Title: Clinical experience with pirfenidone for the treatment of idiopathic pulmonary fibrosis

Body: Background: Idiopathic pulmonary fibrosis (IPF) is a progressive and fatal lung disease with an estimated median survival of only 3 years after diagnosis. Pirfenidone is the only medication approved in the European Union for the treatment of adults with mild to moderate IPF. Aim of the study: To report on safety and efficacy of pirfenidone in the treatment of patients with IPF in our centre. Patients and methods: From 2006 to 2012, 45 patients (28 inside clinical trials, 17 outside) with mild to moderate IPF were treated with pirfenidone. Clinical data, results of lung function tests, and radiological findings as well as data about side effects were collected routinely. Results: The mean duration of treatment per patient was 48 [range 3–321] weeks. 16 patients (35%) received pirfenidone as monotherapy and 29 (65%) in combination with corticosteroids and/or N-acetylcysteine (NAC). At the end of the follow-up period 28 of 40 patients (70%) with treatment duration > 3 months were in a stable condition. 26 patients (58%) suffered from side effects, mostly gastrointestinal (17 [38%]). Pirfenidone was discontinued by six patients (13%) because of side effects. The median survival after the start of pirfenidone was 3.8 years. Conclusion: Pirfenidon alone or in combination with corticosteroids and/or NAC was generally well tolerated. Severe side effects were rare. The course of the disease was stable during treatment with pirfenidone in two out of three patients. Our results are in line with the previous published safety and efficacy data on pirfenidone as treatment for IPF.