

being the first presenting symptom in around 50–70% of cases and affecting up to 95% of patients at some point. The most prevalent manifestations are arthralgia and arthritis (95%), follow by myalgia/myositis (17%), tenosynovitis and bursitis (12%). Deformities arise in 5–15% of patients with no radiographic erosions, as the hallmarks of Jaccoud's arthropathy, whereas radiographic erosions may be detected in less than 5% of patients suggesting the overlap between rheumatoid arthritis and SLE referred as rhupe syndrome. Other comorbidities may be present as musculoskeletal involvement, such as: fibromyalgia (6–32%), fragility fractures (8–12%) and avascular osteonecrosis (2–12%).^{1–4}

Recent insights from US studies show that in large numbers of lupus patients with arthralgia, despite subclinical synovitis, clinical assessment underestimates the level of joint and tendon inflammation compared to ultrasound and magnetic resonance imaging. This has implications for therapeutic choice, evaluation of response or treat-to-target protocols. While prevalence of subclinical synovitis is agreed, it is not yet clear whether it should be treated, however imaging studies suggest potential changes to the classification, assessment and management of patients with inflammatory musculoskeletal lupus.

SLE manifestations are considered refractory when patients are unresponsive to or disease relapses despite treatment with corticosteroids, antimalarials and/or immunosuppressants. Other key issues are that recurrent flares of disease activity are associated with poor long-term outcomes and longstanding overreliance on corticosteroid therapy, which contributes substantially to damage accrual and patient mortality.

Hydroxychloroquine and corticosteroids remain first-line therapies for musculoskeletal manifestations of SLE; however, patients with refractory musculoskeletal disease may require further management with immunosuppressive (methotrexate, leflunomide) or biologic (belimumab, abatacept, rituximab) agents for inflammatory disease control. Mycophenolate mofetil and, to a lesser degree, azathioprine have shown efficacy in the treatment of inflammatory myositis in SLE patients and have demonstrated a steroid sparing effect. Despite current therapy, musculoskeletal manifestations are major contributors to poor quality of life and work instability. There remains an unmet clinical need in SLE, particularly in patients with disease refractory to conventional immunosuppressive therapies.

Learning Objectives

- Explain the main musculoskeletal manifestations in patients with SLE
- Describe potential changes to the classification of musculoskeletal manifestations
- Describe the current recommendations for the treatment of refractory musculoskeletal manifestations
- Discuss new trends in research on new therapies for musculoskeletal manifestations in patients with SLE

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REFRACTORY AND/OR CATASTROPHIC APS IN SLE

Ricard Cervera. *Hospital Clinic, Barcelona, Catalonia, Spain*

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Antiphospholipid syndrome (APS) is an autoimmune disease characterised by vascular thrombosis and pregnancy morbidity in the presence of antiphospholipid antibodies (aPL), mainly lupus anticoagulant (LA), anticardiolipin antibodies (aCL) and anti- β 2-glycoprotein I antibodies (anti- β 2GPI). Precision Medicine can benefit the specific profiles of patients with refractory thrombotic and/or obstetric APS and those with catastrophic APS.

Hydroxychloroquine and low-dose steroid, alone or combined, may be an option for pregnant APS patients with a previous pregnancy refractory to conventional therapy. Intravenous immunoglobulins and plasma exchange, alone or combined, could be considered in refractory high-risk pregnant APS patients.^{1 2}

Evidence on the management of recurrent thrombosis despite vitamin K antagonists (VKA) treatment is limited. After evaluating other risk factors for thrombosis (e.g., traditional cardiovascular risk factors, cancer, other thrombophilic states) and investigating the adherence to VKA treatment, increase of target international normalised ratio (INR) to 3–4, or INR 2–3 with the addition of low dose aspirin, or switching to low molecular weight heparin may be considered. Adjunctive therapy with antimalarials or statins could also be considered.

Management of catastrophic APS is challenging. The higher recovery rate is achieved by the combination of anticoagulation, plus glucocorticoids, plus plasma exchange and/or intravenous immunoglobulins. New therapeutic approaches include rituximab and eculizumab.

Learning Objectives

- Explain the main unmet needs in the management of the APS in SLE
- Describe the options for the treatment of refractory thrombotic and obstetric manifestations of APS
- Discuss the current recommendations for the management of catastrophic APS cases
- Discuss new trends in research on new therapies for APS

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