early identification and management. A multidisciplinary approach, involving cross-specialty collaboration, can facilitate accurate diagnosis and tailored therapy, especially in complex or atypical cases. There is a clear need for larger studies and longitudinal data to better understand the prevalence, mechanisms, and best treatment strategies for carditis in these conditions. By addressing these gaps, we can move toward more proactive, evidence-based care and ultimately improve the quality of life and prognosis for patients with autoimmune-related cardiac involvement.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Sharma RY, Modi KT, Patel MS, Patel KE, Memon SA. The hidden heart of autoimmunity: a scoping review of carditis in rheumatoid arthritis and IgG4-related disease. Int J Res Med Sci 2025;13:5057-66.