Supplementary Tables

Supplementary Table S1. Prevalence of COPD among IPF Patients

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	COPD/ emphysema Case Ascertainment	Prevalence (%)
Collard HR[26]	US	2001–2008	9,286	IPF patients identified from 2 US claims databases	ICD-9 codes	Emphysema (ICD-9 codes)	8
Munson JC[23]	UK	1989–2006	1,126	IPF patients identified from the THIN database	Read codes	Asthma or COPD (ICD-9 codes predating IPF diagnosis)	19
Kurashima K[36]	Japan	1997–2006	660	IPF patients from a respiratory clinic	Presence of reticular abnormalities and traction bronchiectasis with basal and peripheral predominance; presence of honeycombing with basal and peripheral predominance; and absence of atypical features (e,g., micronodules,	Emphysema (CT scan)	34

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	COPD/ emphysema Case Ascertainment	Prevalence (%)
					consolidation, non-honeycomb cysts or extensive ground glass attenuation).		
Ryerson CJ[32][c]	US	2000–2010	365	IPF patients from the UCSF longitudinal and Mayo Clinic Rochester ILD databases	ATS/ERS/JRS/ ALAT	Emphysema (CT scan)	29
						>10% Emphysema	8
Ryerson CJ[27] [c]	US	2000–2010	242	IPF patients from the UCSF longitudinal ILD database	ATS/ERS	COPD (patient questionnaire and chart review)	29
Wells AU[22]	UK	1990–1996	212	Consecutive patients presenting with a clinical diagnosis of IPF	ATS/ERS	Emphysema (CT scan)	36 (median extent of emphysema: 10.5% (range, 1–69%).

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	COPD/ emphysema Case Ascertainment	Prevalence (%)
Cai M[37]	China	1999 – 2007	210	IPF patients identified from the database of Beijing Institute of Respiratory Medicine Interstitial Lung Disease Group, Beijing Chao-Yang Hospital	ATS/ERS	Emphysema (CT scan)	42
Antoniou KM[24]	UK	1991– 1999	186	Review of consecutive IPF patients from an interstitial lung unit who were current or former smokers	ATS/ERS	Emphysema (CT scan)	35
Schmidt SL[30]	US	1995– 2007	169	IPF patients from the University of Michigan ILD database	ATS/ERS	Emphysema (CT scan)	47–51
						Moderate–to-severe	25–27
Mejia M[6]	Mexico	1996– 2006	110	Clinical records of consecutive IPF patients at the National Institute of Respiratory Diseases, Mexico	ATS/ERS	≥10% Emphysema (CT scan)	28

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	COPD/ emphysema Case Ascertainment	Prevalence (%)
Sugino K[38]	Japan	2003–2010	108	Medical records of Consecutive IPF patients admitted to a single hospital	ATS/ERS	Emphysema (CT scan)	43
Hwang J-H[29]	US	1998–2006	97	Review of data from single center, IPF patients who had CT scans available were included	ATS/ERS	Emphysema (CT scan)	36
Araki T[34]	Japan	1978–1997	86	Consecutive autopsy series of IPF patients >65 years	ATS/ERS	Emphysema (greater than moderate severity in macroscopic observations)	65
Kim Y[35][a]	Korea	2006–2010	81	Medical records of patients with IPF/NSIP[b]	NR	Emphysema (CT scan)	40
Mura M[19]	Italy	2005–2007	70	Patients with newly diagnosed IPF	ATS/ERS	Emphysema (NR)	based on newly collected data: 34

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	COPD/ emphysema Case Ascertainment	Prevalence (%)	
			68	Retrospective (validation) study: IPF patients from the University of Siena, Italy			based on existing data:	19
Bodlet A[25]	Belgium	1981 - 2011	56	medical files IPF patients at single university hospital	ATS/ERS/JRS/ ALAT	Radiological emphysema (CT scan; Schmidt et al.'s HRCT scan criteria)	38	
Doherty MJ[20]	UK	1992–1995	48	Medical records of IPF patients	Fine basal inspiratory crackles with or without clubbing, a reduced TLCO, and a chest radiograph suggesting diffuse interstitial fibrosis	Emphysema (CT scan)	19	
Fernandez Perez ER[28]	US	1997–2005	47	Population-based sample of adult patients with IPF in Olmsted County,	ATS/ERS	COPD (ICD-9 codes)	28	

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	COPD/ emphysema Case Ascertainment	Prevalence (%)	
				Minn.				
Desai SR[21]	UK	1991–1999	40	Consecutive IPF patients	ATS/ERS	Emphysema (CT scan)	12	
						>10% emphysema	0	
Simon-Blancal V[18]	France	2002–2009	survivors: n=27; non-survivors: n=10	Medical records of all IPF patients who experienced an AE and were hospitalized	ATS/ERS	Emphysema (NR)	Total	51
							Survivors	48
							Non-survivors	60
Rufino RL*[39]	Brazil	2008–2010	36	IPF patients referred to an outpatient clinic	ATS/ERS	COPD (NR)	6	
Akagi T[33]	Japan	1988–2007	33	Hospital medical files of all patients	ATS/ERS	Emphysema (CT scan)	44	
Aduen JF[31]	US	1990–2000	9	Review based on existing data of the pulmonary function database at the Mayo Clinic in Jacksonville, Fla.; medical record was reviewed. IPF patients with reduced	Exclusion of other known causes of ILD; bibasilar reticular abnormalities with minimal ground-glass opacities	Emphysema (CT scan)	67	

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	COPD/ emphysema Case Ascertainment	Prevalence (%)
				DLCO who had undergone chest CT and echocardiography within 1 month of their PFT were included.	on HRCT; subpleural honeycombing on HRCT; diagnostic lung biopsy showing usual interstitial pneumonia pattern		

[a] abstract

[b] abstract only, does not report number of patients with IPF vs. NSIP

[c] These studies likely have some overlap is patient samples

*of the 38%, it was predominant in the lung apex: 62%; basal: 14%; lateralized: 5%; diffuse: 4%

Abbreviations: AE: Acute exacerbation; ALAT: Latin-American Thoracic Society; CT: computed tomography; DLCO: diffusing capacity of lung for carbon; monoxide; FEV_1: Forced expiratory volume in one second; FVC: Forced vital capacity; ILD: Interstitial lung disease; JRS: Japanese Respiratory Society; NSIP: Non-specific interstitial pneumonia; NR: Not reported; PFT: Pulmonary function testing; THIN: The Health Improvement Network; TLCO: transfer factor for carbon monoxide; UCSF: University of California, San Francisco

Supplementary Table S2. Prevalence of Pulmonary Hypertension among IPF Patients

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	PH Case Ascertainment		Prevalence (%)
Collard HR[26]	US	2001–2008	9,286	Patients identified from 2 claims databases	ICD-9 codes	ICD-9 codes		3
Shorr AF[61]	US	1995–2004	2,525	IPF patients from the UNOS database	NR	PH	mPAP >25 mmHg (from RHC)	46
						Severe PH	mPAP ≥40 mmHg (from RHC)	9
Mathai SC[56],[a]	US	1998–2008	1,848	IPF patients who RHC and lung transplantation were identified using the UNOS database	NR	mPAP >25 mmHg (from RHC)		33
Lederer DJ[52]	US	2004–2005	376	IPF patients from the UNOS database	NR	mPAP >25 mmHg (from RHC)		36
Cai M[37]	China	1999 – 2007	210	IPF patients identified from the database of Beijing Institute of Respiratory Medicine Interstitial Lung Disease Group, Beijing Chao-Yang Hospital	ATS/ERS	sPAP: ≥37 mmHg		29
Tomassetti	Italy	2002–	Not on	Patients evaluated at	ATS/ERS	NR		Not on

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	PH Case Ascertainment	Prevalence (%)
S[43][a]		2009	anticoagulants: 147; on anticoagulants: 35	an Italian IPF clinic[b]			anticoagulants: 36; on anticoagulants: 51
Papakosta D[41]	Greece	2005–2006	139	Patients with IPF referred to 8 departments of pneumonology	ATS/ERS	sPAP >36 mmHg (from ECHO)	55
Rivera-Lebron BN[67]	US	2005–2010	135	Review of IPF patients evaluated for lung transplantation at a university hospital	ATS/ERS	mPAP ≥25 mmHg and pulmonary capillary wedge pressure PWP <15 mmHg	29
Sherbini N[75]	Saudi Arabia	2007–2012	134	IPF patients' data from 2 tertiary care hospitals	ATS/ERS	NR (from ECHO)	12
Song JW[70]	Korea	1996–2008	131	Patients who underwent both ECHO and BNP measurement at a tertiary referral center	ATS/ERS	sPAP ≥40 mmHg (from ECHO)	25
Castria D[44]	Italy	2001–NR	126	Review of medical records of IPF patients from a single center	ATS/ERS	PH: sPAP > 36 mmHg (from ECHO) or mPAP >25 mmHg and PWP <15 (from RHC)	40
Tomassetti	Italy	2000–	122	IPF patients	ATS/ERS	sPAP ≥36 mmHg (from	61

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	PH Case Ascertainment		Prevalence (%)
S[49]		2009		identified from a database of a hospital's pneumology unit[b]		ECHO)		
Hyldgaard C[48]	Denmark	2003 – 2009	121	IPF patients identified from an Interstitial Lung Disease Registry at a University Hospital,	ATS/ERS/JRS/ALAT	tricuspid pressure regurgitation gradient ≥40 mmHg, a tricuspid annular plane systolic excursion <1.8 cm or right ventricular dilatation on ECHO and/or mPAP ≥25 mmHg (from RHC).		21
Minai OA[64]	US	1990– 2007	119	Consecutive IPF patients evaluated for lung transplantation at the Cleveland Clinic	ATS/ERS/JRS/ALAT	PH	mPAP ≥25 mmHg (RHC)	44
						PVH	mPAP ≥25 mmHg and PAOP >15 mmHg (RHC)	13
						PAH	mPAP ≥25 mmHg and PAOP ≤ 15 mmHg (RHC)	29
Nathan SD[59]	US	1997– 2005	118	IPF patients seen at a tertiary referral center with both PFT and RHC data available (potential	ATS/ERS	mPAP ≥25 mmHg (from RHC)		41

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	PH Case Ascertainment		Prevalence (%)
				overlapping samples with Nathan SD[60])				
Gagermeier J[62]	US	NR	117	Patients identified from the database of the Simmons Center for ILD	NR	PH	sPAP ≥30 mmHg (from ECHO)	40
						Moderate to severe PH	Right ventricular systolic pressure ≥45 mmHg (from ECHO)	17
Nathan SD[60]	US	1996–2006	110	IPF patients from 2 large tertiary centers in whom both ECHO and RHC were available (potential overlapping samples with Nathan SD[59])	ATS/ERS	mPAP >25 mmHg (from RHC)		35
Kimura M[74]	Japan	2001–2009	101	IPF patients identified from a database from a single hospital	ATS/ERS	mPAP >25 mm Hg (from RHC)		15
						mPAP >35 mm Hg		4

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	PH Case Ascertainment		Prevalence (%)
Mejia M[6]	Mexico	1996–2006	97	Clinical records for consecutive IPF patients at the National Institute of Respiratory Diseases	ATS/ERS	PAH	sPAP ≥45 mmHg (from ECHO)	80
						Mild/ moderate PAH	sPAP ≥50 mmHg (from ECHO)	67
						Severe PAH	sPAP ≥75 mmHg (from ECHO)	30
Nadrous HF[5]	US	1994–1996	88	IPF patients evaluated at Mayo Clinic in Rochester, who had complete results available from a comprehensive ECHO evaluation within 3 months of their initial visit	ATS/ERS	PH	sPAP >35 (from ECHO)	84
						Mild/moderate PH	sPAP >35–≤50 mmHg (from ECHO)	53
						Severe PH	sPAP >50 mmHg (from ECHO)	31
Swigris JJ[51]	US	2006–2009	82	IPF patients who performed a 6MWT as part of their clinical evaluation and were followed	ATS/ERS	mPAP ≥25 mmHg (from RHC) in the face of a pulmonary artery occlusion pressure that was ≤15 mmHg		26

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	PH Case Ascertainment	Prevalence (%)
				to lung transplantation or death			
Boutou AK[42]	Greece	2002–2007	81	Consecutive IPF patients evaluated over a 6-year period	ATS/ERS	sPAP >35 mmHg (from ECHO)	57
						Severe: sPAP >50 mmHg (from ECHO)	20
						mild/moderate: sPAP 35–50 mmHg (from ECHO)	37
Lettieri CJ[57]	US	1998–2004	79	Medical records from all IPF patients at Walter Reed Army Medical Center, Washington, D.C., who had undergone RHC as part of their evaluation prior to being listed for lung transplantation	ATS/ERS	mPAP >25 mmHg (from RHC)	32
Hamada K[69]	Japan	1991–2004	70	Consecutive IPF patients undergoing initial workup with RHC and PFT	ATS/ERS	mPAP >25 mmHg (from RHC)	8
Zisman DA[55]	US	1999–2006	65	Cross-sectional study of patients with advanced IPF	ATS/ERS	mPAP >25 mmHg (from RHC)	42

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	PH Case Ascertainment	Prevalence (%)
				and available RHC and HRCT			
Carbone R[45]	Italy	1985–2006	59	IPF patients referred to a single center	ATS/ERS	sPAP >55 mmHg (from ECHO)	66
Modrykamien AM[63]	US	1990–2007	58	Lung transplant patients with pre-transplant IPF diagnosis	Pathology reports showing presence of UIP pattern in explanted lung	PAH post-transplant mPAP >25 mmHg (RHC) and PAOP <15 mmHg	43
Ventetuolo CE[50]	US	2007–2009	52	Patients from the New York Presbyterian/Columbia University Medical Center ILD and Lung Transplantation Programs with both hemodynamics and banked plasma	ATS/ERS	mPAP ≥25 mmHg (from RHC)	31
Nakayama I[65],[a]	US	NR (3-year study)	50	Patients seen at 2 hospitals who had an ICD-9 code for IPF, had both ECHO and PFT, and who had not been referred for lung transplantation	ICD-9 code	By tricuspid regurgitation gradient (from ECHO) (specific diagnostic cut-off for PH NR)	50
Andersen CU[40]	Denmark	NR	49	Patients recruited during 16 months at	ATS/ERS	mPAP >25 mmHg (from RHC); TR >40	79

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	PH Case Ascertainment	Prevalence (%)
				a tertiary referral center for evaluation and treatment of ILD		mmHg; TAPSE <1.8 cm or right ventricular dilatation (from ECHO)	
Fernandez Perez ER[28]	US	1997–2005	47	Population-based sample of adult patients with IPF in Olmsted County, Minn.	ATS/ERS	Right ventricular systolic pressure ≥40 mmHg and peak TR ≥2.9 m/s on transthoracic ECHO	53
Alhamad EH[71]	Saudi Arabia	2008–2010	45	Consecutive IPF patients from a single center	ATS/ERS	mPAP ≥25 mmHg (from RHC)	38
Nathan SD[54]	US	2000–2005	44	IPF patients at single hospital who underwent lung transplantation and in whom serial RHCs were available	ATS/ERS	mPAP ≥25 mmHg (from RHC)	At time of transplant evaluation: 39; at time of transplant: 86
Handa T[72]	Japan	2004–2005	39	Consecutively enrolled patients who were being evaluated for PH by Doppler ECHO	Histological confirmation or clinically diagnosed based on typical clinical and radiographic findings	sPAP ≥40 mmHg; mild PH sPAP 40–50 mmHg (from ECHO)	36
Van Der Plas	Not Reported	NR	38	ILD protocol database	NR	sPAP ≥40 mmHg (from ECHO)	29

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	PH Case Ascertainment	Prevalence (%)
MN[47],[a]	(Europe)						
Saggar R[53]	US	2003–2007	38	All lung transplant patients at the UCLA	ATS/ERS	Assessed by RHC, cut-offs for PAH diagnosis NR	42 (pre-transplant)
Simon-Blancal V[18]	France	2002–2009	Survivors: n=27; non-survivors: n=10	Medical records of all IPF patients that experienced an AE and were hospitalized	ATS/ERS	sPAP >50 mmHg (from ECHO)	Survivors: 44; non-survivors: 60
Poor H[66],[a]	US	NR	37	IPF patients from single medical clinic	ATS/ERS	NR	31
Laz [N[73],a]	Egypt	NR	33	NR	On the basis of clinical data, plain chest radiography, HRCT, and presence of restrictive pulmonary dysfunction	sPAP >40 mmHg (from ECHO)	36
Bodlet A[25]	Belgium	1981 - 2011	32	Medical files IPF patients at single university hospital	ATS/ERS/JRS/ALAT	sPAP>30 mmHg (ECHO)	38
Agarwal R[68]	India	NR	25	IPF patients (no other detail provided)	ATS/ERS	sPAP: ≥40 mmHg or pulmonary acceleration time ≤100 milliseconds or 2-dimensional ECHO findings of the right ventricular enlargement	36

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	PH Case Ascertainment	Prevalence (%)
Zisman DA[58]	US	1997–2007	18	Cross-sectional study of IPF patients from 2 institutions	ATS/ERS	mPAP > 25 mmHg (from RHC)	32
Wiggins J[46]	UK	NR	8	Current or ex-heavy smokers with IPF who were referred to single center for further assessment and advice on treatment	"clinical diagnosis"	Clinical evidence of PH supported by ECHO	25

[a] Abstract only; [b] These studies likely have some overlap is patient samples

Abbreviations: 6MWT: Six-minute walk test; AE: Acute exacerbation; BNP: Brain natriuretic peptide; ECHO: Echocardiography; ILD: Interstitial lung disease; NR: Not reported; PAH: Pulmonary arterial hypertension; PAOP: Pulmonary artery occlusion pressure; PFT: Pulmonary function testing; PVH: Pulmonary venous hypertension; PWP: Pulmonary capillary wedge pressure; RHC: Right heart catheterization; TAPSE: Tricuspid annular plane systolic excursion; TR: Tricuspid pressure regurgitation; UIP: usual interstitial pneumonia; UK: United Kingdom; UNOS: United Network for Organ Sharing; US: United States

Supplementary Table S3. Prevalence of Obstructive Sleep Apnea among IPF Patients

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Obstructive Sleep Apnea Case ascertainment		Prevalence (%)
Collard HR[26]	US	2001–2008	9,286	Patients with IPF identified from two US claims databases	ICD-9 codes	ICD-9 codes		6
Lancaster LH[7]	US	2006–2008	50	Patients with IPF who had been followed up in the Vanderbilt Pulmonary Clinic	ATS/ERS	OSA	AHI of ≥5 events per hour	88
						Mild OSA	5.1–15 events/hour	20
						Moderate-to-severe OSA	>15 events/hour	68
Fernandez Perez ER[28]	US	1997–2005	47	Population-based sample of adult patients with IPF in Olmsted County, Minn.	ATS/ERS	NR		17
Mermigkis C[77]	Greece	2007–2009	34	Consecutive IPF patients evaluated at 4 pulmonary departments	ATS/ERS	OSA	AHI of ≥5 events per hour	59
						Mild OSA	5–15 events/hour	44
						Moderate-to-severe OSA	>15 events/hour	15

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Obstructive Sleep Apnea Case ascertainment		Prevalence (%)
Kolilekas L[79]	Greece	NR	31	Consecutive IPF patients referred to an outpatient interstitial lung disease unit who underwent overnight PSG	ATS/ERS	OSA	AHI of ≥5 events per hour	91
						Mild OSA	5–15 events/hour	39
						Moderate-to-severe OSA	>15 events/hour	52
Lee RNC[78, 80]	Ireland	2009–2012	20	IPF Patients selected from a Hospital IPF database who underwent overnight PSG	ATS/ERS	AHI ≥5/hour		45
						AHI ≥ 5 hour & significant daytime sleepiness		10
Mermigkis C[81]	US	2001–2005	18	Patients with IPF admitted to the Cleveland Clinic who had an all-night PSG	ATS/ERS	Reduction in airflow >90% lasting ≥10 s in which there was evidence of persistent respiratory effort (AHI of ≥5 events per hour)		61

[a] Five patients had AHI >5, though only one reported daytime sleepiness and thus was diagnosed with OSA.

Abbreviations: AHI: Apnea-hypopnea index; NR: Not reported; PSG: Polysomnogram

Supplementary Table S4. Frequency of Lung Cancer among IPF Patients

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Lung Cancer Case Ascertainment	Prevalence (%)	Incidence
Collard HR[26]	US	2001–2008	9,286	Patients with IPF identified from 2 claims databases	ICD-9 Codes	ICD-9 Codes	3	11.8/1,000 PY
Lee KJ[89]	Korea	2003–2007	1,685	Medical records of IPF patients from all tertiary and teaching university hospitals of more than 500 beds that employ pulmonary specialists	ATS/ERS	Pathologic biopsies	7	1.03/100 PY
Le Jeune I[82]	UK	NR ("up to 2004")	1,064	IPF patients identified from the THIN database	Read Codes	Read Codes	3	1.12/100 PY
Hubbard R[83]	UK	1988	890	IPF patients identified from the GPRD database	Read Codes	Read Codes	4	NR
Park J[90]	Korea	1989–1998	281	Patients diagnosed with	Diffuse interstitial	Lung biopsies	22	NR

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Lung Cancer Case Ascertainment	Prevalence (%)	Incidence
				IPF at single medical center during the study period	lung disease without known aetiology; compatible clinical findings; and pathological confirmation by surgical lung biopsy, or HRCT			
Kim ES[92]	Korea	2005–2009	268	IPF patients from a tertiary care hospital	ATS/ERS	NR	4	NR
Lee HJ[91]	Korea	1988–1995	244	Consecutive IPF patients evaluated at Seoul National University College of Medicine	CT and clinical findings or histologically	Histologically confirmed based on surgery, sputum cytology, percutaneous needle aspiration biopsy,	13	NR

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Lung Cancer Case Ascertainment	Prevalence (%)	Incidence
						transbronchial lung biopsy, or pleural fluid cytology		
Hyldgaard C[48]	Denmark	2003 – 2009	121	IPF patients identified from an Interstitial Lung Disease Registry at a University Hospital,	ATS/ERS/JRS/ALAT	NR	6	3.6%/year
Xu YX[84]	China	1999– 2009	104	Medical records of all IPF + lung cancer and IPF-alone patients admitted to the Peking Union Medical College hospital	diffuse interstitial lung disease with unknown etiology; compatible clinical findings, such as inspiratory crackles in both lower lung fields; and pathological confirmation	NR	23	NR

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Lung Cancer Case Ascertainment	Prevalence (%)	Incidence
					or HRCT showing typical patterns of IPF			
Ozawa Y[85]	Japan	1986–2005	103	IPF patients without lung cancer at the time of their initial diagnosis	ATS/ERS	NR	20[a]	Cumulative incidence 1 year: 3.3% 5 years: 15.4% 10 years: 54.7%
Nagai A[86]	Japan	1980–1992	99	Patients examined at Tokyo's Women's Medical College hospital	findings of chest radiography, lung biopsy, bronchoalveolar lavage, serum immunological examinations and pulmonary	Histologic examination was performed on specimens obtained by transbronchial lung biopsy, surgical treatment or autopsy	31	NR

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Lung Cancer Case Ascertainment	Prevalence (%)	Incidence
					function tests as well as detailed historical evaluation.			
Araki T[34]	Japan	1978–1997	86	Consecutive autopsy series of IPF patients >65 years	ATS/ERS	NR	22	NR
Matsushita H[88]	Japan	1972–1992	83	Consecutive autopsy cases in Toranomon Hospital	Clinical findings; histological changes and pathological diagnosis from autopsy	Chest X-ray films or CT scans, and macroscopical examinations	48	NR

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Lung Cancer Case Ascertainment	Prevalence (%)	Incidence
Qunn L[87]	Japan	1973–1996	72	Consecutive autopsy cases	based on the clinical symptoms, reduced pulmonary function, reticulonodular shadow on chest radiography, and histopathology of UIP	Lung biopsies	43	NR
Fernandez Perez ER[28]	US	1997–2005	47	Population-based sample of adult patients with IPF in Olmsted County, Minn.	ATS/ERS	ICD-9 Codes	8	NR

[a]Twenty percent of patients developed lung cancer during the study period, post-IPF diagnosis.

Abbreviations: GPRD: General Practice Research Database; HRCT: high-resolution computed tomography; NR: Not reported; THIN: The Health Improvement Network; UIP: usual interstitial pneumonia

Supplementary Table S5. Prevalence of PE among IPF Patients

Reference	Country	Study Period	Sample Size	Sample Source	IPF Diagnostic Criteria	PE Case Ascertainment	Prevalence (%)
Collard HR[26]	US	2001–2008	9,286	IPF patients identified through medical claims	ICD-9 codes	ICD-9 codes	3
Tomassetti S[49]	Italy	2000–2009	122	Patients with IPF identified in a database of a hospital's pneumology unit	ATS/ERS	Recorded in medical record	3
Saydain G[100]	US	1995–2000	38	IPF patients admitted to the ICU	(1) surgical biopsy showing UIP; (2) abnormal pulmonary function studies that included evidence of restriction, and/or increased alveolar-arterial oxygen tension gradient at rest or during exercise, or decreased diffusing capacity for carbon monoxide; and (3) chest radiograph or HRCT suggestive of UIP. In the absence of surgical biopsy,	NR	6

Reference	Country	Study Period	Sample Size	Sample Source	IPF Diagnostic Criteria	PE Case Ascertainment	Prevalence (%)
					patients had to fulfill all of the major criteria and ≥3 of the four minor criteria of the ATS/ERS		

Abbreviation: HRCT: high-resolution computed tomography; NR: Not reported; UIP: usual interstitial pneumonitis

Supplementary Table S6. Prevalence of Gastroesophageal Reflux among IPF Patients

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	GERD Case Ascertainment	Prevalence (%)
Collard HR[26]	US	2001–2008	9,286	IPF patients identified from claims databases	ICD-9 codes	ICD-9 codes	7
Gribbin J[102]	UK	1991–2003	920	IPF patients identified from UK general practices contributing data to THIN	Read codes	Read codes	30
Ryerson CJ[27]	US	2000–2010	242	IPF patients identified from a longitudinal ILD database	ATS/ERS	Patient questionnaire and chart review	36
Lee JS[10]	US	2001–2008	204	IPF patients identified from longitudinal cohorts of patients with ILD from 2 institutions	ATS/ERS/JRS/ ALAT	History of Nissen fundoplication (with indication for the treatment of GERD)	5
						Reported symptoms of heartburn or regurgitation	33
						Patient or physician reporting of GERD diagnosis	45
						Use of proton pump inhibitor or H2 blocker	47
Sherbini N[75]	Saudi Arabia	2007–2012	134	IPF patients' data from 2 tertiary care	ATS/ERS	NR	23

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	GERD Case Ascertainment	Prevalence (%)
				hospitals			
Lamas DJ[108]	US	2007–2010	129	IPF patients recruited from a single center	ATS/ERS	Patient questionnaire and medical record review	<1-year delay to first evaluation after onset of dyspnea: 21
							1-to-2-year delay to first evaluation after onset of dyspnea: 27
							2-to-4-year delay to first evaluation after onset of dyspnea: 28
							4-year delay to first evaluation after onset of dyspnea: 65
Hyldgaard C[48]	Denmark	2003 – 2009	121	IPF patients identified from an Interstitial Lung Disease Registry at a University Hospital,	ATS/ERS/JRS/ ALAT	NR	8
Garcia-Sancho C[106]	Mexico	2007–2009	100	Consecutive, newly diagnosed IPF patients from a single institution	ATS/ERS	Self-report	23

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	GERD Case Ascertainment	Prevalence (%)
Raghu G[4]	US	NR	65	Consecutive, newly referred IPF patients recruited from a single center	ATS/ERS	Self-reported heartburn and regurgitation	47
						On anti-reflux therapy (PPI or H_2 blocker) at time of of evaluation	66
						Abnormal acid GER among patients not receivi ng PPI therap (n=47)	87
						Abnormal distal acid exposure among patients not receivi ng PPI therap (n=47)	76
						Abnormal proximal acid exposure among patients not receivi ng PPI therap (n=47)	63
Vij R[109]	US	2005–2008	58	IPF patients recruited from a single clinic	ATS/ERS	NR	17
Corte TJ[103]	UK	1978–2005	56	IPF patients identified from a hospital database (n=232)	„met the histological criteria at surgical biopsy and the clinical criteria“	NR	38

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	GERD Case Ascertainment	Prevalence (%)
Savarino E[115]	Italy	2007–2011	40	Consecutive IPF patients	Based on the absence of an identifiable aetiology for ILD and a histopathological/radiological pattern of UIP on surgical lung biopsy and HRCT scans	GERD symptoms: heartburn and regurgitation	48
						GERD medications	40
						abnormal distal acid exposure	83
Fahim A[104]	UK	NR	40	IPF patients attending a single hospital clinic	ATS/ERS	Hull Airway Reflux Questionnaire	68
Kolilekas L[79]	Greece	NR	31	Consecutive IPF patients referred to an Outpatient Interstitial Lung Disease Unit who underwent overnight PSG	ATS/ERS	NR	81
Sweet MP[111]	US	1999–2006	30	IPF patients on lung transplant list referred to a single center	surgical lung biopsy; pathologic review of explanted lung; or ATS/ERS	DeMeester score	67
Lozo Vukovac E[114]	Croatia	2006 - 2010	30	Newly diagnosed IPF patients from a single hospital who had esophagogastroscopy (EGS) 1 week after IPF diagnosis	NR	Previous diagnosis	57
						Reflux symptoms	80

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	GERD Case Ascertainment	Prevalence (%)
Bandeira CD [113]	Brazil	2004–2008	28	Consecutive IPF patients who underwent esophageal manometry, 24-h esophageal pH-metry and pulmonary function tests at 2 institutions	ATS/ERS	Abnormal 24-hour esophageal pH-metry	36
D'Ovidio F[105]	Canada	2002–NR	26	Consecutive end-stage IPF patients assessed for lung transplantation	NR	Esophageal manometry, two-channel esophageal 24-hour pH testing, and gastric emptying studies were performed to determine the presence or absence of typical gastroesophageal reflux symptoms: heartburn, regurgitation, and dysphagia	65
Lee RNC[80]	Ireland	2009–2012	20	IPF Patients selected from a Hospital IPF database who underwent overnight PSG	ATS/ERS[a]	NR	0
Salvioli	Italy	NR	18	Consecutive IPF patients	Medical history, pulmonary	GERD – abnormal esophageal acid	67

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	GERD Case Ascertainment	Prevalence (%)
B[101]					function tests, HRCT (specific criteria NR)	exposure. Percent of time with pH lower than 4 >4.7% during the 24-h study period (T %pH<4), interdigestive	
						Typical GERD symptoms: Heartburn, regurgitation	72
Patti MG[110]	US	2003–2004	18	IPF patients on lung transplant list referred to a single center	NR	DeMesteer score >14.7	67
Tobin RW[112]	US	NR	17	Consecutive, newly diagnosed IPF patients recruited from a single center	Medical history with negative exposure history; no serologic evidence of autoimmune disease; chest radiograph with diffuse parenchymal, basilar-predominant infiltrates; pulmonary function tests with restrictive lung defect and decreased diffusing capacity of carbon monoxide corrected to hemoglobin (Dl COc); and histologic features of usual interstitial	pH probe study – Abnormal distal acid exposure only	24
						pH probe study – Abnormal distal and proximal acid exposure	65
						pH probe study – Abnormal acid exposure in the distal and/or proximal esophagus	94
						pH probe study – Abnormal esophageal acid	67

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	GERD Case Ascertainment	Prevalence (%)
					pneumonia on surgical lung biopsy	exposure	
Soares RV[107]	US	2008–2009	16	IPF/NSIP patients prospectively collected from the referrals for esophageal function tests at a single center	ATS/ERS	pH probe study – Abnormal proximal reflux only	23
						pH probe study – Abnormal distal reflux only	56

[a] "confirmed diagnosis of IPF by ATS criteria" Abbreviations: GERD: Gastroesophageal reflux disease; HRCT: high-resolution computed tomography; ILD: interstitial lung disease; NR: Not reported; NSIP: Non-specific interstitial pneumonia; THIN: The Health Improvement Network; UIP: usual interstitial pneumonia

Supplementary Table S7. Prevalence of IPF Patients with Arrhythmia or Atrial Fibrillation

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Arrhythmia, AF Case Ascertainment	Prevalence (%)
Collard HR[26]	US	2001–2008	9,286	IPF patients identified through medical claims databases	ICD-9 codes	ICD-9 codes	AF: 12
Hubbard RB[117]	UK	NR	920	THIN primary care dataset; IPF cases pre-IPF diagnosis	Read codes	Read codes	AF: 6
Sherbini N[75]	Saudi Arabia	2007–2012	134	IPF patients' data from 2 tertiary care hospitals	ATS/ERS	NR	16
Hyldgaard C[48]	Denmark	2003 – 2009	121	IPF patients identified from an Interstitial Lung Disease Registry at a University Hospital,	ATS/ERS/JRS/ALAT	NR	9
Fernandez Perez ER[28]	US	1997–2005	47	IPF patients identified via the REP, Olmsted County, Minn.	ATS/ER	NR	AF: 19
Daniels CE[118]	US	1996–2004	42	Consecutive patients with IPF who underwent a postmortem evaluation	Post mortem evidence of UIP with no connective tissue disease or exposure to fibrogenic	NR	Arrhythmia: 7

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Arrhythmia, AF Case Ascertainment	Prevalence (%)
					drugs or environmental agents having been identified		
Saito Y[96]	Japan	1994–2007	28	IPF patients with stage IA NSCLC	ATS/ERS	NR	Arrhythmia: 7

Abbreviations: NR: Not reported; NSCLC: Non–small cell lung cancer; REP: Rochester Epidemiology Project; THIN: The Health Improvement Network; UIP: usual interstitial pneumonia

Supplementary Table S8. Prevalence of IPF Patients with Cardiac Failure or CHF

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Cardiac Failure Case Ascertainment	Prevalence (%)	
Collard HR[26]	US	2001–2008	9,286	IPF patients identified through ICD-9 codes in medical claims	ICD-9 codes	ICD-9 codes	20	
Tomassetti S[43], [a]	Italy	2002–2009	147	Cases from an IPF clinic at GB Morgagni Hospital	ATS/ERS	NR	IPF patients not on anticoagulants	9
			35				IPF patients on anticoagulants	26
Tomassetti S[49]	Italy	2000–2009	122	IPF patients identified in a database of a hospital's pneumology unit	ATS/ERS	Recorded in medical record	14	
Nadrous HF[5]	US	1994–1996	88	IPF patients evaluated at a tertiary care, referral medical center	ATS/ERS	NR	11	
Rusanov	Israel	2009	61	IPF patients	ATS/ERS	NR	18	

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Cardiac Failure Case Ascertainment	Prevalence (%)
V[119]				referred for lung transplantation			
Fernandez Perez ER[28]	US	1997–2005	47	IPF patients identified via the REP, Olmsted County, Minn.	ATS/ERS	NR	11
Saydain G[100]	US	1995–2000	38	IPF patients admitted to the Mayo Clinic ICU, Rochester, MN	(1) surgical biopsy showing UIP; (2) abnormal pulmonary function studies that included evidence of restriction, and/or increased alveolar-arterial oxygen tension gradient at rest or	≥1 of the following: Heart rate ≤.54/min.; Mean arterial blood pressure ≤.49 mmHg; ventricular tachycardia and/or ventricular Fibrillation; Serum pH ≤.7.24 with a P_aCO_2 of ≤.49 mmHg.	5

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Cardiac Failure Case Ascertainment	Prevalence (%)
					during exercise, or decreased diffusing capacity for carbon monoxide; and (3) chest radiograph or HRCT suggestive of UIP. In the absence of surgical biopsy, patients had to fulfill all of the major criteria and ≥3 of the four minor criteria of the ATS/ERS		

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Cardiac Failure Case Ascertainment	Prevalence (%)
Saito Y[96]	Japan	1994–2007	28	IPF patients with stage IA NSCLC	ATS/ERS	abnormal ECG result, BNP levels exceeding normal reference ranges,or by a clinical evaluation	4
Rangappa P[120]	Australia	1996–2006	24	IPF patients admitted to the ICU	ATS/ERS	ECHO	13

[a] abstract; Abbreviations: BNP: Brain natriuretic peptide; ECG: echocardiogram; ECHO: Echocardiography; NR: Not reported; NSCLC: Non–small cell lung cancer; REP: Rochester Epidemiology Project

Supplementary Table S9. Prevalence of IPF Patients with IHD

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	IHD Case Ascertainment	Prevalence (%)
Collard HR[26]	US	2001–2008	9,286	IPF patients identified through ICD-9 codes in medical claims	ICD-9 codes	(CAD) ICD-9 codes	25
Collard HR[26]	US	2001–2008	9,286	IPF patients identified through medical claims	ICD-9 codes	(MI) ICD-9 codes	3
Kim WY[131]	Korea	2005 – 2009	460	Database of IPF patients from a Medical Center	ATS/ERS/JRS/ALAT	CAD[f]	7
Lederer DJ[52]	US	2004–2005	454	IPF patients listed for lung transplantation with UNOS	NR	CAD (NR)	~4 (average among quintiles of results for patients completing 6MWD)
Park J[129],[a]	Korea	NR	324	IPF patients admitted to a tertiary referral center, diagnosed by coronary angiography	Lung biopsy	IHD (NR)	14
Navaratnam V[132]	UK	2010 – 2012	211	Incident cases of IPF from five teaching hospitals and eight district general hospitals	ATS/ERS	Previous IHD (NR)	31
Tomassetti S[43],[a]	Italy	2002–2009	182	Cases from an IPF clinic at GB Morgagni Hospital	ATS/ERS	IHD (NR)	Not on anticoagulants: 6 On

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	IHD Case Ascertainment	Prevalence (%)
							anticoagulants: 11
Papakosta D[41]	Greece	2005–2006	139	IPF patients referred to 8 departments of pneumonology	ATS/ERS	CAD (NR)	17
Lamas DJ[108]	US	2007–2010	129	IPF patients evaluated at a tertiary care center	ATS/ERS	CAD (NR)	15–26 (dependent on delay in access to care)
Tomassetti S[49]	Italy	2000–2009	122	IPF patients identified in a database of a hospital's pneumology unit	ATS/ERS	CAD Recorded in medical record	20
Tomassetti S[49]	Italy	2000–2009	122	Patients with IPF identified in a database of a hospital's pneumology unit	ATS/ERS	MI Recorded in medical record	12
Hyldgaard C[48]	Denmark	2003 – 2009	121	IPF patients identified from an Interstitial Lung Disease Registry at a University Hospital,	ATS/ERS/JRS/ALAT	IHD (NR)	18
Miyake Y[126]	Japan	2001	104	IPF patients identified across ~20 hospitals	ATS/ERS	CAD[e]	10
Schomberg LEE[127],	UK	2003–2010	96	Consecutive cases of IPF from a health care provider, diagnosed by	Radiology reports	STEMI (NR)	21

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	IHD Case Ascertainment	Prevalence (%)
[a]				radiology reports			
Nadrous HF[5]	US	1994–1996	88	IPF patients evaluated at a tertiary care, referral medical center	ATS/ERS	CAD (NR)	35
Swigris JJ[123]	US	2003–2008	76	IPF patients evaluated at an ILD center	ATS/ERS	CAD (NR)	20
Nathan SD[121]	US	2003–2008	73	IPF patients who completed LHC as part of lung transplant evaluations	ATS/ERS	Non-significant and significant CAD[b]	66
						Non-significant CAD[b]	37
						Significant CAD[b]	29
Rusanov V[119]	Israel	2009	61	IPF patients referred for lung transplantation	ATS/ERS	IHD (NR)	13
Alhamad EH[130]	Saudi Arabia	1996–2005	61	IPF patients admitted to a hospital	ATS/ERS	IHD (NR)	8
Nathan SD[122]	US	2003–2008	57	IPF patients who completed LHC as part of lung transplant evaluations	ATS/ERS	Non-significant CAD[b]	40
						Significant CAD[b]	28
Weir	US	2003–	52	IPF patients who had	NR	Significan	25

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	IHD Case Ascertainment	Prevalence (%)
N[124],[a]		2008		undergone HRCT and LHC		t CAD[c]	
Ponnuswamy A[128]	UK	1999–2004	50	IPF patients diagnosed at a respiratory outpatient clinic	HRCT	IHD (NR)	40
Izbicki G[125]	Israel	1997–2003	49	IPF patients who were candidates for lung transplantation	ATS/ERS	Significant CAD[d]	29
Fernandez Perez ER[28]	US	1997–2005	47	IPF patients identified via the REP, Olmsted County, Minn.	ATS/ERS	CAD (NR)	45
Daniels CE[118]	US	1996–2004	42	Consecutive patients with IPF who underwent a postmortem evaluation	Post mortem evidence of UIP with no connective tissue disease or exposure to fibrogenic drugs or environme	MI (NR)	7

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	IHD Case Ascertainment	Prevalence (%)
					ntal agents having been identified		
Schomberg LEE[127],[a]	UK	2003–2010	38	Consecutive cases of IPF from a health care provider, diagnosed by radiology reports	Radiology reports	IHD (NR)	58
Lee RNC[80]	Ireland	2009–2012	20	IPF Patients selected from a Hospital IPF database who underwent overnight PSG	ATS/ERS[g]	Inactive CAD‘ NR	35

[a]Abstract;

[b] Nonsignificant CAD: <50% occlusion of a major vessel or disease of smaller vessels as diagnosed with LHC and CT scans; significant CAD: a need for an intervention or major vessel with >50% lesion as diagnosed with LHC and CT scans;

[c] Significant CAD: quantification of coronary calcification observed by LHC and HRCT results;

[d] Significant CAD: ≥50% stenosis of one or more coronary arteries, as reported by coronary angiography;

[e] CAD: having received medication for CAD

[f] symptom and the finding of coronary angiography, coronary computed tomography angiography, thallium scan, or exercise treadmill test.

[g] “confirmed diagnosis of IPF by ATS criteria”

Abbreviations: 6MWD: Six-minute walk distance; HRCT: high-resolution computed tomography; ILD: Interstitial lung disease; NR: Not reported; REP: Rochester Epidemiology Project; STEMI: ST-elevation myocardial infarction; UNOS: United Network for Organ Sharing

Supplementary Table S10. Prevalence of IPF Patients with Cerebrovascular Accident, Cerebrovascular Disease, and Stroke

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Cerebrovascular Accident, Cerebrovascular Disease, Stroke Case Ascertainment	Prevalence (%)
Cerebrovascular Accident							
Hubbard RB[117]	UK	NR	920	THIN primary care dataset; IPF cases pre-IPF diagnosis	Read codes	Read codes	6
Hyldgaard C[48]	Denmark	2003 – 2009	121	IPF patients identified from an interstitial lung disease registry at a university hospital,	ATS/ERS/JRS/ ALAT	(cerebrovascular infarction) NR	9
Cerebrovascular Disease							
Collard HR[26]	US	2001– 2008	9,286	IPF patients identified through medical claims	ICD-9 codes	ICD-9 codes	9
Stroke							
Daniels CE[118]	US	1996– 2004	42	Consecutive patients with IPF who underwent a postmortem evaluation	Post mortem evidence of UIP with no connective tissue disease or exposure to fibrogenic drugs or	NR	3

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Cerebrovascular Accident, Cerebrovascular Disease, Stroke Case Ascertainment	Prevalence (%)
					environmental agents having been identified		

Abbreviations: NR: Not reported; THIN: The Health Improvement Network;

Supplementary Table S11. Prevalence of IPF Patients with Arterial Hypertension

Reference	Country	Study Years	Sample Size	Study Population	IPF Diagnostic Criteria	Arterial Hypertension Case Ascertainment	Prevalence (%)
Lederer DJ[133]	US	1995–2003	2,635	IPF patients listed for lung transplantation with UNOS	NR	NR	19
Munson JC[23]	UK	1989–2006	1,126	IPF patients drawn from the THIN primary care dataset	Read codes	NR (Hypertension or Congestive Heart Failure)	32
Kim WY[131]	Korea	2005–2009	460	Database of IPF patients from a Medical Center	ATS/ERS/JRS/ALAT	blood pressure ≥140/90 mmHg, treatment with antihypertensive medication, or history by questionnaire	35
Lederer DJ[52]	US	2004–2005	454	IPF patients listed for lung transplantation with UNOS	NR	NR	15
Park J*[129]	Korea	NR	324	IPF patients admitted to a tertiary referral center	Lung biopsy	NR	34
Papakosta	Greece	2005–	139	IPF patients	ATS/ERS	NR	37

Reference	Country	Study Years	Sample Size	Study Population	IPF Diagnostic Criteria	Arterial Hypertension Case Ascertainment	Prevalence (%)
D[41]		2006		referred to 8 departments of pneumonology			
Sherbini N[75]	Saudi Arabia	2007–2012	134	IPF patients' data from 2 tertiary care hospitals	ATS/ERS	NR	39
Hyldgaard C[48]	Denmark	2003–2009	121	IPF patients identified from an Interstitial Lung Disease Registry at a University Hospital,	ATS/ERS/JRS/ALAT	NR	18
Kim YJ[135]	Korea	2000–2006	114	IPF patients admitted to a hospital	ATS/ERS	(1) Known arterial hypertension with treatment including antihypertensive agent, diet, exercise et al. or (2) systolic BP ≥140 mmHg, or diastolic BP ≥90 mmHg	20
Miyake Y[126]	Japan	2001	104	IPF patients identified across	ATS/ERS	Having received medication for	27

Reference	Country	Study Years	Sample Size	Study Population	IPF Diagnostic Criteria	Arterial Hypertension Case Ascertainment	Prevalence (%)
				~20 hospitals IPF patients		arterial hypertension	
Nadrous HF[5]	US	1994–1996	88	IPF patients evaluated at a tertiary care, referral medical center	ATS/ERS	NR	44
Rusanov V[119]	Israel	2009	61	IPF patients referred for lung transplantation	ATS/ERS	NR	55
Alhamad EH[130]	Saudi Arabia	1996–2005	61	IPF patients admitted to a hospital	ATS/ERS	NR	22
Enomoto T[134]	Japan	1995–2000	52	IPF patients admitted to a hospital	ATS/ERS	Systolic BP >140 mmHg and/or diastolic BP >90 mmHg, and/or patients had a history of treatment with antihypertensive drugs	24

Reference	Country	Study Years	Sample Size	Study Population	IPF Diagnostic Criteria	Arterial Hypertension Case Ascertainment	Prevalence (%)
Izbicki G[125]	Israel	1997–2003	49	IPF patients who were candidates for lung transplantation	ATS/ERS	Arterial BP ≥140/90 mmHg or treatment with ≥1 antihypertensive agents	14
Fernandez Perez ER[28]	US	1997–2005	47	IPF patients identified via the REP, Olmsted County, Minn.	ATS/ERS	NR	66
Schomberg LEE*[127]	UK	2003–2010	38	Consecutive cases of IPF from a health care provider	Radiology reports	NR	71
Rufino RL*[39]	Brazil	2008–2010	36	IPF patients referred to an outpatient clinic	ATS/ERS	NR	44

Abbreviations: BP: Blood pressure; REP: Rochester Epidemiology Project; NR: Not reported; THIN: The Health Improvement Network; UNOS: United Network for Organ Sharing

Supplementary Table S12. Prevalence of Diabetes among IPF Patients

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Diabetes Case Ascertainment	Prevalence (%)
Munson JC[23]	UK	1989–2006	1,126	Patients with IPF identified from THIN database	Read codes	Read codes	14
Gribbin J[102]	UK	1991–2003	920	Patients with IPF identified from THIN database	Read codes	Read codes	10
Kim WY[131]	Korea	2005 – 2009	460	Medical center database of IPF patients	ATS/ERS/JRS/ALAT	FG ≥126 mg/dL, use of oral hypoglycemic agents or insulin, or history by questionnaire	20
Park J[129]	Korea	NR	324	IPF patients from a tertiary referral center	Lung biopsy	NR	24
Kim ES[92]	Korea	2005–2009	268	IPF patients from a tertiary care hospital	ATS/ERS	NR	18
Papakosta D[41]	Greece	2005–2006	139	IPF patients referred to 8 pneumonology departments	ATS/ERS	NR	18
Sherbini N[75]	Saudi Arabia	2007–2012	134	IPF patients' data from 2 tertiary care hospitals	ATS/ERS	NR	42
Lamas DJ[108]	US	2007–2010	129	IPF patients from a tertiary care center stratified by time from symptom onset to care	ATS/ERS	NR	12–39
Hyldgaard C[48]	Denmark	2003 – 2009	121	IPF patients identified from an interstitial lung disease registry at a University Hospital,	ATS/ERS/JRS/ALAT	NR	17
Garcia-	Mexico	2007–	100	Newly-diagnosed IPF	ATS/ERS	Self-reported	30

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Diabetes Case Ascertainment	Prevalence (%)
Sancho C[106]		2009		patients consecutively seen at a single institution		from questionnaire	
Garcia-Sancho Figueroa Mac[136]	Mexico	2000–2005	97	Records of consecutive IPF patients seen at a single institute	ATS/ERS	One of the following: 1) FG >126 mg/dl in the absence of corticosteroids treatment; 2) patient-reported diagnosis from physician; 3) patient received diagnosis of T2DM at clinic during first consult; 4) patient took oral T2DM drugs or insulin	11
Schomberg LEE[127][a]	UK	2003–2010	96	Consecutive IPF patients from a health care provider	Radiology reports	NR	24
Rusanov V[119]	Israel	2009	61	Patients with IPF diagnosed referred for lung transplant	ATS/ERS	NR	27
Alhamad EH[130]	Saudi Arabia	1996–2005	61	IPF patients at a university hospital	ATS/ERS	NR	25
Enomoto	Japan	1995–	52	Patients admitted to Nippon	ATS/ERS	FG>126 mg/dL	33

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Diabetes Case Ascertainment	Prevalence (%)
T[134]		2000		Medical School Hospital with IPF		and/or HbAIc >6%, or DM therapy, including diet, exercise, and/or medication	
Izbicki G[125]	Israel	1997–2003	49	Patients (age >40 years old) with lung fibrosis diagnosed by ATS/ERS criteria who were candidates for lung transplantation	ATS/ERS	FG >126 mg/dl or treatment with one or more oral or parenteral hypoglycemic medications	20
Rufino RL[39], [a]	Brazil	2008–2010	36	IPF Patients referred to an outpatient clinic at the State University of Rio de Janeiro	ATS/ERS	NR	12
Lee RNC[80]	Ireland	2009–2012	20	IPF Patients selected from a Hospital IPF database who underwent overnight PSG	ATS/ERS[b]	NR	20

[a]Abstract

[b] "confirmed diagnosis of IPF by ATS criteria"

Abbreviations: DM: Diabetes mellitus; FG: fasting glucose; HbAIc: Hemoglobin A1c; NR: Not reported; T2DM: Type 2 diabetes mellitus

Supplementary Table S13. Prevalence of Hypercholesterolemia/Hyperlipidemia among IPF Patients

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Hypercholesterolemia/ Hyperlipidemia Case Ascertainment	Prevalence (%)
Gribbin J[102]	UK	1991–2003	920	Patients with IPF identified using Read codes from THIN database	Read codes	Read codes	10
Kim WY[131]	Korea	2005 – 2009	460	Database of IPF patients from a Medical Center	ATS/ERS/JRS/ ALAT	T-chol ≥240 mg/dL or use of lipid-lowering therapy	15
Park J[129][a]	Korea	NR	324	IPF patients from a tertiary referral center	Lung biopsy	NR	30
Kim YJ[135]	Korea	2000–2006	114	IPF patients diagnosed at Gil Hospital and Samsung Medical Center in Korea	ATS/ERS	Either (1) known hyperlipidemia with treatment with any medication for hyperlipidemia, or (2) T-chol ≥200 mg/dL and/or triglyceride ≥150 mg/dL and/or LDL cholesterol ≥100 mg/dL based on ATP III classification	29
Miyake Y[126]	Japan	2001	104	Patients with IPF, aged ≥40 years	ATS/ERS	NR	10
Enomoto	Japan	1995–	52	IPF patients	ATS/ERS	T-chol >240 mg/dL	19

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Hypercholesterolemia/ Hyperlipidemia Case Ascertainment	Prevalence (%)
T[134]		2000		admitted to Nippon Medical School Hospital		and/or triglyceride >150 mg/d, and treatment with any medications for hyperlipidemia	
Izbicki G[125]	Israel	1997–2003	49	IPF Patients (age >40 years) who were candidates for lung transplantation	ATS/ERS	T- chol > 240 mg/dl or treatment with ≥1 lipid-lowering drug	22
Schomberg LEE[127],[a]	UK	2003–2010	38	Consecutive cases of UIP from a healthcare provider	Radiology reports	NR	55

[a] Abstract

Abbreviations: ATP: Adult Treatment Panel; LDL: Low-density lipoprotein; NR: Not reported; T-chol: Total cholesterol; THIN: The Health Improvement Network

Supplementary Table S14. Prevalence of Weight Disorders among IPF Patients

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Metabolic Comorbidity Case Ascertainment	Prevalence (%)
Collard HR[26]	US	2001–2008	9,286	IPF patients in Thomson Reuters MarketScan database	ICD-9 codes	Obesity (ICD-9 codes)	0.6
Munson JC[23]	UK	1989–2006	1,126	IPF patients in THIN database	Read codes	Obesity (Read codes)	18
Kim WY[131]	Korea	2005 – 2009	460	Database of IPF patients from a Medical Center	ATS/ERS/JRS/ALAT	Obesity (BMI ≥25.0)	39
Lee JS[10]	US	2001–2008	204	IPF patients at the University of California San Francisco and the Mayo Clinic	ATS/ERS/JRS/ALAT	Obesity (BMI ≥30)	37
						Overweight (25– 30)	39
						Underweight (BMI <18.5)	0.5
Alakhras M[137]	US	1994–1996	197	IPF patients who had been evaluated at Mayo Clinic Rochester	ATS/ERS	Obesity (BMI ≥30	34
						Overweight (BMI 25–30)	43
Izbicki G[125]	Israel	1997–2003	49	IPF patients (age >40 years old) who were candidates for lung transplantation	ATS/ERS	Obesity (BMI ≥27.0)	33
Enomoto	Japan	1995–	52	IPF patients	ATS/ERS	Obesity	19

Reference	Country	Study Period	Sample Size	Study Population	IPF Diagnostic Criteria	Metabolic Comorbidity Case Ascertainment	Prevalence (%)
T[134]		2000		admitted to Nippon Medical School Hospital		(BMI >25)	

Abbreviations: THIN: The Health Improvement Network