CHEST

Focus on Idiopathic Pulmonary Fibrosis

Advancing Approaches to Diagnosis, Prognosis, and Treatment

Check for updates

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VIDEO

As seen in this CME online activity (available at http://courses.elseviercme.com/694), idiopathic pulmonary fibrosis (IPF) is a specific form of chronic, progressive, fibrotic lung disease of unknown cause that is associated with substantial health-care utilization and high rates of mortality. The clinical symptoms of IPF are nonspecific and overlap with many pulmonary and cardiac diseases making differential diagnosis challenging. The American Thoracic Society/European Respiratory Society/Japanese Respiratory Society/Latin American Thoracic Association (ATS/ERS/JRS/ALAT) guidelines strongly recommend a multidisciplinary approach to the diagnosis of interstitial lung diseases; however, there are several limitations to the feasibility of this approach in clinical practice. Although early diagnosis is relevant to any chronic, progressive, and irreversible disorder, it is crucially important that effective treatments for IPF are prescribed without delay. A window of opportunity may exist during which time treatment can have optimal outcomes. Evidence on the clinical management of IPF is rapidly evolving, and key updates were made to the most recent ATS/ERS/JRS/ALAT guidelines. The widely used combination of prednisone, azathioprine, and N-acetylcysteine has now been associated with increased risk of hospitalization and death compared with placebo in patients with IPF. These treatments and others for IPF have been mostly supportive, but recently pirfenidone and nintedanib have demonstrated efficacy in reducing functional decline and disease progression in IPF. A pooled analysis of three phase 3 studies of pirfenidone found a significant 48% reduction in all-cause mortality, and a pooled analysis of a phase 2 and two phase 3 studies of nintedanib found a significant 43% reduction in on-treatment mortality. As patient exposure to these two new drugs increases, data continue to emerge on how and when to use these medications and on how to manage their side effects. Finally, several medications targeting the fibrotic pathobiology of IPF are currently in development. Given the limited treatment options for IPF, enrollment in a clinical trial may be the best chance to delay or prevent progression of IPF. This CME-certified expert video roundtable from *CHEST* reviews the ATS/ERS/JRS/ALAT guidelines with a specific focus on accurate and timely diagnosis of IPF and the latest data on the treatment of IPF. CHEST 2018; 154(4):978-979

Editor's Note: This is one in a series of Elsevier and CHEST sponsored activities with CME. The readers can view the video and claim CME.

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interest relating to the topics of this educational activity.

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Target Audience

This activity has been designed to meet the educational needs of health-care professionals involved in the diagnosis, treatment, or management of patients with IPF.

Educational Objectives

On completion of this activity, participants will be better able to do the following:

- Implement multidisciplinary clinical and imaging approaches to achieve a more timely and accurate diagnosis of IPF
- Develop IPF treatment strategies based on key guideline recommendations and examine the supporting clinical trial data
- Describe key elements of an individualized approach to IPF treatment that involves shared decision-making

CME Credit (Physicians)

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