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PIRFENIDONE AND NINTEDANIB IN TREATMENT OF IDIOPATHIC PULMONARY FIBROSIS - OUR FIRST CLINICAL EXPERIENCES

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Objective: Objectives: Idiopathic pulmonary fibrosis (IPF) is a chronic progressive interstitial lung disease (ILD) of unknown aetiology with median survival rate of 3-4 years after diagnosis. Two new agents, nintedanib and pirfenidone decrease the rate of progression of IPF measured by longitudinal changes of FVC. Our aim was to analyze efficiacy and safety od pirfenidone and nintedanib in patients with idiopathic pulmonary fibrosis after 6 and 12 months of therapy.
Methods: We retrospectively collected data from our hospital Registry of rare lung diseases for patients who were treated with pirfenidone and nintedanib for IPF for 6 or 12 months. We collected demographic data, adverse events and lung function results before and after therapy.
Results: A total of 12 patients were included (10 males, 2 females) with medain age 72 (56±81). 8 (67%) patients were treated with pirfenidone and 4 (33%) with nintedanib. 7 (59%) were treated for more than 12 months, 4(33%) for 6 months and 1 (8%) patient was discontinued from therapy after 2 months for adverse effects. The average time from the beginning of the disease until therapy was started was 20 months (1±59). Median time of treatment course was 13.5 months (2±33). The average pre-treatment FVC was 69% predicted (45±93%) and DLCO 40% predicted (20±62%). The average FVC in patients after 6 months of therapy was 72% predicted

(54±80%) and DLCO 41% predicted(17±80). In patients who were treated for 12 months the average FVC was 68% predicted (50±88) and DLCO was 40% predicted (28±60). Two patients who were treated with pirfenidone expirienced adverse effects (nausea, reduced apetiteand tiredness) which led to discontinuation of treatment after 2 and 14 months, and 1 patient who was treated with nintedanib developed liver lession an the therapy was discontinued after 20 months of therapy. Only one patient had skin rash grade 1. Two patients had laug transplantation without any perioperative complications.
Conclusion: After 6 and 12 months all patients trated with pirfenidone or nintedanib had satble lung funcion with mild side effects in most of the patients. Our results are comparable with previously published data which showed that these two drugs are safe and effective in tretment of patients with IPF.