Thorax

Editorial

Flying and cystic fibrosis: getting there and back safely

A K Webb

Patients with cystic fibrosis have the same expectations as their healthy peers and this includes holidays abroad with family and friends to far away places which require long haul flights. This expectation is irrespective of the severity of their lung disease. The physician has the responsibility of balancing their wish to enjoy themselves against their medical safety. The risk of flying is related to the fact that barometric pressure reduces with altitude. The partial pressure of oxygen is inversely proportional to altitude. Commercial aircraft fly with an adjusted cabin altitude of 8000 feet which results in an inspired oxygen concentration of approximately 15% compared with a sea level inspired oxygen concentration of 21%. In healthy individuals this reduced inspired oxygen concentration results in a relative fall of only 4–5% oxygen saturation in the blood. As healthy individuals are on the flat part of the oxygen dissociation curve, no respiratory risk to health is incurred. However, patients with established lung disease and associated hypoxaemia at sea level have a significant respiratory risk while flying. On this basis, paediatric and adult respiratory physicians have tried to develop the best method of assessing those patients at risk of flying and advising which patients should be provided with inflight supplementary oxygen. The advent of the 15% normobaric oxygen challenge using a premixed cylinder of oxygen and nitrogen simulates inflight hypoxia over a 20 minute period. The test is time consuming and not all hospitals have the equipment, nor does a single test replicate a 1–2 week holiday when the patient's health may be much worse on the inward flight than the outward journey.

Many papers have been written about the evaluation of inflight hypoxaemia in adults with chronic lung disease.[1–3] It is a reflection of the current uncertainty that the British Thoracic Society set up a Working Party to formulate national recommendations for managing patients with lung disease planning air travel and concluded that there was insufficient evidence to produce formal guidelines.[4] Crucially, there are no established validated methods of accurately quantifying the respiratory risk of flying for patients with different forms of lung disease, and there is even less information on the safety of patients with cystic fibrosis at altitude. Even patients with mild to moderate disease will have a degree of ventilation perfusion mismatch which at sea level places them on the edge of the down slope of the oxygen dissociation curve. Several reports have documented acute right heart failure or death occurring in patients with cystic fibrosis placed at altitude and not protected by oxygen.[5 6]

In view of these remarks, the paper in this issue of *Thorax* by Buchdahl *et al*[7] is a timely evaluation of the pre-flight hypoxic challenge in children with cystic fibrosis. Although a close run thing, the authors conclude that pre-flight spirometric tests are a better predictor of desaturation during flight than the pre-flight hypoxic challenge. The paper is interesting but not clinically helpful in terms of decision making. Patients with more severe disease were excluded, as detailed in the methodology, and presumably the patients were medically supervised at their holiday destination and kept reasonably well for the return flight. In their previous paper, which had design differences from the current study, the authors reached the opposite conclusion—namely, that a pre-flight hypoxic challenge had a better predictive value than spirometric tests.[8] However, a study of fitness to fly in adults with moderate to severe cystic fibrosis concluded that neither resting oxygen saturation Sa_{O_2} nor forced expiratory volume in 1 second (FEV_1) will accurately predict hypoxaemia during simulated flight conditions.[9]

Adult patients with cystic fibrosis want to travel whatever the level of their disease severity, but they are undoubtedly at greater risk than most patients with lung disease. There are additional specific risk factors causing a reduction in arterial Pa_{O_2} other than an inspired oxygen concentration of 15%. Travellers with cystic fibrosis have chronic pulmonary sepsis which can be exacerbated by the development of viral infections acquired on either the outgoing or return flight. Dehydration is a well recognised complication of flying and further dries secretions in cystic fibrosis. A significant number of travellers with cystic fibrosis will have diabetes, and a holiday which consists of sun, alcohol, partying, and dehydration puts them at greater risk on the return flight. Self-care, an essential component of their disease management, is usually totally neglected until the holiday is over!

How can we reduce the risk of air flight for subjects with cystic fibrosis? The small number of peer reviewed publications suggests that this is a wonderful area for clinical research, while we are currently dependent upon making difficult decisions about fitness to fly with no guidelines in a very compromised group of patients and at the same time filling out travel insurance forms with a song and a prayer that we have got it right. On our adult cystic fibrosis unit we recommend inflight oxygen when the Pa_{O_2} falls below 6.6 kPa during a simulated flight oxygen test. This recommendation is currently accepted for safety reasons.[10 11] We also observe the reduction in Pa_{CO_2} as an indicator of how hard the patient is breathing to maintain adequate levels of P_{O_2}. If oxygen is required it is absolutely crucial that the patient uses an airline which allows inflight oxygen. They may have to pay for their own oxygen which can increase the cost of what was initially a cheap holiday. Some airlines refuse to allow inflight oxygen usage, as some patients have discovered to their personal cost when having to rebook on an alternative airline.

A holiday for a subject with cystic fibrosis is about a great deal more than flying and arriving. Even with careful planning things can go unexpectedly wrong in a distant land with no cystic fibrosis centre and, on their return, patients with cystic fibrosis will often need additional treatment.[12] For patients with cystic fibrosis a holiday abroad requires

careful preparation; at the very least, each patient should know whether they require inflight oxygen, have a fitness to fly letter from the consultant, a letter detailing their current medical condition and medication, and a treatment protocol for the usual emergencies if the patient presents to a hospital which has little experience of cystic fibrosis.

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