Ann Med Res 2021;28(5):987-92

Annals of Medical Research

DOI: 10.5455/annalsmedres.2020.09.933

Original Article

# Patients with idiopathic pulmonary fibrosis referred for lung transplantation: Initial institutional experience in Turkey

Mustafa Vayvada, Ayse Nigar Halis, Pinar Atagun Guney, Sevinc Citak, Ali Yiginsu, Atakan Erkilic, Ahmet Erdal Tasci

Department of Transplant Surgery, Kartal Kosuyolu Training and Research Hospital, Istanbul, Turkey



**Abstract**

**Aim:** Lung transplantation is the only therapeutic option that can improve survival and quality of life for idiopathic pulmonary fibrosis (IPF) despite recent advances in medical treatment. This study aimed to analyze mortality during the waiting list in IPF patients listed for lung transplantation since the implementation of the lung transplant program.

**Materials and Methods:** Patients with IPF listed for lung transplantation were retrospectively analyzed between December 2016 and December 2018. Moreover, patients were excluded if they have been referred for lung transplantation and were not suitable after evaluation.

**Results:** A total of 28 patients with IPF were listed for lung transplantation. The waiting list mortality rate was 39.3% (11/28). The median days on the waiting list were 71.2 days (range, 3–206) in patients who underwent lung transplantation and 110 days (range, 14–303) in patients who died on the waiting list. No differences in the demographic and clinical data were observed between both groups. Arterial blood gas saturation was statistically significantly lower in patients who died on the waiting list (74.2% vs. 82.9%, p = 0.046). Two single and 15 bilateral lung transplants were performed. Hospital mortality rate was 23.5% (4/17). The 1- and 3-year survival rates were 70.6%.

**Conclusion:** Lung transplantation is the only treatment option for patients with IPF who are not responsive to medicinal treatment. The late referral may lead to mortality on the waiting list. Thus, patients newly diagnosed with IPF should be promptly referred to a transplant center for evaluation.

**Keywords:** Idiopathic pulmonary fibrosis; lung transplantation; waiting list mortality

## INTRODUCTION

Idiopathic pulmonary fibrosis (IPF), a chronic and progressive fibrosis lung disease of unknown etiology, is the most common form of idiopathic interstitial pneumonia (1). Lung transplantation is a well-accepted treatment option for selected patients with end-stage pulmonary fibrosis. According to the European IPF registry data, the median survival in patients with IPF was 123.1 and 68.3 months during antifibrotic therapy and treatment with other drugs, respectively (2). The antifibrotic drugs pirfenidone and nintedanib slow down the deterioration of lung function. However, these drugs cannot prevent end-stage respiratory failure, secondary pulmonary hypertension, and death in patients. Lung transplantation is the only treatment that has the potential to improve the quality of life and survival of patients when medical treatment is inadequate.

The International Society for Heart and Lung Transplantation recommends criteria for referral of patients with interstitial lung disease (3). Transplant clinics may be exposed to a large number of patients if each patient who meets these criteria is referred. This situation may adversely affect patients in poor condition. The development of important comorbidities such as pulmonary hypertension, coronary artery disease, gastroesophageal reflux, malignancy, and fragility may cause the patient to lose the chance of transplantation with a late referral. Therefore, the treating clinics must refer patients at the most optimal time.

Waiting list mortality is an important problem in end-stage pulmonary diseases, especially in patients with IPF (4). Although IPF is associated with decreased respiratory function over time, many patients may experience acute respiratory deterioration within days to weeks. The median

**Received:** 09.09.2020 **Accepted:** 01.12.2020 **Available online:** 21.05.2021
**Corresponding Author:** Mustafa Vayvada, Department of Transplant Surgery, Kartal Kosuyolu Training and Research Hospital, Istanbul, Turkey **E-mail:** mustafavayvada@gmail.com

survival time after acute exacerbation of IPF is 3 months (5). End-stage patients may not have enough time for lung transplantation mainly because of the lack of donors and late referral.

The current paper reports the initial experience of consecutive patients with IPF listed for lung transplantation. Furthermore, this study aimed to analyze the characteristics of patients with waiting list mortality.

## MATERIALS and METHODS

Twenty-eight patients with IPF listed for lung transplantation were retrospectively evaluated between December 2016 and December 2018. Patients with IPF were determined according to the IPF diagnostic criteria as recommended by the official American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and the Latin American Thoracic Society clinical practice guideline (1).

Patients with IPF were listed for lung transplantation according to the consensus document for the selection of lung transplant candidates from the Pulmonary Transplantation Council of the International Society for Heart and Lung Transplantation (3). Not only patients on the waiting list but also those with acute deterioration at an external center were taken into consideration for patient selection. Age, comorbidities, absence of multi-organ failure, adaptation to post-transplant rehabilitation, neurological status, and absence of active bacteremia were the factors taken into account for patient selection. Patients with IPF were not listed if they have contraindications for lung transplantation (e.g., untreatable coronary artery disease, obesity (BMI >30 kg/m$^2$), severely altered functional status with an inability to rehabilitate, severe psychiatric disorders, and poor social support). These patients were excluded from the study.

Patients with IPF were divided into two groups. Group A (n = 17) were patients who underwent lung transplantation, and group B (n = 11) were patients who died on the waiting list. Data were collected during the evaluation process for lung transplantation. Arterial blood gas, respiratory function test, carbon monoxide diffusion test (DLCO), and 6-min walking test (6MWT) were evaluated. Echocardiography and right heart catheterization were performed in all patients for cardiac evaluation. Patients were stratified based on the global alignment and proportion (GAP) score (6). The clinical and functional status of the patients in both groups was compared. The patients' priority for lung transplantation was determined according to the general condition of the patient and donor suitability. Recipients and donors were matched according to blood group compatibility, urgency status of the patient, measured and calculated total lung capacity, and height.

Clamshell incision and anterolateral thoracotomy were performed for bilateral and single-lung transplantations. Indications for intraoperative extracorporeal membrane oxygenation (ECMO) were hypercapnia (>50 mmHg), arterial saturation (<85%), cardiac index (<2 L min−1 m−2), and mean pulmonary arterial pressure (mPAP; >40 mmHg) in addition to the right ventricular dysfunction. Central venoarterial ECMO was used for intraoperative cardiopulmonary support by cannulating the right atrial auricle and the ascending aorta. A 15–9 French (Fr) arterial cannula is used for the aorta, and a two-stage venous cannula or a 36 Fr curved-tip cannula is used for the right atrium.

Mycophenolate mofetil (1,000 mg) was administered in the operating room at the time of transplant. Methylprednisolone (500 mg, intravenous) was administered just before reperfusion of the transplanted lungs. In immunosuppressive treatment after transplantation, induction therapy with 20 mg of basiliximab was administered on the day of transplantation and 4 days post-transplantation in addition to triplet therapy comprising tacrolimus, mycophenolate mofetil, and prednisolone.

The study was designed in accordance with the principles outlined in the Helsinki Declaration and was approved by the local Clinical Research Ethics Committee at the Kartal Kosuyolu Training and Research Hospital (ID: 2020/6/344).

### Statistical Analysis

The distribution of quantitative data was analyzed using the Kolmogorov–Smirnov test. Data were expressed as a median (minimum–maximum). Mann–Whitney U-test was used to compare characteristics between groups A and B. Qualitative data were compared using Fisher's exact test. The Kaplan–Maier method was used for survival. A value of $P < 0.05$ was considered to be statistically significant. The analysis was performed using the SPSS for Windows software (IBM, Armonk, NY, USA).

## RESULTS

A total of 28 patients with IPF were listed for lung transplantation after evaluation for contraindications and screening. Age distribution was similar between groups A and B (54.7 vs. 54.1, p = 0.747). The difference was not statistically significant although group A had more men (94.1% vs. 63.6%). No significant difference in pO2 and pCO2 values were noted in arterial blood gas evaluations, whereas saturation values were worse in group B (82.9 vs. 74.2, p = 0.046). Pulmonary function test values and 6MWT results were similar between the two groups. Four (23.5%) and three (27.2%) patients in groups A and B, respectively, could not perform the 6MWT. Moreover, the echocardiographic findings of both groups were similar. Right heart catheterization failed to demonstrate a different systolic pulmonary artery pressure, pulmonary artery capillary wedge pressure, cardiac output, and cardiac index. However, mean pulmonary artery pressure was higher in group A than in group B (31.6 vs. 23.9, p = 0.053). Comorbidities such as coronary artery disease and arterial hypertension were more common in group B than in group A. The majority of the patients were classified as stage 2 in the GAP index evaluation in both groups, but the GAP index was similar between the groups (Table 1).

**Table 1. Clinical characteristics of patients at the time of the evaluation for lung transplantation**

| | Group A (n=17) | Group B (n=11) | p |
|---|---|---|---|
| **Age** | 54.7 (45-64) | 54.1 (38-62) | 0.74 |
| **Male** | 16 (94.1) | 7 (63.6) | 0.12 |
| **Female** | 1 (5.9) | 4 (36.4) | |
| **Comorbidities** | 9 (52.9) | 6 (54.5) | |
| Coronary artery diseases | 3 (17.6) | 3 (27.2) | |
| Arterial hypertension | 4 (23.5) | 4 (36.3) | |
| Pulmonary hypertension | 8 (47.1) | 3 (27.2) | 0.93 |
| Diabetes | 2 (11.7) | 1 (9.1) | |
| Gastro-oesophageal reflux | 1 (5.8) | - | |
| Depression | 2 (11.7) | 1 (9.1) | |
| Osteoporosis | 2 (11.7) | 2 (18.1) | |
| **Waiting list time days** | 71.2 (3-206) | 110 (14-303) | 0.23 |
| **BMI kg/m²** | 26.8 (20.2-32.4) | 24.9 (18.7-30.1) | 0.18 |
| **O2-therapy (l/min)** | 3.7 (2-6) | 3.3 (0-7) | 0.76 |
| **Urgent listing** | 3 (17.6) | 1 (9.1) | |
| **Arterial blood gas** | | | |
| pH | 7.41 | 7.4 | |
| pO2 | 53 (45-68) | 51.3 (41-65) | 0.23 |
| pCO2 | 41.2 (37-47) | 43.8 (36-52) | 0.44 |
| sat % | 82.9 (69-93) | 74.2 (75-91) | 0.04 |
| **Pulmonary function test** | | | |
| FVC (% of predicted) | 42.1 (28-70) | 40.8 (22-56) | 0.95 |
| FEV1 (% of predicted) | 44.9 (32-73) | 42.1 (24-56) | 0.68 |
| DLCO (% of predicted) | 27.3 (18-41) | 28.8 (21-37) | 0.56 |
| DLCO unperformed, n | 4 (23.5) | 5 (45.4) | |
| **6-MWT** | | | |
| Distance m | 193 (63-350) | 218 (200-315) | 0.16 |
| Final SpO2 (%) | 81 (65-89) | 85 (71-94) | 0.27 |
| Unperformed | 4 (23.5) | 3 (27.2) | |
| **ECHO** | | | |
| RV dilation,n | 8 (47) | 6 (54.5) | 0.16 |
| TAPSE mm | 20.2 (15-27) | 20.6 (15-28) | |
| **Right heart catheterization** | | | |
| PABs mmHg | 44.7 (21-95) | 43.4 (27-98) | 0.76 |
| PABm mmHg | 31.6 (11-54) | 23.9 (17-52) | 0.05 |
| CO mL/min | 4.2 (2.4-5.5) | 4.8 (3.4-7) | 0.55 |
| Ci L/min/m² | 2.5 (1.8-3.5) | 2.7 (2.2-3.5) | 0.35 |
| PAWP, mmHg | 9.5 (6-14) | 8.9 (5-14) | 0.61 |
| **GAP index** | | | |
| Stage I | 0 | 1 (9.2) | |
| Stage II | 10 (58.8) | 7 (63.6) | 0.40 |
| Stage III | 7 (41.2) | 3 (27.2) | |
| **Antifibrotic use** | 5 (29.4) | 6 (54.5) | 0.40 |

Value are expressed as mean (min.-max. range) or n (%) RV: Right ventricle, RHC: Right heart catheterization, PAPs: Systolic pulmonary arterial pressure, PAPm: Mean pulmonary arterial pressure, PHT: Pulmonary arterial hypertension

The waiting list mortality was 39.3% (11/28). The median waiting time on the list was similar for groups A and B with 71.2 days (range, 3-206 days) and 110 days (range, 14-303 days), respectively. Four patients were listed for urgent lung transplantation in severe acute respiratory failure based on acute exacerbation IPF. All four patients underwent extracorporeal membrane oxygenation as a bridge to lung transplantation. Three patients underwent transplant procedures and one died on the waiting list.

**Post-transplant outcome**

Two single and 15 bilateral lung transplants were performed. Intraoperative ECMO was used in eight (47.1%) cases. Donor variables were presented in Table 2. The mean duration of mechanical ventilation and intensive care unit (ICU) stays were 5.6 and 7.3 days, respectively. Furthermore, the 30-day and 1-year mortalities were 23.5% (4/17) and 29.4% (5/17), respectively. The first patient was a 63-year-old man with acute exacerbation of IPF who was listed as urgent. He was referred from an external ICU to our clinic with extracorporeal membrane oxygenation preoperatively. Bilateral lung transplantation was performed in this patient on the third day. No intraoperative complications were evident, and the patient was extubated on the second postoperative day. In the preoperative period, blood culture taken in the external center was positive and the patient died of sepsis on the 20$^{th}$ day. The other three patients died due to sudden cardiac arrest, hyperacute rejection, and bleeding. One patient died due to sepsis in the fifth month. In this patient, single-lung transplantation was performed on the 55$^{th}$ day of veno-venous ECMO due to intensive intraoperative bleeding. The lesion was radiologically compatible with Aspergillus infection, and the bronchoscopy material showed Aspergillus growth in the donor lung in the fourth month of follow-up. A bronchoscopy lavage culture culture-identified Pseudomonas was noted in a patient with acute deterioration. The patient died of sepsis (Table 3).

**Table 2. Donor variables**

| | |
|---|---|
| **Cause of death** | |
| Intracranial bleed/stroke | 8 (47) |
| Trauma | 4 (23.5) |
| Dilated cardiomyopathy | 1 (5.9) |
| Heart unknown cause | 2 (11.8) |
| Gunshot wound | 1(5.9) |
| Suicidal hanging | 1 (5.9) |
| **Age** | 34.7 (16-55) |
| **PaO2 mm Hg on FiO2 of 1.0** | 379 (278-521) |
| **Intubation time (day)** | 3.93 (1-9) |
| **Heavy smoker (>20 pack/year)** | 6 (35.3) |

Twelve patients who survived the first year were alive without any problems. The mean follow-up period was 25.7 months (range, 13–37 months). The 3-year survival rate was 70.6% (Figure 1).

**Table 3. Intraoperative data and outcomes (n=17)**

| | |
|---|---|
| **Transplantation type** | |
| Bilateral | 15 (88.2) |
| Single | 2 (11.8) |
| **Ischemic time (min)** | |
| First lung | 312 (230-410) |
| Second lung | 415 (340-520) |
| **ECMO as a bridge to lung transplantation** | 2 (11.7) |
| **Intraoperative ECMO** | 8(47.1) |
| **Severe PGD** | 7 (41.2) |
| **Mechanical ventilation, days** | 5.6 (1-21) |
| **Tracheostomy** | 4 (23.5) |
| **ICU stay, days** | 7.3 (1-21) |
| **30 day mortality** | 4 (23.5) |
| **One year mortality** | 5 (29.4) |
| **Cause of mortality** | |
| Sepsis | 2 |
| Sudden cardiac arrest | 1 |
| Hyperacute rejection | 1 |
| Bleeding | 1 |
| **Three year survival %** | 70.6 |

**Value are expressed as mean (min.-max. range) or N (%)**
**ECMO: Extracorporeal membrane oxygenation, PGD: Severe primary graft dysfunction; ICU: intesive care unit**

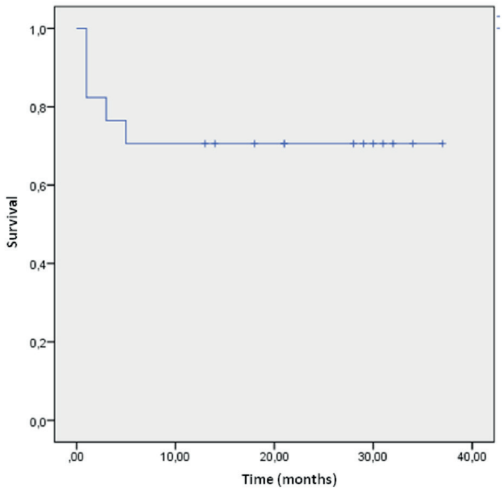


**Figure 1.** Overall survival

## DISCUSSION

Despite the ISHLT indications (3), in our experience, patients with IPF were not referred at the time of diagnosis and were listed for lung transplantation when respiratory function had already deteriorated significantly. All of the patients were already on long-term oxygen therapy, DLCO % was less than 30 % or the 6-MWT was less than 250 meters. In this current study, 1-year mortality was 29.4% and the 3-year survival rate was 70.6%. Our outcomes of lung transplantation were acceptable according to the data of the ISHLT registry, survival was 80% in the first year, 64% in the third year (7).

Pulmonary hypertension (PH) was present in 39.2% (11/28) of patients who were listed for lung transplantation at our institute. PH was higher in patients who underwent lung transplantation compared with patients who died on the waiting list. Lung allocation score (LAS) was not used in the lung transplant recipient selection, but its criteria were considered. The priority of transplantation is given to patients with PH in the presence of more than one patient suitable for a donor lung. Therefore, the presence of PH was higher in the transplant group. Higher mean pulmonary arterial pressure is associated with an early incidence of postoperative mortality and poorer survival (8). PH, which is a progressive disease involving the small vessels of the pulmonary vascular bed and is significantly associated with poor prognosis and mortality in IPF, is the most important comorbidity. PH has been shown to increase for up to 85% in the patients on the waiting list although its prevalence varies between 37% and 41% in patients with IPF (9). The benefit of treatment is still controversial in patients with PH caused by pulmonary fibrosis. However, PH treatment (drugs such as sildenafil and treprostinil) has been shown to preserve or increase the exercise capacity of patients, although it does not contribute to survival (10-11). PH treatment was not given in the patients listed during this study period. It is believed that better pulmonary rehabilitation can be provided if patients with advanced PH are treated on the waiting list. Thus, more studies are needed on this topic.

The shortage of suitable donors increases the waiting list mortality. Data from a 2017 report in the USA showed that the waiting list mortality was 19.7% (12). Mortality varies according to the diagnosis of diseases, and the waiting list mortality is up to 64%, especially in IPF (13). The waiting list mortality has decreased with the development of donor lung allocation strategies. The number of lung transplantation for IPF exceeded the number of patients transplanted due to chronic obstructive pulmonary disease (COPD) since LAS was used in the USA. Furthermore, all transplantation indications increased from 34% to 41%. On the other hand, the waiting list mortality decreased from 21% to 11% (14).

Idiopathic interstitial pneumonia is the second most common indication of lung transplantation following COPD (15). In addition, IPF is the most common and has the worst prognosis in idiopathic interstitial pneumonia. Moreover, lung transplantation is the most common among these diseases (16). The median survival of patients with IPF is 2–5 years. Thus, these patients should be immediately directed to a transplant center when diagnosed (3-17). Poor prognosis parameters in IPF include ≥10% decline in forced vital capacity or ≥15% in DLCO during a 6-month follow-up, worsening blood gas, SaO2 < 88%, increased dyspnea and cough symptoms, 6MWM < 250 m or >50 m reduction in 6 months, diagnosis of PH, worsening of breathing, and hospitalization due to pneumothorax or acute exacerbation (18). Thus, the patients should be referred promptly to a lung transplant clinic in the presence of these parameters.

Some concerns exist on antifibrotic use before transplantation. Pirfenidone delays wound healing by inhibiting transforming growth factor-β and can prevent healing of the bronchial anastomosis. Nintedanib can cause bleeding with tyrosine kinase inhibition and vascular endothelial growth factor receptor inhibition (19). Although limited studies on this subject are available, some have proved the opposite of this idea. Leuschner et al. and Delanote et al. suggested in their studies that previous antifibrotic therapy caused no complications in the post-transplant period (20-21). This study used antifibrotics in five (29.4%) patients who underwent lung transplantation and six (54.5%) patients who died on the waiting list. It is believed that these patients were referred for transplantation too late, and consequently the mortality in patients who used antifibrotic drugs was high. The main topic to be discussed here is whether the use of antifibrotics prevents acute exacerbation. Studies on the success of antifibrotic therapy as a bridge to lung transplantation by preventing acute exacerbation are needed.

Double-lung transplantation is more preferred today than single-lung transplantation (15). However, choosing single-lung transplantation procedures due to donor insufficiency may decrease waiting list mortality. Villavicencio et al. claimed that patients who underwent single-lung transplantation had worse survival if the mean pulmonary artery pressure was higher than 30 mmHg (22). On the other hand, double-lung transplantation had better results than single-lung transplantation in patients with mPAP over 40 mmHg in another study (23). Single- and double-lung transplantations will continue to be discussed due to donor shortage, but it is common to perform double-lung transplantations in the presence of severe PH.

This study is the first to report on lung transplantation for IPF in Turkey although outcomes of lung transplantation for IPF have been published for more than two decades. However, it has some limitations. This is a single-center, retrospective study. The sample size of the present study is low owing to the limited experience of lung transplantation in the region. All patients were seen for the first time in the examination for listing. No patients were placed on the waiting list after the follow-up list, thus the waiting time was very short in both groups. Therefore, factors that increased waiting list mortality were not shown. It was not determined whether deaths on the waiting list were due to acute exacerbation. The prioritization of patients on the waiting lists is controversial. The study could not provide clear information about this issue due to the low number of patients on the waiting list.

## CONCLUSION

In conclusion, lung transplantation should be offered as a treatment option for patients with IPF without absolute contraindications. Patients with IPF should be referred to the transplant clinic as soon as diagnosed. Late referral of patients increases the waiting list and postoperative mortality.

*Competing interests: The authors declare that they have no competing interest.*

*Financial Disclosure: There are no financial supports.*

*Ethical approval: The approval of the Kartal Kosuyolu Training and Research Hospital scientific research and publication ethics board; numbered as 2020/7/344.*

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