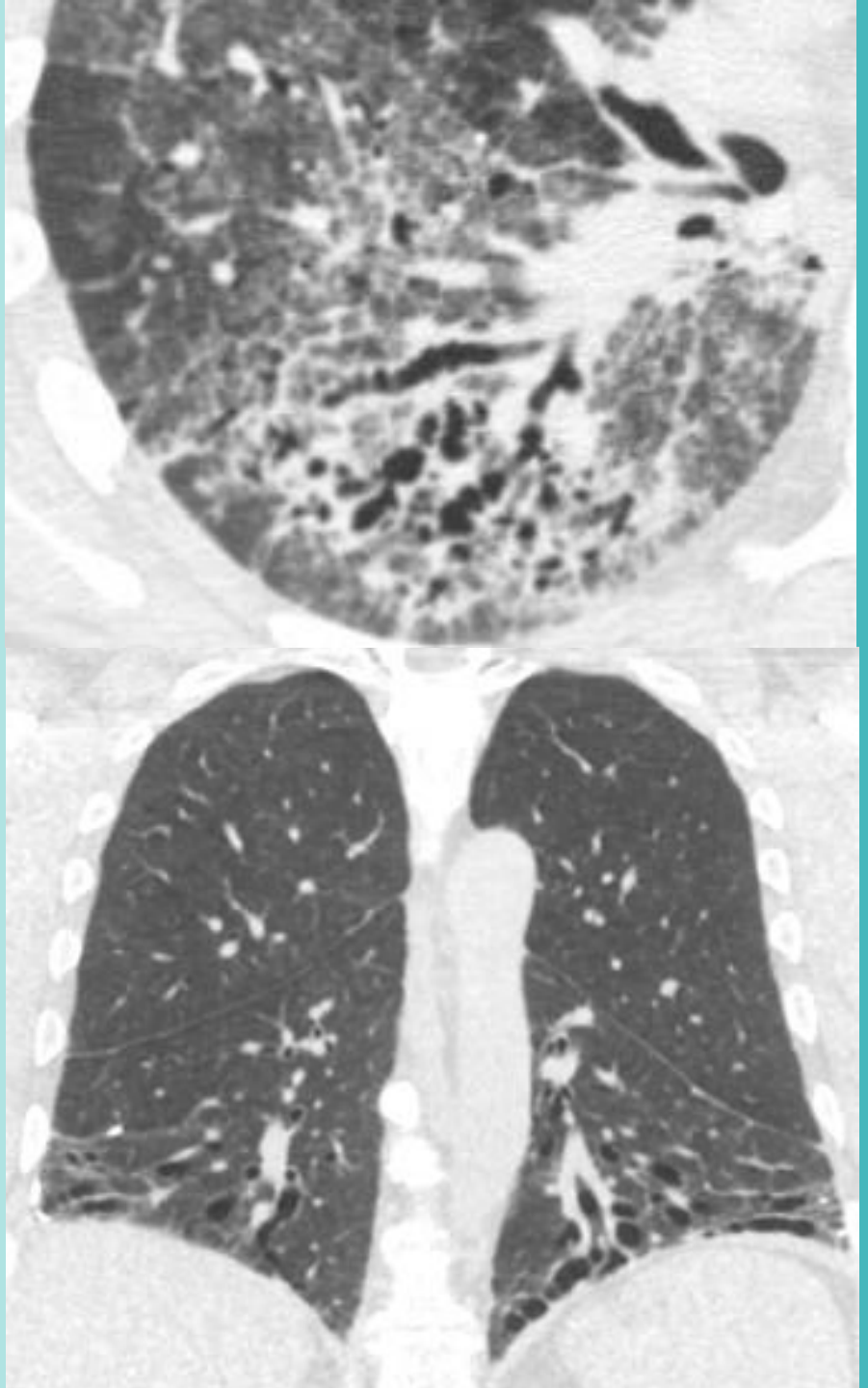
Mass General Brigham

Update of the International Multidisciplinary Classification of the Interstitial Pneumonias

Amita Sharma
Division Chief Thoracic Radiology
Mass General Brigham
Harvard Medical School

April 15, 2026



Disclosures

Clinical Trials research funding to institution from Bristol Myers Squibb, Eli Lilly, Novartis, AstraZeneca, Boehringer Ingelheim, and aTyr Pharma Inc

Stocks as Ownership interest for Small Pharma

Advisory board member for Horizon

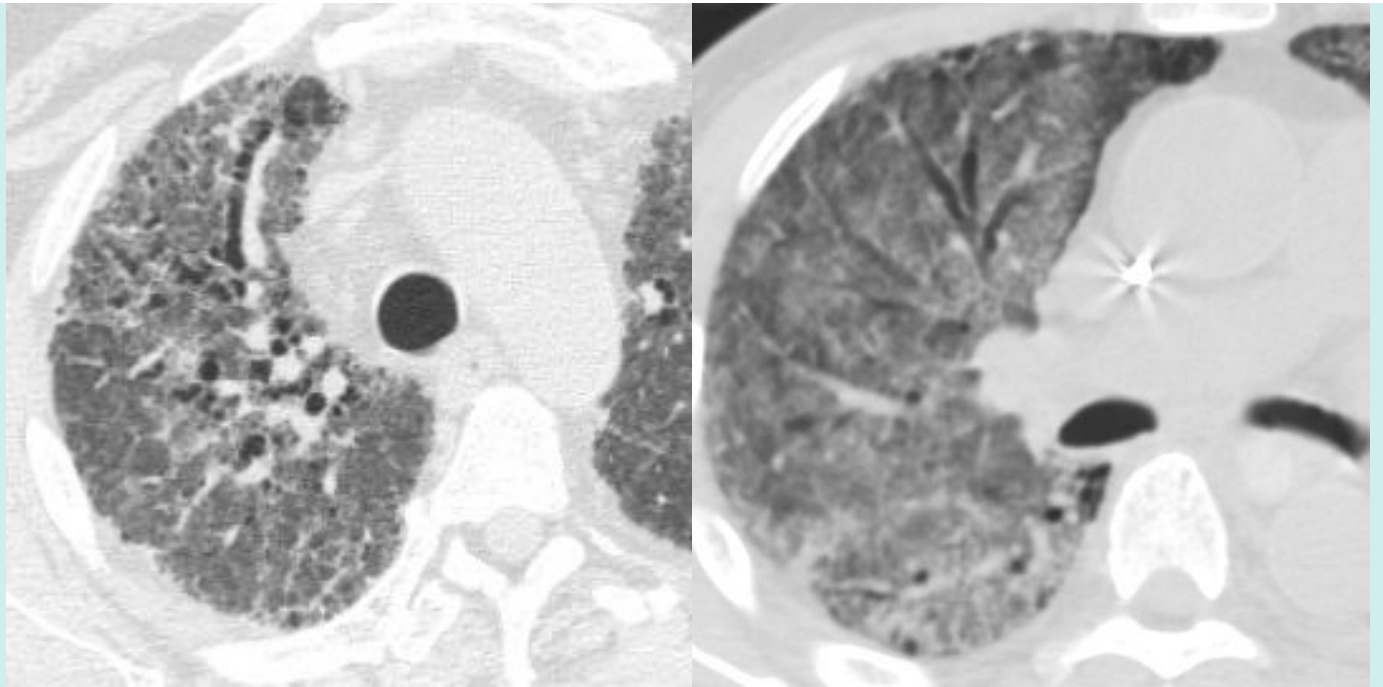
HISTORY
MAJOR CHANGES
SECONDARY /IDIOPATHIC
INTERSTITIAL AND
ALVEOLAR
FIBROTIC/NON-FIBROTIC

OBJECTIVES

EUROPEAN RESPIRATORY JOURNAL
ERS OFFICIAL DOCUMENT
C.J. RYERSON ET AL.

Update of the international multidisciplinary classification of the interstitial pneumonias: an ERS/ATS statement

Christopher J. Ryerson[1,42], Ayodeji Adegunsoye[2,42], Sara Piciucchi[3,4,42], Lida P. Hariri[5], Yet H. Khor[6,7,8,9], Marlies S. Wijsenbeek[10], Athol U. Wells[11], Amita Sharma[12], Wendy A. Cooper[13], Katerina Antoniou[14], Raphael Borie[15], Aurelie Fabre[16,17], Yoshikazu Inoue[18,19], Kerri Johannson[20], Takeshi Johkoh[21], Leticia Kawano-Dourado[22,23,24], Ella Kazerooni[25], Toby M. Maher[26,27], Philip L. Molyneaux[27,28], Raymond Protti[29], Claudia Ravaglia[3,30], Elisabetta A. Renzoni[27,31], Ryoko Saito-Koyama[32], Nicola Sverzellati[33,34], Simon L.F. Walsh[35,36,37], Paul Wolters[38], Soo-Ryum Yang[39], William Travis[40,42] and Andrew G. Nicholson[27,41,42]



American Thoracic Society

American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias

This Joint Statement of the American Thoracic Society (ATS), and the European Respiratory Society (ERS) was adopted by the ATS Board of Directors, June 2001 and by The ERS Executive Committee, June 2001

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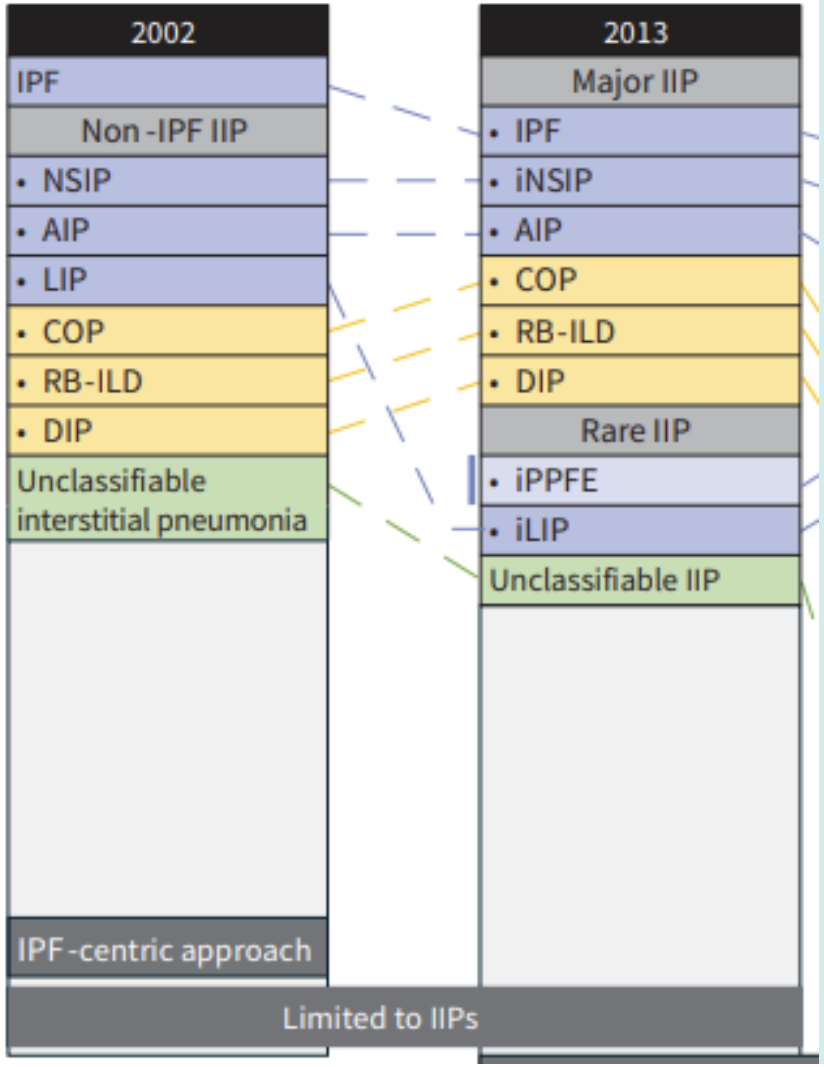
Eur Respir J 2025; 66: 2500158



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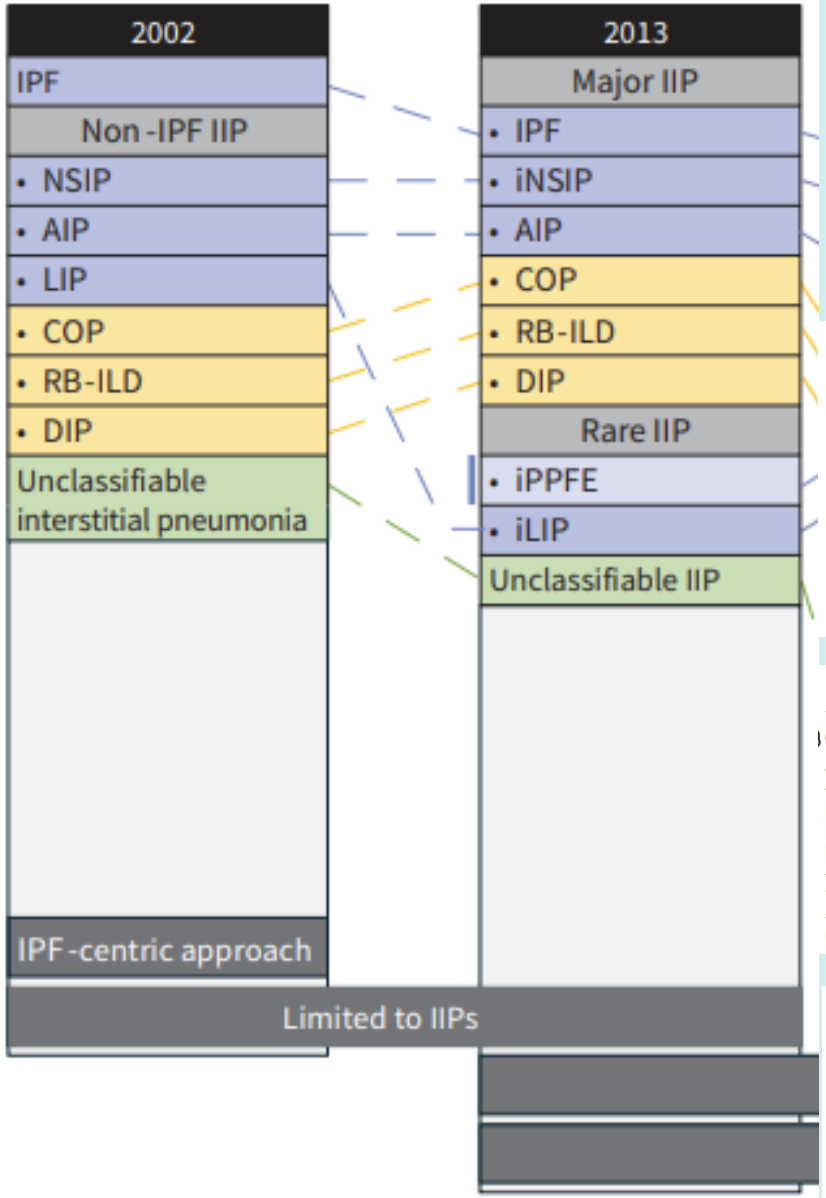

Diagnostic criteria for idiopathic pulmonary fibrosis: a Fleischner Society White Paper

David A Lynch, Nicola Sverzellati, William D Travis, Kevin K Brown, Thomas V Colby, Jeffrey R Galvin, Jonathan G Goldin, David M Hansell, Yoshikazu Inoue, Takeshi Johkoh, Andrew G Nicholson, Shandra L Knight, Suhail Raoof, Luca Richeldi, Christopher J Ryerson, Jay H Ryu, Athol U Wells

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Department of Radiology (Prof D A Lynch MB), Department of Medicine (Prof K K Brown MD), and Library and Knowledge Services (S L Knight MS), National Jewish Health, Denver, CO, USA; Department of Radiology, Ospedale di Circolo, Fondazione Macchi, Università degli Studi dell'Insubria, Varese, Italy (N Sverzellati MD); Department of Radiology, University of Pittsburgh Medical Center, Pittsburgh, PA, USA (W D Travis MD); Department of Radiology, Mayo Clinic, Rochester, MN, USA (J H Ryu MD); Department of Radiology, David Geffen School of Medicine at UCLA, Los Angeles, CA, USA (J G Goldin MD); Department of Histopathology, Royal Brompton Hospital, London, UK (D M Hansell MD); Department of Histopathology, Imperial College London, London, UK (A G Nicholson MD); Department of Pathology, Mayo Clinic, Scottsdale, AZ, USA (T V Colby MD); Department of Radiology, Virginia Commonwealth University, Richmond, VA, USA (J R Galvin MD); Department of Pathology, Yamagata University, Yamagata, Japan (Y Inoue MD); Department of Respiratory Medicine, Kanazawa Medical University, Kanazawa, Japan (T Johkoh MD); Department of Pathology, Fujita Health University, Toyoake, Japan (T Johkoh); Department of Pathology, Duke University Medical Center, Durham, NC, USA (S Raoof MD); Department of Pulmonary Medicine, Cleveland Clinic, Cleveland, OH, USA (L Richeldi MD); Department of Pulmonary Medicine, Thoracic Institute, Cleveland Clinic, Cleveland, OH, USA (C J Ryerson MD); Department of Medicine, Queen's University, Kingston, ON, Canada (C J Ryerson); and Department of Medicine, University of Chicago, Chicago, IL, USA (A U Wells MD)

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Diagnosis of Idiopathic Pulmonary Fibrosis
An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline

Ganesh Raghu, Martine Remy-Jardin, Jeffrey L. Myers, Luca Richeldi, Christopher J. Ryerson, David J. Lederer, Juergen Behr, Vincent Cottin, Sonye K. Danoff, Ferran Morell, Kevin R. Flaherty, Athol Wells, Fernando J. Martinez, Arata Azuma, Thomas J. Bice, Demosthenes Bouros, Kevin K. Brown, Harold R. Collard, Abhijit Duggal, Liam Galvin, Yoshikazu Inoue, R. Gisli Jenkins, Takeshi Johkoh, Ella A. Kazerooni, Masanori Kitaichi, Shandra L. Knight, George Mansour, Andrew G. Nicholson, Sudhakar N. J. Pipavath, Ivette Buendía-Roldán, Moisés Selman, William D. Travis, Simon L. F. Walsh, and Kevin C. Wilson; on behalf of the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Latin American Thoracic Society

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This official ATS/ERS/JRS/ALAT Clinical Practice Guideline was endorsed by the Pulmonary Pathology Society October 2018

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

A Phase 3 Trial of Pirfenidone in Patients with Idiopathic Pulmonary Fibrosis

Talmadge E. King, Jr., M.D., Williamson Z. Bradford, M.D., Ph.D., Socorro Castro-Bernardini, M.D., Elizabeth A. Fagan, M.D., Ian Glaspole, M.B., B.S., Ph.D., Marilyn K. Glassberg, M.D., Eduard Gorina, M.D., Peter M. Hopkins, M.D., David Kardatzke, Ph.D., Lisa Lancaster, M.D., David J. Lederer, M.D., Steven D. Nathan, M.D., Carlos A. Pereira, M.D., Steven A. Sahn, M.D., Robert Sussman, M.D., Jeffrey J. Swigris, D.O., and Paul W. Noble, M.D., for the ASCEND Study Group*

The NEW ENGLAND JOURNAL of MEDICINE
ESTABLISHED IN 1812 MAY 29, 2014 VOL. 370 NO. 22

Efficacy and Safety of Nintedanib in Idiopathic Pulmonary Fibrosis

Luca Richeldi, M.D., Ph.D., Roland M. du Bois, M.D., Ganesh Raghu, M.D., Arata Azuma, M.D., Ph.D., Kevin K. Brown, M.D., Ulrich Costabel, M.D., Vincent Cottin, M.D., Ph.D., Kevin R. Flaherty, M.D., David M. Hansell, M.D., Yoshikazu Inoue, M.D., Ph.D., Dong Soon Kim, M.D., Martin Kolb, M.D., Ph.D., Andrew G. Nicholson, D.M., Paul W. Noble, M.D., Moisés Selman, M.D., Hiroyuki Taniguchi, M.D., Ph.D. Michèle Brun, M.Sc., Florence Le Maulf, M.Sc., Mannaïg Girard, M.Sc., Susanne Stowasser, M.D., Rozsa Schlenker-Herceg, M.D., Bernd Disse, M.D., Ph.D., and Harold R. Collard, M.D., for the INPULSIS Trial Investigators*

[Original Research Diffuse Lung Disease]
CHEST

Diagnostic Ability of a Dynamic Multidisciplinary Discussion in Interstitial Lung Diseases
A Retrospective Observational Study of 938 Cases

Laurens J. De Sadeleer, MD; Caressa Meert, MD; Jonas Yserbyt, MD, PhD; Hans Slabbynck, MD; Johny A. Verschakelen, MD, PhD; Eric K. Verbeken, MD, PhD; Birgit Weynand, MD, PhD; Ellen De Langhe, MD, PhD; Jan L. Lenaerts, MD; Benoit Nemery, MD, PhD; Dirk Van Raemdonck, MD, PhD; Geert M. Verleden, MD, PhD; Athol U. Wells, MD, PhD; and Wim A. Wuyts, MD, PhD

Diagnosis of Hypersensitivity Pneumonitis in Adults
An Official ATS/JRS/ALAT Clinical Practice Guideline

Ganesh Raghu, Martine Remy-Jardin, Christopher J. Ryerson, Jeffrey L. Myers, Michael Kreuter, Martina Vasakova, Elena Bargagli, Jonathan H. Chung, Bridget F. Collins, Elisabeth Bendstrup, Hassan A. Chami, Abigail T. Chua, Tamera J. Corte, Jean-Charles Dalphin†, Sonye K. Danoff, Javier Diaz-Mendoza, Abhijit Duggal, Ryoko Egashira, Thomas Ewing, Mridu Gulati, Yoshikazu Inoue, Alex R. Jenkins, Kerri A. Johannson, Takeshi Johkoh, Maximiliano Tamae-Kakazu, Masanori Kitaichi, Shandra L. Knight, Dirk Koschel, David J. Lederer, Yolanda Mageto, Lisa A. Maier, Carlos Matiz, Ferran Morell, Andrew G. Nicholson, Setu Patolia, Carlos A. Pereira, Elisabetta A. Renzoni, Margaret L. Salisbury, Moises Selman, Simon L. F. Walsh, Wim A. Wuyts, and Kevin C. Wilson; on behalf of the American Thoracic Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax

This guideline is dedicated to the memory of Prof. Jean-Charles Dalphin† (June 2, 1956–October 17, 2019)

This official clinical practice guideline was approved by the American Thoracic Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax May 2020

[Diffuse Lung Disease Guidelines and Consensus Statements]
CHEST

Diagnosis and Evaluation of Hypersensitivity Pneumonitis
CHEST Guideline and Expert Panel Report

Evans R. Fernández Pérez, MD, FCCP; William D. Travis, MD, FCCP; David A. Lynch, MB, BCh; Kevin K. Brown, MD, FCCP; Kerri A. Johannson, MD, MPH; Moisés Selman, MD; Jay H. Ryu, MD, FCCP; Athol U. Wells, MD; Yuh-Chin Tony Huang, MD, MHS, FCCP; Carlos A. C. Pereira, MD, FCCP; Mary-Beth Scholand, MD, FCCP; Ana Villar, MD, PhD; Naohiko Inase, MD, PhD; Richard B. Evans, MD, MPH, FCCP; Stephen A. Mette, MD, FCCP; and Lindsy Frazer-Green, PhD

AMERICAN THORACIC SOCIETY DOCUMENTS

Idiopathic Pulmonary Fibrosis (an Update) and Progressive Pulmonary Fibrosis in Adults
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Ganesh Raghu, Martine Remy-Jardin, Luca Richeldi, Carey C. Thomson, Yoshikazu Inoue, Takeshi Johkoh, Michael Kreuter, David A. Lynch, Toby M. Maher, Fernando J. Martinez, Maria Molina-Molina, Jeffrey L. Myers, Andrew G. Nicholson, Christopher J. Ryerson, Mary E. Strek, Lauren K. Troy, Marlies Wijsenbeek, Manoj J. Mammen, Tanzib Hossain, Brittany D. Bissell, Derrick D. Herman, Stephanie M. Hon, Fayez Kheir, Yet H. Khor, Madalina Macrea, Katerina M. Antoniou, Demosthenes Bouros, Ivette Buendia-Roldan, Fabian Caro, Bruno Crestani, Lawrence Ho, Julie Morisset, Amy L. Olson, Anna Podolanczuk, Venerino Poletti, Moisés Selman, Thomas Ewing, Stephen Jones, Shandra L. Knight, Marya Ghazipura, and Kevin C. Wilson; on behalf of the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax

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Arthritis & Rheumatology
Vol. 76, No. 8, August 2024, pp 1201–1213
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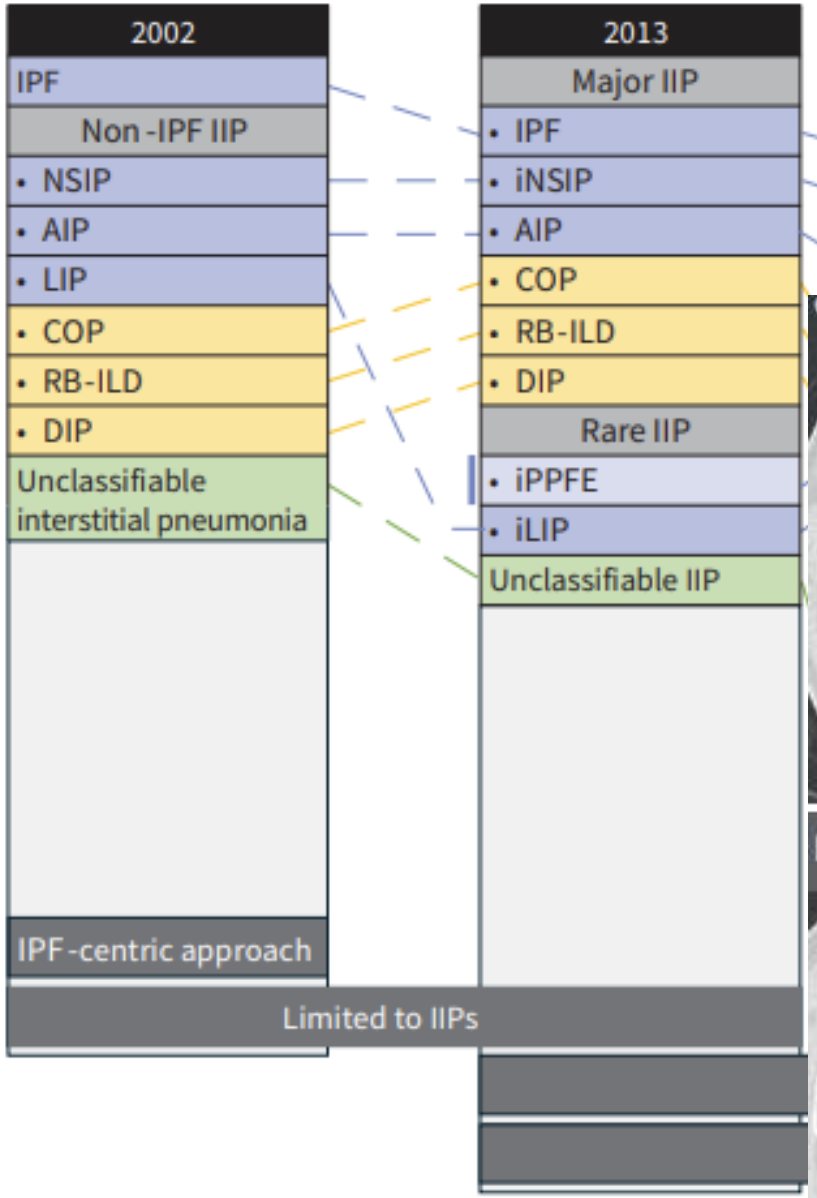
American College of Rheumatology
Empowering Rheumatology Professionals

2023 American College of Rheumatology (ACR)/American College of Chest Physicians (CHEST) Guideline for the Screening and Monitoring of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Diseases

Sindhu R. Johnson,[1,11*] Elana J. Bernstein,[2*] Marcy B. Bolster,[3] Jonathan H. Chung,[4] Sonye K. Danoff,[5] Michael D. George,[6] Dinesh Khanna,[7] Gordon Guyatt,[8] Reza D. Mirza,[8] Rohit Aggarwal,[9] Aberdeen Allen Jr,[10] Shervin Assassi,[11] Lenore Buckley,[12] Hassan A. Chami,[5] Douglas S. Corwin,[13] Paul F. Dellaripa,[14] Robyn T. Domsic,[9] Tracy J. Doyle,[14] Catherine Marie Falardeau,[15] Tracy M. Frech,[16] Fiona K. Gibbons,[3] Monique Hinchcliff,[12] Cheilonda Johnson,[6] Jeffrey P. Kanne,[17] John S. Kim,[18] Sian Yik Lim,[19] Scott Matson,[20] Zsuzsanna H. McMahan,[5] Samantha J. Merck,[21] Kiana Nesbitt,[22] Mary Beth Scholand,[23] Lee Shapiro,[24] Christine D. Sharkey,[17] Ross Summer,[25] John Varga,[7] Anil Warrier,[26] Sandeep K. Agarwal,[27] Danielle Antin-Ozerkis,[12] Bradford Bemiss,[28] Vaidehi Chowdhary,[12] Jane E. Dematte D'Amico,[28] Robert Hallowell,[3] Alicia M. Hinze,[29] Patil A. Injean,[30] Nikhil Jiwrajka,[6] Elena K. Joerns,[31] Joyce S. Lee,[32] Ashima Makol,[29] Gregory C. McDermott,[14] Jake G. Natalini,[33] Justin M. Oldham,[7] Didem Saygin,[9] Kimberly Showalter Lakin,[34] Namrata Singh,[35] Joshua J. Solomon,[36] Jeffrey A. Sparks,[14] Marat Turgunbaev,[37] Samera Vaseer,[38] Amy Turner,[37] Stacey Uhl,[39] and Ilya Ivlev[39]



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See Comment page 82

Department of Radiology (Prof D A Lynch MB), Department of Medicine (Prof K K Brown MD), and Library and Knowledge

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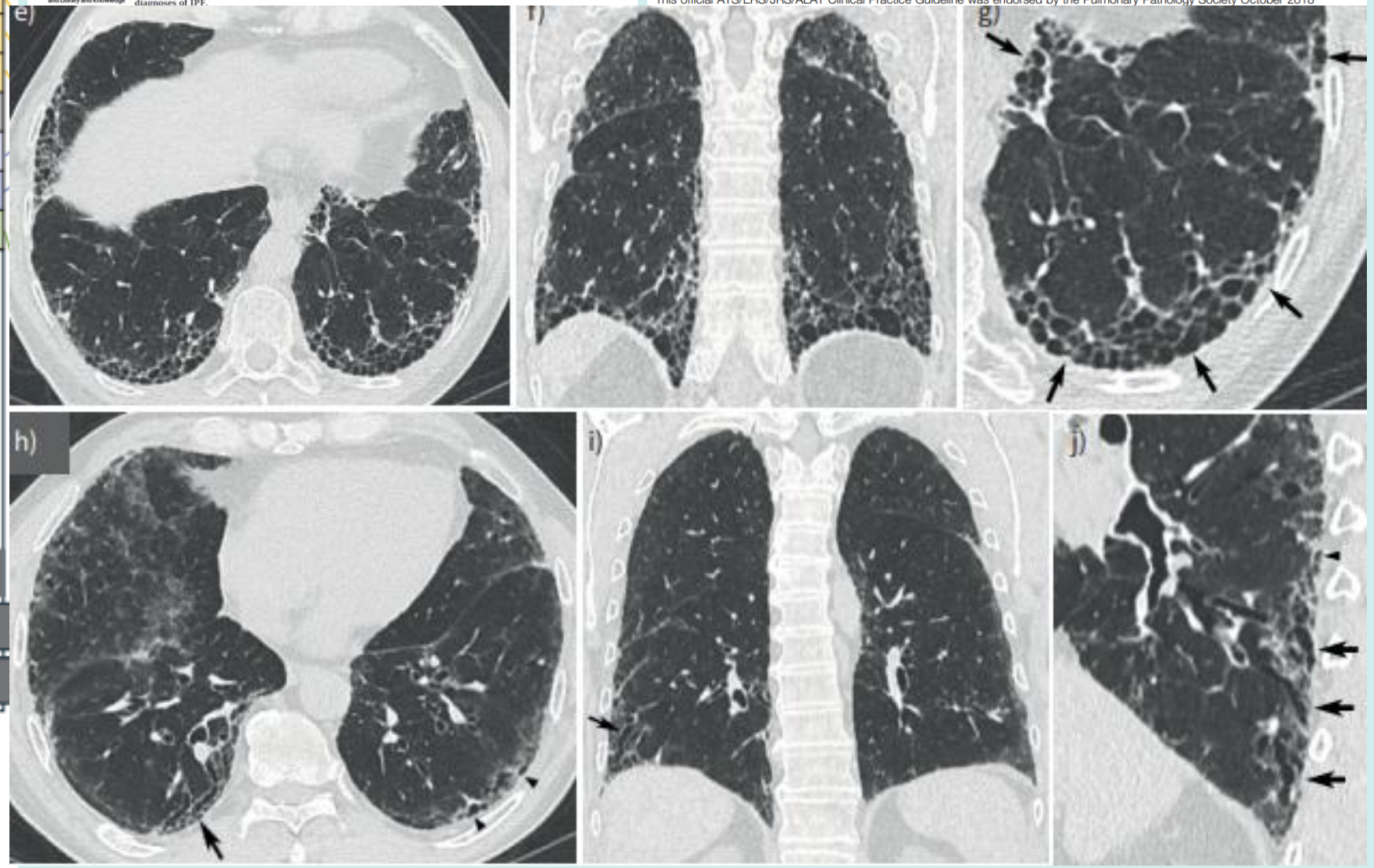
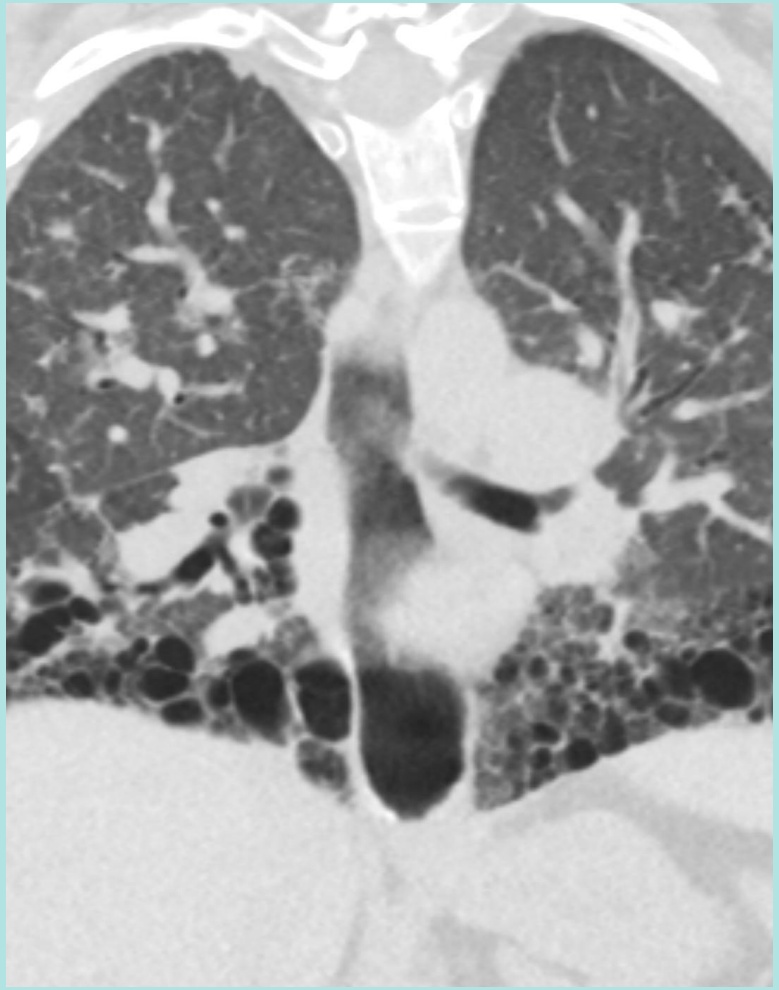
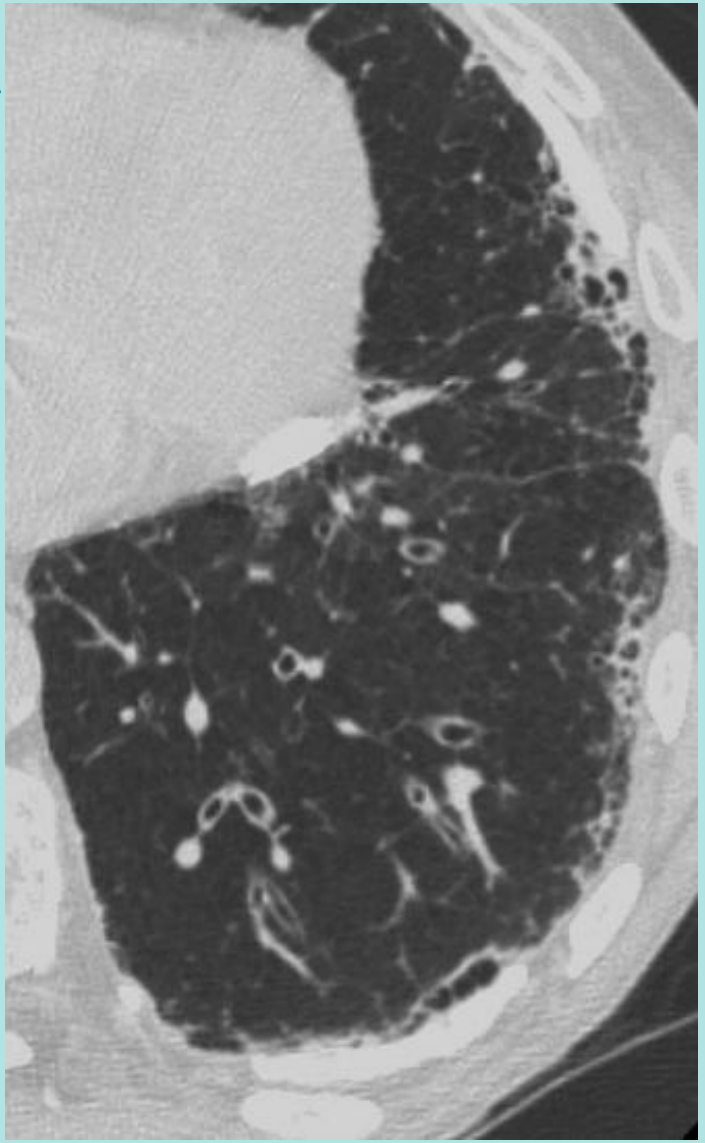


UIP Pattern Etiology Idiopathic>>Secondary



Scleroderma



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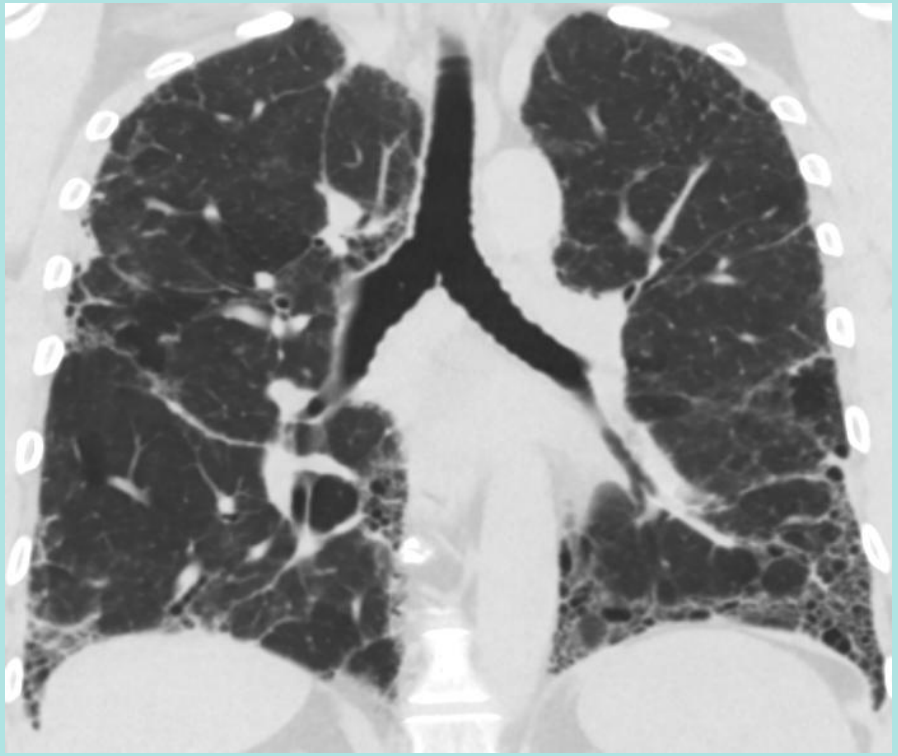
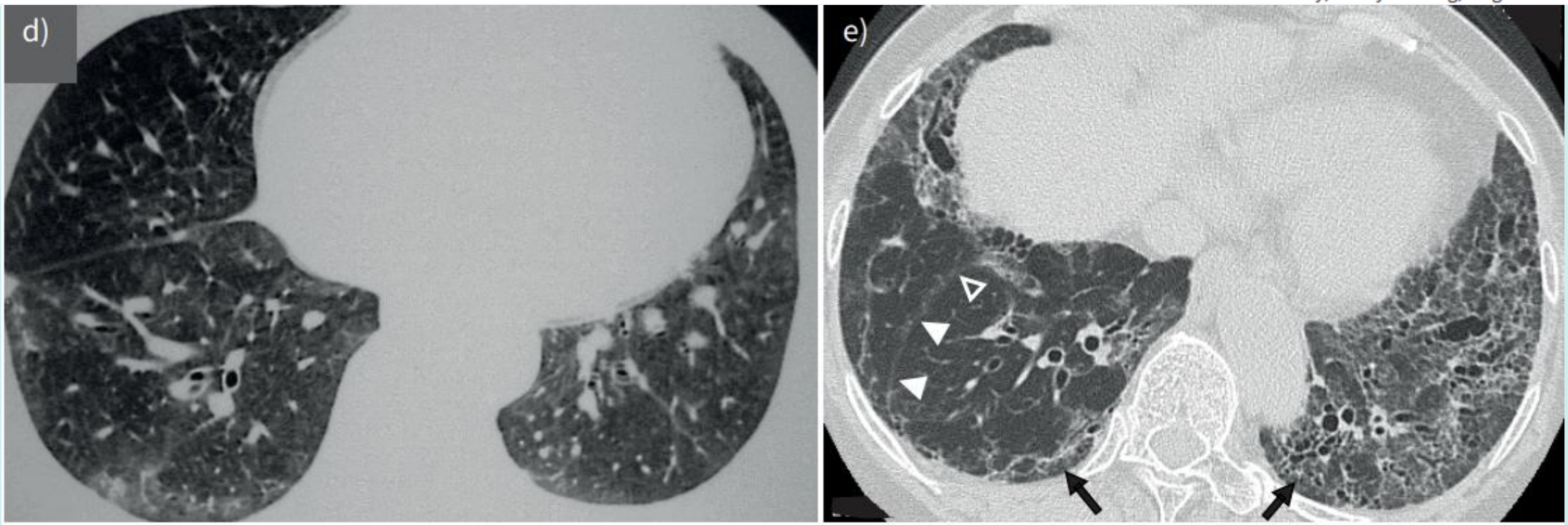


fHP

TABLE 2 Typical pathological, radiological and clinical features of major interstitial pneumonia patterns

	Clinical features	Radiological features	Pathological features
Interstitial patterns			
NSIP	Presentation: chronic onset, but with variable risk of progression. Potential for rapid worsening, especially with concurrent organising pneumonia Risk factors: more common in younger females, often with a background of CTD	Lower-lobe predominance, commonly with subpleural sparing. Ground-glass opacities, fine reticulation, and traction bronchiectasis, without honeycombing	Cellular NSIP: cellular interstitial infiltrate of lymphocytes causing thickening of alveolar walls with no fibrosis or granulomas Fibrotic NSIP: diffuse thickening of alveolar walls by fibrosis with traction bronchiolectasis; no bronchiolocentricity, honeycombing, or granulomas

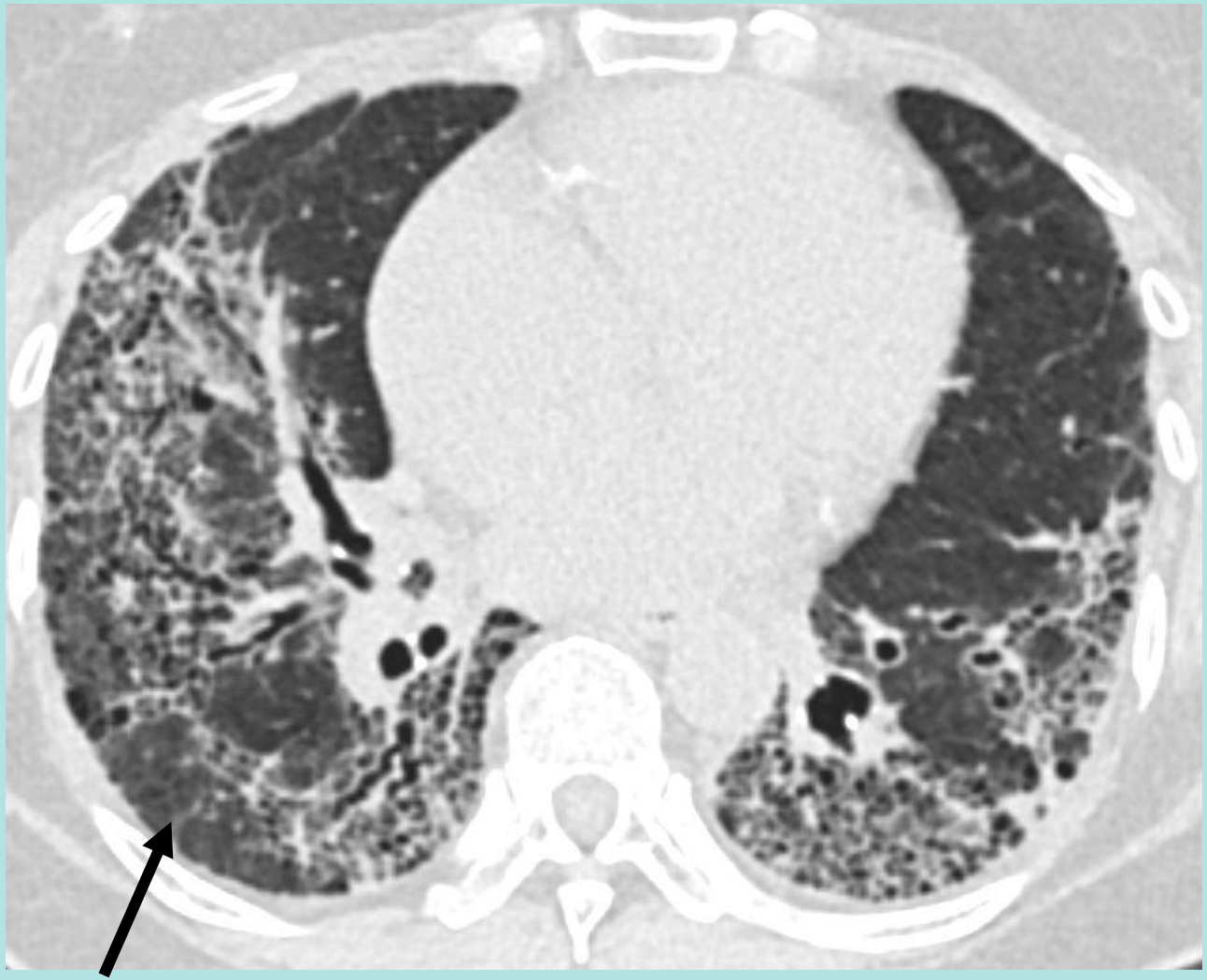


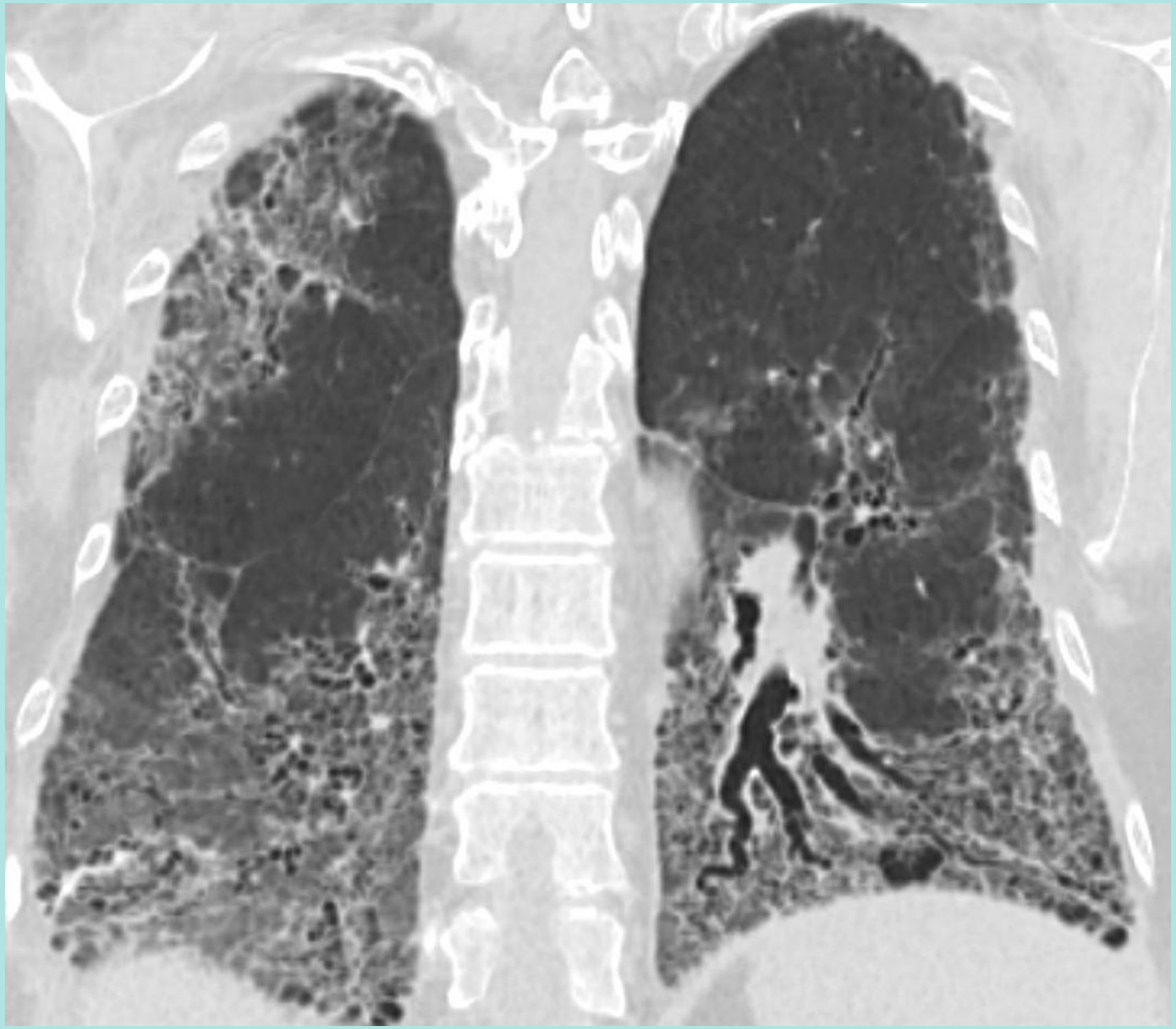
d) Axial computed tomography (CT) of non-fibrotic NSIP shows lower lung predominant ground-glass and minimal reticulation without traction bronchiectasis or other signs of fibrosis, and with subpleural sparing. e) Axial CT of fibrotic NSIP shows lower-lung-predominant reticular abnormality with traction bronchiectasis. Subpleural sparing is present (arrows). There is bilateral lower lobe volume loss with posterior displacement of the right major fissure (arrowheads).

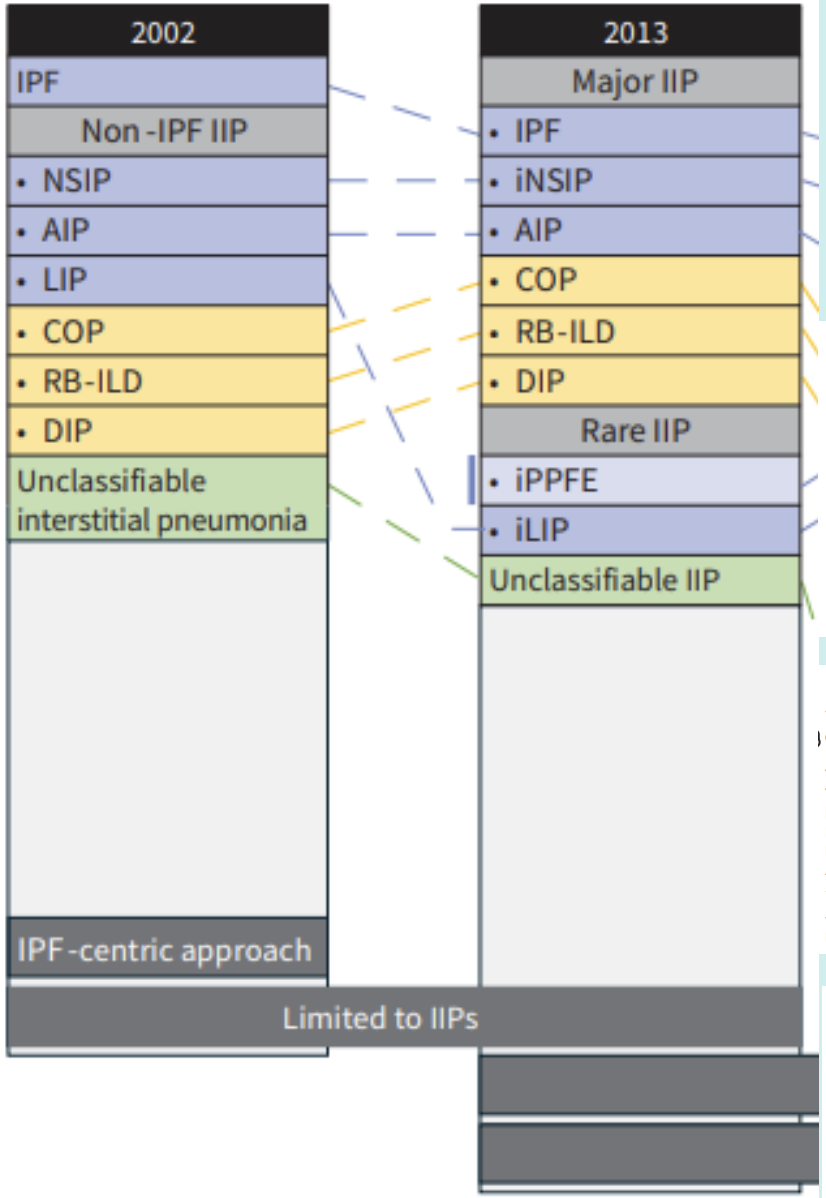
Eur Respir J 2025; 66: 2500158

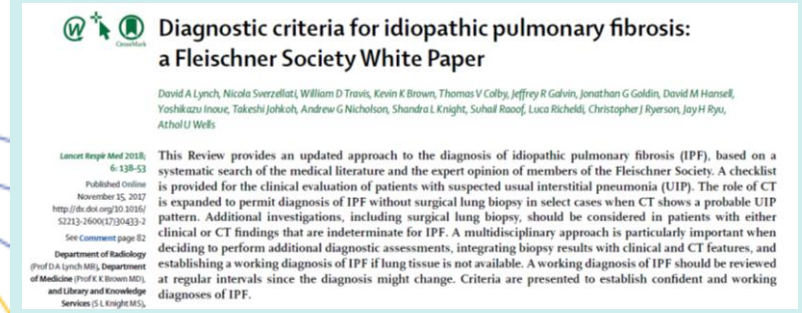
60-year-old female diagnosed with CTD ILD-Sjogrens with myositis. Fibrotic NSIP with subpleural sparing and absence of honeycombing

Am J Resp Crit Care Med 2008: 1338-47









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Lancet Respir Med 2018; 6: 138–53

This Review provides an updated approach to the diagnosis of idiopathic pulmonary fibrosis (IPF), based on a systematic search of the medical literature and the expert opinion of members of the Fleischner Society. A checklist is provided for the clinical evaluation of patients with suspected usual interstitial pneumonia (UIP). The role of CT is expanded to permit diagnosis of IPF without surgical lung biopsy in select cases when CT shows a probable UIP pattern. Additional investigations, including surgical lung biopsy, should be considered in patients with either clinical or CT findings that are indeterminate for IPF. A multidisciplinary approach is particularly important when deciding to perform additional diagnostic assessments, integrating biopsy results with clinical and CT features, and establishing a working diagnosis of IPF if lung tissue is not available. A working diagnosis of IPF should be reviewed at regular intervals since the diagnosis might change. Criteria are presented to establish confident and working diagnoses of IPF.

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

A Phase 3 Trial of Pirfenidone in Patients with Idiopathic Pulmonary Fibrosis

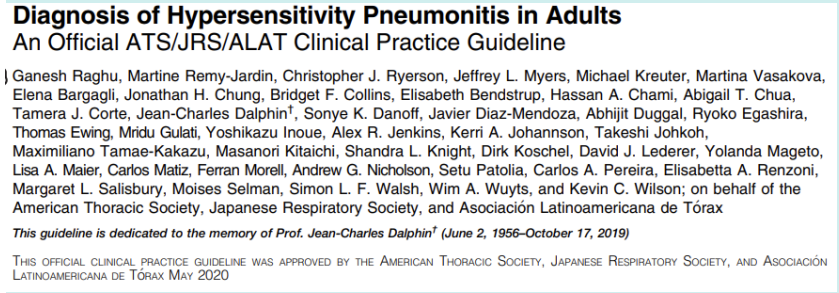
Talmadge E. King, Jr., M.D., Williamson Z. Bradford, M.D., Ph.D., Socorro Castro-Bernardini, M.D., Elizabeth A. Fagan, M.D., Ian Glaspole, M.B., B.S., Ph.D., Marilyn K. Glassberg, M.D., Eduard Gorina, M.D., Peter M. Hopkins, M.D., David Kardatzke, Ph.D., Lisa Lancaster, M.D., David J. Lederer, M.D., Steven D. Nathan, M.D., Carlos A. Pereira, M.D., Steven A. Sahn, M.D., Robert Sussman, M.D., Jeffrey J. Swigris, D.O., and Paul W. Noble, M.D., for the ASCEND Study Group*

The NEW ENGLAND JOURNAL of MEDICINE

ESTABLISHED IN 1812 MAY 29, 2014 VOL. 370 NO.

Efficacy and Safety of Nintedanib in Idiopathic Pulmonary Fibrosis

Luca Richeldi, M.D., Ph.D., Roland M. du Bois, M.D., Ganesh Raghu, M.D., Arata Azuma, M., Kevin K. Brown, M.D., Ulrich Costabel, M.D., Vincent Cottin, M.D., Ph.D., Kevin R. Flaherty, David M. Hansell, M.D., Yoshikazu Inoue, M.D., Ph.D., Dong Soon Kim, M.D., Martin Kolb, M., Andrew G. Nicholson, D.M., Paul W. Noble, M.D., Moisés Selman, M.D., Hiroyuki Taniguchi, M., Michèle Brun, M.Sc., Florence Le Maulf, M.Sc., Mannaïg Girard, M.Sc., Susanne Stowasser, Rozsa Schlenker-Herceg, M.D., Bernd Disse, M.D., Ph.D., and Harold R. Collard, M.D., for the INPULSIS Trial Investigators*



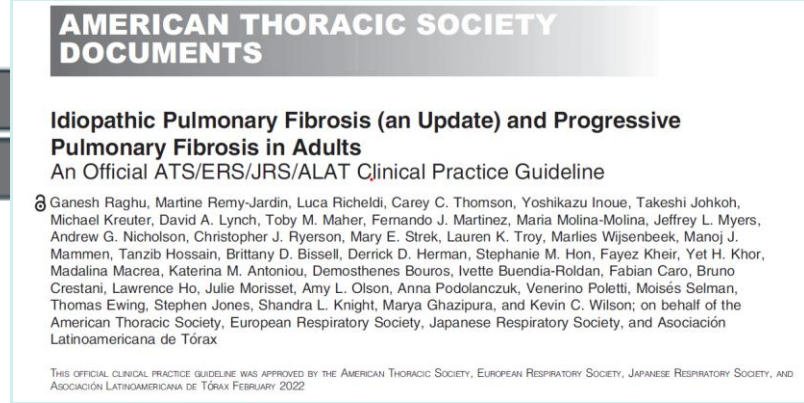
Diagnosis of Hypersensitivity Pneumonitis in Adults

An Official ATS/JRS/ALAT Clinical Practice Guideline

Ganesh Raghu, Martine Remy-Jardin, Christopher J. Ryerson, Jeffrey L. Myers, Michael Kreuter, Martina Vasakova, Elena Bargagli, Jonathan H. Chung, Bridget F. Collins, Elisabeth Bendstrup, Hassan A. Chami, Abigail T. Chua, Tamera J. Corte, Jean-Charles Dalphin†, Sonye K. Danoff, Javier Diaz-Mendoza, Abhijit Duggal, Ryoko Egashira, Thomas Ewing, Mridu Gulati, Yoshikazu Inoue, Alex R. Jenkins, Kerri A. Johannson, Takeshi Johkoh, Maximiliano Tamae-Kakazu, Masanori Kitaichi, Shandra L. Knight, Dirk Koschel, David J. Lederer, Yolanda Mageto, Lisa A. Maier, Carlos Matiz, Ferran Morell, Andrew G. Nicholson, Setu Patolia, Carlos A. Pereira, Elisabetta A. Renzoni, Margaret L. Salisbury, Moises Selman, Simon L. F. Walsh, Wim A. Wuyts, and Kevin C. Wilson; on behalf of the American Thoracic Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax

This guideline is dedicated to the memory of Prof. Jean-Charles Dalphin† (June 2, 1956–October 17, 2019)

This official clinical practice guideline was approved by the American Thoracic Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax May 2020



AMERICAN THORACIC SOCIETY DOCUMENTS

Idiopathic Pulmonary Fibrosis (an Update) and Progressive Pulmonary Fibrosis in Adults

An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline

Ganesh Raghu, Martine Remy-Jardin, Luca Richeldi, Carey C. Thomson, Yoshikazu Inoue, Takeshi Johkoh, Michael Kreuter, David A. Lynch, Toby M. Maher, Fernando J. Martinez, Maria Molina-Molina, Jeffrey L. Myers, Andrew G. Nicholson, Christopher J. Ryerson, Mary E. Strek, Lauren K. Troy, Marlies Wijsenbeek, Manoj J. Mammen, Tanzib Hossain, Brittany D. Bissell, Derrick D. Herman, Stephanie M. Hon, Fayez Kheir, Yet H. Khor, Madalina Macrea, Katerina M. Antoniou, Demosthenes Bouros, Ivette Buendia-Roldan, Fabian Caro, Bruno Crestani, Lawrence Ho, Julie Morisset, Amy L. Olson, Anna Podolanczuk, Venerino Poletti, Moisés Selman, Thomas Ewing, Stephen Jones, Shandra L. Knight, Marya Ghazipura, and Kevin C. Wilson; on behalf of the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax

This official clinical practice guideline was approved by the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax February 2022

Diagnosis of Idiop

An Official ATS/ERS

Ganesh Raghu, Martine Rem
Juergen Behr, Vincent Cottin
Arata Azuma, Thomas J. Bice
Yoshikazu Inoue, R. Gisli Jen
George Mansour, Andrew G.
William D. Travis, Simon L. F
Respiratory Society, Japanes

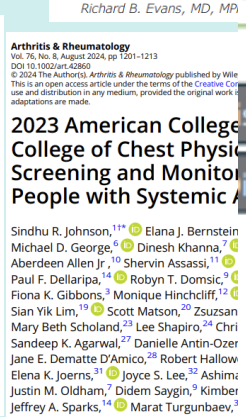
This official clinical practice guidel
Society (JRS), and Latin American

This official ATS/ERS/JRS/ALAT

Diffuse Lung Diseas

Diagnosis a
Hypersensit
CHEST Guideli

Evans R. Fernández Pérez,
Kevin K. Brown, MD, FCCP
Athol U. Wells, MD; Yuh-C
Mary-Beth Scholand, MD,
Richard B. Evans, MD, MP



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Vol. 76, No. 8, August 2024, pp 1201–1213
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2023 American College
College of Chest Physi
Screening and Monito
People with Systemic

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Sian Yik Lim,[19] Scott Matson,[20] Zsuzsan
Mary Beth Scholand,[23] Lee Shapiro,[24] Chri
Sandeep K. Agarwal,[27] Danielle Antin-Ozer
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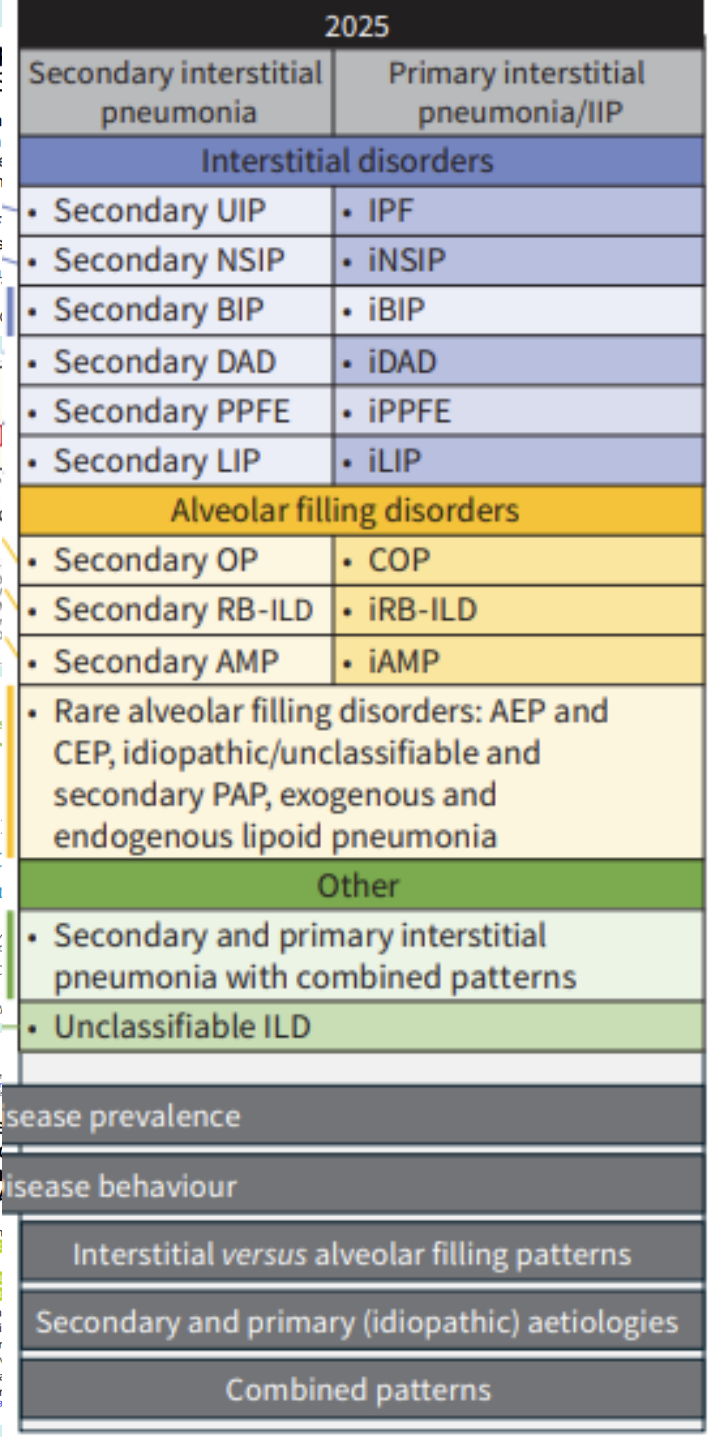
Justin M. Oldham,[7] Didem Saygin,[9] Kimber
Jeffrey A. Sparks,[14] Marat Turgunbaev,[3]



Diagnostic CT categories of fibrotic HP based on CT patterns. From Fernández Pérez ER, Travis WD, Lynch DA, et al. Diagnosis and Evaluation of Hypersensitivity Pneumonitis: CHEST Guideline and Expert Panel Report. *Chest*. 2021;160(2):e97-e156.

TABLE 5] Diagnostic CT Categories of Fibrotic HP Based on CT Patterns

HRCT	Typical Fibrotic HP	Compatible With Fibrotic HP	Indeterminate for Fibrotic HP
Features	CT signs of fibrosis with either of the following: • Profuse poorly defined centrilobular nodules of ground-glass opacity affecting all lung zones • Inspiratory mosaic attenuation with three-density sign ***And*** • Lack of features suggesting an alternative diagnosis	CT signs of fibrosis with any of the following: • Patchy or diffuse ground-glass opacity • Patchy, nonprofuse centrilobular nodules of ground-glass attenuation • Mosaic attenuation and lobular air-trapping that do not meet criteria for typical fibrotic HP ***And*** • Lack of features suggesting an alternative diagnosis	CT signs of fibrosis without other features suggestive of HP

CT signs of fibrosis include any of the following: reticular or ground-glass abnormality with traction bronchiectasis or bronchiolectasis; lobar volume loss; honeycombing. The distribution of fibrotic hypersensitivity pneumonitis (HP) is quite variable and often not diagnostically helpful. However, a mid-lung predominant distribution of fibrosis is suggestive of fibrotic HP, and an upper lobe predominance is much more common in fibrotic HP than in idiopathic pulmonary fibrosis. HRCT = high-resolution CT.

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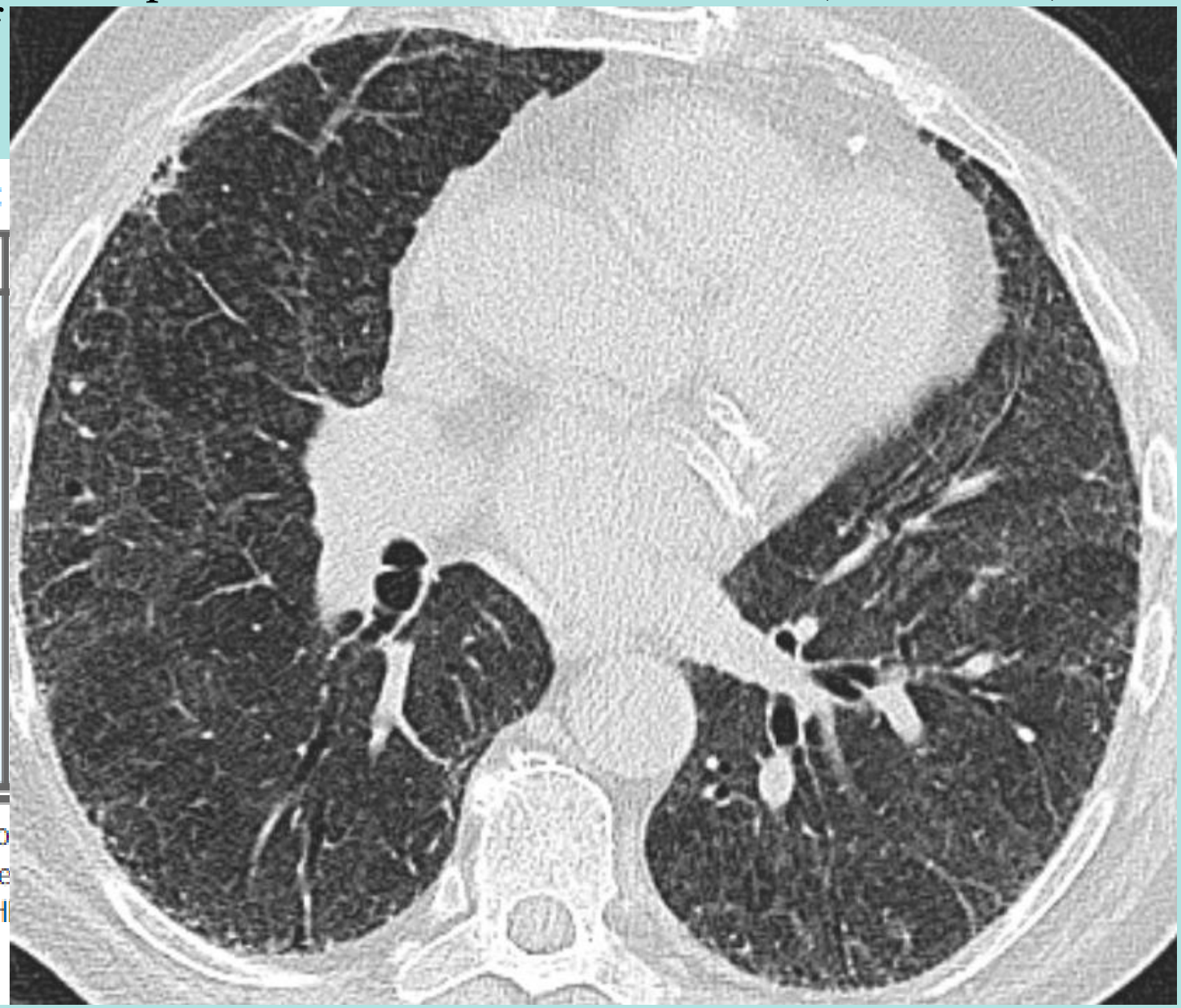

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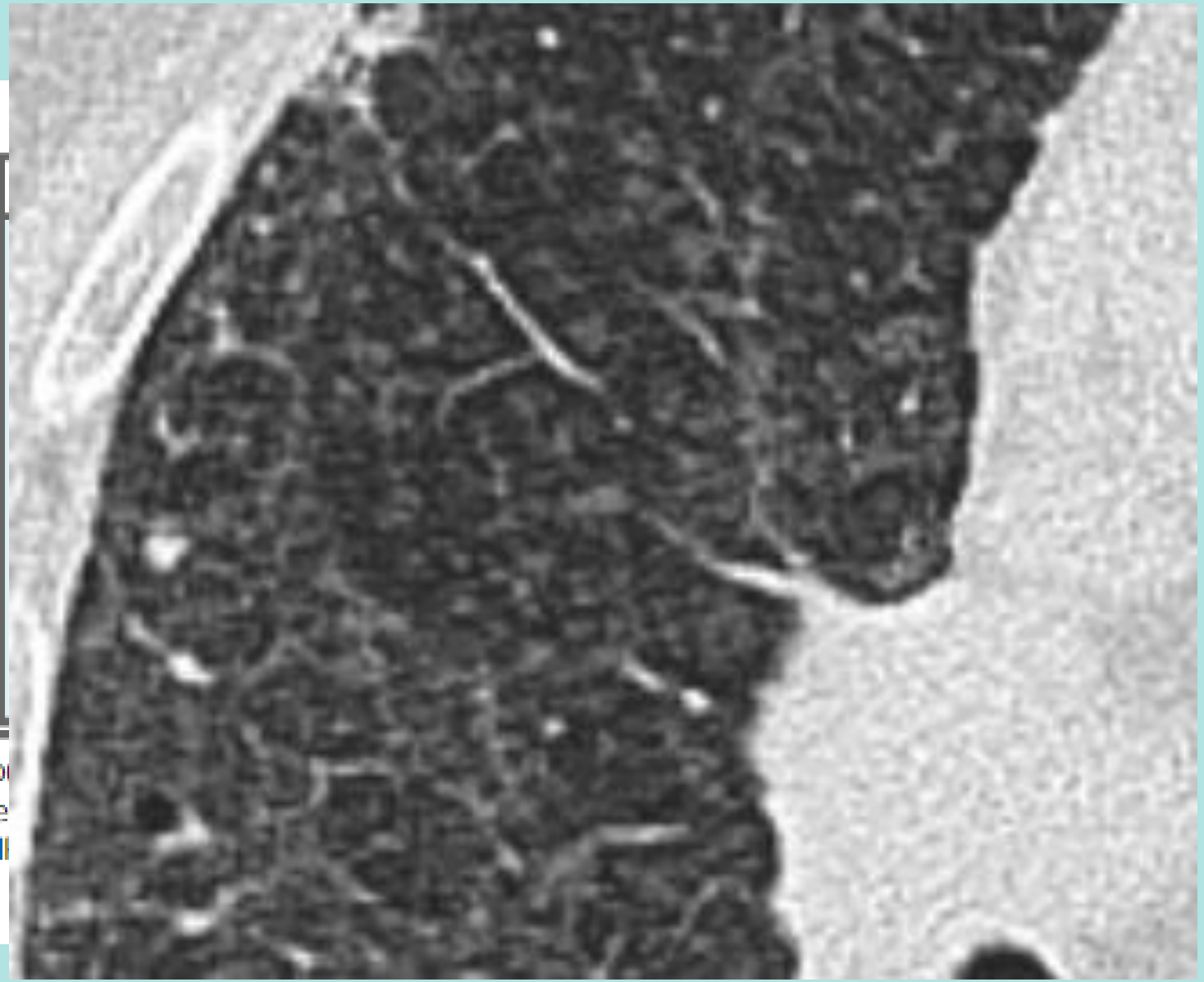
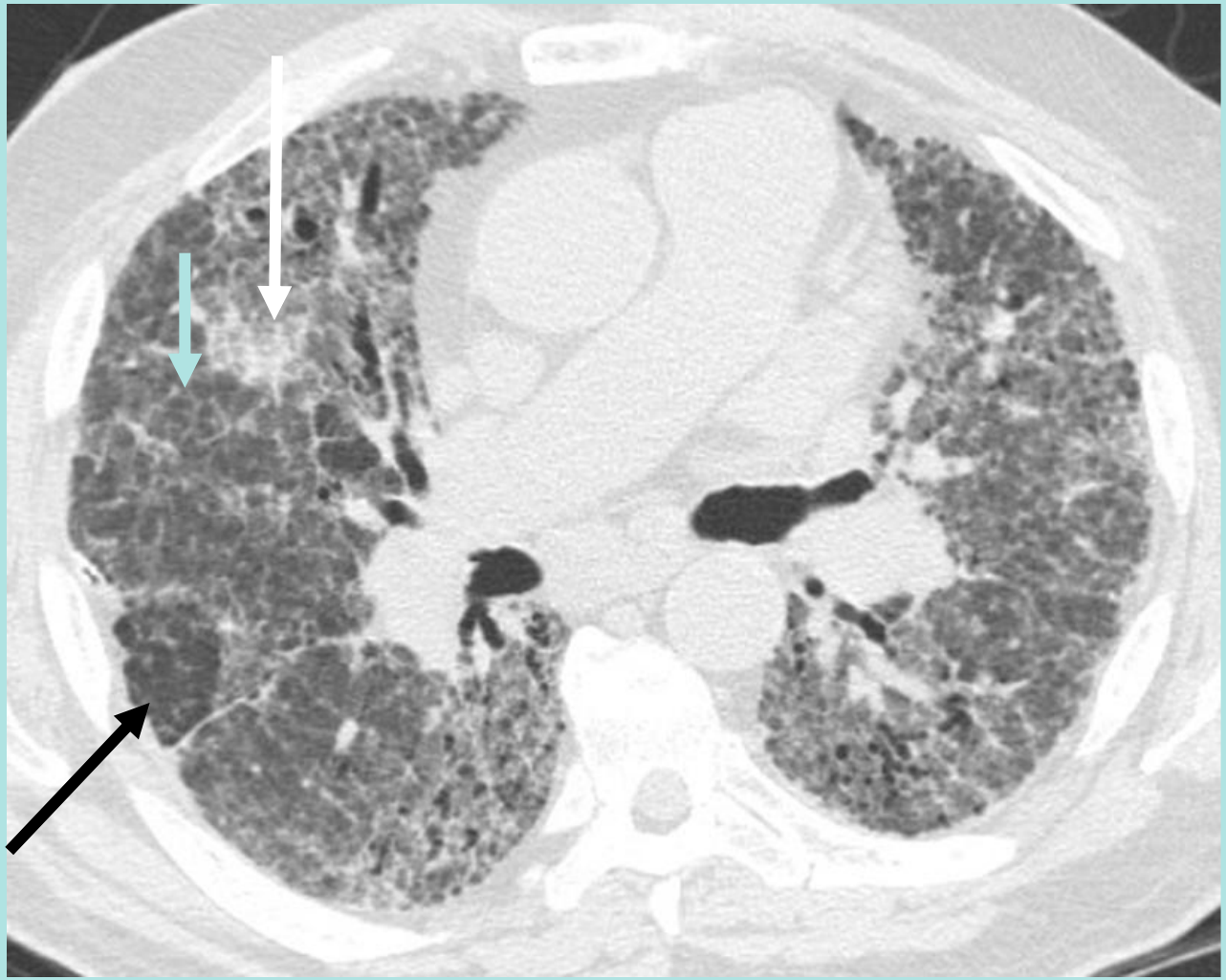
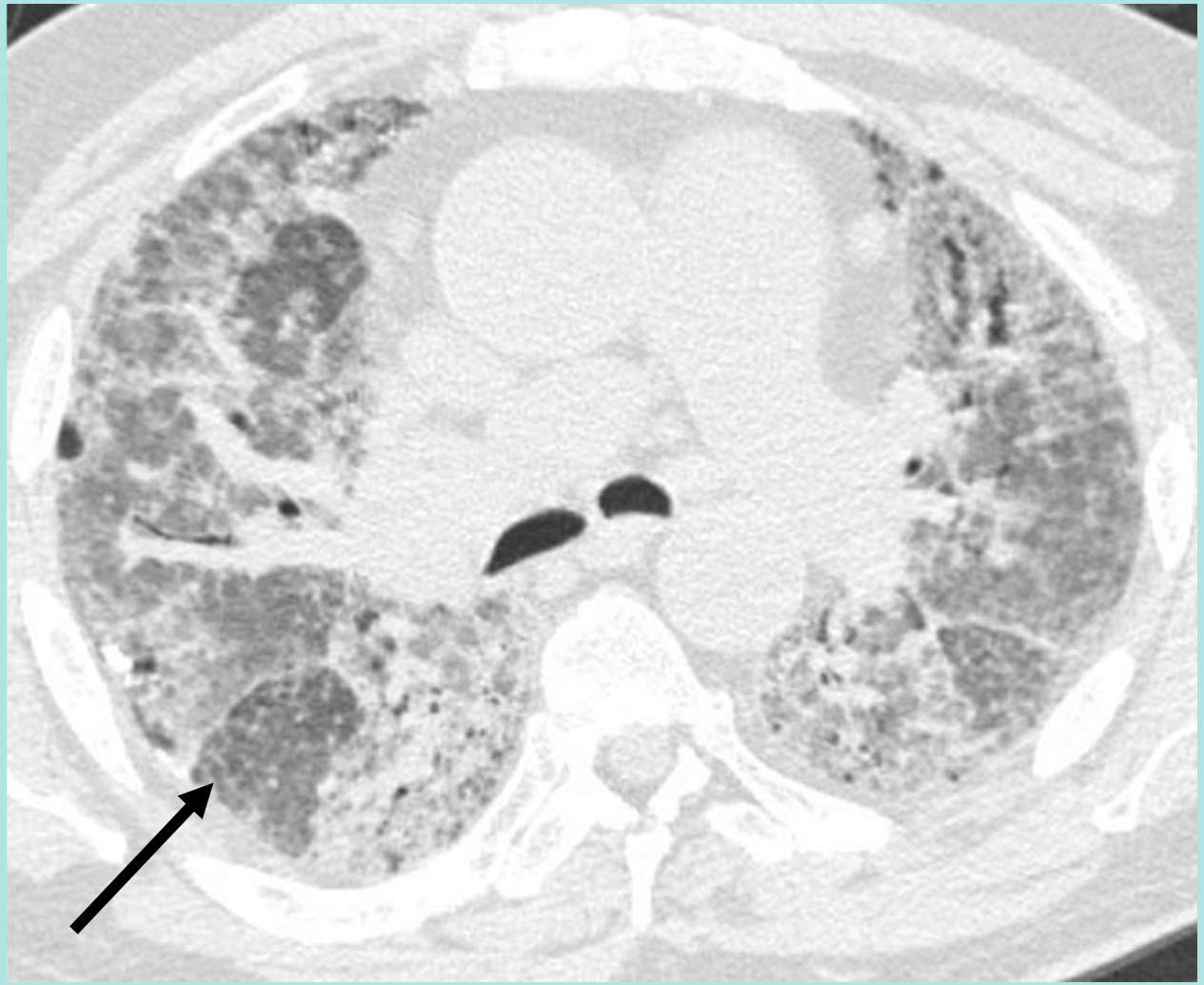
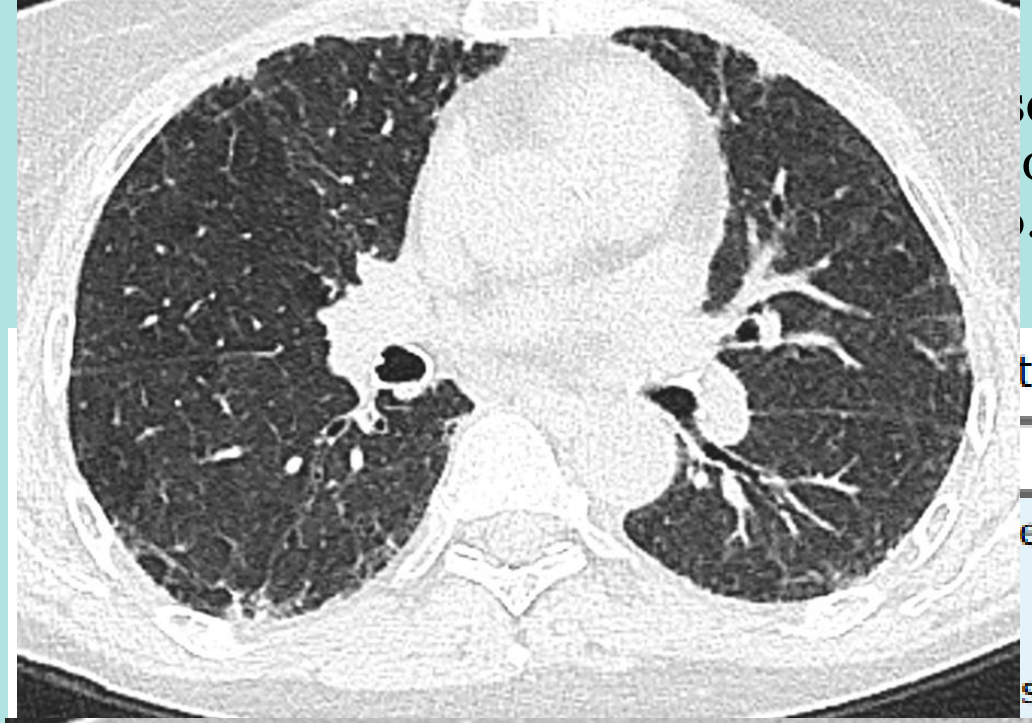
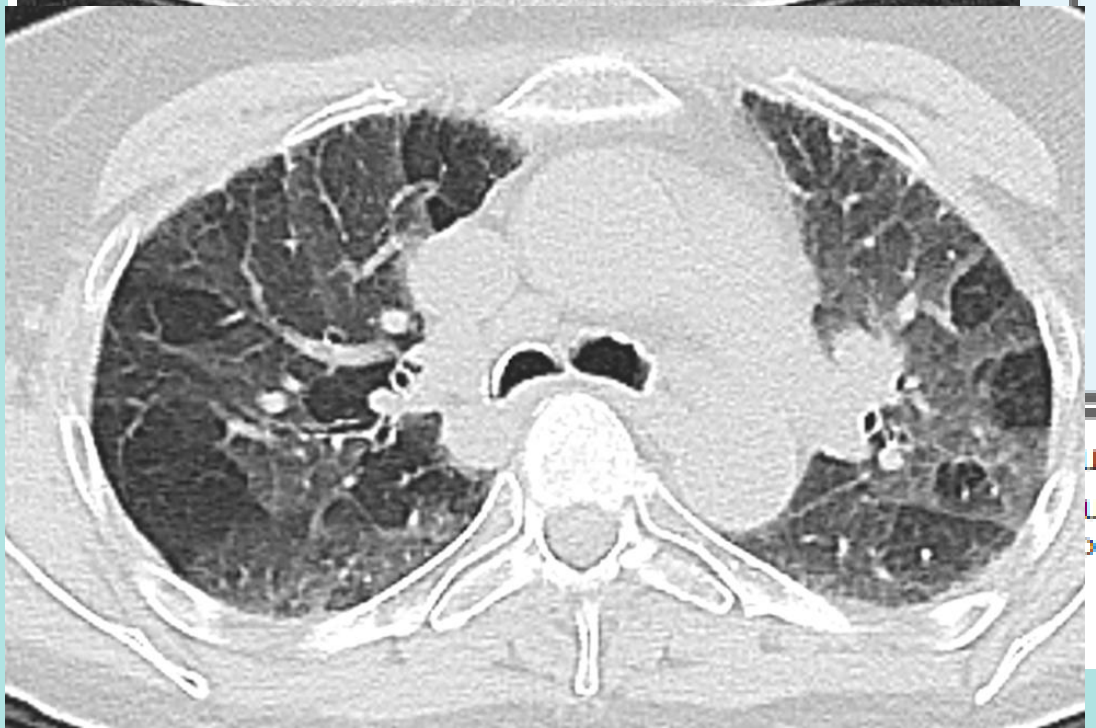


Typical fibrotic HP : Micronodules and three-density sign









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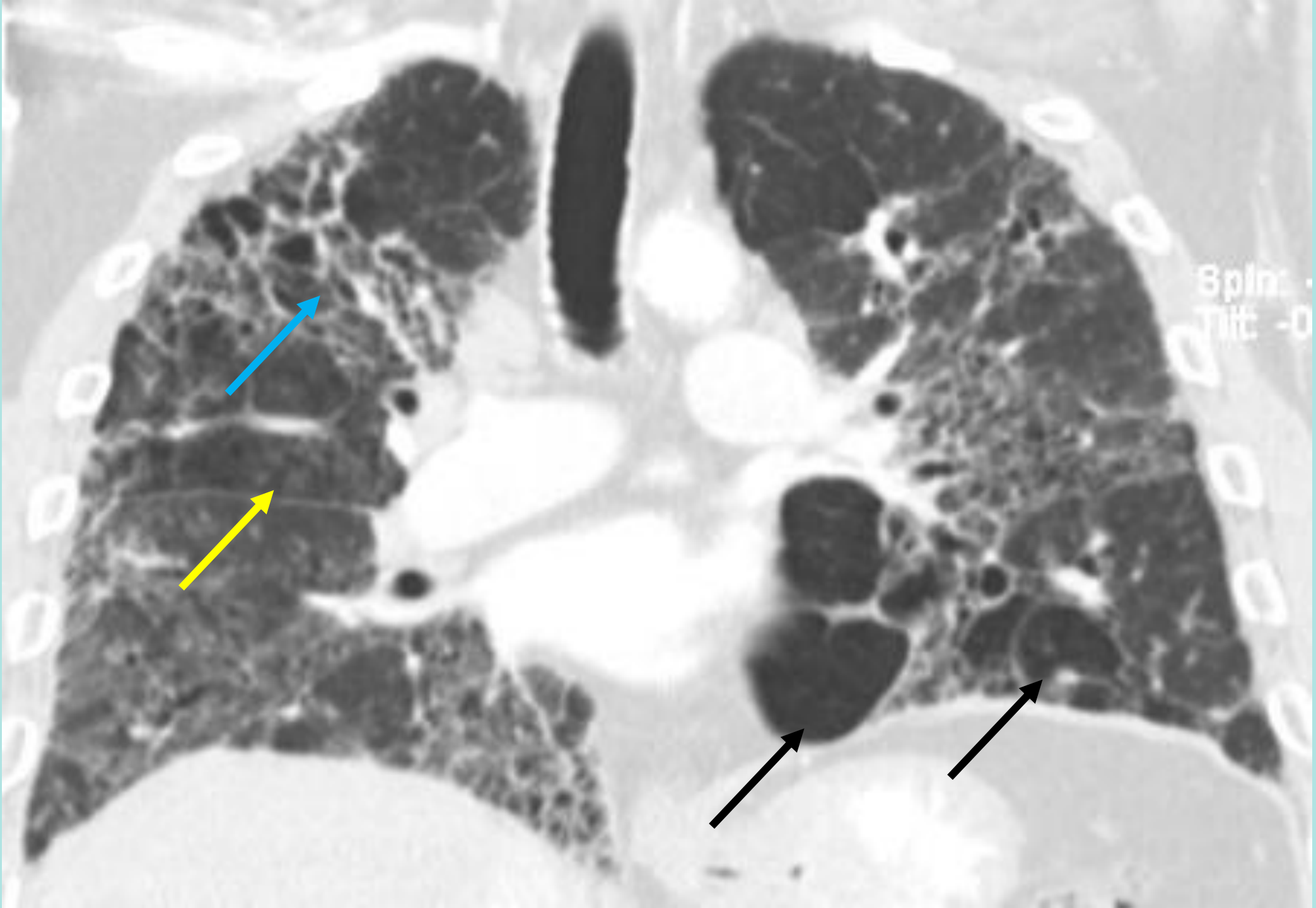
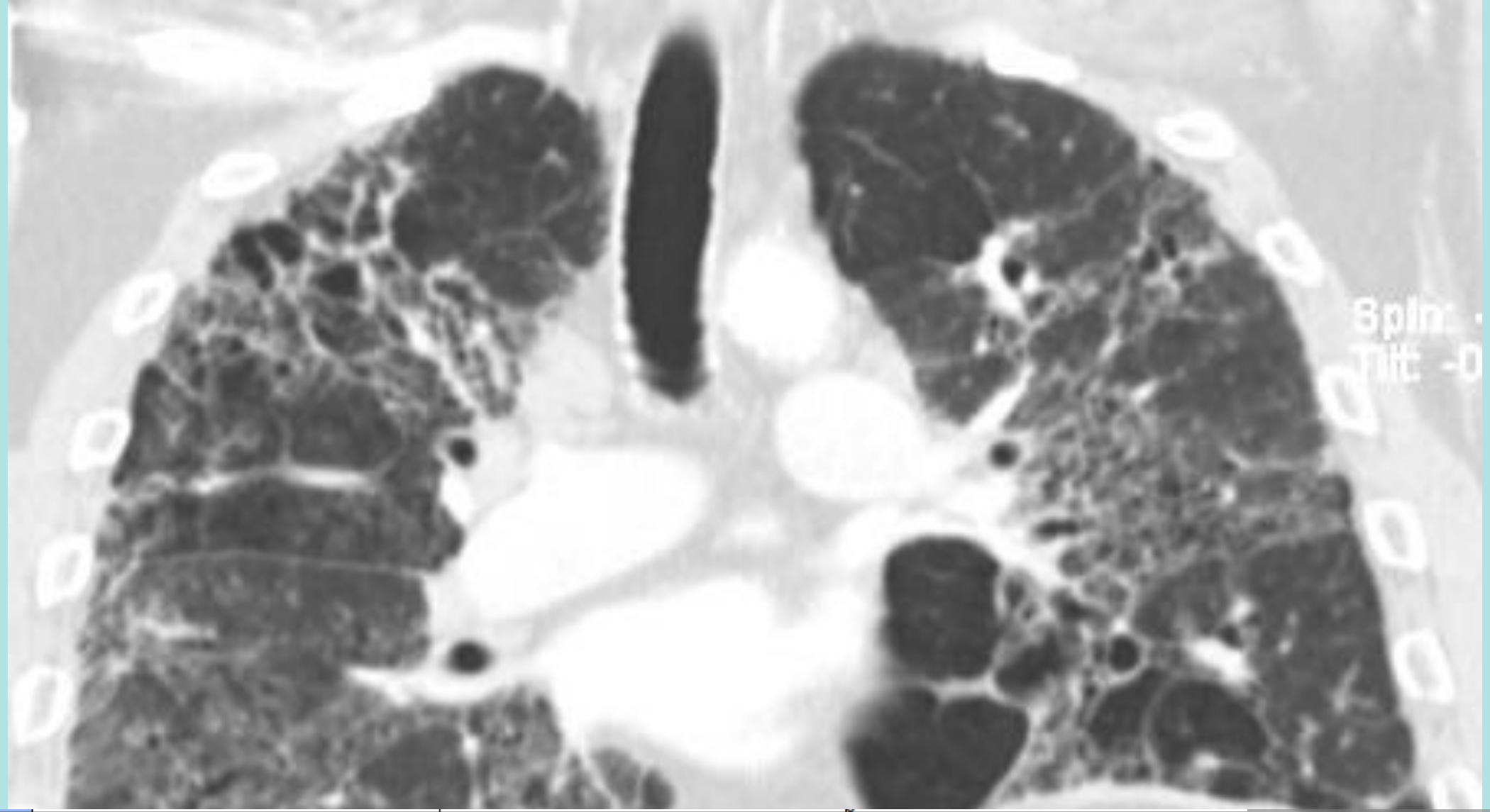
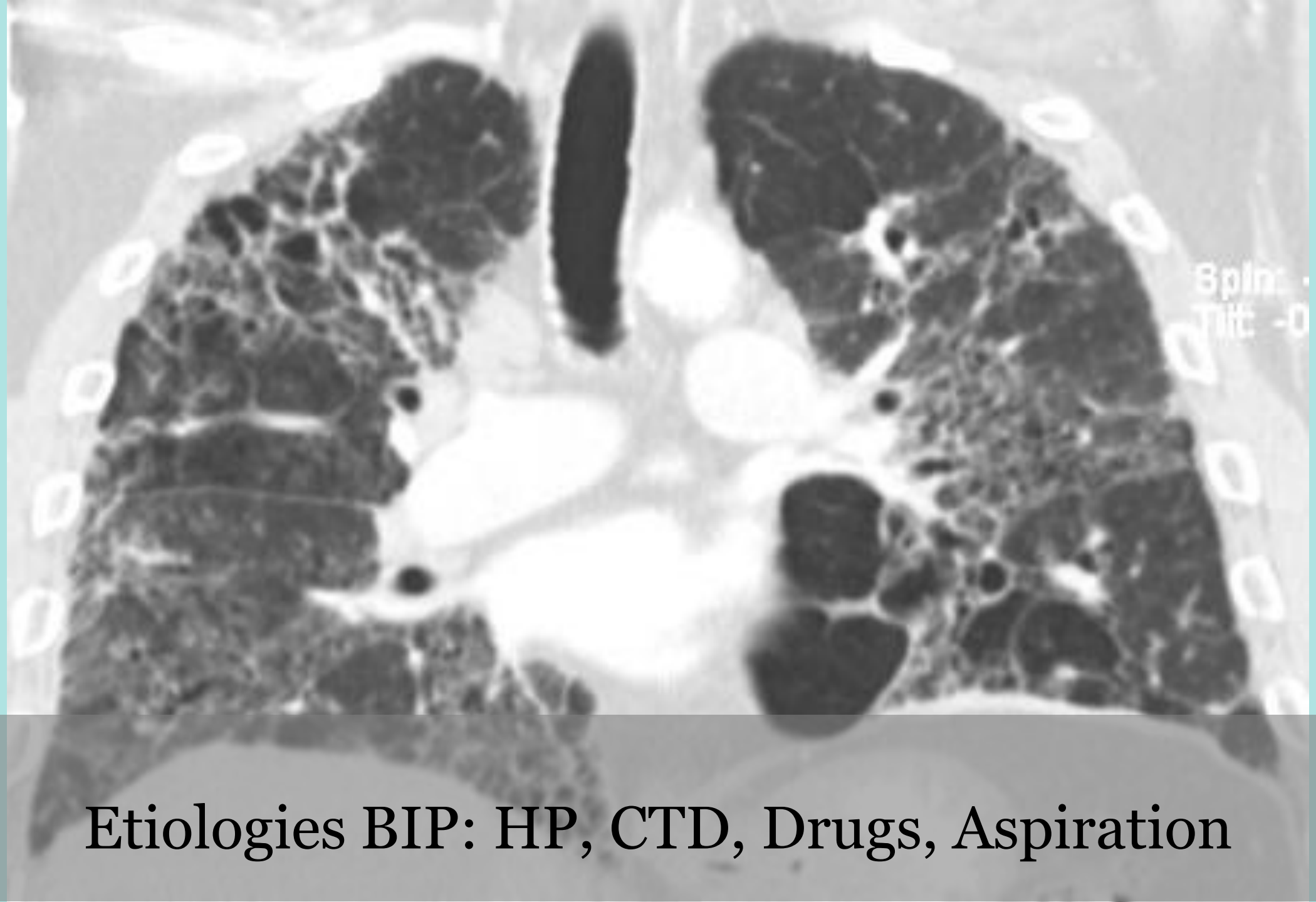


60-year-old woman with Rheumatoid arthritis



BIP	*Presentation:* Non-fibrotic BIP: Acute/Subacute onset frequently with respiratory and systemic symptoms. Fibrotic BIP: Chronic onset but with variable risk of progression. Potential for rapid worsening, especially with ongoing exposure. *Risk factors:* No clear age or sex predilection. Positive exposure history for HP associated antigens.	Non-fibrotic BIP: Centrilobular ground glass nodules, mosaic attenuation, and/or lobular air trapping. Fibrotic BIP: Peribronchovascular ground glass, traction bronchiectasis, mosaic attenuation, lobular air trapping and three-density sign. Features can overlap with non-fibrotic BIP.	Nonfibrotic BIP: Bronchiolocentric chronic inflammation +/- non-necrotising granulomatous inflammation. Fibrotic BIP: Bronchiolocentric fibrosis often with peribronchiolar metaplasia ± bronchiolocentric inflammation.

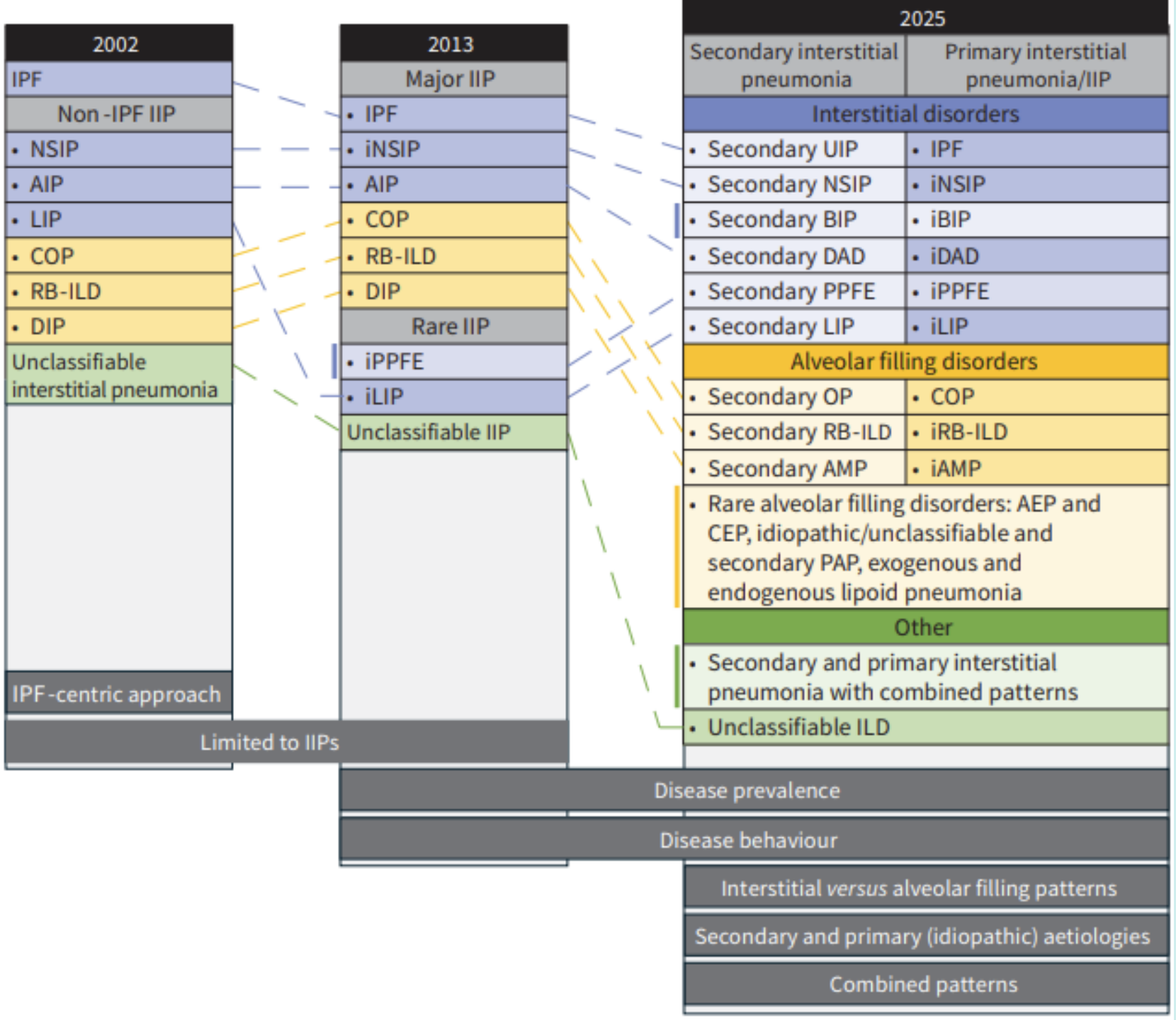


Etiologies BIP: HP, CTD, Drugs, Aspiration

BIP Controversies

What is the radiology correlation of histologic Bronchiolocentric Interstitial Pneumonia?

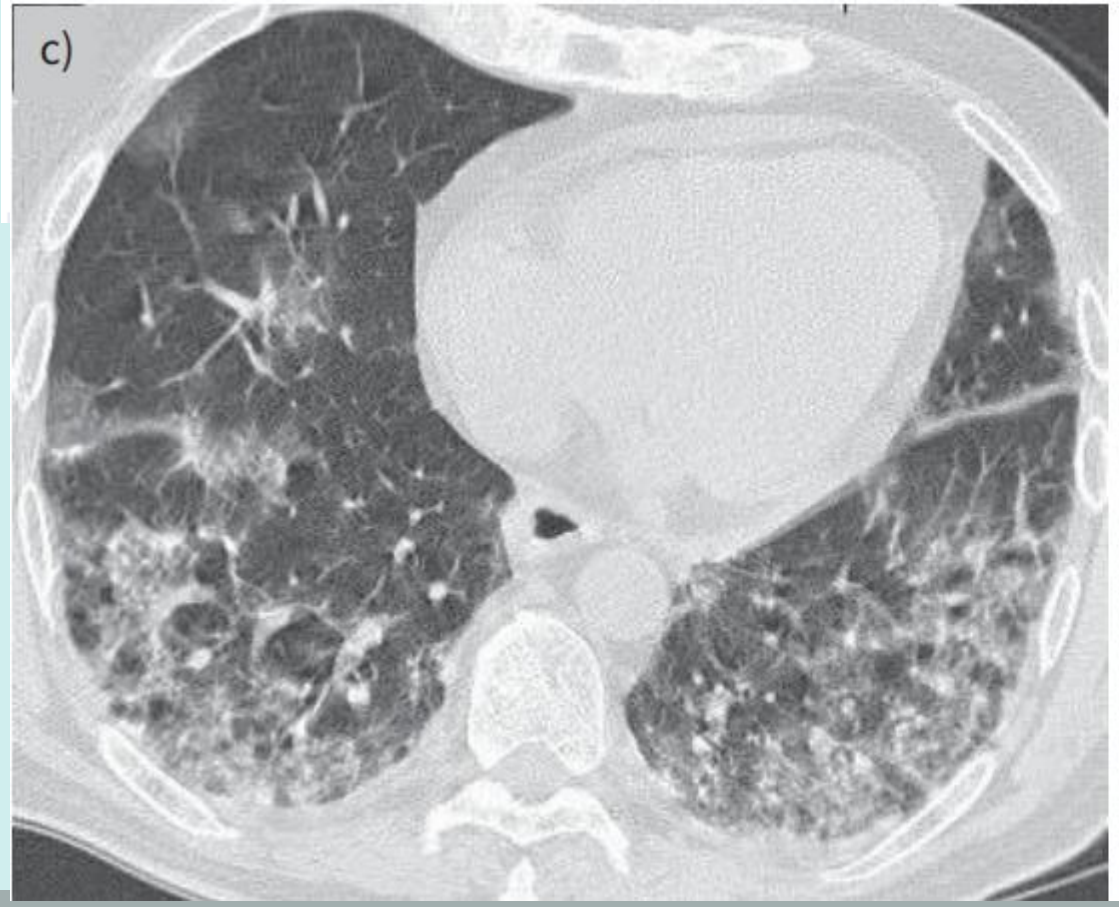
What is Idiopathic BIP? Will this deter motivation for the search for an antigen as a cause of HP?

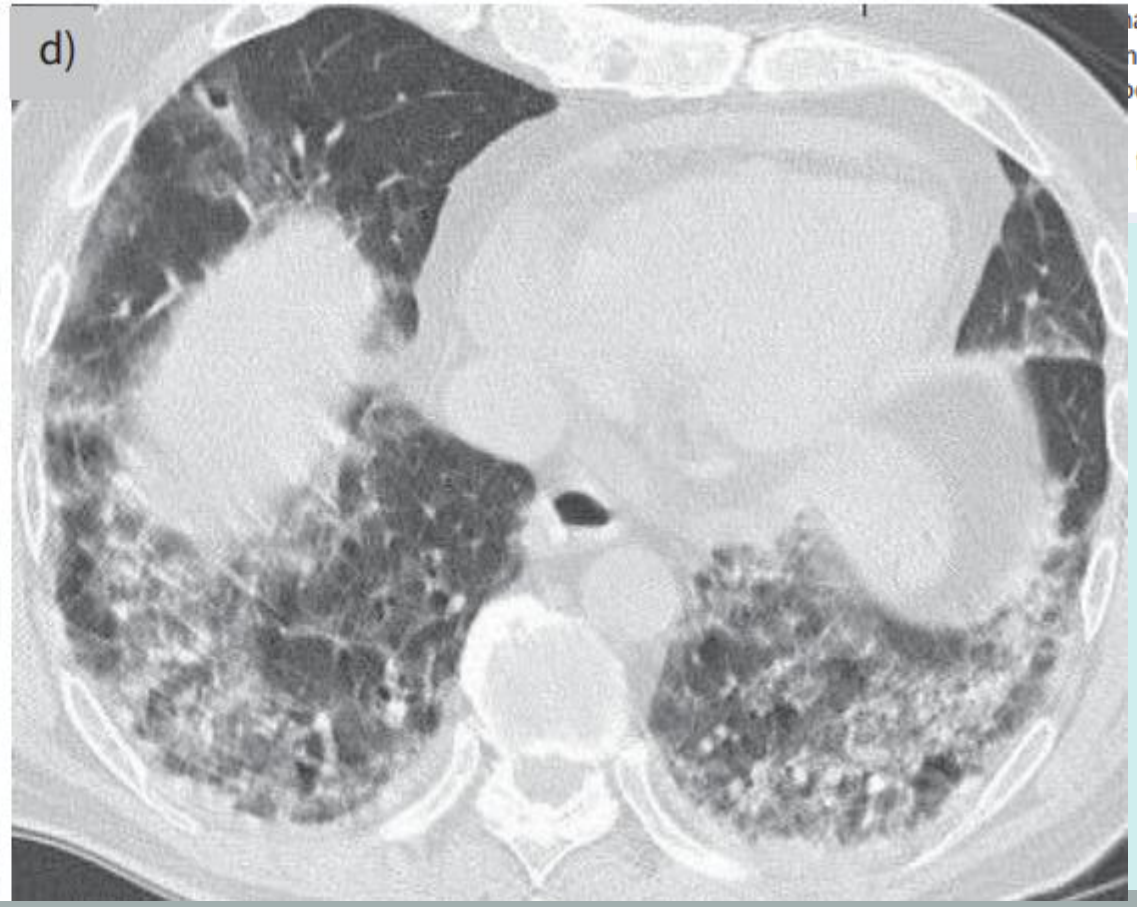


Eur Respir J 2025; 66: 2500158

TABLE 2 Typical pathological, radiological and clinical features of major interstitial pneumonia patterns

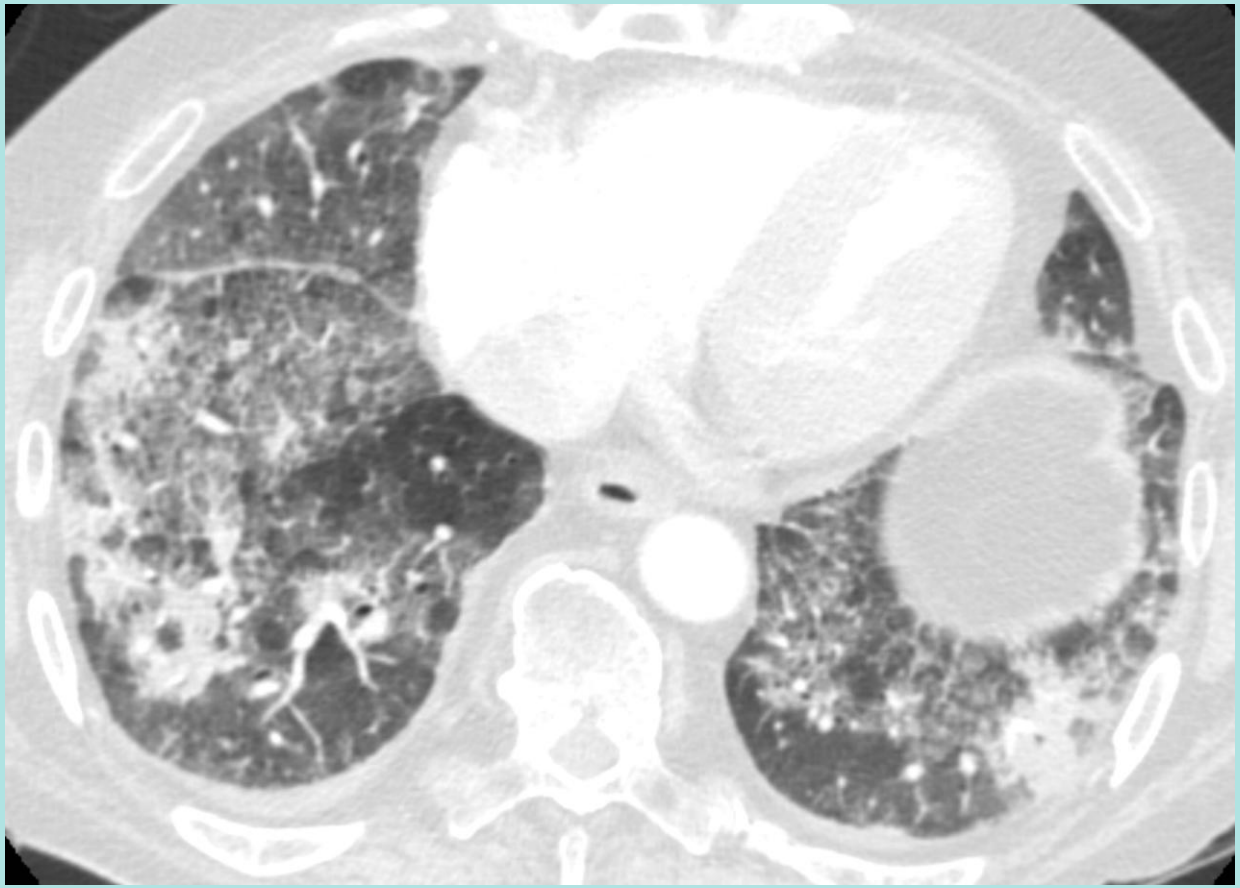
	Clinical features	Radiological features	Pathological features
Interstitial patterns			
DAD	Presentation: acute worsening with high likelihood of residual fibrosis and potential for ongoing progression	Patchy or diffuse ground-glass opacity that may progress to fibrosis over time with significant traction bronchiectasis, architectural distortion and honeycombing	Exudative phase: alveolar septa are thickened by interstitial oedema, pneumocyte hyperplasia and surface hyaline membranes

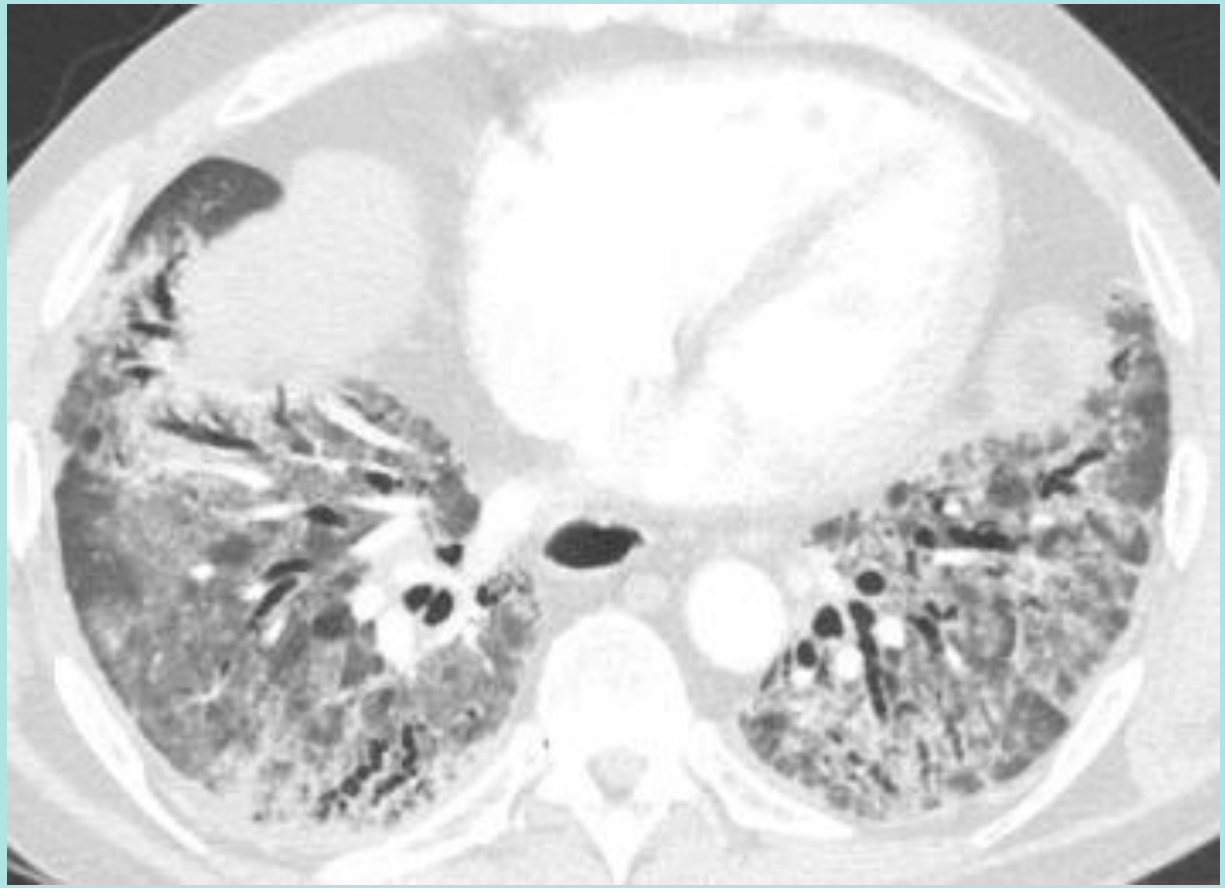




Acute Interstitial Pneumonia (AIP) = Idiopathic Diffuse Alveolar Damage (iDAD)

Idiopathic DAD: 5 days later





Survivors:

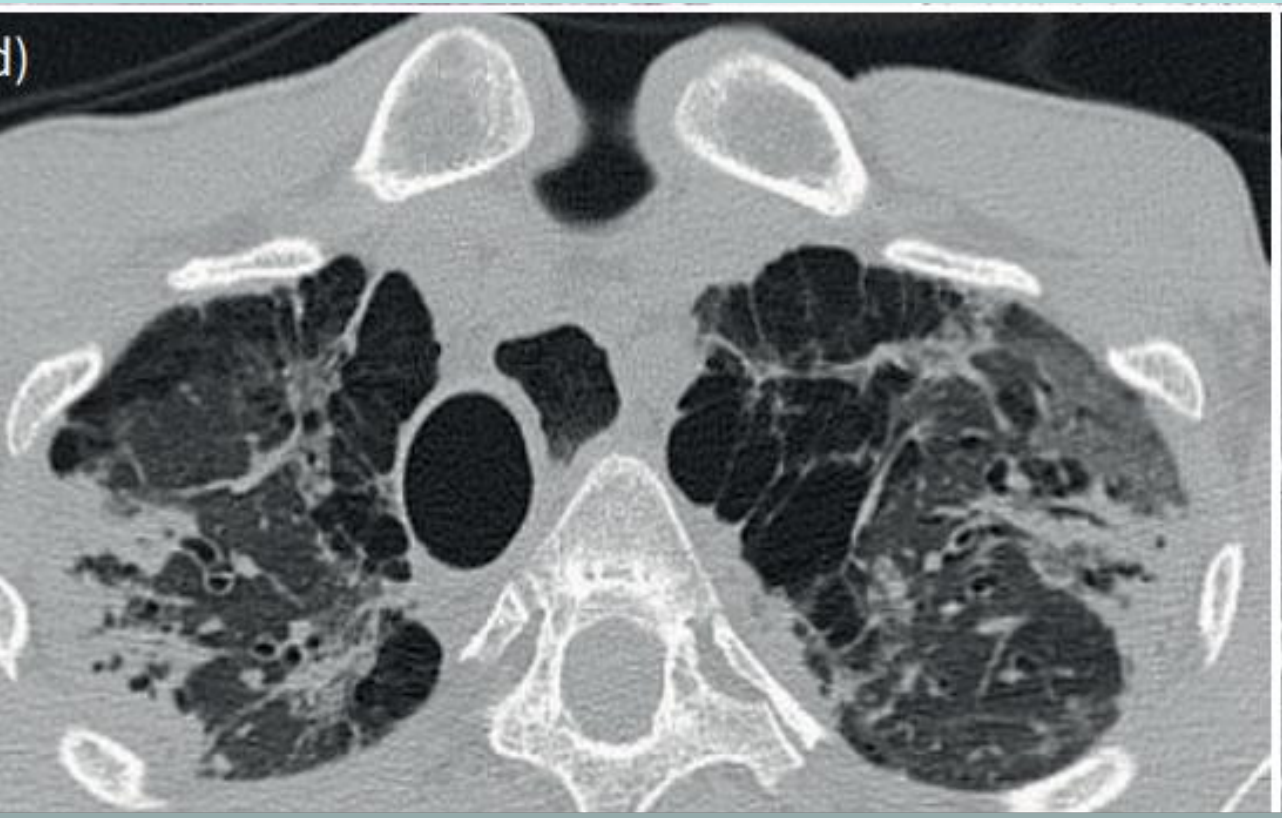
More Ground-glass without associated traction bronchiectasis

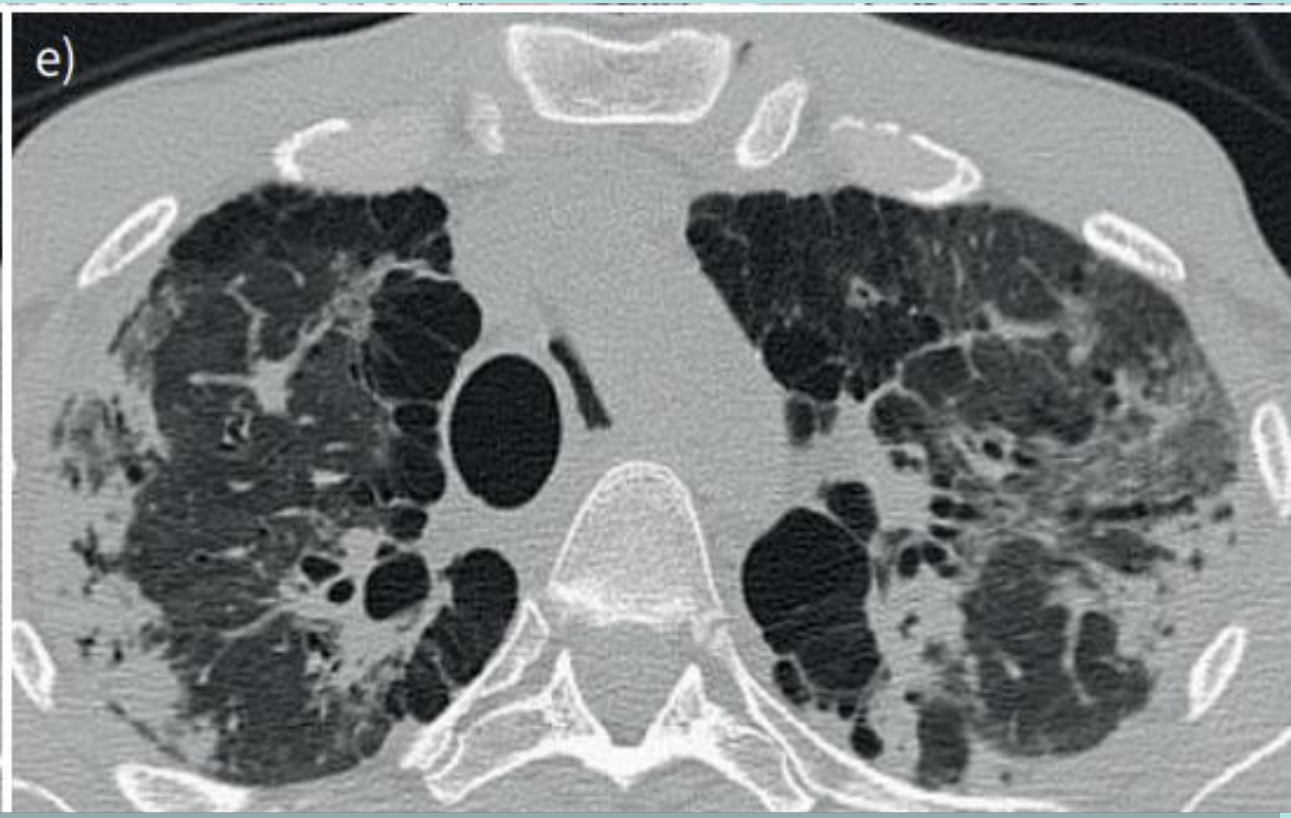
Less architectural Distortion

Am J Respir Crit Care Med 2002;1551-56

TABLE 2 Typical pathological, radiological and clinical features of major interstitial pneumonia patterns

	Clinical features	Radiological features	Pathological features
PPFE	Presentation: chronic onset, but with variable risk of progression. Frequently associated with significant weight loss Risk factors: no clear age or sex predilection	Biapical pleural thickening, dense subpleural confluent fibrosis and traction bronchiectasis with volume loss and upward hilar retraction, which may extend more caudally to involving the major fissures, and occasionally into the middle and lower lobes. May include platythorax, deep sternal notch, and/or tracheal deviation. Often coexists with additional patterns, especially with UIP	Upper zone pleural fibrosis with subpleural intra-alveolar fibroelastosis, often with extension of fibrosis into underlying lung parenchyma. Some cases may lack upper lobe predominance. At least one-third of cases coexist with additional patterns or specific findings characteristic of UIP, NSIP, BIP, granulomas, or mycobacterial or fungal infection

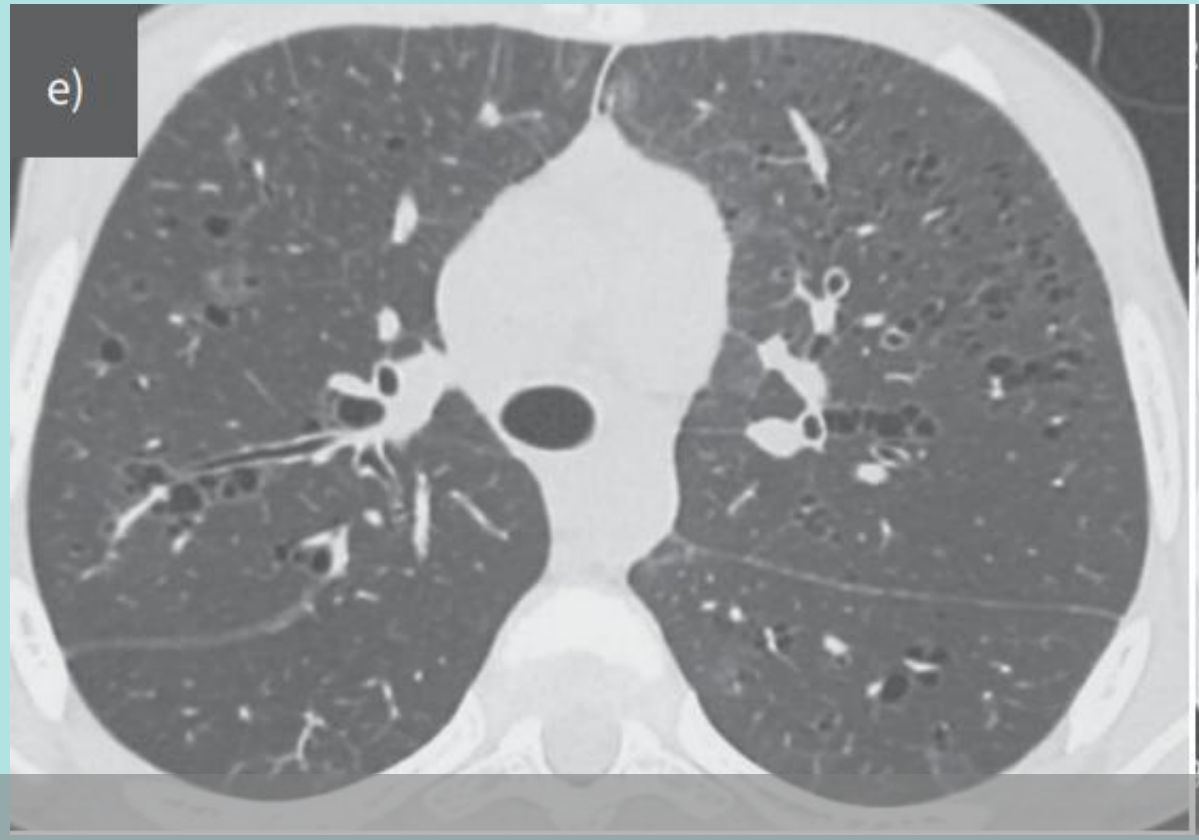


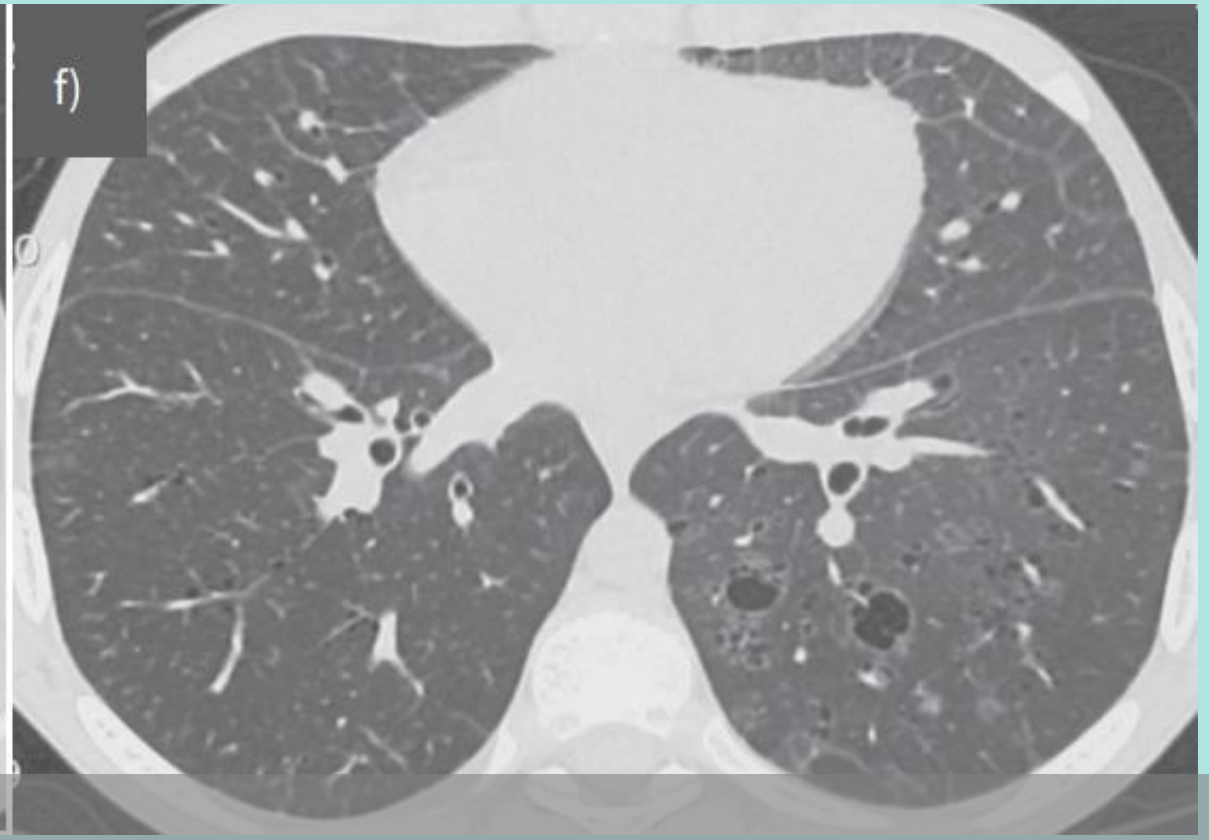


Pleuroparenchymal Fibroelastosis

TABLE 2 Typical pathological, radiological and clinical features of major interstitial pneumonia patterns

	Clinical features	Radiological features	Pathological features
Interstitial patterns			
LIP	Presentation: insidious onset with slow progression Risk factors: no clear age or sex predilection for idiopathic LIP. Risk factors for secondary LIP vary with underlying aetiology	Mid- to lower-lobe-predominant thin-walled lung cysts with reticulation, ground-glass opacity, bronchovascular thickening and perilymphatic interstitial thickening; randomly distributed or lower-lobe-predominant thin-walled cysts may be the only or dominant finding	Dense and diffuse interstitial infiltrate of lymphocytes and plasma cells expanding the alveolar septa, often with peribronchial and peribronchiolar lymphoid follicles, sometimes showing germinal centres. Lymphoplasmacytic infiltrate is polyclonal. Dutcher bodies are absent





Lymphocytic Interstitial Pneumonia (LIP)

Eur Respir J 2025; 66: 2500158

TABLE 2 Typical pathological, radiological and clinical features of major interstitial pneumonia patterns

	Clinical features	Radiological features	Pathological features
Interstitial patterns			
UIP	Presentation: chronic onset with high likelihood of progression and potential for rapid worsening and acute exacerbation Risk factors: more frequent in older males with a smoking history, particularly for IPF	Subpleural and usually lower lung predominance. Fibrotic changes including reticulation, peripheral traction bronchiectasis, and honeycombing	Dense fibrosis, typically including fibroblast foci, with a predilection for subpleural and paraseptal parenchyma; architectural distortion in the form of honeycomb change juxtaposed with relatively unaffected lung parenchyma
NSIP	Presentation: chronic onset, but with variable risk of progression. Potential for rapid worsening, especially with concurrent organising pneumonia Risk factors: more common in younger females, often with a background of CTD	Lower-lobe predominance, commonly with subpleural sparing. Ground-glass opacities, fine reticulation, and traction bronchiectasis, without honeycombing	Cellular NSIP: cellular interstitial infiltrate of lymphocytes causing thickening of alveolar walls with no fibrosis or granulomas Fibrotic NSIP: diffuse thickening of alveolar walls by fibrosis with traction bronchiolectasis; no bronchiolocentricity, honeycombing, or granulomas
BIP	Presentation Non-fibrotic BIP: acute/subacute onset frequently with respiratory and systemic symptoms Fibrotic BIP: chronic onset but with variable risk of progression. Potential for rapid worsening, especially with ongoing exposure Risk factors No clear age or sex predilection. Positive exposure history for HP-associated antigens	Non-fibrotic BIP: centrilobular ground-glass nodules, mosaic attenuation, and/or lobular air trapping Fibrotic BIP: peribronchovascular ground-glass, traction bronchiectasis, mosaic attenuation, lobular air trapping, and three-density sign. Features can overlap with non-fibrotic BIP	Non-fibrotic BIP: bronchiolocentric chronic inflammation ± non-necrotising granulomatous inflammation Fibrotic BIP: bronchiolocentric fibrosis often with peribronchiolar metaplasia ± bronchiolocentric inflammation
DAD	Presentation: acute worsening with high likelihood of residual fibrosis and potential for ongoing progression Risk factors: vary with underlying aetiology (*e.g.* sepsis, aspiration, pneumonia, CTD-ILD)	Patchy or diffuse ground-glass opacity that may progress to fibrosis over time with significant traction bronchiectasis, architectural distortion and honeycombing	Exudative phase: alveolar septa are thickened by interstitial oedema, pneumocyte hyperplasia and surface hyaline membranes Organising phase: evolution into predominant interstitial involvement by loose organising connective tissue and exuberant proliferation of pneumocytes, often with some atypia Fibrotic phase: DAD can progress to interstitial fibrosis with varying features
PPFE	Presentation: chronic onset, but with variable risk of progression. Frequently associated with significant weight loss Risk factors: no clear age or sex predilection	Biapical pleural thickening, dense subpleural confluent fibrosis and traction bronchiectasis with volume loss and upward hilar retraction, which may extend more caudally to involving the major fissures, and occasionally into the middle and lower lobes. May include platythorax, deep sternal notch, and/or tracheal deviation. Often coexists with additional patterns, especially with UIP	Upper zone pleural fibrosis with subpleural intra-alveolar fibroelastosis, often with extension of fibrosis into underlying lung parenchyma. Some cases may lack upper lobe predominance. At least one-third of cases coexist with additional patterns or specific findings characteristic of UIP, NSIP, BIP, granulomas, or mycobacterial or fungal infection
LIP	Presentation: insidious onset with slow progression Risk factors: no clear age or sex predilection for idiopathic LIP. Risk factors for secondary LIP vary with underlying aetiology	Mid- to lower-lobe-predominant thin-walled lung cysts with reticulation, ground-glass opacity, bronchovascular thickening and perilymphatic interstitial thickening; randomly distributed or lower-lobe-predominant thin-walled cysts may be the only or dominant finding	Dense and diffuse interstitial infiltrate of lymphocytes and plasma cells expanding the alveolar septa, often with peribronchial and peribronchiolar lymphoid follicles, sometimes showing germinal centres. Lymphoplasmacytic infiltrate is polyclonal. Dutcher bodies are absent

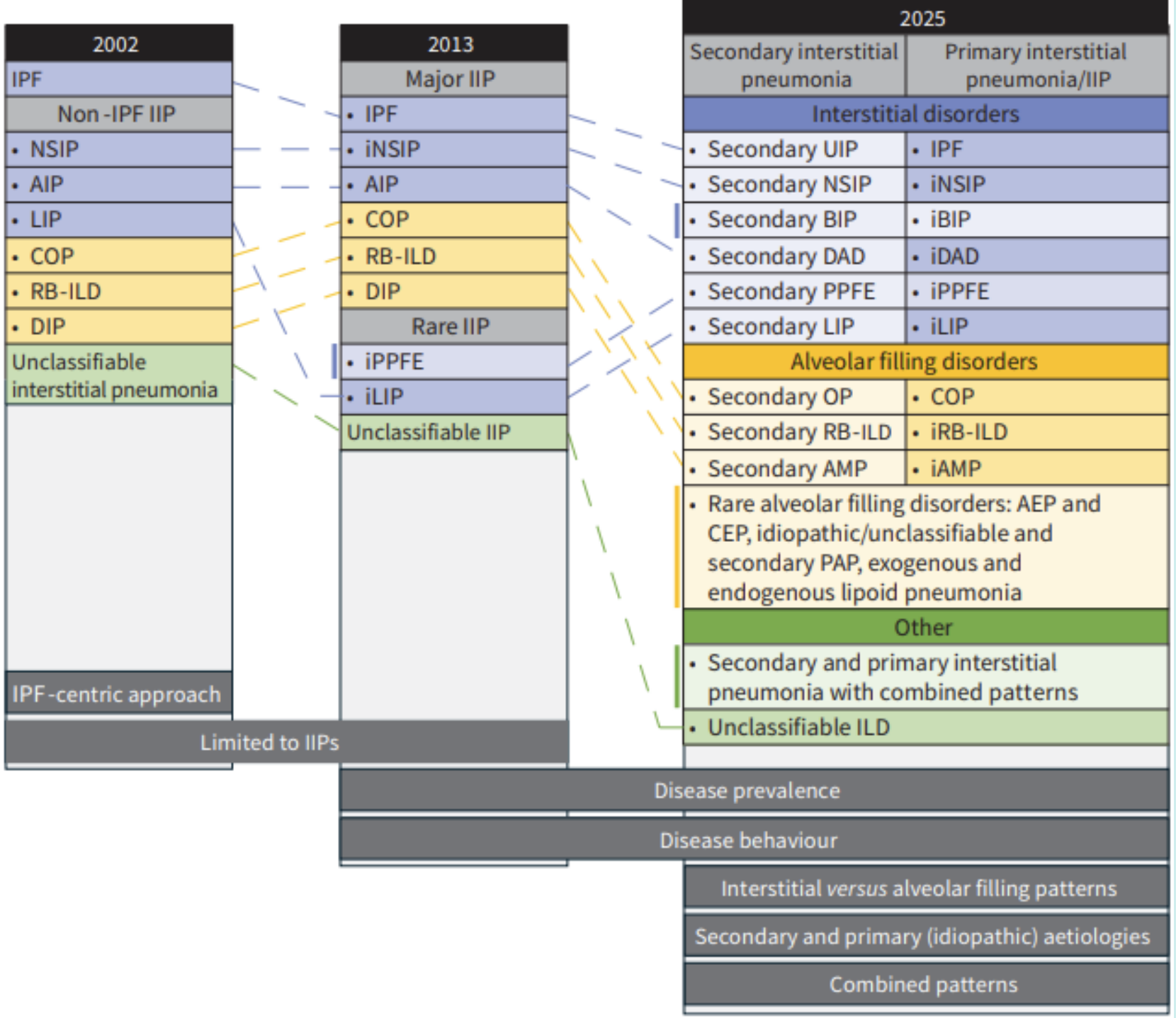
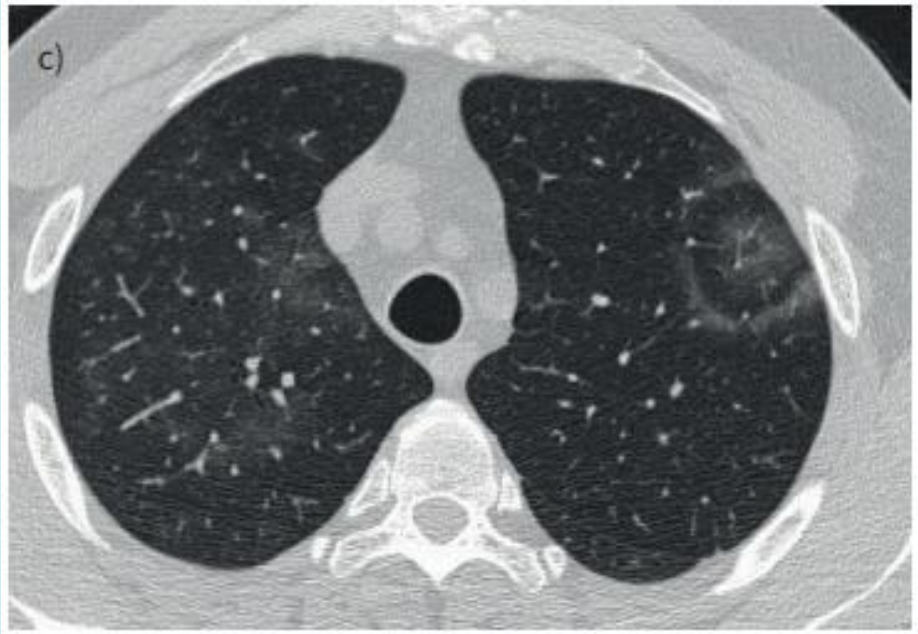
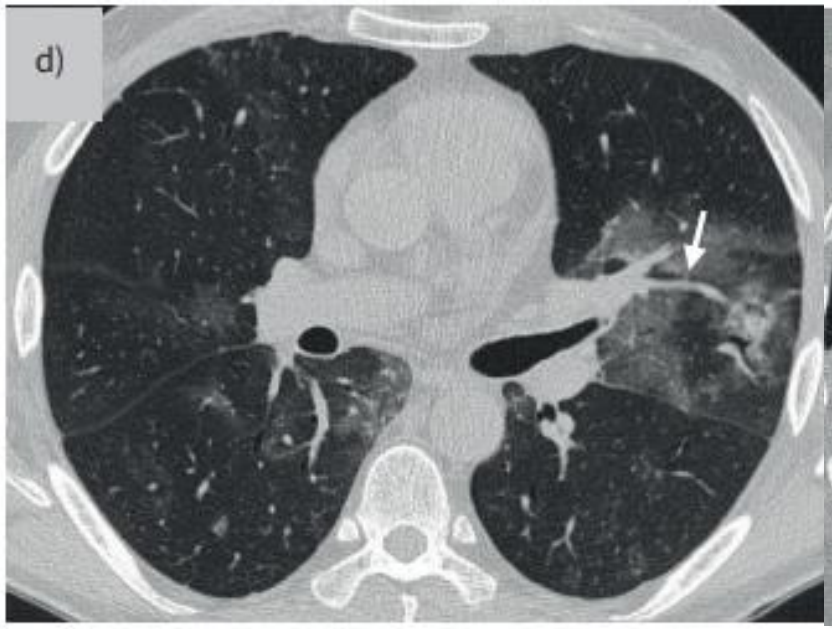

2002
IPF
Non-IPF IIP
• NSIP
• AIP
• LIP
• COP
• RB-ILD
• DIP
Unclassifiable interstitial pneumonia
IPF-centric approach
Limited to IIPs
2013
Major IIP
• IPF
• iNSIP
• AIP
• COP
• RB-ILD
• DIP
Rare IIP
• iPPFE
• iLIP
Unclassifiable IIP
Disease prevalence
Disease behaviour
2025
Secondary interstitial pneumonia
Primary interstitial pneumonia/IIP
Interstitial disorders
• Secondary UIP
• IPF
• Secondary NSIP
• iNSIP
• Secondary BIP
• iBIP
• Secondary DAD
• iDAD
• Secondary PPFE
• iPPFE
• Secondary LIP
• iLIP
Alveolar filling disorders
• Secondary OP
• COP
• Secondary RB-ILD
• iRB-ILD
• Secondary AMP
• iAMP
• Rare alveolar filling disorders: AEP and CEP, idiopathic/unclassifiable and secondary PAP, exogenous and endogenous lipoid pneumonia
Other
• Secondary and primary interstitial pneumonia with combined patterns
• Unclassifiable ILD
Interstitial versus alveolar filling patterns
Secondary and primary (idiopathic) aetiologies
Combined patterns

TABLE 2 Continued

	Clinical features	Radiological features	Pathological features
Alveolar filling patterns			
Organising pneumonia	Presentation: acute or subacute onset of respiratory ± constitutional symptoms. Typically reversible, but potential for residual fibrosis; low potential for ongoing progression for COP, with higher potential for recurrence or progression for some causes, particularly CTD Risk factors: no clear age or sex predilection for COP. Risk factors for secondary organising pneumonia vary with underlying aetiology	Patchy consolidation with air bronchograms, patchy ground-glass opacities, focal nodules, and/or mass-like lesions, sometimes featuring a halo sign, reversed halo sign, perilobular pattern, or air-bronchograms	Intra-alveolar buds of loose connective tissue predominantly within alveoli, alveolar ducts ± respiratory bronchioles. Interstitial inflammation is typically mild in COP. Secondary organising pneumonia can be associated with ancillary findings





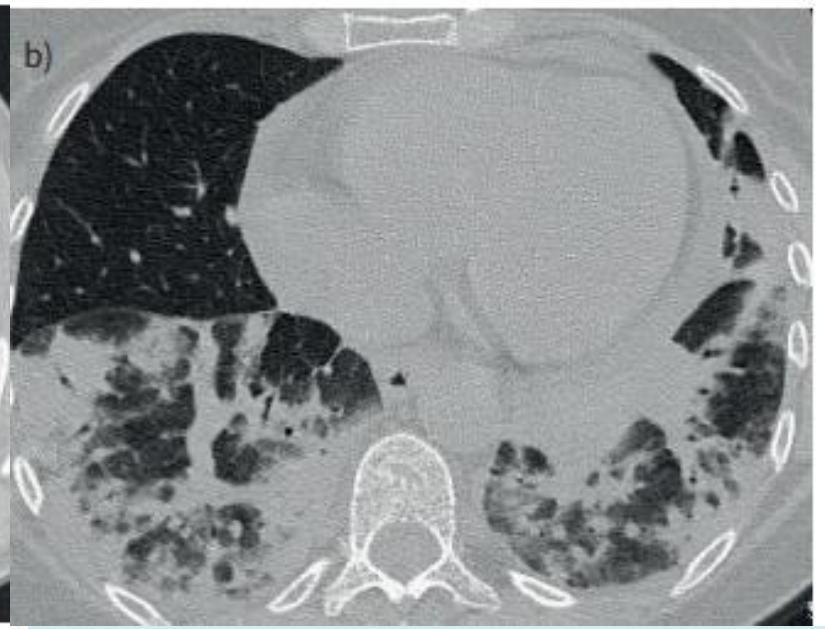


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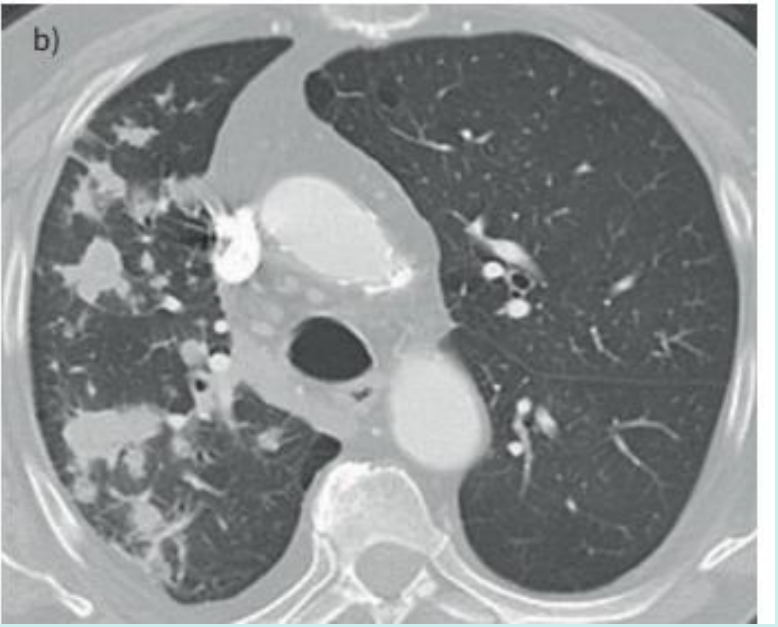
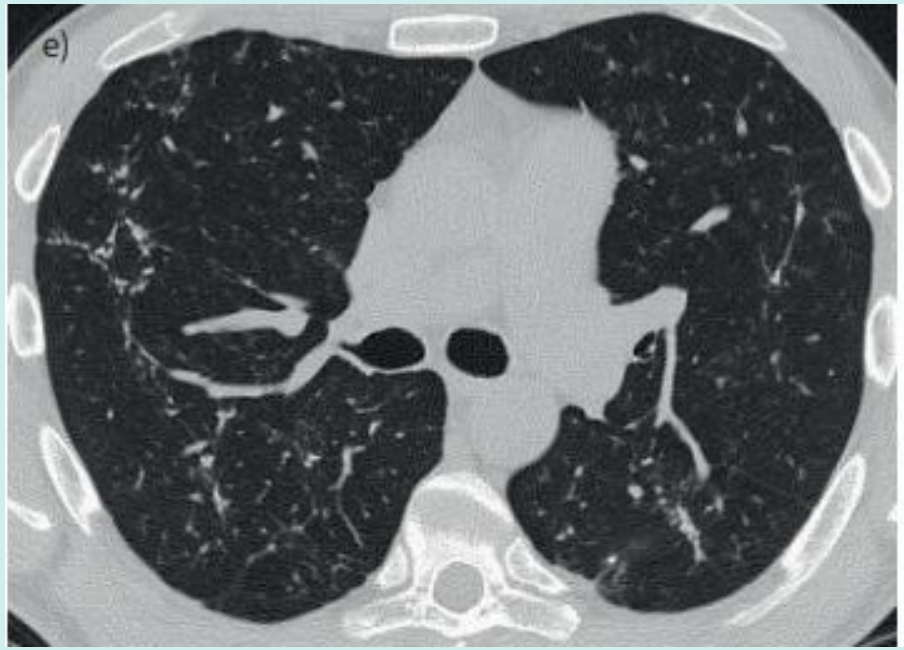
TABLE 2 Continued

	Clinical features	Radiological features	Pathological features
Alveolar filling patterns			
Organising pneumonia	Presentation: acute or subacute onset of respiratory ± constitutional symptoms. Typically reversible, but potential for residual fibrosis; low potential for ongoing progression for COP, with higher potential for recurrence or progression for some causes, particularly CTD Risk factors: no clear age or sex predilection for COP. Risk factors for secondary organising pneumonia vary with underlying aetiology	Patchy consolidation with air bronchograms, patchy ground-glass opacities, focal nodules, and/or mass-like lesions, sometimes featuring a halo sign, reversed halo sign, perilobular pattern, or air-bronchograms	Intra-alveolar buds of loose connective tissue predominantly within alveoli, alveolar ducts ± respiratory bronchioles. Interstitial inflammation is typically mild in COP. Secondary organising pneumonia can be associated with ancillary findings





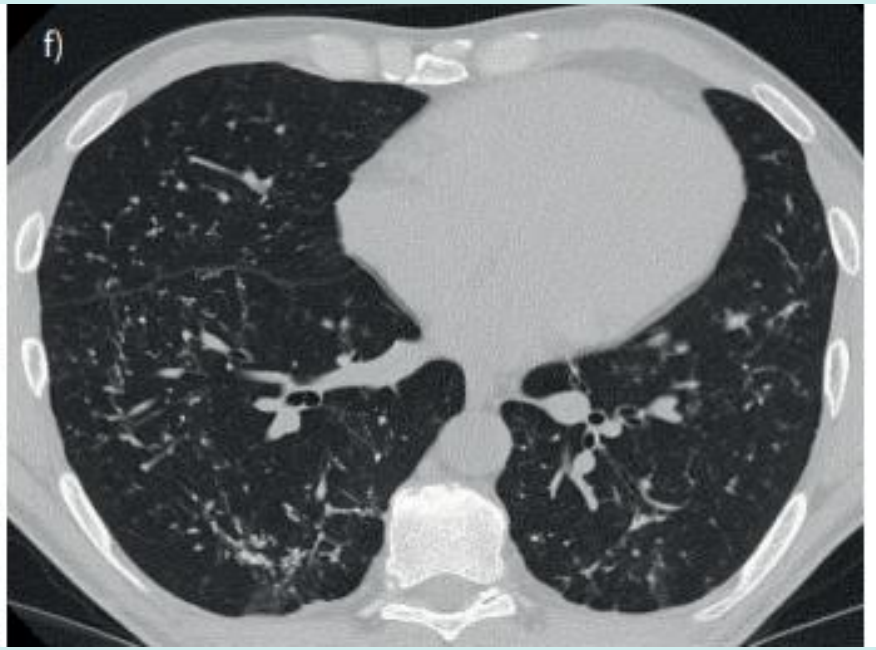


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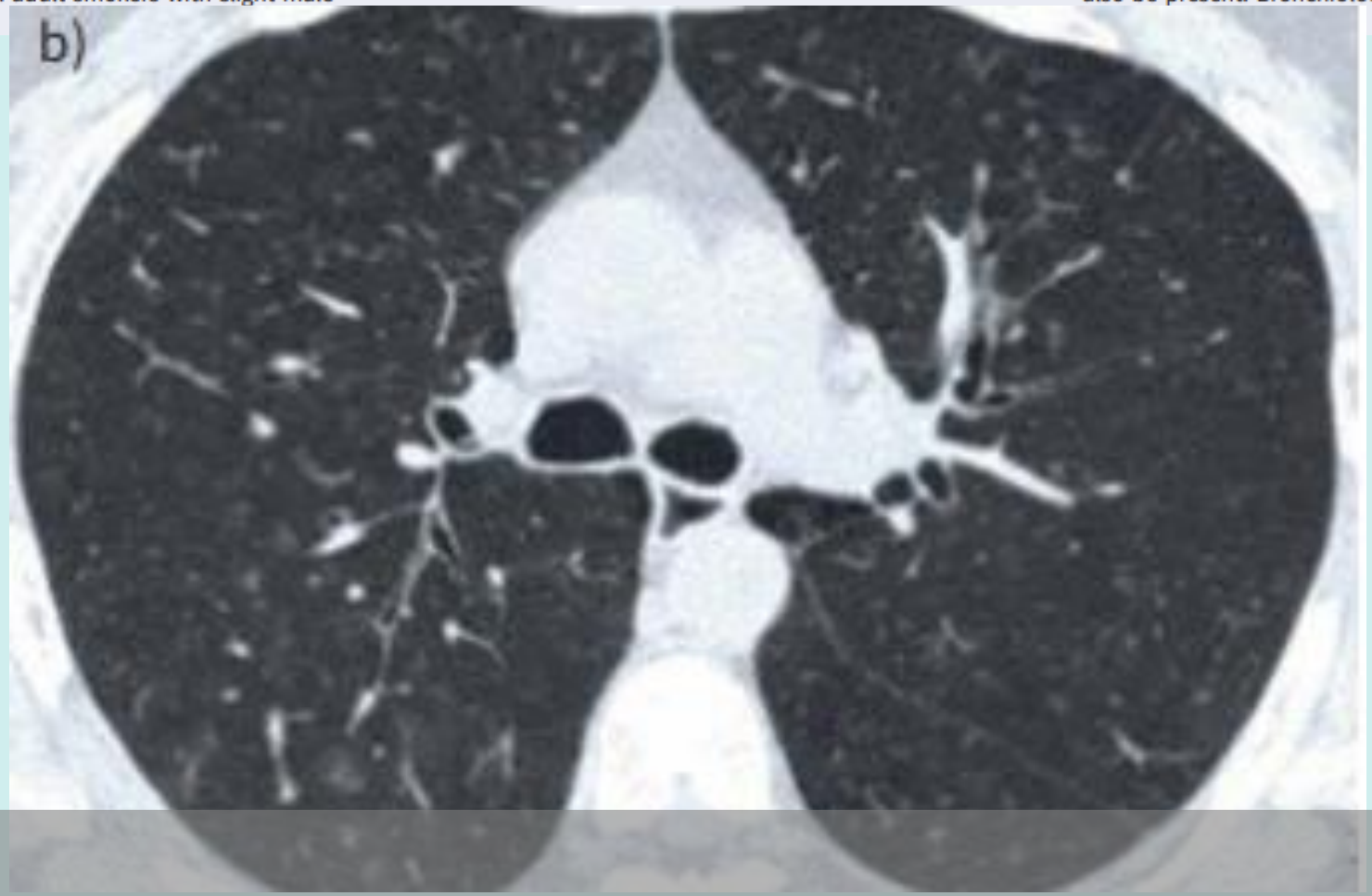
TABLE 2 Continued

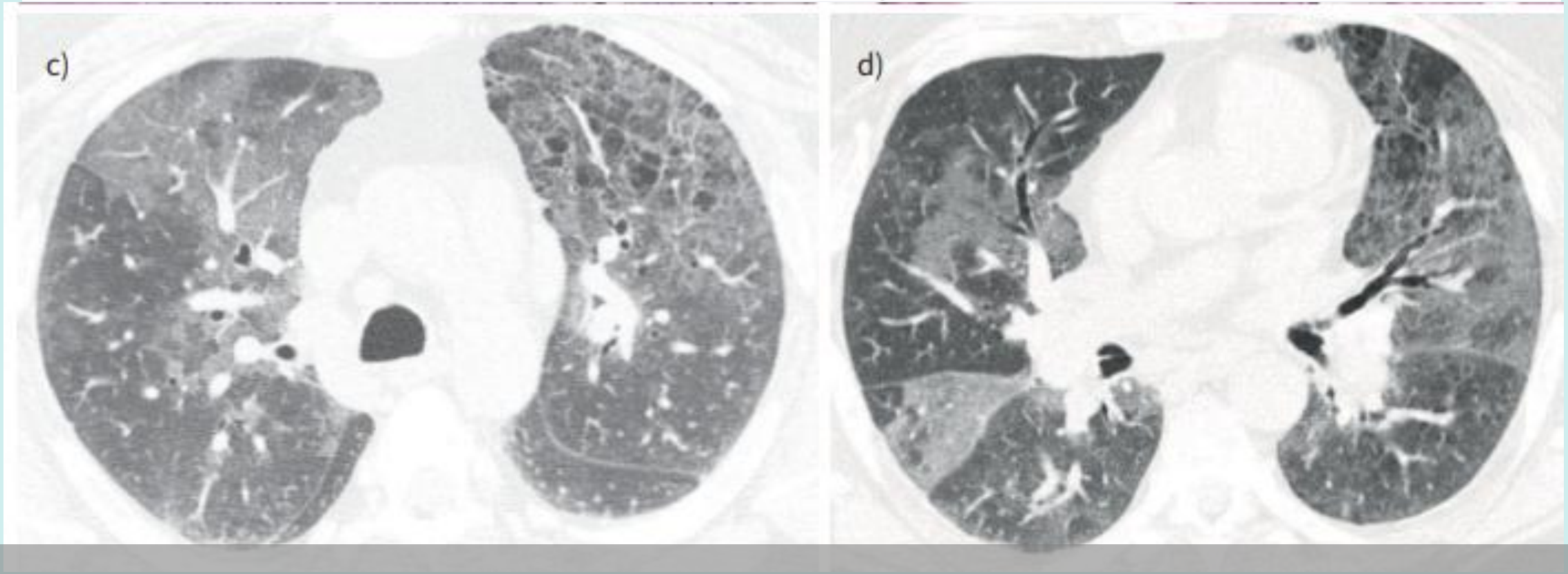
	Clinical features	Radiological features	Pathological features
Alveolar filling patterns			
RB-ILD	Presentation: subacute onset with potential for improvement or resolution with smoking cessation Risk factors: adult smokers with slight male	Upper-lobe-predominant centrilobular ground-glass nodularity	Accumulation of lightly pigmented macrophages in airspaces associated with a mild chronic inflammatory cell infiltrate. Lymphoid aggregates and fibrosis may also be present. Bronchiolocentric distribution



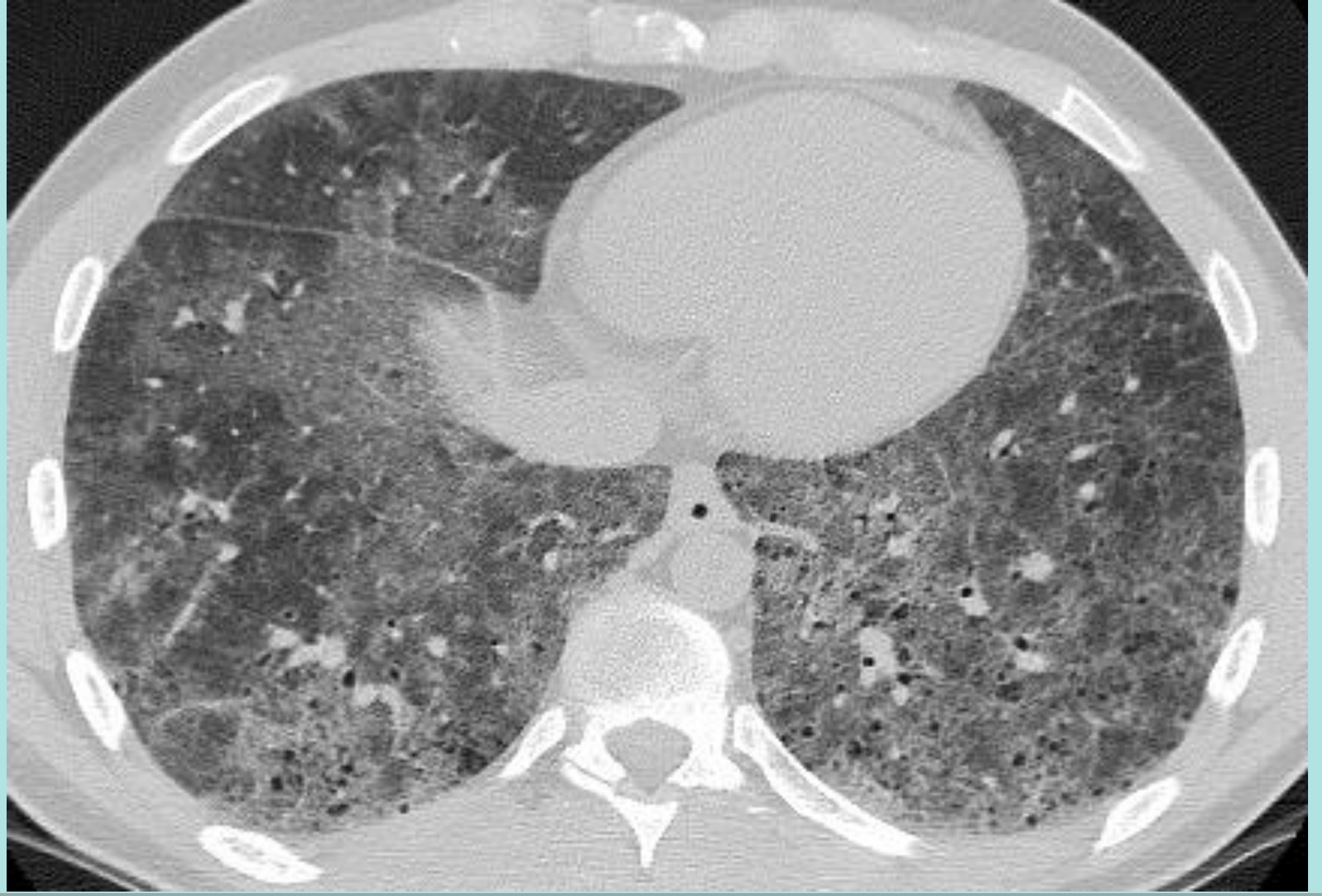
RB-ILD = Respiratory Bronchiolitis-ILD

TABLE 2 Continued

	Clinical features	Radiological features	Pathological features
Alveolar filling patterns			
	predominance		
AMP	Presentation: highly variable onset, progression in a subset Risk factors: adult smokers with slight male predominance, although rare cases have been reported in nonsmokers	Middle- to lower-lung-predominant with variable upper lung involvement, although may be predominantly peripheral. Primarily patchy and confluent ground-glass opacities with smooth reticulation. Cystic spaces may be present within the ground-glass; emphysema, and traction bronchiectasis may also be seen	Accumulation of lightly pigmented macrophages in airspaces associated with a mild chronic inflammatory cell infiltrate. Lymphoid aggregates and fibrosis may also be present. Diffuse distribution



DIP = Alveolar Macrophage Pneumonia (AMP)



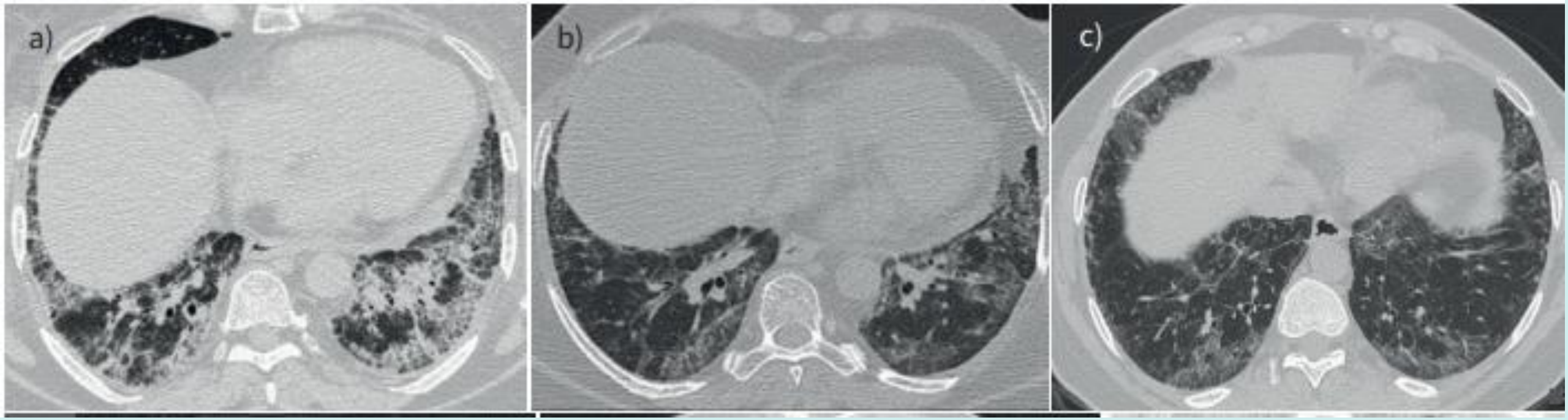
Courtesy of Dr Sara Piciucci

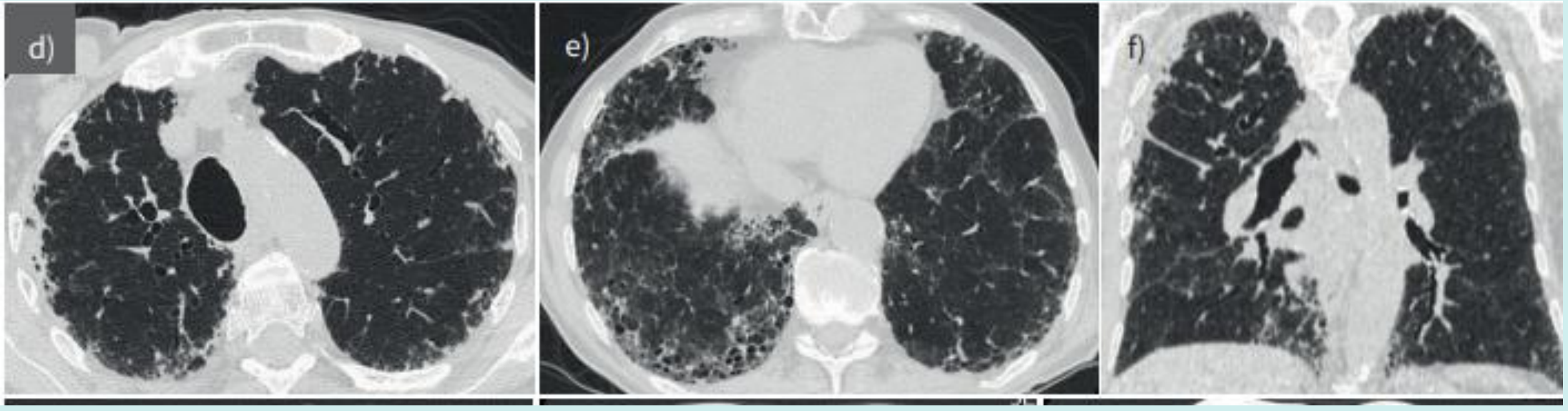
DIP = Alveolar Macrophage Pneumonia (AMP)

TABLE 2 Continued

	Clinical features	Radiological features	Pathological features
Alveolar filling patterns			
Organising pneumonia	Presentation: acute or subacute onset of respiratory ± constitutional symptoms. Typically reversible, but potential for residual fibrosis; low potential for ongoing progression for COP, with higher potential for recurrence or progression for some causes, particularly CTD Risk factors: no clear age or sex predilection for COP. Risk factors for secondary organising pneumonia vary with underlying aetiology	Patchy consolidation with air bronchograms, patchy ground-glass opacities, focal nodules, and/or mass-like lesions, sometimes featuring a halo sign, reversed halo sign, perilobular pattern, or air-bronchograms	Intra-alveolar buds of loose connective tissue predominantly within alveoli, alveolar ducts ± respiratory bronchioles. Interstitial inflammation is typically mild in COP. Secondary organising pneumonia can be associated with ancillary findings
RB-ILD	Presentation: subacute onset with potential for improvement or resolution with smoking cessation Risk factors: adult smokers with slight male predominance	Upper-lobe-predominant centrilobular ground-glass nodularity	Accumulation of lightly pigmented macrophages in airspaces associated with a mild chronic inflammatory cell infiltrate. Lymphoid aggregates and fibrosis may also be present. Bronchiolocentric distribution
AMP	Presentation: highly variable onset, progression in a subset Risk factors: adult smokers with slight male predominance, although rare cases have been reported in nonsmokers	Middle- to lower-lung-predominant with variable upper lung involvement, although may be predominantly peripheral. Primarily patchy and confluent ground-glass opacities with smooth reticulation. Cystic spaces may be present within the ground-glass; emphysema, and traction bronchiectasis may also be seen	Accumulation of lightly pigmented macrophages in airspaces associated with a mild chronic inflammatory cell infiltrate. Lymphoid aggregates and fibrosis may also be present. Diffuse distribution
Rare AFPs (supplementary table S2)			

UIP: usual interstitial pneumonia; NSIP: nonspecific interstitial pneumonia; BIP: bronchiolocentric interstitial pneumonia; DAD: diffuse alveolar damage; PPFE: pleuroparenchymal fibroelastosis; LIP: lymphoid interstitial pneumonia; RB-ILD: respiratory bronchiolitis-interstitial lung disease; AMP: alveolar macrophage pneumonia; AFPs alveolar filling pattern; IPF: idiopathic pulmonary fibrosis; CTD: connective tissue disease; HP: hypersensitivity pneumonitis; COP: cryptogenic organising pneumonia.


a)
b)
c)


d)
e)
f)

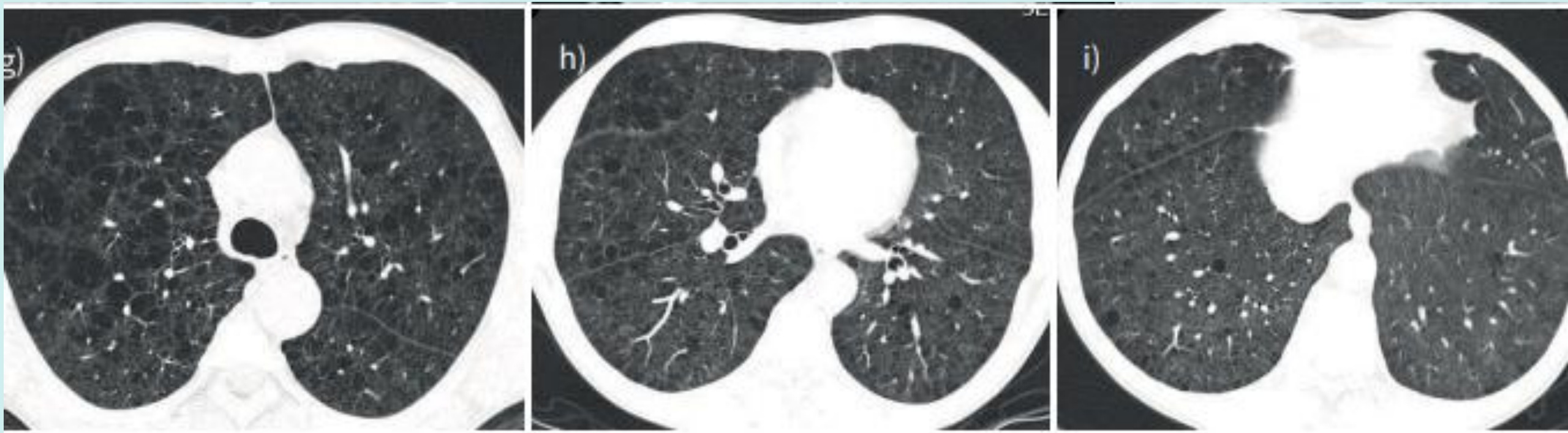
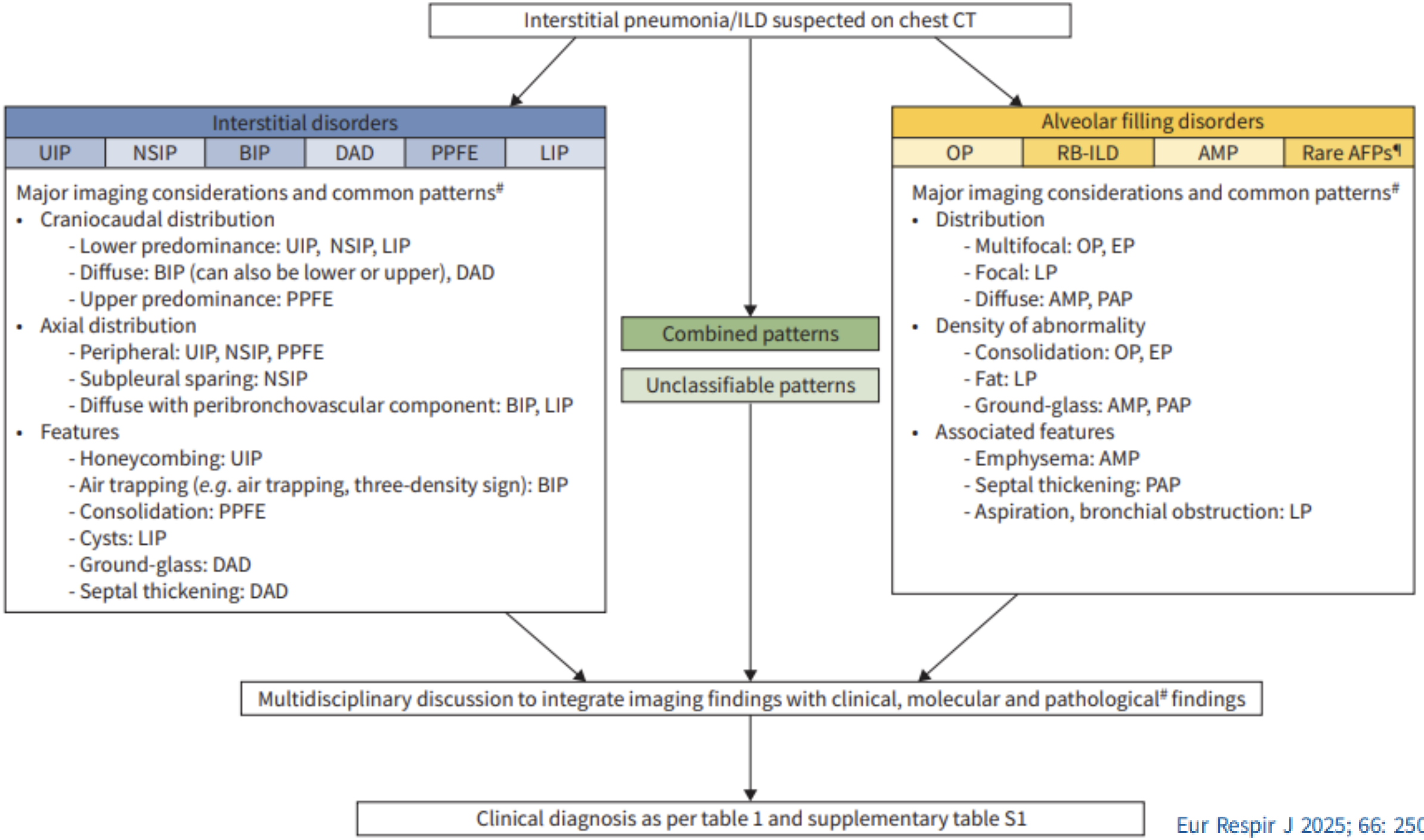


FIGURE 13 Radiological features of combined patterns. a–c) Axial computed tomography (CT) shows a combination of nonspecific interstitial pneumonia (NSIP) and organising pneumonia. a) Axial CT in a patient with antisynthetase syndrome at baseline shows features of both NSIP and organising pneumonia, with ground-glass attenuation and bilateral consolidation in a peribronchovascular and subpleural distribution. Serial CTs at b) 2 and c) 7 years following initiation of immunomodulatory treatment show reduced consolidation with residual ground-glass and mild fibrosis that is more typical of NSIP. d–f) Axial CT shows a combination of pleuroparenchymal fibroelastosis (PPFE) in the upper lobes and usual interstitial pneumonia (UIP) in the lower lobes. d) Axial CT of PPFE shows mild fibroelastosis in both upper lobes, characterised by dense pleural and subpleural fibrosis. e) The lower lobes show a UIP pattern, characterised by subpleural honeycombing and peripheral traction bronchiectasis, while f) the coronal view confirms the combination of patterns. g–i) Axial CT of hypersensitivity pneumonitis (HP) with emphysema in a never-smoker with a strong history of organic dust exposure and lymphocytosis on bronchoalveolar lavage showing both extensive emphysema and scattered thin-walled cysts with centrilobular ground-glass.



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Mass General Brigham

Update of the International Multidisciplinary Classification of the Interstitial Pneumonias

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Mass General Brigham
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