For the week of Idiopathic Pulmonary Fibrosis

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The management of patients with Idiopathic Pulmonary Fibrosis, the most frequent and devastating of all Idiopathic Interstitial Pneumonias, is facing the beginning of a new era. The old therapeutic regimen of treating IPF with corticosteroids and immunosuppressants has at last proved deleterious only for the patient and not at all effective against the disease. The same thing is now starting to become apparent for the use of steroids in the rapidly deteriorating patient with IPF who develops Acute Respiratory Distress Syndrome. We suggest that the so long awaited and absolutely necessary change in the therapeutic approach of IPF patients be based on a so called "strategy of small steps" incorporating the use of the recently approved anti-fibrotic medications expected to slow down the functional progression of the disease, early diagnosis, the systematic management of comorbidities, the prevention and timely treatment of infections as well as the avoidance of risk factors such as smoking, pneumotoxic drugs, deleterious environmental exposures and air pollution. Although all these measures together are not still able to save from the disease, they might improve survival and significantly ameliorate quality of life, a therapeutic target not negligible at all if we consider that in IPF we refer mostly to elderly adult patients.

Taking into consideration all the above as well as the high cost of new treatments, management of IPF patients should take place in Centers of Excellence for interstitial lung diseases under the supervision of specially trained and experienced pulmonary medicine physicians. Finally, thoughtful selection of candidates for lung transplantation and the systematic application of a program of lung transplantation in Greece could represent the ultimate "small", big step that we still owe to our IPF patients.

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