Oxygen therapy for cystic fibrosis

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INTRODUCTION

Oxygen therapy for children with cystic fibrosis (CF) is indicated in a number of situations. During serious lower respiratory tract infections it is useful both acutely and also long-term, when chronic respiratory failure has become established. In addition, it may be required short-term during periods spent in relatively hypoxic environments such as during aircraft flights and on winter sports holidays at altitude. Those who desaturate during exercise may also be helped to maintain a better quality of life with oxygen supplementation during periods of more vigorous physical activity. Nowadays there is quite rightly an increased emphasis on community and home care, but if this is to be provided optimally then the best facilities, particularly in relation to oxygen delivery systems, must be made available in order to ensure the best quality of life for as long as possible.

PHYSIOLOGY

The physiology of oxygen uptake is complex. The basic requirements are known to be increased in CF, although in clinical practice this probably does not cause a significantly increased need in terms of inspired oxygen concentration. The circulating level is based on the oxygen saturation curve which shows that normal healthy values lie above 95%. In clinical practice levels of 92% or more are acceptable and unlikely to cause problems in day-to-day life[1]. Below 92% the desaturation curve is more acute and descent into physiologically abnormal levels becomes more likely. These effects are significantly enhanced during flight in commercial aircraft and especially during holidays at altitude[2]. The practical difficulties of delivering continuous oxygen therapy during normal life, such as at school, must be weighed against the potential advantages. Many children with CF lead a perfectly acceptable social and indeed academic life with oxygen saturations in the range of 89–92%. This has to be balanced against the possible but less proven risk of its contribution to the development of clinically important pulmonary hypertension in the longer term. Nowadays in clinical practice progressively advancing suppurative lung disease and the onset of chronic respiratory failure are more common terminal events in CF than chronic cor pulmonale.

The patient's perception of long-term oxygen therapy is another important factor to be taken into consideration. The effects on self-image, the perceived increase in burden of treatment and insight into decline in health are important issues. The use of chronic therapy implies a loss of control of body image and is certainly not 'sexy' amongst one's peers, particularly during adolescence. Such difficulties contribute to a negative perception towards the initiation of long-term therapy.

During exercise oxygen demand is higher, and this has to be met by increased delivery to the tissues with an associated higher extraction level from the blood. If necessary, this can be compensated for by an increase in the fractional inspired oxygen concentration (FiO_2). These demands become greater if exercise is taken at altitude, which is not uncommon nowadays as winter holidays such as skiing trips become ever more popular. A recent study of adult CF patients undertaking maximum exercise at an altitude of 1500 metres (4920 feet) showed that oxygen saturation dropped below 90% in 29 of 50 patients (58%)[3]. This was most common in those with an FEV_1 of less than 40%, where it was seen in 12 of 14 (86% of those tested), it also occurred in 15 of 25 (60%) of those with FEV_1 between 40 and 70%.

Many overseas journeys involve travel by air with flights of varying duration. The internal environment of modern aircraft is designed as a balance between the normal values of air pressure and humidity at sea level and what it is reasonable to achieve at high altitude (e.g. 35 000 feet). This is limited by the constraints of weight relating to the equipment to provide it, provision of adequate humidity and its effects on fuel consumption. A compromise has therefore been reached such that the average cabin pressure is equivalent to an altitude of 7500 to 8000 feet with a relative humidity of 17%. This equilibrates to a reduction in barometric pressure of approximately 20% compared to that at sea level and a fall in oxygen saturation in an otherwise healthy subject of about 1%. This is equivalent to breathing 15% inspired oxygen at sea level.

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ASSESSMENT

The assessment of extra oxygen requirement in the paediatric age group is principally based on measurements of oxygen saturation and basic lung function such as FEV_1. These are simple non-invasive tests which are easily tolerated. Arterial blood gases may be needed from time to time and capillary samples are useful for the measurement of carbon dioxide levels in those with advanced respiratory disease. Indications for chronic therapy at home include significant hypoxia at rest with a saturation of less that 90% in air, $PaO_2 < 7.3$ kPa, hypercapnia with a $PaCO_2 > 6.0$ kPa and lung function measurements in adults including FVC < 2.0 L and $FEV_1 < 1.5$ L.

Evaluation of the effects of altitude can be performed by allowing the subject to breathe an inspired oxygen concentration of 15% in the laboratory[3], alternatively this can be reproduced within a body plethysmograph[4], (Figure 1) which also allows the measurement of the effects of additional oxygen to correct any induced hypoxaemia. A recent study has confirmed its use during transatlantic flights while children were awake, but showing a less good correlation when they were asleep. During outbound flights when children were awake the laboratory hypoxic challenge accurately predicted resting oxygen saturation (94.4%), but during the return flight when 24 of 44 tested were asleep it underestimated saturation by 1.8% on average. This may not matter greatly during this time, since the patient is physically inactive or asleep[5]. In a further study oxygen saturation fell during flight in six children taken on a skiing holiday, but reverted to baseline even at altitude (Figure 2)[6]. FEV_1 was maintained throughout and no child with a FEV_1 of greater than 40% desaturated significantly at rest (Figure 3).

Exercise hypoxia can be assessed by the use of the 3-minute step test (Figure 4)[7]. This involves a simple protocol which can be used with the minimum of equipment and which is easily portable so that it can be utilized for research purposes during sports activities at altitude. It involves a 3-minute standardized test using a 15 cm step, at a rate of 30 per minute. The rate is controlled by a metronome and 91% of patients are able to undertake the test successfully[7]. It is portable and therefore allows testing inside and outside in any setting, when exercise is being undertaken and at any altitude. It is reasonably predictive of likely hypoxia during aircraft flights and exercise well above sea level (Figure 5). The subject of exercise testing in cystic fibrosis has been reviewed by Orenstein[8].

PRODUCT AVAILABILITY

Provision of oxygen in the community is vital to the maintenance of quality of life and mobility in the seriously ill patient. Products available for the oxygen dependent

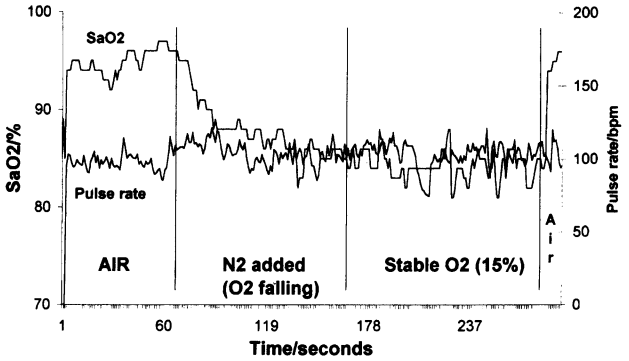

Figure 1 **Assessment of oxygen saturation and pulse rate, breathing 15% oxygen at sea level in the body plethysmograph**

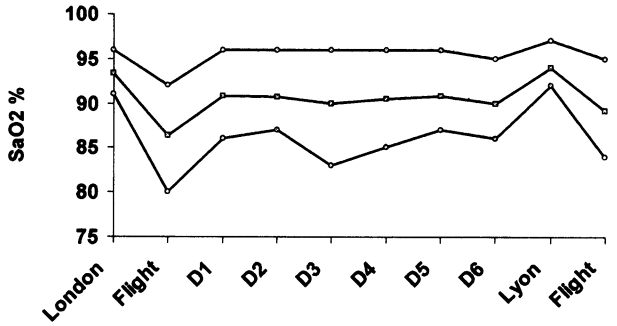

Figure 2 **Oxygen saturation at sea level, during flight and at 1500 m altitude in six cystic fibrosis children during a skiing holiday (minimum, mean and maximum, D=day)**

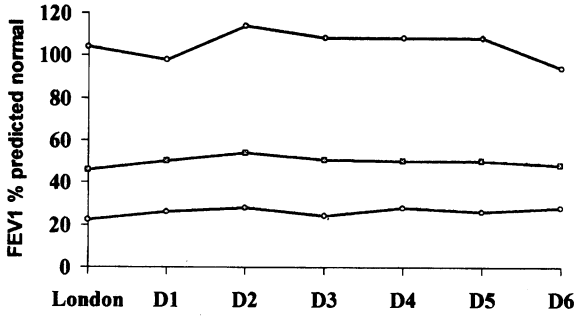

Figure 3 **FEV_1 during a skiing holiday for six cystic fibrosis children at an altitude of 1500 m (minimum, mean and maximum, D=day)**

Figure 4 **Cystic fibrosis patient undertaking a step test at 1500 m altitude**

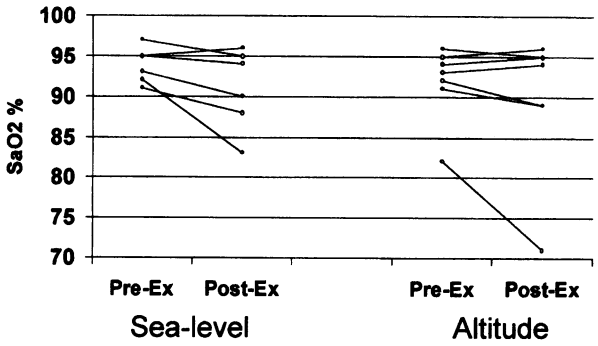

Figure 5 **Effect of 1500 m altitude on 3-minute step test for six cystic fibrosis children**

patient at home have been thoroughly reviewed in the paper by Dodd *et al.*[9]. The products which can be used are mainly prescribable, such as domiciliary oxygen sets, face masks, oxygen concentrator, oxygen cylinder and stand. General practitioners and patients are, however, met by confusing rules when they try to arrange for a suitable system for an active life. Portable oxygen cylinders sizes C and D are prescribable, but the cylinder headset and recharging adaptor are not. These smaller cylinders cannot be used with a headset from a larger one and therefore the appropriate adaptors either have to be lent out by the hospital or bought privately. Consequently, many patients have to rely on their local hospital to ensure that they have an adequately portable system, or they cannot go out.

Unfortunately, other alternatives are not always known about either by the professionals or the support agencies. The only prescribable portable system is the 'PD' oxygen cylinder which holds about 300 L of oxygen and lasts up to 2½ h at a flow rate of 2 L/min. This cylinder will fit the same headset as the larger ones. GPs can prescribe the PD with any of the necessary equipment required and pharmacists can obtain them from the British Oxygen Company (BOC). The pharmacy is charged for the oxygen and a monthly rental for the cylinder.

Oxygen concentrators became available from the USA in the mid-1980s. These require an external power source, so patient mobility is limited. The flow rate provided is up to a maximum of 5 L/min and the oxygen is delivered at the ambient room temperature and humidity. Concentrators are prescribable on FP10 but the commercial suppliers are selected by the National Health Service on a competitive tender basis. The GP gives the prescription form to the patient and telephones the details to the supplier, who then arranges home delivery. After installation the supplier collects the prescription, the cost of which is then met through the Pricing Authority. Oxygen concentrators generate a small amount of noise during continuous use. Their output becomes less accurate at higher flows and it is important to assess this by intermittent measurement of saturation levels during treatment. A backup oxygen cylinder must also be provided in case of power failure.

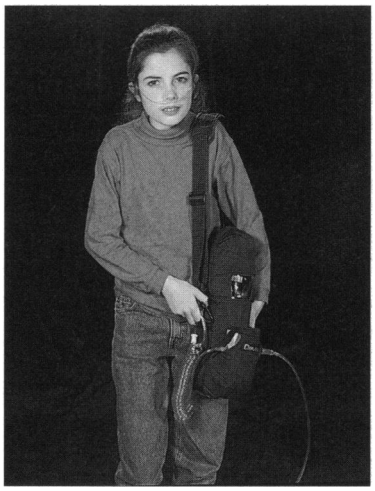

Figure 6 **Oxylite portable oxygen system**

Other more 'patient friendly' alternatives, such as liquid oxygen, are available but non-prescribable. A liquid oxygen system has been developed by BOC which consists of a master cylinder based in the home containing 25 000 L, which can then be downloaded to a portable cylinder of 1.2 L which will deliver 2 L/min of gaseous oxygen for up to 8 h. Portable liquid oxygen systems involve a certain amount of evaporative loss, hence the need for a master tank at home. They are significantly more expensive than cylinders or a concentrator and not all Health Authorities will pay for this system. The Oxylite (Figure 6) is another alternative; light, portable gas-filled cylinders are available which can be filled from another source. These are non-prescribable.

Should oxygen be required during flight a charge for this may be made by the airline[2]. Some of the current costs are set out in Table 1. There appears to be no justification for the huge variation in such costs from one airline to another, since the service provision is similar.

Table 1 **Costs of inflight oxygen**

Airline	*Cost of oxygen*
Air Canada	£38 each way
Cathay Pacific	No charge
Virgin Atlantic	No charge
Caledonian	£42 each way
British Airways	£100 each way
Alitalia	Cost of air fare
Singapore Airlines	50% cost of air fare
Air Malta	No charge
Quantas	£25 each way
Air New Zealand	Cost confirmed at booking

(Source: Isitt J (ed.) Oxygen on flights. *Breathe Easy* 1997;**25**:5)

The Royal College of Physicians currently has a multidisciplinary working party on the provision of domiciliary oxygen and is expected to issue a report to the Department of Health next year.

OXYGEN DELIVERY

Although there are a number of ways to deliver oxygen, in practice patients with CF prefer either nasal cannulae or a face mask. Nasal cannulae are useful in those who require low flows and when there is no significant obstruction, such as with nasal polyps. The cannulae are simple to wear and allow eating, drinking and talking relatively easily. It is difficult to know the precise inspired oxygen concentration delivered by nasal cannulae since the amount provided is diluted by the patient's inspiratory flow rate during tidal breathing. This is also reduced during sleep and there may be an increased requirement at night[10]. Overnight oximetry can be helpful in assessing this need. The provision of humidity is also more difficult with cannulae and there may be a drying effect on the nasal mucosa, although this can be overcome by the use of normal saline nasal drops and the use of a face mask at night, through which increased humidification can be more easily provided.

Face masks are the alternative method of delivery. These can provide an inspired concentration of 24% and 28%. In order to provide this level of inspired oxygen, flow rates of 2–8 L/min may be required. It is useful to assess the requirements in hospital by monitoring the inspired concentration and its effects on oxygen saturation during rest, normal physical activity and procedures such as physiotherapy.

It should also be remembered that should the peak tidal inspiratory flow rate exceed 40–50 L/min then room air is drawn into the mask and the inspiratory concentration will fall. While this may not be common in children it is important in adults[8]. Patients with chronic respiratory failure will also have an elevated carbon dioxide level at rest. This can only be assessed accurately by capillary or arterial blood gases or end tidal measurements during quiet breathing. Many of these patients are driven by a hypoxic response and therefore increasing the inspired concentration and thus the circulating PaO_2 may not always be helpful to respiratory drive.

HUMIDIFICATION

Oxygen supplied from a central hospital supply or via cylinder is relatively cold and non-humidified. This can be uncomfortable for the patient and is potentially irritant to the airways. While it is not easy to moisturize portable oxygen delivery systems it is useful to humidify and also heat the supply of those attached to a static supply while in hospital. A number of humidifiers are available and in common use for this purpose. The provision of adequately humidified inspired gas is particularly important during respiratory exacerbation, when drying of the airway can increase the difficulty in clearing secretions. The provision of adequate humidity to supplemental oxygen in the home is much more difficult, although oxygen from a concentrator will be delivered at room temperature and humidity.

CONCLUSIONS

The provision of oxygen to CF patients is important in the maintenance of long-term stability of lung function and in the quality of day-to-day life. Various fixed and portable systems are now available for use both in the home and outside. If the patient is going to fly then an assessment of need, utilizing a period of inhalation of 15% oxygen, may be helpful. Should this not be readily available current studies would suggest that significant desaturation during flight is unlikely if FEV_1 is greater than 40% and baseline saturation at rest more than 92–94%. If exercise is going to be undertaken, especially at altitude, then a simple exercise test may be helpful in assessing the degree of hypoxia which could be induced by such activity. Overall, however, there is no reason why CF patients should not be able to fly anywhere, with additional oxygen if necessary, and to undertake non-stressful physical activity at altitude within reasonable limits. This approach should maintain quality of life even in those who have relatively advanced lung disease.

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