Assisted ventilation in cystic fibrosis: nursing care

In this article the author explains nasal intermittent positive pressure ventilation (NIPPV) for patients with cystic fibrosis (CF). Indications for use are noted before nursing care of the patient is described in detail.

Date of acceptance: July 9 1998.

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Cystic fibrosis (CF) is the most common lethal genetic disorder affecting Caucasian populations, particularly those of northern European origin. It is a hereditary disease affecting cells of the exocrine glands. The faulty gene responsible is recessive and has been identified as lying on chromosome number 7. Affected individuals lack the protein that enables transport of chloride ions across cell membranes. This results in the production of thick mucus which obstructs the intestinal glands, pancreas and bronchi. Currently, the average age at death is 31 years.

Treatment options have been augmented by opportunities for surgical intervention. Royal Brompton Hospital serves the largest number of CF patients of any unit in Europe. Some of these patients have been accepted for heart-lung transplantation, however, they can develop severe life-threatening respiratory failure before suitable donor organs become available. In this situation nasal intermittent positive pressure ventilation (NIPPV) may be used to keep the patient alive.

NASAL INTERMITTENT POSITIVE PRESSURE VENTILATION

Intermittent positive ventilation through an endotracheal or tracheotomy tube has been the mainstay of respiratory assistance for many years (Branthwaite 1991). With advances in technology, NIPPV became possible in the mid-1980s (Braun 1987). The respiratory condition of CF patients usually worsens, despite intensive treatment comprising physiotherapy, intravenous and nebulised antibiotics, oxygen therapy, nutritional support and bronchodilators. However, NIPPV is effective in such chronic respiratory failure (Baculard et al 1993, Hodson et al 1991).

Observing two CF patients ventilated conventionally before transplantation, Yacoub et al (1990) showed that the patients needed many weeks in intensive care after surgery because of their inability to be weaned off the ventilator and cough effectively. Patients treated with NIPPV before transplantation did not have similar problems. Piper et al (1992) looked at four patients treated with NIPPV nocturnally with a diagnosis of hypercapnic respiratory failure. They reasoned that ventilatory assistance during sleep might improve gas exchange as decreases in arterial oxygen saturation during rapid eye movement sleep have been documented in CF patients and others with severe lung disease (Tepper et al 1983). All patients had failed to respond to conventional therapy. Within a few days of commencing NIPPV, all reported improved length and quality of sleep. There was lessening of the degree of hypercapnia and increase in respiratory muscle strength. The authors hoped that these advantages would benefit particularly those patients awaiting transplantation.

Characteristics of NIPPV

The main characteristics of NIPPV are that it is delivered non-invasively via a nasal mask and that the patient triggers breaths, but the ventilator operates automatically to give missing breaths if inspiratory effort is lacking. Unlike for more invasive ventilation techniques, NIPPV patients do not require sedation, nor do they need to be in an intensive care unit. Therefore, they can eat, drink, talk and move relatively freely and their general physical condition is better maintained than when conventional ventilation is used. This means that NIPPV is cost-effective as well as comfortable for patients.

There are two types of nasal ventilators, volume preset and pressure preset. Both types can be used to control ventilation completely or to augment spontaneous respiratory efforts and both have advantages and disadvantages (Simonds 1996). The pressure preset models are smaller and more compact than volume preset apparatus and, as a consequence, are usually less powerful in terms of flow generation.

Volume preset ventilators

Creation of a slight negative pressure by attempting to breathe triggers the ventilator to deliver a preset tidal volume to the patient. If the ventilator is in the 'assist/control' mode, an expiratory time is set so that if the patient fails to trigger the machine within a predetermined space of time, another breath will be delivered automatically. This ensures that adequate ventilation is maintained.
Pressure preset ventilators Instead of delivering a particular volume of gas to the patient, pressure preset ventilators allow a chosen airway pressure to be achieved before a breath is deemed to be complete. A pressure level is set so that on initiation of a triggered breath, air flows from the ventilator to the patient’s lungs until the pressure in the entire system, that is lungs and ventilator, reaches the preset level – at which point the ventilator moves into the expiration segment of the cycle.

The Brompton PAC Patients awaiting transplantation at Royal Brompton Hospital are treated on the Brompton PAC, a volume preset ventilator. This machine provides high inspiratory flow, corresponding tidal volume and minute ventilation at normal respiratory frequencies. By ‘tuning’ respiratory flow rate, inspiratory time and expiratory time, the ventilatory pattern provided by the Brompton PAC is adjusted to match that of spontaneous breathing and medical requirements. Air leaks set off the low pressure alarm, and the high pressure alarm will sound when there is an obstruction, for example, when the user coughs. Inspiratory pressures are displayed on a colour-coded pressure gauge.

**NURSING CARE**

Once a decision has been taken that NIPPV may be of value to the patient, either immediately or as a future option, the patient should receive all relevant information about NIPPV and be helped by family and friends and the multidisciplinary team to make an informed decision regarding treatment. The patient’s right to refuse treatment must be acknowledged.

Before introducing NIPPV to patients, it is important to establish or perhaps reinforce their knowledge and understanding of the disease and significance of transplantation. When able, patients should be taught how to use and care for the equipment themselves. NIPPV is a simple technique and most patients soon learn to handle the equipment independently, a reassuring fact for anxious patients. Learning to be in charge of the equipment can also be beneficial for building self-esteem (Sim 1993).

It is important to keep a record of the amount of time NIPPV is used. Side effects can occur and nurses must be vigilant to spot them and plan suitable care to cope with them. Possible side effects include: mask discomfort, dry nose, air leaks, eye irritation and gastric distension (Foglio et al 1992).

**Nasal bridge sores/pressure necrosis**

Pressure sore problems related to the mask are easier to prevent than cure. A carefully fitted mask, use of polystyrene wedges or bubble masks to reduce pressure on the nasal bridge, and protective skin dressings, such as Granuflex, are useful preventive measures. If these fail, switching to nasal plugs, which ventilate directly into the nose, allows time for healing without interrupting NIPPV. Simple advice may be all that is needed to avoid obvious problems. For example, the patient should be discouraged from pulling the headgear straps too tightly (Jones et al 1994).

**Rhinitis, nasal dryness and streaming**

Minor nasal symptoms occur frequently in patients new to NIPPV. Some experience nasal dryness, nasal congestion and streaming. Short-term use of 0.5 per cent ephedrine nose drops will reduce nasal congestion, and ipratropium nasal spray can reduce streaming. Long-term nasal steroid spray has been used in patients with allergic rhinitis and persistent nasal symptoms. Adding humidification via the Brompton PAC can also help. Nasal symptoms are exacerbated by mouth leaks and lessen when the leak is corrected (Simonds 1996).

**Mouth leaks**

Mouth leaks during NIPPV occur frequently, both during wakefulness and sleep. Robert et al (1991) carried out polysomnographic monitoring and showed that leaks from the mouth during sleep were responsible for increased arousals and poor ventilatory efficiency. Leaks around the top of the mask can lead to soreness of the eyes. This can be dealt with by improving the fit of the mask. Mouth leaks during sleep can be reduced by the patient using a chin strap. If the patient continually experiences air leaks, a full face mask may be necessary.

**Gastric distension**

Abdominal bloating and gastric distension are often more sporadic than other side effects (Simonds 1996). Charcoal tablets, peppermint water or alginate preparations can be used to treat this.

**Difficulty in maintaining adequate dietary intake**

Some patients become dependent on NIPPV as they wait for transplantation, using it up to 20 hours a day. This can prove a problem when trying to maintain the patient’s calorie intake and weight. For these patients, percutaneous endoscopic gastrostomy feeding may be considered as an alternative (Duncan-Skingle and Foster 1995). Nursing interventions include monitoring patients’ dietary intake, recording weekly weight, prompt reporting of weight loss and close collaboration with dieticians.

When the patient is off NIPPV, high calorie snacks and compliance with pancreatic enzyme regime must be encouraged. Nurses must assess activities of daily living (Roper et al 1993) and plan them in terms of patients’ energy requirements, how much they feel capable of doing, number of rest periods required in between activities, and amount of time required to complete tasks (Williams 1993). The goals set should be short term and realistic.
**Psychological needs** Preparation for transplantation and undergoing a new and potentially alarming therapy concurrently adds to patients’ stress, and resulting psychological needs must be assessed and, if necessary, care planned.

Social isolation and loneliness can result from restricted mobility. Feelings of low self-esteem can become a problem. Guyatt et al (1987) found embarrassment regarding symptoms, disability and medication to be important to many patients. They will experience fear and anxiety through deterioration in their condition. Concern about the future is inevitable, as commencing NIPPV is a sign that the need for transplantation is becoming acute.

It is an integral part of care to help reduce patients’ fear and anxiety (Bott and Moran 1996). Anxiety will increase if patients do not understand their condition or the ventilatory equipment around them. Effective communication, counselling and health education are important measures in alleviating patients’ stress (Sim 1993). As Bott and Moran (1996) advised, throughout treatment careful explanation, constant reassurance and praise, combined with the use of relaxation techniques, are key factors.

Nurses should possess good interpersonal skills, give time, demonstrate willingness and ability to listen, and be able to offer help and advice on how best to cope (McSweeney and Labuhn 1993). As well as looking after the patient, it is important to care for family and friends. These problems can impose major changes on the lifestyle of individuals, family and immediate social contacts (Patrick and Peach 1989).

Listening, clarifying and offering advice and information are major aspects of the nurse’s role (Robinson et al 1991). Carers may feel guilty about their own needs and be reluctant to ask for help (Wade 1983). Nurses should be aware of this and offer support accordingly.

When NIPPV is an elective treatment, it can be introduced gradually. This allows time to familiarise patients with the technique, the equipment and its care.

This also gives time and opportunity for discussion and questions that will help allay both patients’ and carers’ anxiety.

The shortage of donor organs means that some patients will die before transplant. As Hodson et al (1991) stated: ‘It may not be appropriate to use any form of mechanical ventilation in a CF patient in terminal respiratory failure, who has already had maximal medical treatment, if he has not been accepted for transplantation. There would be no way forward and the patient could spend many weeks on the ventilator before death. However, in patients already assessed and accepted for transplantation who suddenly deteriorate with unacceptable hypercapnia and hypoxia, the use of NIPPV until transplantation can be performed, is cost effective and may be a life saving procedure.’

**CONCLUSION**

Advances in the medical care of patients with CF has led to parallel developments in their nursing care. When once palliative care was the only option for patients whose condition was deteriorating, both transplantation and NIPPV have offered new challenges to nurses as well as patients. Care of the patient on NIPPV will continue to evolve as techniques improve and nurses must continue to adapt their care for this patient group. Combining routine and technologically advanced nursing skills, encompassing spiritual and psychological care, presents a significant but potentially rewarding challenge for nurses caring for these patients.

**REFERENCES**


