

# Our Experience with Pirfenidone in Patients with Idiopathic Pulmonary Fibrosis

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Idiopathic pulmonary fibrosis (IPF) is a chronic, progressive, mortal disease of unknown cause that is characterized by an irreversible pulmonary function loss and progressive pulmonary fibrosis. There is no standard treatment for IPF. Pirfenidone, approved by the Food and Drug Administration for the treatment of IPF, is an oral drug having antifibrotic, anti-inflammatory, and antioxidant effects. Pirfenidone limits the decline in pulmonary function tests and disease progression in patients with extended fibrosis (1).

Eight patients diagnosed with IPF for whom pirfenidone was initiated between 2013 and 2015 were included in the study. The results of the patients' pulmonary function test and 6-min walk test (6MWT) before and 1 year after treatment were recorded. The quality of life was evaluated before and 1 year after treatment using the St. George's Respiratory Questionnaire (SGRQ). Descriptive statistical methods were used. The study was approved by the hospital's ethics committee.

Five male and three female patients were enrolled; the mean age was 58.25 years. Six patients had a 30 packs.year smoking history. Five patients with possible UIP pattern on radiological examination were diagnosed with IPF via an open lung biopsy; three were clinically and radiologically diagnosed. Pirfenidone treatment was initiated in patients diagnosed with IPF whose FVC was >50%, DLco was >30% in pulmonary function test, and walking distance was >150 m in 6MWT. For all patients, pirfenidone dosage was 3×1 tablets for the first week, 3×2 for the second week, and 3×3 tablets from the 15<sup>th</sup> day onward. For controls, the patients were examined in the first and second week and first, third, sixth, ninth, and twelfth months following treatment initiation. At treatment initiation and during follow-up, the patients' laboratory tests were normal. The average treatment duration was 12.8 (1–23) months. Mean FVC before and after treatment was 2.17 lt, 63% and 2.33 lt, 67.6% respectively. Mean DLco before and after treatment were 63.4% and 64.4%, respectively. When compared, a 33.2% increase was observed in 6MWT after treatment. During treatment, no exacerbations occurred. The total SGRQ scores before and after treatment were 55.4 and 32.4, respectively (Table 1).

Pirfenidone and nintedanib have been recently approved for the treatment of IPF after demonstrating an efficacy in slowing functional decline and disease progression (2). Pirfenidone demonstrates anti-fibrotic effect via cytokines and transforming growth factors. In the studies performed with pirfenidone, the drug decelerated the disease progression rate and the loss in pulmonary function tests (3–5). Exacerbations were observed less in the group using pirfenidone (3). The survival rate without progression was higher in the group using pirfenidone compared with that in the group using placebo (4, 5). Pirfenidone decelerated the decline in 6MWT (5). The most frequent side effects were skin-related events, gastrointestinal symptoms (nausea, vomiting, and dyspepsia), and increase in liver function tests (3–5). Serious side effects were observed in 1%–5% of patients (5). We observed deceleration in the loss in pulmonary function tests and improvement in 6MWT in our patients. There was a significant improvement in the quality of life according to SGRQ. No exac-



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**Table 1.** Our patients' age and gender and FVC, 6MWT, and SGRQ total score values before and after treatment

Case no	Age (year)/Sex	Before treatment					After treatment						
		FVC L	FVC %	6MWT (m)	SpO <sub>2</sub> %	SGRQ	FVC L	FVC %	6MWT (m)	SpO <sub>2</sub> %	SGRQ	6MWT differences	SGRQ differences
1	61/E	2.13	49	430	98-93	27.39	1.87	43	600	94-82	27.18	-170	0.21
2	53/E	2.82	64	520	97-93	49.25	2.77	63	480	98-94	23	40	2625
3	64/E	2.35	66	200	97-91	98.03	2.2	60					
4	68/K	1.97	72	400	96-92	40.3	2.02	91	450	95-90	39.3	50	1
5	73/E	2.92	75	340	95-85	13	2.91	88	420	90-82	70	80	-57
6	55/K	1.48	69	400	97-92	76.6							
7	51/E	2.16	59	440	94-94	64.2	2.23	61	660	96-90	2.82	220	61.38
8	41/K	1.54	51	390	95-82	74.7							

FVC: Forced vital capacity; SGRQ: St. George's Respiratory Questionnaire; approval forms were taken from the patients; SpO<sub>2</sub>: Oxygen saturation; 6MWT: 6-min walk test

erbatations occurred during the 1-year follow-up period. Dyspepsia was observed in five patients, whereas gastrointestinal bleeding was observed in one patient. Skin-related events were not noted in patients. We observed that pirfenidone decelerated disease progression, reduced exacerbations, and improved the quality of life. Furthermore, we can state that pirfenidone is more reliable than previously used drugs in terms of side effects.

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