Since January 2020 Elsevier has created a COVID-19 resource centre with free information in English and Mandarin on the novel coronavirus COVID-19. The COVID-19 resource centre is hosted on Elsevier Connect, the company's public news and information website.

FEV1% predicted of 61% (range 18–101%). Extensive bronchiectasis was noted in 77.8%. Three patients had no respiratory symptoms. Microbiologic cultures from sputum isolated *Pseudomonas aeruginosa* and *Staphylococcus aureus* in 66.7% and 55.6% respectively. Three patients had documented pulmonary tuberculosis, and atypical mycobacterium was isolated from 1 patient. Nasal polyps were noted in 16.7%. From a nutritional perspective, fecal elastase data was available in 16 patients, and 69% were pancreatic sufficient, 31% were pancreatic insufficient. Median body mass index (BMI) was 14.8 kg/m2 (range 11–25 kg/m^2), and at time of diagnosis 27% had BMI above the 25thpercentile. Four patients had CF-related diabetes.

Conclusion: In the Indian subcontinent, PwCF diagnosed after 10 years of age had mild clinical phenotype in early childhood, often related to uncommon mutations. However, there is a trend toward significant decline in pulmonary disease in a few patients in the second decade. Proactive efforts to identify PwCF with milder phenotypes, including surveillance of siblings, would be important to prevent worsening pulmonary morbidity even though disease progression is gradual in the first decade.

159

Lung function changes following Sars-CoV-2 infection in cystic fibrosis

T. Kremer[1], I. St Onge[1], M. Trivedi[1], F. Dy[1], C. Bielick[2], D. Fish[3], O. Schaefer[2], J. Longtine[4], K. Longtine[5]. *[1]Pediatrics, University of Massachusetts Medical School, Worcester, USA; [2]Medicine, University of Massachusetts Medical School, Worcester, USA; [3]Pediatrics and Internal Medicine, University of Massachusetts Medical School, Worcester, USA; [4]Horae Gene Therapy Center, UMass Memorial Medical Center, Worcester, USA; [5]Horae Gene Therapy Center, University of Massachusetts Medical School, Worcester, USA*

Background: Individuals infected with Sars-CoV-2 commonly have pulmonary manifestations as part of their disease process. There are increasing reports of pulmonary function changes in individuals post Sars-CoV-2 infection. Very little is known about the impact of Sars-CoV-2 infection on lung function in patients with cystic fibrosis. We report on 4 patients with cystic fibrosis who were infected with Sars-CoV-2 and the changes in FVC and FEV1.

Methods: We retrospectively reviewed charts of CF patients at our center since the onset of the COVID pandemic to determine who had been infected with Sars-CoV-2. We then determined which patients had PFTs performed prior to and post infection. We assessed changes in FVC and FEV1 for these patients.

Results: There were 9 patients (age 10 to 42 years) at our center identified as having been infected with Sars-CoV-2 from March 2020 to April 2021. Of these, 4 patients (age 21–40 years) had PFTs prior to and following Sars-CoV-2 infection. There were 3 of these patients being treated with elexacaftor/tezacaftor/ivacaftor as part of their routine care. One patient did receive monoclonal antibody therapy for his infection. Baseline FVC pp ranged from 78 to 97% predicted and baseline FEV1 pp ranged from 59 to 89% predicted. Sars-CoV-2 infection severity for all patients was mild, and no patients were hospitalized as a result of this infection. The percent change in FVC (pre- to post-Sars-CoV-2) for all 4 patients was 1.5%, 10.5%, –7.7% and 0.9%. The percent change in FEV1 (pre- and post-Sars-CoV-2) for all 4 patients was 0.8%, 5%, –2.4% and –0.4%. There was no significant change in the FVC and FEV1 from baseline values following Sars-CoV-2 infection for these 4 patients with mild infection (Table 1).

	Pre FVC (L)	Pre FVC pp	Post FVC (L)	Post FVC pp	Pre FEV1 (L)	Pre FEV1 pp	Post FEV1 (L)	Post FEV1 pp	% Change FVC	% Change FEV1
Patient 1 (31yo)	4.78	97	4.85	98	3.6	89	3.63	89	1.50%	0.80%
Patient 2 (40yo)	2.87	78	3.17	86	1.99	66	2.09	70	10.50%	5.00%
Patient 3 (38yo)	3.39	78	3.13	71	2.07	59	2.02	57	-7.70%	-2.40%
Patient 4 (21yo)	3.23	84	3.26	84	2.85	84	2.84	84	0.90%	-0.40%

Table 1. Sars-CoV-2 pre- and post-spirometry

Conclusion: This small case series of 4 adult patients with CF infected with Sars-CoV-2 noted no significant change in lung function following infection. The range in Sars-CoV-2 infection severity can vary between mild disease with no change in lung function to death. Based on CFF reports, it is recognized that outcomes may depend on baseline lung function and severity of lung disease, as well as if post-lung transplant. Other factors may play a role in COVID-19 outcomes, including use of routine airway clearance, possible treatment with highly effective CFTR modulators, and outpatient therapies for Sars-CoV-2.

160

Using dynamic chest radiography to assess response to treatment in acute pulmonary exacerbations of adult people with cystic fibrosis

T. FitzMaurice[1], C. McCann[2], D. Nazareth[3], M. Walshaw[3], P. McNamara[4]. *[1]Liverpool Heart and Chest Hospital, Liverpool, UK; [2]Department of Radiology, Liverpool Heart and Chest Hospital, Liverpool, UK; [3]Adult CF Unit, Liverpool Heart and Chest Hospital, Liverpool, UK; [4]Department of Child Health, Institute in the Park (University of Liverpool), Alder Hey Children's Hospital, Liverpool, UK*

Background: Using dynamic chest radiography (DCR), a unique real-time X-ray imaging system [1], we have shown variation in diaphragm movement following pulmonary exacerbations (PEx) in adult people with CF (PwCF). Now, to assess the impact of PEx on tidal and deep breathing phases of respiratory physiology, we have utilized DCR to look at their effect on lung area changes.

Methods: Twenty PwCF ([mean ± SD] age 25.2 ± 6.6 years, average ppFEV1 59.5 ± 17.1, BMI 21.1 ± 3.6kgm^{-2}, 14 female) underwent spirometry and DCR before and after inpatient treatment for PEx. Hemidiaphragm speed and change in vertical hemidiaphragm position were measured by DCR during tidal and deep breathing, to assess the impact of PEx on respiratory muscle function, with the posteroanterior projected lung area (PLA) measured as a surrogate for lung volume.

Results: Treatment improved maximum excursion during tidal breathing of both left (12.9 ± 5.1 to 15.9 ± 6.6 mm, $P = 0.01$) and right (11.2 ± 5.2 to 14.5 ± 7.1 mm, $P = 0.02$) hemidiaphragms, as well as maximum tidal inspiratory speed (left 15.8 ± 5.0 to 18.6 ± 6.6 mm/s, $P = 0.04$; right 13.9 ± 5.1 to 16.5 ± 6.9 mm/s, $P = 0.07$). Hemidiaphragm excursion on deep breathing improved on the right (13.4 ± 6.7 to 18.3 ± 8.9 mm, $P < 0.001$) and left (18.2 ± 8.4 to 23.9 ± 9.5 mm, $P = 0.01$), as did maximum passive expiratory speed for right (12.0 ± 4.4 to 19.5 ± 10.4 mm/s, $P = 0.003$) and left (15.6 ± 8.4 to 22.6 ± 11.9 mm/s, $P = 0.008$) hemidiaphragms. The ratio between maximum inspiratory/expiratory PLA correlated with ppFEV1 (r = 0.753, $P < 0.001$). PLA at maximum expiration correlated with the FEV1/FVC ratio (left, r = –0.800, $P < 0.001$; right, r = –0.672, $P < 0.001$; Figure 1). Maximum hemidiaphragm excursion correlated with FVC (left, r = 0.6340, $P < 0.001$; right, r = 0.6927, $P < 0.001$).

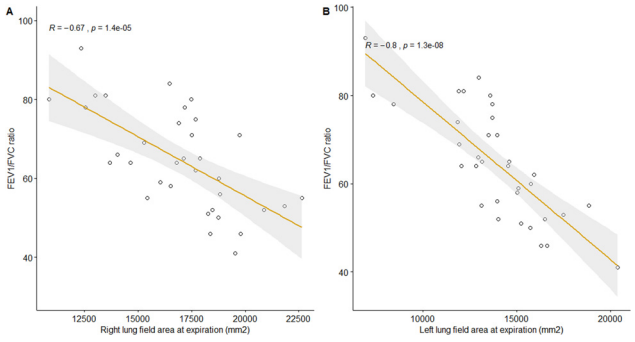

Figure 1. Relationship between FEV1/FVC ratio, and right (A) and left (B) lung field area at end expiration

Conclusion: We have demonstrated improvement in diaphragm excursion and speed during tidal breathing, suggesting individuals are able to take deeper and faster tidal breaths as a result of treatment for PEx. The increase in passive expiratory speed following a deep breath matches our previous work and suggests improvement in the elastic recoil of the lungs. The correlation between spirometric and DCR parameters suggest DCR may be a complementary investigation in measuring lung function changes during PEx, especially in individuals in whom reproducible spirometry is difficult.

Reference

1. FitzMaurice T, Nazareth D, Bedi McCann C, McNamara P, Walshaw M. Dynamic chest radiography is a novel tool to assess chest wall and diaphragm dynamics during pulmonary exacerbations of cystic fibrosis. *Pediatr Pulmonol*. 2020;55(S2):P671,301.