Chronic respiratory insufficiency (CRI) is an inevitable complication in the disease progression of patients with Duchenne muscular dystrophy (DMD). Without mechanical ventilation (MV), morbidity and mortality are highly likely towards the end of the second decade of life. The present review reports evidence and clinical implications regarding DMD patients treated with MV. There is no doubt that nocturnal hypercapnia precedes daytime hypercapnia. Historical comparisons have provided evidence that non-invasive intermittent positive pressure ventilation (NIPPV) at night is effective and improves quality of life and survival by 5–10 years. By contrast, the optimal criteria and timing for initiation of NIPPV are inconsistent. A recent randomized study however demonstrated the benefits of commencing NIPPV as soon as nocturnal hypoventilation is detected (Ward S, et al., Randomised controlled trial of non-invasive ventilation (NIV) for nocturnal hypoventilation in neuromuscular and chest wall disease patients with daytime normocapnia. Thorax 2005; 60: 1019–24). The respective role of the three hypotheses of the indirect action of nocturnal NIPPV on daytime blood gases may be complimentary; the main improvement may be due to improved ventilatory response to CO2. The ultimate time to offer full time ventilation with the most advantageous interface is lacking in evidence. Full time NIV is possible with a combination of a nasal mask during the night and a mouthpiece during the day, however tracheostomy may be provided when mechanical techniques of cough-assistance are useless to treat chronic cough insufficiency. Chronic Respiratory Disease 2007; 4: 167–177

Key words: Duchenne; hypercapnia; neuromuscular diseases; noninvasive ventilation; respiratory failure

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specific studies with DMD patients, but non-specific studies in the neuromuscular (NM) population including DMD patients are used when specific studies are deficient. In addition, this review includes recommendations for further research when the matter is controversial or unexplored.

The need for a protocol to detect CRI

The need for systematic detection of the risks for CRI is evident. We know nocturnal hypoventilation precedes daytime hypoventilation for some years in disease progression of DMD patients (Figure 1). Hypoventilation during REM-sleep was first reported in 1975 in generalized NM patients and in 1984 in DMD patients. After 1987, breathing disorders during sleep were extensively documented. Interestingly, one study investigating daytime MV during wakefulness was able to reverse nocturnal hypoventilation with similar benefits as with ventilation during sleep. This implies that respiratory improvement obtained with daytime ventilation was not directly mediated by an effect on sleep quality. This observation suggests that sleep deterioration may trigger and complicate nocturnal hypoventilation rather than inducing hypoventilation itself. An obvious question is to determine the role of sleep quality monitoring (extensive polysomnography) in the protocol to detect hypercapnia in DMD patients.

Extensive polysomnography

Many studies have reported sleep statistics in DMD patients; alterations in sleep quality (arousals) and architecture (sleep fragmentation). In adolescent DMD patients, 60% of apnoeas are initially obstructive. With disease progression apnoeas are essentially central or pseudo-central (inspiratory effort too weak to be identified). The presence of macroglossia was reported to exacerbate sleep apnoeas. Unfortunately, no studies concluded the impact of sleep deterioration on the decision to initiate nocturnal non-invasive positive pressure ventilation (NIPPV) in DMD patients. The major reason is that sleep

![Diagram](https://example.com/diagram.png)

**Figure 1** This Figure demonstrates the impact of initiation time of non-invasive positive pressure ventilation (NIPPV) on survival of Duchenne muscular dystrophy patients. Ages are given as indicative values; FVC: forced vital capacity; CRI: chronic respiratory insufficiency; Prev. NIPPV: preventive NIPPV initiation (see text); early NIPPV: early NIPPV initiation (see text); late NIPPV: late NIPPV initiation (see text); REM: rapid eye movement; Black vertical arrow: point of time of NIPPV intervention.
apnoeas do not always lead to hypercapnia. As a consequence, there is no need to know that sleep is disrupted to initiate NIPPV. By contrast, there is a need to measure hypercapnia to initiate NIPPV. One must be aware that a full polysomnography is expensive and hospital admissions are considered a major organizational event for DMD patients.

Nocturnal monitoring

Carbon dioxide tension (PCO$_2$) measurement during the night should be considered every year once VC declines, and ultimately when daytime PCO$_2 \geq 45$ mmHg. Nocturnal measurement of PCO$_2$ should always be considered in symptomatic patients in whom CRI is highly likely, however clinically this does not always occur.

PCO$_2$ measurement: invasive or non-invasive technique?

There is no agreement regarding the technique to detect hypercapnia in NM patients. One concern is to know whether transcutaneous PCO$_2$ (TcCO$_2$) is sufficient compared to the arterial PCO$_2$ measurement (PaCO$_2$). PaCO$_2$ has the disadvantage to be measured in the morning during wakefulness therefore not reflecting PCO$_2$ during sleep. Despite slightly overestimating PaCO$_2$, TcCO$_2$ has an excellent agreement with PaCO$_2$ in stable patients with NIPPV ($r$: 0.97, $P < 0.001$) and in patients with critical illness. New technology using earlobe sensor provides an excellent agreement between TcCO$_2$ and PaCO$_2$ ($r$: 0.98, $P < 0.0001$). TcCO$_2$ has the advantage of being non-invasive, and easy to use. However, morning arterial or venous PCO$_2$, pH and base excess should be carried out to ensure accurate baseline TcCO$_2$. Overnight pulse oximetry can be effective but is considered as less sensitive in the earlier stages of DMD disease.

Detection of symptoms

CRI develops insidiously and symptoms of respiratory failure may not be pronounced, especially in patients with nocturnal hypercapnia with low mobility such as in wheelchair bound DMD patients. The first symptoms are related to a poor sleep quality with or without hypercapnia, such as somnolence which is the most common complaint, nightmares, morning headaches and poor concentration have also been reported. Pure respiratory-related symptoms appear very late usually at the time of diurnal hypercapnia such as weariness, dyspnoea, orthopnoea, chest oppression, hand sweating, swallowing disturbance, loss of appetite and moodiness. To date, no validated symptom score is available.

Detection of CRI by daytime measurements

Undoubtedly, lung function best assess the degree of respiratory muscle involvement in DMD patients. Indispensable lung function measurements include VC, the maximal inspiratory and expiratory pressures (MIP and MEP) at the mouth and the peak expiratory flow during coughing (PCF). Sniff nasal inspiratory pressure is reported as an accurate estimate of maximal inspiratory strength. However, it has been shown to underestimate inspiratory strength in the DMD population. It is recommended that VC in sitting and supine position, MIP and MEP, and PCF measurements should be recorded twice a year in non-ambulatory patients. In DMD patients, the age at which VC falls below 1 L is a best predictor of age of death than nocturnal oxygen saturation.

Prediction of hypercapnia by the vital capacity

Few studies have addressed the unique needs of DMD patients. Some authors failed to establish a relationship between VC and hypercapnia while others demonstrated the appropriateness of VC and MIP in predicting nocturnal hypercapnia. At a later stage of disease progression, VC and MIP accurately predicted daytime hypercapnia. The disability score added to VC% provides a better indication of the need for MV than VC% alone.

Prediction of hypercapnia by the breathing pattern

During weaning procedure, the use of the rapid and shallow breathing index (RSBI) and the tension time index (TTI) calculated by Ti/Ttot x Pi/MIP provides prediction of the ability for NM patients to self ventilate. However, additional investigations are needed to detect whether hypercapnia is related to those indexes in DMD patients. The TTI and the endurance tests at constant inspiratory load are promising indices to discriminate those DMD patients at risks for respiratory muscle fatigue approaching the need for full time-assisted ventilation.

Multidisciplinary team

There is a need for an integrated multidisciplinary team in the care of DMD patients. Such a team should include a key physician, particularly between
childhood and adult transition to assure continuity in the detection of CRI.64 There should be compassionate discussions with patients and family about respiratory progression and prognosis and about MV before CRI occurs;24,35 this should be with the most appropriate member of the multidisciplinary team.

The optimum timing for initiation of MV: four options

No ventilation

Assisted ventilation was seldom proposed to DMD patients5 before the 1980’s. The option of no ventilation is now universally judged as unethical in countries where the technology is available. However, in rare cases it may be ethical not to commence NIPPV where this is in accordance with the profound wishes of some well-informed patients.

Preventive use of NIPPV

The initial objective of preventive NIPPV is aimed at lowering the inevitable worsening of VC with disease progression in normocapnic and asymptomatic patients.1,3 Rideau et al.18 showed preventive NIPPV resulted in long-term stabilization of VC in 70% of patients. In 1994 a French multicenter randomized study conducted by Raphael et al.65 investigated the impact of preventive NIPPV on survival in a DMD population versus control without NIPPV. Survival rates were significantly lower in the NIPPV group. As a conclusion, prophylactic use of NIPPV in Duchenne patients was not recommended for DMD patients with VC ranging 20–50% predicted.65 Pure prophylactic introduction of NIPPV was thereafter rejected. However, this study has been criticized for the following reasons: The families in both groups were aware that if the individual in the control arm deteriorated they would receive NIPPV, and the NIPPV arm, should use their ventilator more liberally. This advice may have caused the control arm to seek medical assistance earlier than the NIPPV arm and this may have prevented deaths. In this study, there were no recommendations that the ventilator could be used to assist in secretion clearance. Secondly, cardiac function was significantly worse in the NIPPV arm with only 10 having normal echo versus 20 in the control group, potentially biasing the results. Thirdly, Raphael et al.65 did not measure nocturnal PCO₂ in patients with VC ranging 20–50% although DMD patients with VC < 40% are exposed at high risk of developing nocturnal hypercapnia.5,46,57 Consequently, Raphael et al.65 did not discriminate patients without nocturnal hypercapnia from those with nocturnal hypercapnia, although NIPPV should be considered as a treatment of nocturnal hypercapnia (see below). All these methodological shortcomings make interpretation of the results of this study65 confusing.

‘Early’ initiation of NIPPV with nocturnal hypercapnia alone

Early initiation of NIPPV is a growing approach (Table 1): NIPPV is here proposed in nocturnal hypercapnia alone, even when sleep-related symptoms are not present.22 At this stage of respiratory involvement, DMD patients do not show severe O₂ desaturation at night.42 Soudon et al.15 proposed a very early approach by starting MV as soon as REM sleep hypercapnia was present, considering early MV as a curative treatment of early CRI. These authors however, could not validate their approach. Other groups suggested that a ‘semi-early’ approach was beneficial since hypercapnia was present either during 50%21 or 100% sleep.22,55 Importantly, Ward et al.22 recently investigated the initiation of NIPPV with patients with nocturnal hypercapnia alone in a randomized controlled study; they demonstrated that these patients are likely to deteriorate with the development of daytime hypercapnia within two years. Initiation of nocturnal NIPPV before the development of daytime hypercapnia was beneficial to the patient. Benefits included increased quality of life, relief of the symptoms, reduction of daytime hypercapnia and reduced risk of admission in ICU for decompensated respiratory failure.22 Initiation of NIPPV in the presence of nocturnal hypercapnia alone can delay future daytime hypercapnia which occurs 4–5 years after NIPPV initiation.48

‘Late’ initiation of NIPPV with diurnal hypercapnia

In this approach, NIPPV is established when daytime hypercapnia occurs, meaning that patients have raised CO₂ levels day and night. Most patients are very symptomatic patients66 with VC < 20% predicted7 sometimes resulting in uncontrolled respiratory decompensation.67 At this stage of disease progression, DMD patients will show severe O₂ desaturations at night,11,19,42,68,69 with up to 30% sleeping time with SpO₂ < 90%.8 Published consensuses do not clearly guide clinicians on the correct time for NIPPV initiation. A European consensus considers NIPPV initiation when
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A PaCO$_2$ $\geq$ 45 mmHg.$^{70}$ The American consensus statement$^{23}$ considers two possible criteria; first one should consider NIPPV with the presence of symptoms accompanied by PaCO$_2$ $\geq$ 45 mmHg or nocturnal SpO$_2$ $\geq$ 88% for five consecutive minutes. The second consideration may indicate onset of NIPPV with the presence of symptoms, but accompanied here by less severe criteria such as MIP $\geq$ 60 cm H$_2$O or FVC $\leq$ 50% predicted. As previously stated, Ward et al.$^{22}$ provided powerful arguments in favour of commencing assisted ventilation in the presence of PCO$_2$ $>$ 48.9 mmHg during the night.

Contraindications and precautions for NIPPV

Contraindications are rare and include: lack of consciousness, lack of cooperation from the patient, inability to maintain the airway, presence of severe bulbar impairment and a lack of nasal access.$^{56,57,71}$ Patients should be taught airway clearance techniques prior to the commencement of NIPPV.$^{24,64,72,73-76}$ Education of caregivers and families on airway clearance techniques during home NIPPV is an important factor in the success of NIPPV.$^{76,77}$

### Table 1
Patient group demographics of studies reporting initiation of non-invasive positive pressure ventilation (NIPPV) in DMD patients

<table>
<thead>
<tr>
<th>First author</th>
<th>Year (months)</th>
<th>n DMD</th>
<th>VC (mL)</th>
<th>Age (years)</th>
<th>Option</th>
<th>PCO$_2$ criterion (mmHg)</th>
<th>PCO$_2$ measurement technique</th>
<th>PCO$_2$ (mmHg)</th>
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<td>22</td>
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<td>63$^*$</td>
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<td>14</td>
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<td>19.9</td>
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<td>$&gt;$45$^{**}$ Tc</td>
<td>89$^{**}$</td>
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<tr>
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<td>3</td>
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<td>16.1</td>
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<td>$&gt;$45$^{**}$ Tc</td>
<td>65$^{**}$</td>
<td>47</td>
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<tr>
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<td>19.3</td>
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<td>--</td>
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<td>21</td>
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<td>$&gt;$45$^*$ Art</td>
<td>52$^*$</td>
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<td>48.8$^*$</td>
<td>46</td>
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<td>$&gt;$50.5$^{**}$ ART/Tc</td>
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<td>73</td>
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<td>--</td>
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<td>$&gt;$48.9$^*$ Art/Tc</td>
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<tr>
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<td>$&gt;$45$^*$ Tc</td>
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<td>$&gt;$25</td>
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</tbody>
</table>

VC: vital capacity; L: late NIPPV initiation (see text); E: early NIPPV initiation (see text); P: preventive NIPPV initiation (see text); Art: arterial PCO$_2$ measurement; Tc: transcutaneous PCO$_2$ measurement; *wakeful PCO$_2$ measured during the day; **PCO$_2$ measured during sleep.

### Choice of the ventilator and settings

**Ventilator**

Intermittent positive pressure ventilators supplanted negative pressure ventilators previously used with DMD patients.$^{13,78}$ Two types of positive pressure ventilators were used: volume ventilators and pressure support ventilators. Both systems have similar effects on alveolar hypoventilation,$^{79,80}$ and on (partial) respiratory muscles unloading.$^{80-82}$ Recently, microprocessor-based ventilators were developed aiming at combining the advantages of pressure and volume ventilators.$^{83-85}$ This new generation of ventilators provides leaks compensation, targeted volume$^{86}$ and sensitive triggering for optimal synchronization$^{87}$ and comfort.$^{21,78,88,89}$ During the daytime, volume ventilators are preferable$^{48,77,90}$ for the benefits of air-stacking$^{16,74,90}$ and for the absence of leaks compensations enabling mouthpiece ventilation on demand.$^{48,90}$

**Settings**

Effective ventilation is easier achieved with low-inspiratory positive airway pressures (IPAP) between
10–15 cm H₂O although pressures > 20 cmH₂O have been reported. In NM patients, adequate setting for volume ventilators devices was found at high-respiratory rate (RR: 23/min) and low tidal volume (VT: 14 mL/kg of actual weight). Parameters may extremely vary among individuals.

**Interfaces**

Introduced in the 1980’s by Rideau et al.,1 the nasal mask still remains the first choice of interface for nocturnal ventilation,9,12,16–20,22,47,68,89,95–98 however, there is no agreement concerning the choice of the diurnal interface. Dr Alba introduced mouthpiece ventilation in DMD patients;16 daytime mouthpiece ventilation (Figure 2) has become a growing technique.90 A recent Belgian cohort study reported use of 24 h nasal/mouthpiece ventilation in 42 DMD patients over a period of 9.6 years.48 They concluded that it is a safe and effective form of ventilation for patients requiring 24/24 h NIV; the mean survival was 31 years. Some authors consider tracheostomy as more effective and more secure than non-invasive interfaces,14,20 although a similar risk of death with both non-invasive and invasive ventilation has been reported.90 Tracheostomy offers the possibility for direct suctioning,100 with an additional risk for tracheal erosion,93 mucus hypersecretion, granulation tissue formation,101 tracheobronchomalacia93 and tracheal haemorrhage.93,101,102 This morbidity may decrease the quality of life of the patient and caregivers. Patients are more likely to be cared for in an institution as opposed to the family home,48 while families and caregivers are exposed to greater demands for care.

The appropriate time to offer tracheostomy for 24 h MV remains controversial and depends on individual experiences.76 Traditionally, early tracheostomy is used more commonly in France compared to other countries103 despite requiring more carer and financial resources than NIPPV.106,103 Undoubtedly, tracheostomy should be considered when non-invasive techniques for airway clearance have become ineffective to treat chronic cough insufficiency.

**PCO₂ monitoring during MV**

As suggested by many recent clinical studies,9,21,22,37,40,48,68 TcCO₂ is considered as the first choice of technique to monitor nocturnal blood CO₂ tension in the follow-up of home MV.

**Efficacy of MV**

**Quality of life and symptoms**

DMD patients consider home MV beneficial for independent living and enhancing their overall health while health-related quality of life further decreases without MV.105 Nocturnal NIPPV reverses symptoms associated with poor sleep quality22,47,66 and full time MV reverses symptoms associated with daytime dyspnoea.48,55 Quality of life may be perceived differently in function of the interface. Tracheostomized patients express fewer positive statements than those ventilated non-invasively.104,106

**Blood gas improvement**

A consensus report23 and a Meta-analysis107 validated the short-term benefits of nocturnal MV on nocturnal (direct effect) and diurnal (indirect effect) blood gases in NM patients. Nevertheless, by contrast with other NM disorders,17,20,105 PCO₂ normalization is often suboptimal in DMD patients as suggested by PCO₂ ranging 45–50 mmHg during NIPPV (table 1).8,9,12,17,20 This phenomenon is not explained and warrants further investigation in order to avoid biased additional daytime ventilation to compensate for ineffective nocturnal NIPPV.

**Survival**

Cohorts of DMD patients receiving MV were compared to patients refusing MV.12,15 Survival rates at 24 and 30 months were 100% and 80%, respectively,
while death occurred within 10 months without MV. In two other studies, survival reached 25.3 years\(^{55}\) and 31 years\(^{108}\) with MV compared to 19.3 years\(^{55}\) and 20.4 years\(^{108}\) without MV. In DMD patients, MV is generally considered to provide an additional life’s expectancy of 5–10 years (Figure 1).\(^{9,48,55,108–110}\)

To date, there is no data to support an enhanced survival rate depending on timing of NIPPV initiation.\(^{34}\)

**Ethical considerations**

Quality of life is not estimated as lower by DMD patients after MV initiation, suggesting that MV itself does not adversely affect the perceived health status.\(^{68}\) However, resistances to recommend MV are reported in underinformed medical teams believing that MV brings a new handicap in DMD patients’ life.\(^{111}\) Information given to patients and family is never unbiased and often influences the choice for MV institution and evolution, especially when full time MV is required.\(^{106,111,112}\) The issue of euthanasia has never been reported in end-stage DMD patients.

**Indirect effect of nocturnal MV on gas exchange during spontaneous breath**

Three hypotheses were developed to explain the indirect mechanism of action of nocturnal MV in sustained normocapnia during the spontaneous breathing during wakefulness (SBW).\(^{113}\)

**Respiratory muscle rest**

This hypothesis is controversial and not convincing.\(^{14}\) Undoubtedly, NIPPV has been shown to unload inspiratory muscles,\(^{62,84,114}\) but the relation between muscle fatigue and respiratory failure is not evident.\(^{115}\)

**Improvement of respiratory system compliance**

In NM disorders, short sessions of 20 min per day of insufflations with positive pressure did not change the respiratory system compliance.\(^{116,117}\) The effects on compliance of NIPPV during the night have been reported as variable in DMD patients and require further investigation.

**Resetting of the central respiratory centre sensitivity for CO\(_2\)**

In NM disorders, resetting of the central respiratory centres may be the most important among the three hypotheses.\(^{99,118,119}\) Many studies support the hypothesis\(^{8,19,21,25,27,98,120,121–123}\) that nocturnal NIPPV leads to an upward resetting of the central respiratory centre CO\(_2\) sensitivity, indirectly reducing daytime hypercapnia.\(^{27,80,119}\)

Undoubtedly, future work is required to clarify the role of these three hypotheses at different stages of illness progression.\(^{119}\) Sessions of daytime ventilation are deemed to offer interesting area of research.\(^{27,124,125}\)

**What to do when NIPPV fails?**

All non-invasive techniques should be fully explored before providing tracheostomy. Failure of NIPPV may occur, due to technical issues, patient’s reluctance or due to disease progression.\(^{126}\)

**Technical issues**

Technical problems include mask discomfort, leaking and gastro intestinal bloating. Mask discomfort\(^{126}\) is easily solved by using another mask\(^{64}\) or a custom made mask.\(^{95}\) Leaking around the mask and/or by the mouth is associated with poor ventilation and sleep disruption,\(^{88,127,128}\) and is often related to poor mask fitting,\(^{126}\) to problems of timing of ventilation or to the use of excessive IPAP.\(^{126}\) The strategy to reduce leaking include increasing RR (≥23/min),\(^{94}\) decreasing IPAP (≤15 cm H\(_2\)O)\(^{129}\) or VT (≤13 mL/kg),\(^{94}\) or increasing the inspiratory/expiratory (I/E) ratio (from 1/1.5 to 1/1).\(^{126}\)

Mouth leaking\(^{24}\) and asynchrony\(^{87}\) may be improved by the use of microprocessor-based ventilators, which compensate for leaks. An additional mouth taping\(^{24}\) via a chin strap\(^{127,128}\) or a cervical collar\(^{126}\) is very useful in reducing mouth leaking. Mouthpiece with lip sealing or full face mask ventilation may be used when uncontrolled leaks are present.\(^{10}\) Finally, gastro-intestinal bloating may be a severe complication of NIPPV. Again, limiting IPAP is useful.\(^{126}\)

**Anxiety, lack of confidence, lack of intellectual cooperation**

Anxiety can be limited by educating the patient, family and caregivers. Patients are often more afraid about the idea of NIPPV than NIPPV itself. Anxiety
can easily be reduced by an informal trial during a consultation to actually experience ventilation via a nasal mask. A trial is also useful in patients with learning difficulties, one must bear in mind severe learning difficulties is not a contra-indication to NIPPV, although the process can be challenging.

**Disease progression**

Tracheostomy ventilation has become less common within the disease progression management of DMD patients. As previously said, DMD patients are being managed safely with a series of non-invasive techniques.

**Short-term physiological effects of MV**

In non-NM patients receiving NIPPV, VT and Ti increase, while RR, p0.1 and the tension-time index all reduce during assisted ventilation. During intervals of spontaneous breathing between periods of NIPPV, RR remains constant or increases with increasing p0.1. The impact on the short-term of MV on spontaneous breathing has not been studied in DMD patients.

**Long-term physiological effects of MV**

Long-term physiological outcomes of MV are conflicting in DMD patients: VC have been shown to increase to stabilize, to decline at a slower rate or to decline further similarly to that prior to MV. One explanation for the inconsistent reports may be related to the use of different types of ventilators and settings. Hospitalization rates have been reported as reduced or unchanged after initiation of MV and therefore we would recommend further investigation in this area.

**Full time ventilation**

**When is full time MV required?**

Extension of MV to daytime use is empirically driven. The use of MV from 18 to 20/24 h has been reported with VC ≤ 400 mL to VC < 300 mL, respectively. Ventilator free time was documented as less than 15 min in patients with VC < 10% predicted. The ATS consensus statement for DMD patients recommends daytime MV when PCO₂ > 50 mmHg and/or SpO₂ < 92% while awake. Those criteria were recently reported as difficult to reach in symptomatic patients in whom diurnal PCO₂ > 45 mmHg served as decisive factor to extend NIPPV into the daytime. Worsening of the symptoms during the day suggest a high cost of energy expenditure spent in maintaining normocapnia. Reversal of the symptoms may be expected with a few hours of daytime NIPPV, but this has not yet been demonstrated. Controlled investigations should help clinicians in deciding when and how to start daytime ventilation, possibly by the use of a simple symptom score.

**Outcome of 24 h MV**

There is little information on the outcome of 24 h MV. Three studies reported prolonged survival in DMD patients with a combination of daytime mouthpiece ventilation and nocturnal nasal NIPPV with stabilization of VC over five years. Undoubtedly, tracheostomy and intubations may be avoided on the condition that cough-assistance techniques are available. With mouthpiece ventilation, the challenge of eating together with breathing via the mouth may lead to the loss of weight and consequently to the placement of enteral feeding.

**Conclusions**

Undoubtedly, nocturnal ventilation is safe and effective to treatment in DMD patients with hypercapnia. However, the ideal timing of when to initiate nocturnal assisted ventilation remains debatable. The ultimate timing to offer full time ventilation with the most advantageous interface is also lacking in evidence. Future investigations are required to enable clinicians to refine the introduction and long-term follow-up of both nocturnal and full time-assisted ventilation in DMD patients.

**References**


