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Letter to the Editor

Care of Cystic Fibrosis Children in COVID-19 Pandemic

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The entire world is currently facing a pandemic of coronavirus disease 2019 (COVID-19) caused by severe acute respiratory syndrome coronavirus-2 (SARS-CoV-2) [1]. Cystic fibrosis (CF) is an autosomal recessive multiorgan disease more common in Caucasians with an incidence of 1 in 3,500. The exact prevalence of CF in Asian countries is not well known, but studies have estimated that these regions may constitute a large proportion of patients with CF worldwide [2]. In CF, the lungs are the most commonly and severely involved organ. Here we have highlighted some issues related to the care of children with CF in the COVID-19 pandemic.

CF is considered a high risk for the SARS-CoV-2 infection, which may cause severe illness in patients with CF [3]. However, the impact of this novel virus on patients with CF is still not fully elucidated. A multinational study from Europe has estimated the COVID-19 incidence of 0.07% in the population suffering from CF, which is much lower than the general population. They also observed that the clinical course of COVID-19 in patients with CF was similar to that of the general population [4]. The possible lower rate of SARS-CoV-2 infection in CF may be because of underdiagnoses, relatively younger age, altered airway milieu and immune response, and probably better awareness and practice of cough etiquette and airway clearance in patients with CF [4, 5].

Viruses are a common cause of CF exacerbation in children. Possibly, SARS-CoV-2 infection can also trigger CF exacerbation. Therefore, any case of CF exacerbation should be suspected as COVID-19, especially those coming from high infective areas [5]. All the routine CF medication should be continued as such. To date, there is no approved medication for the prevention of COVID-19 in patients with CF. Instead, experimental medicines may lead to drug interaction and potentially severe adverse reactions in children with CF, and it should not be given without the supervision of the CF care team [6]. The use of nebulizer is essential to the care of patients with CF; however, it is a potential source of aerosol generation and risk of COVID-19 transmission to the healthcare workers and cross-contamination. Therefore, nebulizer use should be restricted in a hospital setting, but its use at home may be continued [6].

Similarly, pulmonary function testing (PFT) is also a potential source of virus transmission due to coughing and droplet formation; thus, routine PFT should be avoided. PFT, which is essential for making treatment decisions, should only be performed after carefully considering the risk of virus transmission to its benefit and taking full protection [7]. All the equipment (facemask, spacer, nebulizer, and other physiotherapy devices) should be cleaned regularly as per the standard guideline [6, 8].

Every effort should be made to communicate with the patients with CF or their caretaker to alleviate their anxiety and inquire about their disease status. Telemedicine may play a significant role in CF care, especially in a patient's triage, and it can avoid unnecessary hospital visits [6]. However, it has some limitations related to the patients, healthcare personnel, and technology, which should also be considered. Nevertheless, devoid of physical evaluation for a more extended period may lead to disease progression, including deterioration in lung function, poor compliance to therapy, and an increase in CF-related complications. Furthermore, it will also hamper the new drug's clinical trial , as no new trial has been started since this pandemic began, and further recruitment to the existing trials has also been jeopardized [6, 9].

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Home monitoring should be encouraged using a pulse oximeter and anthropometric measurement. The concept of home spirometry is emerging and currently in practice at a few CF centers. It may be beneficial in children who required frequent lung function monitoring; however, its accuracy is doubtful. Thus, its value should be interpreted with caution. The sputum or blood samples may also be sent to the respective CF clinics. Patients with CF can share all this information with the CF care team to avoid hospital visits [9].

Patients with CF should strictly adhere to their prescribed medication, airway clearance, adequate diet (rich in fat, energy, and salt), regular exercise, and maintain hydration. Medication should be stored at home for several weeks [3]. Pulmonary exacerbation should be promptly treated at home (with oral or parenteral antibiotics) to avoid hospital visits [9]. It is desirable to strictly follow the general public health care measures, that is, social distancing, handwashing, and cough etiquette [5, 6]. Many countries have adopted the concept of shielding (avoiding face-to-face contact; self-isolation) or cocooning (staying at home and no contact with any other person from outside) to prevent SARS-CoV-2 infection in children with CF [10].

Because lockdown in many parts of the world has adversely affected the family income, patients with CF and their families require continuous motivation and some source for financial support.

In conclusion, although the COVID-19 pandemic has severely handicapped the routine CF services, emphasizing on adherence to medication, general health measures, proper diet, exercise, self-isolation, home monitoring, telemedicine, and maintaining mental peace are the ways to continue CF care.

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