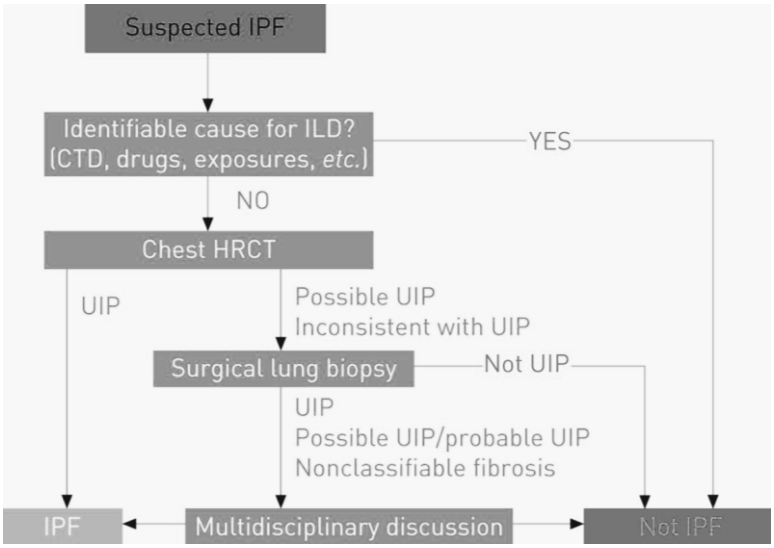
Diagnostic criteria for idiopathic pulmonary fibrosis (IPF)

(Raghu G. et al., Am J Respir Crit Care Med, 183. pp 788–824, 2011.
Accessible at http://www.atsjournals.org/doi/abs/10.1164/rccm.2009-040GL#.VYwfDkauoxI)

Diagnostic algorithm for idiopathic pulmonary fibrosis



IPF, idiopathic pulmonary fibrosis, ILD, interstitial lung disease; CTD, connective tissue disease; HRCT, high-resolution computed tomography; UIP, usual interstitial pneumonia

The diagnosis of IPF requires the exclusion of other known causes of ILD (e.g., domestic and occupational environmental exposures, connective tissue disease, and drug toxicity), the presence of a UIP pattern on HRCT in patients not subjected to surgical lung biopsy and specific combinations of HRCT and surgical lung biopsy pattern in patients subjected to surgical lung biopsy.

High-Resolution Computed Tomography (HRCT) criteria for Usual Interstitial Pneumonia (UIP) pattern

UIP pattern (all 4 features):
- Subpleural, basal predominance
- Reticular abnormality
- Honeycombing with or without traction bronchiectasis
- Absence of features listed as inconsistent with UIP pattern (*)

Possible UIP pattern (all 3 features):
- Subpleural, basal predominance
- Reticular abnormality
- Absence of features listed as inconsistent with UIP pattern (*)

** Inconsistent with UIP pattern (any of the features):*
- Upper or mid-lung predominance
- Peribronchovascular predominance
- Extensive ground-glass abnormality (extent > reticular abnormality)
- Profuse micronodules (bilateral, predominantly upper lobes)
- Discrete cysts (multiple, bilateral, away from areas of honeycombing)
- Diffuse mosaic attenuation/air-trapping (bilateral and in ≥3 lobes)
- Consolidation in bronchopulmonary segment(s)/lobe(s)

Histopathological criteria for Usual Interstitial Pneumonia (UIP) pattern

UIP pattern (all 4 features):

- Evidence of marked fibrosis/architectural distortion, ± honeycombing in a predominantly subpleural/paraseptal distribution
- Presence of patchy involvement of lung parenchyma by fibrosis
- Presence of fibroblast foci
- Absence of features against a diagnosis of UIP suggesting an alternate diagnosis (**)

Probable UIP pattern:

- Evidence of marked fibrosis/architectural distortion, ± honeycombing
- Absence of either patchy involvement or fibroblastic foci, but not both
- Absence of features against a diagnosis of UIP suggesting an alternate diagnosis (**)

OR

- Honeycomb changes only

Possible UIP pattern (all 3 features):

- Patchy or diffuse involvement of lung parenchyma by fibrosis, with or without interstitial inflammation
- Absence of other criteria for UIP (see UIP Pattern)
- Absence of features against a diagnosis of UIP suggesting an alternate diagnosis (**)

*** Not UIP pattern (any of the features):*

- *Hyaline membranes*
- *Organizing pneumonia*
- *Granulomas*
- *Marked interstitial inflammatory cell infiltrate away from honeycombing*
- *Predominant airway centered changes*
- *Other features suggestive of an alternate diagnosis*

Combination of high-resolution computed tomography (HRCT) and surgical lung biopsy for the diagnosis of IPF (requires multidisciplinary discussion)

	HRCT features	Pathological features
Definite IPF	UIP	No biopsy
		Definite UIP
		Probable UIP
		Possible UIP
		Unclassifiable fibrosis
	Possible UIP	Definite UIP
		Probable UIP
Probable IPF	Possible UIP	Possible UIP
		Unclassifiable fibrosis
Possible IPF	Inconsistent with UIP	Definite UIP
Not IPF	UIP	Non-UIP
	Possible UIP	Non-UIP
	Inconsistent with UIP	Probable UIP
		Possible UIP
		Unclassifiable fibrosis
		Not UIP