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The effects of one year of nocturnal cuirass-assisted ventilation in chest wall disease

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ABSTRACT: The effects of one year of nocturnal cuirass-assisted ventilation using individually designed cuirass respirators have been investigated in twenty-five patients with chest wall disease. After one year, 22 (88%) of the patients were alive. Daytime arterial blood gases had improved. Functional residual capacity (FRC) had increased but there was no significant change in other lung volumes. Maximum inspiratory pressure (MIP) improved in the subjects with a scoliosis but not in those with a thoracoplasty or neuromuscular disease. Maximum expiratory pressure (MEP) was unchanged. Maximum voluntary ventilation (MVV), the ventilatory response to carbon dioxide and six minute walking distance had all increased. There was no improvement in respiratory symptoms, but a decrease in depression scores and in the time taken to complete a trail test. The mean (SD) number of days spent in hospital over the year was 21.5 (15.1) per patient, with patients consulting their general practitioners less frequently than in the year prior to commencing nocturnal cuirass-assisted ventilation. The cost of commencing a patient on domiciliary nocturnal cuirass-assisted ventilation is estimated as £2470, and of maintaining them at home for one year as £3302.

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Nocturnal hypoventilation most commonly occurs as a result of upper airway obstruction, and correction of this requires tracheostomy or the application of positive airway pressure applied via a nasal mask. It is also seen in association with abnormalities of the respiratory centres, and in some patients with chest wall and pulmonary disease [13]. Assisted ventilation can be used to correct nocturnal hypoventilation in these patients. Cuirass respirators are one of the simplest and cheapest methods for this purpose [18], and have been shown to improve nocturnal gas exchange in patients with neuromuscular and skeletal chest wall disease [6, 12, 27]. Longer term use of cuirass-assisted ventilation at night leads to improvement in daytime respiratory failure [8, 11, 20, 23, 28]. Respiratory muscle fatigue may contribute to hypoventilation in such patients [26], and nocturnal rest of respiratory muscles during cuirass-assisted ventilation [25] may explain some of the improvement in daytime gas exchange. We have studied the results of one year of nocturnal cuirass-assisted ventilation at home in patients with chest wall disease.

Subjects and methods

Twenty-five patients with chest wall disease were commenced on domiciliary cuirass-assisted ventilation between 1983 and 1985. Upper airway obstruction during sleep was excluded in all patients by recording ribcage and abdominal motion using respiratory inductance plethysmography (Respitrace) and oronasal airflow using thermistors (Ormed). Twelve of the patients were female. Nine patients had a thoracic scoliosis which was congenital in seven patients and followed poliomyelitis in two. One patient had a thoracic kyphosis as a result of spinal tuberculosis. In table 1 the results from this kyphotic patient are included in the scoliosis group. Eight patients had a thoracoplasty which had been performed in the 1950's for pulmonary tuberculosis. In addition to the two patients with poliomyelitis, seven patients had a neuromuscular disease affecting the respiratory muscles, this being muscular dystrophy in four and an unclassified myopathy in three.

Arterial blood was sampled from the radial artery in all patients whilst breathing air and analysed in a Radiometer blood gas analyser. Vital capacity (VC) was measured with a Vitalograph wedge spirometer in twenty patients and functional residual capacity (FRC) by helium dilution in seventeen patients. Residual volume (RV) and total lung capacity (TLC) were calculated from FRC, expiratory reserve volume and inspiratory capacity. Maximum mouth pressures were measured using a pressure transducer (Si Plan Electronics Research Ltd) in conjunction with an occluded mouthpiece with a standard 22 gauge leak. Maximum inspiratory pressure (MIP) was measured at RV and maximum expiratory pressure (MEP) at TLC. Satisfactory recordings of MIP were taken from twenty patients and of MEP from nineteen patients.

Table 1. - Arterial blood gas, lung volume, maximum mouth pressure and 6 minute walking distance before and after a year of nocturnal cuirass-assisted ventilation in patients with chest wall disease (†=p<0.05).

	Scoliosis		Thoracoplasty		Neuromuscular		Total	
	pre	post	pre	post	pre	post	pre	post
$Paco_2$ kPa	8.38 (1.19)	6.23† (0.89)	6.83 (1.22)	6.06† (0.65)	7.92 (2.10)	6.02† (1.00)	7.76 (1.58)	6.12† (0.82)
Pao_2 kPa	6.98 (1.44)	8.68† (1.76)	7.80 (2.30)	8.80 (1.80)	9.65 (2.10)	10.0 (1.48)	7.97 (2.13)	9.08† (1.72)
VC *l*	0.75 (0.31)	0.89 (0.24)	1.36 (0.69)	1.41 (0.54)	1.22 (0.92)	1.02 (0.57)	1.08 (0.67)	1.10 (0.49)
TLC *l*	1.38 (0.22)	1.59 (0.34)	3.16 (1.52)	3.22 (1.26)	2.40 (1.22)	2.28 (0.55)	2.31 (1.31)	2.36 (1.06)
FRC *l*	0.78 (0.14)	1.06 (0.14)	1.83 (0.79)	2.09 (0.79)	1.49 (0.51)	1.56 (0.37)	1.33 (0.67)	1.54† (0.64)
RV *l*	0.68 (0.20)	0.81 (0.22)	1.67 (0.89)	1.72 (0.66)	1.32 (0.74)	1.27 (0.49)	1.72 (0.77)	1.26 (0.61)
MIP cmH_2O	33.1 (14.4)	48.1† (12.5)	48.1 (18.3)	52.3 (15.7)	43.0 (16.0)	35.0 (11.6)	41.4 (16.8)	45.8 (14.6)
MEP cmH_2O	49.3 (11.4)	54.5 (23.4)	69.1 (29.4)	69.9 (28.5)	43.7 (14.1)	39.8 (18.1)	54.8 (22.6)	55.5 (26.0)
MVV $l \cdot min^{-1}$	28.7 (18.5)	29.8 (16.4)	22.3 (7.6)	28.6 (9.2)	22.0 (10.5)	38.7 (17.0)	25.2 (13.8)	31.2† (14.0)
6 minute walk distance m	291 (140)	359 (124)	193 (146)	283 (110)	109 (130)	90 (93)	232 (150)	294† (144)

Mean ± (SD)

Maximum voluntary ventilation (MVV) was measured over 15 s in fifteen patients. The volume of expired gas was measured and the results expressed in litres per minute. In thirteen ambulant patients, a six minute walking distance was recorded. In eight patients, the ventilatory response to carbon dioxide was assessed by hyperoxic rebreathing from a bag containing 7 *l* of oxygen. The volume of each breath was measured by integration of the flow signal from a Fleisch pneumotachograph head connected to a Validyne differential pressure transducer. Instantaneous minute volume (IMV) was computed from total breath time and inspired volume. The concentration of carbon dioxide was measured by a Centronics mass spectrometer and IMV was plotted against the end-tidal carbon dioxide tension recorded from the expiratory phase of the same breath.

Individual cuirass respirators were made for each patient [5]. They were used in conjunction with either a Cape cuirass or Newmarket pump (Si Plan Electronics Research Ltd) [17]. The respiratory rate was set at that which the patient found most comfortable, and the peak negative pressure was set at the lowest (most negative) that the patient could tolerate. For all the patients this was in the range −20 to −40 cm of water. Patients used their cuirass respirator for 6–10 h each night at home, and in addition two thoracoplasty patients used their cuirass for one hour in the afternoon. Patients were assessed before starting cuirass-assisted ventilation and after one year. In the intervening period, patients were admitted routinely every two months for a sleep study, and were also admitted during acute respiratory infections or if adjustments to their cuirass were required.

In twelve patients, breathlessness, tiredness and mood were measured by a 10 cm visual analogue scale. Depression was assessed by completion of a Hamilton questionnaire [14]. These patients also performed simple mental tests of copying, deletion of numbers from a matrix, decoding of letter-number pairs, and a trail test, which involved joining in sequence numbers scattered randomly on a page. The number of days spent in hospital over the year were

obtained from the hospital records, and the cost of home and hospital care estimated from health authority statistics. The general practitioners of fifteen patients were contacted and replies obtained from thirteen. The number of home and surgery consultations and the number of antibiotic prescriptions were calculated from this data, for the year before commencing assisted ventilation at home and for the year of the study.

The cost of hospital care was based on the mean number of days spent in hospital over the year for all twenty-five patients using cuirass-assisted ventilation at home. The depreciation of the assisted ventilation unit buildings and monitoring equipment in a year was shared between all patients admitted to the unit over the year, assuming a life of 50 yr for the assisted ventilation unit and 5 yr for the monitoring equipment. The cost of home care was estimated from the survey of the general practitioners of thirteen patients.

Results

The arterial blood gas, maximum mouth pressure, lung volume and 6 minute walk distance results are given in table 1. The mean (SD) slope of the ventilatory response to carbon dioxide for the eight patients, assessed before and after a year of cuirass-assisted ventilation, increased from 0.47 (1.00) $l \cdot min^{-1} \cdot kPa^{-1}$ to 1.74 (1.94) $l \cdot min^{-1} \cdot kPa^{-1}$.

The symptom and mental function test results are given in table 2. The mean (SD) number of days each patient spent in hospital during the year was 21.5 (15.1). For the thirteen patients for whom details of general practitioner consultations were obtained, the mean (SD) number of consultations per patient was 11.8 (7.3) in the year before and 5.3 (5.0) in the year after commencing cuirass-assisted ventilation at home ($p < 0.05$). For the same group of patients, the mean (SD) number of antibiotic prescriptions per patient was 1.8 (1.4) for the year before and 1.0 (1.1) for the year after commencing cuirass-assisted ventilation at home ($p < 0.05$).

One patient with a myopathy died in hospital during an acute pneumonia. One patient with scoliosis and one with a thoracoplasty died at night, at home, within one month of initial discharge from hospital with a cuirass respirator. Both these patients lived alone, but two other patients living alone have managed nocturnal cuirass-assisted ventilation at home for over 2 yr.

An estimate of the capital and recurrent cost of home cuirass-assisted ventilation is given in table 3. Four patients were using home oxygen prior to commencing home cuirass-assisted ventilation, and this was discontinued in all four.

Table 2. - Mean (SD) visual analogue symptom scores, Hamilton depression scores and time to complete mental function before and after a year of cuirass-assisted ventilation (†=$p<0.05$).

Symptom:	Before	After
Breathlessness (0-100)	24 (25)	29 (16)
Tiredness (0-100)	26 (23)	41 (26)†
Happiness (0-100)	85 (15)	85 (21)
Depression Score (0-62)	5.3 (3.7)	2.4 (2.0)
Test:		
Trail seconds	45 (16)	35 (8)†
Deletion	34 (10)	31 (12)
Copying	38 (9)	34 (8)
Decoding	69 (21)	57 (17)

Discussion

We were unable to include a control group in this study. The patients studied were referred by physicians in other hospitals for treatment, often following an episode of life-threatening respiratory failure. We did not feel justified in withholding assisted ventilation from some of these patients to provide controls,

Table 3. - The cost of nocturnal cuirass-assisted ventilation per patient per annum.

		£
Capital costs		
Cuirass		50
Pump		2400
Home alterations		20
	TOTAL	2470
Recurrent costs		
Hospital admissions		
Assisted ventilation unit depreciation		40
Monitoring equipment depreciation		24
Nursing staff		1604
Medical staff		206
Physiotherapy		201
Technical staff		32
General services and equipment		948
Cuirass repairs		5
Transport		65
Home		
Medical visits		26
Nursing visits		52
Social services		46
Other attenders		18
Electricity for pump		35
	TOTAL (per annum)	3302

whilst less severely affected patients not requiring assisted ventilation would not form a comparable group.

The improvement in daytime arterial blood gases with the use of nocturnal assisted ventilation seen in this study has been reported previously [8, 20, 21, 28]. A lower daytime arterial carbon dioxide tension ($Paco_2$) after nocturnal assisted ventilation, could reflect an increase in the output of the respiratory centres, or improved ventilation without any change in respiratory drive. The latter could result from improved respiratory muscle function, an increase in total respiratory system compliance or a decrease in resistance. An increase in ventilatory response to carbon dioxide accompanying the fall in $Paco_2$ after nocturnal assisted ventilation has also been described previously [2, 10, 24]. This may reflect recovery of respiratory centre sensitivity to carbon dioxide, possibly due to a decrease in cerebrospinal fluid bicarbonate buffering capacity, or a greater rise in cerebrospinal fluid hydrogen ion concentration occurring for the same change in $Paco_2$ [4]. The increase in arterial oxygen tension (Pao_2) could be explained by the rise in FRC, with improved ventilation-perfusion matching in dependent areas of the lung. Although others have reported an increase in VC after cuirass-assisted ventilation [3, 11], no change in VC, TLC or RV was detected in our patients.

Mean VC, RV and TLC decreased in the neuromuscular patients and increased in the other patients. A similar pattern was seen in MIP and MEP. This probably reflects progression of the underlying disease which was clinically apparent in four of the six neuromuscular patients. Formal assessment of non-respiratory muscles was not undertaken. The distance walked in 6 minutes decreased in both of the ambulant neuromuscular patients but in only three of the eleven ambulant scoliotic or thoracoplasty patients after a year of nocturnal cuirass-assisted ventilation.

Only the scoliotic patients showed a significant increase in MIP. The change in MIP did not correlate with the change in either RV or Pao_2, and none of the patients were hypokalaemic or receiving theophyllines or digoxin. It is unlikely that a learning effect would be seen in only the scoliotic patients. Inspiratory muscle activity decreases during cuirass-assisted ventilation [25], and nocturnal rest could explain the increase in daytime function. The relevance of this is unclear, since daytime $Paco_2$ also decreased in the neuromuscular patients despite a decrease in MIP. Increases in MIP after assisted ventilation in restrictive chest wall disease have been documented previously [3, 21], along with an increase in MVV as seen in our patients.

Cuirass-assisted ventilation improves nocturnal gas exchange, and this may increase the amount of time spent in rapid eye movement sleep [6]. Correction of sleep pattern, daytime and arterial blood gases improves mental alertness, and this could contribute some of the physiological changes described above [7, 9]. Patients were more tired after a year of nocturnal cuirass-assisted ventilation, and of the mental function tests only the trail test time improved, which may be a learning effect. The decrease in depression scores may reflect the interest taken in the patient over the year. Both learning and increased motivation may have contributed to the changes in some of the effort related tests.

The one year survival rate of 88% for this study is comparable to that described for patients with chest wall disease using intermittent positive pressure ventilation [1, 18]. Four patients required nocturnal assisted ventilation by a tank ventilator or intermittent positive pressure ventilation prior to commencing cuirass-assisted ventilation and were confined to hospital. For these patients at least, the time spent in hospital was reduced by nocturnal cuirass-assisted ventilation. The majority of the other patients had spent little time in hospital before the acute episode which precipitated their referral to the Assisted Ventilation Unit. The reduction in general practitioner prescriptions and attendances may reflect a decrease in the number of episodes of acute respiratory failure, but must at least partly be due to closer hospital surveillance of the patients. Changes in the quality of a patient's life cannot be measured in cash terms. All patients were able to leave their homes during daytime, and three patients were able to travel away from home for holidays during the course of the year.

Conclusions

Nocturnal cuirass-assisted ventilation improves daytime gas exchange in patients with chest wall disease. This is associated with an increase in FRC, in MVV and in the ventilatory response to carbon dioxide. Only scoliotic subjects show an increase in MIP. Domiciliary cuirass-assisted ventilation should be considered, along with nasal intermittent positive pressure ventilation [16, 19] and external negative pressure ventilation using tank or body respirators [15], as a means of providing long-term nocturnal assisted ventilation for these patients. Cuirass-assisted ventilation requires relatively little surveillance and can improve the quality of a patient's life.

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RÉSUMÉ: Les effets de la ventilation nocturne assistée pendant un an, ntilisant des cuirasses ajustées individueflement, ont été investigués chez 25 sujets avec maladie de la cage thoracique. A un an, 22 patients (88%) sont vivants. Les gaz du sang diurnes sont améliorés; la capacité résiduelle fonctiounelle (FRC) est augmentée mais les autres volumes sont inchangés. Les pressions inspiratoires maximales (MIP) augmentent chez les scoliotiques mais pas chez les malades avec thoracoplastie ou maladie neuromusculaire. Les pressions expiratoires maximales (MEP) sont inchangées. La ventilation maximum volontaire (MVV), la réponse ventilatoire au CO_2 et la distance parcourue en 6 min ont toutes augmenté. Il n'y a pas d'amélioration des symptômes respiratoires, mais bien une diminution des scores de dépression et du temps nécessaire pour compléter un test de trace. La durée moyenne de séjour hospitalier a été de 21.5 (SD: 15.1) jours, et le numbre de consultations chez le médecin généraliste a diminué par rapport à l'année précédente. Le coût de la mise en route d'une ventilation à domicile par cuirasse est estimé à £2470 et le coût de la maintenance à domicile pendant un an est estimé à £3302.