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ORIGINAL ARTICLE

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The success of the Cystic Fibrosis Registry of Turkey for improvement of patient care

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Abstract

Background: Cystic fibrosis (CF) registries play an essential role in improving disease outcomes of people with CF. This study aimed to evaluate the association of newly established CF registry system in Turkey on follow-up, clinical, growth, treatment, and complications of people with this disease.

Methods: Age at diagnosis, current age, sex, *z*-scores of weight, height and body mass index (BMI), neonatal screening results, pulmonary function tests, history of meconium ileus, medications, presence of microorganisms, and follow-up were evaluated and compared to data of people with CF represented in both 2017 and 2019 registry data.

Results: There were 1170 people with CF in 2017 and 1637 in 2019 CF registry. Eight hundred and fourteen people were registered in both 2017 and 2019 of whom *z*-scores of heights and BMI were significantly higher in 2019 ($p = 0.002$, $p = 0.039$, respectively). Inhaled hypertonic saline, bronchodilator, and azithromycin usages were significantly higher in 2019 ($p = 0.001$, $p = 0.001$, $p = 0.003$, respectively). The percent predicted of forced expiratory volume in 1 sec and forced vital capacity were similar in 2017 and 2019 (88% and 89.5%, $p = 0.248$ and 84.5% and 87%, $p = 0.332$, respectively). Liver diseases and osteoporosis were significantly higher, and pseudo-Bartter syndrome (PBS) was significantly lower in 2019 ($p = 0.011$, $p = 0.001$, $p = 0.001$, respectively).

Conclusions: The *z*-scores of height and BMI were higher, the use of medications that protect and improve lung functions was higher and incidence of PBS was lower in 2019. It was predicted that registry system increased the care of people with CF regarding their follow-up. The widespread use of national CF registry system across the country may be beneficial for the follow-up of people with CF.

KEYWORDS
child, cystic fibrosis, registry

1 | INTRODUCTION

A patient registry is an organized data collection tool that includes clinical, sociodemographic, and other information about patients obtained from multiple data sources in the health system.[1] Registries document and standardize the data of patients according to defined questions, and provide continuous and comprehensive monitoring of individual data of patients with any disease in a particular geographic area.[2,3] Other tasks of the registries include evaluating the efficiency in medical care and monitoring patient safety.[2]

Cystic fibrosis (CF) is caused by various mutations in the CF transmembrane conductance regulator gene and every patient develops different clinical manifestations.[1] It is important to understand the heterogeneity of CF through registries to enable advances in clinical care.[3] National CF registries are developed by compiling patient data from specialist CF centers, and they are now available in many countries.[4–7] CF registries play an essential role in improving the quality of life and outcomes of people with this disease.[6] The information in the registry is intended to be used to evaluate the health status of people with CF, reveal care-related deficiencies, assist centers that monitor people, and guide their quality improvement initiatives. CF registries have been viewed as a model for the development and use of patient registries for other diseases.[1,6]

CF care has long been carried out in many clinics, throughout Turkey. Hence, the CF registry of Turkey (CFRT) was established by the "Turkish Pediatric Respiratory Diseases and Cystic Fibrosis Society," which is the first CF society in Turkey, to monitor people with CF in detail and regularly. The CFRT has participated in the European Cystic Fibrosis Society (ECFS) Patient Registry (ECFSPR) since 2016 and patient data are recorded annually by each CF center in a program specially developed for the CFRT since 2017. In 2017, the CFRT began feeding back summary data to CF centers contributing to the registry. The data recorded each year are shared with all CF centers in the form of annual reports and meetings. Annual reports allow each CF center to view and evaluate data of their own and other centers.[7] This may have raised awareness in centers that provide CF care. In this study, we aimed to evaluate the association between the data collection with CFRT and follow-up, clinical features, growth, treatment, and complications of people with CF in our country by comparing the results in the first year of the registry with the results in the third year.

2 | MATERIALS AND METHODS

This was a retrospective study of the CFRT. Annual data of the people with CF in 2017 and 2019 were included in the study. The demographic and clinical data for 2017 and 2019 were obtained from the CFRT, based on the definitions of the ECFSPR.[8] People who met the CF diagnostic criteria according to the ECFS were recorded in the CFRT. Informed consent was obtained from both the people and their families above the age of 6, and only from the families under the age of 6 before they were registered in the registry system.

The annual data in the CFRT consisted of multivariate sections. These six sections were the final status, treatment, microbiology, complications, and transplantation of the patients. Informations about the age at diagnosis, current age, sex, neonatal screening, and history of meconium ileus are also available in the CFRT. Besides the best weight and height in that year and pulmonary function test (PFT) results were recorded. PFT data included forced vital capacity (FVC) and forced expiratory volume in 1 sec (FEV_1) as the percent predicted. Growth of people with CF aged over 2 years was assessed using the body mass index (BMI) *z*-score.[9] Weight, height measurements, and BMI are expressed in terms of *z*-scores by using reference values. The *z*-scores of weight and height were noted for all people; BMI and *z*-scores of BMI were noted for people aged over 2 years. Chronic infection was defined as an infection that persisted despite treatment and the immune or inflammatory response of the host.[10] Chronic growths of microorganisms and nonchronic growths of microorganisms were noted. The treatments used and complications observed and transplantations were also recorded. According to the ECFSPR guidelines, pseudo-Bartter syndrome (PBS) was defined as blood pH >7.45, serum sodium <130 mmol/L, and serum chloride <90 mmol/L.[8]

The people with CF represented in both 2017 and 2019 registry data were evaluated in terms of demographic and clinical features such as sex, age at diagnosis, current age, *z*-scores of weights, height and BMI, neonatal CF screening results, PFTs, history of meconium ileus, medications, presence of microorganisms, complications, and transplantations. The IRT/IRT (immunoreactive trypsinogen) protocol has been used for neonatal CF screening in Turkey since 2015. If the first IRT is 90 μg/L and above which is taken at 72 h of life, the second IRT is applied to the patient in 7–14 days of life. If the second IRT is 70 μg/L and above, the screening test is considered positive.[7] The data of people represented in both years were compared between 2017 and 2019.

This study was conducted in accordance with the amended Declaration of Helsinki and all procedures performed in studies involving human participants were prepared in accordance with the ethical standards of the institutional and/or national research committee (Hacettepe University Ethics Board, Date: April 12, 2007; Reference number: HEK 07/16-21, Date: June 5, 2018, Reference number: GO 18/473-31).

2.1 | Statistical analysis

The IBM SPSS Statistics version 22.0 (IBM) for Windows was used for the statistical analyses. For descriptive statistics, categorical variables are expressed as absolute numbers and percentages, and continuous variables are expressed as medians (min–max). The normality of distribution of continuous variables was tested using visual and analytical methods. For comparisons between two dependent variables, the Wilcoxon signed-rank test was used for data that were not normally distributed, and the paired *t*-test was used for normally distributed data. McNemar tests were used for comparisons of

categorical variables between dependent groups. A value of $p < 0.05$ was considered statistically significant.

3 | RESULTS

There were 1170 people with CF in 2017 and 1637 in 2019 in CFRT. Twenty-three centers recorded data into the registry system in 2017 versus 27 centers in 2019. Two centers in 2017 did not enter data in 2019, and six centers in 2019 were newly added centers.

In 2017 and 2019, 54.3% and 53.2% of all people with CF were male, respectively. The median ages of all were 6.0 years (0–42 years) and 7.0 years (0–44 years) in 2017 and 2019, respectively. Figure 1 shows the histograms presenting number and ages of all people with CF in the 2017 and 2019 registry data and highlighting the subset of people with data for both years.

Eight hundred and fourteen people with CF were represented in both 2017 and 2019 registry data. The median age at diagnosis of these represented in both years was 0.3 years (0.1–22 years) and 431 (52.9%) were male. At birth, 44 (5.4%) had meconium ileus and 29 (3.6%) underwent surgery. The neonatal screening was performed on 193 people, and in 173 (89.6%) of them, the screening result was above the cut-off values, and 20 were false negatives in neonatal screening and were diagnosed by sweat chloride test and/or genetic analysis with clinical findings. The demographic and clinical features of people with data in both years were shown in Table 1. There were significant differences in *z*-scores of heights and BMI; but no difference in *z*-score of weight between 2017 and 2019 ($p = 0.002$, $p = 0.039$, $p = 0.401$, respectively).

In people with data in both years, there were no significant differences in chronic *Pseudomonas aeruginosa*, chronic *Staphylococcus aureus*, chronic *Burkholderia cepacia complex*, *nontuberculous mycobacteria*, and *Stenotrophomonas maltophilia* infections between 2017 and 2019 ($p = 0.798$, $p = 0.825$, $p = 0.625$, $p = 0.688$, and $p = 1.000$, respectively).

The medications and supportive treatments of people with data in both years were shown in Table 2.

The medians of percent predicted of FEV_1 and FVC of these 814 people in 2017 were 88 (19–143) and 84.5 (21–137), respectively. In 2019, the medians of percent predicted of FEV_1 and FVC were 89.5 (15–146) and 87.0 (13–143), respectively. Although it was observed that the percent predicted of FEV_1 and FVC increased after 2 years but there were no statistically significant differences ($p = 0.248$ and $p = 0.332$).

The complications of people with data in both 2017 and 2019 were shown in Table 3. In 2017, there was no pneumothorax but in 2019 pneumothorax was observed in 2 (0.2%) people with CF. In 2017, malignancy was seen in 1 (0.1%) person and were no further reports of malignancy in 2019 data.

4 | DISCUSSION

In the 2-year follow-up of people with CF in the newly established CFRT, an increase in the *z*-scores of height and BMI of them, as well as an increase in the use of medications that promote mucociliary clearance and improve lung function was observed. The frequency of PBS, which was seen as the most common complication of people with CF in the first year of the registry, decreased 2 years later. The registry system can enable us to see the clinical changes of these people by annual reports.

Management of nutrition, which is a risk factor for morbidity and mortality is important in people with CF as lung function, pulmonary muscle function, and exercise tolerance are associated with nutritional status.[11,12] Although many physicians consider malnutrition as

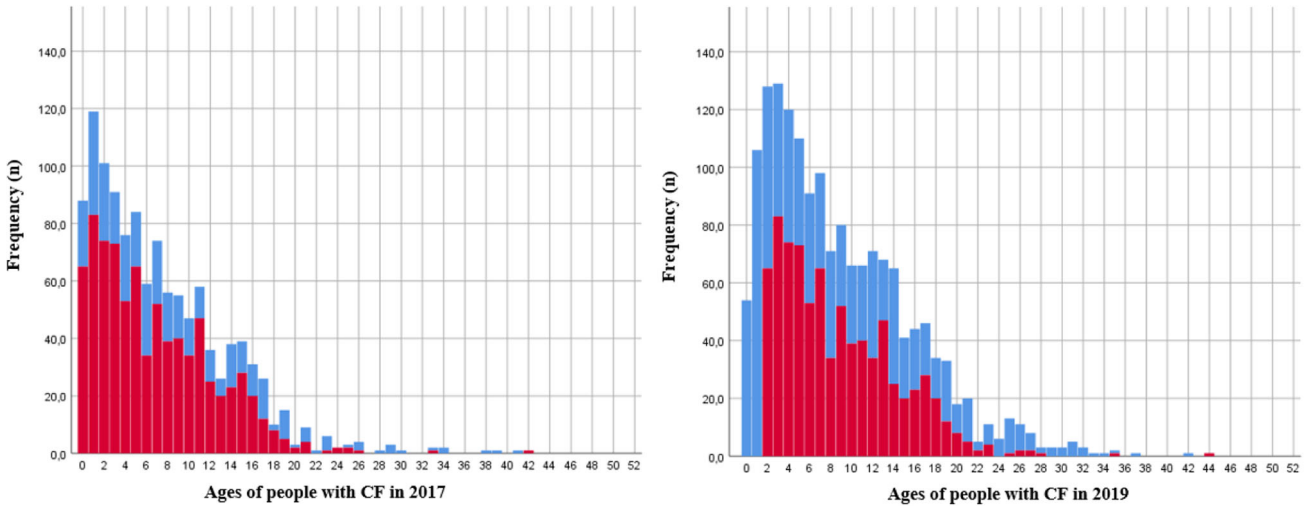

FIGURE 1 The histograms presenting numbers and ages of all people with cystic fibrosis (CF) in the 2017 and 2019 registry data and highlighting the subset of people with CF with data for both years (people with CF with data in both years represented with red and the others with blue) [Color figure can be viewed at wileyonlinelibrary.com]

TABLE 1 The demographic and clinical features of people with cystic fibrosis represented data in both 2017–2019

n = 814	2017 Median (min–max)	2019 Median (min–max)	*p*
Age (years)	5 (0–42)	7 (2–44)	-
Weight *z*-score	−0.89 (−5.48–7.40)	−0.82 (−3.69–3.89)	0.401
Height *z*-score	−0.93 (−10.75–10.14)	−0.78 (−6.65–3.46)	0.002*
BMI (kg/m^2)[a]	15.69 (7.71–39.59)	15.80 (9.60–33.20)	0.109
BMI *z*-score[a]	−0.58 (−5.67–6.02)	−0.46 (−3.26–7.05)	0.039*

[a]For ≥2 years old (*n*: 666 patients).

*Statistically significant.

TABLE 2 Treatments reported in the CFRT, 2017 and 2019

n = 814	2017 *n* (%)	2019 *n* (%)	*p*
rhDNase	750 (92.1)	767 (94.2)	0.106
Pancreatic enzyme replacement	744 (91.4)	736 (90.4)	0.182
Ursodeoxycholic acid	143 (17.6)	174 (21.4)	0.008*
Azithromycin	53 (6.5)	77 (9.5)	0.003*
Inhaled hypertonic saline	68 (8.4)	158 (19.4)	0.001*
Inhaled antibiotics	151 (18.6)	162 (19.9)	0.488
Bronchodilators	151 (18.6)	212 (26.0)	0.001*
Oxygen support	15 (1.8)	27 (3.3)	0.031*

Note: This table represents all people with CF who contributed data to both years.

Abbreviations: CF, cystic fibrosis; CFRT, CF registry of Turkey; rhDNase, recombinant human DNAse.

*Statistically significant.

TABLE 3 Comorbidities reported in the CFRT, 2017 and 2019

n = 814	2017 *n* (%)	2019 *n* (%)	*p*
Allergic bronchopulmonary aspergillosis	12 (1.5%)	15 (1.8%)	0.549
Diabetes mellitus	24 (2.9%)	34 (4.2%)	0.078
Liver diseases	70 (8.6%)	94 (11.5%)	0.011*
Major hemoptysis[a]	2 (0.2%)	1 (0.1%)	1.000
Pseudo-Bartter syndrome	71 (8.7%)	14 (1.7%)	0.001*
Osteoporosis	16 (2.0%)	41 (5.0%)	0.001*

Note: This table represents all people with CF who contributed data to both years.

Abbreviations: CF, cystic fibrosis; CFRT, CF registry of Turkey.

[a]>250 ml hemoptysis.

*Statistically significant.

a result of lung disease, it is a contributing factor to lung disease.[12] In some studies, nutritional status has been associated with survival for people with CF, and stabilization has been observed in malnourished people when their nutritional status improves.[12]

In the ECFSPR, evaluating by age groups, the median *z*-scores for weight and height in 2017 ranged from −1.3 to −1.0 and from −1.1 to −0.9, respectively.[8] In the CFRT, evaluating by age groups in 2017, the median *z*-scores of weights, height, and BMI of all patients ranged from −1.2 to −0.6, −1.4 to −0.6, and −0.8 to −0.3, respectively. In our study, it was observed that height and BMI *z*-score improved over the years in people with data in both years. As seen in the first report of the registry system, and as stated in the study by Dogru et al.,[7] people with CF had low *z*-scores of weights, height, and BMI in 2017. This was thought to raise awareness about the low *z*-scores of people in Turkey as the evaluation of the annual reports and data of them ensured that the necessary follow-up for growth and nutrition was performed well with our registry system.

The severity and deterioration of lung disease are the main factors of morbidity and mortality in CF. The severity of lung disease is also associated with the growth of people. It is known that proper nutritional status and maintenance of growth slow the decline of lung function.[13] Significant associations were identified between nutritional status and PFTs in some studies.[12] In a multicenter study conducted by Zemel et al.,[14] *z*-scores of weight and height of people with CF were positively associated with PFTs, and it was found that growth and nutritional status of people were associated with changes in FEV_1. Thus, it was suggested that nutritional interventions might slow the impaired pulmonary function in CF.[14] In a study comparing two different countries, it was stated that nutrition was associated with PFTs, and additional factors such as different medication usages might impact lung function.[15] In our study, the percent predicted of FEV_1 and FVC were similar in both years. The improvement in the *z*-scores of BMI within 2 years and the medications used for respiratory functions may have preserved these functions.

Recombinant human DNAse (rhDNase) and inhaled hypertonic saline are known to be used in people with CF to increase mucociliary clearance, improve lung function, and reduce pulmonary exacerbations, regardless of severity, and studies showed that they are associated with a decrease in pulmonary function loss and improved survival.[16–18] In a comparative study of two countries, there were large differences between the use of medications such as rhDNase and hypertonic saline and it was thought that the differences affected lung function.[15] Our registry showed that the usage of rhDNase was over 90% in both years. In addition, it was shown that the use of inhaled hypertonic saline was statistically higher 2 years

later. It was predicted that increased use of hypertonic saline with increasing age, in particular, might be related to the reimbursement of the drug aged 6 years and older for the last 2 years in our country and to the easier access to the drug. The use of azithromycin also increased in the follow-up of the people with data in both years in our registry. It was thought that the increase in the use of these medications might also have positively affected the PFTs of the patients. It was observed that the use of bronchodilators also increased over the years. It was suggested that the use of bronchodilators beforehand had an effect on this increase to prevent the side effects such as bronchospasm of inhaled hypertonic saline.

The prevalence of complications has increased with improved life expectancy.[19] In the CFFPR, CF-related liver disease was found in 6.4% and 10.2% in 2017 and 2019, respectively. In addition, osteoporosis was detected in 3.8% and 3.9% in 2017 and 2019, respectively.[20,21] In our study, CF-related liver disease and osteoporosis were found to be increased over the years compared. This was thought to be due to the increasing age of the people because complications may increase with age. Also, the maturation of the CFRT by the time and improved data collection may have caused this. The use of ursodeoxycholic acid increased significantly over 2 years, and this may be related to the increased incidence of CF-related liver disease.

In CF, the balance of sodium plays an important role in weight gain during early life and sodium supplementation is recommended for all patients.[22] It has been widely accepted in studies that PBS tends to occur in infants or young children because they are more susceptible to body fluid loss and infectious diseases.[23] A study conducted in China found that a lack of CF awareness and neonatal screening led to the underdiagnosis of PBS, which is a major complication.[24] As seen in the first report of the registry system, and as stated in the studies in 2017, the most common complication was PBS among people with CF.[7,25] Although the exact mechanism of PBS was not known, it was reported in the study that the high prevalence of PBS in our country might be due to the hot weather conditions, as well as the younger ages of the people.[25] It is pleasing that PBS decreased over the years, according to the results of our study. This may be due to the increased awareness of PBS owing to the CFRT, and therefore, adding regular salt supplementation for our patients.

This study had some limitations. People with CF who were not involved in the registry system were overlooked and the study was retrospective. The CFRT is compatible with the ECFS registry system and only data entered in the CFRT are used. There is no auditing of the CFRT or any other quality control processes to detect data entry errors.

The increase in the *z*-scores of height and BMI of patients, increase in the use of medications that promote mucociliary clearance and improve lung function and decrease in the incidence of PBS show that the registry system may improve the care of people with CF. The widespread use of CF registry system all over our country might be beneficial for the follow-up of our patients. The CFRT is still under development and maturation, but early indications suggest that improved population coverage, data completion, and processes relating to feedback of data to CF centers are associated with improved clinical outcomes and clinical management of Turkish CF population.

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CONFLICT OF INTERESTS

The authors declare that there are no conflict of interests.

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Data is available on request due to privacy ethical restrictions.

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