

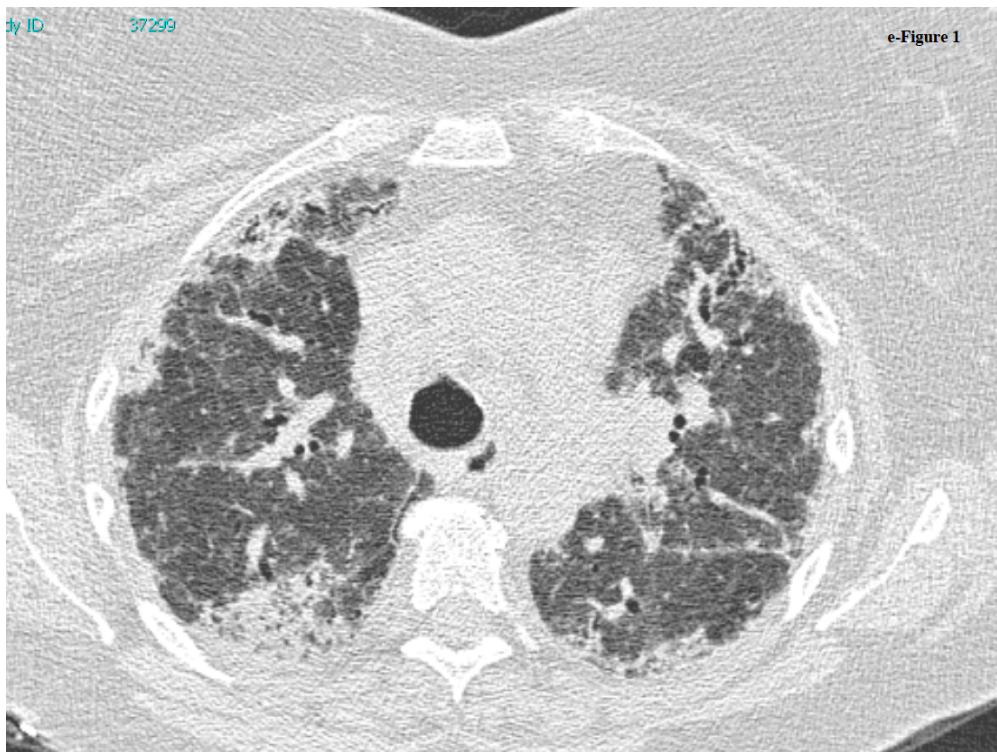
The Idiopathic Pulmonary Fibrosis Clinical Research Network (IPFnet)

Diagnostic and Adjudication Processes

Joao de Andrade, MD; Marvin Schwarz, MD; Harold R. Collard, MD; Tedryl Gentry-Bumpass, BSBE; Thomas Colby, MD; David Lynch, MD; and Robert J. Kaner, MD; for the IPFnet Investigators

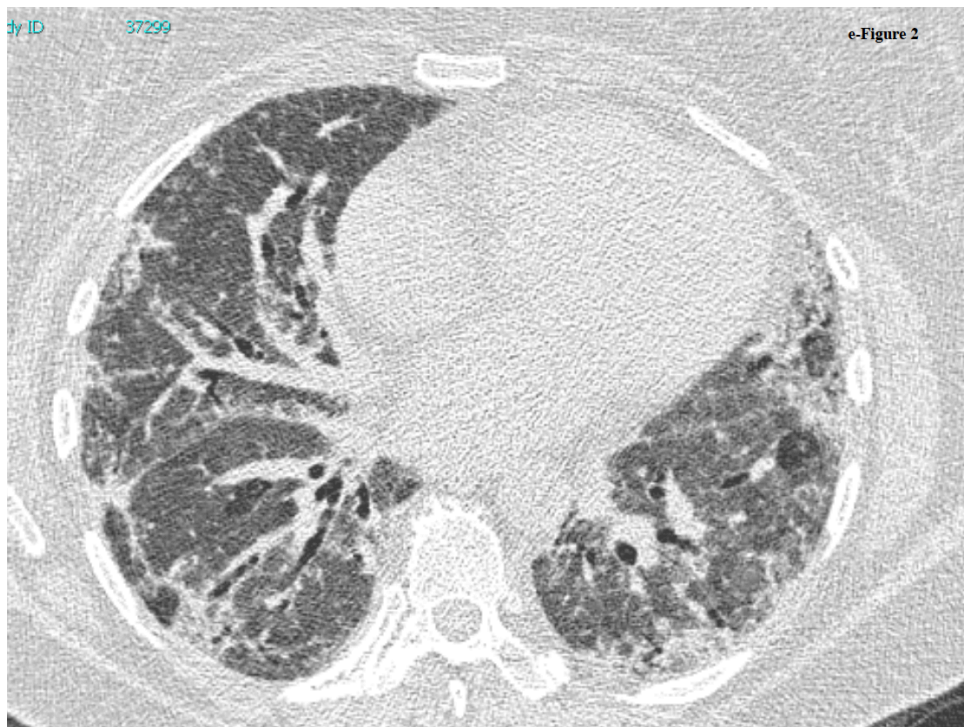
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e-Appendix 1. Chest CT Images



e-Figure 1.

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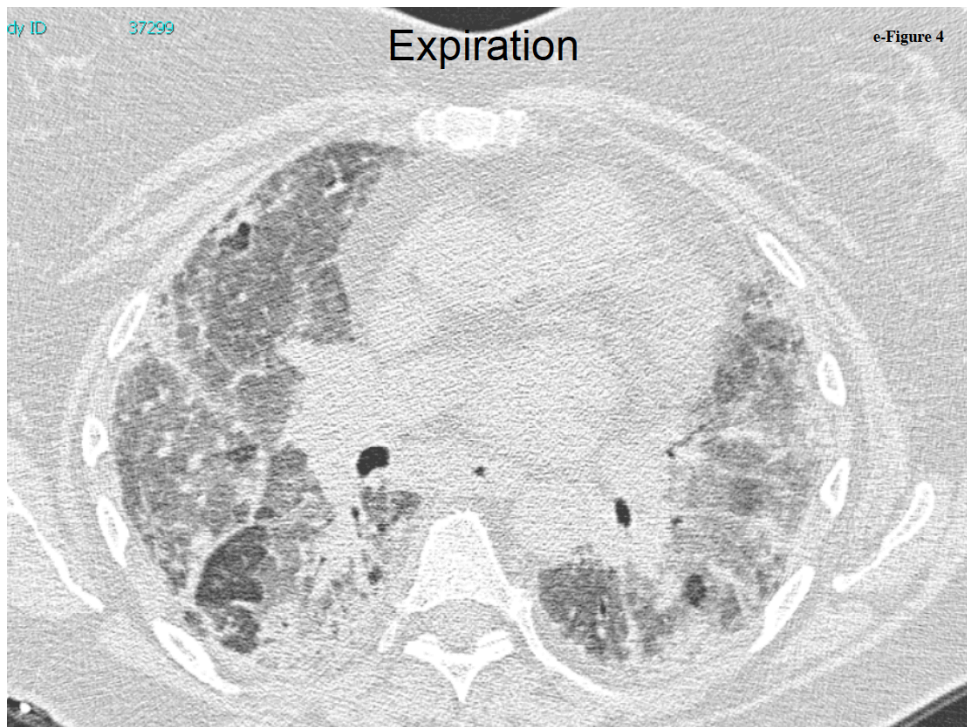


e-Figure 2.

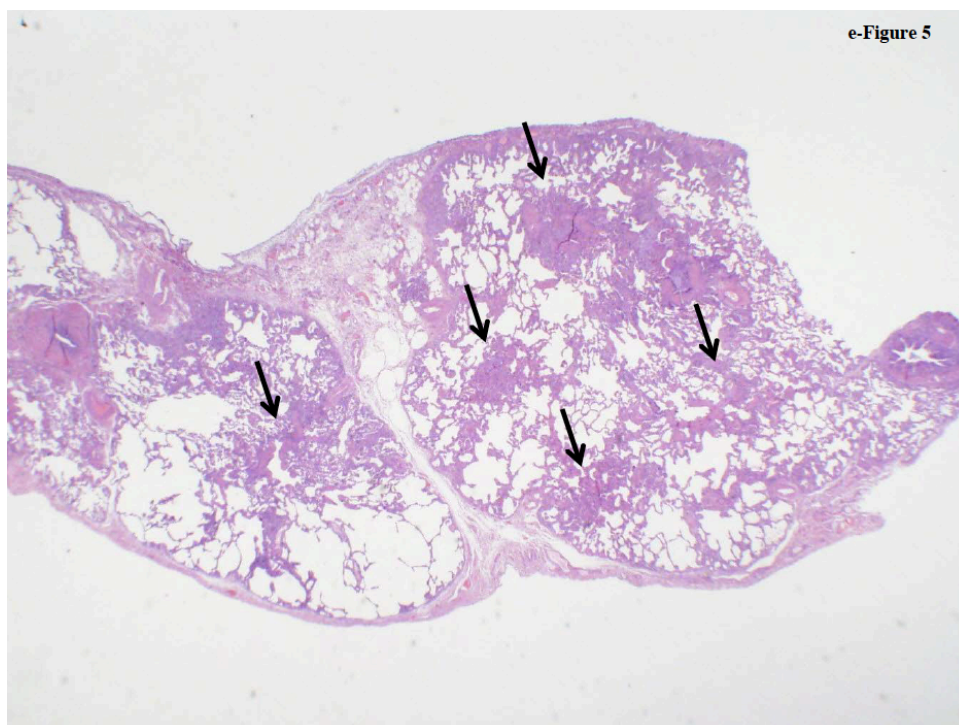


e-Figure 3.

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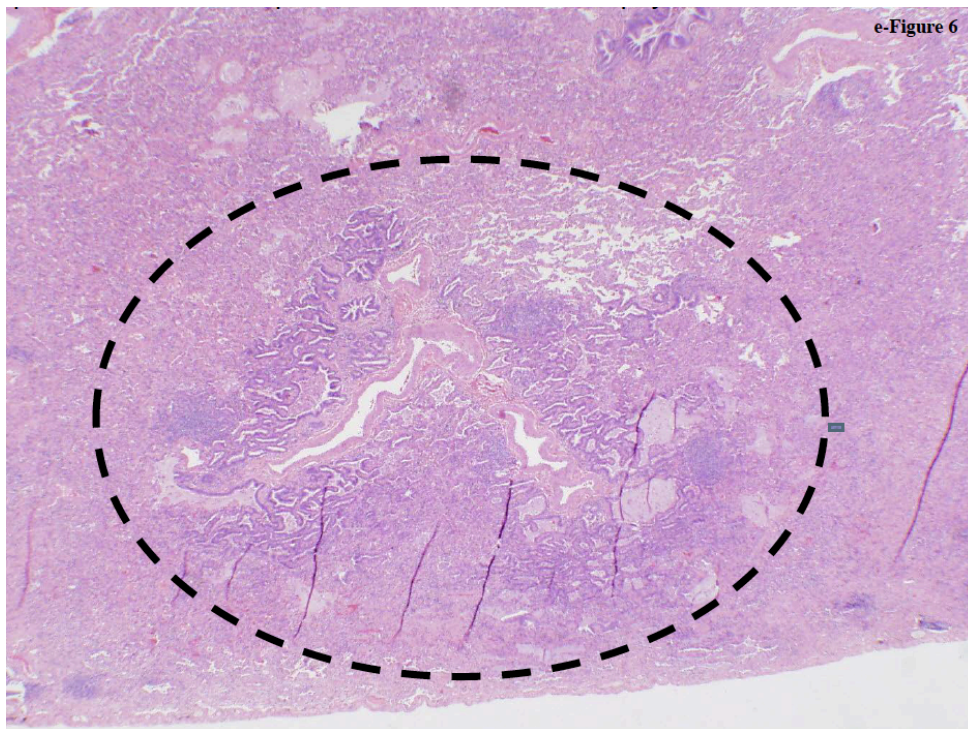


e-Figure 4.

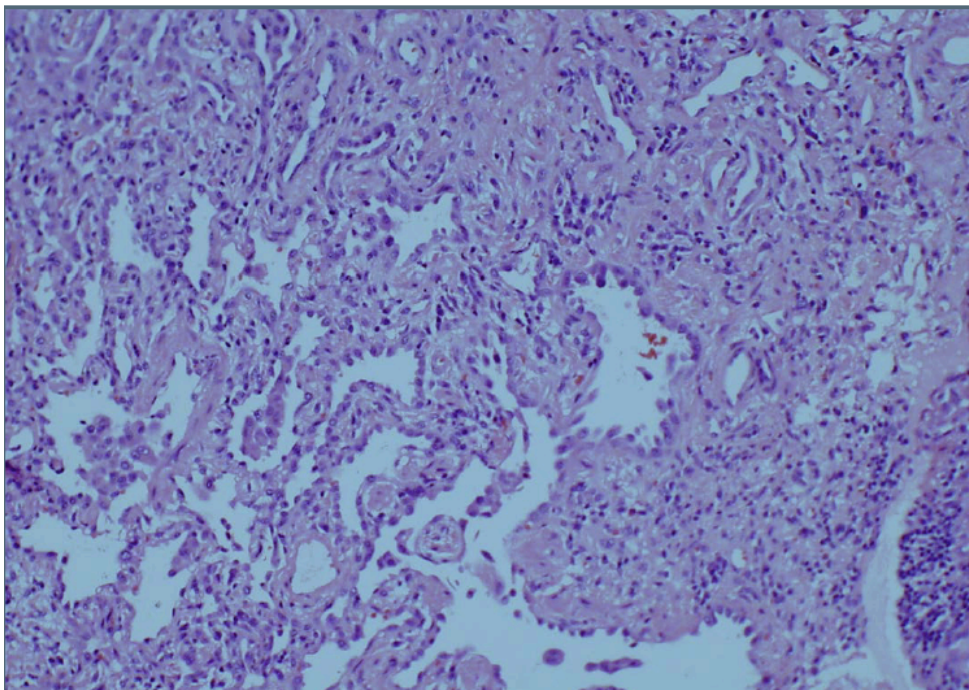


e-Figure 5. Scanning power microscopy showing centrilobular inflammation and scarring in one of the lobar biopsies (arrows)

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e-Figure 6. Scanning power microscopy showing centrilobular scarring with peribronchiolar metaplasia in the other lobar biopsies



e-Figure 7. Medium power microscopy shows interstitial fibrosis; no granulomas are present

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e-Appendix 2. Adjudication Case

The subject is a 56 year old Caucasian woman presenting with 3 years of progressive dyspnea on exertion. Two years prior, she underwent a surgical lung biopsy that was reported to be consistent with usual interstitial pneumonia. She had a history of essential hypertension and gastroesophageal reflux. She was a never smoker, and had one cat and one lovebird (a small parrot) for approximately 7 years. Her physical examination revealed inspiratory dry crackles at both lung bases. She had severe impairment of lung function with FVC 34% of predicted and DLCO was 34% of predicted.

HRCT Images (Appendix e-1, panels 1-4): diffuse fibrotic changes with evidence of mosaic attenuation and subsegmental air trapping in the expiratory views.

Surgical lung biopsy (Appendix e-1, panels 5-7): patchy areas of well established fibrosis that were predominantly centrilobular and peribronchiolar in distribution.

Adjudication: the combination of indoor exposure to a domestic bird, air trapping in the HRCT images, and biopsy findings with pathological changes predominantly centered around airways is most consistent with a final diagnosis of Chronic Hypersensitivity Pneumonitis.

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e-Appendix 3. ACKNOWLEDGEMENTS

The following IPFnet members participated in the **STEP-IPF study**:

Protocol Chairs—*National Jewish Medical and Research Center*: M. Schwarz; *Sansum Clinic, Santa Barbara*: D.A. Zisman. **IPFnet Steering Committee Chair**—*University of Iowa*: G. Hunninghake. **Clinical Centers**—*Cleveland Clinic*: J. Chapman, M. Olman, S. Lubell; *Duke University Medical Center*: L.D. Morrison, M.P. Steele, T. Haram; *Emory University*: J. Roman, R. Perez, T. Perez; *Mayo Clinic, Rochester*: J.H. Ryu, J.P. Utz, A.H. Limper, C.E. Daniels, K. Meiras, S. Walsh. *National Jewish Health*: K.K. Brown, M. Schwarz, MD, C. Bair, D. Kervitsky; *Tulane University*: J.A. Lasky, S. Ditta. *University of Alabama, Birmingham*: J. de Andrade, V.J. Thannickal, M. Stewart; *University of California, Los Angeles*: D.A. Zisman, J. Lynch, E. Calahan, P. Lopez; *University of California, San Francisco*: T.E. King, Jr., H.R. Collard, J.A. Golden, P.J. Wolters, R. Jeffrey; *University of Chicago*: I. Noth, D.K. Hogarth, N. Sandbo, M.E. Streck, S.R. White, C. Brown, I. Garic, S. Maleckar; *University of Michigan*: F.J. Martinez, K.R. Flaherty, M. Han, B. Moore, G.B. Toews, D. Dahlgren; *University of Washington*: G. Raghu, J. Hayes, M. Snyder; *Vanderbilt University*: J.E. Loyd, L. Lancaster, W. Lawson, R. Greer, W. Mason; *Weill Medical College of Cornell University*: R.J. Kaner, V. Monroy, M. Wang. **Core Lab Chairs**—**Radiology**: *National Jewish Health*: D.A. Lynch; **Pathology**: *Mayo Clinic*: T. Colby. **Data Coordinating Center**—*Duke Clinical Research Institute*: K.J. Anstrom, R.C. Becker, E.L. Eisenstein, N.R. MacIntyre, L.D. Morrison, J. Rochon, M.P. Steele, J.S. Sundry, L. Davidson-Ray, P. Dignacco, R. Edwards, R. Anderson, R. Beci, S. Calvert, K. Cain, T. Gentry-Bumpass, D. Hill, M. Ingham, E. Kagan, J. Kaur, C. Matti, J. McClelland, A. Meredith, T. Nguyen, J. Pesarchick, R.S. Roberts, W. Tate, T. Thomas, J. Walker, D. Whelan, J. Winsor, Q. Yang, E. Yow. **NHLBI**—H.Y. Reynolds, X. Tian, J. Kiley.

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