Eur Respir J
1991, 4, 524–527

Non-invasive mechanical ventilation for cystic fibrosis patients - a potential bridge to transplantation

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Non-invasive mechanical ventilation for cystic fibrosis patients - a potential bridge to transplantation. M.E. Hodson, B.P. Madden, M.H. Steven, V.T. Tsang, M.H. Yacoub.

ABSTRACT: The case histories of six cystic fibrosis patients awaiting heart-lung transplantation are reviewed. They all deteriorated with severe hypoxia and hypercapnia before donor organs became available. Nasal intermittent positive pressure ventilation was used in preference to conventional ventilation with excellent results in four patients. There were no episodes of hypotension or toxaemia and the patients were in a stable condition at the time of surgery and made an excellent post-operative recovery. The patients who were transplanted and the patient who died, for whom suitable donor organs did not become available, probably had a more comfortable time than they would have done if treated with conventional ventilation.

This method of ventilation appears to be a useful bridge to transplantation when a patient suddenly deteriorates. It gives them a chance of survival for a few more days or even weeks during which time an urgent search for donor organs can be made. This is also a very cost effective method of ventilation and does not encroach on conventional Intensive Care Unit (ICU) facilities.

Eur Respir J., 1991, 4, 524–527.

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Keywords: Cystic fibrosis; heart-lung transplantation; non-invasive ventilation.

Received: March 19, 1990; accepted after revision February 26, 1991.

Heart-lung transplantation for patients with cystic fibrosis (CF) who have end-stage disease is now becoming established [1] with survival to one year 78% and two years 72% [2]. Nasal intermittent positive pressure ventilation (NIPPV) delivered non-invasively through a well-fitting nasal mask has been shown to be of value in chronic respiratory failure [3, 4] and more recently in acute respiratory failure [5].

We have recently been faced with a number of CF patients who have been accepted for heart-lung transplantation but who developed severe life-threatening respiratory failure before suitable donor organs became available. Donor organs have to be closely matched to the recipient for blood group, size and cytomegalovirus antibody status. In this situation we have used NIPPV as a means of keeping the patient alive in the hope that suitable donor organs would become available. The most recent cases are detailed here. An earlier patient has already been reported [5].

Patients and methods

Six patients with CF (table 1) had been assessed and accepted for a heart-lung transplantation. Their condition deteriorated despite maximum medical treatment including physiotherapy, bronchodilators both by nebulizer and intravenously, appropriate antibiotics, controlled oxygen therapy and nutritional support. When the patient's clinical state and blood gases were such that either their degree of hypoxia or hypercapnia with conventional treatment was considered to be a grave risk to their survival, they were ventilated using the Brompton Pneupac ventilator. The decision to ventilate was a clinical decision taking into account their blood gases, their rate of deterioration, pulse rate, respiratory rate and degree of exhaustion. Some patients were more tolerant of hypoxia and hypercapnia than others.

In addition to gross respiratory failure, Cases 3 and 4 had marked right ventricular failure which could not be controlled with diuretics prior to improving their oxygen saturation using the ventilator.

NIPPV was established using the Brompton Pneupac ventilator (figs 1–3) manufactured by Pneupac Ltd, Luton. The ventilator is a flow-generated, time-cycled machine which delivers a predetermined tidal volume either in response to the initiation of a spontaneous breath by the patient, or automatically. Oxygen was added through a port in the nasal mask. Tidal volumes used were considerably greater than during conventional mechanical ventilation to compensate for leaks through the mask and mouth. Patients needed detailed and careful instruction in the use of the machine and most of them used a chin strap when sleeping to avoid excess

Table 1. – Data for six patients accepted for heart-lung transplantation

			When put on HLT waiting list		Before ventilation			On ventilator			Duration of NIPPV	Outcome
Case	Age yrs	Sex	FEV_1 *l*	FVC *l*	Inspired oxygen	Pao_2 kPa	$Paco_2$ kPa	Added oxygen $l \cdot min^{-1}$	Pao_2 kPa	$Paco_2$ kPa	Days	
1	36	F	600	1210	Air	3.9	9.7	2	10.5	7.7	10	HLT. Extubated at 48 h. Discharged from ICU at 6 days postop.
2	21	M	800	2500	28%	3.9	11.6	2	6.9	11.3	3.5	HLT. Extubated at 36 h. Discharged from ICU at 6 days postop.
3	17	M	520	740	60%	4.3	10.9	2	8.2	8.2	3	HLT. Extubated at 6 h. Discharged from ICU at 13 days postop.
4	29	M	650	900	35%	6.5	8.4	1	10.9	8.7	36	No HLT organs available. Bilateral single lung transplant. Died one week after surgery of bronchial anastomotic problems and sepsis.
5	19	F	560	960	30%	5.3	9.9	2	9.0	8.8	15	No organs available. Died of pulmonary sepsis.
6	27	F	350	970	30%	8.99	14.87	2	13.36	11.95	17	HLT extubated at 18 h. Discharged from ICU 3 days postop.

HLT: heart-lung transplant; NIPPV: nasal intermittent positive pressure ventilation; FEV_1: forced expiratory volume in one second; FVC: forced vital capacity; Pao_2 and $Paco_2$: arterial oxygen and carbon dioxide tension, respectively; F: female; M: male; ICU: Intensive Care Unit.

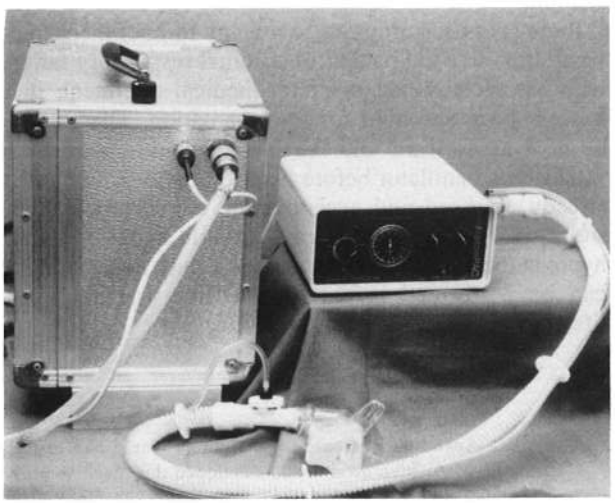

Fig. 1. – Brompton Pneupac ventilator showing the airPAC air compressor and the BromptonPAC ventilator.

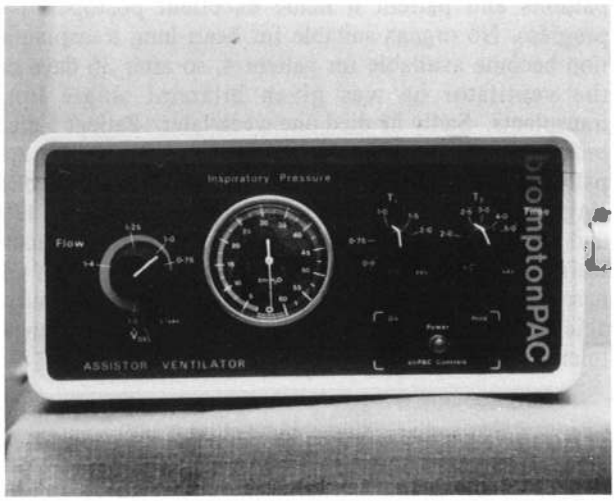

Fig. 2. – A close up view of the BromptonPAC showing the controls for flow, expiratory and inspiratory time.

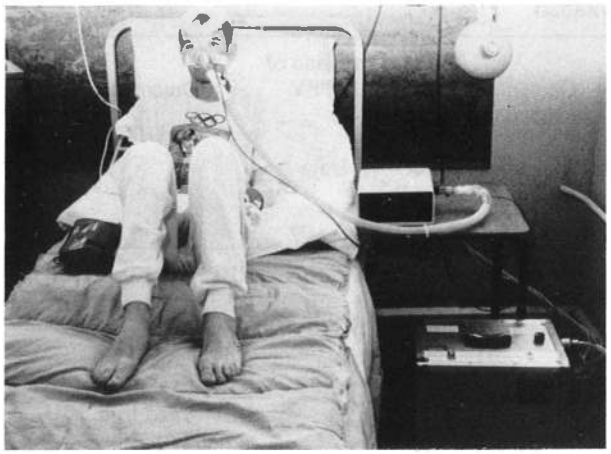

Fig. 3. – Cystic fibrosis patient awaiting heart-lung transplantation using the ventilator.

loss of gas through the mouth. The exact settings for flow rate, inspiratory time and expiratory time were adjusted depending upon the patient's comfort and the blood gases. The patients were monitored using an oximeter together with routine nursing observations. The machine has a high and low pressure alarm which would alert the nursing staff if ventilation became inefficient.

In addition to the cases described here, we attempted to use NIPPV on another male patient who was unable to co-operate despite detailed instruction. He felt claustrophobic and his condition rapidly deteriorated and he died before donor organs became available.

Results

Patients 1–4 and patient 6 would not have survived without assisted ventilation until donor organs became available. Using the Brompton Pneupac ventilator their condition was stable at the time of surgery and we had no episodes of hypotension or toxaemia. The first three patients and patient 6 made excellent postoperative progress. No organs suitable for heart-lung transplantation become available for patient 4, so after 36 days on the ventilator he was given bilateral single lung transplants. Sadly he died one week later. Patient 5 died before suitable donor organs became available. All patients were cared for in the peaceful atmosphere of a single room on a general medical ward. They could talk and move their limbs while on the ventilator. Patients learnt to disconnect their ventilator and use oxygen *via* nasal cannulae for short periods of time so that they were able to eat and drink. They did not require sedation to tolerate the ventilator.

Discussion

The use of a conventional ventilator is not without risk in these patients heavily infected with *Pseudomonas aeruginosa* who have severe airflow obstruction. Previously some patients conventionally ventilated had episodes of hypotension and toxaemia which cause damage to other vital organs and make the patients unsuitable for heart-lung transplantation. Two other CF patients conventionally ventilated before transplantation needed many weeks in the Intensive Care Unit (ICU) after surgery [2] because of their inability to come off the ventilator and cough effectively. The patients treated using NIPPV showed none of these problems.

The use of NIPPV does not require the use of ICU facilities before surgery and is very cost effective. With the current financial restraints preoperative ventilation in ICU for one patient means that other patients are denied elective surgery or transplantation because of the lack of facilities. When a patient is treated by NIPPV he can eat, drink, talk and move his limbs freely and so nutrition and general physical condition are better maintained than when conventional ventilation is used. The use of NIPPV gives a useful but limited period of extra time for suitable donor organs to be found.

These patients were very sick. We chose, therefore, to use nasal ventilation rather than continuous positive airways pressure (CPAP) which has been shown in kyphoscoliotic patients to be less effective, particularly in patients with severe hypercapnia [6].

The shortage of donor organs sadly means that some patients with CF on the heart-lung transplant waiting list will die before transplantation is possible. When NIPPV is used they can stay in their own room on the ward where they know the nurses. They can communicate freely with relatives and friends. If the episode of deterioration turns out to be their final illness their dying is in many ways more dignified than if they died in the middle of a busy ICU. NIPPV techniques of mechanical ventilation avoid endotracheal intubation and this possibly reduces the risk of infection and ischaemia of the airway after transplantation. A patient trained in the use of this machine can use the technique after surgery and so avoid the need for reintubation in the early postoperative period. However, NIPPV has the disadvantages that the airway is not protected and that it provides no access for tracheal suction.

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 a 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 heart-lung transplantation can be performed is a cost effective and may be a life-saving procedure.

Acknowledgements: The authors would like to thank Dr M.A. Branthwaite who developed this machine for her help and advice and they also thank the many senior registrars, senior house officers, physiotherapists and nursing staff whose skills and hard work gave these young patients a chance to survive.

References

1. Scott J, Higenbottam T, Hutter J, Hodson M, Stewart S, Penketh A, Wallwork J. – Heart lung transplantation for cystic fibrosis. *Lancet*, 1988, 192–194.
2. Yacoub MH, Banner NR, Khaghani A, Fitzgerald M, Madden B, Tsang V, Radley-Smith R, Hodson M. – Combined heart and lung transplantation for cystic fibrosis and subsequent "Domino" cardiac transplantation. *J Heart Transplant*, 1990, 9, 459–467.
3. Ellis ER, Bye PTB, Bruderer JW, Sullivan CE. – Treatment of respiratory failure during sleep in patients with neuromuscular disease. *Am Rev Respir Dis*, 1987, 135, 148–152.
4. Carroll M, Branthwaite MA. – Control of nocturnal hypoventilation by nasal intermittent positive pressure ventilation. *Thorax*, 1988, 43, 349–353.
5. Elliott MW, Steven MH, Phillips GD, Branthwaite MA. – Non-invasive mechanical ventilation for acute respiratory failure. *Br Med J*, 1990, 2, 358–360.
6. Ellis ER, Grunstein RR, Chan S, Bye PT, Sullivan CE. – Non-invasive ventilatory support during sleep improves respiratory failure in kyphoscoliosis. *Chest*, 1988, 94, 811–815.

Ventilation mécanique non invasive dans la fibrose kystique - Un pont potentiel vers la transplantation. M.E. Hodson, B.P. Madden, M.H. Steven, V.T. Tsang, M.H. Yacoub.

RÉSUMÉ: Il s'agit d'une revue de l'histoire clinique de 6 patients atteints de fibrose kystique, en attente d'une transplantation cardio-pulmonaire. Tous ont évolué vers une hypoxie sévère avec hypercapnie avant que les organes du donneur ne deviennent disponibles. La ventilation au moyen de pression positive intermittente nasale a été utilisée de préférence à la ventilation conventionnelle, avec d'excellents résultats chez 4 patients. L'on n'a pas noté d'épisode d'hypotension ou de toxémie, et les patients furent en état stable au moment de la chirurgie et ont eu des suites post-opératoires excellentes. Les patients transplantés et le patient décédé pour lequel les organes adéquats du donneur ne sont pas devenus disponibles en temps utile, ont eu une survie plus confortable que cela n'eut été le cas s'ils avaient été traités par ventilation conventionnelle. Cette méthode de ventilation apparaît donc comme un pont utile vers la transplantation en cas de détérioration soudaine du patient. Elle lui donne une chance de survie pour quelques jours ou même semaines supplémentaires, pendant lesquels on peut s'attacher à une recherche urgente d'organes de donneurs. Il s'agit en outre d'une méthode de ventilation dont le rapport coût-efficacité est excellent, et qui n'empiète pas sur les possibilités conventionnelles de soins intensifs.

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