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Pulmonary Fibrosis Identification:
Lessons for Optimizing Treatment

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NOVEMBER 2014

In this Issue

US FDA Approves Two Treatment Options for Patients with IPF

"The FDA approval of two new drugs for the treatment of IPF is a game-changer in many respects. For patients, it represents an opportunity for medical therapy where previously there was none. For physicians, the stakes are raised in terms of making an early and accurate diagnosis of IPF. Ongoing education and disease awareness activities become increasingly important in the context of a treatable disease. Patient expectations will need to be managed, as neither of these drugs is a cure. However, their availability represents the beginning of a new era in IPF management. Hopefully other therapies will soon emerge that will further modify the natural history of this devastating disease."

Dr. Steven D. Nathan
Inova Fairfax Hospital

Publication Review

The Role of Bacteria in the Pathogenesis and Progression of Idiopathic Pulmonary Fibrosis

Molyneaux PL, Cox MJ, Willis-Owen SA, Mallia P, Russell KE, Russell AM, Murphy E, Johnston SL, Schwartz DA, Wells AU, Cookson WO, Maher TM, Moffatt MF. The Role of Bacteria in the Pathogenesis and Progression of Idiopathic Pulmonary Fibrosis. *Am J Respir Crit Care Med*. 2014 Sep 3. [Epub ahead of print]

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Summary

This study investigated the prevalence and burden of bacteria in bronchoalveolar lavage (BAL) fluid from patients with IPF, COPD, and healthy controls. The various types and abundance of bacteria were determined by quantitative PCR and pyrosequencing of DNA coding for bacterial 16S rRNA. Sixty five patients with IPF had double the burden of bacteria in BAL fluid compared to 44 controls. Haemophilus, Streptococcus, Neisseria and Veillonella were more evident in IPF cases than controls, although no association was observed between specific bacteria species and disease progression. Baseline bacterial burden predicted the rate of decline in lung volume, risk of death, and was independently associated with the rs35705950 polymorphism of the MUC5B mucin gene, a susceptibility factor for IPF. Whether or not these bacteria are pathogenic and somehow fuel the disease process remains uncertain, nonetheless this study provides a further basis for trials of antimicrobial therapy in IPF.

Listen to
Dr. Nathan's commentary

Steven D. Nathan, MD
Inova Fairfax Hospital

Publication Review

Increasing Global Mortality from Idiopathic Pulmonary Fibrosis in the 21st Century

Hutchinson JP, McKeever TM, Fogarty AW, Navaratnam V, Hubbard RB. Increasing Global Mortality from Idiopathic Pulmonary Fibrosis in the 21st Century. *Ann Am Thorac Soc*. 2014 Aug 28. [Epub ahead of print]

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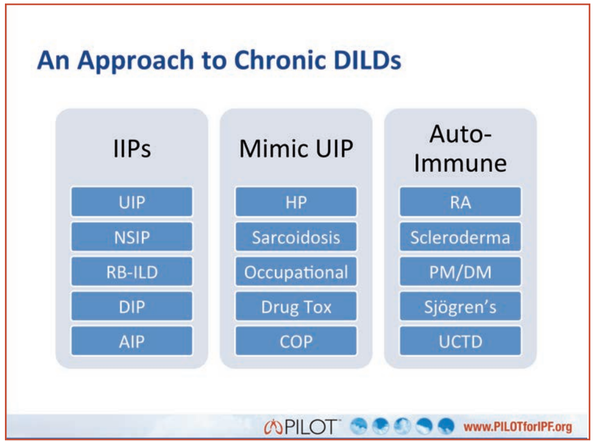
Summary

This research examined global trends in mortality from IPF by collating death certification data from ten countries from 1999 to 2012. Age-standardized mortality ranged between 4.68 per 100,000 population in Sweden to 9.84 per 100,000 in England and Wales, 10.71 per 100,000 in Scotland, and 10.26 per 100,000 in Japan. The variations between countries are less than previously reported, although there was an overall 2–3% annual increase in mortality. All countries had positive associations with male sex and increasing age. The authors project that in 2014 there will be 28,000–65,000 deaths in Europe and 13,000–17,000 deaths in the USA from IPF clinical syndrome. One finding that appeared consistent between all the countries studied was that IPF is an increasingly noted cause of mortality.

Listen to
Dr. Nathan's commentary

Steven D. Nathan, MD
Inova Fairfax Hospital

Diagnostic Dilemmas



Diagnosis is critical in interstitial lung disease, both to select an effective therapeutic approach and to avoid treatment that could be ineffective or harmful. Recent positive clinical results for two emerging drug candidates for IPF underline the importance of accurate diagnosis. Dr. Noth outlines a systematic practical approach to diagnosing idiopathic interstitial pneumonias.

Case Review

Diagnostic Dilemmas in Evaluating Clinical ILD – Imre Noth, MD

Watch Dr. Noth's systematic approach to diagnosing a challenging case.

Imre Noth, MD
The University of Chicago Medicine

PILOTforIPF.org Updates

PILOTforIPF.org supports the global educational initiative with no-cost CME education and resources. New content is always being developed by IPF experts. Recent updates include:

New Slides Available!

"A New Era in IPF: Pathophysiology and Recent Clinical Trials" (pptx) >

"Highlights from ATS 2014" Conference Coverage Series:

- **Update on Predicting Clinical Progression >**
- **Top Line Clinical Trial Results – ASCEND, INPULSIS, PANTHER >**
- **Discussions on Clinical Trial Data >**

A Practical Guide to Idiopathic Pulmonary Fibrosis Monograph

Download PDF >

IPF Trivia

1. Who first described IPF as a distinct form of interstitial lung disease?

a. Anton van Leeuwenhoek (1717)
b. Ludwig von Buhl (1872)
c. Louis Hamman and Arnold Rich (1933)
d. Luca Richeldi (1947)

2. Which description is used for a characteristic radiologic feature of IPF?

a. Honeycombing
b. Hounds tooth reticulation
c. Fractal proliferation
d. Ground glass opacity

3. What location of honeycombing is characteristic of UIP/IPF?

a. Peribronchiolar
b. Subpleural
c. Upper lobe
d. Pericarinal

4. Familial links are observed in approximately 5% of IPF cases. This finding was elucidated in the 1950's; but which researcher published the first description of familial IPF in 1907?

a. Bayer
b. Sandoz
c. Merck
d. Glaxo

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Event Calendar

Upcoming Events:

Friday, November 7
Regional Summit (Yale School of Medicine)
Navigating the New Era of IPF:
Collaborative Patient Care
Westport, Connecticut
Register >

Wednesday, November 12
Hawaii ATS Chapter Lecture
A New Era in IPF
Harold R. Collard, MD
Honolulu, Hawaii
Register >

Friday, November 14
LUNG FORCE Expo
A New Era in IPF
Marilyn K. Glassberg, MD
Orlando, Florida
Register >

Saturday, November 15
Regional Summit (Vanderbilt University Medical Center)
Navigating the New Era of IPF: Collaborative Patient Care
Nashville, Tennessee
Register >

Interested in hosting a live meeting? The PILOT faculty will bring expertise in IPF diagnosis and treatment to your Grand Rounds series, State Chapter Meeting, or other local event. Request a live meeting in your area now!
Request a Meeting >

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