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Title: Pirfenidone in idiopathic pulmonary fibrosis (IPF): Early single centre Irish experience

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Body: Introduction Pirfenidone is an orally bio-available synthetic molecule, which has recently been approved for the treatment of mild to moderate IPF in Europe. It regulates the activity of TGF- β and TNF- α in vitro. Open label pirfenidone prescription commenced in June 2011 in the Republic of Ireland. We report the early experience of a single centre with pirfenidone. Methods We conducted a retrospective review of medical records of those patients who were prescribed pirfenidone. We analysed baseline demographics, symptoms and pulmonary function. Comparisons between groups were conducted using paired t-testing. Results 26 patients (20 male) received the medication. 22 patients remain on the medication. 1 patient died due to an exacerbation of IPF with 3 others discontinuing the medication secondary to side effects. 15 (58%) of patients have reached target dose of medication. 7 subjects continue to take pirfenidone at a reduced dose. 14 participants reported side effects potentially related to pirfenidone. The most commonly reported side effect was fatigue followed by gastro-intestinal disturbance and photo-sensitivity. An increase (< 2 fold) in transaminases was noted in 1 patient. Patients who experienced side effects or required dose reduction were on average older but this did not reach statistical significance. Amongst subjects who had repeated pulmonary function testing (n=14), there was no significant decline in TLCO or FVC between baseline and follow up. Conclusion Pirfenidone is a novel agent for the treatment of limited IPF. The side effect profile in an Irish population appears consistent with recent published data. Further follow up is required to establish efficacy in an Irish population.