

CBLM: RESPIRATORY

INTRODUCTION

Respiratory complications are common in patients with an acute or chronic spinal cord injury (SCI), and are the leading cause of mortality following SCI.¹ This module will focus on respiratory complications that arise in patients with SCI outside of the acute post-injury setting.

Objectives:

This module aims to help clinicians:

- Gain awareness of the common respiratory complications in patients with SCI.
- Promptly identify and manage these complications.
- Outline an approach to monitoring respiratory status in patients with SCI.

CASE

Jordan, age 34

Jordan is new to your practice. He moved to the city to be closer to his family and supports after sustaining an All-Terrain Vehicle accident four years ago that left him with a complete SCI at the C5 level. He initially required mechanical ventilation, but during the weeks following his accident, he was able to wean off respiratory support. At his Periodic Health Exam today, Jordan reports feeling generally well, but does endorse feeling slightly more short of breath with talking, dressing and transfers in the last few months.

With regards to his respiratory status, what additional information would you like to know?

- *On history, it would be important to ask Jordan more about his symptoms (onset, triggers, associated symptoms, etc.) and explore risk factors for respiratory complications such as smoking status (cigarettes and/or marijuana), chronic lung conditions (e.g., asthma/COPD), as well as review history of pulmonary embolism or recurrent respiratory infections/hospitalizations.*
- *Any changes in neurologic function [change in sensory level or change in motor function might be suggestive of syringomyelia (cyst within spinal canal that can occur after SCI and may cause further neurological decline)] or bodily changes (eg. weight gain)*
- *It would be useful to know when Jordan last had his pulmonary function assessed with spirometry or PFTs and if he is aware of any trends in his results.*
- *Asking about any use of respiratory treatments (abdominal binder, puffers), assistive breathing and/or secretion clearance techniques (such as assisted coughing, breath stacking, chest physiotherapy, etc.) would provide a better understanding of Jordan's respiratory status and self-management.*
- *Additionally, it would be important to ask Jordan about the impact of dyspnea on his functioning and quality of life as well as screen for sleep-disordered breathing by asking about snoring, morning headaches and daytime drowsiness.*
- *Physical examination should include vital signs including oxygen saturation if available (especially if looking for signs of infection or impending respiratory failure), examination of the respiratory system and assessment for edema.*

INFORMATION SECTION

Pathophysiology:

All patients with a lesion above T12 will have some form of respiratory dysfunction. Respiratory complications are related to the level and completeness of the injury (*see Appendix 1 for more detail*) and factors such as pre-morbid conditions (e.g., asthma and COPD), and history of previous or current smoking have an impact.²

Both muscles of inspiration and expiration are important in respiratory health. Impairment in inspiration leads to decreased vital capacity and gas exchange, and can ultimately lead to respiratory failure. Normal expiration is passive and does not require muscle use; however, expiratory muscles are required for forced exhalation such as during exercise or coughing. Expiratory muscle dysfunction severely compromises the ability to generate an effective cough and clear respiratory secretions, which in turn predisposes to atelectasis and respiratory infections. Expiratory muscles also exert an indirect effect on inspiration and vital capacity by stabilizing the rib cage and contributing to normal function of the diaphragm.

Inspiration impaired → ↓ vital capacity and gas exchange → respiratory failure
Expiration impaired → ineffective cough → atelectasis → respiratory infections

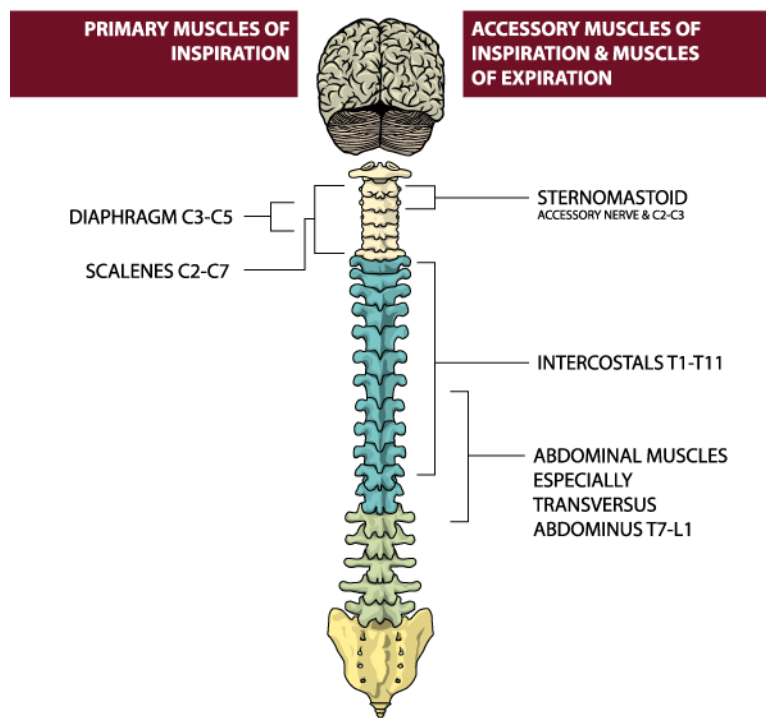


Figure 1: Muscle innervation reproduced from <http://eprimarycare.onf.org/RespiratoryComplications.html>

Secondary consequences of SCI occurring within the lungs include:

- Reduction in lung and chest wall compliance
- Changes in ventilatory control
- Airflow limitation and bronchial hyperresponsiveness

Risk Factors:

Risk factors for respiratory complications in patients with spinal cord injury include:³

- Higher level of complete neurological impairment (i.e. cervical)
- Age > 50 years
- Recent hospital admission
- Smoking
- Chronic lung disease (e.g., Chronic Obstructive Pulmonary Disease [COPD])
- Severe postural deformity
- Obesity
- Decrease in pulmonary function tests

Respiratory Complications:

Respiratory complications include:

- Respiratory insufficiency
- Atelectasis
- Pulmonary infection
- Pulmonary thromboembolism
- Dyspnea
- Sleep-disordered breathing
- Pulmonary edema
- Dysphonia

Respiratory Insufficiency:

Respiratory insufficiency requiring mechanical ventilation occurs in about one in five patients within the first week of SCI, and can also occur later in the presence of contributing factors such as:⁴

- Atelectasis
- Pneumonia (most frequent cause of late ventilatory failure)⁵
- Secretion retention
- New onset cardiopulmonary disease (COPD, heart failure)
- Restrictive pulmonary disease secondary to progressive kyphoscoliosis
- Post-traumatic syringomyelia (fluid filled cavity in the spinal cord)
- Cervical spinal stenosis with compression
- Subdiaphragmatic problems (i.e abscess or fluid collection) that impair motion of the diaphragm
- Obesity

Clinical Pearl:

Signs and symptoms of impending respiratory failure include:

- Hypoxia
- Tachypnea
- Cyanosis
- Altered level of consciousness
- Accessory muscle use
- Increasing O₂ requirement (i.e worsening dyspnea at rest)
- Tachycardia

- Fatigue
- Decrease in vital capacity to < 15 mL/kg ideal body weight

Clinical Pearl: Respiratory failure can occur quickly in patients with SCI so the threshold for referral to the ED should be low.

Atelectasis:

Because of impaired cough and difficulty mobilizing lung secretions, patients with SCI are at increased risk for atelectasis (partial collapse of lung) and subsequent pulmonary infections.

Signs and Symptoms of atelectasis may include:

- Tachypnea
- Tachycardia
- Decreased oxygen saturation
- Shortness of breath
- Cough
- ↑ Anxiety
- ↑ Volume or thickness of secretions
- ↓Vital Capacity (VC) ↓Peak Expiratory Flow (PEF)

Investigations:

As with respiratory insufficiency, monitoring vital capacity or peak expiratory flow rate can be used to detect early problems/atelectasis.

Routine screening CXR is not recommended. However, if there is a clinical suspicion for infection, investigations should be promptly initiated.

Prevention/Management:

Atelectasis needs to be recognized and treated early to avoid further complications. Interventions such as chest physiotherapy and/or respiratory and pulmonary clearance techniques can help prevent and/or treat atelectasis and reduce the risk of pneumonia following SCI and are recommended in all patients with cervical or thoracic SCI^{6,7}. The aim is to maximize lung expansion and mobilize and clear secretions. Patients should be referred to an allied health professional (physiotherapist, respiratory therapist) with experience in chest physiotherapy and the family physician should encourage more simple measures such as deep breathing and incentive spirometry. (*See Assistive Devices and Respiratory and Pulmonary Clearance Techniques below for more details.*)

Pulmonary Infection:

Pneumonia is the leading cause of death for all persons with SCI. Although the incidence of pneumonia is highest in the first year following SCI, these patients remain at increased risk over their lifetime.⁸

The clinical presentation of pneumonia is similar to that of atelectasis (*see Signs and symptoms of atelectasis above*) but the symptom onset is generally more acute and the patient more unwell. Fever may also be

present in pneumonia. Atypical presentations such as autonomic dysfunction (see *Autonomic Dysreflexia*) are also possible.

Clinical Pearl: Viral respiratory infections commonly precede or precipitate hospitalizations, especially in patients with chronic underlying pulmonary conditions. Nearly 45% of patients hospitalized with acute respiratory conditions have evidence of recent respiratory tract viral infections.³⁵ Given the high rate of and seriousness of pneumonia in SCI this is a concern.

Investigations:

If pneumonia is a consideration, then chest x-ray should be obtained.

Other possible investigations for pneumonia include:

- Computed tomography (CT)
- Sputum Gram stain and culture
 - If increased sputum or change in sputum

Clinical Pearl: Sputum Gram stain and culture are of low diagnostic yield but should be strongly considered as they can potentially identify an unsuspected highly resistant organism. Also, if a low virulence organism is identified, the antibiotic spectrum may be narrowed to avoid promoting antibiotic resistance.

Respiratory infections can rapidly progress to respiratory insufficiency and should be identified and managed promptly.

Prevention:

Preventative measures for pneumonia include: prevention and treatment of atelectasis as outlined above, infection control measures (e.g. hand hygiene, avoidance of sick contacts, etc.), compliance with respiratory treatments, education and vaccination.

Vaccination:

Vaccination is a critical role of primary care. Pneumococcal vaccination (with Pneu-23) at the time of injury is recommended. Some experts suggest patients with SCI receive Pneu-13 followed by Pneu-23 eight weeks later, if financially possible (if Pneu-23 given first, do not give Pneu-13 until 1 year later).³⁶

The Canadian Immunization Guide recommends one dose of Tdap if has not received it in adulthood (18 years of age and older) in order to protect against pertussis.³⁶

Annual influenza vaccination is recommended.^{9,10,11,12} It is also recommended that family members and caregivers receive annual influenza vaccinations.

Management:

Management of pneumonia includes prompt recognition, antibiotics, and optimization of secretion mobilization. Emergency department or hospital referral should be considered in individuals at high risk of worsening respiratory function.

Clinical pearl: When determining whether to refer for hospitalization, consider the severity of illness, level of respiratory

compromise, use of assistive breathing equipment/techniques, history of respiratory complications, assistance available at home, the skill of the patient or caregivers with secretion mobilization, the likelihood of compliance with therapy, and the availability and accessibility of follow-up care.

Antibiotics:

Empiric antibiotics should be promptly administered. When choosing an antibiotic, evaluate risk factors for resistant organisms and consider anti-pseudomonal coverage.

Antibiotics for Community Acquired Pneumonia in individuals with Comorbidities (modified from Anti-infective guidelines for community-acquired infections)³⁷

		Antibiotic	Dosage
S. pneumoniae M. pneumoniae C. pneumoniae H. influenzae	FIRST LINE	Amoxicillin	1g TID
		Amoxicillin/Clavulanate	500mg TID or 875mg BID
		Cefprozil	500mg BID
		Cefuroxime-AX	500mg BID
ANY ONE of the beta-lactam agents above PLUS ONE of the following:			
		Azithromycin	500mg daily on first day then 250mg daily x 4 days
		Clarithromycin	500mg BID or 1000mg (extended release once daily)
		Doxycycline	100mg BID first day then 100 mg OD
OR any ONE of the following:			
		Levofloxacin	750mg OD x 5 days
		Moxifloxacin	400mg OD
Polymicrobial Oral anaerobes Gram –ve bacilli	FIRST LINE	Amoxicillin/Clavulanate	500mg TID or 875mg BID
		Clindamycin	300-450mg QID

Table 1: Antibiotic treatment for CAP in SCI

“Comorbidities include hospitalization in the past 3 months, and/or chronic heart, lung, liver or renal disease, diabetes mellitus, alcoholism, malignancies, asplenia and immunosuppression. Anaerobic coverage is indicated in classic aspiration pleuropulmonary syndrome. Consider aspiration pneumonia in patients with difficulties swallowing who show clinical signs of a lower respiratory tract infection.”³⁷

Optimization of Secretion Mobilization:

- Multimodal treatment
- “Quad coughing” (manually assisted coughing); may precede with insufflation.
- Mechanical insufflator-exsufflator (CoughAssist).
- See Table 1

Pulmonary Thromboembolism:

Deep venous thromboembolism and pulmonary embolism are common early complications of SCI, especially in the first two weeks and up to three months, and are major causes of morbidity and mortality.¹³ The risk of venothromboembolic disease remains above the general population for at least one year following injury.¹⁴

The clinical diagnosis of venous thromboembolism is difficult in patients with SCI¹⁵ but signs and symptoms may include (remember due to level of lesion sensation can be altered!):

- Swelling, redness, and pain in the leg
- Shortness of breath, chest pain

- Unusual symptoms such as autonomic dysreflexia (AD) (see *AD CBLM*), unexplained fever, or altered mental state

Investigations:

Like for the general population, D-Dimer has good negative predictive value for DVT. However, D-Dimer has also been found to be elevated in many non-DVT patients with SCI with conditions such as pressure injuries, or infections.¹⁶ When clinical suspicion for DVT/PE is high, further investigation (e.g. Doppler ultrasonography, computed tomographic pulmonary angiography) may be necessary.

Prophylaxis:

Prophylactic use of low-molecular-weight heparin is generally recommended for 8 to 12 weeks after initial SCI provided there is no contraindication. Venous thromboembolism prophylaxis can generally be discontinued after the initial three months following SCI except when otherwise indicated (such as during a hospitalization).¹⁷

Dyspnea:

Dyspnea is a common complaint in patients with chronic SCI and is related to the level of the SCI (prevalence ranging from 21 to 33 percent in tetraplegia, to 2 to 11 percent in lower level injuries).^{18,19} Dyspnea can have a significant impact on a patient's function and quality of life affecting activities such as talking, eating, dressing/undressing, or leaving the house.²⁰

Physical activity may improve dyspnea and should be encouraged in all SCI patients.²¹ Inspiratory muscle training) may also be considered although to date, research has shown it improves pulmonary function, but not breathlessness.²² Referral to a respirologist, respiratory therapist, physiatrist or physiotherapist might be indicated.

Acute worsening of dyspnea should be assessed thoroughly (see *Respiratory Insufficiency*).

Sleep-Disordered Breathing:

The prevalence of obstructive sleep apnea in SCI is high (40-80% in cervical SCI) and obesity (which is common) is a risk factor. The use of muscle relaxants may also contribute. Nocturnal hypoxemia is also common. Monitoring of snoring, apnea, daytime sleepiness, obesity is important. Polysomnography (or overnight oximetry) should be strongly considered in individuals with SCI.²³

Long-Term Ventilation

Due to improved survival rates with high level SCI, more individuals survive requiring long-term ventilation. This requires a comprehensive team to monitor and manage the unique needs. There needs to be planning for back-up equipment and unpredictable events (power failures). The primary care provider is instrumental in helping to coordinate these services. Many centres will have a long-term ventilation clinic. (Berlowitz,

Assistive Devices and Respiratory and Pulmonary Clearance Techniques:

The following devices/techniques will be prescribed/initiated by specialists or allied health professionals. It is helpful for primary care providers to have a general understanding of their use so as to help counsel patients:

Mechanical Ventilation	Typically injuries above C3 will require chronic ventilation. Complete injuries at and below C5 can likely be weaned off ventilator.
Intermittent Positive Pressure Ventilation	Short-term breathing treatment where increased pressures are delivered via ventilator to help treat atelectasis, clear secretions, or deliver aerosolized medications. ²⁴
Electrical Stimulation	Electrical phrenic nerve and diaphragmatic stimulation are used in some individuals with SCI to improve inspiratory function and cough effectiveness. (More information can be found at scireproject.com) ²⁵
Abdominal Binders	Used in some individuals with lesions above T6 to support lack of abdominal muscle tone and keep contents from falling forward, thereby improving diaphragm position and inspiratory ability. ²⁶
Inspiratory Muscle Training	Similar to improving limb muscle strength and endurance, respiratory muscles can be trained using inexpensive, commercially available hand-held devices to help decrease dyspnea and respiratory infections. (More information can be found at scireproject.com). All patients should be taught inspiratory muscle training techniques.
Breath Stacking (Lung Volume Augmentation)	Technique involving the use of an Ambu bag to gently push more air into lungs after inspiration which helps stretch lungs and move secretions ²⁷ (See spinalcordessentials.ca for an excellent patient handout on this technique)
Assisted Cough Technique	Often used with breath stacking, technique that involves an assistant applying pressure below the patient’s rib cage in order to produce a stronger cough ²⁸ (See spinalcordessentials.ca for an excellent patient handout on this technique)
Mechanical Insufflation/Exsufflation (Cough Assist) Device	Device that delivers gradual positive air pressure during inhalation followed by a rapid shift to negative air pressure to help stimulate a deep cough
Chest physiotherapy	Generally includes techniques of manual percussion, vibration well and postural drainage, as well as breath stacking and assisted cough to facilitate airway clearance.

Table 2: Assistive devices and respiratory and pulmonary clearance techniques

Monitoring/Follow-Up Recommendations:²⁹

There are currently no widely accepted clinical practice guidelines for the long-term respiratory management of the patient with SCI. Based on the best available information at this time, we recommend annual assessments of respiratory function for individuals with SCI at T12 and above include:

- History:

- Respiratory complaints (SOB, secretion clearance, aspiration risk)
- Respiratory infection history (frequency, ER/hospitalization, treatment)
- Compliance with respiratory treatments
- Functional change
- Exercise
- Physical examination:
 - Respiratory rate and pattern
 - Continuous pulse oximetry
 - Physical examination of the respiratory system, assessment of edema
- Investigations:
 - ⊖ Spirometry has been recommended annually, if normal consider repeat in 5 years.
 - Polysomnography or nocturnal oximetry testing if indicated
- Referrals:
 - Respiriology should be considered in any patient with:
 - A high level lesion (cervical)
 - Requiring ventilation
 - Difficulty with inspiration/cough
 - History of recurrent infections or hospitalizations
 - Worsening respiratory function
 - Physiotherapy or Respiratory Therapy:
 - Assess, educate and manage respiratory procedures
- Counselling:
 - Smoking cessation
 - Chest physiotherapy/exercises and daily respiratory muscle training in patients with injuries above T6
 - Physical activity is recommended to help improve respiratory function
- Immunizations:
 - Annual influenza vaccination
 - Pneumococcal vaccination at the time of injury
 - Pertussis

Clinical Pearl: All patients with a cervical or thoracic SCI will have some form of respiratory impairment. The higher the level of injury, the more muscles of respiration will be impaired and the more severe the dysfunction will be.

Jordan, age 34

Part 2

Jordan has been feeling progressively more short of breath over the past six months with activities that he was not before and sometimes with lying down. He does not recall a triggering cause such as an illness or trauma. He denies associated chest discomfort, wheezing, tightness, cough or fever. He does feel he is not able to cough as strong or bring up phlegm. He reports no issues with swallowing. He has never smoked. Apart from receiving treatment during his initial hospitalization post SCI, he has not been treated or hospitalized for a pulmonary infection, nor has he had a blood clot. He has no history for any respiratory conditions such as asthma or COPD. He has no recent hospitalizations. He had spirometry prior to discharge from rehab four years ago and he is unsure of the results. He denies snoring, morning headache and daytime somnolence.

Physical examination reveals: HR 80, BP 98/66, RR 20/min, O₂ sat 95% on room air, temperature 36.8 °C. Auscultation reveals a few crackles at the lung bases.

What would be your approach to the investigation of this patient?

- *It would be appropriate to order new spirometry (and compare to previous if available), especially as it has been over 1-2 years since his last pulmonary function assessment. A chest x-ray may also be indicated.*

Part 3

Spirometry reveals a decrease in FVC consistent with moderate restriction but no obstruction. CXR report indicates atelectasis and otherwise normal.

What recommendations would you make to Jordan?

- *A referral to a physiotherapist or respiratory therapist experienced in managing respiratory issues in SCI for assessment and management would be beneficial if able. Breath stacking, assistive coughing maybe procedures that can be useful and taught.*
- *Referral to a respirologist may be considered for assessment and monitoring as he has a high level lesion.*
- *Physical activity should also be encouraged.*
- *Preventative health such as immunizations should definitely be addressed in primary care.*
- *A sleep study should be considered even though he is not having obvious symptoms.*
- *Regular monitoring of pulmonary function via spirometry should be considered.*
- *Since respiratory complications are the leading cause of mortality, especially with higher level complete SCI, Jordan should be monitored closely over time and advised as to signs and symptoms to seek medical attention.*

SUMMARY

- Respiratory complications are common post SCI and are the leading cause of death in spinal cord injured individuals.³⁰
- The most common respiratory complications are atelectasis, pulmonary infections and respiratory failure. Other complications include pulmonary thromboembolism, pulmonary edema, dyspnea, sleep-disordered breathing and dysphonia.
- A downward trend in vital capacity may be sign of impending respiratory failure and should be fully investigated.
- Atelectasis is the most common respiratory complication in patients with SCI and can predispose to respiratory infections.
- Respiratory infections should be treated promptly with antibiotics and optimization of secretion mobilization.

Appendix 1. Respiratory dysfunction based on level of injury (complete lesion)³¹

Level of Injury	Dysfunction
C1-C3	<p>Complete paralysis of all muscles involved with respiration. Vital capacity ~5-10% of normal. Cough absent.</p> <p>Generally, fatal. Immediate and likely permanent ventilatory support needed in order to sustain life.</p>
C3-C4	<p>Diaphragm function impaired reducing tidal volume and vital capacity (~20% of normal). Cough weak and ineffective.</p> <p>Respiratory failure requiring mechanical ventilation common initially following SCI, but eventually, periods of ventilator-free time may be possible as accessory muscles are recruited and strengthened.</p>
C5	<p>Diaphragm function intact but intercostal and abdominal muscle paralysis results in decreased lung volumes, and cough strength and effectiveness.</p> <p>Respiratory failure requiring mechanical ventilation is common initially following SCI, but independent respiration often possible in the long-term.</p>
C6-C8	<p>Independent breathing.</p> <p>People with lesions caudal to C7 typically can augment inspiration and cough with accessory muscles (pectoralis major and minor).</p>
T1-T4	<p>Inspiratory capacity and forced expiration supported by intercostal activity. Vital capacity ~30-50% of normal. Cough efficacy remains reduced secondary to abdominal muscle weakness.</p>
T5-T12	<p>Progressive relative improvement in muscle strength at descending lesion levels.</p>
T12	<p>Respiratory function essentially comparable to that of an able-bodied person</p>

Appendix 2. Indications for polysomnography in patients with neuromuscular disease:³²

Consider referral for polysomnography if any of the following are present:

- Symptoms of sleep disordered breathing
 - Excessive daytime somnolence
 - Disrupted nocturnal sleep
 - Changes in personality, concentration, or memory
 - Snoring
 - Awakening with shortness of breath or choking
 - Morning headache
- Arterial blood gases showing hypoventilation ($\text{PaCO}_2 > 45$ mmg Hg) (how are we getting this?)
- $\text{FVC} < 50$ percent predicted (?)
- Severely reduced PI_{max} (maximal inspiratory pressure at the mouth)
- Unexplained cor pulmonale
- Resistant hypertension

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- ³ <http://eprimarycare.onf.org/RespiratoryComplications.html>
- ⁴ https://www.uptodate.com/contents/respiratory-complications-in-the-adult-patient-with-chronic-spinal-cord-injury?source=search_result&search=respiratory%20complications%20sci&selectedTitle=1~150
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