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Pericarditis in Patients with Autoimmune Disease: Insights into Prevalence and Optimal Management

REVIEW

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ABSTRACT

Initially recognized as a key cardiac feature of systemic lupus erythematosus and rheumatoid arthritis, autoimmune pericarditis has gained increasing attention given the recent advances in cardiac imaging, biomarker assessment, and understanding of immune-mediated mechanisms. The prevalence of pericardial involvement varies considerably among autoimmune diseases. Patients may have a small pericardial effusion that remains clinically silent but could also present with acute, recurrent, or chronic pericarditis. A minority develop severe complications such as tamponade or constrictive physiology. Characterizing autoimmune pericarditis is critical, particularly in an era of expanding immunomodulatory therapies. This review summarizes current knowledge on prevalence, pathophysiology, clinical presentation, and diagnostic strategies, with a particular focus on emerging therapeutics in pericardial disease associated with autoimmune disorders. Integrating rheumatology and cardiology expertise is essential to optimize the care of this heterogeneous patient population.

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INTRODUCTION

About one in ten people are affected by an autoimmune disease, and the burden continues to grow.[1] Autoimmune pericarditis refers to pericardial inflammation occurring in the context of systemic autoimmune disease and represents a distinct phenotype within the broader spectrum of pericardial disorders. Unlike idiopathic, post-surgical, or infectious etiologies, autoimmune pericarditis is driven by dysregulated immune mechanisms linked to the underlying systemic condition.[2,3]

Pericardial involvement has long been recognized in autoimmune disease. Initial descriptions date back to the late 1800s, when Charcot reported pericardial pathology in rheumatoid arthritis (RA).[3] Since then, pericarditis has been described in a wide range of autoimmune disorders, including systemic lupus erythematosus (SLE), RA, systemic sclerosis (SSc), Sjögren's syndrome, mixed connective tissue disease (MCTD), various vasculitides, sarcoidosis, inflammatory bowel disease, and autoimmune thyroid disease.[3] Autoinflammatory syndromes such as Familial Mediterranean Fever and TNF-α (Tumor Necrosis Factor alpha) receptor-associated periodic syndrome (TRAPS) are also associated with recurrent pericarditis, particularly in the pediatric population.[4]

Despite its long historical recognition, autoimmune pericarditis remains less characterized than idiopathic or viral forms.[2] Its clinical presentation is highly heterogeneous, influenced by the type of underlying autoimmune disease, immune-mediated mechanisms, and variable responses to immunosuppression. Patients may develop acute pericarditis, recurrent or persistent inflammatory flares, chronic effusions, or, rarely, tamponade and constrictive pericarditis.[2,3,5] Symptoms frequently overlap with systemic autoimmune manifestations, further complicating timely diagnosis.[3]

Interest in autoimmune pericarditis has expanded in recent years, driven by rising rates of autoimmune disease, improved detection of subclinical pericardial involvement through advanced imaging, a clearer understanding of immune pathways, and the emergence of targeted immunomodulatory therapies. This review provides a contemporary overview of autoimmune-associated pericarditis, emphasizing its prevalence across autoimmune diseases and recent advances in management.

DISEASE SPECIFIC EPIDEMIOLOGY

Pericarditis is an inflammatory process affecting the pericardial sac and is widely prevalent across systemic autoimmune conditions. A rheumatic etiology is identified in approximately 2% to 7% of acute pericarditis cases and up to 10% of recurrent cases.[3]

SYSTEMIC LUPUS ERYTHEMATOSUS

Disease-specific epidemiological data suggests that pericardial involvement was present in around 50% of patients with SLE.[6] Specifically, pericarditis was one of the most common cardiac manifestations, affecting around 20% to 25% of SLE patients.[6,7] The Hopkins Lupus Cohort study showed that recurrence occurred in 20.3% of patients following an initial episode of pericarditis, with 49.2% experiencing multiple recurrences.[7]

The reported risk factors for an acute episode of SLE-associated pericarditis include the presence of autoantibodies such as anti-smooth muscle and anti–double-stranded DNA antibodies as well as proteinuria and hemolytic anemia.[8] Additionally, increased recurrence rates were associated with younger age, active disease, and prednisone use, whereas nephrotic syndrome, proteinuria, and pulmonary hypertension were associated with decreased recurrence rates.[7]

RHEUMATOID ARTHRITIS

Rheumatoid arthritis affects 1% of the population and manifests with numerous extra-articular findings including pericardial disease.[6] Between 30% and 50% of patients with RA develop pericardial disease, typically presenting as asymptomatic effusion.[9,10] Infrequently, RA patients may present with symptomatic pericarditis (< 10%), with cardiac tamponade and constrictive pericarditis described in a few cases.[6,11,12]

In patients with RA, severity of disease as well as higher levels of anti-cyclic citrullinated peptide (anti-CCP) antibodies and rheumatoid factor (RF) are positively associated with pericarditis.[13]

OTHER AUTOIMMUNE CONDITIONS

Pericarditis was also described in SSc, Sjögren's syndrome, vasculitis (eg, anti-neutrophil cytoplasmic antibodies-associated), and other connective tissue diseases. In SSc, pericardial involvement is often asymptomatic but was detected in autopsy series (70%).[3] The true prevalence in living patients is less clearly defined but thought to correlate with disease severity and pulmonary hypertension.[3] There is an estimated 8-fold increased risk of pericardial disease in SSc patients relative to the general population, with symptomatic effusions only present in 5% to 16%.[3]

In vasculitis, pericardial involvement is less common (1–3%) relative to other cardiac manifestations such as myocarditis and coronary or aortic disease.[3,14] The correlation between vasculitis and pericarditis may relate to the utilization of immunosuppressive therapy which

predisposes to viral infections and can consequently lead to secondary acute or recurrent pericarditis.[3]

AUTOINFLAMMATORY SYNDROMES

Pericarditis may occur in autoinflammatory syndromes, genetically mediated periodic fever disorders driven by dysregulated innate rather than adaptive immune mechanisms. Familial Mediterranean Fever and TRAPS are the most frequently described. Although disease-associated mutations are rare among patients with recurrent pericarditis, pericardial involvement has been reported in both conditions, often during systemic inflammatory flares.[15] Prevalence estimates vary widely and are largely based on clinically apparent cases, while episodic disease, pediatric predominance, small cohorts, and limited systematic cardiac imaging likely lead to underestimation of true burden. Collectively, these factors suggest that pericardial involvement in autoinflammatory syndromes, particularly subclinical or transient disease, remains epidemiologically underdefined.

Epidemiological Challenges

Epidemiologic characterization of pericardial involvement in autoimmune disease remains a challenge (Table 1). A substantial number of cases are subclinical, identified incidentally on echocardiography or cardiac magnetic resonance (CMR), leading to underestimation of the true prevalence.[3] Conversely, studies may have misestimated rates of autoimmune pericarditis due to inconsistent diagnostic criteria and nonsystematic screening. These limitations highlight the need for prospective multidisciplinary cohort studies, integrating rheumatology and cardiology expertise, to systematically screen patients with autoimmune disease, including asymptomatic patients, using standardized modern echocardiographic and CMR-based protocols.

IMMUNOPATHOGENESIS

Autoimmune pericarditis arises from a dysregulated immune response (Table 1). Mechanistically, it often follows a cascade that begins with antigen exposure or immune complex formation, proceeds through complement activation and recruitment of innate immune cells, and culminates in a cytokine-driven inflammatory reaction, notably IL-1 (Interleukin-1), IL-6 (Interleukin-6), IL-17 (Interleukin-17), and TNF-α, which together exacerbate pericardial inflammation and tissue injury.[3,16]

In addition, many recurrent and autoinflammatory pericarditis cases appear to be driven by an IL-1–focused inflammasome-mediated pathway. When the NLRP3 inflammasome is activated in pericardial macrophages or mesothelial cells, it triggers maturation and release of IL-1, which in turn promotes neutrophil recruitment, fever, and a broader inflammatory response.[3,17] More recently, large-scale human genomic data have linked variation at the IL-1 locus with pericarditis risk. This provides population-level support that IL-1 signaling is not just associated with the disease but is likely mechanistically important, thus reinforcing the rationale for IL-1 inhibition as a targeted therapy in select patient phenotypes.[17]

DISEASE-SPECIFIC MECHANISMS

Systemic Lupus Erythematosus

Pericarditis in SLE is largely driven by immune-complex deposition and complement activation, particularly complement component 3 (C3).[3,18] Immunopathologic studies revealed prominent deposition of immunoglobulin in pericardial tissue and myocardial vessels.[18] The degree of deposition was directly related to increased serological evidence of disease and clinical severity.[17]

Additionally, variable involvement of IL-1/IL-6/IL-17 pathways modify disease course.[3] A single-nucleotide polymorphism in TNF Receptor-Associated Factor 3 Interacting Protein 2 (TRAF3IP2)—which encodes NF-κB Activator 1 (Act1), a central adaptor in IL-17-mediated immune signaling—has been linked to an increased risk of pericarditis in SLE.[19] This shows that genetic predisposition plays a key role in risk stratification, offering a deeper understanding of susceptibility patterns.

Rheumatoid Arthritis

Early studies on rheumatoid pericarditis showed plasma cell involvement and immune deposition in pericardial vessels including IgG, IgM, IgA, and C3.[20] More recent case reports point to a proinflammatory cytokine-driven mechanism rather than classic immune-complex deposition. In one reported RA case complicated by tamponade, pericardial fluid showed elevated IL-6 levels, implicating IL-6 as a major driver of pericardial inflammation.[21] Moreover, chronic serosal inflammation in RA reflects high-circulating RF/anti-CCP titers.[3]

RA-related pericarditis likely reflects dual mechanisms: immune complex-mediated complement activation in some patients, and a dominant pro-inflammatory cytokine surge (IL-1, IL-6, TNF-α) during active disease in others.

Other Autoimmune Conditions

In SSc, pericarditis is characterized less by immune complex deposition and more by microvascular injury, ischemia, and progressive fibrosis of pericardial layers.[3] Moreover, macrophage activation and profibrotic cytokines contribute to pericardial thickening and restrictive physiology.[3] This suggests that pericarditis is part of a more global end-organ fibrotic process rather than a classic serositis.

AUTOIMMUNE CONDITION	ESTIMATED PREVALENCE OF PERICARDIAL INVOLVEMENT	TYPICAL PERICARDIAL MANIFESTATION	PATHOPHYSIOLOGIC DRIVER OF PERICARDIAL DISEASE	STANDARD MANAGEMENT	EMERGING FUTURE THERAPIES
Systemic Lupus Erythematosus	50% asymptomatic pericardial involvement; 20-25% pericarditis	Asymptomatic involvement more prevalent than symptomatic disease. Acute or recurrent pericarditis; often painless effusions; rare tamponade or constriction	Immune-complex deposition, complement activation and interleukin-dependent cytokine amplification pathways	1st line: NSAIDs + colchicine 2nd line: steroids 3rd line: azathioprine, mycophenolate mofetil, belimumab, and anifrolumab 4th line: IL-1 inhibitors such as rilonacept and anakinra, rituximab, cyclophosphamide, intravenous immunoglobulins	IL-1 inhibition (anakinra, rilonacept)
Rheumatoid Arthritis	30-50% pericardial involvement; < 10% symptomatic pericarditis	Asymptomatic effusions more prevalent than acute disease. Acute or recurrent pericarditis; rare tamponade or constriction	Interleukin-dependent cytokine amplification pathways ± immune-complex deposition	1st line: NSAIDs + colchicine 2nd line: steroids RA-directed DMARDs	IL-1 inhibition (anakinra, rilonacept); possible IL-6 inhibition
Systemic Sclerosis	Detected in autopsy series (70%); 8-fold increased risk of pericardial involvement with symptomatic effusions present in 5-16%	Small chronic effusions more common than symptomatic or acute disease	Microvascular injury, ischemia, and progressive fibrosis	1st line: NSAIDs + colchicine 2nd line: steroids, but reduced dose for concern for scleroderma renal crises 3rd line: mycopheonlate, cyclophosphamide, rituximab, tocilizumab	Anti-fibrotic and vascular-targeted therapies
Mixed Connective Tissue Disease	Variable; limited epidemiologic data	Variable	Overlapping autoimmune features (eg, anti-ribonucleoprotein antibodies, interferon)	1st line: NSAIDs + colchicine 2nd line: steroids 3rd line: mycophenolate, azathioprine, methotrexate 4th line: rituximab, cyclophosphamide, IVIG	IL-1 inhibition (anakinra, rilonacept)
Vasculitis	Uncommon overall; 1-3% pericardial involvement; myocarditis and valvular disease are more common	Inflammation of pericardial walls causing acute presentation	Less defined; can be cell or immune complex mediated	1st line: NSAIDs + colchicine 2nd line: steroids Treat underlying vasculitis	IL-1 inhibition (anakinra, rilonacept)
Autoinflammatory Conditions (eg, FMF, TRAPS)	Rare but recognized	Recurrent inflammatory pericarditis	Innate immune activation	1st line: colchicine for FMF, colchicine/NSAIDs/steroids for TRAPS	IL-1 inhibition (anakinra, rilonacept)

Table 1 Summary of key features of autoimmune pericarditis across the most common autoimmune conditions. NSAIDs: nonsteroidal anti-inflammatory drugs; RA: rheumatoid arthritis; DMARD: disease-modifying antirheumatic drug; IVIG: intravenous immunoglobulin; FMF: familial Mediterranean fever; TRAPS: tumor necrosis factor receptor-associated periodic syndrome

Mixed connective tissue disease demonstrates overlapping autoimmune features (anti-ribonucleoprotein antibodies, interferon) that can manifest with serositis.[3] In vasculitis, pericardial inflammation is less common than myocarditis or vascular complications.[3] When present, it likely reflects shared vascular inflammatory pathways.[3] Pericardial involvement in vasculitis is rare and poorly understood, with no single immunopathogenic pathway identified, underscoring the need for individualized evaluation using imaging, fluid analysis, and biopsy when indicated, particularly in cases of pericarditis arising during immunosuppression or with unexplained effusions.

DISEASE COURSE

Autoimmune pericarditis can present in many ways, from acute symptomatic episodes to recurrent or persistent flares, chronic low-grade effusions, life-threatening tamponade, and less commonly fibrosing constrictive pericarditis.[3] Its clinical course is influenced by the underlying autoimmune disease, the dominant immune pathway, whether autoantibody or autoinflammatory, and prior immunomodulatory treatments.[3] Some patients show mixed patterns, shifting over time, for example, acute flares driven by immune complexes alternating with IL-1 mediated relapses.[3]

Although acute pericarditis is widely observed in the context of systemic autoimmune disease, it rarely presents as the sole initial manifestation.[15,22-24] In SLE, pericardial involvement, particularly acute pericarditis, is rarely the first or only presenting feature, typically occurring after other characteristic manifestations such as rash, arthritis, cytopenias, or renal disease.[15,22-24] In most other autoimmune conditions, such as RA, Sjögren's syndrome, or SSc, clinically overt pericarditis is uncommon as a presenting feature and more often manifests as incidentally detected, small, asymptomatic effusions in advanced disease.[3,15,22-24]

Accordingly, broad autoimmune testing after a first, uncomplicated episode of typical acute pericarditis is usually low yield and may lead to false positives and unnecessary follow-up. A focused autoimmune evaluation is better reserved for patients with concerning features, including recurrent or treatment-resistant disease, steroid dependence, large effusions or tamponade, or accompanying characteristic systemic signs, including a classic rash, inflammatory arthritis, cytopenias, renal involvement, or sicca/Raynaud symptoms.[3,15,22-24]

RISK OF TAMPONADE AND CONSTRICTION

Contemporary registry data show that although many patients have only mild or subclinical effusions, a meaningful minority, particularly those with SLE or advanced RA, develop large effusions that require pericardiocentesis or surgical drainage.[25] Case reports of SLE highlight that tamponade can occasionally be the initial manifestation.[25] The risk of progression to constrictive pericarditis is nontrivial in autoimmune disease, yet it remains underexplored. Chronic inflammation, repeated flares, delayed diagnosis, and inadequate immunosuppression likely contribute to fibrotic remodeling, but further research is necessary to characterize this phenomenon.

DIAGNOSTIC APPROACH

In patients with suspected autoimmune pericarditis, diagnostic evaluation should be guided by clinical history and laboratory evidence of systemic inflammation, with targeted immunologic testing performed when an underlying autoimmune disease is suspected. Cardiac imaging is used to support the diagnosis, assess disease extent, and identify complications, not for first-line etiologic classification.[15] CRP and ESR remain the most practical markers for detecting active pericardial inflammation and for monitoring treatment response.[3,6] Elevated CRP indicates ongoing inflammation, and persistently high or rising levels despite therapy should raise concern for smoldering disease or early relapse.[3,5]

Serologic screening is recommended when clinical history or disease course suggests a systemic autoimmune etiology. Antinuclear antibodies is the preferred first-line test, followed by disease-specific antibodies based on the clinical scenario, including anti-double stranded DNA for SLE, anti-ribonucleoprotein for MCTD, anti-CCP and RF for RA, anti-SSA and anti-SSB for Sjögren's, anti-topoisomerase I antibodies and anti-RNA polymerase III antibodies for SSc.[3,6] A positive autoantibody profile supports an autoimmune etiology and helps guide long-term management.[3,6]

THE ROLE OF MULTIMODALITY IMAGING

Echocardiography remains the first-line modality because it is widely available, noninvasive, and effective for assessing effusion size, hemodynamic impact, and the need for drainage.[26] CMR is suitable to confirm pericardial inflammation in uncertain cases, and for follow-up and assessment of treatment response and remission (usually at baseline, then following 6-month intervals).[26] Computed tomography (CT) is most valuable for identifying pericardial thickening, calcification, and for preoperative planning in patients with prior cardiac surgery.[26] Compared with echocardiography and CT, CMR uniquely detects inflammation and fibrosis via the late gadolinium enhancement technique.[27]

Cardiac fluorodeoxyglucose positron emission tomography (FDG-PET) is not routinely recommended for the diagnosis of pericarditis and is limited by the need for specialized dietary preparation and practicality compared with CMR, but it may serve as an adjunctive tool in selected cases, particularly when assessment of active inflammation is needed and conventional imaging is inconclusive, or when systemic inflammatory disease is suspected.[26] Cardiac PET is useful in characterizing pericardial involvement in patients with lymphoma, where increased FDG uptake may correlate with disease burden and cancer staging.[15] It may also be useful in differentiating select cases of tuberculous pericarditis, as these typically demonstrate higher FDG uptake compared with idiopathic pericarditis.[15] However, the overall role of cardiac PET in autoimmune pericarditis remains limited, and further studies are needed to better define its diagnostic and prognostic utility.

Recent international recommendations underscore the importance of multimodality imaging for diagnostic assessment, prognostic evaluation and treatment planning, particularly when using IL-1 targeted therapies that rely on imaging evidence of active inflammation.[15,26]

GENERAL MANAGEMENT PRINCIPLES

Management of pericarditis in autoimmune conditions should be tailored to the underlying disease, symptom severity, and clinical/laboratory systemic disease activity (Table 1). Incidental or minimally symptomatic effusions often require no specific therapy beyond optimizing control of the underlying autoimmune condition.[6] Symptomatic pericarditis is generally treated first-line with nonsteroidal anti-inflammatory drugs (NSAIDs) and colchicine, as in idiopathic pericarditis, although trials supporting colchicine largely involved idiopathic pericarditis, with autoimmune cases representing only a small subset. In addition, efficacy of traditional regimens may be lower when systemic inflammation predominates, often prompting earlier steroid initiation and disease-specific considerations.[6]

Second-line therapy includes steroids, especially when NSAIDs/aspirin and colchicine combination therapy fails, in severe flares, or when accompanied by systemic disease activity such as in SLE or vasculitis.[3,6] Dosing of steroids in autoimmune-associated pericarditis is disease-specific. When steroids are used to primarily treat pericardial inflammation in the absence of significant concurrent systemic autoimmune activity, the usual regimen includes initiating oral prednisone at 0.2 mg/kg to 0.5 mg/kg daily for 2 to 4 weeks (for acute or recurrent episodes), followed by a gradual taper over 2 to 3 months.[3,6]

Higher steroid doses (1 mg/kg of prednisone or 1g/day for 3 days of pulse intravenous methylprednisolone) are reserved for pericarditis that occurs in the setting of active organ-threatening systemic autoimmune disease, most commonly SLE or systemic vasculitis, and are used to control systemic disease manifestations rather than purely isolated pericardial inflammation.[6] The steroid tapering schedule may be longer in such cases, as compared to idiopathic pericarditis, and close collaboration with rheumatology is essential.[6]

However, high-dose steroids are not always recommended in autoimmune conditions. In SSc, high-dose steroids are avoided due to the risk of precipitating scleroderma renal crises.[6] Prophylaxis for *Pneumocystis jirovecii* pneumonia should be considered if taking more than 20 mg or more daily for over 4 weeks.[28] Prolonged steroid therapy should also include bone protection with calcium and vitamin D, with consideration of bisphosphonate therapy based on fracture risk, and gastrointestinal prophylaxis with a proton pump inhibitor in high-risk patients, particularly those receiving concomitant NSAIDs or antiplatelet therapy.[29,30] Careful monitoring for side effects, along with regular follow-up with rheumatology, should be maintained.[6] Long-term steroid use raises recurrence risk in idiopathic acute and recurrent pericarditis, but the recurrence risk in autoimmune pericarditis remains uncertain.[3]

Immunosuppressive therapy with azathioprine, mycophenolate mofetil, methotrexate, rituximab, cyclophosphamide or intravenous immunoglobulins could be used in severe cases that are refractory to steroids and not suitable for IL-1 inhibitors.[3,6] In SLE, specific therapies such as belimumab and anifrolumab are also considered. Alternatively, IL-1 inhibitors such as rilonacept and anakinra are used in patients with multiple recurrences or refractory pericarditis.[3,6] Data on rilonacept in autoimmune-associated pericarditis are limited, and current use is largely extrapolated from studies in recurrent pericarditis. Nevertheless, rilonacept is increasingly used off-label in select cases of recurrent, inflammatory autoimmune pericarditis.

THE RISE OF IL-1 INHIBITORS

IL-1 inhibition has emerged as a major therapy for recurrent or refractory pericarditis, although evidence specific to autoimmune pericarditis is still limited.

Anakinra, an IL-1 receptor antagonist, rapidly reduced symptoms and decreased recurrences in colchicine-resistant or steroid-dependent patients with recurrent idiopathic pericarditis (Anakinra-Treatment of Recurrent Idiopathic Pericarditis, AIRTRIP trial), with injection-site reactions as the main side effect and a low risk of serious infections.[31] Small series in SLE suggest it can safely reduce flares when added to background immunosuppression. A notable case described a young man with SLE

whose pericarditis persisted despite treatment with multiple immunosuppressants including cyclophosphamide, mycophenolate, cyclosporine, belimumab, and colchicine. After beginning anakinra (100 mg/day) in combination with prednisone and colchicine, he achieved full remission and remained flare-free for at least 6 months while maintained on low-dose prednisone and anakinra.[32]

Rilonacept is another IL-1 blocker approved by the US Food and Drug Administration for treatment and prevention of recurrent pericarditis. The efficacy of rilonacept in autoimmune pericarditis remains uncertain since the RHAPSODY (Study to Assess the Efficacy and Safety of Rilonacept Treatment in Participants With Recurrent Pericarditis) trial excluded patients with autoimmune pericarditis.[33] In a case series involving four women with refractory SLE-associated pericarditis, two of whom relapsed after anakinra, rilonacept led to complete symptom resolution in three patients and partial improvement in one within 2 weeks.[34] Multicenter registry data and prospective safety/efficacy studies are needed in this patient population.

As previously discussed, autoimmune-associated pericarditis may result from IL-1 driven autoinflammatory pathways.[3,16,17] In this context, disease phenotyping may help guide therapy—particularly in recurrent or refractory cases with persistent inflammation despite adequate control of the underlying autoimmune disease. IL-1 inhibition may be appropriate in patients with frequent recurrences, steroid dependence, or colchicine resistance, especially when pericarditis appears to have activity independent of systemic disease activity or is the primary manifestation of the autoimmune disorder. Available case series suggest that IL-1 inhibitors can be used safely in conjunction with background immunosuppression, although high-quality prospective data in autoimmune cohorts are lacking.[32,34] In patients where recurrent pericarditis is the main manifestation of the disease process, achieving control of disease with IL-1 inhibition may be pursued, followed by gradual withdrawal of other disease-modifying agents, particularly to reduce the risk of infection or other side effects. However, these approaches have not been tested prospectively. Further studies are needed to define optimal patient selection, timing, and long-term safety of IL-1 inhibition in autoimmune-associated pericarditis.

DISEASE-SPECIFIC THERAPY

Systemic Lupus Erythematosus

Management of SLE-associated pericarditis depends on the severity of the presentation. Mild pericarditis without major organ involvement (renal/central nervous system) can be treated with a longer course of NSAIDs and colchicine due to a higher risk of recurrence.[6,35] Steroids play a key role in SLE-associated pericarditis, but dosing and duration should be minimized whenever possible to avoid side effects.[6] In refractory or recurrent cases, and in pregnant patients, azathioprine has the strongest evidence with a starting dose of 1 mg/kg/day, increased to 2 to 3 mg/kg/day.[3,6] Effect onset is slow (1–2 months), but > 50% achieve durable remission.[3,6] Mycophenolate mofetil is also used in recurrent refractory pericarditis at a starting dose of 500 mg twice daily and gradually increased to a maximum dose of 1000 mg to 1500 mg twice daily.[6] Also, intravenous immunoglobulins can be used in refractory cases when steroid-sparing is desirable.[6,36] The dose of 2 g/kg is given intravenously over the course of 3 to 5 days with caution in patients with a history of heart failure and thromboembolism.[6,36] Fourth-line agents such as cyclophosphamide and rituximab are typically reserved for organ-threatening lupus flares and may be considered in severe pericardial involvement.[6,37] Belimumab, an anti-BAFF monoclonal antibody approved for active SLE, and anifrolumab, an anti-type I interferon receptor antibody approved for moderate-to-severe SLE, have demonstrated overall disease-modifying effects, with limited case-level evidence suggesting benefit in steroid-resistant SLE-associated pericarditis.[37-39] However, data specific to pericardial involvement remain sparse, underscoring the need for further targeted studies. Lastly, the use of IL-1 inhibitors such as anakinra and rilonacept is limited to a few case studies.[6,34]

Rheumatoid Arthritis

Management of RA-associated pericarditis should integrate treatment of pericardial inflammation with control of systemic RA activity. Initial therapy may include NSAIDs and colchicine when renal function and comorbidities permit; however, lack of improvement, persistent symptoms, or progression of pericardial effusion should prompt reassessment of RA disease activity rather than escalation of pericarditis-directed therapy alone.[2,3,6] Colchicine may reduce recurrence and support steroid minimization, although RA-specific data remain limited.[2,3,6] When steroids are required for refractory pericarditis or concurrent RA flare, the lowest effective dose with slow tapering is recommended given the increased recurrence risk with prolonged or high-dose exposure.[2,3,6] In recurrent or steroid-dependent disease, optimization of disease-modifying antirheumatic drugs, in close collaboration with rheumatology, remains the preferred steroid-sparing strategy.[2,3,6] Although cytokine-targeted therapies show inconsistent benefit, limited case-based evidence suggests that B-cell–directed therapy with rituximab may be effective in severe, refractory RA-associated pericarditis, highlighting a potential role for alternative immunologic targets in this uncommon but life-threatening manifestation.[40]

CONCLUSION

Autoimmune-associated pericarditis remains clinically important yet understudied, with no disease-specific randomized trials across SLE, RA, SSc, MCTD, or vasculitis. Diagnosis requires close rheumatology-cardiology collaboration using inflammatory markers, complement levels, autoantibody profiles, and selective echocardiography or CMR to identify pericardial inflammation during systemic flares. NSAIDs and colchicine remain first line, steroids are crucial although long-term therapy is limited due to side effects, and recurrent or steroid-dependent disease should prompt early use of steroid-sparing immunosuppressants or IL-1 inhibitors. Future progress hinges on autoimmune-specific trials, prospective cohorts, and long-term biologic registries to better define risk, guide therapy, and personalize care.

KEY POINTS

- Pericardial involvement in autoimmune disease is common, often subclinical and underrecognized.
- Pathogenesis and disease course are highly variable.
- While nonsteroidal anti-inflammatory drugs and colchicine are often the first line of therapy followed by corticosteroids in many patients, disease-specific steroid-sparing immunomodulatory therapy has become essential in many conditions.
- IL-1 inhibition has become established strategy in relapsing auto-inflammatory pericarditis, but autoimmune-pericarditis-specific data remain limited.
- Multidisciplinary care and use of multi-modality imaging are key in the care of patients with autoimmune pericarditis.

COMPETING INTERESTS

The authors have no competing interests to declare.

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