Systemic Sclerosis: EULAR 2026 Highlights Diagnosis, Prognosis

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Systemic sclerosis (SSc) presents a significant clinical challenge due to its heterogeneous manifestations and variable disease course, often leading to delayed diagnosis and suboptimal management. The EULAR 2026 session on SSc highlighted the imperative for earlier, more precise identification of disease subtypes and risk stratification to enable timely, targeted therapeutic strategies.

KEY TAKEAWAYS

- The Pivot: Evolving diagnostic criteria and prognostic models are refining SSc classification beyond traditional diffuse versus limited subtypes.
- The Data: Early identification of specific autoantibody profiles and organ involvement patterns correlates with distinct disease trajectories and treatment responses.
- The Action: Clinicians should integrate updated diagnostic algorithms and risk stratification tools to guide patient management and intervention.

Systemic sclerosis (SSc) is a complex autoimmune connective tissue disease characterised by widespread vascular damage, immune activation, and progressive fibrosis of the skin and internal organs. The clinical presentation is highly variable, ranging from mild cutaneous involvement to severe, life-threatening organ complications, including interstitial lung disease (ILD), pulmonary arterial hypertension (PAH), and renal crisis. This heterogeneity complicates early diagnosis and accurate prognostication, often leading to delayed therapeutic intervention and irreversible organ damage. The EULAR 2026 session, 'New dimensions in systemic sclerosis: evolving concepts in diagnosis, prognosis and intervention,' addressed these challenges by presenting updates on diagnostic methodologies, risk stratification, and emerging therapeutic approaches.1

Historically, SSc classification has relied on the extent of skin involvement, broadly categorising patients into limited cutaneous SSc (lcSSc) and diffuse cutaneous SSc (dcSSc). While useful, this dichotomous system does not fully capture the prognostic diversity or guide specific therapeutic choices for all patients. Recent advancements emphasise the importance of integrating autoantibody profiles, specific organ involvement patterns, and genetic markers to refine diagnosis and predict disease progression. For instance, the presence of anti-topoisomerase I (Scl-70) antibodies is strongly associated with dcSSc and a higher risk of ILD, while anti-centromere antibodies (ACA) are characteristic of lcSSc and predict PAH.2

Evolving Diagnostic and Prognostic Frameworks

The EULAR 2026 discussions underscored the utility of updated diagnostic criteria that incorporate not only clinical features but also serological markers and capillaroscopic abnormalities. Early detection of SSc-specific autoantibodies, even in the absence of overt clinical symptoms, can identify individuals at risk of developing definite SSc. This pre-sclerotic phase, often characterised by Raynaud's phenomenon and abnormal nailfold capillaries, represents a critical window for potential preventative or early disease-modifying interventions.3

Prognostic models are also evolving to provide more granular risk stratification. Beyond the traditional lcSSc/dcSSc distinction, new models integrate factors such as the rate of skin thickening, the presence and severity of specific organ involvement (e.g., forced vital capacity percentage predicted for ILD, mean pulmonary arterial pressure for PAH), and specific autoantibody subsets. For example, patients with rapidly progressive skin disease and anti-RNA polymerase III antibodies are at increased risk of renal crisis and severe skin involvement.4 These refined prognostic tools aim to identify high-risk patients who may benefit from more aggressive or targeted therapies earlier in their disease course. The session highlighted the potential for artificial intelligence and machine learning algorithms to integrate these diverse data points, providing more accurate individualised risk predictions.5

Interventional Strategies and Future Directions

Interventional strategies in SSc continue to focus on managing organ-specific complications and modulating the underlying immune and fibrotic processes. For SSc-ILD, antifibrotic agents such as nintedanib and pirfenidone have demonstrated efficacy in slowing the decline in forced vital capacity. Immunosuppressants, including mycophenolate mofetil and cyclophosphamide, remain cornerstones for managing inflammatory manifestations and progressive ILD. For SSc-PAH, endothelin receptor antagonists, phosphodiesterase-5 inhibitors, and prostacyclin analogues are established therapies.6

The EULAR 2026 session also touched upon emerging therapeutic targets, including pathways involved in B-cell activation, T-cell dysregulation, and fibroblast activation. Biologic agents targeting specific cytokines or cell surface receptors are under investigation, with some showing promise in early-phase trials. The concept of 'treat-to-target' in SSc, similar to its application in rheumatoid arthritis, is gaining traction. This approach involves setting specific treatment goals based on disease activity and organ involvement, with regular assessment and adjustment of therapy to achieve these targets. However, defining and validating appropriate targets and outcome measures in SSc remains an ongoing challenge due to the disease's heterogeneity.7

Limitations in current SSc management include the lack of truly disease-modifying therapies that can halt or reverse fibrosis, the need for better biomarkers to predict treatment response, and the challenges of conducting large-scale clinical trials in a rare and heterogeneous disease. Future research directions include the development of novel antifibrotic and immunomodulatory agents, the identification of predictive biomarkers, and the implementation of precision medicine approaches tailored to individual patient profiles. The integration of multi-omics data (genomics, proteomics, metabolomics) is expected to provide deeper insights into disease pathogenesis and identify new therapeutic avenues.8

> **CLINICAL IMPLICATIONS**
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> The EULAR 2026 session on systemic sclerosis underscores a clear message for clinicians: the era of broad, symptomatic management is receding. The emphasis on refined diagnostic criteria and prognostic

indicators demands a more granular approach to patient assessment. Relying solely on the diffuse versus limited classification is increasingly insufficient; clinicians must integrate autoantibody profiles and specific organ involvement patterns from the outset. This shift requires a proactive stance, moving beyond reactive treatment of established complications to earlier identification of at-risk individuals and targeted intervention.
For the pharmaceutical industry, the implications are equally direct. The demand for therapies that address specific SSc subtypes or pathways is growing. Broad-spectrum immunosuppressants, while useful, may not be the optimal long-term solution for all patients. Investment in developing agents that target specific fibrotic pathways or immune dysregulations identified by advanced phenotyping, such as those associated with anti-topoisomerase I or anti-RNA polymerase III antibodies, will be critical. Furthermore, the development of robust biomarkers to predict treatment response and disease progression is paramount to demonstrating efficacy in a heterogeneous patient population.
Ultimately, patients stand to benefit from this evolving understanding. Earlier, more precise diagnosis and risk stratification mean the potential for earlier, more effective interventions, potentially mitigating irreversible organ damage. However, this also places a greater onus on healthcare systems to ensure access to advanced diagnostic tools, such as detailed autoantibody panels and high-resolution imaging for organ screening. The promise of personalised medicine in SSc is contingent not just on scientific advancement, but also on the practical implementation of these 'new dimensions' into routine clinical practice, ensuring that the benefits reach every patient, not just those in specialist centres.

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