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Acute exacerbation of idiopathic pulmonary fibrosis: international survey and call for harmonisation

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Lack of focussed international guidelines for management of acute exacerbation of IPF results in global variability in prevention, diagnosis and treatment strategies. Global trials are urgently needed

to inform international specific guidelines for AE-IPF. http://bit.ly/3a8FB5i

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ABSTRACT Acute exacerbation of idiopathic pulmonary fibrosis (AE-IPF) is an often deadly complication of IPF. No focussed international guidelines for the management of AE-IPF exist. The aim of this international survey was to assess the global variability in prevention, diagnostic and treatment strategies for AE-IPF.

Pulmonologists with ILD expertise were invited to participate in a survey designed by an international expert panel.

509 pulmonologists from 66 countries responded. Significant geographical variability in approaches to manage AE-IPF was found. Common preventive measures included antifibrotic drugs and vaccination. Diagnostic differences were most pronounced regarding use of Krebs von den Lungen-6 and viral testing, while high-resolution computed tomography, brain natriuretic peptide and D-dimer are generally applied. High-dose steroids are widely administered (94%); the use of other immunosuppressant and treatment strategies is highly variable. Very few (4%) responders never use immunosuppression. Antifibrotic treatments are initiated during AE-IPF by 67%. Invasive ventilation or extracorporeal membrane oxygenation are mainly used as a bridge to transplantation. Most physicians educate patients comprehensively on the severity of AE-IPF (82%) and consider palliative care (64%).

Approaches to the prevention, diagnosis and treatment of AE-IPF vary worldwide. Global trials and guidelines to improve the prognosis of AE-IPF are needed.