

Acute Exacerbation in Idiopathic Pulmonary Fibrosis

- Although idiopathic pulmonary fibrosis (IPF) is a chronic progressive disorder abrupt worsening can occur during the clinical course. This has been termed “acute exacerbation” of IPF. Occasionally, acute exacerbation may be the presenting feature of IPF.
- Acute exacerbation in IPF is defined by worsening of dyspnea within days to weeks (generally <30 d) accompanied by evidence of worsening gas exchange and new radiologic opacities in the absence of an identifiable cause including infection, left heart failure, and pulmonary embolism.
- The incidence of acute exacerbation in IPF is estimated to be in the range of 5% to 19% of patients per year.
- Acute exacerbation is the cause of death in 30% to 50% of patients with IPF.
- The cause of this sudden worsening is not known but the lung biopsy usually reveals diffuse alveolar damage (DAD) superimposed on usual interstitial pneumonia (UIP).
- Radiologic findings include multifocal or diffuse ground-glass opacities and consolidation superimposed on underlying reticular opacities and honeycombing.
- Acute exacerbation of IPF is associated with a high mortality rate (80% to 90%). Optimal clinical management of this phenomenon remains to be defined.

References

1. American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias. *Am J Respir Crit Care Med* 2002;165:277-304.
2. Kondoh Y, Taniguchi H, Kawabata Y, et al. Acute exacerbation in idiopathic pulmonary fibrosis: analysis of clinical and pathologic findings in three cases. *Chest* 1993;103:1808-1812.
3. Collard HR, Moore BB, Flaherty KR, et al. Acute exacerbations of idiopathic pulmonary fibrosis. *Am J Respir Crit Care Med* 2007;176:636-643.
4. Hyzy R, Huang S, Myers J, et al. Acute exacerbation of idiopathic pulmonary fibrosis. *Chest* 2007;132:1652-1658.
5. Martinez FJ, Safrin S, Weycker D, et al. The clinical course of patients with idiopathic pulmonary fibrosis. *Ann Intern Med* 2005;142:963-967.
6. Parambil JG, Myers JL, Ryu JH. Acute exacerbation as presenting or complicating feature of idiopathic pulmonary fibrosis. *Chest* 2006;130:553-558.
7. Daniels CE, Yi ES, Ryu JH. Causes of mortality and associated findings in patients with idiopathic pulmonary fibrosis: study of 42 consecutive autopsies. *Eur Respir J* 2008;32:170-174.
8. Akira M, Kouka T, Yamamoto S, Sakatani M. Computed Tomography Findings in Acute Exacerbation of Idiopathic Pulmonary Fibrosis. *Am J Respir Crit Care Med* 2008; 178:372-378.