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Figure E1 Search Strategies for EMBASE

Overall ILD or cryptogenic fibrosing alveolitis search term

epidemiology.mp.

**epidemiology/*

**public health/*

1 or 2 or 3

incidence.mp.

prevalence.mp.

**mortality/ or *mortality risk/ or *mortality rate/*

**prevalence/ or epidemiological data/ or epidemiology/ or statistical concepts/ or statistical parameters/*

5 or 6 or 7 or 8

4 or 9

ILD.mp.

**interstitial lung disease/ or interstitial lung diseases*.mp.

11 or 12

10 and 13

limit 14 to human

limit 15 to english language

limit 16 to yr="2009 - 2019"

Asbestosis or Silicosis search terms

epidemiology.mp.
**epidemiology*/
**public health*/
1 or 2 or 3
incidence.mp.
prevalence.mp.
**mortality*/ or **mortality risk*/ or **mortality rate*/
**prevalence*/ or *epidemiological data*/ or *epidemiology*/ or *statistical concepts*/ or *statistical parameters*/
5 or 6 or 7 or 8
4 or 9
asbestosis.mp. or **asbestosis*/
silicosis.mp. or **silicosis*/
**asbestosis*/ or **silicosis*/
11 or 12 or 13
10 and 14
limit 15 to human
limit 16 to english language
limit 17 to yr="2009 - 2019"

Pneumoconiosis search term

epidemiology.mp.
**epidemiology*/
**public health*/
1 or 2 or 3
incidence.mp.
prevalence.mp.
**mortality*/ or **mortality risk*/ or **mortality rate*/
**prevalence*/ or *epidemiological data*/ or *epidemiology*/ or *statistical concepts*/ or *statistical parameters*/
5 or 6 or 7 or 8
4 or 9
ILD.mp.
**interstitial lung disease*/ or *interstitial lung diseases*.mp.
11 or 12
10 and 13
pneumoconiosis.mp. or *exp pneumoconiosis*/
10 and 15
limit 16 to human
limit 17 to english language
limit 18 to yr="2009 - 2019"

Respiratory bronchiolitis search term

epidemiology.mp.
**epidemiology*/

**public health/
1 or 2 or 3
inciden*.mp.
prevalen*.mp.
*mortality/ or *mortality risk/ or *mortality rate/
*prevalence/ or epidemiological data/ or epidemiology/ or statistical concepts/ or statistical parameters/
5 or 6 or 7 or 8
4 or 9
ILD*.mp.
interstitial lung disease/ or interstitial lung diseas.mp.
11 or 12
10 and 13
exp bronchiolitis/ or Respiratory bronchiolitis.mp.
10 and 15
limit 16 to human
limit 17 to english language
limit 18 to yr="2009 - 2019"*

Hypersensitivity pneumonitis or Extrinsic alveolar alveolitis search terms

epidemiology.mp.
*epidemiology/
*public health/
1 or 2 or 3
inciden*.mp.
prevalen*.mp.
*mortality/ or *mortality risk/ or *mortality rate/
*prevalence/ or epidemiological data/ or epidemiology/ or statistical concepts/ or statistical parameters/
5 or 6 or 7 or 8
hypersensitivity pneumonitis.mp. or exp allergic pneumonitis/
*hypersensitivity pneumonitis/ or *extrinsic alveolar alveolitis/ or *EAA/ or *HP/
10 or 11
4 or 9
12 and 13
limit 14 to human
limit 15 to english language
limit 16 to yr="2009 - 2019"*

Cystic lung disease

epidemiology.mp.
*epidemiology/
*public health/
1 or 2 or 3
inciden*.mp.*

prevalen.mp.*

**mortality/ or *mortality risk/ or *mortality rate/*

**prevalence/ or epidemiological data/ or epidemiology/ or statistical concepts/ or statistical parameters/*

5 or 6 or 7 or 8

4 or 9

cystic lung diseas.mp.*

cystic lung diseas.mp. or exp lymphangiomyomatosis/ or exp Birt Hogg Dube syndrome/*

**birt hogg dube/ or *LAM/ or *PLCH/ or *LIP/ or *lymphangiomyomatosis/ or *pulmonary Langerhans cell histiocytosis/ or *lymphoid interstitial pneumonia/ or*

**Birt-Hogg-Dube/*

11 or 12 or 13

10 and 14

limit 15 to human

limit 16 to english language

limit 17 to yr="2009 - 2019"

Sarcoidosis search term

epidemi.mp.*

**epidemiology/*

**public health/*

1 or 2 or 3

inciden.mp.*

prevalen.mp.*

**mortality/ or *mortality risk/ or *mortality rate/*

**prevalence/ or epidemiological data/ or epidemiology/ or statistical concepts/ or statistical parameters/*

5 or 6 or 7 or 8

4 or 9

**lung sarcoidosis/ or *sarcoidosis/ or sarcoidosis*.mp.*

10 and 11

limit 12 to human

limit 13 to english language

limit 14 to yr="2009 - 2019"

Idiopathic interstitial lung disease

epidemi.mp.*

**epidemiology/*

**public health/*

1 or 2 or 3

inciden.mp.*

prevalen.mp.*

**mortality/ or *mortality risk/ or *mortality rate/*

**prevalence/ or epidemiological data/ or epidemiology/ or statistical concepts/ or statistical parameters/*

5 or 6 or 7 or 8

4 or 9

idiopathic interstitial pneumon.mp.*

**idiopathic interstitial pneumonia/ or *IIP/ or *idiopathic interstitial/*

**idiopathic interstitial pneumonia/ or idiopathic interstitial pneumon*.mp. or *Idiopathic Interstitial Pneumonias/*

11 or 12 or 13

10 and 14

limit 15 to human

limit 16 to english language

limit 17 to yr="2009 - 2019"

Idiopathic pulmonary fibrosis

epidemi.mp.*

**epidemiology/*

**public health/*

1 or 2 or 3

inciden.mp.*

prevalen.mp.*

**mortality/ or *mortality risk/ or *mortality rate/*

**prevalence/ or epidemiological data/ or epidemiology/ or statistical concepts/ or statistical parameters/*

5 or 6 or 7 or 8

4 or 9

**idiopathic pulmonary fibrosis/*

**lung fibrosis/ or *fibrosing alveolitis/ or idiopathic pulmonary fibrosis*.mp.*

11 or 12

10 and 13

limit 14 to human

limit 15 to english language

limit 16 to yr="2009 - 2019"

Autoimmune ILDs

epidemi.mp.*

**epidemiology/*

**public health/*

1 or 2 or 3

inciden.mp.*

prevalen.mp.*

**mortality/ or *mortality risk/ or *mortality rate/*

**prevalence/ or epidemiological data/ or epidemiology/ or statistical concepts/ or statistical parameters/*

5 or 6 or 7 or 8

4 or 9

ILD.mp.*

**interstitial lung disease/ or interstitial lung diseas*.mp.*

11 or 12

10 and 13

exp autoimmune disease/ or autoimmune.mp.*

**autoimmune/ or *RA/ or *Rheumatoid arthritis/ or *rheumatoid arthritis/ or *scleroderma/ or *Scleroderma/ or *systemic sclerosis/ or *Systemic Sclerosis/*

15 or 16

10 and 13 and 17

limit 18 to human

limit 19 to english language

limit 20 to yr="2009 - 2019"

Other ILDs

epidemiology.mp.*

**epidemiology/*

**public health/*

1 or 2 or 3

incidence.mp.*

prevalence.mp.*

**mortality/ or *mortality risk/ or *mortality rate/*

**prevalence/ or epidemiological data/ or epidemiology/ or statistical concepts/ or statistical parameters/*

5 or 6 or 7 or 8

4 or 9

ILD.mp.*

**interstitial lung disease/ or interstitial lung disease*.mp.*

11 or 12

cryptogenic organizing pneumonia.mp.

exp bronchiolitis obliterans organizing pneumonia/ or exp cryptogenic organizing pneumonia/ or cryptogenic organizing pneumonia.mp.*

exp bronchiolitis obliterans organizing pneumonia/ or exp bronchiolitis obliterans/ or bronchiolitis obliterans.mp.*

exp antidepressant agent/ or antidepressive.mp.*

exp hyperthermic intraperitoneal chemotherapy/ or exp maintenance chemotherapy/ or exp chemotherapy/ or exp induction chemotherapy/ or exp cancer chemotherapy/ or exp adjuvant chemotherapy/ or Chemotherapy.mp. or neoadjuvant chemotherapy/ or exp combination chemotherapy/*

**Amiodarone/ or *Fluoxetine/ or *Nitrofurantoin/ or *amiodarone/ or *fluoxetine/ or *nitrofurantoin/*

14 or 15 or 16 or 17 or 18 or 19

10 and 13 and 20

limit 21 to human

limit 22 to english language

limit 23 to yr="2009 - 2019"

Figure E2: ILD Aetiological Classification

ILD Subgroups

Individual ILDs

| | |
|---|---|
| Occupational or Exposure related ILD | Asbestosis or silicosis |
| | Pneumoconiosis |
| | Respiratory bronchiolitis (RB-ILD) and other smoking related ILD |
| | Hypersensitivity pneumonitis (HP) |
| Idiopathic interstitial pneumonia | Idiopathic pulmonary fibrosis (IPF) |
| | Idiopathic non-specific interstitial pneumonia (NSIP) |
| | Desquamative interstitial pneumonia (DIP) |
| | Cryptogenic organizing pneumonia (COP) |
| Autoimmune ILD | Rheumatoid arthritis associated ILD (RA-ILD) |
| | Systemic sclerosis associated ILD (SSc-ILD) |
| | Sjogren's syndrome associated ILD (SS ILD) |
| | Inflammatory myositis associated ILDs and anti-synthetase syndromes |
| | Other autoimmune ILDs |
| Pulmonary sarcoidosis | Pulmonary sarcoidosis |
| Unclassifiable ILD | Unclassifiable ILD |
| Cystic lung disease | Birt Hogg-Dube (BHD) |
| | Lymphangioleiomyomatosis (LAM) |
| | Pulmonary Langerhans cell histiocytosis (PLCH) |
| | Lymphoid interstitial pneumonia (LIP) |
| Other ILDs | Obliterative bronchiolitis |
| | Drug induced ILD (eg. Amiodarone, Nitrofurantoin, Fluoxetine, Chemotherapy induced) |
| | Radiation induced ILD |

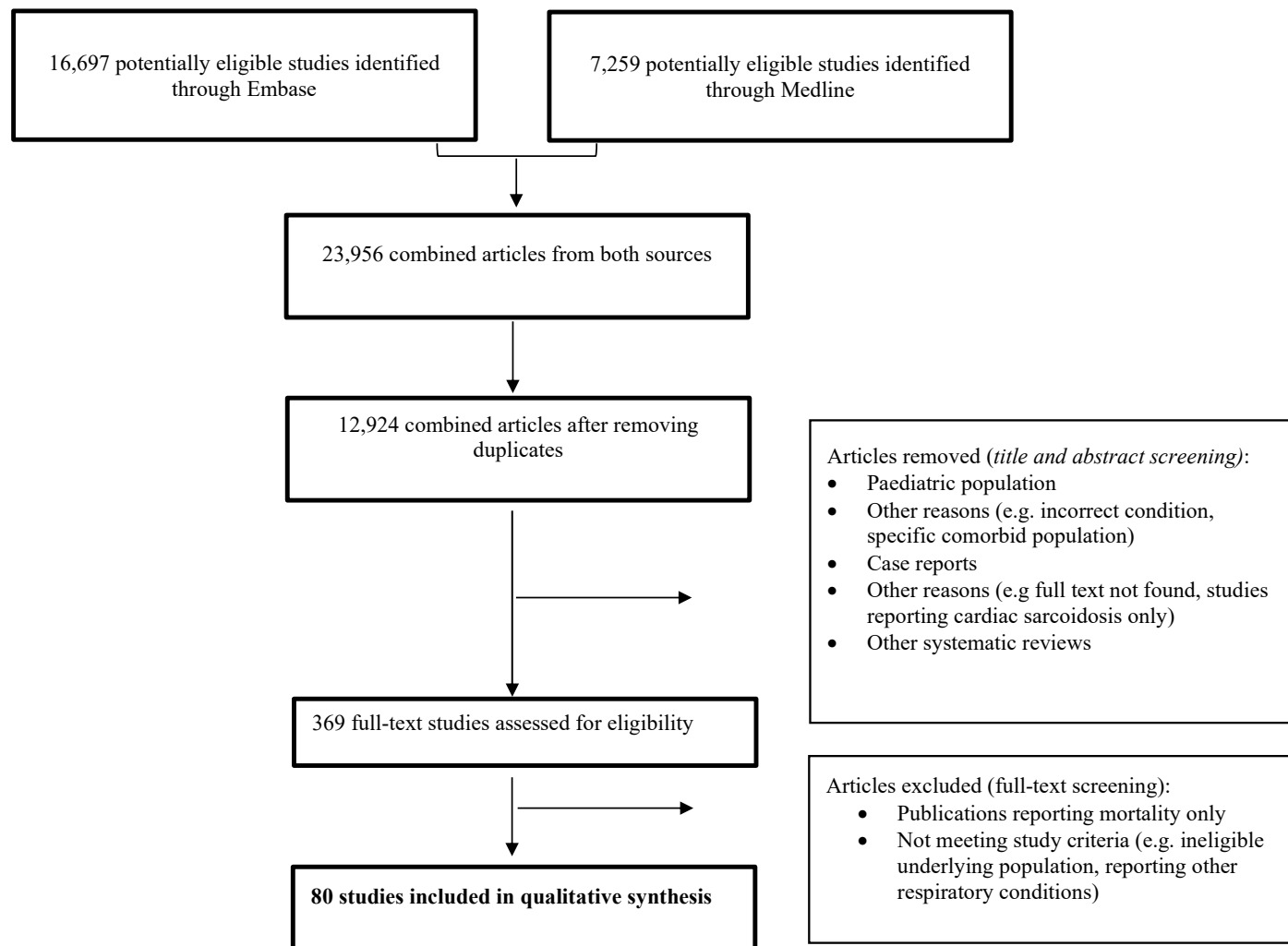
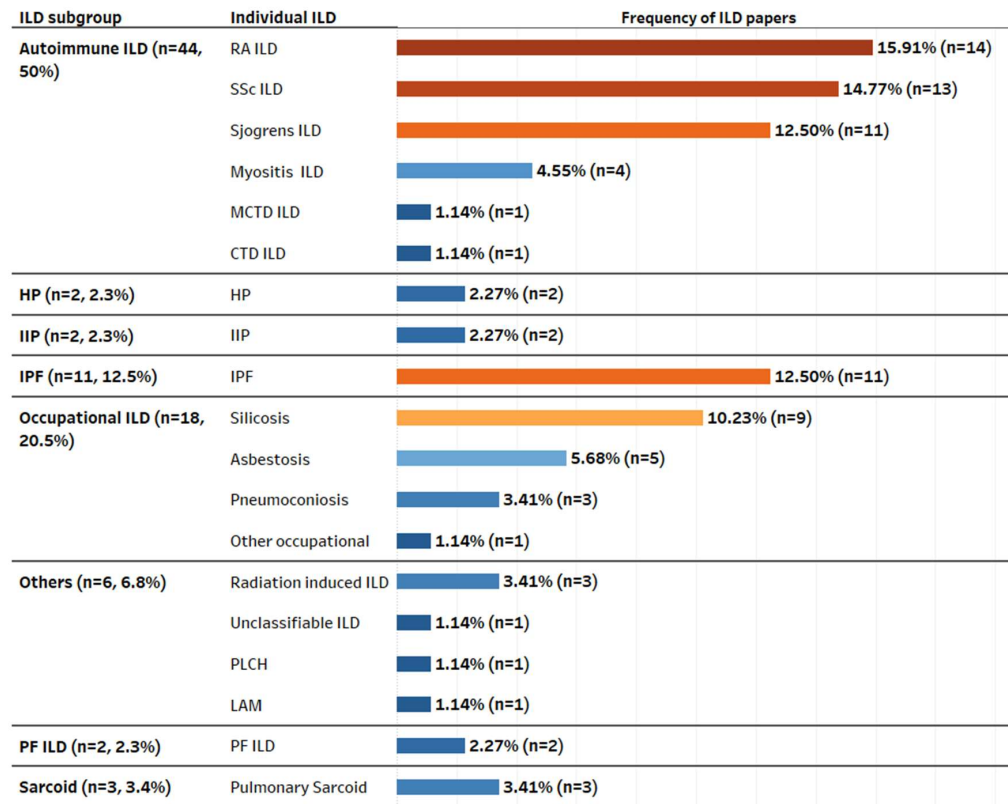
Figure E3 PRISMA Flow Diagram to Demonstrate Studies Identified, Screened And Included

Figure E4 Distribution of Included Publications by ILD Subgroups and Individual Disorder Type



RA ILD, rheumatoid arthritis; SSc, systemic sclerosis; MCTD, mixed connective tissue disorder; CTD, connective tissue disorder; HP, hypersensitivity pneumonitis; IIP, idiopathic interstitial pneumonia; IPF, idiopathic pulmonary fibrosis; PLCH, pulmonary Langerhans cell histiocytosis; LAM, lymphangioleiomyomatosis; PF, progressive fibrosing

Figure E5 Publications reporting non-IPF-ILD prevalence per 100,000 persons

| ILD condition | Country | Author, Year | Prevalence per 100,000 persons |
|------------------------------|---------------|---------------|--------------------------------|
| SSc ILD | Canada | Pope 2021 | 2.3 |
| | USA | Li 2021 | 19 |
| RA ILD | USA | Raimundo 2019 | 6 |
| | | Sparks 2021* | 2 |
| Progressive fibrosing ILD | France | Nasser 2021 | 19.4 |
| | USA | Olson 2021** | 70.3 |
| HP | United States | Perez 2018 | 2.7 |
| IIP | South Korea | Lee 2016 | 81.86 |
| Silicosis | USA | Casey 2019† | 20.1 |

SSc ILD, Systemic sclerosis ILD; RA ILD, Rheumatoid arthritis ILD; HP, Hypersensitivity pneumonitis; MCTD, mixed connective tissue disorder; IIP, Idiopathic interstitial pneumonia; USA, United States of America

*Unit of reporting is percentage of the study population (%); **Age and sex adjusted estimate; † Narrow silicosis definition used: Medicare beneficiaries with any claim that included ICD-9-CM code 502, pneumoconiosis due to other silica or silicates, listed in any position during 1999-2014, with at least one inpatient, skilled nursing or home health agency claim, or at least two outpatient provider claims within 365 days of each other and cases with a chest X-ray or CT scan 30 days before or 30 days after a silicosis claim.

Tables E1-E31: Characteristics of Included Studies (n=88)

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Glossary Of Abbreviations And Definition Of Terms

| | |
|------------------|--|
| AHP | Acute hypersensitivity pneumonitis |
| ATS/ERS | American Thoracic Society /European Respiratory Society |
| ATS/ERS/JRS/ALAT | American thoracic society/ European respiratory society/ Japanese Respiratory Society/ Latin American Thoracic Society |
| CHP | Chronic hypersensitivity pneumonitis |
| CIHI | Canadian Institute for Health Information |
| CLD | Cystic lung disease |
| CPRD | Clinical practice research datalink |
| CS | Cross sectional study design |
| CT | Computed tomography |
| CXR | Chest x-ray |
| DAD | Discharge Abstract Database |
| DLCO | Diffusing capacity of the lungs for carbon monoxide |
| dcSSc | Diffuse cutaneous systemic sclerosis |
| FEV | Forced expiratory volume |
| FHP | Fibrotic hypersensitivity pneumonitis |
| FLD | Farmers lung disease |
| FVC | Forced vital capacity |
| HIRA | Health Insurance Review and Assessment Service |
| HP | Hypersensitivity pneumonitis |
| HRCT | High-resolution computed tomography |
| ICD | International code for diagnosis |
| IIP | Idiopathic interstitial pneumonia |
| ILD | Interstitial lung disease |
| ILO | International labor organization |
| IPF | Idiopathic pulmonary fibrosis |
| LAM | Lymphangioleiomyomatosis |
| lcSSc | Limited cutaneous systemic sclerosis |
| MDD | Multidisciplinary discussion |
| NACRS | National Ambulatory Care Reporting System |
| NIH | National insurance of health |
| NR | Not reported |
| NSIP | Non-specific interstitial idiopathic pneumonia |
| PFT | Pulmonary function tests |
| PH | Pulmonary hypertension |
| PLCH | Pulmonary Langerhans cell histiocytosis |
| Prosp | Prospective study design |
| py | Person-years |
| RA | Rheumatoid arthritis |
| RID | Rare and Intractable Diseases |
| Rtr | Retrospective study design |
| SLB | Surgical lung biopsy |

| | |
|-------|---|
| pSS | Primary Sjogren's syndrome |
| sSS | Secondary Sjogren's syndrome |
| SSc | Systemic sclerosis |
| ssSSc | Sine scleroderma |
| TLC | Total lung capacity |
| TLB | Transbronchial biopsy |
| UIP | Usual interstitial idiopathic pneumonia |
| UK | United Kingdom |
| USA | United States of America |

Table E1: List of Publications Reporting Prevalence of IPF, Sorted By Publication Year

| Author, year | Study design | Country | Study population | ILD assessment | Sample size | No. of IPF cases by ILD definition | Mean age; Female (%) | Reported Prevalence (per 100,000 persons) |
|----------------|--------------|---------|--|--|--------------------|--|------------------------|--|
| Kaunisto 2015 | CS | Finland | University hospital | <ul style="list-style-type: none"> ICD codes J84.1 and J84.9 Pulmonary physician re-evaluated the patients 'data by reading through the patient charts.70–80 % of patients 'diagnoses did not meet the clinical criteria of idiopathic pulmonary fibrosis. Baseline CT scans were centrally re-evaluated by chest radiologist and radiology resident. HRCT scans were evaluated and SLB was performed in 27 (22 %) patients. | NR | 111 | 74;40 | 8.6 |
| Lee 2016 | Rtr | Korea | HIRA database | Patients with IPF were defined as those with a disease code of K-J84.18. | 51,026,868 | 2010, 2011, 2012, 2013;16,325 | NR | 30.4,30.1,32.4,35 |
| Harari 2016 | Rtr | Italy | Healthcare administrative database of Lombardy | <u>GCD</u> : Defined as IPF cases all individuals with at least one hospitalization with diagnosis of IPF or at least one outpatient visits with diagnosis of IPF (ICD-9-CM code 516.3) during the study period | Approx. 10 million | 5,441 | Majority >65 years; 46 | 35.51 (35.02–36.00) |
| | | | | <u>BCD</u> : Defined IPF cases those patients that satisfied the GCD and had no claims (inpatient or outpatient) with a diagnosis code for any other type of ILDs on or after the date of the last claim with IPF diagnosis | | 3,573 | Majority >65 years; 43 | 22.39 (21.9-22.8) |
| | | | | <u>NCD</u> : Defined IPF cases those patients that satisfied the BCD and had one or more claim with a procedure code for surgical lung biopsy, transbronchial lung biopsy or computed tomography of the thorax, on or before the date of the last claim with a diagnosis code for IPF. | | 2,097 | Majority >65 years; 43 | 12.55 (12.3-12.8) |
| Hopkins 2016 | Rtr | Canada | National databases: DAD and NACRS | <u>Broad definition</u> : by excluding cases with an ICD-10 CA code for another ILD after the J84.1 code, like other studies | NR | 14,259 | NR | 41.8 |
| | | | | <u>Narrow definition</u> further excluded cases that did not have chest CT, bronchus or lung biopsy, or bronchoscopy prior to establishing a J84.1 code. | | 6,390 | NR | 20 |
| Raghu 2016 | Rtr | USA | Optum Clinformatics | <u>Primary cohort</u> : IPF patients were first identified if they had at least one claim with ICD-9-CM diagnosis code 516.3 (idiopathic fibrosing alveolitis, which is used by providers to identify IPF) during the study period. Two subgroups of the primary cohort were further identified by applying additional criteria to form more restrictive cohorts of prevalent IPF patients: | 9,375,020 | 2005, 2006, 2007, 2008, 2009,2010; 7,671 | NR | 15.1, 16.9, 17.5, 18.4, 18.2, 13.4 |
| | | | | <u>Broad case definition</u> excluded patients if they had a claim with the diagnosis code 515 after the last diagnosis code 516.3. | | 2005, 2006, 2007, 2008, 2009,2010; 2,670 | NR | 8.4, 9.7, 10.6, 10.8, 11.6, 11.3 |
| | | | | <u>Narrow case definition</u> further restricted the broad case subgroup patients by requiring a claim for a surgical lung biopsy, transbronchial lung biopsy, or computed tomography scan of the thorax prior to the last diagnosis code 516.3. | | 2005, 2006, 2007, 2008, 2009,2010; 1,685 | NR | 4.6, 5.6, 6.1, 6.2, 6.9, 6.7 |
| Raimundo 2016 | CS | USA | Optum Humedica | Patients with at least one IPF inpatient claim, or two IPF outpatient claims with [ICD-9-CM] code 516.3 anytime in that calendar year (with no other ILD claim) | 4,138,796 | 2009, 2010, 2011; 1,191, 1292, 1136 | 70-71;49-52 | 28.8, 28.1, 19.8 |
| Strongman 2018 | Rtr | UK | CPRD | <u>Broad IPF -clinical syndrome</u> definition included the following three additional Read codes: H563100 (Diffuse pulmonary fibrosis), H563200 (Pulmonary fibrosis), and H563.11 (Hamman-Rich syndrome). | 9,748,108 | 2010 to 2013;4,527 | NR | 19.94 (18.48, 21.47) to 38.82 (37.04, 40.66) |

| Author, year | Study design | Country | Study population | ILD assessment | Sample size | No. of IPF cases by ILD definition | Mean age; Female (%) | Reported Prevalence (per 100,000 persons) |
|--------------|--------------|---------|---|--|--------------|------------------------------------|----------------------|---|
| Zhang 2021 | Rtr | USA | Veterans Health Administration (VHA) electronic health record | <p>Broad: Patients who had a diagnosis code for IPF (ICD-9-CM code 515, 516.3, 516.31 or ICD10-CM code J84.111, J84.112, J84.89, J84.9, J84.10, J84.17) recorded during study period</p> <ul style="list-style-type: none"> • Patients were considered to have IPF if they did not have any other diagnosis code for an alternative interstitial lung disease | 10.7 million | NR | NR | 1,160 |
| | | | | <p>Narrow: Patients who had a diagnosis code for IPF (ICD-9-CM code 515, 516.3, 516.31 or ICD10-CM code J84.111, J84.112, J84.89, J84.9, J84.10, J84.17) recorded during study period</p> <ul style="list-style-type: none"> • Patients were considered to have IPF if they did not have any other diagnosis code for an alternative interstitial lung disease • Patients who had procedure code for a lung biopsy or a CT scan of the thorax before the last IPF diagnosis | | NR | NR | 725 |

GCD, general case definition; BCD, broad case definition; NCD, narrow case definition

Table E2: List of Publications Reporting Incidence of IPF, Sorted By Publication Year

| Author, year | Study design | Country | Study population | ILD assessment | Sample size | No. of IPF cases by ILD definition | Mean age; Female (%) | Reported Incidence |
|-----------------|--------------|-------------|--|--|--------------------|------------------------------------|------------------------|---------------------------------|
| Gjonbrataj 2015 | Rtr | South Korea | HIRA | • Definition 1: code J84, ICD-10 code; (BAL) or lung biopsy; | 36,765,374 | 21,805 | NR; 38 | 48.5 per 100,000 |
| | | | | • Definition 2: code J84 and HRCT, bronchoalveolar lavage or lung biopsy | | 13,680 | NR; 25 | 32.2 per 100,000 |
| | | | | • Definition 3: code J84.1, ICD-10 code; | | 5,881 | NR; 12 | 16.2 per 100,000 |
| | | | | • Definition 4: code J84.1 and HRCT, BAL or lung biopsy; and | | 4,219 | NR: 8.5 | 11.4 per 100,000 |
| | | | | • Definition 5: code J84.1A, based on the 2011 international statement. | | 615 | NR; 0.9 | 1.7 per 100,000 |
| Lee 2016 | Rtr | South Korea | HIRA | Patients with IPF were defined as those with a disease code of K-J84.18. | 51,026,868 | 2011,2012;657 | NR | 13.1 and 12.9 per 100,000 |
| Harari 2016 | Rtr | Italy | Healthcare administrative database of Lombardy | <p>IPF was defined as:</p> <p>GCD: Defined as IPF cases all individuals with at least one hospitalization with diagnosis of IPF or at least one outpatient visits with diagnosis of IPF (ICD-9-CM code 516.3) during the period from 1st January 2000 to 31st December 2010.</p> | Approx. 10 million | 2,951 | Majority >65 years; 43 | 5.24 (5.06-5.44) per 100,000 py |
| | | | | <p>BCD: Defined IPF cases those patients that satisfied the GCD and had no claims (inpatient or outpatient) with a diagnosis code for any other type of ILDs on or after the date of the last claim with IPF diagnosis</p> | | 2,093 | Majority >65 years; 40 | 3.74 (3.58-3.90) per 100,000 py |
| | | | | <p>NCD: Defined IPF cases those patients that satisfied the BCD and had one or more claim with a procedure code for surgical lung biopsy, transbronchial lung biopsy or computed tomography of the thorax, on or before the date of the last claim with a diagnosis code for IPF.</p> | | 1,309 | Majority >65 years ;41 | 2.33(2.20-2.46) per 100,000 py |
| Hopkins 2016 | Rtr | Canada | National databases: DAD and NACRS | Broad definition: by excluding cases with an ICD-10 CA code for another ILD after the J84.1 code, like other studies. | NR | 6,390 | NR | 18.7 per 100,000 |

| Author, year | Study design | Country | Study population | ILD assessment | Sample size | No. of IPF cases by ILD definition | Mean age; Female (%) | Reported Incidence |
|----------------|--------------|-------------|---|---|--|------------------------------------|----------------------|---|
| | | | | <u>Narrow</u> definition further excluded cases that did not have chest CT, bronchus or lung biopsy, or bronchoscopy prior to establishing a J84.1 code. | | 3,057 | NR | 9 per 100,000 |
| Raghu 2016 | Rtr | USA | Optum Clinformatics | <u>Primary</u> cohort: IPF patients were first identified if they had at least one claim with ICD-9-CM diagnosis code 516.3 (idiopathic fibrosing alveolitis, which is used by providers to identify IPF) during the study period. Two subgroups of the primary cohort were further identified by applying additional criteria to form more restrictive cohorts of prevalent IPF patients: | 9,375,020 | 4,206 | 53;51 | 5.8 per 100,000 |
| | | | | <u>Broad</u> case definition excluded patients if they had a claim with the diagnosis code 515 after the last diagnosis code 516.3. | | 2,670 | 53;52 | 3.6-5.1 per 100,000 |
| | | | | <u>Narrow</u> case definition further restricted the broad case subgroup patients by requiring a claim for a surgical lung biopsy, transbronchial lung biopsy, or computed tomography scan of the thorax prior to the last diagnosis code 516.3. | | 1,685 | 54;53 | 2.4-2.9 per 100,000 |
| Strongman 2018 | Rtr | UK | CPRD | <u>Narrow</u> IPF-CS case definition were H563.00 (Idiopathic fibrosing alveolitis), H563.12 (Cryptogenic fibrosing alveolitis), H563z00 (Idiopathic fibrosing alveolitis NOS), H563300 (Usual interstitial pneumonitis), and H563.13 (Idiopathic pulmonary fibrosis). | 9,748,108 | 1,491 | NR | 2.85 (2.7-3) per 100,000 py |
| | | | | <u>Broad</u> IPF-CS definition included the following three additional Read codes: H563100 (Diffuse pulmonary fibrosis), H563200 (Pulmonary fibrosis), and H563.11 (Hamman-Rich syndrome). | | 4,527 | | 8.7 (8.4-8.9) per 100,000 py |
| Lim 2019 | Rtr | South Korea | Korean National Health Insurance Service (NHIS) database for patients registered with the co-payment assistance policy for rare and incurable diseases. | ICD codes | 2011: 50,908,646, 2012: 51,169,141, 2013: 51,448,491, 2014: 51,757,146 2015: 52,034,424 | NR | NR | 2011: 27.85 2012: 28.36 2013: 31.28 2014: 32.21 2015: 35.82 Cumulative 2011-2015: 31.03 per 100,000 |
| Belbasis 2021 | Prosp | UK | Biobank, self-reported (all unrelated individuals of White ethnicity participating in the UK Biobank) | <u>Broad</u> included all individuals with an ICD, Tenth Revision, diagnosis of J84.1 and J84.9 or an ICD, Ninth Revision, diagnosis of 516.3. | 437,453 individuals | 1,234 | 63,41 | 282.1 per 100,000 |
| | | | | <u>Narrow</u> : individuals with pulmonary fibrosis excluding other medical conditions that cause pulmonary fibrosis or interstitial lung disease | | 802 | NR | 183.3 per 100,000 |
| Zhang 2021 | Rtr | USA | Veterans Health Administration (VHA) electronic health record | <u>Broad</u> : Patients who had a diagnosis code for IPF (ICD-9-CM code 515, 516.3, 516.31 or ICD10-CM code J84.111, J84.112, J84.89, J84.9, J84.10, J84.17) recorded during study period • Patients were considered to have IPF if they did not have any other diagnosis code for an alternative interstitial lung disease | 10.7 million | 139,116 | 71;7 | 331 per 100,000 |
| | | | | <u>Narrow</u> : Patients who had a diagnosis code for IPF (ICD-9-CM code 515, 516.3, 516.31 or ICD10-CM code J84.111, J84.112, J84.89, J84.9, J84.10, J84.17) recorded during study period • Patients were considered to have IPF if they did not have any other diagnosis code for an alternative interstitial lung disease • Patients who had procedure code for a lung biopsy or a CT scan of the thorax before the last IPF diagnosis | | 82,557 | 707 | 210 per 100,000 py |

GCD, general case definition; BCD, broad case definition; NCD, narrow case definition

Table E3: List of Publications Reporting Prevalence of IIP, Sorted by Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | Type of IIP | No. of ILD cases | Mean age; Female (%) | Prevalence |
|----------------|---------|--------------|--|--|----------------------|-------------|------------------|----------------------|---------------------------|
| Lee 2016 | Korea | Rtr | HIRA database | Diagnosis codes; Patients with IIP were defined as those with two or more medical visits or at least one hospitalisation with an IIP diagnostic code | 51,026,868 | IIP | 41,770 | NR; 50 | 81.86 per 100,000 persons |
| Duchemann 2017 | France | CS | Sources of case identification from separate sources of patients were used: <ul style="list-style-type: none"> Public and private hospitals Community Pulmonologists and General Practitioners | <ul style="list-style-type: none"> A MDD of clinicians, radiologist and pathologists discussed each case, after a systematic review of medical charts, HRCT, and SLB, when applicable. ILD aetiology was determined when a diagnosis could be attributed in consensus according to the current guidelines, including 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF. ILD aetiology remained undetermined when a diagnosis could not be established based on available investigations after MDD. For unreviewable cases, information on the aetiological diagnosis was taken from the completed questionnaires or the ICDs-10 codes. | 1,194,601 | IIP | 145 | 56;50 | 12.14 per 100,000 persons |
| | | | | | | NSIP | 20 | | 1.7 per 100,000 persons |
| | | | | | | DIP | 10 | | 0.8 per 100,000 persons |
| | | | | | | OP | 9 | | 0.8 per 100,000 persons |
| | | | | | | RB-ILD | 2 | | 0.2 per 100,000 persons |

Table E4: List of Publications Reporting Incidence of IIP, Sorted by Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | Type of IIP | No. of ILD cases | Mean age; Female (%) | Incidence |
|----------------|---------|--------------|--|--|----------------------|-------------|------------------|----------------------|---------------------------------|
| Lee 2016 | Korea | Rtr | HIRA database | Diagnosis codes; Patients with IIP were defined as those with two or more medical visits or at least one hospitalisation with an IIP diagnostic code | 51,026,868 | IIP | 17,826 | NR; 50 | 34.93 per 100,000 persons |
| Duchemann 2017 | France | CS | Sources of case identification from separate sources of patients were used: <ul style="list-style-type: none"> Public and private hospitals Community Pulmonologists and General Practitioners | <ul style="list-style-type: none"> A MDD of clinicians, radiologist and pathologists discussed each case, after a systematic review of medical charts, HRCT, and SLB, when applicable. ILD aetiology was determined when a diagnosis could be attributed in consensus according to the current guidelines, including 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF. ILD aetiology remained undetermined when a diagnosis could not be established based on available investigations after MDD. For unreviewable cases, information on the aetiological diagnosis was taken from the completed questionnaires or the ICDs-10 codes. | 1,194,601 | IIP | 52 | 56;50 | 4.4 per 100,000 person per year |
| | | | | | | NSIP | 10 | | 0.8 per 100,000 person per year |
| | | | | | | DIP | 3 | | 0.3 per 100,000 person per year |
| | | | | | | OP | 1 | | 0.1 per 100,000 person per year |

Table E5: List of Publications Reporting Prevalence of Systemic Sclerosis-ILD, Sorted By Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | Type of SSc | No. of ILD cases | Mean age; Female (%) | Prevalence |
|-------------------|-----------|--------------|--|---|----------------------|--------------|------------------|----------------------|------------------------|
| Simeon-Aznar 2015 | Spain | Rtr | RESCLE registry | Pulmonary involvement is defined by the presence of ILD or PH. ILD was established if any of the following criteria were identified: <ul style="list-style-type: none"> restrictive pulmonary pattern with FVC below 80% of expected value on PFTs, pulmonary interstitial pattern evidenced by chest radiograph HRCT scan, alveolitis confirmed by bronchoalveolar lavage (defined as neutrophilia of $\geq 3\%$, eosinophilia of $\geq 2\%$, or lymphocytosis $\geq 15\%$). | 879 | Overall SSc | 421 | 44.4;85 | 47.8% |
| Wangkaew 2016 | Thailand | Prosp | Maharaj Nakorn Chiang Mai Hospital | <ul style="list-style-type: none"> All HRCTs images were reviewed by an experienced thoracic radiologist We used HRCT to categorize pattern of the lung parenchyma findings that represented ILD, which were unexplainable by any other causes. | 113 | Overall SSc | 80 | 53.4;58 | 69.1% |
| | | | | | 89 | dcSSc, | 70 | 54.1;51 | 78.7% |
| | | | | | 24 | LcSSc, | 11 | 50.6;88 | 45.8% |
| Tomiyama 2016 | Japan | Rtr | SSc who visited hospital department | ILD were evaluated by radiologists using high-resolution computed tomography (HRCT). Follow-up CT scans were performed when patients experienced a worsening of respiratory symptoms | 139 | Overall SSc | 66 | 49.2;82 | 47.5% |
| | | | | | 116 | lcSSc | 48 | NR | 41.4% |
| | | | | | 23 | dcSSc | 18 | NR | 78.3% |
| | | | | | 1,374 | Overall SSc | 595 | 57;86 | 43.3% |
| Sanchez-cano 2018 | Spain | CS | RESCLE is a nation-wide, multi-center registry | ILD: evidence consistent with pulmonary fibrosis present on chest radiograph or HRCT. Additionally, FVC value was considered to categorise as ILD severity, we took a 70% value as cutoff, between mild and moderate-severe ILD. | 1,374 | lcSSc | 316 | 60;87 | 53.1% |
| | | | | | 1,374 | dcSSc | 240 | 52;85 | 40.3% |
| | | | | | 1,374 | ssSSc | 39 | 58;80 | 6.6% |
| Janardana 2019 | India | Rtr | Department of Clinical Immunology & Rheumatology, Christian Medical College | <ul style="list-style-type: none"> The diagnosis of ILD was made HRCT of the chest reported by radiologists at our institute in majority of the cases. Only for occasional patients, the diagnosis was based on clinical examination findings and CXR | 327 | Overall SSc | 288 | 35.1,8 | 88.1% |
| Morrisroe 2019 | Australia | Prosp | SSc patients enrolled in the Australian Scleroderma Cohort Study (ASCS), | ILD was defined by characteristic changes on HRCT. | 1,727 | Overall SSc | 489 | 46.6;86 | 28.3% |
| Noviani 2019 | Singapore | Prosp | Three tertiary rheumatology centres in Singapore | <ul style="list-style-type: none"> ILD was diagnosed by HRCT and the severity of ILD was determined by PFT. HRCT was performed in patients with clinical symptoms (dyspnea or cough) or signs suggestive of ILD (end-inspiratory bibasilar crepitations), chest radiographs suspicious of ILD or restrictive pattern on PFT with FVC or TLC $< 80\%$ predicted or DLCO $< 75\%$ predicted. Significant ILD was defined by FVC $< 70\%$ predicted and categorised under the ILD group | 490 | Overall SSc | 135 | 49; 83 | 27.6% |
| Bauer 2020 | France | Rtr | Patients treated at Nancy University Hospital | <ul style="list-style-type: none"> Pulmonary involvement was determined with functional measurement in terms of obstructive and restrictive defects and decreased diffusing capacity of the lungs for carbon monoxide DLCO ($< 80\%$) With a specific examination of the CT scan by an experienced radiologist | 77 | Overall SSc | 24 | NR | 31.2% |
| | | | | | 58 | lcSSc | 14 | | 24.1% |
| | | | | | 19 | dcSSc | 10 | | 52.6% |
| Jaafar 2021 | USA | Prosp | Multicentre national Prospective Registry of Early Systemic Sclerosis (PRESS) registry | Not clearly reported. Based on the results, ILD was diagnosed using HRCT and significant functional progression of ILD was defined as an absolute FVC decline of $\geq 10\%$ as compared to baseline FVC during the whole course of the study. | 239 | Overall SSc | 128 | 51,70 | 53.6% |
| Li 2021 | USA | Rtr | IBM MarketScan claims database | ICD codes | 78,964,708 | Overall, SSc | 15,005 | NR | 19 per 100,000 persons |

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | Type of SSc | No. of ILD cases | Mean age; Female (%) | Prevalence |
|--------------|-----------|--------------|--|---|----------------------|-------------------------------------|------------------|---|--|
| Pope 2021 | Canada | Rtr | SSc Patients were identified using the CIHI, NACRS, DAD. | ICD 10 codes | NR | Overall, SSc For year: (2017/18) | 257 | Age groups: 18-29: 0 30-50:1.7 51-64:4.2 65+:3.2; 3.6 | 2.3 per 100,000 persons (12% SSc cases with ILD) |
| Fairley 2021 | Australia | Prosp | Consecutive patients with SSc who prospectively enrolled in the Australian Scleroderma Cohort Study (ASCS) | <ul style="list-style-type: none"> HRCT were used to diagnose ILD These were usually performed in response to clinical examination findings (chest crepitations) or abnormal respiratory function testing | 1,505 | SSc only | 393 | NR | 26.1% |
| Carton 2021 | Belgium | Rtr | Consecutive SSc patients, included in two Flemish cohort | <ul style="list-style-type: none"> Each HRCT was centrally analysed by an experienced investigator who was blinded. Patients were classified into three subgroups ("no ILD", limited ILD or extensive ILD), according to the simplified flow diagram described by Goh et al | 722 | Overall, SSc | 243 | NR | 34% |

Table E6: List of Publications Reporting Incidence of Systemic Sclerosis-ILD, Sorted By Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | Type of SSc | No. of ILD cases | Mean age; Female (%) | Incidence rate (per 100,000 py) |
|---------------|----------|--------------|---|---|----------------------|--------------|------------------|----------------------|---------------------------------|
| Wangkaew 2016 | Thailand | Prosp | Maharaj Nakorn Chiang Mai Hospital, Chiang Mai University | <ul style="list-style-type: none"> All HRCTs were obtained with one of two MDCT platforms All images were reviewed by an experienced thoracic radiologist blinded to clinical and laboratory data. HRCTs were categorized by the pattern of the lung parenchyma findings that represented ILD, | 89 | dcSSc | NR | 53.4;58 | 58.8 |
| Wangkaew 2016 | | | | | 24 | lcSSc | NR | 53.4;58 | 17.3 |
| Carton 2021 | Belgium | Rtr | Consecutive SSc patients, included in two Flemish cohort | <ul style="list-style-type: none"> Each HRCT was centrally analysed by an experienced investigator who was blinded. Patients were classified into three subgroups ("no ILD", limited ILD or extensive ILD), according to the simplified flow diagram described by Goh et al | 722 | SSc, overall | 39 | | 2,570 (1,800-3,560) (8%) |
| | | | | | 281 | lcSSc | 26 | | 2,300 (1,500-3,360) (9%) |
| | | | | | 64 | dcSSc | 10 | | 3,730 (1,790 – 6,850) (16%) |
| Li 2021 | USA | Rtr | IBM MarketScan claims database | ICD codes | 78,964,708 | Overall, SSc | 8,252 | 58,82 | 4.3 (4.2 -4.4) |

Table E7 List of Publications Reporting Prevalence of Rheumatoid Arthritis - ILD, Sorted By Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age; Female (%) | Unit of measurement | Prevalence |
|----------------|-------------|--------------|---|---|------------------------|------------------|----------------------|------------------------------|----------------------|
| Md Yosuf 2017 | UK | Prosp | Patients with moderate to severe RA who were treated with RTX at participating site | <ul style="list-style-type: none"> PFT collected at 6–12 months pre-RTX, at the time of treatment with RTX, 6–12 months post-RTX and at the most recent follow-up. HRCT scans were acquired (when clinically indicated) in patients with worsening dyspnoea and/or deterioration in lung function using a standardised method. The scans were scored independently by two radiologists, both blinded to lung function information and the sequence of scans. In order to account for missing PFT data of those with severe ILD who were unable to perform the test, data from the HRCT and survival status were incorporated into the overall lung response. | 700 | 56 | 64;98 | Percentage of study cohort | 8 |
| Kim 2017 | South Korea | Prosp | RA Patients from the Korean Observational study Network for Arthritis cohort | <ul style="list-style-type: none"> ILD was diagnosed based on the results of CXR or CT scans. The CXRs and CT scans were read by expert radiologists at each hospital and the chest imaging records were collected from each hospital. The ILD group consisted of all patients whose chest image reports contained key words such as “pulmonary fibrosis”, “interstitial fibrosis”, “interstitial pneumonia”, “interstitial lung disease” and “ILD”, USA in addition to a descriptions of patterns of the lung disease such as “usual interstitial pneumonia” and “nonspecific interstitial pneumonia”. | 3,555 | 64 | 63.2;70 | Percentage of study cohort | 1.8 (95% CI 1.4–2.3) |
| Zhang 2017 | China | Rtr | Patients who were diagnosed with RA at the Capital Medical University, Beijing Chaoyang Hospital, during the study period | Not reported, ILD cases were reported as seen in the medical history | 550 | 32 | 57.6;63 | Percentage of study cohort | 5.8 |
| Sakr 2018 | Egypt | Rtr | RA patients attending the Rheumatology outpatient clinic, Kasr Alainy Hospital, Cairo University | ILD presence was extracted from patient’s medical history, details of ILD assessment are not reported. | 3,219 | 27 | NR | Percentage of study cohort | 0.8 |
| Duarte 2019 | UK | Rtr | RA patients that attended the Rheumatology Department of University College London Hospital | <ul style="list-style-type: none"> Types of lung involvement were based on HRCT, with the date of the exam being considered the date of lung disease diagnosis. Subsequent HRCT results were also recorded. PFTs including gas transfer (transfer factor for carbon monoxide), were collected for patients with lung involvement | 1,129 | 45 | 63.2;67 | Percentage of study cohort | 3.98 |
| McFarlane 2019 | USA | CS | RA inpatient discharges during study period, that took place at NY hospitals | <ul style="list-style-type: none"> A chest radiologist, pulmonary specialist, and a rheumatologist i.e. MDD reviewed the clinical course, management, pulmonary function tests, and chest CT images for each patient during the multidisciplinary reviews. | 503 | 32 | 64;88 | Percentage of study cohort | 6.4 |
| Kiely 2019 | UK | Prosp | The study used data from ERAS (1986–2001) and ERAN (2002–2012), two multicentre early RA inception cohorts. | The diagnosis of ILD at each centre was according to standard practice, with confirmatory evidence from standard investigations including pulmonary function tests, chest radiographs and HRCT scans. ILD was deemed to be present if the terms pulmonary fibrosis or ILD were listed on the CRF or the death certificate using ICD codes. | 1114 | 53 | NR | Percentage of study cohort | 4.8 |
| Raimundo 2019 | USA | Rtr | Truven Health MarketScan Commercial and Medicare Supplemental | <p>Patients meeting ≥ 1 of the following criteria were identified as potential RA-ILD cases:</p> <ul style="list-style-type: none"> ≥ 2 claims with an International Classification of Diseases-9-Clinical Modification (ICD-9-CM) diagnosis code for pulmonary disease (fibrosis: ICD-9-CM 515, 516.3, 516.31; rheumatic lung disease: ICD-9-CM 714.81) on | over 150 million lives | 648 | 66;67 | 2004, per 100,000 RA persons | 3.2 (95% CI 3.0–3.4) |

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age; Female (%) | Unit of measurement | Prevalence |
|---------------|-------------|----------------------|--|--|--|------------------|----------------------|---|----------------------|
| Raimundo 2019 | | | health insurance databases | <p>different days, with the second claim occurring within 12 months of the first plus ≥ 2 claims with a diagnosis of RA (ICD-9-CM 714.xx, excluding 714.3x and 714.4x) on different days in the 12 months before or after the first claim for pulmonary disease;</p> <ul style="list-style-type: none"> ≥ 2 claims with a diagnosis of rheumatic lung disease (ICD-9-CM 714.81) on different days, with the second claim occurring within 12 months of the first based on a previously published algorithm. | | 1,575 | 65;70 | 2013, per 100,000 RA persons (general population) | 6.0 (95% CI 5.7–6.2) |
| Ke 2021 | China | Rtr case control | Subjects were selected from RA diagnosed patients who were hospitalized in the Department of Rheumatology and Immunology; there 2 cohorts: i) patients aged above 60 years, ii) patients between 18 to 59 years. | <ul style="list-style-type: none"> Not reported (self-report) | 142 (79 EORA ie elderly onset patient developed RA after the age of 60 years and 63 YORA (developed RA at a younger age) | 21 | NR | Percentage of study cohort | 14.8 |
| Kronzer 2021 | Sweden | Matched case control | RA patients enrolled in EIRA (Epidemiological investigation of RA) which is a population-based, case-control study of incident RA in central and southern Sweden that began in 1995 | <ul style="list-style-type: none"> Diagnosis code in linked EHR record (ie the Swedish National Patient Register) | 1631 | 10 | NR | Percentage of study cohort | 0.6 |
| Paulin 2021 | Argentina | CS | RA patients recruited from 2 rheumatology outpatient clinics with less than 2 years of RA diagnosis (Early RA study) | <ul style="list-style-type: none"> Scans HRCT used to detect abnormalities | 83 | 6 | NR | Percentage of study cohort | 7.5 |
| Samy 2021 | Egypt | CS | RA Patients were recruited from the outpatient clinic and the inpatient Department of Rheumatology at a university hospital | <ul style="list-style-type: none"> RA-ILD patients had any of the following HRCT abnormalities: ground glass opacities, septal lines, reticulation, subpleural fibrosis, traction bronchiectasis, architectural distortion, and/or honey combing. Radiographic abnormalities could occur with/without clinical symptoms of dyspnea and cough or significant PFTs abnormalities. | 160 | 102 | 45;86 | Percentage of study cohort | 63.7 |
| Shin 2021 | South Korea | Rtr | RA patients were enrolled into the KORean Observational study Network for Arthritis (KORONA) database by rheumatologists from 23 centres | <ul style="list-style-type: none"> Self-report (questionnaire responses at study enrolment) | 5,376 | 45 | NR | Percentage of study cohort | 0.84 |
| Sparks 2021 | USA | Rtr | Medicare claims data that includes nearly all | <ul style="list-style-type: none"> Among patients ever identified with RA, we identified ILD by the presence of two or more ICD-9 or ICD-10 codes for ILD in any position by a | 509,787 | 10,306 | 73;73 | Percentage of study cohort | 2 |

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age; Female (%) | Unit of measurement | Prevalence |
|--------------|---------|--------------|--|--|----------------------|------------------|----------------------|---------------------|------------|
| | | | Americans 65 years of age and some younger individuals with disabilities certain disabilities. | rheumatologist or pulmonologist separated by 7–365 days, as previously validated (PPV 72.4%) <ul style="list-style-type: none"> RA-ILD date was defined as the second billing code for ILD that fulfilled the algorithm. Incident RA-ILD was defined as occurring after the RA index date. | | | | | |

AD, airway disease; NY, New York

Table E8 List of Publications Reporting Incidence of Rheumatoid Arthritis - ILD, Sorted By Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age; Female (%) | Unit of measurement | Incidence |
|---------------|---------|--------------|--|---|------------------------|------------------|----------------------|---|--|
| Zhang 2017 | China | Rtr | Patients who were diagnosed with RA at the Capital Medical University, Beijing Chaoyang Hospital, during the study period | Not reported, ILD cases were reported as seen in the medical history | 550 | 205 | 57.6;63 | Percentage of RA study cohort | 37.3 |
| Raimundo 2019 | USA | Rtr | Truven Health MarketScan Commercial and Medicare Supplemental health insurance databases | Patients meeting ≥ 1 of the following criteria were identified as potential RA-ILD cases: <ul style="list-style-type: none"> ≥ 2 claims with an International Classification of Diseases-9-Clinical Modification (ICD-9-CM) diagnosis code for pulmonary disease (fibrosis: ICD-9-CM 515, 516.3, 516.31; rheumatic lung disease: ICD-9-CM 714.81) on different days, with the second claim occurring within 12 months of the first plus ≥ 2 claims with a diagnosis of RA (ICD-9-CM 714.xx, excluding 714.3x and 714.4x) on different days in the 12 months before or after the first claim for pulmonary disease; ≥ 2 claims with a diagnosis of rheumatic lung disease (ICD-9-CM 714.81) on different days, with the second claim occurring within 12 months of the first based on a previously published algorithm. | over 150 million lives | NR | NR | 2004, Per 100,00 persons | 2.7 (95% CI 2.5-2.9) |
| | | | | | | NR | NR | 2013, Per 100,00 persons (general population) | 3.8 (95% CI 3.5-4.0) |
| Sparks 2021** | USA | Rtr | Medicare claims data that includes nearly all Americans 65 years of age and some younger individuals with disabilities certain disabilities. | <ul style="list-style-type: none"> Among patients ever identified with RA, we identified ILD by the presence of two or more ICD-9 or ICD-10 codes for ILD in any position by a rheumatologist or pulmonologist separated by 7–365 days, as previously validated (PPV 72.4%) RA-ILD date was defined as the second billing code for ILD that fulfilled the algorithm. Incident RA-ILD was defined as occurring after the RA index date. | 499,481 | 13,372 | NR | Per 100,000 person-years among RA patients | 714 (700-730) per 100,000 persons (2.7%) |

** Adjusted for age and sex

Table E9 List of Publications Reporting **Prevalence of Myositis ILD**, Sorted By Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age (years); Female (%) | Prevalence (%) |
|---------------|-----------|--------------|--|--|----------------------|------------------|------------------------------|----------------|
| Ishizuka 2016 | Japan | Rtr | Enrolled patients with PM/DM who were referred to our department | The presence of ILD was evaluated by computed tomography (CT). | 124 | 76 | 53.5;65 | 61.3 |
| Ishizuka 2016 | | | | | 46 (PM) | 25 | NR | 54.3 |
| Ishizuka 2016 | | | | | 78 (DM) | 51 | NR | 65.4 |
| Gomez 2021 | Argentina | CS | Argentine registry of IMM that includes patients older than 18 years who have been diagnosed with myositis ILD | HRCT or biopsy | 360 | 99 | NR | 27.5 |
| Huang 2021 | Taiwan | Rtr | Patients who underwent complete myositis autoantibody serology examination at university hospital | NR | 97 | 46 | 56;80 | 47.4 |

Table E10 List of Publications Reporting **Incidence of Myositis ILD**, Sorted By Publication Year

None reported

Table E11 List of Publications Reporting **Prevalence of Sjogren's Syndrome ILD**, Sorted By Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age (years); Female (%) | Type of SS | Prevalence (%) |
|-----------------|---------|--------------|---|--|---|------------------|------------------------------|------------|----------------|
| Kvarnstorm 2015 | Sweden | Prosp | All incident patients referred to the dept. of Rheumatology at the Karolinska University Hos. | <ul style="list-style-type: none"> ILD assessment not reported | 199 | 2 | 55;94 | p-SS | 1 |
| Zhao 2015 | China | CS | Patients from 16 Chinese medical centers nationwide | <ul style="list-style-type: none"> Pulmonary involvement was indicated by persistent cough and/or dyspnea with chronic diffuse interstitial infiltrates on X-rays, altered patterns on pulmonary function tests, and/or evidence of lung alveolitis or fibrosis in computed tomography (CT) scans. | 317 | 59 | 49;94 | p-SS | 18.6 |
| Manfredi 2017 | Italy | Prosp | All consecutive patients diagnosed as pSS during the study period | <ul style="list-style-type: none"> ILD assessment was based on HRCT and PFTs. HRCT was categorized as definite, possible, or inconsistent with usual interstitial pneumonia (UIP) pattern according to the 2011 ATS/ERS/JRS/ALAT Guidelines for Diagnosis and Management of IPF | 77 | 13 | 67;77 | p-SS | 16.9 |
| Roca 2017 | France | Rtr | Medical records from 2 participating institutional centers | <ul style="list-style-type: none"> ILD was investigated by PFTs, and HRCT scan of the lungs; no patient underwent lung biopsy. | 263 | 11 | 63;86 | p-SS | 4.2 |
| Gao 2018 | China | Rtr | Hospitalized patients admitted to Peking | <ul style="list-style-type: none"> Diagnosis of pSS-ILD was made according to HRCT abnormalities and/or impaired PFTs, with or without pulmonary signs | 853 | 165 | 61;92 | p-SS | 19.3 |
| | | | | | 488 | 126 | 61;92 | s-SS | 25.8 |
| Kampolis 2018 | Greece | CS | Department of Pathophysiology (Medical School, University of Athens) | <ul style="list-style-type: none"> HRCT scan of the lungs was performed in all patients at the day of pulmonary function testing. Abnormal HRCT findings were classified into four major categories: reticular (septal thickening, honeycombing), nodular (multiple large or small nodules), high attenuation | Overall, 384; Asymptomatic patients who | 7 | 63;95 | p-SS | 17.9 |

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age (years); Female (%) | Type of SS | Prevalence (%) |
|--------------|--------------|--------------|--|--|----------------------|------------------|------------------------------|------------|----------------|
| | | | | (consolidation, ground glass opacities) and low attenuation (air trapping, lung cysts) pattern. CT images were independently evaluated by two experienced radiologists. | underwent HRCT: 39 | | | | |
| Wang 2018 | China | Prosp | Newly diagnosed pSS were recruited from Beijing Chao-Yang Hospital | <ul style="list-style-type: none"> All patients underwent HRCT scans. The scans included both lungs in the field of view. Each HRCT scan was reviewed independently by two experienced thoracic radiologists blinded to the clinical data before the therapeutic interventions. 65% pSS-ILD patients underwent percutaneous lung biopsy or bronchoscopy, including bronchoalveolar lavage total cell counts and cell differentials and transbronchial lung biopsy. PFTs were performed according to the guidelines. | 201 | 158 | 62;85 | p-SS | 78.6 |
| Gao 2021 | China | Rtr | pSS patients admitted to Peking university hospital | <ul style="list-style-type: none"> Expert radiologist re-assessment of the imaging | 934 | 178 | 62;87 | p-SS | 19.1 |
| Kam 2021 | Singapore | Rtr | pSS between 1993 and 2013, seen at Tan Tock Seng Hospital | <ul style="list-style-type: none"> NR | 102 | 2 | NR | p-SS | 1.96 |
| Nilsson 2021 | Sweden | CS | Patients with pSS had been previously investigated for pulmonary involvement at university hospital | <ul style="list-style-type: none"> HRCT, pulmonary function tests | 40 | 15 | NR | p-SS | 38 |
| Omair 2021 | Saudi Arabia | CS | Study participants were recruited from patients with pSS attending the rheumatology and pulmonary clinics at a university hospital | <ul style="list-style-type: none"> NR | 41 | 36 | NR | p-SS | 87.8 |

Table E12 List of Publications Reporting Incidence of Sjogren's Syndrome ILD, Sorted By Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age (years); Female (%) | Type of SS | Incidence (%) |
|--------------|---------|--------------|--|---|----------------------|------------------|------------------------------|------------|---------------|
| Roca 2017 | France | Rtr | Medical records from 2 participating institutional centers | <ul style="list-style-type: none"> ILD was investigated by PFTs, and HRCT scan of the lungs; no patient underwent lung biopsy. | 263 | 9 | 63;86 | p-SS | 3.4 |

Table E13 List of Publications Reporting Prevalence/Incidence of Other Autoimmune Conditions

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age (years); Female (%) | Prevalence (%) | Incidence (%) |
|------------------|---------|--------------|---|---|----------------------|------------------------------|------------------------------|----------------|---------------|
| Reiseter 2018 | Norway | Rtr | Nationwide MCTD cohort from depts of Rheumatology | <ul style="list-style-type: none"> CT scans and PFTs | 119 | Prevalent: 47 Incident: 2 | 91;38 | 40% | 1.7% |
| Olaosebikan 2021 | Nigeria | CS | CTD patients managed at the Rheumatology Unit of Lagos State University Teaching Hospital | <ul style="list-style-type: none"> Clinical information including socio-demographics, signs and symptoms of ILD, pattern(s) of CTD, serology, pulmonary function, and radiology findings as well as treatment outcomes were retrieved from patients' records | 318 | Prevalent: 23 Incident: 8 | 39;90 | 7.2% | 2.5% |

Table E14 List of Publications Reporting Prevalence of Hypersensitivity pneumonitis, Sorted By Type of Hypersensitivity pneumonitis and Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | Type of HP; No. of cases | Unit of Prevalence | Prevalence |
|----------------|---------------|---------------------|--|--|----------------------|------------------------------|------------------------------|--|
| Duchemann 2017 | France | CS | Sources of case identification from separate sources of patients were used: <ul style="list-style-type: none"> Public and private hospitals Community Pulmonologists and General Practitioners | <ul style="list-style-type: none"> MDD of clinicians, radiologist and pathologists discussed each case, after a systematic review of medical charts, HRCT, and SLB, when applicable. ILD aetiology was determined when a diagnosis could be attributed in consensus according to the current guidelines, including 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF. ILD aetiology remained undetermined when a diagnosis could not be established based on available investigations after MDD. For unreviewable cases, information on the aetiological diagnosis was taken from the completed questionnaires or the ICDs-10 codes. | 1,194,601 | HP; 28 | Per 100,000 persons per year | 2.3 |
| Perez 2018 | United States | Rtr | Truven Health Market Scan Commercial and Medicare | <ul style="list-style-type: none"> This research developed novel claims-based coding algorithms to identify HP, CHP, FHP cases, these algorithms validity and reliability were assessed with clinical data from National Jewish Health. The case validation procedure involved medical record review performed using the electronic medical records. An expert chest radiologist reviewed CT scans, blinded to subjects' clinical information and the primary radiologist's interpretation, to determine whether pulmonary fibrosis was present and whether the CT met criteria for consistent or possible HP or was inconsistent with HP. Last, a clinical-radiological consensus diagnosis of HP or not HP was applied. | 158,270,350 | HP; 7,498 | Per 100,000 persons | 1.67 (95% CI, 1.5 - 1.8) - 2.71 (95% CI, 2.6 -2.9) |
| | | | | | 4,093 | FLD; 157 | % of the HP cohort | 3.8 |
| | | | | | 4,093 | Bagassosis; 11 | % of the HP cohort | 0.3 |
| | | | | | 4,093 | Bird fancier's; 89 | % of the HP cohort | 2.2 |
| | | | | | 4,093 | Suberosis; 71 | % of the HP cohort | 1.7 |
| | | | | | 4,093 | Malk workers; 21 | % of the HP cohort | 0.5 |
| | | | | | 4,093 | Mushroom workers; 55 | % of the HP cohort | 1.3 |
| | | | | | 4,093 | Maple bark strippers; 26 | % of the HP cohort | 0.6 |
| | | | | | 4,093 | Ventilation pneumonitis; 201 | % of the HP cohort | 4.9 |
| | | | | | 4,093 | Other Spec; 482 | % of the HP cohort | 11.8 |
| | | | | | 4,093 | Unspec; 3,015 | % of the HP cohort | 73.7 |
| | | | | | 4,093 | CHP; 2,308 | Per 100,000 persons | 0.91 (95% CI, 0.8-1.0) - 1.7 (95% CI, 1.6-1.8) |
| 4,093 | FHP; 941 | Per 100,000 persons | 0.41 (95% CI, 0.3-0.5) - 0.8 (95% CI, 0.7- 0.9) | | | | | |

Table E15 List of Publications Reporting Incidence of Hypersensitivity pneumonitis, Sorted By Type of Hypersensitivity pneumonitis and Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | Type of HP; No. of cases | Unit of Prevalence | Prevalence |
|----------------|---------------|--------------|--|--|----------------------|--------------------------|----------------------|---|
| Duchemann 2017 | France | CS | Sources of case identification from separate sources of patients were used: <ul style="list-style-type: none"> Public and private hospitals Community Pulmonologists and General Practitioners | <ul style="list-style-type: none"> MDD of clinicians, radiologist and pathologists discussed each case, after a systematic review of medical charts, HRCT, and SLB, when applicable. ILD aetiology was determined when a diagnosis could be attributed in consensus according to the current guidelines, including 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF. ILD aetiology remained undetermined when a diagnosis could not be established based on available investigations after MDD. For unreviewable cases, information on the aetiological diagnosis was taken from the completed questionnaires or the ICDs-10 codes. | 1,194,601 | HP; 11 | per 100,000 per year | 0.9 |
| Perez 2018 | United States | Rtr | Truven Health Market Scan Commercial and Medicare | <ul style="list-style-type: none"> Novel claims-based coding algorithms to identify HP, CHP, FHP cases The case validation procedure involved medical record review performed using the electronic medical records An expert chest radiologist reviewed CT scans, blinded to subjects' clinical information and the primary radiologist's interpretation, to determine whether pulmonary fibrosis was present and whether the CT met criteria for consistent or possible HP or was inconsistent with HP. Last, a clinical-radiological consensus diagnosis of HP or not HP was applied. | 158,270,350 | HP; 3405 | Per 100,000 | 1.28-1.9 (1.7 – 2.2) |
| | | | | | 158,270,350 | CHP; NR | Per 100,000 | 0.63 (95% CI, 0.5– 0.7) – 1.08 (95% CI, 0.9– 1.3) |
| | | | | | 158,270,350 | FHP; NR | Per 100,000 | 0.29 (95% CI, 0.2–0.4) - 0.43 (95% CI, 0.4– 0.5) |

Table E16 List of Publications Reporting Prevalence of Cystic Lung Disease, Sorted By Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | Type & No. of ILD cases | Prevalence |
|----------------|---------|--------------|--|--|----------------------|-------------------------|-------------------------|
| Duchemann 2017 | France | CS, Rtr | Sources of case identification from separate sources of patients were used: <ul style="list-style-type: none"> Public and private hospitals Community Pulmonologists and General Practitioners | <ul style="list-style-type: none"> MDD of clinicians, radiologist and pathologists discussed each case, after a systematic review of medical charts, HRCT, and SLB, when applicable. ILD aetiology was determined when a diagnosis could be attributed in consensus according to the current guidelines, including 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF. ILD aetiology remained undetermined when a diagnosis could not be established based on available investigations after MDD. For unreviewable cases, information on the aetiological diagnosis was taken from the completed questionnaires or the ICDs-10 codes. | 1,194,601 | LAM, 9 | 0.8 per 100,000 persons |
| Duchemann 2017 | | | | | | PLCH, 4 | 0.3 per 100,000 persons |

Table E17 List of Publications Reporting Incidence of Cystic Lung Disease, Sorted By Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | Type & No. of ILD cases | Incidence |
|----------------|---------|--------------|--|---|----------------------|-------------------------|----------------------------------|
| Duchemann 2017 | France | CS | Sources of case identification from separate sources of patients were used: <ul style="list-style-type: none"> Public and private hospitals Community Pulmonologists and General Practitioners | A multi-disciplinary team of clinicians, radiologist and pathologists discussed each case, after a systematic review of medical charts, HRCT, and surgical lung biopsy (SLB), when applicable. The aetiology of ILD was determined when a diagnosis could be attributed in consensus according to the current guidelines, including 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF. The aetiology of ILD remained undetermined when a diagnosis could not be established based on available investigations after multidisciplinary discussion (MDD). For unreviewable cases, information on the aetiological diagnosis was taken from the completed questionnaires or the ICDs-10 codes. | 1,194,601 | LAM, 4 | 0.3 per 100,000 persons per year |
| Duchemann 2017 | | | | | | PLCH,2 | 0.2 per 100,000 persons per year |

Table E18 List of Publications Reporting Incidence and Prevalence of Unclassifiable ILD, Sorted By Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Prevalence | Incidence |
|----------------|---------|--------------|--|--|----------------------|-------------------------------|-----------------------|----------------------------------|
| Duchemann 2017 | France | CS | Sources of case identification from separate sources of patients were used: <ul style="list-style-type: none"> Public and private hospitals Community Pulmonologists and General Practitioners | <ul style="list-style-type: none"> A MDD of clinicians, radiologist and pathologists discussed each case, after a systematic review of medical charts, HRCT, and SLB, when applicable. ILD aetiology was determined when a diagnosis could be attributed in consensus according to the current guidelines, including 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF. ILD aetiology remained undetermined when a diagnosis could not be established based on available investigations after MDD. For unreviewable cases, information on the aetiological diagnosis was taken from the completed questionnaires or the ICDs-10 codes. | 1,194,601 | Prevalent: 60 Incident: 22 | 5 per 100,000 persons | 1.8 per 100,000 persons per year |

Table E19 List of Publications Reporting Prevalence of Silicosis, Sorted By Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age; Female (%) | Exposure duration | Prevalence |
|----------------|--------------|--------------|--|---|----------------------|------------------|----------------------|--|------------|
| Knight 2015 | South Africa | Prosp | Baseline survey using a random sample of approximately 1,000 miners in each cluster (a subset of the total workforce) was conducted, staggered over calendar time (2006–2009). | <ul style="list-style-type: none"> Radiographs were read for silicosis according to the ILO Classification. All radiographs were read by an experienced nursing health professional for silicosis. For the purpose of the silicosis study an experienced occupational medicine physician and trained reader read All radiographs reported as silicosis ILO grade $\geq 0/1$ by reader I, | 11,557 | 1,374 | NR | NR | 11.9% |
| Siribadda 2016 | Srilanka | CS | Workers at a silica factory in Matale District | <ul style="list-style-type: none"> A screening questionnaire was used to assess symptoms of silicosis as well as tuberculosis which is a recognized complication of silicosis. The chest radiographs were reported according to the guidelines of ILO and the diagnosis of silicosis was made according to the following criteria as per the guidelines of the ILO Chest imaging (usually a conventional chest radiograph) that shows opacities consistent with silicosis. | 250 | 14 | 30;13 | < 5 yrs: 6 5 - 10 yrs: 7 > 10 yrs: 1 | 5.6% |

| | | | | | | | | | |
|------------------------|-----------|-----|--|--|-------------|---------|---------------------|---|------------------|
| Silanum 2017 | Thailand | CS | Stone carving workers | <ul style="list-style-type: none"> The questionnaire (that was developed from reviewing silicosis relevant documents) was performed by interviewing prior to performing the chest x-ray. Reading and interpreting chest films were done by NIOSH B reader according to ILO film. | 315 | 80 | NR | NR | 25.4% |
| Souza 2017 | Brazil | CS | Mineworkers in semi-precious stone mining | <ul style="list-style-type: none"> The diagnosis of silicosis was made by a physician based on exposure history and alterations typical of the disease shown on the chest X-ray. The chest X-rays were interpreted independently by a radiologist and a pulmonologist according to the ILO of Pneumoconioses. Silicosis was diagnosed in those patients whose chest X-ray showed rounded opacities with profusion equal to or greater than 1/0. | 348 | 129 | 48; NR | Duration of exposure (years): 28.3 (10.4) years; Avg time working per day, hours: 8.7 (2.3) hours | 37% |
| Casey 2019 | USA | Rtr | Medicare beneficiaries | <p>Three case definitions were used to identify beneficiaries with silicosis:</p> <ul style="list-style-type: none"> Broad: Any claim that included ICD-9-CM code 502, pneumoconiosis due to other silica or silicates, listed in any position during 1999-2014 Intermediate: Among broad definition cases those who used i) at least one inpatient, skilled nursing, or home health agency claim or ii) at least two outpatient provider claims within 365 days of each other. Narrow: Among intermediate cases, a narrow definition applied included, that cases have a chest X-ray or CT scan 30 days before or 30 days after a silicosis claim. | 4,99,23,987 | Broad: | 65-84 years: 84; 19 | NR | 39.5 per 100,000 |
| | | | | | | Interm: | 65-84 years: 85; 11 | NR | 23 per 100,000 |
| | | | | | | Narrow: | 65-84 years: 84; 16 | NR | 20.1 per 100,000 |
| Hoy 2021 | Australia | Rtr | V- SHARP is a population-based health assessment programme for current and former stonemasons | <ul style="list-style-type: none"> HRCT, lung function tests | 239 | 86 | | | 36% |
| Poinen-Rughooputh 2021 | China | CS | Silica dust-exposed workers employed for at least 1 year during the study period in any of eight Chinese pottery factories | <ul style="list-style-type: none"> Diagnosis of silicosis were based on 1986 Chinese pneumoconiosis Roetgen diagnostic criteria which classified silicosis as stages I, II or III similar to ILO of 1/1 or greater | 8,887 | 1,219 | 40;25 | Cum. Exp: 5.7 (mg/m ³ yr); Net years if dust exp: 25.8 | 13.7% |
| Requena-Mullor 2021 | Spain | CS | Workers in the stone sector handling artificial stone or silica compacts that were exposed to crystalline silica dust daily | <ul style="list-style-type: none"> NR | 311 | 64 | 39;0 | with the average number of years exposed being 8.40 (6.75) years for workers with silicosis | 20.6% |
| Monteiro 2021 | Brazil | CS | The targeted population was composed of 776 workers from 277 mines registered in A cooperative that organizes the mining operation in the region | <ul style="list-style-type: none"> NR, self-reported | 258 | 27 | 53;0 | Median crude dust exposure estimated by ART was 13.2 mg/m ³ | 10.5% |

ART, Advanced REACH (Registration, Evaluation, and Authorization of Chemicals) Tool

Table E20 List Of Publications Reporting **Incidence of Silicosis**, Sorted By Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age; Female (%) | Incidence |
|--------------|---------|--------------|------------------------------|---|----------------------|-------------------|-----------------------|------------------------------|
| Casey 2019 | USA | Rtr | Medicare beneficiaries | Three case definitions were used to identify beneficiaries with silicosis: <ul style="list-style-type: none"> • Broad: Any claim that included ICD-9-CM code 502, pneumoconiosis due to other silica or silicates, listed in any position during 1999-2014 • Intermediate: Among broad definition cases those who used i) at least one inpatient, skilled nursing, or home health agency claim or ii) at least two outpatient provider claims within 365 days of each other. • Narrow: Among intermediate cases, a narrow definition applied included, that cases have a chest X-ray or CT scan 30 days before or 30 days after a silicosis claim. | 4,99,23,987 | Broad: 19,696 | 65-84 years: 80;19 | 32.1 per 100,000 |
| | | | | | | Interm: 11,469 | 65-84 years: 81;19 | 19.1 per 100,000 |
| | | | | | | Narrow: 10,026 | 65-84 years: 67;19 | 16.6 per 100,000 |
| | | | | | | Avg. annual | | 2.4-4.6 cases per 100 000 |

Duration of exposure for ILD patients is not reported for papers reporting Silicosis incidence

Table E21 List of Publications Reporting **Prevalence of Asbestosis**, Sorted by Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age; Female (%) | Exposure duration | Prevalence |
|-----------------------|-------------|--------------|--|---|----------------------|------------------|--|---|------------|
| Lee 2015 | South Korea | CS | Residents from 5 villages that are within 2km from mines | <ul style="list-style-type: none"> • Chest radiographs were evaluated by radiologists • Reports were classified by ILO classification of pneumoconioses | 35 | 27 | Age (yr) 40-49: 0 50-59: 11% 60-69: 37% 70-79: 48% > 80: 4%; 48 | Duration of exposure (yr) 10-19:1 (4) 20-29:2 (7) 30-39: 3 (11) 40-49: 6 (22) 50-59: 8 (30) 60-69: 7 (26) | 77.1% |
| Wickram-atillake 2019 | Sri Lanka | CS | Participants from 9 provinces where asbestos factories, tsunami clean-up, construction industry were located | <ul style="list-style-type: none"> • Chest radiographs were independently • Reports were classified by ILO classification of pneumoconioses | 250 | 16 | NR | 13 out of 16 people were exposed to asbestos at work for more than 10 years | 7% |

Table E22 List of Publications Reporting Incidence of Asbestosis, Sorted by Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean/Median age(years); Female (%) | Prevalence |
|--------------------------|---------|--------------|---|--|-----------------------------|--|------------------------------------|-------------------------------|
| Szeszenia-Dąbrowska 2015 | Poland | Rtr | Based on data obtained from former asbestos company records and the Central Register of Occupational Diseases, this study included 18 large state-owned asbestos processing enterprises | <ul style="list-style-type: none"> Chest radiography and detection of the changes in the X-ray chest image are crucial in diagnosing asbestosis. The diagnosis of asbestosis relies on detecting the presence of X-ray changes as small irregular s-, t-, u-type opacities with density starting from 1/0. It is in accordance with the radiological classification of pneumoconiosis developed by ILO. | 43,650 | 2,160 | NR | 49.8 per 100,0 workers (4.9%) |
| DeBono 2021 | Canada | Longitudinal | Workers employed in over 900 industry and occupation groups as part of ongoing surveillance program | <ul style="list-style-type: none"> Cases were defined as those who had any of the following: (1) two or more physician visits with a diagnosis of ICD-9 code 501 (asbestosis), (2) two or more visits to an ambulatory care center with a main diagnosis of ICD-10 code J61 (pneumoconiosis due to asbestos and other mineral fibers), or (3) one or more hospital discharges with a main diagnosis of ICD-10 code J61. | 1.76 million | 737 | 66;44 | 0.042% |
| Thomsen 2021 | Denmark | Longitudinal | This study took advantage of the new nationwide DOC*X Project research database, which has been approved by the Danish Data Protection Agency, self-reported | <ul style="list-style-type: none"> On 1 January 1980, we ascertained all hospital contacts up to and including the year 2015 for asbestosis (ICD- 8 code 515.2; ICD- 10 code J61) Pathological confirmation remains the gold standard for diagnosis, 19 35 but lung biopsy is not routinely recommended in Denmark | 138,276 (Vehicle mechanics) | 47 (Number at risk: 138,226 Person-years at risk: 2,751,423) | NR | 0.03% |

Table E23 List of Publications Reporting Prevalence of Pneumoconiosis, Sorted by Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Prevalence |
|----------------|---------|--------------|--|--|----------------------|------------------|---------------------------|
| Duchemann 2017 | France | CS | Sources of case identification from separate sources of patients were used: <ul style="list-style-type: none"> Public and private hospitals Community Pulmonologists and General Practitioners | <ul style="list-style-type: none"> A MDD of clinicians, radiologist and pathologists discussed each case, after a systematic review of medical charts, HRCT, and SLB, when applicable. ILD aetiology was determined when a diagnosis could be attributed in consensus according to the current guidelines, including 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF. ILD aetiology remained undetermined when a diagnosis could not be established based on available investigations after MDD. For unreviewable cases, information on the aetiological diagnosis was taken from the completed questionnaires or the ICDs-10 codes. | 1,194,601 | 42 | 3.5 per 100,000 |
| Simsek 2021 | Turkey | CS | Workers from 19 workplaces that were randomly selected that use hard metal tools in metal machining processes in organized industrial zone | <ul style="list-style-type: none"> CXR's were evaluated by two readers separately and independently according to the ILO international classification of radiographs of pneumoconiosis, | 139 | 54 | 38.8% of study population |

Table E24 List of Publications Reporting Incidence of Pneumoconiosis, Sorted by Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Exposure duration | Incidence |
|----------------|---------|--------------|--|--|----------------------|------------------|----------------------------------|--|
| Cui 2015 | China | Rtr | Workers registered in the selected 4 colliery groups in North China | <ul style="list-style-type: none"> Qualified experts who were all members of the Pneumoconiosis Diagnosis Committee independently read the chest radiographs of CWP patients and other investigated workers If there was difference among the experts on the diagnosis, the diagnosis judgment principle was that the minority should be subordinate to the majority | NR | 2,873 | Datong: 19.8 (7.2) years | Avg: 4.06% Cum (1980-1970): 6.6-14.1% |
| | | | | | | | Kailuan: 24.9 (7.1) years | Avg: 4.9% Cum (1980-1970): 2.9-8.5% |
| | | | | | | | Fuxin: 23.5 (5.1) years | Avg: 1.4% Cum (1980-1970): 1.6-77.3% |
| | | | | | | | Tiefa: 20.5 (6.2) years | Avg: 0.3% Cum (1980-1970):0.4-3.6% |
| Duchemann 2017 | France | CS | Sources of case identification from separate sources of patients were used: <ul style="list-style-type: none"> Public and private hospitals Community Pulmonologists and General Practitioners | <ul style="list-style-type: none"> A MDD of clinicians, radiologist and pathologists discussed each case, after a systematic review of medical charts, HRCT, and SLB, when applicable. ILD aetiology was determined when a diagnosis could be attributed in consensus according to the current guidelines, including 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF. ILD aetiology remained undetermined when a diagnosis could not be established based on available investigations after MDD. For unreviewable cases, information on the aetiological diagnosis was taken from the completed questionnaires or the ICDs-10 codes. | 1,194,601 | 9 | NR | 0.8 per 100,000 per year |

Table E25 List of Publications Reporting Prevalence of Other Occupational / Exposure Related ILD

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age; Female (%) | Exposure duration | Prevalence |
|--------------|-------------|--------------|--|---|----------------------|------------------|----------------------|--|-------------------------------|
| Choi 2015 | South Korea | Longitudinal | Indium-handling workers from seven factories | <ul style="list-style-type: none"> PFTs and HRCTs to confirm ILD | 50 | 12 | 36;16 | mean exposure duration was 44 (range 2-144) months. ILD diagnosed mean exposure 9.6 (4-17) | 24% of Indium exposed workers |

Table E26 List of Publications Reporting Prevalence of Pulmonary Sarcoidosis, Sorted by Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Prevalence |
|----------------|------------|--------------|--|--|----------------------|------------------|--|
| Duchemann 2017 | France | CS | Sources of case identification from separate sources of patients were used: <ul style="list-style-type: none"> Public and private hospitals Community Pulmonologists and General Practitioners | <ul style="list-style-type: none"> A MDD of clinicians, radiologist and pathologists discussed each case, after a systematic review of medical charts, HRCT, and SLB, when applicable. ILD aetiology was determined when a diagnosis could be attributed in consensus according to the current guidelines, including 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF. ILD aetiology remained undetermined when a diagnosis could not be established based on available investigations after MDD. For unreviewable cases, information on the aetiological diagnosis was taken from the completed questionnaires or the ICDs-10 codes. | 1,194,601 | 361 | 30.2 per 100,000 |
| Coquart 2015 | Guadeloupe | Rtr | 2 hospitals | <ul style="list-style-type: none"> Cases were identified from medical records using search term 'sarcoidosis' | 72 | 49 | 68.1%; 14.4 per 100,000 inhabitants (95% CI, 10.8 to 17.8) |

Table E27 List of Publications Reporting Incidence of Pulmonary Sarcoidosis, Sorted by Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Incidence (100,000 persons per year) |
|----------------|-------------|--------------|--|--|----------------------|------------------|--------------------------------------|
| Duchemann 2017 | France | CS | Sources of case identification from separate sources of patients were used: <ul style="list-style-type: none"> Public and private hospitals Community Pulmonologists and General Practitioners | <ul style="list-style-type: none"> A MDD of clinicians, radiologist and pathologists discussed each case, after a systematic review of medical charts, HRCT, and SLB, when applicable. ILD aetiology was determined when a diagnosis could be attributed in consensus according to the current guidelines, including 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF. ILD aetiology remained undetermined when a diagnosis could not be established based on available investigations after MDD. For unreviewable cases, information on the aetiological diagnosis was taken from the completed questionnaires or the ICDs-10 codes. | 1,194,601 | 58 | 4.9 |
| Jeon 2020 | South Korea | Rtr | NHI linked to RID | <ul style="list-style-type: none"> ICD/KCD diagnosis | 402,345,679.01 | 1,955 | 0.48 |

Table E28 List of Publications Reporting Prevalence of Radiation -Induced ILD, Sorted by Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Prevalence |
|--------------------------------------|---------|--------------|--|--|----------------------|------------------|-----------------|
| Radiation-induced pneumonitis | | | | | | | |
| Duchemann 2017 | France | CS | Sources of case identification from separate sources of patients were used: <ul style="list-style-type: none"> Public and private hospitals Community Pulmonologists and General Practitioners | <ul style="list-style-type: none"> MDD of clinicians, radiologist and pathologists discussed each case, after a systematic review of medical charts, HRCT, and SLB, when applicable. ILD aetiology was determined when a diagnosis could be attributed in consensus according to the current guidelines, including 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF. ILD aetiology remained undetermined when a diagnosis could not be established based on available investigations after MDD. For unreviewable cases, information on the aetiological diagnosis was taken from the completed questionnaires or the ICDs-10 codes. | 1,194,601 | 7 | 0.6 per 100,000 |

Mean age, sex and exposure duration for ILD patients is not reported for papers reporting radiation induced ILD prevalence

Table E29 List of Publications Reporting Incidence of Radiation -Induced ILD, Sorted by Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age; Female (%) | Exposure duration | Incidence |
|--------------------------------------|---------|--------------|--|--|----------------------|------------------|----------------------|---|---|
| Radiation induced BOOP | | | | | | | | | |
| Murofushi 2015 | Japan | Prosp | Breast cancer patients treated with RT at a Japanese cancer institute hospital | <ul style="list-style-type: none"> Routinely collected chest x-rays In addition, when patients developed consistent respiratory symptoms or fever, lasting longer than 2 or more weeks, CT was performed | 1,175 | 16 | 52;100 | Median: 4.9 months (range, 2.3 to 8.6 months) | 1.4% of patients who underwent breast cancer RT |
| Sato 2018 | Japan | Rtr | Newly diagnosed breast cancer patients who received WBRT at Fukushima Medical University Hospital | <ul style="list-style-type: none"> Puchest X-ray and/or chest CT | 665 | 9 | 52;100 | Median: 4 months (range, 2–12 months) | 1.4% of patients who underwent breast cancer RT |
| Radiation induced pneumonitis | | | | | | | | | |
| Duchemann 2017 | France | CS | Sources of case identification from separate sources of patients were used: <ul style="list-style-type: none"> Public and private hospitals Community Pulmonologists and General Practitioners | <ul style="list-style-type: none"> A MDD of clinicians, radiologist and pathologists discussed each case, after a systematic review of medical charts, HRCT, and SLB, when applicable. ILD aetiology was determined when a diagnosis could be attributed in consensus according to the current guidelines, including 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF. ILD aetiology remained undetermined when a diagnosis could not be established based on available investigations after MDD. For unreviewable cases, information on the aetiological diagnosis was taken from the completed questionnaires or the ICDs-10 codes. | 1,194,601 | 1 | NR | NR | 0.1 per 100,000 persons-years |

Table E30 List of Publications Reporting Prevalence of Progressive Fibrosing ILD, Sorted by Publication Year

| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Mean age (yrs.); Female (%) | Prevalence (per 100,000 persons) |
|--------------|---------|--------------|--------------------------------|--|----------------------|------------------|-----------------------------|----------------------------------|
| Olson 2021** | USA | Rtr | IBM MarketScan claims database | <ul style="list-style-type: none"> ICD codes | 37,565,644 | NR | NR | 70.3 (69.3-71.3) |
| Nasser 2021 | France | Rtr | French healthcare database | <ul style="list-style-type: none"> PF-ILD case definition was based on three algorithms modified from Olson et al. 2020 using diagnosis codes | 538,472 | 14,413 | 68;48 | 6.6 -19.4 (18.9 – 19.8) |

**Adjusted for age and sex

Table E31 List of Publications Reporting Incidence of Progressive Fibrosing ILD, Sorted by Publication Year





| Author, year | Country | Study design | Study population description | ILD assessment | Study population (N) | No. of ILD cases | Incidence (per 100,000 persons) |
|--------------|---------|--------------|--------------------------------|--|----------------------|------------------|---------------------------------|
| Olson 2021** | USA | Rtr | IBM MarketScan claims database | <ul style="list-style-type: none"> ICD codes | 37,565,644 | NR | 32.6 (32 – 33.1) |
| Nasser 2021 | France | Rtr | French healthcare database | <ul style="list-style-type: none"> PF-ILD case definition was based on three algorithms modified from Olson et al. 2020 using diagnosis codes | 538,472 | 14,119 | 4.0 to 4.7 |

**Adjusted for age and sex

Figure E6 Quality Assessment for Studies Reporting Incidence

| ILD condition | Study author | Risk of Bias Assessment | Ascertainment of ILD |
|------------------------|--------------------------|-------------------------|---|
| Asbestosis | Thomsen 2021 | Low Risk | Multidisciplinary assessment |
| Asbestosis | Szeszenia-Dąbrowska 2015 | Medium Risk | HRCT scan |
| Asbestosis | DeBono 2021 | Medium Risk | Diagnosis codes (example: ICD) |
| Asbestosis | DeBono 2021 | Medium Risk | CXR findings or method of ILD ascertainment unclear or not stated |
| CTD ILD | Olaosebikan 2021 | High Risk | |
| Cystic Lung disease | Duchemann 2017 | Medium Risk | |
| HP | Duchemann 2017 | Medium Risk | |
| HP | Perez 2018 | Medium Risk | |
| IIP | Lee 2016 | Low Risk | |
| IIP | Duchemann 2017 | Medium Risk | |
| IPF | Gjonbrataj 2015 | Medium Risk | |
| IPF | Harari 2016 | Medium Risk | |
| IPF | Hopkins 2016 | Medium Risk | |
| IPF | Lee 2016 | Low Risk | |
| IPF | Raghu 2016 | Medium Risk | |
| IPF | Belbasis 2021 | High Risk | |
| IPF | Strongman 2018 | Low Risk | |
| IPF | Lim 2019 | Medium Risk | |
| IPF | Zhang 2021 | Medium Risk | |
| MTCO ILD | Reiseter 2018 | Low Risk | |
| Pneumoconiosis | Cui 2015 | Medium Risk | |
| Pneumoconiosis | Duchemann 2017 | Medium Risk | |
| PF ILD | Olson 2021 | Medium Risk | |
| PF ILD | Nasser 2021 | Low Risk | |
| Pulmonary Sarcoid | Duchemann 2017 | Medium Risk | |
| Pulmonary Sarcoid | Jeon 2020 | Medium Risk | |
| Pulmonary Sarcoid | Coquart 2015 | Medium Risk | |
| RA ILD | Zhang 2017 | Low Risk | |
| RA ILD | Raimundo 2019 | Medium Risk | |
| RA ILD | Sparks 2021 | Medium Risk | |
| Radiation Induced | Duchemann 2017 | Medium Risk | |
| Radiation Induced | Murofushi 2015 | Medium Risk | |
| Radiation Induced | Sato 2018 | Medium Risk | |
| Silicosis | Casey 2019 | Medium Risk | |
| Sjogren's Syndrome ILD | Roca 2017 | Low Risk | |
| SSc ILD | Wangkaew 2016 | Low Risk | |
| SSc ILD | Carton 2021 | Low Risk | |
| SSc ILD | Li 2021 | Medium Risk | |
| Unclassifiable ILD | Duchemann 2017 | Medium Risk | |

ILD diagnosis

| | |
|---|---|
|  | Multidisciplinary assessment |
|  | HRCT scan |
|  | Diagnosis codes (example: ICD) |
|  | CXR findings or method of ILD ascertainment unclear or not stated |

Risk of Bias

| | |
|---|-------------|
|  | High Risk |
|  | Medium Risk |
|  | Low Risk |

Figure E7 Quality Assessment for Studies Reporting Prevalence

| ILD condition | Study author | Risk of Bias Assessment | Ascertainment of ILD |
|----------------------------|---------------------|-------------------------|----------------------|
| Asbestosis | Wickramatillake2019 | | |
| Asbestosis | Lee 2015 | | |
| CTD ILD | Duchemann 2017 | | |
| CTD ILD | Olaosebikan 2021 | | |
| HP | Duchemann 2017 | | |
| HP | Perez 2018 | | |
| IIP | Lee 2016 | | |
| IIP | Duchemann 2017 | | |
| IPF | Harari 2016 | | |
| IPF | Hopkins 2016 | | |
| IPF | Kaunisto 2015 | | |
| IPF | Lee 2016 | | |
| IPF | Raghu 2016 | | |
| IPF | Raimundo 2016 | | |
| IPF | Strongman 2018 | | |
| IPF | Zhang 2021 | | |
| LAM | Duchemann 2017 | | |
| MCTD ILD | Reiseter 2018 | | |
| Myositis ILD | Ishizuka 2016 | | |
| Myositis ILD | Gomez 2021 | | |
| Myositis ILD | Huang 2021 | | |
| Other Exposure related ILD | Choi 2015 | | |
| PLCH | Duchemann 2017 | | |
| Pneumoconiosis | Simsek 2021 | | |
| Pneumoconiosis | Duchemann 2017 | | |
| Progressive fibrosing ILD | Nasser 2021 | | |
| Progressive fibrosing ILD | Olson 2021 | | |
| Pulmonary sarcoidosis | Duchemann 2017 | | |
| Pulmonary sarcoidosis | Coquart 2015 | | |
| RA ILD | McFarlane 2019 | | |
| RA ILD | Kim 2017 | | |
| RA ILD | Shin 2021 | | |
| RA ILD | Sakr 2018 | | |
| RA ILD | Ke 2021 | | |
| RA ILD | Kiely 2019 | | |
| RA ILD | Paulin 2021 | | |
| RA ILD | Raimundo 2019 | | |
| RA ILD | Duarte 2019 | | |
| RA ILD | Zhang 2017 | | |
| RA ILD | Samy 2021 | | |
| RA ILD | Kronzer 2021 | | |
| RA ILD | Sparks 2021 | | |
| RA ILD | Md Yosuf 2017 | | |

| ILD condition | Study author | Risk of Bias Assessment | Ascertainment of ILD |
|-----------------------|------------------------|-------------------------|----------------------|
| Radiation induced ILD | Duchemann 2017 | | |
| Silicosis | Casey 2019 | | |
| Silicosis | Hoy 2021 | | |
| Silicosis | Knight 2015 | | |
| Silicosis | Poinen-Rughooputh 2021 | | |
| Silicosis | Requena-Mullor | | |
| Silicosis | Silanum 2017 | | |
| Silicosis | Siribadda 2016 | | |
| Silicosis | Souza 2017 | | |
| Silicosis | Souza 2021 | | |
| Sjogrens ILD | Gao 2018 | | |
| Sjogrens ILD | Gao 2021 | | |
| Sjogrens ILD | Kam 2021 | | |
| Sjogrens ILD | Kampolis 2018 | | |
| Sjogrens ILD | Kvarnstorm 2015 | | |
| Sjogrens ILD | Manfredi 2017 | | |
| Sjogrens ILD | Nikson 2021 | | |
| Sjogrens ILD | Omair 2021 | | |
| Sjogrens ILD | Roca 2017 | | |
| Sjogrens ILD | Wang 2018 | | |
| Sjogrens ILD | Zhao 2015 | | |
| SSc ILD | Janardana 2019 | | |
| SSc ILD | Sanchez-cano 2018 | | |
| SSc ILD | Morrisroe 2019 | | |
| SSc ILD | Noviani 2019 | | |
| SSc ILD | Bauer 2020 | | |
| SSc ILD | Fairley 2021 | | |
| SSc ILD | Jaafar 2021 | | |
| SSc ILD | Simeon-Aznar 2015 | | |
| SSc ILD | Tomiyama 2016 | | |
| SSc ILD | Wangkaew 2016 | | |
| SSc ILD | Li 2021 | | |
| SSc ILD | Pope 2021 | | |
| SSc ILD | Carton 2021 | | |
| Unclassifiable ILD | Duchemann 2017 | | |