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activity. Indeed, mitochondrial activity that is altered in CF plays a crucial role in epithelial repair, and monitors cellular ATP and pH, both involved in ciliary activity and mucin structure.

Methods: The studies were performed on primary culture of nasal epithelial cells (hNEC) from patients with CF and CFBE410- bronchial epithelial cell line. We used multiscale differential dynamic microscopy to assess cilia beat frequency and coordination. Mucins production and localisation were investigated using RT-qPCR and confocal microscopy. Airway epithelium repair was studied using the Incucyte life-cell imaging analyser. Mitochondrial activity was investigated using the Seahorse extracellular flux analyser.

Results: Our results suggest that SPMs increased ciliary beat frequency in CF hNEC primary cultures. Several SPMs stimulated CFBE410- cells repair with various efficacy. SPMs did not significantly affect basal mitochondrial respiration of CFBE410- cells. However, specific SPMs restored mitochondrial respiration after TNF- α induced inflammation and alteration of mitochondrial activity.

Conclusion: Our first results provided evidence of a role for several SPMs in enhancing mucociliary clearance, epithelial repair, and mitochondrial activity with various efficacy. This highlights a possible therapeutic benefit of some SPMs in the CF airway disease.

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SARS-CoV-2 infection in cystic fibrosis during the first pandemic wave in Italy: a multi-centre prospective study with a control group

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Background: Patients with cystic fibrosis (CF) are at high risk of developing severe forms of viral respiratory infections. This study aimed at comparing symptoms and clinical course of SARS-CoV2 infection with other respiratory infections in patients with CF.

Methods: We carried out a prospective multicentre cohort study within the Italian CF Society involving 32 CF centres following 6,597 patients. CF centres were contacted to collect baseline and follow-up data of all patients who had reported symptoms suggestive of COVID-19 or who had had contact with a positive/suspected case between the end of February and July 2020. Symptoms and clinical course of the infection were compared between patients who tested positive by molecular testing (cases) and those who tested negative (controls).

Results: Thirty patients were reported from the centres, 16 of whom tested positive and 14 negative. Fever, cough, asthenia and dyspnea were the most frequently reported symptoms and their frequency were not significant different between groups. Eight cases (50%) were hospitalised but none required ICU admission. Two adults with a history of lung transplant required non-invasive ventilation; none required ICU admission. All patients fully recovered without short-term sequelae. Changes in FEV₁ (percent of predicted) after recovery were not significantly different between groups (median, interquartile range: 3.0%, -1.5, 5.5 among cases and -3.0%, -8.5, 6.3 among controls, $P = 0.48$).

Conclusions: Symptoms and clinical course of SARS-CoV-2 infection in our patients was not significantly different from other respiratory infections. The clinical course of COVID-19 was relatively favourable, however CF patients with severely impaired respiratory function and organ transplant

may develop complications and a negative outcome. The study is ongoing, and we are recruiting patients during the second wave of the pandemic.

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SARS-COV-2 infection in patients with cystic fibrosis

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Introduction: Patients suffering from cystic fibrosis (CF) are more susceptible to recurrent or persistent pulmonary infections, which limit their lungs' capacity and can endanger their life. The evolution of SARS CoV-2 infection in patients with CF is a subject of interest and needs further research.

Aim: Evaluation of clinical signs of SARS CoV-2 in patients with CF.

Methods: We evaluated 3 patients with CF, who supported COVID-19 during the July 2020 - January 2021 period. Patients were tested by RT-PCR test when they presented symptoms.

Results: The COVID-19 diagnosis has been confirmed in two male patients (7 and 10 years old) and a female patient of 30 years old. Fever, cough, and dyspnea were the common signs, which indicated an exacerbation of pulmonary infection. The child of 7 years with CF developed a mild form of infection and was treated at home, while the other two patients were admitted to hospital with severe infectious exacerbations. The hospitalised child developed SARS CoV-2 infection in the context of chronic infection with *Pseudomonas aeruginosa* and required O₂-therapy due to decreased SpO₂ (92–93%). The adult woman with CF and chronic lung infection with *Staphylococcus aureus* showed a severe form of COVID-19 and pulmonary exacerbation. All the cases presented had a favorable evolution.

Conclusions: The presence of comorbidities in patients with SARS CoV-2 is a major risk factor, and in CF patients with chronic pulmonary manifestations, infection with SARS CoV-2 infection support severe course of disease with intensive inpatient treatments.

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Relationship between cystic fibrosis disease severity and susceptibility to COVID-19 infection

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Objective: Since cytokine storm and hyperinflammation play a key role on CF disease severity, severe CF patients should be considered to have an increased risk of developing severe symptoms of COVID-19. In this preliminary study, we evaluated whether there was a relationship between clinical severity of CF siblings and their susceptibility to COVID-19 infection.

Method: In our preliminary study, we used a targeted transcriptomic approach (CF Profiler Array) obtained from nasal samples of three families who had CF siblings harbouring same mutation but showing different severity of CF phenotype. The siblings were classified as severe or mild CF according to recurrent lung infection, hepatic involvement, and FEV₁.

Results: In severe CF patients (n=4) compared to mild patients (n=3) CXCL1 (FC:-3.53), CXCL2 (FC:-2.16), CXCL8 (FC:-5.41), IL1B (FC:-2.61), SERPINA1 (FC:-2.54), TNFSF10 (FC:-1.73) were found to be downregulated. CXCL1, CXCL2, CXCL8 play critical role during infection control in neutrophils that release other chemotactic mediators and recruit leukocytes. Additionally IL1B and TNFSF10 also affect activation of leukocytes. In the case of COVID-19 infection, the expression of these genes increases and leads to a cytokine storm. However our results show that CXCL1, CXCL2, CXCL8, IL1B genes which have a function in IL-17, NFKB, NLRP3 signaling pathways are downregulated in severe CF patients. Significant evidence supports the role of IL-1B, NLRP3-dependent inflammasome activation which is a central mediator of severe COVID-19 in the pathogenesis of acute lung injury. However, downregulation of inflammatory pathways is detected in severe forms of CF.

Conclusion: The results of our preliminary study strengthens the hypothesis that severe forms of CF may constitute an advantage to mild forms of CF in susceptibility to COVID-19 and CXC inhibitors may be a

promising therapeutic option for COVID-19 in the future. Further analyses should be performed with larger sample sizes.

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COVID-19 in cystic fibrosis patients with and without lung transplantation: the Zurich cohort

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Objectives: Reports on short- and long-term presentations of COVID-19 in CF patients is limited. We aimed to describe clinical features and outcomes of all our CF patients with laboratory confirmed COVID-19 between March 2020 and January 2021 (ongoing observation).

Methods: Retrospective review of clinical data and treatment of CF patients with COVID-19 confirmed by RT-PCR or serological evidence (n = 11).

Results: Mean age at presentation was 32 (23–48) years, 27% (n = 3) were female. Six patients (55%) had a previous lung transplantation (post-Ltx). Two patients were on modulator therapy (Trikafta[®] and Symdeco[®]). The most common presenting symptoms were cough (36%), fever (27%), headache (27%) and dyspnea (18%). Nine patients (82%) had mild disease and were treated as outpatients. Two patients (18%) were post-LTx and were hospitalised with severe disease (1 on the normal ward, 1 in the intensive care unit). The most notable laboratory findings were lymphopenia, and elevated levels of C-reactive protein. In the two hospitalised patients, computed tomography of the chest showed ground-glass opacities with consolidations; 1 patient additionally had a small pleural effusion. These 2 patients were treated with remdesivir, as well as broad-spectrum antibiotics (meropenem). The patients with mild disease were treated with co-amoxicillin (n = 3, 27%). Dexamethasone was given in selected cases. Mechanical ventilation was not necessary for any of these patients. The hospitalised patients received oxygen by nasal cannula and high-flow oxygen therapy. All patients recovered. Residual symptoms are being monitored.

Conclusion: This is the first study of an adult CF-COVID-19 cohort in Switzerland, which included patients who underwent lung transplantation. Cough, fever, headache and dyspnea were the most common symptoms. Two patients (27%) had severe disease. The majority had a benign course and long-term symptoms are still under investigation.

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Rapid implementation of virtual clinics during the COVID-19 pandemic

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Introduction: At the start of the UK COVID-19 pandemic, people with cystic fibrosis (pwCF) were designated “extremely clinically vulnerable,” underwent shielding, with face-to-face clinical contact postponed. Prior to lockdown we had cancelled routine CF clinics and used this unique opportunity to rapidly redesign outpatient services providing remote video-assisted consultations (VAC) with digital technology. VAC also enabled vulnerable shielding MDT members or those self-isolating through COVID contact to provide care. We describe our experience in rolling out VAC.

Methods and results: We initially used AccurX[®], a healthcare provider (HCP) text message-initiated video call service. From March 20th–April 17th 2020, 192 physician-led VACs were completed to support pwCF with the lowest FEV1% in our clinic. From April 18th we moved to the NHS England and NHS Improvement supported secure Attend Anywhere[®] service with pre-arranged appointment times, constructing 10 bespoke MDT waiting area ‘clinics’ with 44 active MDT users, including an all-discipline MDT clinic. Over 8 months, 1,348 consultations (602 hours) took place with most activity (1,163 consultations; 86%; 557 hours) in the MDT clinic. Other areas used were CF Nurse Specialist (n = 86), CFRD (n = 21), Dietetics (n = 12), Physiotherapy (n = 31) and Psychology (n = 19). Since August 2020, we have reinstated routine face-to-face clinics for emergencies and Annual Review, but >90% of our pwCF are now managed virtually via VAC with remote spirometric monitoring (Nuvoair[®]).

Conclusions: MDT members and pwCF report a high satisfaction with VAC with an increased number of pwCF using it. VAC has improved pwCF’s access to MDT services, abolishing the need to travel for and the risk of

cross-infection associated with physical routine review, whilst improving appointment convenience. There has also been improved collaboration between professionals with this new way of working, and we continue to adapt our services to the benefit of pwCF.

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Evaluating the impact of a telemedicine service during the COVID-19 pandemic in people with cystic fibrosis

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Introduction: To provide continuity of care during the COVID-19 pandemic, our large adult CF unit implemented a telemedicine service in order that people with cystic fibrosis (pwCF) – categorised as ‘extremely clinically vulnerable’ in the UK – could access clinicians during periods of shielding. Here we report their opinions of and engagement with the service during the early pandemic.

Methods: A total of 44 consecutive pwCF attending virtual clinics completed a 23-part survey composed of free text and rating scale questions to evaluate the impact of the telemedicine service and the pandemic on their CF care. We also compared telemedicine and face-to-face (FTF) appointment attendance rates during the pandemic.

Results: Feedback was generally positive (see Table). The main areas of constructive criticism revolved around audio and visual quality, as well as connectivity issues associated with use of mobile devices to access the telemedicine portal. Positive comments of the telemedicine clinic included no travel requirements, and no risk of cross-infection. Between July and October 2020, attendance was higher at FTF appointments than telemedicine (166/186 vs 417/513, Chi-squared p = 0.01).

Rating scale question	Strongly disagree (%)	Disagree (%)	Neither (%)	Agree (%)	Strongly agree (%)
Satisfied with the clinics	0	0	7	61	32
Efficient	0	2	12	42	44
Save time	0	5	9	50	36
Easy to access	0	0	9	55	36
Meeting health needs	0	5	9	52	34
Preferable to FTF	2	7	5	43	43
Want to continue	2	0	14	41	43
Confidentiality/privacy respected	0	0	9	52	39
Satisfied with audio/visual quality	2	7	23	48	20

Conclusions: The telemedicine service is generally popular with pwCF, and engagement with the service has been good. Clinicians need to be aware that technology issues may hamper appointment attendance and interaction for some users.

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TeCC (TeleMedicine, Cystic Fibrosis, Corona-Virus) study in a previous telemedicine-naïve centre: clinical challenges, outcomes, and user experience in the first six months of a global pandemic

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Background: COVID-19 made it necessary to establish telemedicine as a first default for reviews in a previously telemedicine-naïve clinic.

Objectives: To establish and assess telemedicine in the first 6 months of COVID-19 pandemic.

Methods: Utilising a multidisciplinary team (MDT) approach, we established a ‘Covid Pack’ of medical equipment (sent to each patient) and a suitable video conference platform to replicate the in-person clinic format. The virtual clinic was then rolled out (94 patients reviewed in the first 6