BRIEF REPORT

Telemonitoring in Cystic Fibrosis: a single centre experience in a Tertiary Paediatric Hospital

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ABSTRACT

Telemedicine during Covid-19 pandemic became a necessity to reach and follow up patients with chronic diseases. During this emergency period patients with cystic fibrosis required a follow-up reorganization to stay safe and to prevent cross-infections. The aim of this study was to explore the adherence to a telemonitoring program, patients' clinical status and to measure the rate of satisfaction related to our telemedicine program thought for patients with cystic fibrosis enrolled in a tertiary paediatric hospital.

Fifty patients (M 29 age mean 35,6 y) were enrolled in a study period from 2020 to 2022 with a rate of transmission data of twice a week; 30 of them were monitored also for lung function with the MIR Spirobank (spirometer).

The majority of data sent was during the Covid-19 pandemic with 1605 records forwarded; adherence was better particularly in females, and patients who lived farther away from our center. The FEV_1 mean value was 74.8%, 74.2% and 74.0% in 2020, 2021 and 2022 respectively. Average response rate about satisfaction and utility of the offered telemedicine service was respectively 4,03 and 4,5 on a 5-point Likert scale.

Thanks to transmission of spirometric values in 30 patients, we have intercepted indirectly 14 potential bronchopulmonary exacerbations, studying the delta variation between spirometries executed along the pandemic period and the previous consecutive values, preventing the use of unnecessary antibiotics.

Telemedicine could be a useful tool to be included in the follow-up of CF patients that could help families lowering the costs for frequent travels and containing the burden of healthcare management. Future efforts in chronic disease management should improve the use of telemedicine.

IMPACT STATEMENT: This brief report has the aim to underline the role of telemedicine in cystic Fibrosis not only during Covid-19 pandemic period.

KEY WORDS
Telemonitoring; Cystic fibrosis; Covid-19; telemonitoring; telemedicine; sensor technology.

BACKGROUND

Cystic fibrosis (CF) is the most common autosomal recessive genetic pathology of the Caucasian race and it affects nearly 100,000 people worldwide (many have not been diagnosed) and, in Italy, we count around 6000 patients (1). Functional failure of CF leads to multisystemic dysfunction, such as lungs, gastrointesti-

nal tract, liver and pancreatic gland. Impaired mucociliary clearance and dense secretions primarily result in chronic pulmonary inflammation and infections, irreversible lung architecture modification, respiratory failure and death (2). Most people with CF will require lifelong treatment involving frequent hospital visits and admissions and rigorous daily therapy regimens (oral treatment, aerosol therapy, airway cleaning, and also physical activity rehabilitation) (3). Since many of these treatments necessitate frequent and continuous hospital visits for assessment and ongoing management, telemedicine and telemonitoring may be useful. Telemedicine is defined as a direct, synchronous, or remote communication between a physician and a patient (4). Telemedicine could potentially supply remote specialistic performance such as cardiological, pulmonary or rehabilitation service. In respiratory diseases, the telemonitoring was demonstrated to be able to identify early changes in patients' conditions; this is important to guarantee specific interventions and to avoid pulmonary exacerbations (5, 6). The role of remote telemonitoring had been of crucial importance during Covid-19 (7, 8, 9, 10, 11). The pandemic reality has underlined the extreme necessity of promoting remote monitoring for chronic conditions like cystic fibrosis, and it has shown how the use of telemedicine could prove to be extremely useful in this emergency (8, 9).

The primary objective was to compare the adherence to our telemedicine program during the whole study period, including the national pandemic of Covid-19. The secondary aims were to assess the clinical status of patients in relation to: lung function, number of hospitalizations for bronchopulmonary exacerbation and the need for extra visits or hospitalizations from 2020 to 2022, and in details during Covid-19 pandemic. Finally, we aimed to explore impression and satisfaction of patients related to our telemedicine program through a questionnaire distributed to patients.

MATERIALS AND METHODS

A longitudinal observational study was conducted at Bambino Gesù Children's Hospital during a period from 2020 to 2022.

Patients with Cystic Fibrosis (pwCF) were assisted by home tele-monitoring twice a week with remote transmissions of spirometric values and remote consultations with a specialized physician, respiratory physioterapist and research nurse. Demographic data entered by the user is used to calculate percent predicted values based on the Global Lung Function Initiative (GLI) (12).

Acceptable tests were compared to one another and were graded according to ATS guidelines (13).

The MIR Spirobank was used for telemonitoring in our patients, a portable spirometer that allows the transmission of data regarding the respiratory function to health specialists (nurses and physicians) and in some cases also tablet connected with Bluetooth and pulse oximeter. The program of remote telemonitoring includes two trasmissions per week and after each, the research nurse could call the patients or their parents to discuss the values of the spirometry executed.

RESULTS

Characteristics of 50 pwCF enrolled in the study are shown in **Table 1**. A total of 30/50 pwCF sent telemonitoring data regularly, also during Covid-19 pandemia.

In 2020, at the beginning of Covid-19 pandemic we recorded a peak of the number of telemedicine transmissions with a total of 1605 records submitted (**Figure 1**). On the contrary, during the last year of follow-up patients sent spirometries less frequently.

Looking at average trasmissions during and after the Covid-19 period (2020-2021) we found that the aver-

Table 1. *Patients characteristics.*

	Mean and/or N (%)
Patients characteristics	
Number of patients (M/F)	50 (39 M/21 F)
Age (years)	30,1
Staphylococcus colonization	36 (71%) 12 (24%)
Pseudomonas aeruginosa colonization	12 (24%)
Burkholderia Cepacia colonization	2 (5%)
Modulator	
Elexacaftor-Tezacaftor-Ivacaftor	27 (47%)
Compassionate use of Kaftrio	1 (3%)
Ivacaftor	8 (18%)
Without CFTR modulator	14 (28%)
Provenance	
Lazio region	34 (68%)
Other regions	16 (32%)

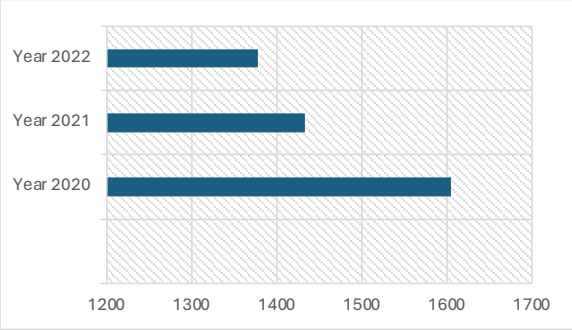

Figure 1. *Number of telemonitoring transmissions from 2020 to 2022.*

age of transmissions during the pandemia was 1,76 per week with a progressive decrease coming out from the pandemic period arriving at 0,86 (average of transmissions, data from 2022) (**Figure 2**).

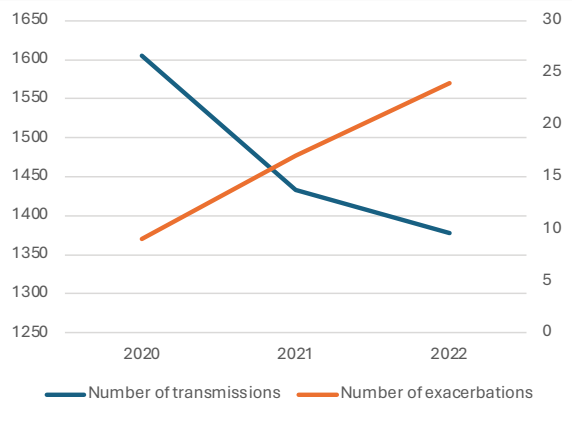

Figure 2*. Average number of transmissions and pulmonary exacerbations from 2020 to 2022.*

Adherence to trasmission was higher in females than males, 76% and 62,5% respectively. Patients living in other regions than Lazio had a higher average rate of transmission (1,54 transmission per week versus 1,21 transmission per week respectively).

Parents of paediatric patients sent transmissions more frequently than adolescents and adults. Patients living closer to the hospital (Lazio Region) progressively reduced the number of transmissions with a 25% reduction in particular after the period of Covid-19 pandemia).

The FEV_1 mean value was 74.8%, 74.2%, 74.0% and 76.9% respectively during 2020, 2021, 2022 and in the first 3 months of 2023.

Considering lung colonization, we recorded a lower FEV_1 in patients with *Pseudomonas aeruginosa* compared to patients with Staphylococcus aureus: FEV_1 mean 72,4 and 78%, respectively.

In 2021 we registered a decreased need for outpatients visits (-30%) comparing with 2022 year when there was a reduction of remote data transmission.

Thanks to the available spirometric values, we intercepted 9 pulmonary exacerbations during 2020, 17 during 2021 and finally 24 during 2022.

Regarding pwCF (30/50) who were in follow-up in telemedicine during Covid-19 period, a significant number (13,43%) was from other regions than Lazio (17,57%), and in particular from Calabria (3,10%), Molise (2,7%), Puglia (1,3%), Abruzzo (2,7%), Umbria (1,3%), and Campania (3,10%).

All those patients (30/30) could perform a home-spirometry, and 7 patients (23%) also had pulse oximeter and tablet for data transmission and to access to telemedicine platform; on the other hand 3 (10%) patients had only spirometer and tablet. 11/30 (36,6%) completed data trasmission twice a week, 12/30 (40%) patients once a week and only one patient (3%) sent spirometry every day. The rest of 6 patients (20%) preferred to send data after medical contact or for clinical need. Of these subgroups of 30 patients: 15 (50%) are in our telemedicine program from less than 3 years; 8 (26,7%) patients between 4 and 6 years, and 7 patients (23,3%) for over 6 years.

Those 30 patients were asked to answer to a specific questionnaire to explore their satisfaction and if they want to change something in telemedicine/telemonitoring service. 23/30 (76,6%) patients considered that telemonitoring service helped them to improve compliance to their daily therapy. Moreover, 26 patients (86,6%) reported that this program is helpful to intercept bronchopulmonary exacerbations.

18/30 (60%) would not want to change anything about the service, only 5 patients (16,6%) want to change the platform because considered it unclear and 7 patients (23,3%), instead, wanted to change both spirometer and platform. During the pandemic period the majority of patients (25, 83,3%) referred to feel more safe being part of the telemonitoring program which could them to prevent possible infections.

Finally, 24 patients (80%) reported that the telemedicine program improved their quality of life and that they felt more safe.

Average response rate about the satisfaction of the service was 4,03 on a 5-point Likert scale.
Average response rate about the utility of telemonitoring service was 4,5 always using the Likert scale.

DISCUSSION

The use of telemonitoring contributed to improve the quality of care during the pandemic period, and allowed our pwCF to reduce their need of hospital visits avoiding the risk of contracting the infection. As expected, number of patients admission after outbreak Covid-19 returned at the same level but during Covid-19 period telemedicine was a valid instrument to rationalize hospital access.
Telemonitoring could change the approach and management to chronic pulmonary diseases, and this seems valid also for pwCF. In fact, due to telemonitoring spirometric values, we intercepted 14 possible bronchopulmonary exacerbations, in particular considering a significant decrease of FEV_1 values between two consecutive spirometries. We could prevent the use of unnecessary antibiotics (intravenous or oral) in pwCF, stimuling patients to do respiratory physiotherapy and also underling the importance of physical activity increasing volume and number of training sessions (14). In particular these interventions could prevent a possible pulmonary damage. One important aspect about this tele-monitoring project is rationalization of hospital access, in fact patients decreased their hospital access, and in some cases, they transformed intravenous antibiotic treatment with outpatient access because the medical team stopped previously the possible exacerbations (15).
Probably the peak of adherence during the first year of pandemic underlines the priority for patients to stay safe and away from hospital, if their clinical conditions allowed it. The most important aspect was to intercept pulmonary exacerbations, in particular for patients with a severe lung damage. This is the most important result that underlines how this service can improve care for pwCF. In fact pulmonary exacerbations in CF are crucial events which progressively determine a loss of respiratory function, worsening of the quality of life and negatively impact overall survival. According to studies, lung function fails to return to baseline value in up to 25% of CF pulmonary exacerbations, despite a prompt antibiotic treatment (16, 17).
Patients thanks to telemonitoring have addressed more secure the pandemic years.
Telemedicine system could become a useful tool to monitor patients in a new CFTR modulator era where there are a reduced number of pulmonary flare ups and a new approach to disease with more number of outpatient controls (18). In particular, in Italy, Cystic Fibrosis centers are distributed mostly in large cities, and the use of telemedicine could cut the economic costs of patients travel and thus increase adherence to not postponeable checkups.
Despite the recommendation of physicians to transmit regularly, some patients were scared from the possible results obtained, infact FEV_1 deterioration in some patients brings a lot of psycological distress.
On the contrary, home monitoring might bear the risk of increased anxiety caused by increased awareness of deterioration in lung function in some patients (19). However, our experience showed that patients with CF considered our program to be useful to keep up with their daily treatment and let them feeling safer and more controlled.

CONCLUSION

In conclusion new forms of health care are to be evaluated for their impact on patients' health and also for their cost-effectiveness compared to traditional care, and telemedicine with tele-monitoring could be a passage essential to change the management of chronic diseases like Cystic Fibrosis. In summary, this experience offers the opportunity and consciousness to improve the use of telemedicine and to evaluate the use of telemedicine like a fundamental tool to manage pwCF in routine care, also reducing cost-effectiveness of medical treatments and high burden of health-care management.

COMPLIANCE WITH ETHICAL STANDARDS

Conflict of interests

The Authors declare no conflict of interests.

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Authorship

Matteo De Marchis and Alessandra Federici.

Author contributions

Conceived the report: MDM, AF and SB; collected data: MDM and AF; wrote the manuscript: MDM, AB and NU; conceived and directed the work thanks to their knowledge of the subject matter: AGF and RC. All

Authors analyzed and interpreted patients' data read and approved the final manuscript.

Ethical approval

Human studies and subjects

Ethics approval for this study was obtained from Bambino Gesù Children's Hospital IRCCS, Rome, Italy.

Animal studies

N/A.

Data sharing and data accessibility

The data presented in this study are available on request from the Corresponding Author.

Publication ethics

Plagiarism

All original studies are cited as appropriate.

Data falsification and fabrication

All the data correspond to the real.

Manipulation of images

All images are original.

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