Pulmonary Pathology Society Survey on Practice Approaches in the Histologic Diagnosis of Fibrotic Interstitial Lung Disease: Consensus and Opportunities

Supplemental Digital Content.

PPS Fibrotic ILD/UIP Survey

Introduction: This survey is designed to assess the current clinical practice of PPS members with regard to fibrotic interstitial lung disease and specifically usual interstitial pneumonia. There are no right or wrong answers. The PPS working group on fibrotic ILD thanks you for your time and efforts. Our goal is to present this data for discussion at the upcoming 2022 PPS meeting.

Thank you!

Background Questions:

Practice Setting	◯ Teaching/Academic institution ◯ Private practice ◯ Other, specify
Other, specify	_____
What is your current practice status?	◯ Trainee ◯ Practicing Pathologist ◯ Other, specify
If trainee, what is your current trainee level?	◯ First year resident ◯ Second year resident ◯ Third year resident ◯ Fourth year resident ◯ Fellow
If practicing Pathologist, how many years have you been in practice?	_____
If practicing Pathologist, have you completed a fellowship in pulmonary pathology?	◯ Yes ◯ No
Other, specify	_____
Region of practice	◯ Asia ◯ Africa ◯ Oceania ◯ Europe ◯ Middle East ◯ North America ◯ South America
Approximate number of fibrotic ILD cases encountered per month:	_____
How many pathologists with pulmonary pathology expertise are in your practice, including yourself?	_____
Do you have direct access to colleagues in your practice with pulmonary pathology expertise to share difficult cases?	◯ Yes ◯ No
If yes, how frequently do you share ILD cases with your colleagues? (Enter percentage of cases)	_____ ((%))

Do you receive ILD cases in consultation from outside institutions?	◯ Yes ◯ No
If yes, approximately how many consultation cases do you personally review each month?	_____

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Histologic Guidelines for UIP:

Do you use or reference published guidelines for the histologic assessment of UIP in your clinical reports for cases with fibrotic ILD?

- ◯ Yes
- ◯ Sometimes
- ◯ No

If yes or sometimes, which guidelines do you use?

- ◯ 2018 ATS/ERS/JRS/ALAT
- ◯ 2011 ATS/ERS/JRS/ALAT
- ◯ 2018 Fleischer Society
- ◯ Other, specify

Other, specify

_____

If yes or sometimes, what level of adherence best describes your use of the guidelines strictly?

- ◯ Strict adherence to criteria
- ◯ Moderate adherence to criteria
- ◯ Loose adherence to criteria
- ◯ Gestalt approach
- ◯ Other, specify

Other, specify

_____

If yes or sometimes, do you include the guideline categorization in the diagnostic line or in the comment?

- ◯ Diagnostic line
- ◯ Comment/Note section
- ◯ Microscopic Description
- ◯ I don't include the categorization in my pathology reports
- ◯ Other, specify

Other, specify

_____

	They are complete and usable5	4	3	2	They are incomplete and have no role in pathology1
What is your impression of the completeness and usability of the current histologic 2018 ATS/ERS/ALAT/JRS UIP guidelines	◯	◯	◯	◯	◯

The ATS/ERS/ALAT/JRS guidelines refer to UIP secondary to another cause for cases that have UIP like fibrosis but also other histology to suggest a specific etiology aside from IPF. What is your concept of secondary UIP? (Select all that apply)

- ☐ Secondary UIP is a useful term for pathologists
- ☐ Secondary UIP is a useful term for clinicians
- ☐ Secondary UIP might be a useful term, but requires a more clear definition amongst the ILD community as a whole
- ☐ Secondary UIP is not a useful term and should not be used
- ☐ UIP pattern is a better term for such cases
- ☐ Other, specify
- ☐ None of the above

Other, specify

_____

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Clinical and radiologic history:

How do you incorporate clinical and radiologic history into your diagnosis/report	◯ I do not incorporate clinical and radiologic history. My diagnosis is based solely on what I see on the slides. ◯ I look at the slides first and then seek additional information if it is available. ◯ I may alter my pathologic diagnosis based on additional clinical and radiologic history if it is pertinent
Do you have access to discuss cases with ILD clinicians/pulmonologists?	◯ Yes ◯ No
If yes, how frequently do you discuss cases with clinicians/pulmonologists? (Enter percentage of cases)	_____ ((%))
Do you have access to discuss cases with thoracic radiologists?	◯ Yes ◯ No
If yes, how frequently do you discuss cases with radiologists? (Enter percentage of cases)	_____ ((%))
Do you have a formal MDD at your institution?	◯ Yes ◯ No

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Diagnostic line:

Which of the following is closest to how you sign out cases that meet your criteria for histologic UIP?

- ◯ UIP
- ◯ UIP pattern
- ◯ UIP with a comment
- ◯ Advanced fibrosing interstitial pneumonia consistent with UIP
- ◯ Advanced fibrosing interstitial pneumonia with a UIP pattern
- ◯ UIP compatible with IPF
- ◯ Other, specify

Other, specify

_____

How do you sign out cases of fibrosing interstitial pneumonia that do not have all the features of histologic UIP, but UIP is your leading diagnosis, ie probable UIP

- ◯ Fibrosing ILD most consistent with UIP
- ◯ Fibrosing ILD with some features of UIP
- ◯ A descriptive diagnosis would be warranted in this situation, with the differential diagnosis on the comments/note section.
- ◯ Other, specify

Other, specify

_____

How do you sign out cases of fibrosing interstitial pneumonia that have advanced fibrosis, fibroblast foci, spared areas of normal lung, and honeycomb but also contain features that suggest an alternative etiology

- ◯ UIP
- ◯ UIP pattern
- ◯ Secondary UIP
- ◯ UIP with additional feature
- ◯ UIP pattern with additional feature
- ◯ Advanced fibrosing ILD with additional feature
- ◯ Other, specify

Other, specify

_____

What is your impression of UIP

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
Histologic UIP is a pattern of injury seen in several fibroinflammatory diseases	◯	◯	◯	◯	◯
Histologic UIP is a pathologic diagnosis, regardless of etiology	◯	◯	◯	◯	◯
Histologic UIP is a pathologic diagnosis that requires clinical and radiologic consultation	◯	◯	◯	◯	◯
Histologic UIP is diagnostic of IPF	◯	◯	◯	◯	◯
Histologic UIP is a useful pathologic term	◯	◯	◯	◯	◯

Histologic UIP is also a radiologic diagnosis	○	○	○	○	○

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
Cases with perfect/classic histologic UIP (2018 criteria) are highly specific for the clinical syndrome IPF	○	○	○	○	○
Most cases signed out by the field as histologic UIP are likely IPF	○	○	○	○	○
Perfect histologic UIP can be seen in chronic HP	○	○	○	○	○
Perfect histologic UIP can be seen in CTD-ILD	○	○	○	○	○
Perfect histologic UIP can be seen in pneumoconioses	○	○	○	○	○

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Specific Histologic Features: Section 1

Please evaluate each of the following statements regarding fibroblast foci (FF) in the setting of fibrotic ILD

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
FF are required for a histologic UIP diagnosis.	○	○	○	○	○
FF may be seen in the setting of interstitial lung diseases other than UIP (NSIP, HP, etc.).	○	○	○	○	○
The microscopic distribution of the FF is an important factor in my diagnostic consideration.	○	○	○	○	○
The pattern of fibrosis in which I encounter a FF changes my impression of the significance of the FF.	○	○	○	○	○
Distinguishing between FF and polyps of OP is an important factor in my diagnostic consideration.	○	○	○	○	○

Please evaluate each of the following statements regarding honeycomb lung

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
Honeycomb lung is likely histologic UIP	○	○	○	○	○
Based on the guidelines, I categorize cases with honeycomb lung only as Probable UIP	○	○	○	○	○
Honeycomb lung is equivalent to end-stage lung which could be from any etiology. I still look for additional features (inflation, granulomas, food, etc.) in the honeycomb lung to assess for etiology	○	○	○	○	○
Honeycomb is easy to recognize histologically	○	○	○	○	○
Honeycomb can be difficult to distinguish from traction bronchiectasis and peribronchiolar metaplasia (PBM)	○	○	○	○	○

Honeycomb cysts must be embedded in advanced fibrosis	○	○	○	○	○
Honeycomb can be seen in any chronic fibrosing lung disease	○	○	○	○	○
Honeycomb requires multiple clustered cysts in multiple foci within the biopsy	○	○	○	○	○
In the appropriate histologic setting, even a single honeycomb cyst is sufficient to support a histologic UIP diagnosis	○	○	○	○	○

Please evaluate each of the following statements regarding areas of normal lung in cases of fibrotic ILD.

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
I require areas of perfectly normal alveolar walls to diagnose histologic UIP	○	○	○	○	○
I allow mild interstitial fibrotic expansion in histologic UIP	○	○	○	○	○
I allow mild chronic inflammation in the interstitium of "normal lung" in histologic UIP	○	○	○	○	○

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Specific Histologic Features: Section 2

Please evaluate each of the following statements regarding granulomas and giant cells

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
Giant cells do not influence my diagnosis of histologic UIP	○	○	○	○	○
The number of granulomas and giant cells impact my assessment for histologic UIP	○	○	○	○	○
Even a single giant cell in dense fibrosis pushes me away from histologic UIP	○	○	○	○	○
I ignore giant cells in honeycombed airspaces	○	○	○	○	○
I allow a rare giant cell in histologic UIP, especially if in the airspace or deep in the fibrosis	○	○	○	○	○
I allow many giant cells in histologic UIP, as long as they are not coalescing into a granuloma	○	○	○	○	○
I allow many giant cells in histologic UIP, as long as they are not peribronchiolar in distribution	○	○	○	○	○
Granulomas should raise suspicion for a diagnosis other than histologic UIP or an additional diagnosis	○	○	○	○	○
The histologic features of granulomas (well vs. poorly focused, necrosis vs. not, etc.) impact their importance in potential histologic UIP cases	○	○	○	○	○

Please evaluate each of the following statements regarding airway centered fibrosis

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
I find it difficult to recognize airway centered fibrosis	○	○	○	○	○
I don't think airway centered fibrosis exists	○	○	○	○	○

I consider chronic inhalational diseases like chronic HP, aspiration, and PLCH when I see airway centered fibrosis.	○	○	○	○	○
Airway centered fibrosis can be seen in histologic UIP.	○	○	○	○	○
I only consider airway centered fibrosis significant if there are associated granulomas.	○	○	○	○	○

Please evaluate each of the following statements regarding peribronchiolar metaplasia (PBM) and mucostasis in fibrotic ILD

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
PBM and mucostasis are commonly seen in UIP	○	○	○	○	○
PBM and mucostasis do not influence my diagnosis of UIP	○	○	○	○	○
Extensive PBM and mucostasis causes me to consider other inhalational etiologies	○	○	○	○	○
I compare the degree of PBM and mucostasis to the degree of fibrosis, allowing more PBM and mucostasis in UIP as long as there is significant fibrosis and honeycomb	○	○	○	○	○

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Specific Histologic Features: Section 3

Please evaluate each of the following statements regarding lymphoid hyperplasia in the setting of a fibrotic ILD

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
The definition of "prominent lymphoid hyperplasia" is clear	○	○	○	○	○
The amount of inflammation required for "prominent lymphoid hyperplasia" is clear	○	○	○	○	○
When I encounter what I consider prominent lymphoid hyperplasia, I suggest a non-IPF diagnosis	○	○	○	○	○
I ignore lymphoid hyperplasia in areas of advanced scarring	○	○	○	○	○
The definition of "cellular inflammatory infiltrate away from honeycombing" is clear	○	○	○	○	○
The amount of inflammation required for "cellular inflammatory infiltrate away from honeycombing " is clear	○	○	○	○	○
When I encounter what I consider cellular inflammatory infiltrate away from honeycombing, I suggest a non-IPF diagnosis	○	○	○	○	○
I ignore cellular inflammatory infiltrates in areas of advanced scarring	○	○	○	○	○
The definition of "lymphoid follicles with secondary germinal centers" is clear	○	○	○	○	○
The amount of inflammation required for "lymphoid follicles with secondary germinal centers " is clear	○	○	○	○	○
When I encounter what I consider lymphoid follicles with secondary germinal centers, I suggest a non-IPF diagnosis	○	○	○	○	○

I ignore lymphoid follicles with secondary germinal centers in areas of advanced scarring	○	○	○	○	○

Please evaluate each of the following statements regarding chronic fibrous pleuritis in the setting of a fibrosing ILD

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
The amount of chronic pleuritis in a biopsy is important in considering a UIP diagnosis.	○	○	○	○	○
The amount of chronic pleuritis needed to consider diagnoses other than UIP is clear	○	○	○	○	○
Chronic pleuritis on a biopsy leads to me consider CTD-associated ILD	○	○	○	○	○

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Specific Histologic Features: Section 4

How do you diagnose cases with histologic UIP and superimposed OP, fibrin, and/or hyaline membranes (select all that apply).

- ☐ I diagnose UIP in acute exacerbation as UIP
- ☐ Based on the guidelines, I categorize cases that I think are UIP in acute exacerbation as Indeterminate for UIP.
- ☐ Diagnose as "acute on chronic fibrosing ILD" and explain the differential diagnosis.
- ☐ Diagnose as acute lung injury (OP, DAD, AFOP) with background UIP
- ☐ I am unclear how to diagnose these cases.

Please evaluate the following regarding fibrosing/chronic organizing pneumonia

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
Fibrosing/chronic organizing pneumonia alone can result in advanced fibrosis with architectural distortion similar to UIP	○	○	○	○	○
Fibrosing/chronic organizing pneumonia pattern excludes UIP/IPF	○	○	○	○	○
Fibrosing/chronic organizing pneumonia is not an ILD	○	○	○	○	○

Please evaluate each of the following statements regarding cases with overlapping features of smoking-related ILD and UIP

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
I find these entities difficult to distinguish	○	○	○	○	○
The location of the biopsies (upper vs lower lobe) and the pattern of injury can be helpful	○	○	○	○	○
Advanced fibrosis and fibroblast foci should not be seen in smoking related ILD	○	○	○	○	○
Honeycomb lung should not be seen in smoking related ILD	○	○	○	○	○

Please evaluate each of the following statements regarding cases with overlap between chronic aspiration and UIP

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
I find these entities difficult to distinguish	○	○	○	○	○

The location of the biopsies (upper vs lower lobe) and the pattern of injury can be helpful	○	○	○	○	○
Aspiration alone does not result in advanced fibrosis, fibroblast foci, and honeycomb lung.	○	○	○	○	○

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Comments

Is there anything else regarding UIP/IPF and fibrotic ILD you would like to share or discuss at the PPS meeting?

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