Cervical spinal cord injury (CSCI) is a rare but devastating condition, with immense implications for patient and health care resources. In most cases, spinal cord injury (SCI) is associated with radiological abnormalities and the spinal cord may appear compressed, contused or oedematous on magnetic resonance imaging (Figure 1). Respiratory complications are the leading cause of morbidity and mortality in the short and long term after injury.1-4

The incidence of traumatic SCI is 15 per million per year in Australia, 16 in Europe and 39 in North America.4 In Australia, 300–400 cases of SCI are reported every year.5 CSCI accounts for 40%–60% of all SCI.4

If the neurological level of injury (NLOI) is at or above C5, the diaphragm will be affected. Intercostal and abdominal muscles are paralysed in complete CSCI. As a result, both inspiratory and expiratory function may be severely compromised. At presentation, most patients with complete CSCI require endotracheal intubation and mechanical ventilation (MV).6 Injury above C5, complete injury, atelectasis, copious respiratory secretions and pneumonia predict the need for MV in the first week after injury.1

Most patients with complete injury at or above C5 will also require tracheostomy to facilitate MV or protect the airway.5,7 The time to weaning from MV after CSCI usually varies from weeks to months.8-10 Up to 40% of patients with complete injuries at or above C4 can become ventilator-dependent.2 There are limited high-quality data to guide MV and weaning in patients with CSCI. In this article we review the pathophysiology of breathing in patients with CSCI, and strategies for MV and weaning.

Abbreviations

ASIA American Spinal Injury Association
CSCI Cervical spinal cord injury
EMG Electromyography
FEV1 Forced expiratory volume in 1 second
FVC Forced vital capacity
IMT Inspiratory muscle training
LOS Length of stay
MV Mechanical ventilation
NIV Non-invasive ventilation
NLOI Neurological level of injury
PEEP Positive end-expiratory pressure
SCI Spinal cord injury
SDB Sleep-disordered breathing
VC Vital capacity
VFB Ventilator-free breathing
VT Tidal volume

ABSTRACT

Respiratory complications following cervical spinal cord injury are common and are the leading cause of morbidity and mortality after this type of injury. Impaired mechanics of ventilation, poor cough, increased secretions and bronchospasm predispose to atelectasis, pneumonia and exacerbations of respiratory failure. Prolonged mechanical ventilation and tracheostomy are often required. This review discusses the relevant pathophysiology, various ventilatory strategies and timing of tracheostomy, and examines the evidence surrounding physiotherapeutic and pharmacological treatment options.
Physiology of normal breathing

In humans, inspiration is an active process, and quiet expiration occurs due to elastic recoil of lungs and chest wall. The nerve supplies of the key muscles of ventilation are listed in Table 1. The diaphragm is the main muscle of inspiration. The phrenic nerve, which innervates the diaphragm, originates predominantly from the C4 spinal segment, with a variable contribution from C3 and C5. Contraction of the diaphragm increases the vertical dimension of the thoracic cavity and the anteroposterior diameter of the lower rib cage by elevating the ribs at its insertion. The resultant negative intrathoracic pressure draws air into the lungs. The external intercostals and the parasternal component of the internal intercostal muscles are also active during inspiration. In the neck, the scalene muscles are obligatory muscles of inspiration that are active even in quiet breathing. The sternocleidomastoid is an accessory muscle of inspiration that contracts only during deep or forceful breathing. The muscles active in forced expiration or cough include the abdominals, the interosseous part of the internal intercostals, triangularis sterni, serratus posterior inferior, latissimus dorsi and quadratus lumborum.

Respiratory dysfunction after cervical spinal cord injury

The degree of respiratory dysfunction after CSCI depends on the neurological level and completeness of injury. Severity of injury is commonly assessed by using the American Spinal Injury Association (ASIA) International Standards for Neurological Classification of Spinal Injury (http://www.asia-spinalinjury.org/publications/59544_sc_Exam_Sheet_r4.pdf). This should be accurately documented at admission for comparative and prognostic purposes. Paralysis of the diaphragm may result from injury to the spinal cord or the phrenic nerve itself. In injuries above C5, paralysis of the diaphragm results in reduced tidal volume (VT) and vital capacity (VC). Abdominal compliance is increased due to loss of tone in paralysed abdominal muscles. In complete injuries, the sternocleidomastoid and scalene muscles become more important for inspiration and the medial part of pectoralis major becomes important for expiration. The NLOI correlates inversely with a reduction in VC. Lung and chest wall compliance is also reduced within a month of injury, which most likely is due to airway secretions, mucus plugging, atelectasis and altered proprioceptive reflex control of chest wall muscles. Chest x-ray often shows widespread atelectasis (Figure 2). Later on, progressive ankylosis of chest wall joints becomes important. Abdominal compliance is increased due to decreased abdominal muscle tone. The mechanics of breathing are disordered after CSCI. Among patients with CSCI in whom the diaphragm is preserved (ie, below C4) but the other muscles of inspiration are paralysed, the upper rib cage retracts inwards with each inspiration due to paralysis of the intercostal muscles. This leads to reduced efficiency of ventilation, with a lower VT for a given diaphragmatic excursion. The resting position of the diaphragm is affected by the pressure exerted by the abdominal viscera. In a supine position, the diaphragm is at a higher position in the rib cage, and therefore has greater excursion than in upright position. This results in greater VC and improves comfort in the supine position.

![Figure 2. Bilateral extensive atelectasis resulting from poor tidal volume, reduced vital capacity and retained secretions](image-url)
patients with CSCI typically feel more breathless when upright than when supine.

Cough is impaired due to paralysis of the abdominal musculature. Effective cough also requires deep inspiration, which is impaired after CSCI. This can cause retained secretions and subsequent respiratory tract infections, which are the leading acute cause of death in patients with CSCI. The clavicular portion of pectoralis major plays an important role in effective coughing after quadriplegia. Training the pectoralis major can improve the strength of cough.

Sympathetic denervation to the lungs after CSCI leads to relative parasympathetic overstimulation, resulting in an increase in quantity of secretions and bronchial tone. Sleep-disordered breathing (SDB) is common after CSCI. It is present as early as 2 weeks after or as late as 6 months after the injury; up to 80% of patients may have SDB. The reason for the increased prevalence of SDB in the CSCI population is unclear and certainly is not explained by a hypothesis solely involving diaphragmatic weakness. It is possible that the (non-diaphragmatic) inspiratory muscle afferent projections from an intercostal muscle group facilitate the cranial nerve motor neurones subserving upper airway stabilisation during inspiration.

In the first year after injury, the forced vital capacity (FVC) progressively improves. Maximum improvement occurs within the first 5 weeks, with more gradual improvement thereafter. Multiple factors contribute to improvement of respiratory function. Diaphragmatic function improves progressively over time, and the contribution of intercostals and neck muscles also improve. An increase in tone of the abdominal muscles reduces the abdominal compliance, while spasticity of the intercostal muscles stabilises the chest wall. This reduces retraction of the upper thoracic wall during inspiration, thereby improving efficiency of ventilation. A history of smoking, obesity and age over 60 years are associated with accelerated decline in forced expiratory volume in 1 second (FEV1) and FVC after CSCI. The changes in respiratory function, along with their proposed mechanism, are listed in Table 2.

### Endotracheal intubation after cervical spinal cord injury

When endotracheal intubation is required for a patient with suspected or confirmed cervical spinal instability, there is a justifiable fear that secondary neurological injury may be caused by the process of intubation. Immobilisation of the spine in at-risk patients at the time of first system contact is accepted as the standard of care. There are a variety of techniques that are employed to secure endotracheal intubation.

In time-critical circumstances, rapid sequence induction and intubation using direct laryngoscopy with manual in-line stabilisation appears safe and appropriate. During direct laryngoscopy and intubation, most cervical motion occurs at the craniocervical junction, and the subaxial cervical segments subjacent to and including C4 are minimally displaced. Manual in-line stabilisation does not eliminate all movement, but there is no evidence that this technique produces inferior neurological outcomes to any other.

If sufficient time and expertise exist, awake fibre-optic intubation has the potential advantages of minimising any cervical spinal movement and being able to perform neurological assessment immediately after intubation. Of note, the success rate of awake fibre-optic intubation in trauma has been cited at 83%, although failure is associated with morbidity and mortality. With improving technology, several new indirect laryngoscopic devices are being widely used. These include Storz V-Mac and C-Mac, GlideScope, the McGrath laryngoscope, Pentax Airway Scope, Airtraq Optical Laryngoscope and Bullard Laryngoscope. None of these devices have been shown to improve neurological outcomes. However, for patients in cervical immobilisation, the GlideScope and the Storz V-Mac have been shown to improve glottic visualisation, and the airway scope improved Cormack–Lehane grade, intubation difficulty score and decreased upper cervical movement compared with direct laryngoscopy.

### Table 2. Changes in respiratory function after cervical spinal cord injury

<table>
<thead>
<tr>
<th>Pathophysiological change</th>
<th>Mechanism</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reduced vital capacity</td>
<td>Paralysis of inspiratory muscles, Distortion of upper rib cage during inspiration, Reduced lung compliance</td>
</tr>
<tr>
<td>Reduced chest wall compliance</td>
<td>Increased tone of intercostal muscles, Altered articulation of ribs to sternum and spine</td>
</tr>
<tr>
<td>Reduced lung compliance</td>
<td>Low functional residual capacity, Recurrent respiratory infections, Altered properties of surfactant</td>
</tr>
<tr>
<td>Impaired cough</td>
<td>Paralysis of abdominal muscles, Reduced vital capacity</td>
</tr>
<tr>
<td>Increased secretions and bronchial tone</td>
<td>Decreased sympathetic tone</td>
</tr>
</tbody>
</table>

Table 2. Changes in respiratory function after cervical spinal cord injury
Mechanical ventilation after cervical spinal cord injury

After CSCI, MV is usually initiated to manage respiratory failure. It may also be required to aid tracheal toilet or to provide airway protection if the level of consciousness is impaired. Specific risk factors for intubation related to SCI include NLOI above C5 and complete injury, with 74% of all SCI patients requiring intubation. Excessive respiratory secretions and pneumonia are independent predictors for the need for MV. Many patients will need intubation for spinal surgery. Extubation of these patients may not be straightforward.

In high SCI, the duration of MV may be months to years and patients may remain ventilator-dependent. The duration of intensive care admission may be affected by availability of an appropriate rehabilitation bed, which has significant resource implications. Management of MV after CSCI therefore requires patience, and appreciation of the time frame required for improvement and the nuances of weaning a patient with SCI.

In the acute stage, the mode and setting for MV in CSCI patients is dictated by associated conditions, such as the need for carbon dioxide control in traumatic brain injury or hypoxaemia secondary to acute respiratory distress syndrome, pulmonary contusion or infection. Once the acute issues have been resolved and weaning from the ventilator is the predominant problem, MV should be tailored to CSCI. While a patient is on a ventilator, baseline spirometry measurements including maximum inspiratory pressure and VC should be obtained at the earliest time possible.

Tidal volume

This is a controversial topic with differing schools of thought and a paucity of evidence. Following Acute Respiratory Distress Syndrome Network trials, conventional wisdom in ventilating general ICU patients is that a lower (6–8 mL/kg ideal body weight) VT reduces ventilator-associated lung injury, and there is evidence that ventilation with a VT of 10 mL/kg compared with 6 mL/kg in all comers to ICU is associated with sustained plasma cytokine production and may contribute to the development of lung injury in patients without acute lung injury at the onset of MV.

However,Gattinoni and Pesenti’s concept of the “baby lung” does not intuitively apply to the generally healthy lungs of patients with CSCI. Tetraplegic patients often experience air hunger when lower-VT ventilation is used, even with a normal PaCO2, and there is some evidence that high-VT ventilation may improve success of weaning from MV.

Peterson and colleagues retrospectively evaluated the effect of two high-VT strategies in patients with C3–C4 tetraplegia, who were successfully weaned in a spinal injury centre. All patients received VT >10 mL/kg at the start of the study. Atelectasis was treated by increasing VT. VT was recorded at 2 weeks after the day of admission. Based on the VT at a point 2 weeks after admission, patients were divided into those receiving high VT (mean, 1.7 L) and those receiving low VT (mean, 1.18 L). The time to wean was less in high-VT patients compared with low-VT patients. Interestingly, there were no complications related to barotrauma. There are several important limitations of this study. It was small, single centre, and looked retrospectively only at the patients who were weaned successfully. Overall success rate was not reported. There were no patients with a VT less than 10 mL/kg, which is no longer representative of how other patients in ICU are ventilated. These findings have never been confirmed in a prospective, multicentre, randomised trial.

There are several other case series from spinal cord rehabilitation facilities reporting ventilation with VT 10–15 mL/kg as standard practice, and many experts are convinced from personal experience that this strategy reduces atelectasis and particularly upper lobe collapse with prolonged MV of patients with SCI. As tone returns to the intercostal muscles and they form a rigid, fixed thoracic wall, higher VT can help the intrathoracic volume to be fixed at a more advantageous size.

Among patients with CSCI who are ventilated and have healthy lungs, high VT in the range of 10–15 mL/kg can be considered. If high VT is used, additional dead space may have to be introduced into the circuit to prevent hypocarbia, which can reduce the ventilatory drive.

Positive end-expiratory pressure

The optimal positive end-expiratory pressure (PEEP) in ventilated patients with CSCI is unknown. Too much PEEP may lead to excessive flattening of the diaphragm due to high abdominal compliance, resulting in mechanical disadvantage when the diaphragm is the predominantly active muscle of inspiration. Too little PEEP may cause atelectasis, particularly in the dependent portions of the lungs and left lower lobe. The optimal PEEP has not been studied in a controlled fashion. Ventilation strategies using high VT generally use low or no PEEP and rely on the high VT to prevent atelectasis. There are no studies comparing high-PEEP, low-VT with low-PEEP, high-VT strategies. Without significant atelectasis, a low PEEP is reasonable.

Extubation or tracheostomy

Patients with low CSCI (C5 or below) or incomplete injuries may be candidates for early extubation. The risk of failed extubation is high due to pulmonary secretion retention and inadequate cough. Failed extubation is associated with increased risk of pneumonia, prolonged
Tracheostomy was required for 58% of patients with a complete SCI and 48% with incomplete injuries. Berney and colleagues reported that 60% of patients with CSCI, 67% of patients require tracheostomy. Early identification of patients who will require tracheostomy is important but difficult. One case series has shown that a tracheostomy fashioned before Day 7 reduced the duration of MV and ICU LOS. In another study, early tracheostomy (within the first 7 days after intubation) was associated with a shorter duration of MV after tracheostomy and a shorter ICU stay. In 45% of cases, tracheostomy was placed percutaneously. There was no difference in the complications after surgical compared with percutaneous tracheostomy.

Early tracheostomy should therefore be the goal in patients for whom the need for tracheostomy is inevitable. Early tracheostomy, within the first 10 days after anterior cervical spine fixation, does not increase the risk of wound or implant infection. Percutaneous dilatational tracheostomy is safe even after an anterior fixation of cervical spine.

Weaning from mechanical ventilation

The slow nature of weaning from MV among patients with CSCI presents unique issues for the ICU. When several specialists from ICU and other specialties are involved, it becomes essential that a long-term weaning strategy is agreed upon, documented and adhered to.

In a prospective case series, Gutierrez and colleagues used an elegant resistance and endurance protocol (REP) to

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### Box 1. Checklist before extubation

- No further surgery anticipated
- No further procedures or imaging outside the intensive care unit anticipated
- No significant hypoxia or reversible respiratory pathology
- Patient awake and able to cooperate with physiotherapy and non-invasive ventilation (NIV)
- No contraindication for assisted cough or chest physiotherapy (eg, rib fractures, major laparotomy)
- No contraindication for NIV (eg, facial fractures)
- Minimal bronchial secretions
- Negative inspiratory pressure > 20 cm H₂O
- Vital capacity > 10 mL/kg
- Cuff leak present (particularly after spinal stabilisation by anterior approach)
- Fluid balance optimised
- Airway plan and equipment prepared for reintubation if required

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The most common cause of failed extubation is retained secretions in the airways due to weak cough. The importance of good physiotherapy in the immediate postextubation period cannot be overemphasised. In the first 48 hours after extubation, physiotherapy is usually required around the clock. Exubation outside of usual working hours is not advised if experienced physiotherapists are not available.

Non-invasive ventilation (NIV) is often routinely used immediately after extubation and there is some evidence supporting this practice. In a case series, Bach and colleagues reported extubation of patients with neuromuscular disorders, 11% of whom had SCI, directly to NIV. NIV and aggressive chest physiotherapy led to successful extubation in 95% of cases. All extubations were successful if assisted peak cough flow rate was greater than 160 L/min. Bilevel NIV should be used rather than single level, as inspiration must be assisted. NIV is typically required for several days after extubation and a choice of different interfaces should be available. Protective dressing over the nasal bridge should be used to prevent pressure areas from the interface.

Patients who fail extubation once are unlikely to be extubated successfully again and will usually require tracheostomy. Box 1 provides a list of variables that should be considered before extubation.

### Timing of tracheostomy

Early identification of patients who will require tracheostomy is important but difficult. One case series has shown that a tracheostomy fashioned before Day 7 reduced the duration of MV and ICU LOS. In another study, early tracheostomy (within the first 7 days after intubation) was associated with a shorter duration of MV after tracheostomy and a shorter ICU stay. In 45% of cases, tracheostomy was placed percutaneously. There was no difference in the complications after surgical compared with percutaneous tracheostomy.

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Duration of MV, prolonged intensive care unit length of stay (LOS) and increased hospital mortality. However, early extubation and aggressive respiratory management can prevent the need for tracheostomy and its associated complications. Tracheostomy rates after CSCI vary widely, from 20% to 60%. The NLOI and completeness of injury are the most important predictors of requirement for tracheostomy. Berney and colleagues conducted a survey of clinical experts in Australia to identify factors used to decide between extubation or tracheostomy, then used these factors for data collection in a prospective observational study and for development of a classification and regression tree. FVC less than 11.9 mL/kg, endotracheal suction more than every hour and PaO₂/FiO₂ less than 189 mmHg were predictive of tracheostomy. Extubation failure occurred in 8.7% (4/47) of patients, all of whom subsequently required a tracheostomy.
wean patients with incomplete CSCL. Although there were only seven patients in the case series, the REP is worthy of detailed review. Before weaning exercises were started, patients were preoptimised. Respiratory optimisation was done by tracheal suctioning in the Trendelenburg position, use of bronchodilators and transient hyperinflation of lungs. For inspiratory resistance training, patients breathed through a fixed inspiratory resistor applied at the mouth for 10 seconds, four times a day. Resistance was progressively increased as tolerated. This was followed by expiratory resistance training by breathing through an expiratory resistor. Endurance training was initially done on ventilator by gradually reducing the support. When patients were ready, ventilator-free breathing (VFB) was used for endurance training and time off the ventilator was gradually increased. Using this three-step approach, both high cervical (C2; n = 2) and low cervical (C4–C7; n = 5) injury patients gained muscle strength. C4–C7 tetraplegic patients were weaned off the ventilator in 1–2 months. C2 tetraplegic patients were able to tolerate a short period off the ventilator, which increases safety in situations like accidental disconnection. This was an uncontrolled case series. However, the results of the series are compelling and their protocol deserves to be prospectively studied in a larger study. The same authors have also shown in a randomised crossover trial that chest optimisation in the Trendelenburg position, compared with a supine position, prolongs the duration of a spontaneous breathing trial.

Using a protocol of progressive VFB, Atito-Narh and colleagues reported 69% success in weaning patients with CSCL. Most patients with an injury at or above C3 could not be weaned. In this study, the tracheostomy cuff was deflated and speech valve was used during VFB. The mean time to wean patients was 56 days. The consistent factors underpinning successful weaning after spinal cord damage were accurate neurological assessment; prevention of pulmonary atelectasis by physiotherapy; VFB graduated according to VC; rest periods with controlled ventilation; cuff deflation with translaryngeal airflow, and regular tracheostomy tube changes.

Trendelenburg or supine positions are usually better tolerated than sitting-up position during weaning exercises for the reasons previously described. An abdominal binder can compensate for low abdominal muscle tone and aids weaning in sitting position. The binder must be sufficiently narrow that it does not rest over the pelvis or the lower margin of rib cage.

The alleviation of patient anxiety and the treatment of depression are important, as these conditions can contribute to weaning failure if left untreated. During VFB, exhaustion should be assessed by monitoring of respiratory rate, hypoxia, hypercarbia and drowsiness. Intercostal reces-
methacholine. Several small studies have shown modest increased responsiveness to agents like histamine and an anabolic effect on the respiratory muscles. Airway FVC and peak expiratory flow rate, and may have induced showed that 4-week administration of salmeterol improved quad cough and the use of a mechanical insufflation/exsufflation device, but whether this translates to meaningful clinical end points is yet to be shown definitively.

Prediction of success of weaning
If patients who will require lifelong MV can be identified from the outset, it may be possible to decrease their ICU and hospital LOS and expedite the prescription of their necessary equipment. NLOI and completeness of injury are the strongest predictors of weaning success, which emphasises the importance of accurate neurological assessment. Patients with complete injury at C3 or above are unlikely to be weaned, and the chance of requiring regular MV increases as the NLOI ascends. Unfortunately for many patients with a complete NLOI from C3 to C6, it is hard initially to exclude the possibility of weaning success and the individual’s circumstances must be examined carefully. Other factors that can help predict the likelihood of long-term ventilator dependence include the absence of diaphragm motor unit recruitment with invasive electromyography (EMG) and spirometry. Diaphragmatic EMGs are not universally available, may be affected by critical illness-associated weakness (carrying a different prognosis), and alone are not sufficiently sensitive to exclude the possibility of weaning. The role of transcutaneous diaphragmatic EMG, a much simpler investigation than invasive EMG, in predicting suitability for weaning is yet to be clarified.

Pharmacological adjuncts in respiratory care
Patients with CSCI have increased airway resistance due to loss of sympathetic nerve supply. The airways have increased responsiveness to agents like histamine and methacholine. Several small studies have shown modest but significant short-term improvements in pulmonary function with ipratropium and metaproterenol (orciprenaline) in tetraplegic patients. A randomised controlled trial showed that 4-week administration of salmeterol improved FVC and peak expiratory flow rate, and may have induced an anabolic effect on the respiratory muscles. Airway hyperresponsiveness in tetraplegia may also be modulated by baclofen and oxybutynin, both commonly prescribed in SCI for muscle and bladder spasticity, respectively. Contradictory findings in small studies suggest these medicines should not be prescribed solely to attenuate airway hyperresponsiveness. Small animal studies have suggested that theophylline may improve respiratory function after hemisection of spinal cord, but a human study using oral theophylline failed to show any benefit.

Testosterone derivatives such as oxandrolone have been investigated as adjunct to improve respiratory function by increasing respiratory muscle mass and strength. An initial small, uncontrolled study examined 10 patients with complete C4–C5 tetraplegia who received oxandrolone for 1 month. Significant improvements were seen in FVC, FEV1, and maximum inspiratory pressure, along with a significant decrease in dyspnoea. In a subsequent small, uncontrolled study, oxandrolone (20 mg/day) did not lead to any significant improvement in pulmonary function tests, but there were elevations in liver enzymes and low density lipoprotein. With minimal evidence of benefit and significant potential for harm, the use of anabolic steroid in this context is currently considered investigational.

Pacing as an alternative to long-term ventilation
Patients with complete injury at or above C3 are likely to become ventilator-dependent. Such patients may benefit from electrical diaphragmatic pacing to provide liberation from the ventilator. Stimulating the phrenic nerve directly in the neck or chest is the most common alternative method of managing inadequate spontaneous ventilation. Long-term studies suggest this is a realistic long-term ventilation option. Hirschfeld and colleagues compared 32 patients with phrenic nerve pacing to conventional ventilation and found that the initial cost of the phrenic pacing was high, but there was lower rate of respiratory infection, better speech and lower running costs. Sensation of smell is restored and patients report a more natural quality of breathing. Sufficient numbers of intact phrenic axons are required bilaterally, so candidates for phrenic pacing have both an SCI above C4 and a largely intact C4 cord segment. There are several newer methods of electrically stimulated breathing. The modality with most applicability to the ICU setting is external diaphragmatic pacing. This system uses electrodes endoscopically implanted directly into the diaphragm. The electrodes are connected to an external stimulator by wires that exit through the skin. There is a possibility that this system could be used earlier after SCI and hence could have implications for the acute management of ventilator-dependent tetraplegia in the ICU setting, but additional data are required.

Summary
Respiratory complications following CSCI are common and serious, but may be anticipated and prevented. Patients with high and complete CSCI are likely to need initial and
prolonged MV. An understanding of CSCI explains the predictability of early respiratory complications, the efficacy of supine positioning, early physiotherapy, bronchodilators and abdominal binders.

In the initial phase, the mode and settings of MV will be dictated by any associated pathological features. Later, a strategy including \( V_{\text{t}} \) of 10−15 ml/kg and lower PEEP may be considered, although there is minimal evidence to support this. The need for tracheostomy may be predicted by factors including NLOI, FVC, \( \text{PaO}_{2}/\text{FiO}_{2} \) ratio and the amount of tracheal suctioning required, and percutaneous dilatational tracheostomy is safe even after anterior fixation of the cervical spine.

Physiotherapy is essential, but the specific components which are most beneficial have not been elucidated. \( \beta \)-Agonist bronchodilators should be used if pulmonary function tests suggest an obstructive airways limitation but there is currently insufficient evidence to recommend theophylline or oxandrolone.

Diaphragmatic pacing may have a role in ventilator-dependent patients, even while still in ICU. Early involvement of a multidisciplinary team to instigate physiotherapy and occupational therapy, coordinate ventilator weaning, identify rehabilitation needs, and manage the tracheostomy, is strongly recommended.

Competing interests
None declared.

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