SCIE DOI: 10.14744/scie.2022.34635
South. Clin. Ist. Euras. 2022;33(3):225-230

Original Article

Candidate Listing and Lung Transplantation in Cystic Fibrosis: A Single-Center Experience from Turkey

Muhammet Ali Beyoğlu, Mehmet Furkan Şahin, Sinan Türkkan, Erdal Yekeler

Department of General Thoracic Surgery and Lung Transplantation, University of Health Sciences, Ankara City Hospital, Ankara, Türkiye

Submitted: 20.07.2022
Accepted: 25.07.2022

Correspondence: Muhammet Ali Beyoğlu, Sağlık Bilimleri Üniversitesi, Ankara Şehir Hastanesi, Göğüs Cerrahisi ve Akciğer Nakli Kliniği, Ankara, Türkiye
E-mail: muhammetalibeyoglu@gmail.com

Keywords: Lung transplantation; referral and consultation; waiting lists.

ABSTRACT

Objective: This study aimed to analyze candidate listing and lung transplant outcomes in cystic fibrosis (CF) patients referred to a lung transplant center in Turkey.

Methods: Between January 2015 and July 2022, 27 patients were evaluated retrospectively in our lung transplant center for end-stage lung disease due to CF. The characteristics of the patients at the time of admission, their status on the list, survival, and lung transplantation results were recorded.

Results: Of the total patients, 14 (51.9%) of the patients were females and 13 (48.1%) were males. The average age was 17.81±7.24 years, and the mean body mass index (BMI) was 15.5±2.0 years. Six patients had cystic fibrosis-related diabetes (22.2%). Twenty-five patients (92.6%) were receiving supplemental oxygen. Low BMI was present in 23 (85.2%) patients, osteopenia in 8 patients (29.6%), and osteoporosis in 15 patients (55.6%). Four patients had liver fibrosis (14.8%). Twenty-five patients (92.6%) were added to the waiting list. Because of the early case and low BMI, one patient from each was omitted. The mean overall waiting time was 544.0±376.5 days. Nine patients (45%) on the waiting list died. Lung transplantation (4 bilateral pulmonary + 1 bilateral lobar) was performed on 5 patients (20%) on the waiting list. One of the lung recipients died in the early postoperative period.

Conclusion: Contrary to developed countries, patients referred to our lung transplant center due to CF consisted of pediatric and young adult patients. Patient referral should be planned considering the long waiting period and the shortage of donors in Turkey, and the need for time to manage modifiable comorbidities. Patients with CF should be referred early to the transplantation center to obtain the best benefit from lung transplantation.

INTRODUCTION

Cystic fibrosis (CF) is an autosomal recessive genetic disorder affecting the electrolyte transport of epithelial cells. The disease frequently affects the lungs, pancreas, liver, intestine, sinuses, and reproductive organs.[1] Individuals with CF live longer due to advances in CF care and the introduction of newer CF disease-modifying therapies in developed countries.[2] Nevertheless, advanced CF lung disease is common and is the leading cause of death in CF patients.[3]

As in other end-stage lung diseases, lung transplantation is successfully applied in CF patients for whom medical treatment is insufficient. CF-related lung disease has been the third most common indication for adult lung transplantation for decades and by far the most common indication in children and adolescents.[4] Developing countries, on the other hand, lag behind these developments for various reasons. Delay in diagnosis, lack of regular use of maintenance therapy, and less access to new and costly CF modulator drugs are the leading causes of poor survival in patients with CF in developing countries. Also, the lung transplant program has been slow to be implemented in developing countries. There are few studies in the literature on the lung transplantation process in patients with CF in developing countries. This study aimed to analyze the candidate listing and lung transplant results in patients with CF in a developing country.

MATERIALS AND METHODS

Institutional Ethics Committee approved this single-center, retrospective, and observational study (approval no. E1-22-2666). Because the patients' personal information was not shared, informed consent was waived. All authors confirmed adherence to the World Medical Association Declaration of Helsinki on the ethical conduct of human subjects research.

The patients hospitalized as lung recipient candidates in our center between January 2015 and July 2022 were evaluated retrospectively. Patients with a diagnosis of CF were included in the study. Patients with noncystic bronchiectasis and other indications were excluded from the study. The following diagnostic criteria were met by all CF patients: (a) two sweat tests with chloride concentrations greater than 60 mmol/L and (b) one with chloride concentrations greater than 60 mmol/L and DNA analysis—two identified disease-causing CF mutations. If the sweat value was less than or equal to 60 mmol/L, the following conditions were met: (a) DNA analysis revealed two disease-causing CF mutations and (b) clinical presentation revealed typical CF features.[1]

A retrospective review of electronic and physical patient files was performed. Age, gender, body mass index (BMI), CF-related diabetes (CFRD), other comorbid diseases, need for oxygen supplement, pulmonary function test parameters, 6-minute walk test (6MWT) result, and laboratory and imaging test results were all recorded. Nose, throat, urine, stool, blood cultures, sputum acid-fast bacillus staining, and quantiferon-tb tests were all performed. Weight (kg)/height $(m)^2$ was used to calculate the BMI. Values below 18 are classified as low BMI. CFRD was defined as hyperglycemia based on oral glucose tolerance testing results or random blood sugar measurements necessitating insulin therapy. The 6MWT was defined as the distance the patients walked on a hard and flat surface in 6 min. Hospitalization for frequent lung infections, intravenous antibiotic use, enteral nutrition support, and osteopenia–osteoporosis treatment status were all documented. Bone mineral densitometry results between 1 and 2.5 were classified as osteopenia, and those below 2.5 were classified as osteoporosis.[5] The International Cystic Fibrosis Society criteria were used to classify advanced CF-associated lung disease. ACFLD is defined as FEV1 < 40% predicted when "stable" (not during a pulmonary exacerbation), or referred for lung transplantation evaluation, or one or more of the following characteristics: previous intensive care unit admission for respiratory failure, hypercarbia, daytime oxygen requirement at rest (excluding nocturnal use only), pulmonary hypertension, severe functional impairment from respiratory disease (New York Heart Association Class IV), 6MWT distance <400 m.[3] Patients with CF were listed for lung transplant based on the International Society for Heart and Lung Transplantation's timing of listing criteria (Table 1).[6] The dates on which the patients were placed on the lung transplant waiting list were recorded. The current status information of the patients was obtained from the application records of the lung transplantation center outpatient clinic, phone calls, and the national electronic patient record system. The waiting times of the patients on the lung transplant waiting list were calculated according to the date of death, the date of a lung transplant, or July 1, 2022, from the listing date.

Statistical analysis

All analyses of the research were evaluated with the SPSS 24.0 package program. Descriptive statistics were given a number of units (n), percent (%), and mean and standard deviation.

RESULTS

In our lung transplantation center, the first recipient candidate with the diagnosis of CF was examined in December 2015. Until July 2022, 27 patients with CF were examined as lung recipient candidates. Of these patients, 14 (51.9%) were females and 13 (48.1%) were males. The mean age was 17.81±7.24 years (min–max: 7–30). Six patients had CFRD. The mean BMI of the patients was 15.5±2 (min–max: 11.4–19.4). Percutaneous endoscopic gastrostomy was performed in 8 patients due to low BMI. Of the total patients, 25 (92.6%) followed pancreatic enzyme replacement therapy. Baseline patient demographics are presented in Table 2.

Table 1. Criteria for the lung waiting list in cystic fibrosis patients

1. Chronic respiratory failure with hypoxia (PaO_2 <60 mmHg) and/or hypercapnia (pCO_2 >50 mmHg)
2. Inability to wean from mechanical ventilation support
3. Development of pulmonary hypertension
4. Frequent hospitalization due to exacerbations
5. Rapid decline in pulmonary function tests
6. World Health Organization functional classification is IV

Table 2. Baseline characteristics of the patients

Variables	n Mean	% SD
Gender		
Female	14	52
Male	13	48
Age		
<18 (years)	13	48
≥18 (years)	14	52
BMI		
<18	23	85.2
≥18	4	14.8
Supplemental oxygen requirement		
Not required	2	7.4
8 h	1	3.7
12 h	7	25.9
24 h	17	63
CFRD	6	22
Pancreatic enzyme replacement	25	92.6
Frequent hospitalization	10	37
Hemoptysis	1	3.7
Pneumothorax	2	7.4

BMI: Body mass index; CFRD: Cystic fibrosis-related diabetes.

When the indications for referral to the lung transplantation center were examined, it was determined that 25 patients (92.6%) were referred for advanced CF-related lung disease, 1 (3.7%) for recurrent pneumothorax, and 1 (3.7%) for massive hemoptysis (>240 mL). When the patients were examined according to the need for oxygen support, 17 (63.0%) of 25 patients (92.6%) using long-term oxygen were using oxygen for 24 h, 7 (25.9%) for 12 h, and 1 (3.7%) for 8 h. The pulmonary function test, 6MWT, laboratory, and imaging results of the patients are shown in Table 3.

Twenty-five (92.6%) patients were placed on the lung waiting list. One patient was excluded from the list because of persistent low BMI despite supportive treatment, and 1 patient was excluded because it was an early case. Lung transplantation for CF was performed for the first time in our center in November 2018. Lung transplantation was performed in 5 (20%) of 25 patients on the lung waiting list due to CF until July 2022. While bilateral sequential pulmonary transplantation was performed in 4 patients (80%), bilateral sequential lobar transplantation was performed in 1 patient (20%). Of the 20 patients on the waiting list who could not be transplanted, 9 (45%) died due to respiratory failure. The mean age of the patients who died on the waiting list is 22.0±7.5 years. The patient who was not included in the list due to low BMI died 3 months after the examination, again due to respiratory failure. One lung recipient died early postoperatively. Liver enzyme elevation was not detected in the pre- and post-listing follow-ups of the patient with liver fibrosis. The degree of liver fibrosis was evaluated as low in clinical controls. Before listing, the council, which included gastroenterology, determined that liver fibrosis would not impede lung transplantation. Cerebral hypoxia and brain death occurred after coagulation-related bleeding caused by a postoperative liver failure in the patient. The patient died 30 days after the lung transplant. The other 4 (80%) patients who underwent lung transplantation healthily continue their lives. Pulmonary function test and 6MWT results of patients who underwent lung transplantation are given in Table 4. The average overall waiting time was calculated as 544.0±376.5 days, 692.0±437.5 days for those who died while on the list and 358.6±378.6 days for those who underwent lung transplantation.

Table 3. Clinical, laboratory, and invasive test results of the patients

Variables	n Mean	% SD
Pulmonary function test results (%)		
FEV_1	26.6	9.0
FVC	33.6	11.6
DLCO	39.1	20.3
6MWT (m)	257.1	136.6
Heart catheterization (mmHg)		
Mean PAP	18.9	8.4
PCWP	11.9	3.4
BMD	-2.42	1.38
Osteopenia	8	29.6
Osteoporosis	15	55.5
Microbiology		
Pseudomonas aeruginosa	8	29.6
Klebsiella Pneumonia	2	7.4
Corynebacterium pseudodiphtheriticum	2	7.4
Staphylococcus aureus	1	3.7
Candida tropicalis	1	3.7
Sinusitis or polip on paranasal CT	19	70.3
Hepatic fibrosis	4	14.8

FEV_1: Forced expiratory volume in 1 s; FVC: Forced vital capacity; DLCO: Diffusing capacity of the lung for carbon monoxide; 6MWT: 6-minute walk test; PAP: Pulmonary artery pressure; PCWP: Pulmonary capillary wedge pressure; BMD: Bone mineral densitometry; CT: Computed tomography.

Table 4. Postoperative pulmonary function parameters of lung recipients

	Recipient 1	Recipient 2	Recipient 3	Recipient 4
FEV_1 (%)	81	76	84	86
FVC (%)	93	73	86	90
6MWT (m)	550	500	600	550

FEV_1: Forced expiratory volume in 1 s; FVC: Forced vital capacity; 6MWT: 6-minute walk test.

DISCUSSION

This study showed essential results about lung transplantation activities in CF patients in developing countries, which are few in the literature. Contrary to developed countries, half of the patients referred to the lung transplant center due to CF in Turkey are pediatric patients, and the other half are young adult patients. Pathologies that increase morbidity and mortality after lung transplantation, such as malnutrition, osteoporosis, and liver fibrosis, are frequently observed in these patients. However, lung transplantation can be successfully applied in developing countries.

As consanguineous marriage is very common in Turkey, the prevalence of autosomal recessive CF is expected to be high. The incidence of CF has been reported to be 0.029% in previous studies.[7] The first report of the Turkish National CF Registry includes 1170 patients, which is estimated to comprise 30% of the number of CF patients in the country.[8] There are several reasons for the low number of registered patients. Initially, newborn screening (NBS) for CF began in 2015 as two repetitive immunoreactive trypsinogen tests. The national CF registration system has been in operation since 2017. In addition, it is thought that the lack of knowledge and diagnosis about CF among healthcare workers in rural areas is effective in the low number of cases. According to NBS and the national registry system, patients with CF can be referred to a lung transplant center at a later age and without delay, owing to earlier diagnosis and appropriate supportive care.

Advances in CF care have resulted in significant adult populations in developed countries. In the European Cystic Fibrosis Society Patient Registry Annual Data Report, more than half of all patients with CF are over 18 years of age.[9] Based on the ISHLT Thoracic Transplant Registry analysis, approximately 90% of CF patients undergoing lung transplantation were reported to be over 18 years of age and 17.2% over 40 years of age.[4] However, the mean age of CF patients still alive in Turkey in 2017 was 7.3 years, and only 4.6% were adults.[8] In a Turkey-based study in which 14 patients evaluated for lung transplantation due to CF were analyzed, the mean age of the patients was presented as 22.8 years (range 11–41).[10] Similarly, the fact that the patients referred to our transplant clinic are pediatric and young adults indicates that CF maintenance therapy is ineffective.

We found that 85.2% (n=23) of the study patients had a BMI <18. Similarly, in the Turkish National CF Registry Report, patients' BMI median z scores were lower than in European countries.[8] The low BMI in our patients indicates that we should pay attention to monitoring the nutritional status of our patients and providing the necessary nutritional support, which seems to be a neglected issue in our CF care. Also, previous studies have found low body weight to be an independent risk factor for death after lung transplantation.[11,12] Improving the nutritional care of patients will improve respiratory functions, increase survival, and the success of lung transplantation.

A triple immunosuppressant regimen, including corticosteroids, is routinely administered after lung transplantation. The presence of osteopenia and osteoporosis in transplant candidates predisposes to pathological fractures with long-term use of corticosteroids after the lung transplant. BMI measurement is routinely performed on all patients examined for the waiting list. In our study, osteopenia or osteoporosis was detected in 70% (n=23) of the patients, and appropriate treatment was initiated.

Pseudomonas and *Aspergillus* infections are associated with chronic lung allograft dysfunction, particularly in CF patients.[13] Because *Pseudomonas* colonization is common in allografts following lung transplant, pretransplant use of inhaled antibiotics is necessary. In addition, the paranasal sinuses may act as a reservoir for allograft colonization in CF patients.[13] Paranasal sinusitis or polyp findings were found in 70.4% (n=19) of the study patients. Pretransplant sinus surgery may be beneficial in patients with CF with evidence of paranasal sinusitis or polyps. The prevalence of chronic *Pseudomonas aeruginosa* in CF patients in Turkey has been reported as 22.82%.[8] Similarly, our study's prevalence of chronic *P. aeruginosa* was 29.6%.

The government covers almost all drugs in Turkey, including inhaled antibiotics, inhaled bronchodilators, azithromycin, mannitol, hypertonic saline, noninvasive mechanical ventilation, pancreatic enzyme, nutritional supplements, and recombinant human DNAse. However, new and high-cost CF modulator drugs are not covered by the government. In a study of 1351 CF patients who underwent genetic analysis in Turkey, only 313 (23.2%) of these patients were found suitable for currently licensed CF modulator therapies.[14] Similarly, 31 distinct mutations were discovered in CF patients of Turkish origin examined in European CF centers.[15] Although successful results of modulator treatments in patients with specific mutations have been reported recently, only a quarter of patients in Turkey seem to benefit from these treatments. Studies evaluating the effectiveness of modulatory drugs in different mutations detected in Turkish patients are needed. For this reason, maintenance therapy and lung transplantation stand out as the most realistic treatments in three-quarters of CF patients who are not yet suitable for modulator therapy.

Savi et al.[16] reported that after 2011, the median lung waiting time for patients with CF was 17.6 months in their study in which they shared their 20-year experience. Similarly, in our study, the waiting time in patients who underwent lung transplantation was calculated as 358.6±378.6 days. Overall waiting time was determined as 544.0±376.5 days. The waiting time for patients with CF who died on the list was calculated as 692.0±437.5 days. When the reasons for the long waiting period are evaluated, the COVID-19 pandemic draws attention in addition to the current shortage of donors. The low number of donors and lung transplants performed in Turkey due to COVID-19 was presented in our previous study.[17] Considering the long waiting times and the unpredictability of the pandemic, we think earlier referral of CF patients to transplant centers would be beneficial. The life expectancy of CF patients in Turkey cannot be predicted because the national registry system has started recently, and the records are not yet sufficient. The mean age of the 15 patients who died in the national CF registry report was 13.5±9.9 years.[8] In our study, 9 patients died on the waiting list. The mean age of these patients was calculated as 22.0±7.5 years. Vayvada et al.[10] reported 1-year mortality of 28.6% (n=2) in 7 patients who underwent lung transplantation for CF. Similarly, mortality was calculated as 20% in our study's patients who underwent lung transplantation.

A prospective cohort study of 3328 patients with CF reported an incidence of CF-related liver disease (CFLD), increasing by approximately 1% per year after the age of 5 years, reaching 32% by age 25 years. The same study reported that the incidence of severe KFLD, such as cirrhosis or portal hypertension, reached 10% by age 30 years.[18] A recently published systematic review reported that CF-related liver disease triples the risk of death.[19] Hepatic fibrosis was detected in 4 (14.8%) of the study patients. One of these patients underwent a lung transplant, and hemorrhagic shock and ischemic cerebral edema occurred due to coagulopathy due to liver failure in the early postoperative period. In the per-operative period of a lung transplant, the hemodynamic status is relatively labile. Liver hypoxia should also be considered when patients with CF-related liver fibrosis are evaluated for lung transplantation. When patients with CF-related liver fibrosis are evaluated for lung transplantation, the risk of liver ischemia due to labile hemodynamic status should also be considered.

The limited number of cases stands out as the most significant limitation of our study. We also believe that the COVID-19 pandemic, which has been ongoing for over 2 years, increases the waiting time. Despite these shortcomings, we think that the waiting list and transplant results in CF patients will be a guide for clinics that will initiate a lung transplant program under similar conditions.

CONCLUSION

Coordination of CF centers with lung transplant clinics is essential to improve care and survival in patients with CF. Although the criteria for referral for lung transplantation are specific, regional conditions should also be considered when determining the referral timing. Due to the shortage of donors and the ongoing pandemic in Turkey, the waiting time is quite long. A comprehensive evaluation for lung transplantation and early referral to the transplantation center are necessary to inform the patient and family and avoid lung transplantation-specific contraindications.

Ethics Committee Approval

This study approved by the Ankara City Hospital Clinical Research Ethics Committee (Date: 15.06.2022, Decision No: E1/2666/2022).

Informed Consent

Retrospective study.

Peer-review

Internally peer-reviewed.

Authorship Contributions

Concept: M.A.B.; Design: M.F.Ş.; Supervision: E.Y.; Fundings: S.T., E.Y., M.F.Ş.; Materials: M.A.B.; Data: S.T.; Analysis: E.Y.; Literature search: M.A.B.; Writing: M.A.B.; Critical revision: E.Y.

Conflict of Interest

None declared.

REFERENCES

1. De Boeck K, Vermeulen F, Dupont L. The diagnosis of cystic fibrosis. Presse Med 2017;46:e97–e108. [CrossRef]
2. Stephenson AL, Ramos KJ, Sykes J, Ma X, Stanojevic S, Quon BS, et al. Bridging the survival gap in cystic fibrosis: An investigation of lung transplant outcomes in Canada and the United States. J Heart Lung Transplant 2021;40:201–9. [CrossRef]
3. Kapnadak SG, Dimango E, Hadjiliadis D, Hempstead SE, Tallarico E, Pilewski JM, et al. Cystic Fibrosis Foundation consensus guidelines for the care of individuals with advanced cystic fibrosis lung disease. J Cystic Fibrosis 2020;19:344–54. [CrossRef]
4. Benden C, Goldfarb SB, Stehlik J. An aging population of patients with cystic fibrosis undergoes lung transplantation: An analysis of the ISHLT Thoracic Transplant Registry. J Heart Lung Transplant 2019;38:1162–9. [CrossRef]
5. Lakey WC, Spratt S, Vinson EN, Gesty-Palmer D, Weber T, Palmer S. Osteoporosis in lung transplant candidates compared to matched healthy controls. Clin Transplant 2011;25:426–35. [CrossRef]
6. Orens JB, Merlo CA. Selection of candidates for lung transplantation and controversial issues. Semin Respir Crit Care Med 2018;39:117–25. [CrossRef]
7. Hangül M, Pekcan S, Köse M, Acıcan D, Şahlar TE, Erdoğan M, et al. The incidence of cystic fibrosis in central Anatolia region of Turkey in 2015 and 2016. Balkan Med J 2019;36:179–83. [CrossRef]
8. Dogru D, Çakır E, Şişmanlar T, Çobanoğlu N, Pekcan S, Cinel G, et al. Cystic fibrosis in Turkey: First data from the national registry. Pediatr Pulmonol 2020;55:541–8. [CrossRef]
9. ECFS Patient Registry. Available at: https://www.ecfs.eu/ecfspr/. Accessed Jul 17, 2022.
10. Vayvada M, Halis AN, Saribas E, Uygun Kizmaz Y, Çardak ME, Erkilic A, et al. Lung transplantation for cystic fibrosis in Turkey: First report. Exp Clin Transplant 2021;19:481–8. [CrossRef]
11. Lederer DJ, Wilt JS, D'Ovidio F, Bacchetta MD, Shah L, Ravichandran S, et al. Obesity and underweight are associated with an increased risk of death after lung transplantation. Am J Respir Crit Care Med 2009;180:887–95.
12. Allen JG, Arnaoutakis GJ, Weiss ES, Merlo CA, Conte JV, Shah AS. The impact of recipient body mass index on survival after lung transplantation. J Heart Lung Transplant 2010;29:1026–33.
13. Morrell MR, Pilewski JM. Lung transplantation for cystic fibrosis. Clin Chest Med 2016;37:127–38. [CrossRef]
14. Çobanoğlu N, Özçelik U, Çakır E, Şişmanlar Eyüboğlu T, Pekcan S, Cinel G, et al. Patients eligible for modulator drugs: Data from cystic fibrosis registry of Turkey. Pediatr Pulmonol 2020;55:2302–6.
15. Lakeman P, Gille JJ, Dankert-Roelse JE, Heijerman HG, Munck A, Iron A, et al. CFTR mutations in Turkish and North African cystic fibrosis patients in Europe: implications for screening. Genet Test 2008;12:25–35. [CrossRef]
16. Savi D, Mordenti M, Bonci E, Troiani P, Giordani B, D'Alù V, et al. Survival after lung transplant for cystic fibrosis in Italy: a single center experience with 20 years of follow-up. Transplant Proc 2018;50:3732–8. [CrossRef]
17. Sahin MF, Beyoglu MA, Turkkan S, Tezer Tekce Y, Yazicioglu A, Yekeler E. Donor lung evaluation and lung transplantation in the COVID-19 Era. Exp Clin Transplant. 2021 Apr 29. doi:10.6002/ect.2020.0567. [Epub ahead of print].
18. Boëlle P-Y, Debray D, Guillot L, Clement A, Corvol H. Cystic fibrosis liver disease: outcomes and risk factors in a large cohort of French patients. Hepatology 2019;69:1648–56. [CrossRef]
19. Sasame A, Stokes D, Bourke B, Connolly L, Fitzpatrick E, Rowland M. The impact of liver disease on mortality in cystic fibrosis-A systematic review. J Cyst Fibros. 2022;21:202–11.

Kistik Fibroziste Aday Listeleme ve Akciğer Nakli: Türkiye'den Tek Merkez Deneyimi

Amaç: Bu çalışmanın amacı, Tükiye'deki bir akciğer nakli merkezine sevk edilen kistik fibrozis hastalarda aday listeleme ve akciğer nakli sonuçlarını analiz etmektir.

Gereç ve Yöntem: Ocak 2015–Temmuz 2022 yılları arasında kistik fibrozise bağlı son dönem akciğer hastalığı nedeniyle akciğer nakli merkezimizde detaylı tetkik edilen 27 hasta geriye dönük olarak değerlendirildi. Hastaların başvuru anındaki özellikleri, listeye alınma durumu, sağ kalım ve akciğer nakli sonuçları kaydedildi.

Bulgular: Hastaların 14'ü (%52) kadın, 13'ü (%48) erkekti. Ortalama yaş 17.81±7.24, ortalama VKİ 15.5±2.0 idi. Altı hastada (%22) KF ilişkili diabet mevcuttu. Yirmi beş hasta (%92.6) uzun süreli oksijen tedavisi almaktaydı. Yirmi üç (%85.2) hastada düşük BMI, sekiz hastada (%29.6) osteopeni 15 hastada (%55.5) osteoporoz mevcuttu. Dört hastada (%14.8) karaciğer fibrozisi saptandı. Yirmi beş hasta (%92.6) bekleme listesine alındı. Bir hasta (%3.7) erken olgu bir hasta (%3.7) da düşük VKİ nedeniyle listeye alınmadı. Overall bekleme süresi ortalama 544.0±376.5 gün idi. Bekleme listesinde dokuz hasta (%45) hayatını kaybetti. Bekleme listesindeki hastalardan beşine (%20) akciğer nakli (4 bilateral pulmoner+1 bilateral lober) uygulandı. Akciğer nakli yapılan hastalardan biri erken dönemde hayatını kaybetti.

Sonuç: Gelişmiş ülkelerin aksine ülkemizde KF nedeniyle akciğer nakli merkezine yönlendirilen hastalar pediatrik ve genç erişkin hastalardan oluşmaktadır. Akciğer naklinden en iyi faydayı sağlamak için Kistik fibrozisli hastalar, uzun bekleme süresi, donör kıtlığı, modifiye edilebilen ko-morbiditelerin tespit ve tedavisi için transplantasyon merkezine erken dönemde sevk edilmelidir.

Anahtar Sözcükler: Akciğer transplantasyonu; bekleme listeleri; sevk ve konsültasyon.