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LETTER TO THE EDITOR

Clinical characteristics and outcome of SARS —CoV-2 infection in patients with cystic fibrosis managed at home

Lung disease in Cystic Fibrosis (CF) is characterised by bronchiectasis with persistent airways-based infection and inflammation and remains the main cause of morbidity and mortality.[1]

Severe acute respiratory syndrome coronavirus-2 (SARS-CoV-2), associated with the ongoing coronavirus disease 2019 (COVID-19) pandemic, has had a huge impact on world population. The presence of co-morbidities, such as CF, has been identified as a risk factor for severe disease.[2,3] The incidence is higher in people with CF versus the age-matched general population and significantly higher rates of admission to hospital and higher rates of intensive care have been recently reported, especially in patients receiving an organ transplant.[3] Mild illness was reported in CF children who did not have pre-existing severe lung disease.[4] Furthermore, the COVID-19 pandemic has created great interest in the use of telemedicine in CF patients since it can be a valid tool to assess their clinical condition.[5]

In this paper we evaluated clinical presentation, management and outcomes of CF patients with SARS-CoV-2, managed at home thanks to telemedicine.

We retrospectively reviewed clinical charts of all CF patients diagnosed with SARS-CoV-2 infection via a positive nasal/throat polymerase chain reaction (PCR) test and followed-up at CF centre of Florence, Italy, where we take care of both paediatric and adult patients.[6] Cases were recorded up to 30 June 2021. We enrolled only CF patients managed at home, thanks to telemedicine consultations. Diagnostic tests were performed where there were symptoms or in asymptomatic cases if the patients were at risk for positive familial or at work contact. We also compared, pre and post infection, the trend of body mass index (BMI, expressed as centile in patients younger than 20 years), or of the weight/length centile for infants and percentage predicted forced expiratory volume in one second (FEV_1) for patients aged 6 years and older. Data collected also included CFTR genotype, pancreatic and microbiological status, age at SARS-CoV-2 infection, pre-existing CF related diabetes (CFRD).

The study was approved by the Ethics committee (Florence, Ethics Clearance number 217/2021, on 7 September 2021) and we obtained from all patients (or from their legal guardian) their informed consent to allow the use of anonymous clinical data for research purposes.

Telemedicine consultation took place immediately following COVID-19 diagnosis and during the course of the infection [phone call, monitoring of oxygen saturation (SpO2) and screening for pulmonary symptoms suggestive of COVID-19]. In absence of criteria for hospitalisation, we advised the isolation of the patient and his family, the use of a home pulse oximeter and in the case of a reading of less than 92% or respiratory distress signs, the need for hospitalisation.

Eighteen (5.1%) out of 352 CF patients followed at our Regional centre suffered from SARS-CoV-2 infection. Thirteen (72.2%, 10 males, mean age at SARS-CoV-2 infection: 27 years, range 3 months-59 years) out of 18 were managed at home. We excluded 5 patients, 2 adults who needed hospitalisation due to lung transplant in 2 and 3 more cases with persistent fever with SpO2 $<$ 92% .

Key characteristics and outcomes of enrolled CF patients diagnosed with SARS-CoV-2 infection are reported in Table 1. Nine (69%) out of 13 had pancreatic insufficiency. No patients had CFRD.

We compared the FEV_1 values and BMI or BMI centile obtained at a mean period of 33 days (range 17-50 days) before infection and at the first visit after recovery (negative PCR test), performed after a mean period of 50 days (range 7-116 days). No significant worsening was reported (Table 1).

Unlike the children described by Bain R et al,[4] only one child aged 4 years needed antibiotic and corticosteroid medication for increased cough and wheezing in the first 24 hours. Similarly, two adult patients were given antibiotic therapy because of increased coughing. All enrolled patients had normal values of SpO2 (96-98%).

Male gender, CFRD and being over 50 years of age have been shown to be associated with more severe SARS-CoV-2 infection in the general population.[2,7,8] In this small cohort we report a mild course of SARS-CoV-2 infection, despite 3 patients in our cohort being older than 50 years and 10 (77%) out of 13 patients being males. In addition, we report 7 more cases of CF patients with asymptomatic SARS-CoV-2 infection,[4] among them an adolescent and an adult patient, both with severe lung disease (case 2 and 3 of Table 1).

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Table 1 key characteristics and outcomes of enrolled CF patients diagnosed with SARS-CoV-2 infection.

Patient	First CFTR variant	Second CFTRvariant	Age atSARS-CoV-2 infection(years)	Symptoms	Microbiological status	Pre infection FEV_1 (%)	Post infection FEV_1 (%)	Pre infection BMI [a]	Post infection BMI [a]
1	2789+5G>A	1602delCT	54	Fever	*Burkholderia gladioli*	33	36	27.67	27.67
2	F508Del	CFTR Dele 2	14	None	MSSA	40	37	36.88	80.5
3	F508Del	N1303K	26	None	MSSA	53	53	25.51	25.21
4	E585X	Dele 22-24	10.5	Cough	MSSA	86	95	52.33	60.42
5	R347H	G542X	59	Myalgia, fever	MSSA	51	63	21.89	21.74
6	F508Del	A1006E	4.5	Cough, wheezing	MSSA	na[b]	na[b]	61.68	50.75
7	F508Del	(TG)12T5	10	None	MSSA	126	98	84.14	76.53
8	F508Del	G542X	30	None	*Stenotrophomonas maltophilia*.	72	76	19.72	20.26
9	G178R	L1065P	40	Cough, fever myalgia	MSSA	87	92	26.96	26.6
10	F508Del	G542X	3 months	None	Normal flora	na[b]	na[b]	0.65[c]	1.24[c]
11	F508Del	D192G	57.5	Cough	*Pseudomonas aeruginosa*	44	42	19.14	19.53
12	F508Del	F508Del	31	None	MSSA	61	65	20.27	20.13
13	F508Del	L1065P	13.8	None	MSSA	72	77	65.09	52.79

Abbreviations: SARS-CoV-2: severe acute respiratory syndrome coronavirus-2; MSSA: Methicillin-susceptible *Staphylococcus aureus*; $FEV_{1:}$ predicted forced expiratory volume in one second; BMI: body mass index; CFTR: cystic fibrosis transmembrane conductance regulator; na: not available

[a] we report BMI data in patients younger than 20 years as BMI centile

[b] children aged < 6 years

[c] we refer to weight/length centile given the age of the child

Finally, we highlight a higher prevalence of SARS-CoV-2 infection, constantly increasing, in CF patients compared to previous studies.[2,3] However, no cases of infection occurred in the early period of the pandemic, probably due to the greater restrictive measures adopted in Italy in that period.

Management at home reduced the risk of hospital cross-infection and avoided hospital overcrowding.

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Conflict of Interest Disclosures

The authors declare no conflicts of interest relevant to this article to disclose.

A data availability statement

All reported data are available.

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