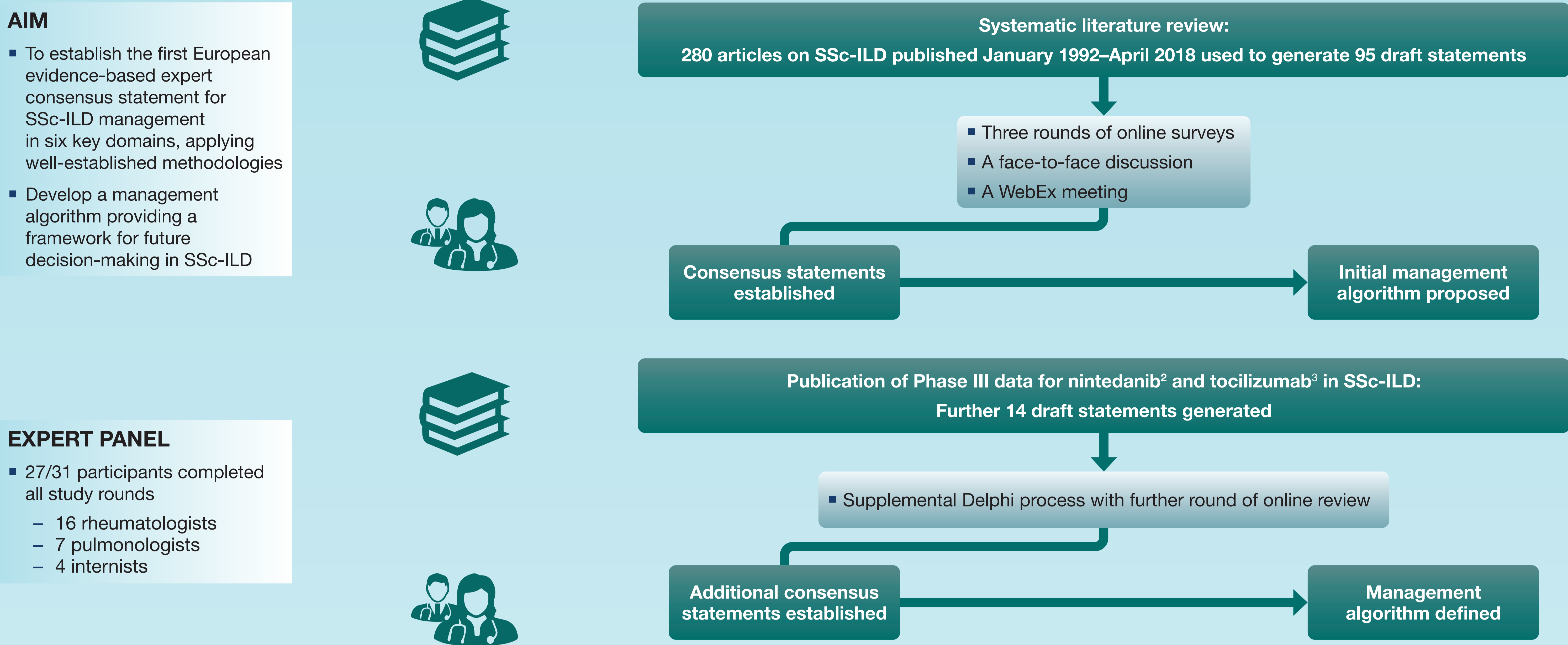


Evidence-based consensus statements for the identification and management of interstitial lung disease in systemic sclerosis

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MODIFIED DELPHI PROCESS¹ TO DEVELOP EVIDENCE-BASED CONSENSUS STATEMENTS



STATEMENTS IN SIX KEY DOMAINS







1. Risk factors that affect the likelihood of developing SSc-ILD⁴⁻⁹

	Respiratory symptoms	More likelihood of ILD
	Smoking history	
	Ethnicity (Native American; African heritage)	
	Male gender	
	Diffuse cutaneous SSc	
	Anti-topoisomerase I antibodies	Less likelihood of ILD
	Anti-centromere antibodies	

2. Screening for SSc-ILD¹⁰⁻¹⁴

	All patients should be screened at baseline using HRCT
	Pulmonary function testing provides baseline parameters <ul style="list-style-type: none">FVCDL_{CO}Auscultation

3. SSc diagnosis^{15,16} and severity assessment^{17,18}

	The primary tool for diagnosing ILD in patients with SSc is HRCT		
	Supporting diagnostic tools Pulmonary function tests and clinical assessment of respiratory symptoms ¹⁸		
More than one measure should be used to determine severity			
			
HRCT findings	Respiratory symptoms	Exercise-induced oxygen desaturation	Quality of life

4. SSc-ILD treatment initiation¹⁹ and options^{2,20-22}

Drivers of treatment initiation	Pharmacological treatment options
<ul style="list-style-type: none">QoLClinical guidelines/experiencePatient's symptomsEfficacySafety/tolerability	<p>"No pharmacological treatment" is an option for some</p> <ul style="list-style-type: none">Patients should be followed up regularly and treatment initiated in case of progression <p>All cases of severe ILD should be offered treatment</p> <ul style="list-style-type: none">Mycophenolate mofetil, cyclophosphamide and nintedanib are effective

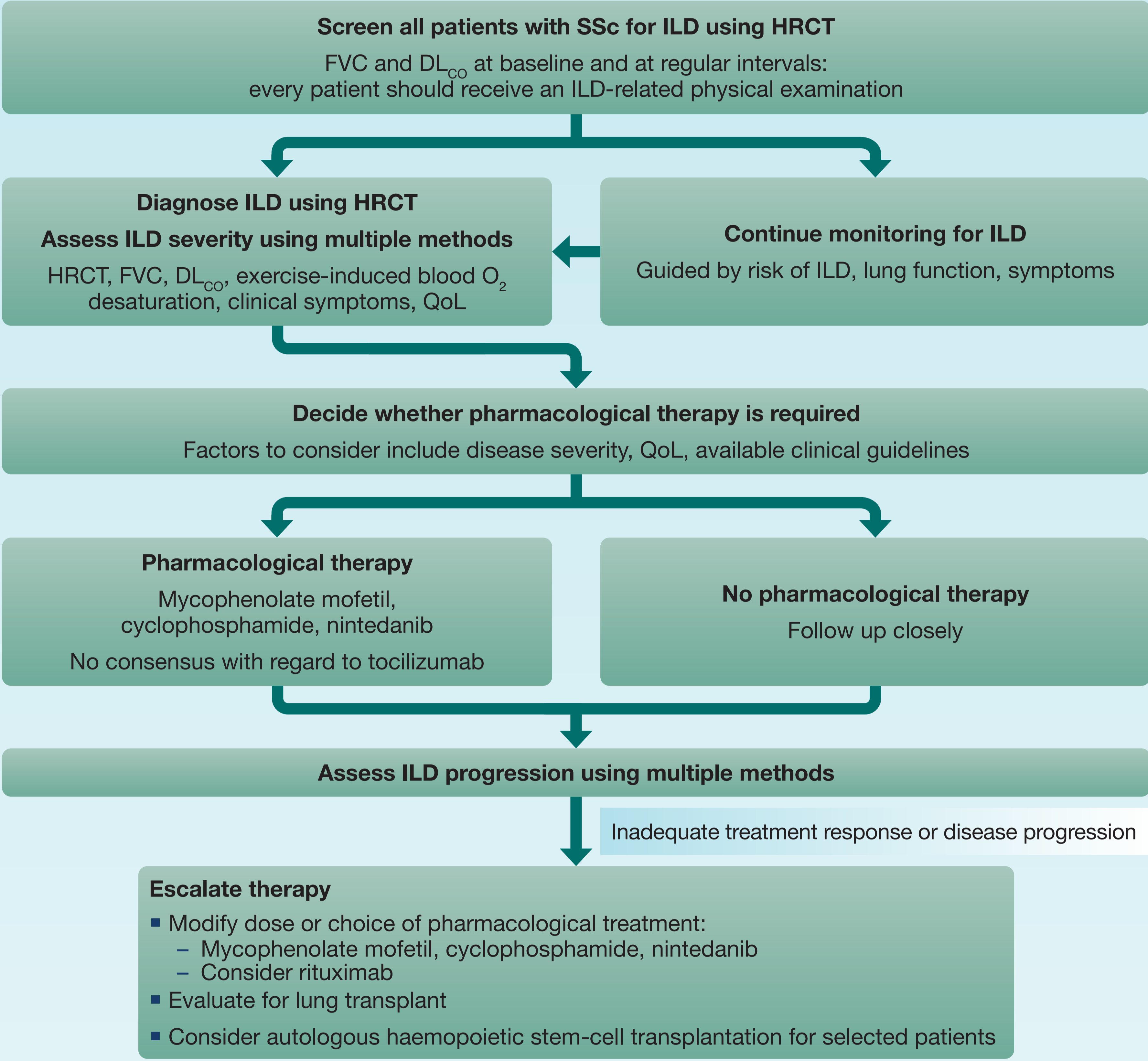
5. SSc-ILD disease progression assessment^{18,23}

More than one measure should be used to assess progression	
	Changes in extent of fibrosis or pattern on HRCT
	Pulmonary function tests (FVC and DL _{CO} absolute value, or FVC decline)
	Exercise-induced oxygen desaturation
	Worsening of clinical symptoms

6. SSc-ILD treatment escalation and options^{2, 24-29}

Drivers of escalation	Available treatment options
<ul style="list-style-type: none">Pace of progressionDisease severity	<p>All patients with severe or progressive SSc-ILD should be offered pharmacological treatment</p> <ul style="list-style-type: none">Available options include: mycophenolate mofetil, cyclophosphamide, nintedanib, combination nintedanib and mycophenolate mofetil, rituximab <p>Autologous haemopoietic stem-cell transplant</p> <p>Lung transplant (evaluate suitability early)</p>

MANAGEMENT ALGORITHM



CONCLUSIONS

- This multidisciplinary modified Delphi study provides evidence-based expert consensus statements for SSc-ILD management across six key domains
- An SSc-ILD management algorithm for use in clinical practice is also provided
- These consensus statements and the clinical management algorithm provide important clinical guidance for the early identification and medical management of SSc-ILD, and offer a framework for future treatment decision-making

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Abbreviations

DL_{CO}, diffusing capacity of the lungs for carbon monoxide; FVC, forced vital capacity; HRCT, high-resolution computed tomography; ILD, interstitial lung disease; QoL, quality of life; SSc, systemic sclerosis.