Idiopathic pulmonary fibrosis (IPF)

Is a devastating condition

- barely 1/2 of patients surviving the 3 years

Carries a prognosis

- worse than that of many cancers.

1. King TE, Pardo A, Selman M: Idiopathic pulmonary fibrosis. Lancet 2011, 378:1949-61.
2. Ganesh Raghu et al :An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline: Treatment of Idiopathic Pulmonary Fibrosis. Am J Respir Crit Care Med Vol 183. pp 788–824, 2011
3. Spagnolo et al : Idiopathic pulmonary fibrosis: diagnostic pitfalls and therapeutic challenges . Multidisciplinary Respiratory Medicine 2012, 7:42
4. Athol U. Wells, Ulrich Costabel et al: Challenges in IPF diagnosis, current management and future perspectives. Sarcoidosis ,vasculitis and diffuse lung diseases 2015; 32; Suppl. 1: 28-35
5. Ganesh Raghu et al :An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline: Treatment of Idiopathic Pulmonary Fibrosis An Update of the 2011 Clinical Practice Guideline , Am J Respir Crit Care Med Vol 192, Iss 2, pp e3–e19, Jul 15, 2015
6. Fidler, Lee et al: Diagnostic disparity of previous and revised American Thoracic Society guidelines for idiopathic pulmonary fibrosis/La disparité diagnostique entre les lignes directrices passées et révisées de l'American Thoracic Society à l'égard de la fibrose pulmonaire idiopathique .Canadian Respiratory Journal22.2 . 2015 : 86-90
7. https://www.blf.org.uk/Page/IPF-patient-charter

Suspicion of IPF
often months/years after the initial manifestations

The most common presenting symptoms

- dry cough
- exertional dyspnea

wrongly attributed to
smoking habits
or aging.

1. Flaherty KR et al: Idiopathic interstitial pneumonia: do community and academic physicians agree on diagnosis? Am J Respir Crit Care Med 2007, 175:1054–1060.
2. Thomeer M et al: Idiopathic Pulmonary Fibrosis International Group Exploring N-Acetylcysteine I Annual (IFIGENIA) study group: Multidisciplinary interobserver agreement in the diagnosis of idiopathic pulmonary fibrosis. Eur Respir J 2008, 31:585–591
3. Spagnolo et al : Idiopathic pulmonary fibrosis: diagnostic pitfalls and therapeutic challenges . Multidisciplinary Respiratory Medicine 2012, 7:42
4. Wells A. The revised ATS/ERS/JRS/ALAT diagnostic criteria for idiopathic pulmonary fibrosis (IPF) – practical implications. Respiratory Research 2013; 14 (Suppl 1): S2.
5. Athol U. Wells, Ulrich Costabel et al: Challenges in IPF diagnosis, current management and future perspectives. Sarcoidosis ,vasculitis and diffuse lung diseases 2015; 32; Suppl. 1: 28-35

IPF -Diagnostic challenges

Delay in diagnosis

Alternative diagnosis

IPF and other forms of ILD ?

Special conditions

Acute exacerbation of IPF

Unclassifiable ILD

Obtaining a surgical lung biopsy

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Voicu Tudorache

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RARE DISEASES

a challenging activity for physicians