**Supplementary material**

**Table S1**. ICD codes used to classify ILDs in the analysis of claims data.

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| **ILD type** | **Classification based on ILD code alone** | **Classification based on systemic condition code and ILD code** |
| **ILD ICD-9 or ICD-10 code** | **Systemic condition ICD-9 or ICD-10 code** | **ILD ICD-9 or ICD-10 code** |
| SSc-ILD | 517.2: Lung involvement in systemic sclerosisM34.81: Systemic sclerosis with lung involvement | 710.1: Systemic sclerosisM34.0: Progressive systemic sclerosisM34.1: CR(E)ST syndromeM34.9: Systemic sclerosis, unspecifiedM34.89: Other systemic sclerosis | Any ILD code (see table below) |
| RA-ILD | 714.81: Rheumatoid lungM05.10: Rheumatoid lung disease with rheumatoid arthritis of unspecified site | 714.0: Rheumatoid arthritis714.1: Felty's syndrome714.2: Other rheumatoid arthritis with visceral or systemic involvement714.4: Chronic postrheumatic arthropathy714.89: Other specified inflammatory polyarthropathies714.9: Unspecified inflammatory polyarthropathiesM06.9: Rheumatoid arthritis, unspecifiedM05.00: Felty's syndrome, unspecified siteM05.30: Rheumatoid heart disease with rheumatoid arthritis of unspecified siteM05.60: Rheumatoid arthritis of unspecified site with involvement of other organs and systemsM12.00: Chronic postrheumatic arthropathy [Jaccoud], unspecified siteM06.4: Inflammatory polyarthropathyM06.09: Rheumatoid arthritis without rheumatoid factor, multiple sitesM05.79: Rheumatoid arthritis with rheumatoid factor of multiple sites without organ or systems involvement M05.89: Other rheumatoid arthritis with rheumatoid factor of multiple sitesM06.89: Other specified rheumatoid arthritis, multiple sitesM06.00: Rheumatoid arthritis without rheumatoid factor, unspecified siteM05.40: Rheumatoid myopathy with rheumatoid arthritis of unspecified siteM05.70: Rheumatoid arthritis with rheumatoid factor of unspecified site without organ or systems involvementM05.741: Rheumatoid arthritis with rheumatoid factor of right hand without organ or systems involvementM06.041: Rheumatoid arthritis without rheumatoid factor, right handM05.742: Rheumatoid arthritis with rheumatoid factor of left hand without organ or systems involvementM06.80: Other specified rheumatoid arthritis, unspecified siteM05.69: Rheumatoid arthritis of multiple sites with involvement of other organs and systemsM06.042: Rheumatoid arthritis without rheumatoid factor, left hand | Any ILD code (see table below) |
| Sjogren's syndrome ILD\* | n/a | 710.2: Sicca SyndromeM35.00: Sicca syndrome, unspecifiedM35.01: Sicca syndrome with keratoconjunctivitisM35.09: Sicca syndrome with other organ involvement | Any ILD code (see table below) |
| SLE-ILD\* | n/a | 710.0: Systemic lupus erythematosusM32.10: Systemic lupus erythematosus, organ or system involvement unspecifiedM32.19: Other organ or system involvement in systemic lupus erythematosus | Any ILD code (see table below) |
| PM/DM-ILD\* | n/a | 710.3: Dermatomyositis710.4: PolymyositisM33.90: Dermatopolymyositis, unspecified, organ involvement unspecifiedM33.20: Polymyositis, organ involvement unspecifiedM33.22: Polymyositis with myopathy | Any ILD code (see table below) |
| MCTD-ILD\* | n/a | 710.8: Other specified diffuse diseases of connective tissueM35.5: Multifocal fibrosclerosisM35.1: Other overlap syndromes | Any ILD code (see table below) |
| Non-specified CTD-ILD\* | n/a | 710.9: Unspecified connective tissue diseaseM35.9: Systemic involvement of connective tissue, unspecified | Any ILD code (see table below) |
| HP | 495.0: Farmer's lung495.1: Bagassosis495.2: Bird-fanciers' lung495.3: Suberosis495.4: Malt workers' lung495.5: Mushroom workers' lung495.6: Maple bark-strippers' lung495.7: Ventilation pneumonitis495.8: Other specified allergic alveolitis and pneumonitis495.9: Unspecified allergic alveolitis and pneumonitisJ67.0: Farmer's lungJ67.1: BagassosisJ67.2: Bird-fanciers' lungJ67.3: SuberosisJ67.4: Malt workers' lungJ67.5: Mushroom workers' lungJ67.6: Maple bark-strippers' lungJ67.7: Air conditioner and humidifier lungJ67.8: Hypersensitivity pneumonitis due to other organic dustsJ67.9: Hypersensitivity pneumonitis due to unspecified organic dust | n/a | n/a |
| Sarcoidosis ILD | D86.0: Sarcoidosis of the lungD86.2: Sarcoidosis of the lung with sarcoidosis of the lymph nodes | 135: SarcoidosisD86.9: Sarcoidosis, unspecifiedD86.89: Sarcoidosis of other sitesD86.3: Sarcoidosis of skinD86.1: Sarcoidosis of lymph nodes | 515: Postinflammatory pulmonary fibrosis517.8: Lung involvement in other diseases classified elsewhereJ84.10: Pulmonary fibrosis, unspecifiedJ84.89: Other specified interstitial pulmonary diseasesJ84.17: Other interstitial pulmonary diseases with fibrosis in diseases classified elsewhereJ99: Respiratory disorders in diseases classified elsewhere |
| iNSIP | 516.32: Idiopathic NSIPJ84.113: Idiopathic non-specific interstitial pneumonitis | n/a | n/a |
| Other specified non-IPF ILDs | 516.30: Idiopathic interstitial pneumonia, not otherwise specified516.33: Acute interstitial pneumonitis516.34: Respiratory bronchiolitis interstitial lung disease516.35: Idiopathic lymphoid interstitial pneumonia516.36: Cryptogenic organizing pneumonia516.37: Desquamative interstitial pneumonia516.4: Lymphangioleiomyomatosis516.5: Adult pulmonary Langerhans cell histiocytosisJ84.111: Idiopathic interstitial pneumonia, not otherwise specifiedJ84.114: Acute interstitial pneumonitisJ84.115: Respiratory bronchiolitis interstitial lung diseaseJ84.2: Lymphoid interstitial pneumoniaJ84.116: Cryptogenic organizing pneumoniaJ84.117: Desquamative interstitial pneumoniaJ84.81: LymphangioleiomyomatosisJ84.82: Adult pulmonary Langerhans cell histiocytosis500: Coal workers' pneumoconiosis501: Asbestosis502: Pneumoconiosis due to other silica or silicates503: Pneumoconiosis due to other inorganic dust504: Pneumonopathy due to inhalation of other dust505: Pneumoconiosis, unspecified506.0: Bronchitis and pneumonitis due to fumes and vapors506.3: Other acute and subacute respiratory conditions due to fumes and vapors 506.4: Chronic respiratory conditions due to fumes and vapors508.0: Acute pulmonary manifestations due to radiation508.1: Chronic and other pulmonary manifestations due to radiation508.8: Respiratory conditions due to other specified external agents508.9: Respiratory conditions due to unspecified external agentJ60: Coalworker pneumoconiosisJ61: Pneumoconiosis due to asbestos and other mineral fibresJ62.8: Pneumoconiosis due to other dust containing silica J63.0: Aluminosis (of lung) J63.1: Bauxite fibrosis (of lung) J63.2: BerylliosisJ63.3: Graphite fibrosis (of lung)J63.4: SiderosisJ63.5: StannosisJ63.6: Pneumoconiosis due to other specified inorganic dustsJ66.0: ByssinosisJ66.1: Flax-dresser diseaseJ66.2: CannabinosisJ64: Unspecified pneumoconiosisJ68.0: Bronchitis and pneumonitis due to chemicals, gases, fumes and vapoursJ68.3: Other acute and subacute respiratory conditions due to chemicals, gases, fumes and vapoursJ68.4: Chronic respiratory conditions due to chemicals, gases, fumes and vapoursJ70.0: Acute pulmonary manifestations due to radiationJ70.1: Chronic and other pulmonary manifestations due to radiation J70.8: Respiratory conditions due to other specified external agentsJ70.9: Respiratory conditions due to unspecified external agentJ92.0: Pleural plaque with presence of asbestosJ62.0: Pneumoconiosis due to talc dustJ65: Pneumoconiosis associated with tuberculosis | n/a | n/a |
| Non-specified ILDs | 517.8: Lung involvement in other diseases classified elsewhere515: Postinflammatory pulmonary fibrosis516.8: Other specified alveolar and parietoalveolar pneumonopathies516.9: Unspecified alveolar and parietoalveolar pneumonopathyJ99: Respiratory disorders in diseases classified elsewhereJ84.10: Pulmonary fibrosis, unspecifiedJ84.89: Other specified interstitial pulmonary diseasesJ84.17: Other interstitial pulmonary diseases with fibrosis in diseases classified elsewhereJ84.09: Other alveolar and parieto-alveolar conditionsJ84.9: Interstitial pulmonary disease, unspecified | n/a | n/a |

Patients were classified based on the ILD code only if the code specified the ILD subtype. If patients had one of the ILD codes and a systemic condition code, they were classified as having ILD associated with that systemic condition. If a patient had claims with ILD or systemic codes that would meet the criteria for multiple subtypes, the patient was categorized based on the hierarchy in the subtype classification table under the ILD subtype highest on the list.

\*Sjogren's syndrome ILD, SLE-ILD, PM/DM-ILD, MCTD-ILD, and non-specified CTD-ILDs were grouped as ‘other CTD-ILDs’ in the results.

CR(E)ST syndrome: combination of calcinosis, Raynaud phenomenon, esophageal dysfunction, sclerodactyly, telangiectasia.

DM, dermatomyositis; MCTD, mixed connective tissue disease; PM, polymyositis; SLE, systemic lupus erythematous.

**Table S2.** ICD-9 and ICD-10 ILD codes.

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| **ICD-9 codes** | **ICD-10 codes** |
| 500: Coal Workers' Pneumoconiosis | 495.7: Ventilation pneumonitis | J60: Coal Workers' Pneumoconiosis | J70.9: Respiratory conditions due to unspecified external agent | J84.9: Interstitial pulmonary disease, unspecified | M05.142: RLD with RA in left hand  |
| 501: Asbestosis | 495.8: Other specified allergic alveolitis and pneumonitis | J61: Pneumoconiosis due to asbestos and other mineral fibers  | J67.0: Farmer's lung | J84.114: Acute interstitial pneumonitis | M05.149: RLD with RA in unspecified hand |
| 502: Pneumoconiosis due to silica | 495.9: Unspecified allergic alveolitis and pneumonitis | J62.8: Pneumoconiosis due to other dust containing silica | J67.1: Bagassosis | J84.115: Respiratory bronchiolitis interstitial lung disease | M05.151: RLD with RA in right hip  |
| 503: Pneumoconiosis due to other inorganic dust | 714.81: Rheumatoid Lung | J63.0: Aluminosis (of lung) | J67.2: Bird-fanciers' lung | J84.2: Lymphoid interstitial pneumonia | M05.152: RLD RA in left hip  |
| 504: Pneumoconiosis due to the inhalation of other dust | 517.2: Lung involvement in systemic sclerosis | J63.1: Bauxite fibrosis (of lung) | J67.3: Suberosis | J84.116: Cryptogenic organizing pneumonia | M05.159: RLD with RA in unspecified hip  |
| 505: Pneumoconiosis, unspecified | 517.8: Lung involvement in other diseases classified elsewhere  | J63.2: Berylliosis | J67.4: Malt workers' lung | J84.117: Desquamative interstitial pneumonia | M05.161: RLD with RA in right knee  |
| 506.0: Bronchitis and pneumonitis due to fumes and vapors | 515: Postinflammatory pulmonary fibrosis | J63.3: Graphite fibrosis (of lung) | J67.5: Mushroom workers' lung | J84.81: Lymphangioleiomyomatosis | M05.162: RLD with RA in left knee  |
| 506.3: Other acute and subacute respiratory conditions due to fumes and vapors | 516.30: Idiopathic interstitial pneumonia, NOS | J63.4: Siderosis | J67.6: Maple bark-strippers' lung | J84.82: Adult pulmonary Langerhans cell histiocytosis | M05.169: RLD with RA in unspecified knee  |
| 506.4: Chronic respiratory conditions due to fumes and vapors | 516.31: Idiopathic pulmonary fibrosis | J63.5: Stannosis | J67.7: Air conditioner and humidifier lung | J92.0: Pleural plaque with asbestosis  | M05.171: RLD with RA in right ankle and foot |
| 508.0: Acute pulmonary manifestations due to radiation  | 516.32: Idiopathic NSIP | J63.6: Pneumoconiosis due to other specified inorganic dusts | J67.8: Hypersensitivity pneumonitis due to other organic dusts | J62.0: Pneumoconiosis due to other talc dust | M05.172: RLD with RA in left ankle and foot  |
| 508.1: Chronic and other pulmonary manifestations due to radiation | 516.8: Other specified alveolar and parietoalveolar pneumonopathies | J66.0: Byssinosis | J67.9: Hypersensitivity pneumonitis due to unspecified organic dust | J65: Pneumoconiosis associated with tuberculosis | M05.179: RLD with RA in unspecified ankle and foot  |
| 508.8: Respiratory conditions due to other specified external agents | 516.9: Unspecified alveolar and parietoalveolar pneumonopathy | J66.1: Flax-dressers' disease | M05.10: RLD with RA of unspecified site | M05.111: RLD with RA in right shoulder  | M05.19: RLD with RA of multiple sights |
| 508.9: Respiratory conditions due to unspecified external agents | 516.33: Acute interstitial pneumonitis | J66.2: Cannabinosis | M34.81: Systemic sclerosis with lung involvement | M05.112: RLD with RA in left shoulder  | M32.13: Lung involvement in systemic lupus erythematosus |
| 495.0: Farmer's lung | 516.34: Respiratory bronchiolitis interstitial lung disease | J66.8: Airway disease due to other specific organic dusts | J99: Respiratory disorders in diseases classified elsewhere | M05.119: RLD with RA in unspecified shoulder  | M35.02: Sicca syndrome with lung involvement  |
| 495.1: Bagassosis | 516.35: Idiopathic lymphoid interstitial pneumonia | J64: Unspecified pneumoconiosis | J84.10: Pulmonary fibrosis, unspecified | M05.121: RLD with RA in right elbow  | M33.01: Juvenile dermatopolymyositis with respiratory involvement |
| 495.2: Bird-fanciers' lung | 516.36: Cryptogenic organizing pneumonia | J68.0: Bronchitis and pneumonitis due to chemicals, gases, fumes and vapors | J84.89: Other specified interstitial pulmonary diseases | M05.122: RLD with RA in left elbow  | M33.11: Other dermatopolymositis, with respiratory involvement  |
| 495.3: Suberosis | 516.37: Desquamative interstitial pneumonia | J68.3: Other acute and subacute respiratory conditions due to chemicals, gases, fumes and vapors | J84.17: Other interstitial pulmonary diseases with fibrosis in diseases classified elsewhere | M05.129: RLD with RA in unspecified elbow  | M33.21: Polymyositis, with respiratory involvement |
| 495.4: Malt workers' lung | 516.4: Lymphangioleiomyomatosis | J68.4: Chronic respiratory conditions due to chemicals, gases, fumes and vapors | J84.111: Idiopathic interstitial pneumonia, not otherwise specified | M05.131: RLD with RA in right wrist  | M33.91: Dermatomyositis, unspecified, with respiratory involvement |
| 495.5: Mushroom workers' lung | 516.5: Adult pulmonary Langerhans cell histiocytosis | J70.0: Acute pulmonary manifestations due to radiation | J84.112: Idiopathic pulmonary fibrosis | M05.132: RLD with RA in left wrist | D86.0: Sarcoidosis of the lung  |
| 495.6: Maple bark-strippers' lung |  | J70.1: Chronic and other pulmonary manifestations due to radiation | J84.113: Idiopathic non-specific interstitial pneumonitis | M05.139: RLD with RA in unspecified wrist  | D86.2: Sarcoidosis of the lung with sarcoidosis of the lymph nodes |
|  |  | J70.8: Respiratory conditions due to other specified external agents | J84.09: Other alveolar and parieto-alveolar conditions | M05.141: RLD with RA in right hand  |  |

RLD, rheumatoid lung disease.

**Figure S1. Specialty that filed first ILD insurance claim.** Data based on US insurance claims from 36,579 patients who had ILD (defined as ≥2 claims with an ILD diagnosis and ≥1 visit to a pulmonologist between 2014 and 2016), whose first ILD claim was made in 2015 or 2016 and who had a claim for any indication ≤180 days prior to the first ILD claim.



**Figure S2. Frequency of follow-up visits and HRCT scans in patients with ILDs.** Data from online survey of physicians (pulmonologists, n=243; rheumatologists, n=203; internists, n=40). Survey questions: *In patients with ILD where you manage / help manage the ILD, on average how frequently do you have follow-up visits with the patient? In patients with ILD where you manage / help manage the ILD, on average how frequently do you check the status of the patients’ ILD with HRCT?*

 

**Figure S3. Number of visits to a pulmonologist or rheumatologist for ILD in 2014.** Data based on US insurance claims from 30,090 patients who had ILD (defined as ≥2 claims with an ILD diagnosis and ≥1 visit to a pulmonologist between 2014 and 2016) and had ≥1 ILD claim in each of 2014, 2015 and 2016, and had ≥1 visit to a pulmonologist or rheumatologist, respectively, between 2014 and 2016.



**Figure S4. Specialties that led treatment decisions for patients with progressive fibrosing ILDs.** Data from online survey of physicians (pulmonologists, n=243; rheumatologists, n=203; internists, n=40). Survey question: *In your estimation, for patients with a non-IPF ILD that has fibrosis and is progressing, in what percentage of patients do the following specialties lead the decision-making on how to treat (i.e. drug treatment) the ILD?* Examples of autoimmune ILDs provided to physicians participating in the survey were RA-ILD, SSc-ILD, and other CTD-ILDs. Examples of non-IPF non-autoimmune ILDs provided were iNSIP, HP, and sarcoidosis-ILD. Rheumatologists were only asked this question in relation to autoimmune ILDs.



**Figure S5. Action taken by pulmonologists managing a patient with a non-IPF progressive fibrosing ILD.** Data from online survey of 243 pulmonologists. Survey question: *What do you typically do if you determine that a patient you are managing has a non-IPF non-autoimmune / autoimmune ILD that has fibrosis and is progressing?* Examples of autoimmune ILDs provided were RA-ILD, SSc-ILD, and other CTD-ILDs. Examples of non-IPF non-autoimmune ILDs provided were iNSIP, HP, and sarcoidosis-ILD.



**Figure S6. Action taken by rheumatologists managing a patient with a non-IPF progressive fibrosing ILD.** Data from online survey of 203 rheumatologists. Survey question: *What do you typically do if you determine that a patient you are managing has an autoimmune ILD that has fibrosis and is progressing?* Examples of autoimmune ILDs provided were RA-ILD, SSc-ILD, and other CTD-ILDs.



**Figure S7.** **Agents used as first-, second- and third-line treatments for fibrotic autoimmune ILDs by region.** Data from online survey of physicians. Survey question: *For the following types of ILDs where patients also have lung fibrosis, please indicate your preferred first, second, and third line treatments for the respective ILD.*

**Physicians in the US:**



**Physicians in Europe (France, Germany, Italy, Spain, UK):**



**Physicians in Japan:**

