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CURRENT AND NOVEL THERAPIES FOR IDIOPATHIC PULMONARY FIBROSIS

Debreslioska A., Jovskovska Kaeva B.; Stojkovic J., Arsovski Z., Angelovska I.

University clinic of pulmoallergology, Skopje

Idiopathic pulmonary fibrosis (IPF) is increasingly recognised as a well-known clinical and highly complex entity associated with poor prognosis and a median survival of 3–5 yrs after the diagnosis is made. The first guideline on the clinical management of IPF was published in 2000, and identified clinical awareness of IPF as a distinct entity, provided clinical criteria for diagnosis of IPF, and suggested recommendations for treatment. Although it was based on the consensus of the opinions of a few experts, as the evidence available at that time was minimal, the statement was very useful to the pulmonary community at large as clinical studies emerged. This provided the official evidence based guidelines for diagnosis and management of IPF in 2011, and subsequent updates summarizing the new reported evidence on the subject.

The diagnosis of IPF requires the following:

1. Exclusion of other known causes of ILD (e.g., domestic and occupational environmental exposures, connective tissue disease, and drug toxicity).
2. The presence of a UIP pattern on HRCT in patients not subjected to surgical lung biopsy.
3. Specific combinations of HRCT and surgical lung biopsy pattern in patients subjected to surgical lung biopsy.

The pharmacologic therapy is proven benefit, with the following novel agents found to be effective:

- a. Nintedanib, a tyrosine kinase inhibitor that targets multiple tyrosine kinases
- b. Pirfenidone

There is a supporting data against the use of anticoagulation, combination prednisone, azathioprine and N- acetylcysteine, the selective tyrosine kinase inhibitor – Imatinib, and the

selective endothelin receptor antagonist (ambrisentan), colchicines, Cyclosporin A, interferon, etanercept. It is also not recommended to use acetylcysteine monotherapy. The use of corticosteroids as monotherapy is not recommended, but has been proven effective in some patients during acute exacerbations.

The antiacid therapy should be used only in selected patients, as the pulmonary hypertension should not be treated in majority of patients with IPF.

Oxygen therapy should be used if there is persistent hypoxemia, with mechanical

ventilation applied in selected patients. The pulmonary rehabilitation should be used in most of the patients with IPF. At the end, lung transplantation is strongly recommended in patients with IPF, but additional evidence should be evaluated to guide this clinical decision.

The treatment regimen should be a decision for each patient, taking into account individual clinical circumstances.