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EDITORIAL

Home mechanical ventilation

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Mechanical ventilation originated as an in-hospital service but there is now an increasing demand for provision at home [1]. This is a consequence of social pressures, a better understanding of the significance of nocturnal hypoventilation in the pathogenesis of respiratory failure, and an awareness of respiratory muscle fatigue. It is important to recognize that there are two quite different circumstances in which respiratory support may be needed: 1) the patient with a complete inability to breathe, for example after high cervical cord transection; 2) the patient unable to sustain adequate spontaneous ventilation indefinitely, in whom assistance during sleep is all that is required.

Pathophysiology

The onset of sleep is associated with loss of accessory muscle activity and a diminution in ventilatory responsiveness [2, 3]. Intercostal activity is also reduced or absent, during rapid eye movement (REM) sleep and, although this is of no consequence in the normal subject, it results in inadequate gas exchange in patients in whom the work of breathing is markedly increased or diaphragmatic performance markedly impaired.

The disability is often multifactorial in origin, *e.g.* the work of breathing is increased in the scoliotic subject because of deformity and rigidity of the chest wall as well as reduced lung compliance [4]. In addition, the diaphragm may be weak because of primary neuromuscular disease, operating at a mechanical disadvantage because of an abnormal configuration, fatigued by the excess of respiratory work to which it has been subjected [5], or by the hypoxia, hypercapnia or impaired perfusion caused by the acute and chronic consequences of inadequate ventilation [6, 7].

If ventilation is enhanced by mechanical measures during sleep, arterial blood gas tensions can be held within the normal range, so preventing any increase in pulmonary vascular resistance [8] and permitting resolution of right heart failure. The respiratory muscles can be rested so that fatigue is alleviated [9] and, by maintaining normoxia and normocapnia by night as well as by day, the sensitivity of the respiratory centre can be restored [10].

The sceptic may question whether in practice the results conform to this hypothesis. Evidence from a number of centres confirms the success of home ventilation as a means of alleviating chronic respiratory or cardiorespiratory failure, permitting respiratory muscle rest, prolonging life and improving its quality [11–13].

Indications

Current indications include central sleep apnoea, respiratory failure caused by deformity of the chest wall, static or only slowly progressive neuromuscular disease, and healed pulmonary tuberculosis treated surgically with resulting restriction. The role of home mechanical ventilation in the management of rapidly progressive neuromuscular disease is more questionable, bearing in mind the limited prognosis and presence of other symptoms and disabilities. It is too soon to predict its value in the management of chronic obstructive pulmonary disease [14].

Patients who will benefit are best identified on the basis of diurnal symptoms and signs in the presence of an appropriate antecedent. Unfortunately, the symptoms are nonspecific and so insidious that they are often missed by patient and doctor alike until some relatively minor event precipitates a respiratory or even cardiorespiratory crisis. Increasing daytime dyspnoea and lethargy, early morning headache and sleep disruption are all the immediate consequence of nocturnal hypoventilation, and there is often a history of recurrent respiratory tract infections which fail to clear.

In patients with a pure restrictive defect such as kyphoscoliosis, residual volume and total lung capacity may fall before there is any further reduction in vital capacity, perhaps reflecting the tendency to atelectasis and reduced compliance which are known to occur with time [4], in turn a likely consequence of repeated episodes of hypoventilation during sleep [15, 16]. The presence of risk factors for the development of respiratory failure in patients with extrapulmonary restriction should lower the threshold at which the decision is taken to record gas exchange during sleep. These risk factors include onset before the age of 7 yrs [17], a high dorsal curve, marked rotation of the spine, associated rib anomalies, or the presence of respiratory muscle weakness. Patients who suffered poliomyelitis in early childhood often fulfil many of these criteria [18].

Not all patients with documented nocturnal hypoventilation require mechanical ventilation at home.

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Protriptyline reduces the time spent in REM sleep and, if tolerated, is effective in patients with either restrictive or obstructive pulmonary disease as a means of curtailing REM-related nocturnal hypoventilation [19–21]. Arterial blood gas status by day improves as well, and many patients remain in good health for long periods on this regime alone. This treatment is most likely to be successful when there is little or no daytime respiratory failure. A brief initial period of in-hospital treatment with noninvasive ventilation may be necessary to restore normal arterial blood gas tensions and pH. It is worth noting that long-term oxygen therapy is of little value, because worsening hypercapnia is almost inevitable.

Patients whose respiratory failure is inadequately controlled by these means should be considered for a trial of home ventilation, especially if they are symptomatic by day. Unlike those who require ventilatory support for at least some part of the day as well as during sleep, patients who only require mechanical assistance at night do not need a tracheostomy and, indeed, may fare better without, unless they also suffer from bulbar weakness. The choice for noninvasive ventilation lies between the application of negative pressure to the thoraco-abdominal wall or intermittent positive pressure delivered noninvasively through a well-fitting nasal mask (occasionally a mouthpiece). Equipment based on the application of negative pressure was popular at one time [22] but is clumsy, inconvenient, relatively inefficient and accentuates any tendency to upper airway obstruction [23]. Noninvasive positive pressure ventilation by nasal mask is now the preferred option in most centres for all but a minority of applications [24–28].

Equipment

Any ventilator for home use must be robust, simple, inexpensive and easy to maintain. Portability and a battery or mains operated power supply are other desirable considerations. The fact that it is used by unsedated patients, both while fully conscious and during sleep, means that the pattern of inspiration should match as nearly as possible the characteristics of the spontaneously breathing subject, and the machine should be capable of following the patient's own respiratory rhythm (the assist or trigger function) as well as providing a predetermined respiratory cycle during periods of apnoea or extreme hypopnoea. High flow rates are also needed, to achieve inflation of the lungs when in series with the deadspace of a distensible upper airway, and to allow for losses around the nasal mask or through the mouth. These requirements limit the choice of suitable equipment but several models are now available which fulfil the specification.

The choice of nasal mask is also critical; it must fit snugly without creating undue pressure, particularly on the bridge of the nose, the upper lip or the gums and teeth. Great care is needed to avoid pressure necrosis of the skin of the nose, particularly in patients unable to raise their hands to adjust the mask by themselves. The system used to secure the mask to the face should not be heavy or clumsy but must remain in place, even when patients turn from side to side during sleep. Finally, it is often necessary to use a chin-strap to keep the mouth lightly closed, usually securing it to the sides of the head-gear.

The cost of medical care is increasing worldwide but treatment at home for those in need of ventilatory support is undoubtedly less expensive than providing the same service in hospital [29], especially for otherwise able-bodied patients who can handle the apparatus independently and who do not have all the additional requirements associated with a permanent tracheostomy. The initial cost of a noninvasive home ventilation system in the UK is approximately £3,000, and patients treated electively can be acclimatized in less than a week in hospital. After-care is equally important and, if this new medical development is to achieve its full potential, we should follow the lead of those nations which have already developed a system of comprehensive respiratory care for patients at home [30].

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