# DISTINCTIVE CLINICAL FEATURES AND OUTCOMES OF LUPUS MYOCARDITIS

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## INTRODUCTION

**Myocarditis** is a severe complication of SLE with potentially life-threatening outcomes. Despite its clinical significance, its prevalence, clinical presentation, and prognosis remain poorly understood. This study aimed to characterise **lupus-related myocarditis (LM)** in patients treated at our institution.

## METHODS

We conducted a retrospective review of our hospital database to identify all SLE patients diagnosed at our centre between 1980 and 2024 who experienced at least one episode of clinically suspected LM.

The diagnosis of LM was based on a compatible clinical presentation and non-invasive findings, including elevated serum troponin I levels and functional or structural abnormalities detected via echocardiography or cardiac magnetic resonance imaging (CMRI).

## RESULTS

Of a total of 697 patients with a diagnosis of SLE, 11 (1.6%) presented 15 episodes of clinical suspicion of lupus myocarditis (LM). The main data of the patients and the 15 episodes are summarized in Tables 1 and 2. Seventy-three percent were women, with an average age of 37 ± 12 years and a median duration of lupus of 36 months.

Table 1. Demographic and clinical characteristics of the SLE study cohort.

| | N=11 |
|---|---|
| Women | 8 (73%) |
| Ethnic groups: Caucasian / Afroamerican / Hispanic | 8 (73%)<br>1 (9%)<br>2 (18%) |
| Age, mean ± SD | 37 ± 12 |
| Median disease duration, months | 36 (8–120) |
| **Cardiovascular risk factors** | |
| Smoker (ever) | 5 (45.5%) |
| Hypertension | 4 (36%) |
| Dyslipidemia | 5 (45.5%) |
| Diabetes | 2 (18%) |
| **Clinical features of SLE** | |
| Musculoskeletal disease | 10 (91%) |
| Mucocutaneous | 8 (73%) |
| Raynaud phenomenon | 3 (27%) |
| Nephritis | 6 (55%) |
| Hematologic manifestations | 8 (73%) |
| Neuropsychiatric manifestations | 2 (18%) |
| Serositis | 6 (55%) |
| **Immunological laboratory** | |
| Anti-DNA | 11 (100%) |
| Anti-Sm | 4 (36%) |
| Anti-URNP | 5 (45.5%) |
| Anti-SSA/Ro | 6 (55%) |
| Anti-SSB/La | 2 (18%) |
| Lupus anticoagulant | 4 (36%) |
| Anticardiolipin positivity | 5 (45.5%) |
| Anti-beta 2 glycoprotein | 5 (45.5%) |
| Hypocomplementemia | 9 (82%) |
| **Treatments prior to lupus myocarditis** | |
| Hydroxychloroquine | 7 (64%) |
| Methotrexate | 3 (27%) |
| Mycophenolate mofetil | 5 (45.5%) |
| Azathioprine | 3 (27%) |
| Belimumab | 1 (9%) |
| Rituximab | 2 (18%) |
| Anifrolumab | 1 (9%) |

Table 2. Clinical presentation, laboratory findings, complementary tests and management of LM

| | N=15 |
|---|---|
| **Clinical manifestations** | |
| Chest | 10 (67%) |
| Dyspnea | 6 (40%) |
| Arrhythmias | 8 (53%) |
| Syncope | 0 (0%) |
| **Laboratory findings** | |
| Raised ESR | 11 (73%) |
| Raised C reactive protein | 9 (60%) |
| Hypocomplementemia | 10 (67%) |
| CPK (U/L), mean ± SD | 740 ± 884.5 |
| Troponin I (ng/L), mean ± SD | 851 ± 1026 |
| Pro-BNP | 6894 ± 11638 |
| **Complementary tests** | |
| ECG abnormalities (N=15) | 8 (53%) |
| Echocardiographic findings (N=13) | |
| Valvular dysfunction only | 3 (23%) |
| Ventricular disfunction only | 4 (31%) |
| Valvular and ventricular disfunction | 2 (15%) |
| Pericardial effusion | 4 (31%) |
| Cardiac MRI findings (N=10) | |
| Myocardial edema | 10 (100%) |
| Ischemic areas | 2 (10%) |
| Fibrosis | 1 (10% |
| **Treatment** | **N=11** |
| Glucocorticoids | 11 (100%) |
| Intravenous MPDN boluses | 7 (64%) |
| Immunosuppressants | 8 (73%) |
| Cyclophosphamide | 4 (36%%) |
| Mycophenolate mofetil | 6 (54.5%) |
| Azathioprine | 1 (9%) |
| Biologics agents | 6 (54.5%) |
| Anifrolumab | 3 (27%) |
| Rituximab | 2 (18%) |
| Belimumab | 1 (9%) |
| Hydroxychloroquine | 7 (64%) |
| Median follow-up, months | 24 (9–72) |
| Full recovery of cardiac function | 10 (91%) |
| Improvement without complete recovery of LVEF | 1 (9%) |

The most frequent symptoms were chest pain (67%), arrhythmias (53%), and dyspnea (40%). Echocardiographic abnormalities were detected in nearly half of the cases, and cardiac MRI showed myocardial edema in all episodes where it was performed (67% of the total). Cardiac biomarkers were markedly elevated. The average ejection fraction was 51.5%.

Concomitant lupus activity was present in all patients, with musculoskeletal, mucocutaneous, hematologic, and renal involvement in most. The positivity of anti-DNA (100%) and antiphospholipid antibodies was significantly more frequent in LM cases compared to SLE patients without cardiac involvement.

All patients received glucocorticoids, with the use of immunosuppressants in 73% and biologic drugs in 54.5%. During follow-up, 91% achieved complete cardiac recovery.

## CONCLUSIONS

**The prevalence of clinical suspicion of LM in our cohort was 1.6%.** It usually appears in the early stages of the disease and in a context of systemic lupus activity. Clinical manifestations are highly variable. Early diagnosis and immediate treatment are essential to avoid fatal consequences, highlighting the importance of maintaining a high level of clinical suspicion regarding this complication.

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