Domiciliary nasal respiratory support  
- first experiences in Malta

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ABSTRACT: Nasal respiratory support is a non-invasive alternative to conventional assisted ventilation with endotracheal intubation, or the more cumbersome negative pressure ventilators. The two main types of this relatively new therapy are nasal intermittent positive pressure ventilation [NIPPV] and nasal continuous positive airway pressure [NCPAP] respiratory support, which are mostly used in chronic hypoventilatory states and obstructive sleep apnoea [OSA] respectively. We have introduced these two types of respiratory support to five patients suffering from neuromuscular disorders and twenty-four patients with OSA with marked improvement in the quality of life of all patients concerned. Our experiences with these patients should hopefully lead to further development in the diagnostic and therapeutic facilities in this field in Malta.

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Introduction

Until recently, patients with ventilatory problems needing long-term artificial assisted ventilation, initially required endotracheal intubation and later a permanent tracheostomy or the cumbersome 'cuirass'. Since the advent of nasal respiratory support, physicians can treat these patients without having to resort to such invasive measures, thereby improving the patient's quality of life markedly.

Nasal intermittent positive pressure ventilation [NIPPV] and nasal continuous positive airway pressure [NCPAP] are the two most widely used modes of this type of non-invasive respiratory support, and when used in the appropriate cases, can transform the patient's lifestyle in the short term1,2 and improve the prognosis of his ventilatory condition together with its secondary effects, in the long term3,4.

Nasal respiratory support has been used in Malta in the last two years and we have succeeded in starting a good number of patients on this type of treatment from which they are still benefiting today.

The start of nasal respiratory support in Malta

Up to two years ago the only Maltese patient utilising domiciliary NIPPV was a young girl suffering from Ondine's curse secondary to damage of her respiratory centre by severe pertussis infection. She had initially been treated in an 'iron lung' in the intensive care unit at St. Luke's Hospital, after unsuccessful attempts at weaning her off invasive ventilation. Following transfer to the UK, she was started on an NIPPV ventilator which she subsequently used every night because of nocturnal hypoventilation. She forms part of our series of patients described below. In November 1994 a fourteen year old male suffering from Duchenne muscular dystrophy, who was admitted to hospital 'in extremis', was the first patient to be started on NIPPV at St. Luke's Hospital. He had been complaining of morning headaches for the weeks preceding admission and his parents had noted that he was becoming more drowsy and cyanosed as time went by. Ultimately he lapsed into unconsciousness on the day of admission when his arterial blood gases revealed a PaCO₂ of 94.2mm Hg and a PaO₂ of 65mm Hg. Within two hours of commencing NIPPV he had regained consciousness and his arterial blood gases had improved markedly [PaCO₂ 62mm Hg, PaO₂ 82mm Hg]. This young man is still alive today and is using NIPPV nightly to compensate for nocturnal hypoventilation and daytime hypoxia and hypercapnia. The success of these two cases prompted the application of nasal respiratory support for other such patients and also for other indications.

Nasal intermittent positive pressure ventilation [NIPPV]

Nocturnal hypoventilation is the major indication for this type of ventilation in which the airway pressure changes phasically throughout the cycle, delivering the entire tidal volume and then allowing passive exhalation. NIPPV is thought to help these patients by resting fatigued muscle5, increasing lung compliance6, and restoring some of the blunted CO₂ sensitivity of the respiratory centre7. Like other types of nasal respiratory support, it is administered through a small ventilator/blower which may either be volume or pressure-dependent. The positive pressure is instituted through a tight-fitting nasal mask which forms a seal by being held to the face by an appropriate type of head-gear8 (See Fig 1). This assembly transmits the pressure or volume generated by the ventilator to the airways. The development of silicone masks have made these much
Fig. 1 - A patient with the nasal mask and head gear used in nasal support

more comfortable while creating better seals. Improvements are being made continuously such as full face masks for patients who have mouth-leaks and small nasal cushion masks for those who find the bigger masks claustrophobic.

Nasal continuous positive airway pressure [NCPAP]

With NCPAP positive airway pressure is maintained, relative to atmospheric pressure, throughout the cycle. NCPAP was introduced by Dr. Colin Sullivan in Australia as a means of splinting the upper airways during sleep in patients with obstructive sleep apnoea [OSA]. Nasal bi-level positive airway pressure [NBIPAP] is a modification of NCPAP in that the positive pressure is decreased slightly during the expiratory phase of the cycle so as to make the whole process more comfortable for the patient thus, presumably, increasing the compliance rate.

The commonest symptoms of obstructive sleep apnoea are excessive daytime somnolence and irregular pattern of snoring, which latter, in the majority of cases, is due to repeated collapse of the oropharyngeal musculature during sleep. This results in complete or partial obstruction of the subject's airway accompanied by a reduction in blood oxygen saturation and followed by waking to breathe. These repeated episodes lead to sleep fragmentation with the deeper stages of sleep [rapid eye movement stages] being the most affected.

It should be distinguished from the much less common central sleep apnoea which is a neurological disturbance resulting in a cessation of respiratory effort in sleep. OSA has come to the medical forefront in the last few years as the commonest type of sleep-related breathing disorder. Many sufferers are unaware of the cause of their excessive daytime sleepiness and pathological tendency to fall asleep, sometimes even while driving their car, leading to fatal or near-fatal accidents. Their sleeping partner complains about their loud snoring interrupted by long breathing pauses and excessive body-movements. This often leads to marital disharmony, mental depression, lack of libido and impotence. OSA adversely affects the quality of life of the subject and can decrease the life expectancy of a 50 year old sufferer by 50%. The effects of OSA on the cardiovascular system include life-threatening arrhythmias, systemic hypertension and myocardial ischaemia, while the recurrent hypoxic episodes can also lead to pulmonary vascular disorders.

The diagnosis is made from a suggestive history and a sleep-study (polysomnogram) in order to differentiate OSA from other sleep-related disorders and simple snoring. During polysomnography a number of the following parameters are monitored: electroencephalogram (EEG), electrocardiogram (ECG), ocular and genohyoid electromyogram (EMG), respiratory effort and air flow, oxygen saturation, apnoea/hypopnoea time and frequency, body position, limb activity and snoring. An important analysis is the apnoea index which is the number of breathing pauses lasting 10 seconds or longer per hour of sleep. An apnoea index of more than 5 is considered pathological by some investigators but others feel that 10 - 15/hr is a more specific measure for diagnosis. Oxygen desaturations of >4% of baseline during hypopnoeas (a reduction in airflow associated with a fall in oxygen saturation and an arousal from sleep) also points towards a diagnosis of OSA. Difficulties in defining the diagnosis may explain the differences in prevalence rates between studies but the range is between 0.5 - 5% of the population.

This paper describes how we screened, diagnosed and treated the first Maltese patients with OSA and nocturnal hypoventilation using nasal ventilation. The future of this modality of treatment in Malta is then discussed.

Method

1) NIPPV

Subjects

Since 1994, four patients suffering from neuromuscular conditions with secondary nocturnal hypoventilation were started on NIPPV. Three presented in acute Type II respiratory failure. A young girl was started on NIPPV for Ondine's curse following severe pertussis affecting her respiratory centre, and continued to be followed up. (Table 1)

Ventilators

A bulky Bromptonpac ventilator (Pneumopac Ltd., Luton) was initially used on two of these patients and later changed to a pressure-dependent NIPPV ventilator Thomas Respiratory Systems UK) weighing around 5 kgs. Two of the other patients are also on such a ventilator and all these four patients have had their inspiratory pressures set at around 10 cms H2O.

Table 1 - Patients on NIPPV

Subject | Sex | Age (yrs) | Condition leading to res. failure
------- |-----|----------|-------------------------------------
1       | F   | 16       | Central Res. Hypoventilation        
2       | M   | 14       | Duchenne Muscular Dystrophy         
3       | F   | 42       | Limb-Girdle Muscular Dystrophy      
4       | F   | 37       | Limb-Girdle Muscular Dystrophy      
5       | M   | 17       | Duchenne Muscular Dystrophy         

2) NCPAP

Subjects

Twenty-four patients (21 male and 3 female) with a mean age +/- SEM 43.4 +/- 2.9 yrs (range 19 - 66 yrs) all suffering from obstructive sleep apnoea were treated on NCPAP. All had a neck size of more than 17 inches in circumference (range 17 - 19.5 inches). Most were above the ideal weight for their height except two, who had an element of micrognatia. They all came from different walks of life but the majority (14 out of 24) were of professional or executive status.

Diagnosis of obstructive sleep apnoea

These twenty-four patients were diagnosed using a combination of history, clinical examination, blood investigations and a partial sleep study.

History

The help of the patient's sleeping partner was sought in taking a history where we specifically enquired about cardinal features such as excessive snoring with apnoeic spells, nocturnal choking sensation, nocturia, reduced libido, morning headaches, increased limb movement in sleep, lack of daytime concentration and general well-being. A modification of the Epworth sleepiness scale was used to try and grade the patients' daytime sleepiness in specific situations such as when driving, reading, watching TV or talking to someone. Seventeen of the patients were asked to put a score of 1 to 10 on a number of these symptoms before and after starting to use NCPAP so that we could grade any improvement the patient might experience. Enquiry was also made about alcohol or sedative ingestion, smoking, sleep hygiene, shift work, attempted weight reduction, nasal blockage and symptoms suggestive of hypothyroidism.

Clinical examination

During the clinical examination emphasis was put on nasal patency, the state of their soft palate and uvula, presence of micrognatia, neck size, blood pressure and any clinical signs suggestive of hypothyroidism and acromegaly.

Blood investigations

Apart from the routine blood tests the patients were screened for hypothyroidism and acromegaly in cases where these conditions were suspected.

Partial sleep studies

These studies were carried out at the coronary care unit at St. Luke's Hospital, when a monitor bed was available for the night in question. Using these monitors, continuous percutaneous oxygen saturation, ECG, blood pressure, respiratory rate and apnoea frequency were studied. The nurses on night duty and one of the investigators (SM) observed the patient from time to time during the night to try and observe the snoring pattern, apnoeas, limb movements and body position during these events. In all we screened 38 patients, 24 of which met most of the criteria we could monitor for OSA. When possible or in cases where the initial study revealed heart rhythm problems, we carried out another study during the first night on the machine.

NCPAP machines

In order to try and maximise the compliance rate we opted to use NBIPAP on most of the patients so as to make this treatment as comfortable as possible. In fact 11 of our patients are on Respironics BIPAPS machines with the pressures used ranging between 10 - 18 cms H2O for the inspiratory positive airway pressure and 5 - 14 cms H2O for the expiratory positive airway pressure. The machine was set on spontaneous mode so that the patient would trigger off the machine and not encounter undue resistance on expiration. Once adequate experience had been gained, it was decided to start the next patients on NCPAP (a mix of Respironics Remstar, DeVilbiss, Healthdyne and ResMed machines), which proved to be sufficient for the rest of the patients and considerably less expensive than the NBIPAP machines.

Statistical analysis

The symptom scores before and after NCPAP were compared using the Wilcoxon paired rank test. A p value of <0.05 was taken to represent a significant association or difference.

Results

NIPPV

All five patients have noted a very marked subjective improvement in their quality of life since initiation of treatment and this was confirmed by their relatives. One of the young men is back attending school while another is now much more responsive and in good spirits than before treatment. The latter initially had some difficulty with increased oral secretions on using the ventilator but this has decreased since being started on inhaled Ipratropium Bromide 2 inhalations nocte through a spacer device. He is now also having some swallowing difficulties which are probably secondary to his muscular dystrophy. In the past, these patients would have succumbed to chest problems before encountering these difficulties. Similar improvement was noted in the other two female patients, both of which have returned to their jobs. All these patients are very compliant with treatment and are on the ventilator for at least six hours every night. Some even use it when experiencing problems in breathing during the day. At first some, especially the younger ones, felt claustrophobic with the nasal mask, but soon, all got used to it.

The girl with the Ondine's curse was recently sent to the UK for a full sleep study and told that she could now stop using the NIPPV on a regular basis.

NCPAP

Of the 38 patients screened, 24 had enough evidence for a diagnosis of OSA. None of the patients screened
had any endocrine or major nasal problem to explain their complaints, while those who imbibed alcohol with their evening meal (four) or used sleeping-pills (three) stopped doing so. These measures, however, did not improve their sleep-related breathing disorder. All the overweight subjects had tried to lose weight but only two managed to lose a substantial amount and even these did not improve enough to render treatment with nasal CPAP unnecessary. There was no great difference in the sleepiness scale between the patients who ended up on nasal respiratory support and those who did not. However there was a marked difference before and after starting nasal ventilation in how sleepy the patient felt during the day (mean score before vs mean score after) (7.8 vs 1.6) and during driving (6.9 vs 0) (Fig 2). This improvement was also noted for nocturnal choking sensation (4.4 vs 0), concentration (5.1 vs 8.2) and general well-being (4.2 vs 8.8). The snoring in the treated group was noted to be as loud as in the group who were subsequently labelled as simple snorers. However, the snoring was more irregular, reached a crescendo more often and was interspersed with apnoeic spells. Much to the relief and appreciation of the patients’ sleeping partners the patients on nasal NCPAP stopped snoring almost completely (9.3 vs 0.5). All these changes reached statistical significance (p<0.05). These symptom scores were not carried out by our seven most recent patients as it was too early to grade any improvement in concentration and well-being but they too improved in the rest of the symptoms.

weeks time to see whether the arrhythmogenicity of their myocardium will have decreased following the use of NCPAP for this period of time.

The patients on whom NCPAP or NBIPAP was instituted, were all admitted to hospital for their first night on the ventilator. All but two accepted the nasal mask very well. One of these two patients who continued to find difficulties was given a nasal bubble-cushion mask (Monarch mask; Respironics, USA) and has adapted to this very well. The other patient has decided to postpone regular use of NCPAP for some time. Once he decides to initiate treatment we plan to use the Monarch mask again so as to increase the chances of acceptance. The pressure settings of the machine were adjusted according to the build of the patient as we did not have the facility to titrate airway pressures in our partial sleep studies. During this first night we decreased the pressures gradually till the patient either started snoring or breathing irregularly and then increased again to just above the breakthrough pressure level. All the patients, without exception, commented favourably about how much better they felt on waking up after that first night and their subjective improvement continued to increase over the first few nights. Subsequent improvement decreased when patients defaulted treatment for as little as 2-3 nights. The most common complaints were airway dryness, nasal bridge soreness and facial marks from the mask, breathing warm air on hot nights and the aesthetics of the whole process.

Discussion

In this paper we have demonstrated the introduction of
a novel mode of treatment for patients with nocturnal hypoventilation secondary to neuromuscular conditions and for OSA sufferers in Malta. Nasal respiratory support has improved the quality of life in all these patients, indeed for three patients with neuromuscular disease, this proved to be life-saving. This improvement in quality of life is the short term result of the treatment. The long term benefits which have been shown to occur in major studies, will hopefully be experienced by our patients as well.

In the past, most patients suffering from muscular dystrophy, would die prematurely from respiratory problems - usually due to acute or chronic respiratory failure precipitated by a chest infection. Preceding this, these patients usually develop cor pulmonale from hypoventilation due to their muscular weakness and thoracic skeletal malformations. Now, these acute and chronic chest problems can all be helped by NIPPV as demonstrated in this series of patients. Similar patients presenting in acute respiratory failure usually present a difficult dilemma to the admitting physician as whether to intubate and ventilate, knowing that their chances of being weaned off are slim. Despite a poor prognosis, these patients would occupy a precious ITU bed for a long time. Nasal ventilation offers a useful alternative to full ventilatory support. Indeed, this was successful in the three acute situations encountered. In the chronic situation, all these patients felt better during their waking hours as their hypercapnic and hypoxic state was ameliorated.

NIPPV can also be used in other cases of chronic respiratory failure such as multiple sclerosis, central hypoventilation syndrome, motor neurone disease and chest wall deformities, but its use in chronic obstructive airway disease is still controversial. The current position is that NIPPV seems to benefit only those cases of COAD who have high daytime PaCO2 (>55mmHg) and may also be considered in acute exacerbations in patients who either refuse intubation or who would not otherwise be considered ideal for endotracheal intubation and ventilation.

NCPAP and NBIPAP, the other mode of nasal respiratory support utilised in this series of patients, also proved to be of major benefit to the patients involved. OSA is a debilitating condition with implications and negative effects on the patient's personal, social, professional and physical lifestyle. Again the long-term effects of this condition can also lead to an untimely death due to cardiac arrhythmias and ischaemia together with the complications encountered in chronic respiratory failure and systemic hypertension. The real life caricature of an overweight man who dozes off easily and snores loudly presents a serious medical problem. OSA is little known among lay people and doctors alike, and must be 'publicised' so as to be recognised and treated more often. The number of patients presenting to us is now increasing partly as a result of the positive feedback from successfully treated patients. Our patients have all experienced a great improvement in their quality of life and this, in turn, is improving their compliance to treatment.

The fact that accidents such as motor-vehicle collisions can affect third parties, makes the situation even more worrisome. The condition is much more common than one expects with 2 - 4% of the American population thought to suffer from various grades of OSA. The fact that the average Maltese person is rather overweight makes it likely that the prevalence of OSA in our country is at least just as high.

Great changes are being made in the field of development of NCPAP machines as these ventilators are now becoming more 'user-friendly', comfortable, lighter and thus more portable. Features such as ramping, which allows a slow build-up of pressure after sleep onset, demand CPAP and AutoCPAP in which the machine senses when the patient does not require CPAP and switches off, have become available in the last couple of years. All these are designed to increase patient compliance. Options such as drastic surgery as in uvulopalatopharyngoplasty (UVPP) have not been very successful, while newer laser surgery is still unproven. Other studies have looked at tracheostomies and oral appliances but these are not very popular. In children, where sleep apnoea is also encountered, surgery in the form of adenoidotonsillectomy tends to correct the majority of OSA cases. NCPAP is only required in children with craniofacial anomalies, trisomy 21 and skeletal dysplasia syndromes, who have not responded fully to surgery.

In our opinion, the setting up of a sleep laboratory is badly needed at our hospital as evidenced by the above results. The condition is common and treatable, but the current facilities are not sensitive or specific enough for us to be sure that we are detecting all cases of OSA amongst the patients we screen. At the present time we are only able to monitor few of the criteria required for a diagnosis and even these are not being sampled frequently enough. We are also currently unable to titrate NCPAP pressures and have to use a rather crude method to decide pressure settings for different patients. The institution of a sleep laboratory does not entail a lot of expense apart from the initial expenditure and the training of a sleep technician. If space is a problem there are new polysomnograms available on the market which the patient can take home with him for monitoring in his usual nocturnal surroundings. Another facility that would obviously be required is a sleep clinic where these patients would be seen, screened and followed up. Such a clinic would also be able to carry out research and try these modes of therapy for new indications such as acute left ventricular failure.

In conclusion we have reviewed the first series of patients which has been initiated on respiratory support for different conditions in Malta. We have discussed the positive impact of this treatment on these patients and their condition and demonstrated that this is a field which is worth pursuing. The development of domiciliary respiratory support can offer significant benefits to patients with respiratory insufficiency and should be pursued by clinicians and the Health Division alike.

References
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