**Additional file 1**

**Expert consensus on the management of systemic sclerosis-associated interstitial lung disease**

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**Table S1. SSc-ILD Delphi Questionnaire 3 results**

This table contains the questions from Questionnaire 3, the mean and SD of the Likert scale results, and whether consensus was reached or not reached.

|  |  |  |  |
| --- | --- | --- | --- |
| **Statements** | **Mean** | **SD** | **Consensus** |
| **How do you screen for SSc-ILD?** | | | |
| **Which of the following would you likely perform to screen the general scleroderma population for ILD?** | | | |
| Spirometry with DLCO | 4.36 | 1.04 | For |
| Full PFT | 4.16 | 1.70 | For |
| Chest x-ray | 0.40 | 3.23 | No |
| HRCT | 4.08 | 1.63 | For |
| Biomarker tests | −0.32 | 2.70 | No |
| Manometry data | −1.92 | 2.14 | No |
| Autoantibody testing | 2.96 | 1.79 | For |
| Echocardiogram | 0.32 | 3.29 | No |
| Doppler | −0.68 | 3.09 | No |
| 6MWD | 2.04 | 2.54 | No |
| Chest auscultation | 4.32 | 1.18 | For |
| **Which patients do you screen for SSc-ILD?** | | | |
| All scleroderma patients | 3.72 | 2.19 | For |
| Patients with symptoms | 4.92 | 0.28 | For |
| High-risk patients (e.g.: dcSSc, +Scl-70 antibodies, African American ethnicity, and/or a high mRSS) | 4.96 | 0.20 | For |
| **Who do you treat?** | | | |
| **When deciding whether to treat patients for ILD do you consider:** | | | |
| Extent of ILD or fibrosis on HRCT | 4.56 | 1.08 | For |
| Autoantibody status | 2.44 | 1.50 | No |
| Baseline PFT values | 4.08 | 0.95 | For |
| Clinically meaningful change in PFT values | 4.76 | 0.52 | For |
| Duration and degree of dyspnea | 3.76 | 1.36 | For |
| Length of disease | 2.36 | 1.38 | No |
| Potential contribution of reflux | 2.52 | 1.58 | For |
| Patient age | 1.28 | 2.46 | No |
| Comorbidities | 2.40 | 1.56 | No |
| Presence of pulmonary hypertension | 3.08 | 1.63 | For |
| **Based on HRCT, do you treat patients who have:** | | | |
| Worsening HRCT with symptoms or declining PFTs | 4.84 | 0.37 | For |
| >20% involvement on HRCT with normal PFTs | 3.04 | 1.57 | For |
| >20% involvement on HRCT with abnormal PFTs | 4.48 | 0.82 | For |
| >10% involvement on HRCT with abnormal PFTs | 3.52 | 1.30 | For |
| High-risk patients (early diffuse disease) with evidence of mild ILD (<10%) and abnormal PFTs | 4.08 | 1.12 | For |
| High-risk patients (early diffuse disease) with evidence of mild ILD (<10%) | 3.24 | 1.51 | For |
| **Based on FVC and symptom status (assume all patients have ILD on HRCT), do you treat patients who have:** | | | |
| FVC >80% with ILD on HRCT in a high-risk patient (early diffuse disease, Topo+) | 3.72 | 1.51 | For |
| FVC >80% with ILD on HRCT in a low-risk patient (early limited disease, centromere+) | 0.72 | 1.93 | No |
| FVC <80% with any degree of ILD on HRCT | 2.68 | 1.80 | For |
| FVC >80% and dyspnea | 2.36 | 1.68 | No |
| FVC <80% and dyspnea | 3.44 | 1.50 | For |
| FVC <70% and dyspnea | 4.12 | 1.24 | For |
| Decline in FVC by greater than measurement error (5–7%) | 4.16 | 0.90 | For |
| Decline in FVC by >10% in 1 year | 4.48 | 0.87 | For |
| Regardless of FVC once other causes of dyspnea are excluded | 2.20 | 1.85 | No |
| **In deciding to initiate treatment for SSc-ILD, how important are other parameters besides HRCT and PFTs?** | | | |
| 6MWD | 2.00 | 1.83 | No |
| Exertional desaturation on SpO2 | 3.28 | 1.54 | For |
| Echocardiogram | 1.68 | 2.58 | No |
| Cough consistent with ILD | 1.52 | 1.53 | No |
| Presence of active cutaneous disease | 1.88 | 1.94 | No |
| Presence of reflux | 1.00 | 1.96 | No |
| Scl-70 antibody status | 2.04 | 1.72 | No |
| Elevated BNP | 0.08 | 2.14 | No |
| Evidence of Velcro crackles on lung examination | 2.08 | 1.91 | No |
| **To determine the phenotype of patients that are likely to respond to treatment, which strategy or strategies are you likely to employ?** | | | |
| Autoantibody status | 2.24 | 1.94 | No |
| Cutaneous disease status | 2.28 | 1.88 | No |
| Findings or changes on HRCT | 3.84 | 1.31 | For |
| Findings or changes on PFTs | 3.68 | 1.35 | For |
| Duration of symptoms | 2.72 | 1.54 | For |
| Lung biopsy specimen demonstrating cellular/non-fibrotic NSIP | 1.64 | 2.75 | No |
| Lung biopsy specimen demonstrating UIP | 0.64 | 2.58 | No |
| Concomitant pulmonary vascular disease | 1.20 | 2.35 | No |
| I don’t believe we have enough reliable information at this point to determine the phenotype of patients likely to respond to treatment | 1.48 | 2.55 | No |
| **The following patients should NOT be treated for SSc-ILD:** | | | |
| Preserved PFTs with mild ILD on HRCT | 1.68 | 2.08 | No |
| Patients with history of chronic lung infections, including MAI and aspergillus | 2.16 | 2.27 | No |
| Patients with longstanding disease (close to 10 years) with stable PFTs and no progression of ILD over last few years | 3.52 | 1.81 | For |
| Centromere+ patients | −0.12 | 2.52 | No |
| Stable PFTs over 1 year | 1.32 | 2.14 | No |
| Advanced/end-stage lung disease | 1.84 | 2.61 | No |
| Patients with UIP pattern on HRCT | −0.92 | 2.57 | No |
| Patients who are older | −0.96 | 2.35 | No |
| Patients with significant comorbidities | 0.32 | 1.99 | No |
| Patients with significant recurrent clinical aspiration | 0.28 | 2.07 | No |
| Patients with pulmonary arterial hypertension | −1.44 | 2.22 | No |
| Patients with advanced liver or renal disease who are at risk for more complications related to medical therapies | 1.36 | 2.66 | No |
| **Regarding consideration of autoantibodies in deciding whether to treat patients for SSc-ILD at initial presentation:** | | | |
| I do not consider autoantibodies in deciding whether to treat patients for SSc-ILD | −0.48 | 2.60 | No |
| I consider the presence of anti-Scl-70 and anti-nucleolar pattern on ANA in deciding to treat patients for SSc-ILD | 1.64 | 1.96 | No |
| I consider presence of RNA polymerase III antibodies in deciding to treat patients for SSc-ILD at initial presentation | 1.52 | 2.02 | No |
| Patients with other antibodies than U1 RNP or no specific antibodies have to be taken individually as we don't know how aggressive their disease will be | 2.76 | 2.19 | For |
| In patients with centromere+ antibodies, I am less likely to treat them for SSc-ILD | 0.12 | 2.91 | No |
| In patients with RNA polymerase III+ antibodies, I am less likely to treat them for SSc-ILD | −0.84 | 2.61 | No |
| **“At initial presentation in patients with SSc, this condition would cause me enough concern about near-term ILD that I would start treatment right away”** | | | |
| Moderate-to-severe ILD on HRCT | 4.00 | 1.12 | For |
| FVC and/or DLCO <LLN | 2.72 | 1.90 | For |
| Moderate-to-severe symptoms | 3.68 | 1.07 | For |
| Early, rapidly progressive dcSSc even with mild abnormalities on HRCT chest scan | 3.64 | 1.15 | For |
| Early, rapidly progressive dcSSc even with mild abnormalities on PFT | 3.60 | 1.29 | For |
| Early, rapidly progressive dcSSc even with mild abnormalities on HRCT chest scan AND mild abnormalities on PFT | 4.20 | 0.87 | For |
| Presence of anti-Scl-70 | 1.56 | 1.73 | No |
| HRCT showing ILD >20% lung involvement | 3.80 | 0.96 | For |
| Hypoxemia at rest | 3.92 | 1.12 | For |
| Desaturation on exercise | 3.84 | 1.11 | For |
| **How do you treat?** | | | |
| **What initial therapy do you use once you have decided to treat SSc-ILD?** | | | |
| MMF | 4.72 | 0.74 | For |
| CYC | 0.36 | 2.84 | No |
| Rituximab | 0.52 | 2.66 | No |
| Azathioprine | −0.28 | 2.41 | No |
| Methotrexate | −2.84 | 2.48 | Against |
| Prednisone | −0.60 | 2.75 | No |
| **What is your typical/target dose for MMF?** | | | |
| 1000 mg daily | −2.40 | 2.45 | No |
| 1500 mg daily | −1.88 | 2.67 | No |
| 2000 mg daily | 2.68 | 1.82 | For |
| 3000 mg daily | 4.44 | 0.77 | For |
| I do not utilize MMF | −4.56 | 1.39 | Against |
| **What is your typical/target dose for azathioprine?** | | | |
| 2–3 mg/kg/day | 2.88 | 2.46 | For |
| 50–150 mg/day | 1.04 | 3.34 | No |
| Up to 200 mg/day | 1.04 | 3.39 | No |
| I do not utilize azathioprine | −2.56 | 2.97 | No |
| **What is your typical/target dose for rituximab?** | | | |
| 1 g on days 0 and 15 | 2.00 | 2.89 | No |
| 1 g on days 0 and 15, then every 6–12 months | 2.76 | 2.49 | For |
| 2 g every 6 months | −0.40 | 2.99 | No |
| I do not utilize rituximab | −2.76 | 2.70 | Against |
| **Use of antifibrotic drugs** | | | |
| I see antifibrotic drugs fitting into the management of SSc-ILD after CYC/MMF | 2.36 | 2.46 | No |
| I see antifibrotic drugs fitting into the management of SSc-ILD concomitant to CYC/MMF | 2.96 | 2.37 | For |
| I see antifibrotic drugs fitting into the management of SSc-ILD as determined based on decline in lung function and/or HRCT | 2.64 | 1.96 | For |
| I do not see antifibrotic drugs fitting into the management of SSc-ILD | −4.24 | 1.17 | Against |
| **Use of nintedanib [following publication of SENSCIS trial results]** | | | |
| **I would use nintedanib for treating patients with SSc-ILD under the following clinical conditions:** | | | |
| Patients with progressive fibrotic ILD despite immunosuppressive therapy | 4.28 | 0.79 | For |
| Patients with progressive fibrotic ILD in combination with MMF/CYC | 4.00 | 0.96 | For |
| Patients who have contraindications to or are unable to tolerate immunosuppression | 3.71 | 1.55 | For |
| In combination with immunosuppressive agents (MMF/CYC) for patients with aggressive ILD, advanced disease at initial presentation, or significant disease progression | 3.68 | 1.28 | For |
| Any patient with CTD with clinically significant or worsening ILD | 1.96 | 1.81 | No |
| Based on lack of effective response or improvement with immunosuppressive agents (MMF/CYC) as defined by lack of symptom improvement | 0.96 | 1.65 | No |
| Based on lack of effective response or improvement with immunosuppressive agents (MMF/CYC) as defined by lack of improvement of ILD on HRCT | 1.28 | 1.67 | No |
| Based on lack of effective response or improvement with immunosuppressive agents (MMF/CYC) as defined by lack of improvement of lung function | 1.76 | 1.76 | No |
| Based on lack of effective response or improvement with immunosuppressive agents (MMF/CYC) as defined by a combination of the above | 2.52 | 1.73 | For |
| Based on active worsening of patient condition as defined by worsening of symptoms | 1.76 | 1.42 | No |
| Based on active worsening of patient condition as defined by worsening of ILD on HRCT | 3.00 | 1.29 | For |
| Based on active worsening of patient condition as defined by worsening of lung function | 3.33 | 0.87 | For |
| Based on active worsening of patient condition as defined by a combination of the above | 3.72 | 0.79 | For |
| Based on inability to continue CYC/MMF due to adverse effects | 2.80 | 1.35 | For |
| Based on inability to continue CYC/MMF due to lack of achievement of effective dose with CYC/MMF | 2.00 | 1.53 | No |
| Based on inability to continue CYC/MMF due to prolonged use | 1.21 | 1.93 | No |
| **Nintedanib fits into the management of SSc-ILD as:** | | | |
| Initial monotherapy in cases with significant lung fibrosis (e.g. >10% total lung involvement on HRCT) with preserved FVC and DLCO | −0.88 | 2.13 | No |
| Initial monotherapy in patients with contraindication to or problems tolerating immunosuppressive agents | 1.32 | 2.50 | No |
| Initial monotherapy in patients with >20% total lung involvement on HRCT, indicative of significant fibrosis | 0.04 | 2.14 | No |
| Initial monotherapy in patients with longstanding SSc (>5 years) with ILD and evidence of progression for whom immunosuppression would not be recommended | 2.60 | 1.63 | For |
| Dual initial therapy in combination with MMF/CYC | 0.92 | 2.20 | No |
| Add-on therapy after failure of MMF/CYC | 3.40 | 1.71 | For |
| Add-on therapy to MMF/CYC | 3.36 | 0.91 | For |
| **Based on the following response to SSc-ILD treatment, please indicate your likely course of action:** | | | |
| With progression/worsening of ILD, I would switch to another agent | 2.64 | 2.80 | No |
| With progression/worsening of ILD, I would add another agent | 2.92 | 2.66 | For |
| With progression/worsening of ILD, I would continue treatment as is | −1.44 | 2.84 | No |
| With stability of ILD, I would switch to another agent | −3.12 | 1.88 | Against |
| With stability of ILD, I would add another agent | −3.28 | 1.88 | Against |
| With stability of ILD, I would continue treatment as is | 3.56 | 1.85 | For |
| **How long do you treat?** | | | |
| 2 years | 2.20 | 2.60 | No |
| 5 years | 2.44 | 2.42 | No |
| Continue until stabilization of PFTs and symptoms | 2.28 | 2.94 | No |
| Continue lifelong | 0.28 | 3.39 | No |
| **What circumstances would prompt you to consider weaning a patient from therapy?** | | | |
| Mild-to-moderate disease | 0.56 | 2.52 | No |
| Comorbidities | 2.08 | 1.94 | No |
| Toxicity to the drug (including side effects and adverse events) | 4.08 | 1.00 | For |
| Stability for ≥2 years in lung as well as skin | 3.04 | 2.30 | For |
| Patient's strong desire to discontinue treatment | 3.60 | 1.66 | For |
| Lack of efficacy | 3.20 | 2.26 | For |
| **How do you wean patients from therapy?** | | | |
| Taper/wean over 1–2 years, monitor PFTs every 6 months, with or without low maintenance dose of MMF | 3.56 | 1.66 | For |
| Taper/wean over months to a year to a lower maintenance dose | 2.60 | 1.47 | For |
| Taper/wean over months to a year to off | 1.20 | 2.53 | No |
| Stop therapy quickly (over weeks or “cold turkey”) | −3.28 | 2.51 | Against |
| **What is progression to you, and how do you monitor it?** | | | |
| Absolute values in PFTs (FVC or DLCO) | 1.60 | 2.75 | No |
| Changes in PFTs over time (FVC or DLCO) | 4.56 | 0.58 | For |
| Features on HRCT (ILD pattern or extent of fibrosis) | 2.80 | 2.43 | For |
| Changes in HRCT over time | 4.28 | 0.79 | For |
| Changes in symptoms over time | 3.28 | 1.99 | For |
| 6MWD | 1.12 | 2.49 | No |
| Patient demographics (age, gender, race) | 0.36 | 2.50 | No |
| Patient autoantibody profile | 0.96 | 2.35 | No |
| Cutaneous disease status/activity | 1.24 | 2.51 | No |
| Exertional hypoxemia | 2.64 | 1.96 | For |
| Development of concomitant GI disease (GERD, microaspiration) | 0.60 | 2.18 | No |
| Development of concomitant pulmonary vascular disease (pulmonary arterial hypertension) | 1.12 | 2.44 | No |
| **What is success to you?** | | | |
| FVC stabilization | 3.76 | 0.88 | For |
| FVC improvement | 4.72 | 0.46 | For |
| DLCO stabilization | 3.68 | 0.85 | For |
| DLCO improvement | 4.60 | 0.58 | For |
| HRCT improvement | 4.60 | 0.65 | For |
| HRCT stabilization | 3.96 | 0.84 | For |
| Symptom stabilization/improvement | 4.12 | 0.83 | For |
| 6MWD stabilization/improvement | 2.92 | 1.71 | For |
| O2 saturation with exercise | 3.00 | 1.26 | For |
| mRSS stabilization/improvement | 0.92 | 2.27 | No |
| Functional status (NYHA FC or CPET) | 2.52 | 1.90 | For |

6MWD, 6-minute walk distance; ANA, antinuclear antibodies; BNP, brain natriuretic peptide; CPET, cardiopulmonary exercise testing; CTD, connective tissue disease; CYC, cyclophosphamide; dcSSc, diffuse cutaneous systemic sclerosis; DLCO, diffusing capacity of the lungs for carbon monoxide; FCV, forced vital capacity; GERD, gastroesophageal reflux disease; GI, gastrointestinal; HRCT, high-resolution computed tomography; ILD, interstitial lung disease; LLN, lower limit of normal; MAI, *mycobacterium avium-intracellulare*; MMF, mycophenolate mofetil; mRSS, modified Rodnan skin score; NSIP, non-specific interstitial pneumonia; NYHA FC, New York Heart Association Functional Classification; PFT, pulmonary function test; RNP, ribonucleoprotein; SD, standard deviation; SpO2, peripheral capillary oxygen saturation; SSc-ILD, systemic sclerosis-associated ILD; TCZ, tocilizumab; Topo, topoisomerase; UIP, usual interstitial pneumonia.

**Table S2. SSc-ILD Delphi Supplemental Questionnaire 2 results**

This table contains the questions from Supplemental Questionnaire 2, the mean and SD of the Likert scale results, and whether consensus was reached or not reached.

|  |  |  |  |
| --- | --- | --- | --- |
| **Statements** | **Mean** | **SD** | **Consensus** |
| **How do you treat?** | | | |
| **Use of antifibrotic drugs** | | | |
| I see antifibrotic drugs fitting into the management of SSc-ILD after CYC/MMF | 3.91 | 0.61 | For |
| I see antifibrotic drugs fitting into the management of SSc-ILD concomitant to CYC/MMF | 3.00 | 1.07 | For |
| I see antifibrotic drugs fitting into the management of SSc-ILD after TCZ | 2.00 | 2.07 | No |
| I see antifibrotic drugs fitting into the management of SSc-ILD concomitant to TCZ | 0.68 | 2.36 | No |
| I see antifibrotic drugs fitting into the management of SSc-ILD before TCZ | 0.27 | 2.49 | No |
| I see antifibrotic drugs fitting into the management of SSc-ILD as determined based on decline in lung function and/or HRCT | 3.82 | 0.91 | For |
| I do not see antifibrotic drugs fitting into the management of SSc-ILD | -4.68 | 0.57 | Against |
| **Use of nintedanib** | | | |
| **I would use nintedanib for treating patients with SSc-ILD under the following clinical conditions:** | | | |
| Patients with progressive fibrotic ILD despite immunosuppressive therapy (MMF/CYC/TCZ) | 4.55 | 0.80 | For |
| Patients with progressive fibrotic ILD in combination with MMF/CYC/TCZ | 4.23 | 0.87 | For |
| Patients who have contraindications to or are unable to tolerate immunosuppression (MMF/CYC/TCZ) | 4.32 | 0.65 | For |
| In combination with immunosuppressive agents (MMF/CYC/TCZ) for patients with aggressive ILD, advanced disease at initial presentation, or significant disease progression | 3.95 | 0.84 | For |
| Any patient with CTD with clinically significant or worsening ILD | 2.14 | 1.75 | No |
| Based on lack of effective response or improvement with immunosuppressive agents (MMF/CYC/TCZ) as defined by lack of symptom improvement | 2.05 | 0.95 | No |
| Based on lack of effective response or improvement with immunosuppressive agents (MMF/CYC/TCZ) as defined by lack of improvement of ILD on HRCT | 1.82 | 1.71 | No |
| Based on lack of effective response or improvement with immunosuppressive agents (MMF/CYC/TCZ) as defined by lack of improvement of lung function | 2.55 | 1.68 | For |
| Based on lack of effective response or improvement with immunosuppressive agents (MMF/CYC/TCZ) as defined by a combination of the above | 3.05 | 1.84 | For |
| Based on active worsening of patient condition as defined by worsening of symptoms | 2.14 | 1.08 | No |
| Based on active worsening of patient condition as defined by worsening of ILD on HRCT | 3.59 | 0.67 | For |
| Based on active worsening of patient condition as defined by worsening of lung function | 3.86 | 0.56 | For |
| Based on active worsening of patient condition as defined by a combination of the above | 4.05 | 0.49 | For |
| Based on inability to continue CYC/MMF/TCZ due to adverse effects | 3.14 | 0.94 | For |
| Based on inability to continue CYC/MMF/TCZ due to lack of achievement of effective dose with CYC/MMF/TCZ | 2.68 | 0.99 | For |
| Based on inability to continue CYC/MMF/TCZ due to prolonged use | 1.18 | 1.82 | No |
| **Nintedanib fits into the management of SSc-ILD as:** | | | |
| Initial monotherapy in cases with significant lung fibrosis (e.g. >10% total lung involvement on HRCT) with preserved FVC and DLCO | –1.55 | 1.22 | No |
| Initial monotherapy in patients with contraindication to or problems tolerating immunosuppressive agents | 2.27 | 1.80 | No |
| Initial monotherapy in patients with >20% total lung involvement on HRCT, indicative of significant fibrosis | 0.59 | 2.26 | No |
| Initial monotherapy in patients with longstanding SSc (>5 years) with ILD and evidence of progression for whom immunosuppression would not be recommended | 3.05 | 1.21 | For |
| Dual initial therapy in combination with MMF/CYC | 1.59 | 1.50 | No |
| Dual initial therapy in combination with TCZ | -0.05 | 2.19 | No |
| Add-on therapy after failure of MMF | 3.82 | 0.66 | For |
| Add-on therapy after failure of CYC | 3.64 | 0.79 | For |
| Add-on therapy after failure of TCZ | 3.00 | 1.93 | For |
| Add-on therapy after failure of MMF and CYC | 3.82 | 0.73 | For |
| Add-on therapy after failure of TCZ and CYC | 3.59 | 0.73 | For |
| Add-on therapy after failure of TCZ and MMF | 3.64 | 0.66 | For |
| Add-on therapy after failure of TCZ and  MMF and CYC | 3.64 | 0.58 | For |
| Add-on therapy to MMF/CYC | 3.77 | 0.81 | For |
| Add-on therapy to TCZ | 2.18 | 2.26 | No |
| **Use of TCZ** | | | |
| **I would use TCZ for treating patients with SSc-ILD under the following  clinical conditions:** | | | |
| Patients with early SSc and ILD with progressive skin disease | 1.45 | 2.13 | No |
| Patients with early SSc and ILD with anti-topoisomerase antibodies | 2.45 | 1.01 | No |
| Patients with early SSc and ILD with elevated acute-phase reactants | 3.18 | 0.96 | For |
| Any patient with early SSc and ILD | –0.23 | 2.18 | No |
| Patients with clinical ILD (according to symptoms, HRCT, FVC and/or DLCO) with active extrapulmonary manifestations | 0.32 | 2.10 | No |
| Patients with clinical ILD (according to symptoms, HRCT, FVC and/or DLCO) with quiescent extrapulmonary manifestations | 0.77 | 2.02 | No |
| Patients with progressive ILD despite antifibrotic therapy | 1.14 | 2.27 | No |
| Patients with progressive ILD despite immunosuppressive therapy (MMF/CYC) | 1.59 | 2.17 | No |
| Patients with progressive ILD in combination with MMF/CYC | 0.18 | 2.02 | No |
| Patients with progressive ILD in combination with antifibrotics | 1.82 | 2.63 | No |
| Patients who have contraindications to or are unable to tolerate immunosuppression (CYC/MMF) | 1.95 | 2.10 | No |
| Patients who have contraindications to or are unable to tolerate antifibrotics | 2.00 | 2.43 | No |
| In combination with immunosuppressive agents (MMF/CYC) for patients with aggressive ILD, advanced disease at initial presentation, or significant disease progression | 0.68 | 2.19 | No |
| In combination with antifibrotics for patients with aggressive ILD, advanced disease at initial presentation, or significant disease progression | 1.55 | 2.11 | No |
| Any patient with CTD with clinically significant or worsening ILD | –0.50 | 2.06 | No |
| Based on lack of effective response or improvement with immunosuppressive agents (MMF/CYC) or antifibrotics as defined by lack of symptom improvement | 0.64 | 2.13 | No |
| Based on lack of effective response or improvement with immunosuppressive agents (MMF/CYC) or antifibrotics as defined by lack of improvement of ILD on HRCT | 1.14 | 2.27 | No |
| Based on lack of effective response or improvement with immunosuppressive agents (MMF/CYC) or antifibrotics as defined by lack of improvement of lung function | 1.50 | 2.37 | No |
| Based on lack of effective response or improvement with immunosuppressive agents (MMF/CYC) or antifibrotics as defined by a combination of the above | 1.82 | 2.36 | No |
| Based on active worsening of patient condition as defined by worsening of symptoms | 0.73 | 1.83 | No |
| Based on active worsening of patient condition as defined by worsening of ILD on HRCT | 1.64 | 2.32 | No |
| Based on active worsening of patient condition as defined by worsening of lung function | 2.23 | 2.18 | No |
| Based on active worsening of patient condition as defined by a combination of the above | 2.36 | 2.22 | No |
| Based on inability to continue CYC/MMF/antifibrotics due to adverse effects | 2.59 | 1.97 | For |
| Based on inability to continue CYC/MMF/antifibrotics due to lack of achievement of effective dose with CYC/MMF/antifibrotics | 2.45 | 2.20 | No |
| Based on inability to continue CYC/MMF/antifibrotics due to prolonged use | 1.09 | 2.02 | No |
| **TCZ fits into the management of SSc-ILD as:** | | | |
| Initial monotherapy in patients with contraindication to or problems tolerating antifibrotics | –0.36 | 2.04 | No |
| Initial monotherapy in patients with contraindication to or problems tolerating immunosuppressive agents (CYC/MMF) | 1.68 | 2.03 | No |
| Initial monotherapy in patients with any SSc-ILD | –1.09 | 2.20 | No |
| Initial monotherapy in patients with early SSc and ILD | –0.05 | 2.08 | No |
| Dual initial therapy in combination with antifibrotics | –0.32 | 2.10 | No |
| Dual initial therapy in combination with MMF/CYC | –0.68 | 1.76 | No |
| Add-on therapy after failure of MMF as initial therapy, before changing MMF to CYC | 1.95 | 1.89 | No |
| Replacement therapy following failure of MMF as initial therapy, before changing to CYC | 2.14 | 2.03 | No |
| Add-on therapy after failure of CYC as initial therapy | 0.45 | 2.09 | No |
| Add-on therapy after failure of antifibrotics | 0.91 | 2.07 | No |
| Add-on therapy after failure of MMF and CYC | 1.64 | 1.97 | No |
| Add-on therapy after failure of MMF and antifibrotics | 1.64 | 1.97 | No |
| Add-on therapy after failure of CYC and antifibrotics | 1.32 | 1.99 | No |
| Add-on therapy after failure of MMF and CYC and antifibrotics | 1.59 | 2.13 | No |
| Add-on therapy to MMF/CYC | 0.91 | 1.87 | No |
| Add-on therapy to antifibrotics | 1.36 | 2.08 | No |
| **What is your typical/target dose for nintedanib?** | | | |
| I do not utilize nintedanib | –4.73 | 0.55 | Against |
| 100 mg twice daily | 0.50 | 2.24 | No |
| 150 mg twice daily | 4.68 | 0.57 | For |
| **What is your typical/target dose for TCZ?** | | | |
| I do not utilize TCZ | –3.36 | 1.89 | Against |
| 162 mg/week | 4.18 | 1.44 | For |
| **When do you screen for pulmonary hypertension in patients with SSc-ILD?** | | | |
| I do not screen for pulmonary hypertension | –4.77 | 0.53 | Against |
| At every patient consultation | –0.14 | 2.51 | No |
| Once per year | 4.41 | 0.85 | For |
| Every 2 years | 0.32 | 2.48 | No |
| I routinely screen for pulmonary hypertension | 4.59 | 0.73 | For |
| I screen for pulmonary hypertension in patients aged >50 years | 2.00 | 2.29 | No |
| I screen for pulmonary hypertension when shortness of breath is not explained by progression of ILD | 4.86 | 0.47 | For |

CTD, connective tissue disease; CYC, cyclophosphamide; DLCO, diffusing capacity of the lungs for carbon monoxide; FCV, forced vital capacity; HRCT, high-resolution computed tomography; ILD, interstitial lung disease; MMF, mycophenolate mofetil; SD, standard deviation; SSc, systemic sclerosis; TCZ, tocilizumab.