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Letter to Editor Open Access

New heights: travelling with cystic fibrosis to high altitude destinations

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Dear Editor,

Cystic fibrosis (CF) is an autosomal recessive disorder affecting mainly the lungs, pancreas and intestines, which is caused by mutations in both copies of the gene for the cystic fibrosis transmembrane conductance regulator (CFTR) membrane protein and chloride channel. Patients expectorate large volumes of thick mucus which leads to recurrent respiratory tract infections, bronchiectasis and, in some patients, secondary pulmonary hypertension. People living with CF are believed to have a poor aerobic exercise performance at high altitude.[1] CF tends to be included in the general advice provided to travellers with cardiorespiratory disease.[2] Recommendations for air travel, including the need for fitness-to-fly assessments using hypoxic challenge testing, are discussed elsewhere.[3] The leading textbook of travel medicine merely advises that CF patients at altitude "should be monitored and might require oxygen therapy".[1] We consider how an amateur trekker with CF who wishes to climb a high mountain such as Kilimanjaro (5895m) should be advised.

Following the success of Nick Talbot, a British patient living with CF, in summiting Everest in 2016, there has been greater interest in high-altitude travel among the CF patient community. The advent of cystic fibrosis transmembrane conductance regulator (CFTR) modulators has transformed patient care, with significant improvements in lung function, nutritional status, exacerbation frequency, and quality of life, making high-altitude trekking an attainable goal for most patients living with CF. Stable transplanted CF patients who achieve the necessary level of aerobic fitness have been found to perform as well as or even better than average at high altitude.[4]

While trekkers with chronic lung disease may have a degree of adaptation to chronic hypoxia, they still experience profound arterial oxygen desaturation as they ascend. At the summit of Kilimanjaro, an individual with normal sea-level capillary oxygen saturation will desaturate to approximately 80% at rest. The value will be even less in CF patients with lower baseline oxygen saturation. CF patients with secondary pulmonary hypertension are at greater risk of developing high altitude pulmonary oedema (HAPE). This risk may be mitigated by encouraging them not to overexert themselves at high altitude and to warm their inspired air as much as possible, for example with a silk scarf.

Since symptomatic pulmonary hypertension is a contraindication to high altitude travel[2], CF patients with pulmonary hypertension should use supplemental oxygen and take nifedipine (30-60mg controlled release once daily starting 24 hours before ascent) for HAPE prophylaxis. Nifedipine acts by reducing pulmonary artery systolic pressure. Tadalafil 20mg daily is an alternative and inhaled salmeterol, which may be part of the CF trekker's daily treatment regime, may also be used in the management of HAPE. It acts by improving

clearance of fluid from the alveolar spaces. There is no contraindication to the use of prophylactic acetazolamide in CF. It produces a mild metabolic acidosis to offset the respiratory alkalosis resulting from carotid body-induced hyperventilation. Acetazolamide at an oral dose of 125mg twice daily decreases hypobaric hypoxaemia, reduces the incidence of acute mountain sickness and HAPE, and improves sleep quality.

With the success of CFTR modulators, it may no longer be necessary to optimize the CF patient's clinical status with an inpatient course of intravenous antibiotics in advance of high-altitude travel. Chronic upper respiratory symptoms are common in this patient population. Nasal polyps and chronic sinusitis may compromise respiration further and these should be managed with a corticosteroid nasal spray well in advance of travel. Owing to the expansion of gasses at altitude, CF patients with a history of spontaneous pneumothoraces should be aware of the risk of recurrence, even at the cruising altitude of a pressurized aircraft cabin.

High-altitude trekkers with CF should use volumatic spacers with their inhalers and portable nebulizers. They should continue to take their regular medications, including mucolytic agents and prophylactic oral antibiotics. Monitoring oxygen saturation using a pulse oximeter may be useful but there would be greater benefit in having an expedition physician in the trekking party who can respond appropriately to excessively low saturations in a symptomatic patient based on knowledge of the expected values at a given elevation. It will not be feasible on most high-altitude treks to use non-invasive ventilation devices, including BiPAP machines.

Where possible, a respiratory therapist should accompany the CF trekker to provide chest physiotherapy to aid in the clearance of thickened airway mucus but vigorous self-administered chest physiotherapy may be a feasible alternative. Commercial trekking companies are increasingly carrying portable hyperbaric chambers[5] and there should be a low threshold for using these to simulate descent in a CF patient with severe AMS or HAPE. Decisions on timely descent should not be delayed, however, and trekking leaders should be very familiar with the location of medical aid posts at lower campsites. On the Kilimanjaro trek, a longer climbing route such as Lemosho is preferred as it allows more time for acclimatization to hypoxia.[6]

Table 1 summarizes key practical recommendations when counselling CF patients travelling on high-altitude trekking expeditions. As with all travellers with chronic medical conditions, CF trekkers should obtain comprehensive travel insurance that includes repatriation coverage and follows a full disclosure of their medical history. A formal medical fitness-to-travel assessment by a respiratory physician may be mandated by insurance companies.

In conclusion, CF patients may travel safely to very high or even extreme altitude destinations given appropriate pre-travel preparation and support during travel. Clinicians who counsel high-altitude travellers with CF should be familiar with the specific health risks involved and their mitigation. Close liaison between respiratory and travel medicine specialists will best serve the needs of high-altitude trekkers with CF.

Conflict of interest

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Table 1. Practical travel health recommendations for trekkers to Kilimanjaro with cystic fibrosis

Travel health issue	Practical recommendations and rationale
Air travel	Wear correctly fitting FFP2 or N95 masks.
Dietary issues	Trekkers require a high-caloric intake and CF patients should match their use of pancreatic enzyme supplements to the amount of fat in their diet. Carbohydrates should be encouraged as they increase the respiratory quotient.
Vaccinations	The live attenuated yellow fever vaccine (YFV) is generally not recommended for travel to Tanzania but would be required in arriving passengers who transit for more than 12 hours in another yellow fever-endemic country. CF travellers who have received a lung transplant should plan air travel to avoid this, as they cannot receive YFV owing to their immunocompromised status. A medical exemption certificate will be indicated in this case.
Dehydration	Dehydration is common at high altitude and should be avoided as it causes drying of mucous membranes and thickened airway secretions. It increases the risk of developing distal intestinal obstruction syndrome and accompanying medical personnel and mountaineering guides should be alert to this complication. Alcohol should be avoided. Excessive sweating may necessitate oral electrolyte replacement.
Trekkers with diabetes	CF trekkers with diabetes should consult their dialectologist or travel health physician for specific advice around the use of insulin, avoidance of hypoglycaemia and foot care at altitude.

FFP: filtering facepiece; **CF**: cystic fibrosis