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CLINICAL CORRESPONDENCE

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Collaboration between two CF centers; one in USA and one in Turkey before and during CoV2 pandemic

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Abstract

To address the discrepancy in the quality of care and outcomes between cystic fibrosis centers (CFCs) in high-income countries and limited resources countries (LRCs), a collaboration between our team at the University of Michigan CFC (UMCFC) and a CF center in Turkey (Marmara University CFC [MUCFC], Istanbul) was established. The collaboration included evaluation of all aspects of care and initiation of quality improvement (QI) measures. Teaching and implementing QI tools has led to start of improvement in MUCFC care. Close monitoring and sharing resources like UMCFC algorithms, protocols, and QI processes were done.

KEYWORDS
BMI, cystic fibrosis, FEV1, high-income countries, infection prevention & control, low resources countries, quality improvement

Cystic fibrosis (CF) has been shown to affect people from all ethnic backgrounds.[1] In Turkey, there are approximately 3000 people with CF (pwCF). Cystic fibrosis centers (CFCs) are supported by the Turkish government. Marmara University CFC (MUCFC) has been reporting to the European CF Society Patient Registry since 2015 and is one of the largest CFC in Turkey. CF clinical outcomes have been lower than the Western European Countries with disproportionate rate of morbidity and mortality, as indicated by the majority of pwCF are <18 years old (297 patients are 0–17 years old and 70 patients are ≥18 years old in 2020 for a total of 367).[2]

The most common mutations in Turkey are F508del, 1677delTA, N1303K. Median age at diagnosis and follow-up duration were 0.3 (interquartile range [IQR]: 0.2–0.9) and 11.9 (IQR: 5.4–16.1) years, respectively. Most CF medications are available in Turkey, and free for patients. There is a Turkish CF patient and family organization (KIFDER).

For comparison, University of Michigan CFC (UMCFC) follows approximately 275 pediatric patients and over 600 total pw CF including adults. The center data have been above average for US centers and are well supported with adequate staffing. The center is very active in research and quality improvement (QI) work.

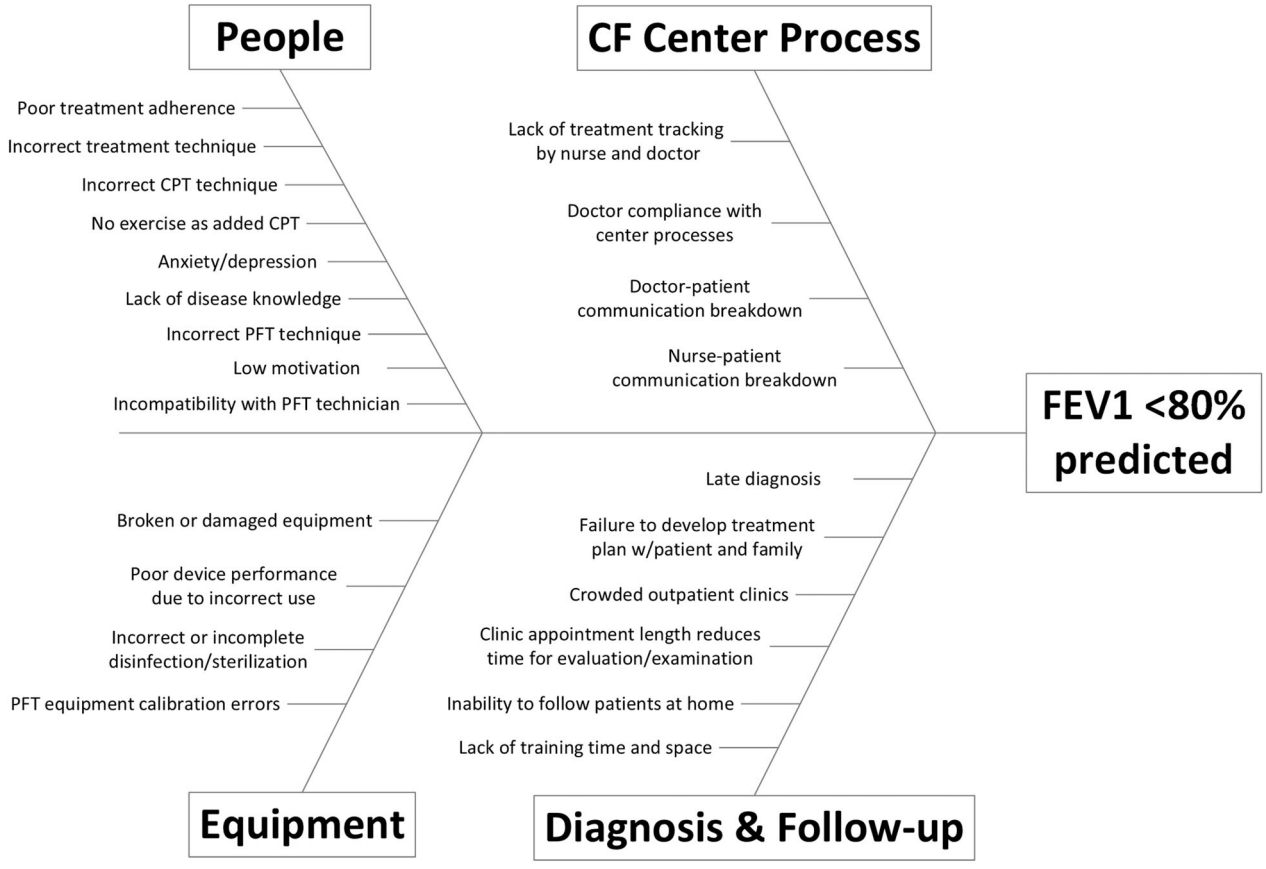
The collaboration between the two centers has been focusing on aspects of care that are identified as needing improvement and started with a site visit in 2018.[2] The following were noted:

1. A high number of pwCF had low body mass index (BMI) *Z*-scores: Median *Z*-score was −1.2 (25th and 75th: −2.2 to −0.2), −0.2 (25th and 75th: −1.5 to 0.4), and −0.6 (25th and 75th: −1.3 to 0.1) in patients 2–5, 6–11, 12–17 years old, respectively. The BMI *Z*-score was lower in all age groups compared with peers registered in ECFSPR.[2]
2. Median forced expiratory volume at 1 second (FEV1) pp was 86.1 (25th and 75th%:70.5–97.8) in 6–17 years compared with 60.8 (25th and 75th: 40.5–73.3) in patients 18–24 years old.
3. Inadequate integration of MUCFC with Physical Therapy and Rehabilitation Department.

4. The center had no infection prevention and control policies in place.
5. Although there was an outpatient clinic schedule in place, patients with mild to moderate CF exacerbation were seen without a scheduled appointment.
6. Inadequate Allied Health professionals (nurses, dietitian, and physical therapy) support to the center.

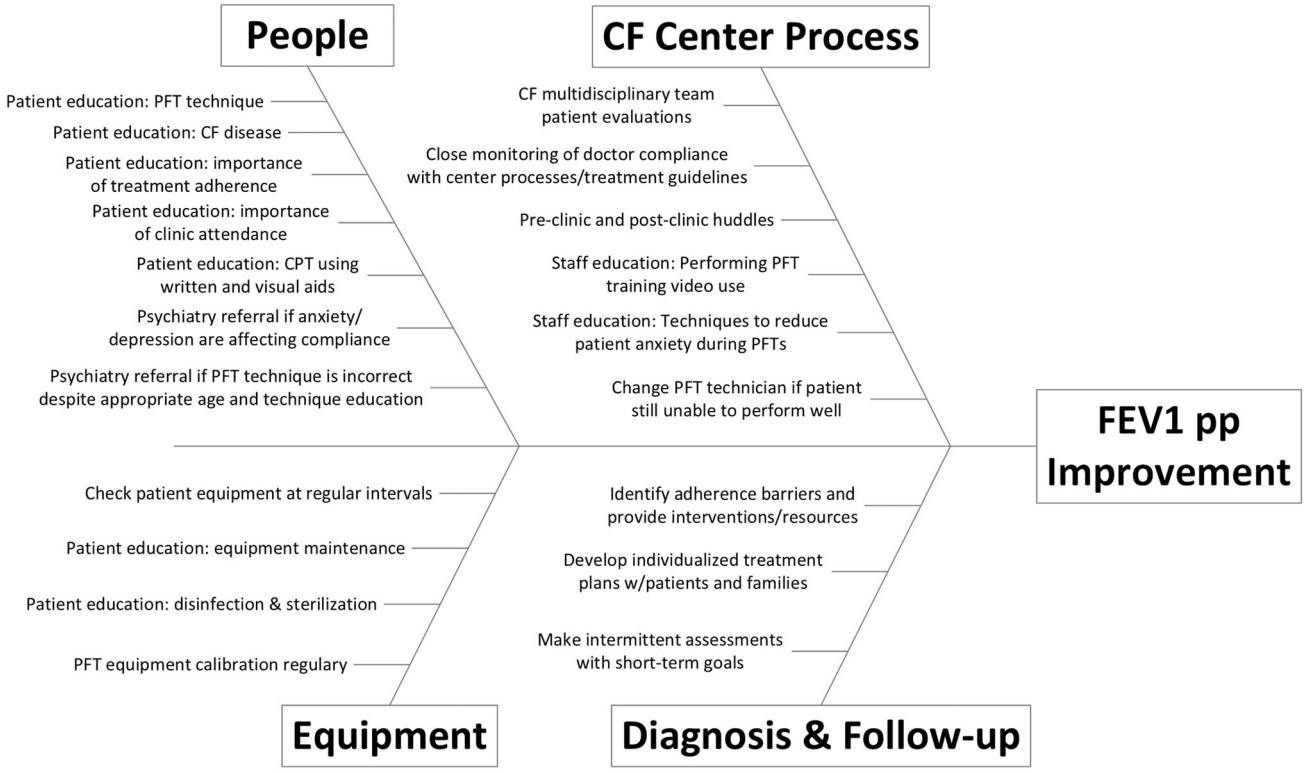
The strategies used throughout the collaboration were based on QI principals. Areas that needed improvement were pointed out and solutions, based on their resources and practices were proposed including creating Fishbones, flow charts, Plan Do Study Act (PDSA) cycles, PDSA ramps, and other QI tools to address each deficiency. Training and engaging the MUCFC team in QI methodology were done. Creating and implementing a standardized CF care algorithm and individualized treatment plan for each patient were developed, to address barriers to adherence that may have contributed to poor outcomes. Weekly case review of patients with BMI ≤ 50% and FEV1 < 80 pp and adjustment of the PDSA cycles were done. Two fishbone diagrams (Figures 1 and 2) were created, to identify the root cause of the low FEV1 and how the FEV1pp was improved. The QI methods used were UMCFC' algorithms and protocols.[3,4] Engaging and educating pwCF and their families as partners in their care through collaboration with KIFDER was crucial to the success of this study.

The collaboration included in person site visit by the UMCF center director in 2018 followed by a visit from the team (center director, dietician, respiratory therapist, and physical therapist) to MUCFC in 2019, visit from the MUCFC team to the UMCFC (center director, three pediatric pulmonologists, dietician, and physical therapy and rehabilitation specialist) in late 2019 and a rotation for the MUCFC fellow to train at the UMCFC in 2019. In 2020 and 2021, due to the coronavirus virus 2 (CoV2) pandemic, webcast meetings have been ongoing between the two centers to continue with follow-up on progress and QI efforts.



CPT – Chest physiotherapy
PFT – Pulmonary function test
CF – Cystic fibrosis
FEV1 – Forced expiratory volume in 1 second

FIGURE 1 Identification of factors causing FEV1 < 80% predicted



FEV1 – Forced expiratory volume in 1 second
PFT – Pulmonary function test
CF – Cystic fibrosis
CPT – Chest physiotherapy
pp – Percent predicted

FIGURE 2 Ways to improve FEV1 percent predicted

Table 1 summarizes the patient population demographics and clinical characteristics from 2020 European Cystic Fibrosis Society Registry. MU Institutional Ethics approval was obtained.

After the initial UMCFC director meeting, appropriate examination rooms were allocated to the center. A strict IP&C policy was initiated, in the outpatient and inpatient settings and a second CF nurse was added to the team. Even though the importance of teamwork is best learned by the whole team, the fellow's visit to UMCFC was a good opportunity for training and working with the UMCFC multidisciplinary team. The process was relatively time consuming at the first year of the collaboration. That included analyzing MUCFC's data, pointing out to areas that needed improved, teaching them QI processes, sharing algorithms and Clinical Practice Guidelines, developing flow charts and help design QI projects, and reassessing and readjusting them moving forward. Following that, regular interactions were done virtually to follow up on the progress. To address the decreased FEV1 for pwCF, UMCFC standardized patient respiratory care protocols and algorithms were adjusted and implemented. The focus of the first QI project was CF patients 6–18 years old with FEV1 pp <80. The project was done from June 2019 through October 2020. Baseline, 6th and 12th months mean FEV1pp was 63.7 ± 14.6, 66.9 ± 16.6, 70.4 ± 19.2, respectively with improvement of 5.0% in 6 months and 10.5% in 12 months ($p = 0.004$).

With the increase in the dietitian's support, they learned additional evaluation tools for a more in-depth assessment of patients' nutritional status, including pediatric malnutrition criteria. In addition, a QI project was started to help improve the nutritional status of pwCF. Recommendations for increasing caloric intake were made as indicated, after evaluation of the food consumption records of the patients. Oral nutritional supplements were recommended, pancreatic enzyme replacement therapy was maximized, and nutrition education was given to pwCF and their families.

Cyproheptadine as an appetite stimulant was started to be used for children in the higher risk group as defined by the QI project protocol.[3] When enteral nutrition was indicated, pwCF and families were informed and the plan was discussed in detail with them to gain their agreement, however, this intervention was not well received. Table 2 shows the summary of the results for BMI percentile and FEV1 pp at baseline, 6, and 12 months, respectively. The table included only pwCF that completed both QI projects combined. Not all pwCF were included in the QI projects. Because of the pandemic,

TABLE 1 Demographic and clinical characteristics of pw CF at MUCFC (2020 ECFSR data) (n = 368)

Median age (IQR)	11.9 (5.4–16.1)
Median age at diagnosis (IQR)	0.3 (0.2–0.9)
Male/female *n* (%)	205 (55.7)/163 (44.3)
Children/adult *n* (%)	297 (81.0)/70 (19.0)
Median FEV1pp (IQR)	
Age groups (years)	
6–17	89.3 (73.5–125.4)
≥ 18	55.6 (40.9–83.3)
Median Z-score for BMI (IQR)	
Age groups (years)	
2–5	−0.4(−1.2–0.3)
6–11	−0.5 (−1.4–0.4)
12–17	−0.3 (−1.1–0.4)

Abbreviations: BMI, body mass index; CF, cystic fibrosis; ECFSR, European Cystic Fibrosis Society Registry; IQR, interquartile range; MUCFC, Marmara University Cystic Fibrosis Center; pwCF, people with CF.

TABLE 2 Comparison of pwCF that completed both QI projects combined at baseline, 6th, and 12th month weight, weight percentiles (weightp), body mass index (BMI), BMI percentile (BMIp), forced expiratory volume in 1 s percent predicted (FEV1pp), and FEV1pp < 80

	Numbers of pwCF completed the three visits	Baseline	6th month	12th month	*p*
Weight	109	29.1	32.4	34.0	**<0.001**
Weightp	109	28.8	33.9	38.5	**0.017**
BMI	109	15.9	16.6	18.7	**0.001**
BMIp	109	24.6	33.8	37.0	**<0.001**
FEV1pp	60	82.4	82.1	86.8	**0.03**
FEV1 < 80 pp	14	59.9	61.8	63.3	0.13

Note: Bold values are statistically significant $p < 0.05$.

Abbreviations: QI, quality improvement; pwCF, people with CF.

only the repeated measures for the patients who were seen at the three points were analyzed using Friedman's analysis measures.

To address the poor adherence to airway clearance techniques, patients' low activity levels, and poor adherence to the percussion and postural drainage, the two teams worked together to improve patient information materials, including developing booklets for patient education that also included section on improving hygiene and nebulizer cleaning and disinfection.[5] The physical therapy team started to follow patients closely in the outpatient and inpatient settings and added evaluation, counseling, and treatment of vestibular dysfunction symptoms and bowel and bladder incontinence symptoms.

Annual depression and anxiety screening of pwCF and their caregivers was started via standardized questionnaires and patients with a high level of depression and/or anxiety levels were referred to the child psychiatry department for evaluation and treatment.[6]

The collaboration has been successful because of the two teams' willingness to work together to create an effective model of collaboration. Each team brought varying strengths and resources to the partnerships, but they also brought their own organizational cultures, regulations, and expectations, that led to the adjustment of the guidelines and protocols to accommodate for their needs. MU administration was also willing to work with the teams to improve care which led to providing MUCFC with much needed resources.

The use of mobile technology, especially during the CoV2 pandemic was crucial, to the continuation of the collaboration between the two teams and to the facilitation and improvement of the communication between the MUCFC team and their patients and families. The extension of the collaboration beyond the scheduled 2 years has been fruitful in strengthening the relationship and the QI work.

To create a model of care at MUCFC, the in-person visits by both teams were important in evaluating the areas that needed improvement and following the progress at MUCFC and provided a look for MUCFC at our center's structure and operation.

Data sharing between the two centers was an important part of the collaboration. Teaching and implementing QI tools have led to improvement in all MUCFC aspects of care and the improvement has been continuing.

Next steps in the collaboration are: (1) to work with the Turkish Thoracic Society and Ministry of Health to update the national CF guideline, that was recently done and will be shared with other centers across the country; (2) to create National CF Center Network in Turkey, using evidence-based, state-of-the-art healthcare delivery, operating under quality improvement principles; (3) to function as a resource to other centers in Turkey and in the region. This partnership can be viewed as an example of collaboration that could be carried out to a certain degree in other Middle East Countries and limited resources countries (LRCs) to deliver better CF care. We believe that more collaborations between CF centers in high-income countries and LRC could lead to significant improvement in CF care worldwide.

AUTHOR CONTRIBUTIONS

Samya Z. Nasr: Conceptualization, manuscript writing, and literature review. All other authors: Helped in manuscript writing, and literature review and critically reviewed the final version of the manuscript and approved for the publication.

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CONFLICT OF INTEREST

All authors declare no conflict of interest.

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