Online Data Supplement

Code-Based Diagnostic Algorithms for Idiopathic Pulmonary Fibrosis: Case Validation and Improvement

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Table E1. Exclusionary diagnostic codes

ICD-9 Code	Description
135	Sarcoidosis
237.7	Neurofibromatosis
272.7	Lipidoses
277.3	Amyloidosis
277.8	Other specified disorders of metabolism—includes eosinophilic granuloma
446.21	Goodpasture's syndrome
446.4	Wegener's granulomatosis
495	Extrinsic allergic alveolitis
500	Coal workers' pneumoconiosis
501	Asbestosis
502	Pneumoconiosis due to other silica or silicates
503	Pneumoconiosis due to other inorganic dust
504	Pneumoconiosis due to inhalation of other dust
505	Pneumoconiosis, unspecified
506.4	Chronic respiratory conditions due to fumes or vapors
508.1	Chronic and other pulmonary manifestations due to radiation
508.8	Respiratory conditions due to other specified external agents
516.0	Pulmonary alveolar proteinosis
516.1	Idiopathic pulmonary hemosiderosis
516.32-516.37	Idiopathic nonspecific interstitial pneumonia, acute interstitial pneumonia, respiratory bronchiolitis interstitial pneumonia, idiopathic lymphoid interstitial pneumonia, cryptogenic organizing pneumonia, and desquamative interstitial pneumonia
516.2	Pulmonary alveolar microlithiasis
516.8	Other specified alveolar and parietoalveolar pneumonopathies
516.9	Unspecified alveolar and parietoalveolar pneumonopathies
517.0	Lung involvement in conditions classified elsewhere
517.2	Lung involvement in systemic sclerosis
517.8	Lung involvement in other diseases classified elsewhere
518.3	Pulmonary eosinophilia
555	Regional enteritis
710.0	Systemic lupus erythematosus
710.1	Systemic sclerosis
710.2	Sjögren's disease
710.3	Dermatomyositis
710.4	Polymyositis
714.81	Rheumatoid lung
720	Ankylosing spondylitis
759.5	Tuberous sclerosis

Table E2. Procedure codes for lung biopsy and chest CT scan

Procedure	ICD-9-CM	CPT-4
Chest CT	87.41	71250, 71260, 71270
Surgical lung biopsy	33.20 33.28 34.21	32095-97, 32100-32160, 32602, 32607-8,
Transbronchial lung biopsy	33.27	31628, 31629, 31632

Table E3. Outcome of the ILD expert diagnosis case adjudication procedure in the first 75 cases meeting criteria for the IPF algorithm

	Radiology Review**					
Clinical* Review	Definite UIP	Possible UIP	Inconsistent with UIP	No ILD	No CT	**Total**
Likely IPF	15[a]	12[a]	9[b]	0	0	**36**
Insufficient	0	1[c]	2[c]	0	4[d]	**7**
Unlikely IPF	3[e]	5[f]	3[g]	1[g]	0	**12**
Not IPF	0	0	4[g]	16[g]	0	**20**
Total	**18**	**18**	**18**	**17**	**4**	**75**

*Based on clinician chart review, cases were categorized as Likely IPF, Insufficient, Unlikely IPF, and Not IPF
**Expert chest radiologist categorized CT patterns as "No IlD" or, if ILD was present, then as Definite, Possible, or Inconsistent with UIP based on consensus criteria

Case adjudication as IPF, unclassifiable, or Not IPF:
a. All cases were considered IPF.
b. Three cases were considered IPF, 6 cases were considered unclassifiable.
c. All cases were considered unclassifiable.
d. These cases were not evaluable due to lack of a CT scan available for review.
e. All cases were considered not IPF. Diagnoses included drug toxicity from nitrofurantoin (n=1) & rheumatoid arthritis-associated ILD (n = 2).
f. Two cases were considered unclassifiable. Three cases were considered not IPF, and diagnoses included asbestosis (n = 1), chemotherapy/radiation-induced ILD (n = 1), and emphysema with minimal smoking-related fibrosis (n = 1).
g. All cases were considered not IPF.

Abbreviations: IPF = idiopathic pulmonary fibrosis; UIP = usual interstitial pneumonia; ILD = interstitial lung disease; CT = computed tomography

Study ID: _____

Data Collection Form: Chart Review

Demographics Age at diagnosis: _____ Sex: M | F	**Bronchoscopy** Bronchoscopy: Y | N TBB: Y | N Interpretation:_____ BAL cell diff:_____
Chart Diagnosis Seen by pulmonologist? Yes | No Pulmonary diagnosis IPF | Other:_____	**Exposures** Smoker: Current | Former | Never PPY:_____ Asbestos: Y | N, Comment:_____ HP exposure: Y | N, Comment:_____ Radiation: Y | N, Comment:_____ Drug: Y | N, Comment:_____ Other:_____
Pulmonary Function Tests PFTs done: Y | N TLC % predicted: _____% FVC % predicted: _____% FEV:FVC ratio: _____ DLCO % predicted: _____%	**Associated Conditions** List relevant:
Imaging CT chest: Y | N HRCT? Y | N Report classification: Definite | Possible | Inconsistent | No ILD TU classification: Definite | Possible | Inconsistent | No ILD	**Serologies** ANA: Y / N | pos / neg | titer:_____ RF: Y / N | pos / neg | titer:_____ CCP: Y / N | pos / neg | titer:_____ CK: Y / N | level: _____ Other serologies:_____
Biopsy Surgical lung biopsy: Y | N Interpretation: _____ _____	**Other Comments:**

Confidence in IPF based on Chart Review:

Likely IPF | Insufficient | Unlikely IPF | Not IPF

1. **Likely IPF** – evidence of ILD AND no persuasive alternative cause of def/prob UIP pattern
2. **Insufficient** – inadequate information to evaluate ILD (e.g. no CT scan)
3. **Unlikely IPF** – persuasive alternative cause for a def/prob UIP pattern (e.g. RA, drug)
4. **Not IPF** – evidence against presence of any ILD or clear alternative diagnosis

Figure E1. Data collection form used for medical record data extraction

Clinical review	Radiology Review					
	Definite UIP	Possible UIP	Incon. with UIP	No ILD	No CT	**Total**
Likely IPF						**36**
Insufficient						**7**
Unlikely IPF						**12**
Not IPF						**20**
Total	**18**	**18**	**18**	**17**	**4**	**75**

IPF Adjudicate Not IPF Exclude

Figure E2. ILD expert diagnosis algorithm using information from medical record review and chest CT scan pattern

Abbreviations: IPF = idiopathic pulmonary fibrosis, ILD = interstitial lung disease, UIP = usual interstitial pneumonia, Incon. = inconsistent, CT = computed tomography

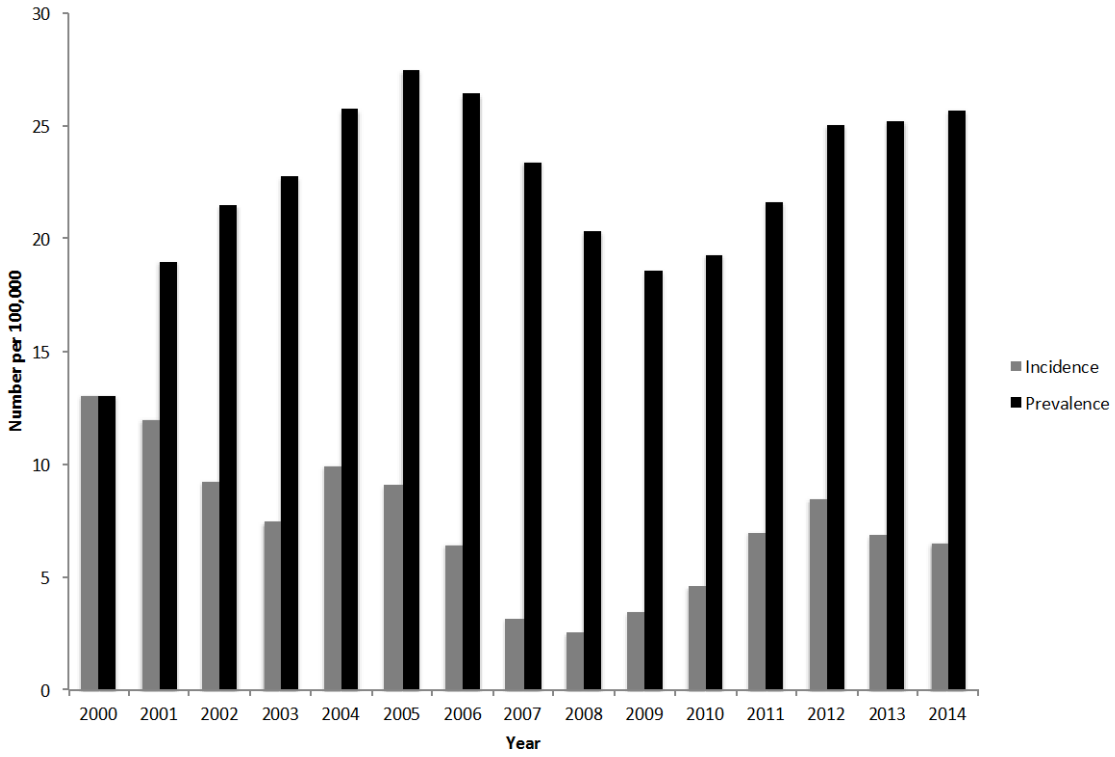

Figure E3. Annual incidence and cumulative prevalence of cases identified by the idiopathic pulmonary fibrosis (IPF) algorithm