

LONG TERM VENTILATION IN NEUROGENIC RESPIRATORY FAILURE

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Respiratory failure, often acutely precipitated by aspiration and/or infection, is a common cause of death in advanced nervous system disease. Importantly, chronic respiratory failure, or symptoms relating to respiratory limitation, may occur before generalised disability. For such patients, providing ventilatory support, usually at night, has long been known to improve daytime symptoms, sleep quality, and prognosis in stable or slowly progressive neurological disease.¹ The increased availability and convenience of modern non-invasive ventilation (NIV) has led to a dramatic change in the management of chronic neurogenic ventilatory failure.^{2–5}

Respiratory symptoms, such as cough, choking, dyspnoea, recurrent infection, and respiratory failure may develop for a number of reasons. In respiratory failure the principal problem is usually muscle weakness leading to respiratory pump failure. Although respiratory rate may increase, there is alveolar hypoventilation because of a fall in tidal volume and the arterial P_{CO_2} rises (type 2 respiratory failure). Other factors, such as paralytic scoliosis in the younger patient, significant upper airway obstruction in conditions involving the bulbar muscles, or truncal obesity may further compromise respiratory function. A secondary fall in residual lung volume, micro-atelectasis from ineffective sigh function, basal lung collapse, and a poor cough result in impaired alveolar gas exchange and hypoxaemia. This is easily corrected by oxygen supplementation. Hence the importance of even minor falls in oxygen saturation in neurological disease and the potential danger of treating the oximeter reading rather than the patient.

Alveolar hypoventilation usually initially occurs at night. Eventually, nocturnal hypoventilation is sufficiently severe that respiratory failure persists while the patient is awake. It is important to recognise patients at risk of respiratory failure. Apart from providing NIV, which reduces both morbidity and mortality,^{1–5} other treatments may be needed to improve cough efficiency, avoid aspiration or prevent infection. The decision to assist ventilation in acute neurological disorders is usually straightforward. It is not uncommon, however, that mechanical ventilation is initiated as a result of a respiratory crisis despite warning features.^{6–8} In this situation, both an accurate diagnosis and an estimate of prognosis is essential for careful consideration of the possible benefits and disadvantages of ventilatory support.

CLINICAL FEATURES OF RESPIRATORY INSUFFICIENCY AND FAILURE

Respiratory failure often develops insidiously and symptoms may not be pronounced or complained about (box 1). Breathlessness is particularly unreliable in the context of a patient with limited mobility. Nocturnal hypoventilation and/or recurrent upper airway obstruction with sleep disruption may initially cause no symptoms. Eventually patients or carers notice insomnia, daytime hypersomnolence and lethargy, morning headaches, reduced mental concentration, and depression, anxiety or irritability. In addition, there may be a complaint of abnormal sleep movements, nocturnal confusion, or vivid or distressing dreams. In isolated bilateral diaphragm paralysis, patients may be aware of breathlessness sitting or standing in water, when lying down or during jerky motion, as occurs driving over rough surfaces. In all of these situations, there is upward displacement of abdominal contents that limit the effectiveness of the accessory muscles of respiration.⁹ Orthopnoea may be severe, patients often refusing to lie supine for clinical examination, sometimes even claiming that the supine position causes backache, claustrophobia or a panic attack. There may be few signs until the patient develops respiratory failure or suffers a respiratory arrest. Clues such as an unexplained tachypnoea, the use of accessory muscles or abdominal paradox may be present. The latter consists of inward movement of the abdomen on inspiration while the upper chest expands. It may be more obvious with the patient supine during a sniff manoeuvre. When upright, active abdominal muscle contraction during expiration may mistakenly suggest preserved diaphragmatic function as the diaphragm passively descends at the beginning of inspiration.¹⁰ It is an important, and often missed, sign of profound respiratory muscle weakness. Another important sign is a raised jugular venous pressure indicating pulmonary hypertension caused by recurrent and severe nocturnal hypoxaemia. It is important to recognise that the pharyngeal reflex is not a good guide to the risk of aspiration.

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Box 1: Symptoms and signs of sleep disordered breathing

- ▶ Symptoms
 - may be none
 - dyspnoea
 - excessive daytime sleepiness
 - difficulty sustaining sleep
 - witnessed apnoea or abnormal sleep movements
 - lethargy, morning headache, and poor concentration
 - depression, anxiety, and irritability
 - orthopnoea or immersion intolerance
 - reduced libido or impotence
- ▶ Signs
 - tachypnoea and use of accessory muscles
 - paradoxical movement of the abdominal wall
 - raised jugular venous pressure or other evidence of heart failure
 - evidence of upper airway obstruction

INVESTIGATIONS

In progressive neuromuscular disease, vital capacity and residual lung volume fall because of respiratory muscle weakness and a reduction in chest wall and lung compliance that results from atelectasis and restriction of chest wall movement. Significant diaphragm weakness is associated with a pronounced fall (> 25%) between vital capacity sitting and lying. A normal supine vital capacity is valuable in excluding clinically relevant inspiratory muscle weakness. Low vital capacity values may reflect difficulty in performing a maximal test, due to poor lip seal if there is facial muscle involvement, or pre-existing lung or chest wall disease. In these circumstances, more specific tests of respiratory muscle strength may be required.¹¹ Maximal mouth pressures can be useful but the maximal sniff nasal inspiratory pressure (SNIP) is easier to measure and more repeatable. Normal values are > 80 cm H₂O (> 70 in females) with a lower limit of 35 cm H₂O indicating the need for more sophisticated tests such as measuring transdiaphragmatic or oesophageal pressure, phrenic nerve conduction or stimulation studies, and diaphragmatic electromyogram (EMG).¹¹

Chest radiographs may show unsuspected unilateral diaphragmatic paresis, small lung volumes in global muscle weakness or evidence of aspiration pneumonitis. Fluoroscopic or ultrasound screening may demonstrate paradoxical upward movement of the paralysed diaphragm during inspiration or with a sharp maximal sniff. False negatives and positives are, however, well recognised with this assessment. Waking arterial blood gas tensions (ABGs) are often normal during the early stages of neurogenic respiratory insufficiency. An elevated bicarbonate and mild alkalosis are an indication of significant nocturnal hypoventilation. Overnight oximetry may reveal recurrent desaturations with two characteristic patterns. Repeated arousal from recurrent obstructive apnoeas causes minor dips in oxygen saturation that are rapidly corrected. More severe hypoventilation is indicated by more prolonged and severe desaturations, initially confined to periods of REM (rapid eye movement) sleep, but eventually occurring throughout sleep. In specialist centres, screening with home oximetry has therefore become the investigation of choice in at risk patients. More detailed analysis of the mechanisms of sleep induced respiratory failure requires full polysomnography.

A summary of these investigations is presented in box 2.

Box 2: Investigations of patients at risk of developing neurogenic respiratory failure

- ▶ Essential
 - forced vital capacity—erect and supine
 - chest radiograph
 - venous HCO₃ or arterial blood gas tensions (ABGs)
 - nocturnal oximetry
- ▶ As indicated
 - diaphragm screening
 - maximal mouth inspiratory and expiratory pressures
 - maximal nasal sniff pressure (SNIP)
 - transdiaphragmatic pressure with voluntary manoeuvres
 - polysomnography
 - central motor conduction (magnetic stimulation to cortex or cervical cord)
 - phrenic nerve conduction studies
 - diaphragmatic electromyogram (EMG)

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PATHOPHYSIOLOGY

Adequate ventilation is dependent on respiratory muscle pump, the ventilatory load placed upon it, and the central drive.¹² In patients with neuromuscular disease, more than one factor is often present. A reduction in central drive occurs in sleep, even in health, when respiration is dependent on diaphragm function, especially during REM sleep when the accessory muscles become inactive. At this time, airway resistance also increases, particularly of the upper airways. These factors may thus combine to cause hypoventilation. Sleep is a critical period for respiratory compromise and sudden death in patients with neuromuscular disease or with central disorders of breathing is well recognised.¹³

Respiratory muscles

The relative contribution made by respiratory muscle weakness to neurogenic respiratory failure varies with the particular condition. For example, in motor neurone disease (MND) and acid maltase deficiency the diaphragm is preferentially affected, while in Duchenne and myotonic dystrophy, expiratory muscle dysfunction develops before inspiratory muscle weakness occurs, leading to impaired cough and risk of aspiration and pneumonia.¹⁴ The effect of respiratory muscle weakness may be potentiated by a number of other problems, summarised in box 3.

Sleep apnoea and alveolar hypoventilation

Episodic apnoea is divided into obstructive, central or both. In obstructive sleep apnoea (OSA) there is upper airway obstruction despite normal movement of the intercostals and diaphragm. In central apnoea, all respiratory phased movements are absent. Central alveolar hypoventilation is characterised by a reduced ventilatory response to hypoxia and CO₂ and consequent CO₂ retention in the absence of respiratory pump disease. There is a reduction in the tidal volume during

Box 3: Factors which may precipitate respiratory failure in at risk patients

- ▶ Pulmonary infection including viral
- ▶ Progressive obesity
- ▶ Abdominal distension—for example, ileus or pregnancy
- ▶ Surgery (particularly abdominal)
- ▶ Sedative drugs including opiates

sleep and reduced hypoxic and hypercapnic drive which culminates in central hypopnoea or apnoea. These effects occur primarily during sleep but hypercapnia usually persists by day.

Central control

Neural control of respiration depends on three largely anatomically and functionally independent, though related, pathways. Automatic (metabolic) respiration is the homeostatic system by which ventilation may be altered to maintain acid–base status and oxygenation to the metabolic requirements. It originates in localised areas of the dorsolateral tegmentum, the pons and medulla in the region of the nucleus tractus solitarius and retroambiguus, descending via pathways in the ventrolateral columns of the spinal cord. It has been suggested that the abnormal patterns associated with brainstem lesions may be of localising value, but there is considerable overlap in patterns and it is often impossible to exclude coexisting pulmonary pathology in the acutely ill. Voluntary (behavioural) respiration operates during consciousness and allows modulation of ventilation in response to voluntary actions such as speaking, singing, breath holding, and straining. This system originates in the contralateral cortex, descending via the corticospinal tract to the segmental level. Voluntary control may be impaired by bilateral lesions affecting the descending corticospinal or corticobulbar tract; it is particularly seen in association with destructive lesions of the basal pons or of the medullary pyramids and adjacent ventromedial portion, which may result in the “locked in” syndrome. Limbic (emotional) control accounts for the preservation of respiratory modulation to emotional stimuli including laughing, coughing, and anxiety despite loss of voluntary control. This implies that descending limbic influences on automatic respiration are anatomically and functionally independent of the voluntary respiratory system.¹⁵

NEUROLOGICAL DISORDERS WHICH LEAD TO CHRONIC RESPIRATORY FAILURE REQUIRING LONG TERM VENTILATION

The causes and consequences of acute neurogenic respiratory failure have been reviewed elsewhere in this issue, and the following descriptions concentrate on those conditions leading to long term respiratory impairment.

Central disorders

The effects of brainstem dysfunction on respiration depend on the pathology, localisation, and speed of onset of the lesion. In patients with bulbar disease, particularly vascular or demyelinating, the combination of impaired swallow, abnormalities of the respiratory rhythm, reduced vital capacity, and reduced or absent triggering of a cough reflex all increase the risk of aspiration pneumonia. Nocturnal upper airway occlusion may also contribute to respiratory impairment. The most common cause is cerebrovascular disease. Unilateral or bilateral lateral tegmental infarcts in the pons (at or below the level of the trigeminal nucleus) leads to apneustic breathing and impairment of CO₂ responsiveness, while similar lesions in the medulla (for example, lateral medullary syndrome) may result in acute failure of the automatic respiration. Infarction of the basal pons (“locked in” syndrome) or of the pyramids and the adjacent ventromedial portion of the medulla may lead to a highly regular breathing pattern but a complete inability to initiate any spontaneous respiratory movements.¹⁶ Respiratory failure requiring long term ventilatory support has also been

described as a result of post-rubella and varicella encephalomyelitis and acute disseminated encephalomyelitis. Multiple sclerosis is occasionally associated with central disorders of respiratory rate and rhythm.¹⁷ Multisystem atrophy is characterised by paresis of the vocal cord abductors (posterior cricoarytenoids), leading to severe upper airway limitation during sleep and giving rise to the characteristic presenting feature of severe nocturnal stridor. These patients occasionally require long term domiciliary ventilatory support, and tracheostomy may be necessary if stridor threatens airway obstruction. Traumatic, demyelinating or vascular lesions of the spinal cord, particularly at high cervical levels, may selectively affect respiratory control. Lesions of the anterior pathways, as may occur following cordotomy, lead to loss of automatic control and sudden nocturnal death from apnoea.

Cervical cord

The respiratory effects of traumatic or vascular lesions of the spinal cord depend on the timing of onset and the extent of involvement of the phrenic nerve supply (C3–C5). Complete lesions usually lead to sudden respiratory arrest and death unless immediate resuscitation is available. Patients with lesions at or above C3 and some patients with lesions at a lower level may require prolonged or even permanent ventilator support.¹⁸ Tetraparesis with levels below C4 lose intercostal and abdominal muscle function while maintaining diaphragm and spinal accessory muscles.

Anterior horn cell

During acute poliomyelitis respiratory insufficiency occurs as a result of respiratory muscle weakness or involvement of the central respiratory control mechanisms. Respiratory insufficiency may develop many years after poliomyelitis, even in the absence of any obvious respiratory involvement during the acute illness or convalescent phase.¹⁹ Respiratory insufficiency is the common terminal event in MND caused either by respiratory muscle or bulbar weakness leading to hypoventilation, aspiration, bronchopneumonia or defects in central control. However, an important proportion of patients with MND may develop respiratory insufficiency early in the course of the disease, and may present with respiratory failure or even respiratory arrest.⁷ The provision of NIV can provide symptomatic relief and increase life expectancy. These benefits must be balanced against the difficulties of compliance in severely disabled patients, the demands on carers and relatives, the practical problems of increasing dependence on ventilatory support, and possible difficulties in managing terminal care. While many of these difficulties can be avoided by careful discussion with patients and their carers, some patients may wish to avoid embarking on ventilatory support.

NIV improves quality of sleep, and other aspects in quality of life scores, and may improve survival in MND by preserving lung function, providing better secretion clearance (see later) or preventing decline in vital capacity and gas exchange. With disease progression, increasing periods of NIV may be required or may become semi-continuous. In these circumstances, different methods of providing NIV (negative, as well as positive, pressure) may prevent the progression to tracheostomy ventilation although, for some, this is preferred. Tracheostomy ventilation inevitably results in difficulties in domiciliary management, not least in the funding and provision of carers, and carries a risk of complications while recurrent aspiration may occur even with a cuffed tube. There is also a concern that tracheostomy ventilation may lead to prolonged, and possibly unwanted, survival in the face of severe disability.²⁰ Ventilatory

failure requiring long term support has been described in all three types of proximal spinal muscular atrophy and occurs as a consequence of weakness of respiratory and bulbar muscles, scoliosis, and a possible central component.^{21 22}

Neuropathy

Approximately one third of patients with acute inflammatory demyelinating polyneuropathy (Guillain-Barré syndrome) develop respiratory failure requiring mechanical ventilation.²³ Ventilatory failure is primarily caused by inspiratory muscle weakness although weakness of the abdominal and accessory muscles of respiration, retained airway secretion leading to aspiration, and atelectasis are all contributory factors. The associated bulbar weakness and autonomic instability contribute to the necessity for control of the airway and ventilation. The decision to intubate may be in anticipation of further deterioration and a vital capacity < 15 ml/kg has been suggested as a good guide. In one study⁸ many patients were intubated “out of hours” and in 16% as an emergency, indicating that care of at risk patients may be best in the intensive care unit. The requirement for long term respiratory support is unusual but occurs in patients with acute motor axonal neuropathy causing severe generalised paralysis of all volitional musculature. Weaning of such patients may still be possible and transfer to a weaning centre may be appropriate. Long term domiciliary support with NIV is occasionally necessary in chronic idiopathic demyelinating polyneuropathy, multifocal motor neuropathy, and hereditary motor and sensory neuropathy.²² Neuralgic amyotrophy may present with dyspnoea and orthopnoea, due to selective or isolated involvement of the phrenic nerve causing unilateral or bilateral diaphragm paresis. This is probably the most common cause of ventilatory failure due to isolated phrenic nerve palsy. The prognosis for recovery is variable and many patients continue to require long term ventilatory support.

Neuromuscular junction

Respiratory failure in myasthenia gravis often results from a myasthenic crisis (usually precipitated by infection) and occasionally a cholinergic crisis, thymectomy or steroid myopathy. However, diaphragm weakness may occur even when there is only mild peripheral muscle weakness, and long term domiciliary ventilatory support may be required.

Muscle

Respiratory muscle weakness is a common cause of morbidity and mortality in the muscular dystrophies.^{14 24–27} In Duchenne muscular dystrophy respiratory failure develops late in the course of the disease and is associated with intercostal and expiratory muscle weakness, as well as kyphoscoliosis, reduced lung compliance, and bulbar weakness causing aspiration and repeated infection. Progressive respiratory muscle weakness culminates in nocturnal hypoventilation while expiratory muscle weakness leads to poor cough. The relatively late occurrence of respiratory failure, often in association with bulbar weakness, makes it desirable to avoid intermittent positive pressure ventilation via a tracheostomy despite the advantages of providing airway access. NIV is well tolerated and leads to a significant improvement in the quality of life.^{14 27} Respiratory involvement in Becker’s muscular dystrophy, the limb girdle dystrophies, is compounded by scoliosis, with respiratory muscle weakness usually occurring together with limb weakness. In facioscapulothoracic dystrophy respiratory involvement is less common but there may be selective diaphragm involvement and respiratory tract infections are common despite the absence of

Abbreviations

ABGs: arterial blood gas tensions
CPAP: continuous positive airway pressure
EMG: electromyogram
IPPV: intermittent positive pressure ventilation
MND: motor neurone disease
NIV: non-invasive ventilation
OSA: obstructive sleep apnoea
PECF: peak cough expiratory flow rate
REM: rapid eye movement
SNIP: sniff nasal inspiratory pressure

clinically overt bulbar weakness. In contrast acid maltase deficiency is characterised by early and selective diaphragm weakness, often with minimal involvement of other respiratory and bulbar musculature.²⁸

Congenital myopathies, including mitochondrial and nemaline myopathy, may present with respiratory insufficiency caused by diaphragm weakness or develop alveolar hypoventilation early in the course of the disease. This may be associated with skeletal dysmorphism (pectus excavatum and pes cavus), kyphoscoliosis contractures, and progressive limb and trunk weakness. Progressive scoliosis and restrictive ventilatory insufficiency is an important complication of Emery-Dreifuss muscular dystrophy and the rigid spine syndrome, and ventilatory support is occasionally necessary. In dystrophia myotonica respiratory involvement is multifactorial. Respiratory muscle weakness may affect both the diaphragm and expiratory muscles leading to a poor cough, restrictive lung defect, and alveolar hypoventilation. There is little evidence that myotonia of the respiratory muscles is a significant factor. A central abnormality contributes to both alveolar hypoventilation and the hypersomnolence of the condition. There may be an undue sensitivity to anaesthetics and sedatives. Long term ventilatory support with NIV may be necessary but these patients are difficult to treat, partly because of cognitive problems.^{29 30} Respiratory muscle weakness requiring long term ventilation also occurs in the inflammatory myopathies such as polymyositis and dermatomyositis. This is caused by generalised respiratory muscle weakness with or without bulbar weakness. There may be co-existing interstitial pulmonary involvement leading to fibrosis.

MANAGEMENT OF CHRONIC RESPIRATORY FAILURE OF NEUROMUSCULAR ORIGIN

In contrast to acute respiratory failure, the respiratory failure associated with chronic neuromuscular disease can be managed at home using a variety of methods of mostly non-invasive techniques of assisting ventilation. Before dealing with these, the indications for starting support need consideration.

Indications for instituting ventilatory support

Respiratory crises, usually infective, are not always preventable but it is still important to anticipate the risk of ventilatory failure at an early stage. Some studies have reported improved survival in MND when NIV is started before daytime respiratory failure is present,³¹ while early “prophylactic” ventilation resulted in a worse prognosis in one study in Duchenne’s muscular dystrophy.²⁶ Despite these contrasting studies, there is general agreement that when symptoms of sleep disordered ventilation are present, such as daytime sleepiness, morning headaches or cognitive impairment, **and** there is evidence of nocturnal arousals or hypoventilation, NIV should be

recommended.¹⁻⁵ Symptom scores, such as the Epworth sleepiness score, are of less use in neuromuscular disease than in conventional non-neurological OSA. This is partly because of acclimatisation to symptoms, partly the expectation of deterioration in the more progressive diseases, and also because OSA is associated with more sleep arousals leading to sleep deprivation. OSA occurs more frequently in neuromuscular disease than in the normal population. For instance, we found that > 40% of polio survivors referred with fatigue and other symptoms of the post-polio syndrome have OSA caused by occult or mild bulbar weakness. OSA is also more common in other neurological diseases, especially with bulbar involvement.

Continuous positive airway pressure (CPAP) is effective in treating OSA. It stents open the upper airway and, by reducing arousals, provides relief of daytime fatigue. CPAP therapy does not correct alveolar hypoventilation and in progressive neurological disease we would initiate NIV in the absence of symptoms if significant nocturnal hypoventilation is revealed by oximetry or the daytime P_{CO_2} is > 7.5 kPa. Risk factors for respiratory failure include clinical evidence of diaphragmatic weakness, such as paradox or orthopnoea, a vital capacity < 50% predicted, or SNIP < 30 cm H₂O. As the vital capacity falls progressively, in many conditions, serial measurements can guide both discussion of NIV and its initiation. Once a decision has been made to recommend ventilatory assistance, it is important to tailor the method with respect to the severity of associated non-respiratory disability, the availability of family support, or suitable helpers, and the home living conditions. For example, a cuirass may be more suitable if upper limb function is poor (applying or removing a mask may be difficult or impossible), while a rocking bed is ideal in isolated diaphragmatic failure. Although mechanical and negative pressure techniques were commonly used in the past, the convenience, reliability, and ease of use of positive pressure ventilators make these the first choice in the majority.

Glossopharyngeal (frog) breathing

Glossopharyngeal (frog) breathing involves incremental inflation of the lungs by gulping air into the oropharynx, closing the mouth and soft palate, opening the larynx, and forcing the air from the pharynx into the trachea. This can more than double vital capacity and assist in producing an adequate cough. It originated in the acute poliomyelitis period and many survivors with limited ventilatory function are able to manage for long periods of the day by glossopharyngeal breathing alone. Acquiring the skill to perform frog breathing semi-automatically is quite demanding and teaching it is sadly a dying art.

Diaphragm pacing

The phrenic nerve may be electrically stimulated to cause diaphragmatic contraction. Its use is largely limited to spinal cord injury. There remain significant technical issues but silver electrodes have overcome the problem of decreasing response to stimulation with time while improved electronics have increased durability. Pacing can provide an alternative to intermittent positive pressure ventilation (IPPV), but a tracheostomy is still required and the high cost limits application.³² Its value is in freeing the individual from the encumbrance of a mechanical ventilator, so increasing mobility.

Negative pressure ventilation

This was, for many years, the mainstay of treatment for patients with chronic neuromuscular respiratory failure. Although there are several methods of delivering negative pressure ventilation, all rely on the principle of enclosing the



Figure 1 Iron lung.

chest and abdomen in an airtight rigid chamber from which air is intermittently evacuated. The resulting sub-atmospheric pressure around the lower thorax and abdomen causes air to be drawn into the lungs. Examples of negative pressure devices include tank (iron lung) (fig 1), jacket (Tunnicliffe), and cuirass ventilators. Although these devices are cumbersome, the main problem is the promotion of upper airway obstruction.³³ Home use is now confined to patients who cannot tolerate mask NIV. We continue to use tanks in the respiratory unit to facilitate extubation or as a temporary means of providing ventilation if continuous mask ventilation has caused significant facial skin trauma. A cuirass ventilator can also be used to provide respite from mask ventilation.

Rocking bed

Breathing can be assisted by rocking a patient consecutively head up and head down. The technique is surprisingly effective and well tolerated, especially in patients whose muscle weakness is predominantly diaphragmatic. It is less effective at increasing alveolar ventilation than either negative or positive pressure techniques, and dysynchrony between spontaneous breathing effort and the timing of the rock can occur. It is the only method of respiratory assistance to leave the patient unconnected to apparatus. Disadvantages include non-portability, high maintenance costs, and engineering problems in achieving a smooth motion.³⁴

Non-invasive intermittent positive pressure ventilation

Non-invasive IPPV (conventionally now abbreviated to NIV) is the most efficient method of increasing alveolar ventilation without intubation. Portable electronic ventilators provide a preset tidal volume or pressure. As NIV is inevitably associated with leak, pressure controlled devices are better able to compensate for this and so ensure adequate support to spontaneous breaths. Sensitive flow triggers enable normal breathing to be supported, increasing comfort, while a back up mandatory pattern of breathing is usually employed to ensure ventilation overnight when reduced respiratory effort may fail to trigger a breath. NIV is applied via a tightly fitting nasal or facial mask or nasal "pillows". Comfort with minimal leak can be achieved in all patients with the range of masks now available. Continuous or semi-continuous NIV can be provided by using two or more different interfaces, and varying the sites of pressure can avoid skin necrosis. This is particularly important for periods of increased dependency, such as during a respiratory infection. Mouth leak is common during sleep and may impair ventilator triggering. Air leak is reduced, by forward



Figure 2 Duchenne full face mask

displacement of the tongue, when positive pressure is also applied during expiration (so called bi-level pressure support), but a chin strap is often also required. When commencing NIV, a full face mask may be best to ensure adequate support (fig 2), switching to a nasal mask (fig 3) when acclimatised. NIV is usually provided using a single ventilator tube fitted with an exhalation port, or valve, to reduce rebreathing.

The adequacy of overnight ventilation will be indicated by relief of sleep related symptoms. Commonly patients, and carers, will report a considerable improvement in daytime functioning including general health, muscle strength, and energy.³⁵ Breathlessness is not always improved. More objective measures of control include oximetry, transcutaneous CO₂ monitoring, and daytime ABGs. Common side effects include a dry mouth, nasal discharge (or drying), and abdominal bloating from air being swallowed. Although topical steroid sprays can be effective, humidification is needed in about 50% of cases. A choice of several different machines and interfaces is available and some are better suited for wheelchair mounting, while external battery packs increase mobility and can make foreign travel possible. Other machines have automatic power switching in the event of mains electricity failure. This is essential in the semi-continuous NIV or tracheostomy ventilated user.

Tracheostomy ventilation

Complexity of care is much increased by tracheostomy ventilation and may prevent discharge to the home. In the past, it was



Figure 3 Nasal mask.

used routinely in the more rapidly progressive conditions, especially in the USA and Japan, but its use was more directed to the spinal injured patient or those with slowly progressive conditions such as old polio, non-Duchenne muscular dystrophy or the myopathies in much of Europe. Quality of life scores, by both patients and carers, are consistently better with NIV than tracheostomy ventilation. However, it may not prove possible to wean a patient to NIV once a tracheostomy has been performed and, in some conditions such as MND, the diagnosis may only be made following a respiratory crisis.^{6,7} In the progressive conditions, once NIV becomes semi-continuous (> 18 hours per day), tracheostomy ventilation may offer advantages and yet retain a good quality of home life. By employing non-cuffed tubes, speech can be preserved and, with better access to the lower respiratory tract for suctioning, minor aspiration can be managed without serious consequences. It is not an easy choice and we advise considerable counselling for patients considering switching to invasive ventilation.

Cough and cough assist techniques

While alveolar hypoventilation results from weakness of the inspiratory muscles, an effective cough is dependent on adequate inspiratory capacity, rapid contraction of the expiratory muscles, and sufficient bulbar control to allow an explosive expulsion of air. In some neuromuscular disease, early abdominal or bulbar muscle involvement may increase the risk of acute episodes of respiratory failure despite adequate nocturnal ventilation. In such individuals, improving cough may be important in preventing lower respiratory tract infection and possibly delaying the onset of respiratory failure. Cough is not commonly quantified and is most commonly assessed simply by listening. It is poor when vital capacity is < 1.5 litres and ineffective when < 850 ml. Some authorities suggest that when the peak cough expiratory flow rate (PECF) is < 250 litres/min, cough will be ineffective, and that when < 160 litres/min the risk of repeated infections demands the regular use of cough assist techniques.³⁶ When measured with a hand held peak flow meter, we have not found these cut offs useful in risk assessment as many patients with PECF < 160 litres/min remain well without specific attention. More usefully, a history of repeated minor chest infections, especially if there is bulbar dysfunction, would indicate the need for further assessment. An expulsive cough, capable of clearing secretions in the larger airways, may be possible by manually assisting spontaneous coughing effort with abdominal pressure. Carers can easily learn the technique. Although it may not be effective in all patients, effectiveness compared with unassisted cough has been demonstrated.³⁷

Breath “stacking”, using a simple volume ventilator (Cape Minor), can be used to augment cough (and also to intermittently augment spontaneous breathing by day in the more dependent). Breath stacking can also be performed using the normal night time ventilator. Indeed, one of the benefits of providing NIV may be an improvement in ventilation to otherwise collapsed lung units and a consequent reduction in lower respiratory tract infections. Another technique to improve clearance of secretions involves hyperinflation followed by application of negative pressure, effectively sucking air out of the chest. The switch from positive to negative pressure occurs within 200 ms and is made to coincide with patient effort. Higher maximal expiratory flows have been demonstrated with this technique and case series have suggested an important role for the use of the CoughAssist MI-E (Emerson, Massachusetts, USA) in the prevention of respiratory morbidity in neuromuscular disease.³⁸ A recent

controlled study confirmed improved peak expiratory flow rate³⁹ and we have found mechanical insufflation/exsufflation clinically useful during infective illness.⁴⁰ At present, the value of providing cough machines for domiciliary use has not been established, although it is common practice in the USA.

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CONCLUSION

Many patients dependent on long term mechanical ventilation can be well maintained in the home environment. Such domiciliary ventilation requires detailed assessment and attention to the availability of home care, equipment maintenance, and hospital access. Suitability for domiciliary support is determined by a number of factors including the degree of respiratory dependence, associated non-respiratory disability, and the family back up. It is necessary to provide patients and carers with detailed education in both the techniques and other equipment such as suction and communication aids. In the more dependent patient, a back up ventilator and provision of emergency power may be required while regular medical review (including annual influenza vaccination) and planned access to the respiratory unit for intercurrent infection is necessary for all home mechanical ventilation patients. Equipment maintenance can be provided by the manufacturers, but larger units usually provide in-house technical support backed up by medical and nursing advice by telephone. Regional centres, with appropriate arrangements for funding the cost of the equipment, its maintenance, and supervision, are therefore preferred. The provision of domiciliary ventilation is increasingly well organised with both national and international organisations to oversee development. Home mechanical ventilation can offer patients and their families a good quality of life and is a cost effective alternative to prolonged hospitalisation.

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