VII DISORDERS OF THE CHEST WALL

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The chest wall consists of the parietal pleura, rib cage, and muscles. The abdominal contents and abdominal wall function as part of the rib cage in that they influence the resting position and movement of the diaphragm.

The respiratory pump apparatus is composed of the rib cage and its musculature, including the intercostal muscles, diaphragm, and accessory muscles of respiration. Optimal pumping action requires structural integrity and the synchronized contraction of the intercostal muscles and diaphragm. Respiratory pump function may be impaired by mass loading from obesity, skeletal abnormalities, neuromuscular disorders, or restriction of lung movement from pleural disease. Cervical strap or abdominal muscles may be recruited for assistance when respiratory pump function is impaired. Because the diaphragm is the major power generator for the respiratory pump, loss of rib cage function alone may be insufficient to cause ventilatory failure; ankylosis and paralysis of the rib cage are often associated with a normal resting carbon dioxide tension (PCO2).

Respiratory Pump Dysfunction

Respiratory pump dysfunction may vary from trivial to severe. Severe dysfunction restricts lung expansion and may cause dyspnea and hypercapnic ventilatory failure because of the small tidal volumes that increase the proportion of wasted ventilation per breath, despite a compensatory increase in the rate of respiration [see Table 1]. The result is a decrease in alveolar ventilation with hypercapnia and hypoxemia. Hypercapnia is often most severe during sleep because of a decrease in ventilatory drive and a sleep-associated increase in upper airway resistance. Hypercapnia and the associated hypoxia may in turn cause vasoconstrictive pulmonary hypertension and cor pulmonale. Weakness of expiratory muscles from neuromuscular disease may produce ineffective cough and result in recurring atelectasis or infections. Severe respiratory pump dysfunction differs from alveolar and interstitial lung disease, in which ventilation abnormalities result from alterations in the lung parenchyma [see Table 2].

When the respiratory pump is impaired, ventilation will be determined by (1) the efficiency of the inspiratory muscles, (2) the strength of the inspiratory muscles, and (3) the impedance to the pumping action of these muscles. The efficiency of the respiratory muscles is determined by their length and by the resulting mechanical action on the pump apparatus. Shortening of the inspiratory muscles from hyperinflation or chest wall deformity reduces their pumping efficiency. Paralysis of either the chest wall or the diaphragm produces an observable paradoxical movement of the paralyzed component during inspiration; this movement results in inefficiency of the pump apparatus. The strength of the inspiratory muscles may be reduced by neuromuscular disease or metabolic disturbances, such as hypokalemia or hypophosphatemia. Ventilatory ability is proportional to the remaining respiratory muscle strength but may be disproportionately reduced if there are concomitant mechanical problems of the respiratory system. Finally, either increased airway resistance or decreased respiratory system compliance may impede the pumping action of the inspiratory muscles and reduce ventilation. Respiratory system compliance may be reduced by morbid obesity, chest wall deformity, circumferential pleural disease, or parenchymal disease. Patients with a poorly compliant respiratory system must exert more effort than healthy patients to achieve equivalent tidal volumes [see Figure 1], so they take smaller breaths to minimize respiratory muscle fatigue but must compensate by increasing their breathing rate.

Obesity and Its Impact on Respiratory Function

Obesity imposes a restrictive load on the thoracic cage, both directly because weight has been added to the rib cage and indirectly because of the large abdominal panniculus, which impedes the motion of the diaphragm when the person is supine. In addition, obese patients, particularly males, may experience increased respiratory resistance and resultant airflow limitation that may be related to breathing at lower lung volumes, increases in pulmonary blood volume, or both.

Obesity characteristically causes a decrease in functional residual capacity that becomes significant only in the presence of coexisting conditions such as obstructive lung disease, in which airway closure occurs at lower lung volumes, leading to hypoxemia.

Obesity in otherwise healthy patients causes little interference with lung function at rest. Generally, vital capacity and total lung capacity remain normal except in the most severe instances of morbid obesity.

In patients with impaired ventilatory drive, the mechanical work load imposed by obesity may not be countered by in-

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**Table 1** Relations among Tidal Volume, Respiratory Frequency, and Arterial and Alveolar Oxygen Tension

<table>
<thead>
<tr>
<th>Volume (L)</th>
<th>V̇f/V̇t</th>
<th>f (L/min)</th>
<th>V̇a (L/min)</th>
<th>ṖCO_2 (mm Hg)</th>
<th>ṖO_2 (mm Hg)</th>
<th>A-aDO_2 (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal respiratory function</td>
<td>0.50</td>
<td>0.35</td>
<td>0.15</td>
<td>30%</td>
<td>12/min</td>
<td>4.20</td>
</tr>
<tr>
<td>Restrictive disorder</td>
<td>0.25</td>
<td>0.10</td>
<td>0.15</td>
<td>60%</td>
<td>30/min</td>
<td>3.00</td>
</tr>
</tbody>
</table>

Note: This table demonstrates how restrictive disorders of the respiratory system resulting from neuromuscular or chest wall disease may produce hypercapnia and hypoxemia by decreasing the amount of ventilation per breath.

*The elevated ṖCO_2 has resulted in arterial and alveolar hypoxia. However, in the absence of atelectasis or another concomitant disease that would increase the ventilation-perfusion mismatch, the A-aDO_2 remains normal.
A-aDO_2—alveolar-arterial difference in oxygen f—respiratory frequency ṖCO_2—arterial carbon dioxide tension ṖO_2—arterial oxygen tension V̇a—alveolar portion of the tidal volume V̇d—dead space portion of the tidal volume V̇f/V̇t—ratio of functional dead space volume to tidal volume, or the wasted ventilation ratio V̇t—tidal volume

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December 2001 Update
increased respiratory effort. Under such circumstances, chronic daytime hypercapnia may develop, most commonly in the setting of obstructive sleep apnea but also in the absence of sleep-disordered breathing. The pathogenetic role of obesity in obstructive sleep apnea [see 14:VI Ventilatory Control during Wakefulness and Sleep and 11:XIII Disorders of Sleep] may relate in part to fatty encroachment on the upper airways.

Obese patients may experience significant dyspnea during exercise because of the increased work required to move the heavy chest and abdomen and because of overall poor conditioning. The tachypneic shallow breathing pattern during exercise in morbidly obese patients reflects the combined effects of this mass loading and diminished compliance of the respiratory system [see Figure 1].

Weight loss is the most important therapy for patients with respiratory problems related to obesity. For patients with associated sleep disorders of breathing, appropriate treatment of obesity may alleviate hypercapnia [see 14:VI Ventilatory Control during Wakefulness and Sleep and 11:XIII Disorders of Sleep]. Noninvasive ventilation may help decrease the symptoms of daytime hypercapnia.4

**Skeletal Abnormalities That Affect Respiratory Function**

Deformities of the costovertebral skeletal structures may affect compliance of the thoracic cage, its shape and volume, and, ultimately, pulmonary compliance. Effects vary from undetectable to severe.

**KYPHOSCOLIOSIS**

The two basic types of costovertebral skeletal deformity—scoliosis, a lateral curvature with rotation of the vertebral column, and kyphosis, an anterior flexion of the spine—are usually found in combination. Approximately 80% of cases of kyphoscoliosis are idiopathic. Idiopathic kyphoscoliosis commonly begins in late childhood or early adolescence and may progress in severity during these years of rapid skeletal growth. The incidence of kyphoscoliosis in females is four times higher than that in males. The remaining 20% of cases of kyphoscoliosis are found in association with neuromuscular disorders (e.g., syringomyelia, neurofibromatosis, or poliomyelitis), congenital vertebral defects (e.g., hemivertebrae), acquired vertebral abnormalities (e.g., tuberculous spondylitis or osteomalacia), or deforming chest wall processes (e.g., sequelae of empyema or as a result of thoracoplasty).

**Respiratory Compromise**

Of the various chest deformities that produce ventilatory failure, kyphoscoliosis is the most common. The degree to which ventilation is reduced is determined by the severity of deformity and the degree of neuromuscular weakness. A standard technique for measuring the degree of curvature was developed by Cobb in 1948 [see Figure 2]. Mild to moderate deformities (scoliotic angle < 60°) are associated with minimal to mild restrictive ventilatory defects.

Dyspnea may occur during exercise and is most often caused by deconditioning and lack of regular aerobic exercise rather than by any alteration of lung function.2 As the scoliotic curvature worsens, vital capacity and total lung capacity decline significantly, and dyspnea on mild or moderate exertion becomes a common complaint. Kyphoscoliosis may distort the respiratory pump, so that inspiratory power becomes limited even in the absence of a neuromuscular disease, such as poliomyelitis. The severity of hypercapnia is therefore related to both the severity of deformity and the degree of inspiratory muscle weakness.

Severe deformities (scoliotic angle > 100°) can be associated with prominently restricted lung volumes; typically, total lung capacity is reduced to 50% or less of the predicted value. Such restriction may lead to chronic alveolar hypoventilation, hypoxemia, pulmonary hypertension, and, ultimately, cor pulmonale. Long-term follow-up of patients with kyphoscoliosis suggests that those with a vital capacity less than 45% of the predicted value and a scoliotic angle greater than 110° are at the greatest risk for respiratory failure.7 Kyphoscoliosis of such severity may cause compression of underlying lung tissue, thereby elevating the alveolar-arterial difference in oxygen

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December 2001 Update
(A-aDO₂). In most cases of kyphoscoliosis, however, the A-aDO₂ remains at normal or near-normal levels, and significant hypoxemia is present only when hypercapnia develops. These findings contrast with restrictive ventilatory disorders caused by diffuse parenchymal lung disease, in which the A-aDO₂ is characteristically elevated and hypoxemia is often associated with hypocapnia until late in the course of disease.

Treatment

Acute complications Patients with severe kyphoscoliosis may live for many years without succumbing to respiratory insufficiency. Such patients, however, are vulnerable to any respiratory tract infection or central nervous system depressant. Because breathing is chronically restricted, increased central neural output and physical work are required to maintain ventilation; relatively minor insults, such as bacterial or viral bronchitis or pneumonia, may represent an increment in load sufficient to produce frank respiratory failure. In addition, standard doses of narcotics or sedatives may suppress chronically hyperactive control mechanisms to a level sufficient to precipitate acute respiratory failure.

Thus, immunization with influenza and pneumococcal vaccines, early treatment of respiratory tract infections, and strict avoidance of CNS depressants are important in the management of kyphoscoliosis. Episodes of hypercapnic respiratory failure precipitated by reversible conditions respond well to short-term supportive measures, including bronchopulmonary drainage, mechanical ventilatory support, and oxygen supplementation.

Chronic complications Chronic respiratory insufficiency may ensue after several years. Older patients with kyphoscoliosis are at risk for respiratory failure because the angle of curvature typically worsens with age. Although most cases of idiopathic scoliosis stabilize just after puberty, further spinal deformity may result from the osteoporosis, vertebral body weakening, and loss of muscle tone that accompany older age. Surgical procedures to straighten and stabilize the vertebral column usually fail to restore ventilatory capacity. Such procedures are useful early in the course of kyphoscoliosis, when they may prevent progression of the deformity before respiratory compromise develops.

Supportive measures may sustain meaningful life for many years, even in patients with chronic respiratory failure. Many can adapt very well to a state of chronically disordered gas exchange. Chronic hypoxemia accompanied by secondary erythrocytosis, worsening of pulmonary hypertension, and cor pulmonale should be treated with supplemental oxygen administration. Such therapy can be augmented by nocturnal ventilatory support.

Although kyphoscoliosis is not a primary sleep-related breathing disorder, the degree of oxygen desaturation that occurs during sleep is greater than that observed in other types of lung disease, possibly because the baseline hypoxemia and hypercapnia are more severe and lung volumes are smaller.8 Thus, nocturnal oxygen therapy and mechanical ventilatory support during sleep often improve functional status and symptoms. In fact, many patients who achieve normal levels of arterial carbon dioxide tension (P_{a}CO₂) during sleep by means of mechanical ventilators can sustain normal or near-normal arterial blood gas levels throughout the day. Devices used for mechanical ventilation in such patients include the iron lung, specially fitted thoracoabdominal negative-pressure ventilators, and positive-pressure ventilators applied via tracheostomy or nasal mask.9

ANKYLOSING SPONDYLITIS

Ankylosing spondylitis may affect the thoracic cage because of arthritic involvement of the costovertebral articulations[see 15:III Seronegative Spondyloarthropathies]. The chest may become relatively fixed in a hyperexpanded position, leading to an elevated midposition lung volume. The reduced compliance of the chest wall causes moderate restriction of vital capacity and total

Figure 1  When the respiratory pump is encumbered by structural changes such as obesity, kyphoscoliosis, or ankylosing spondylitis (blue line), the work of breathing at rest is higher than it is for a healthy person (black line). The work of breathing required to maintain the tidal volumes needed during exercise may be prohibitive for patients with these disorders, forcing them to adopt a shallow tachypneic breathing pattern in response to an increased ventilatory requirement.

Figure 2  In this radiograph of the spine in a patient with kyphoscoliosis, straight lines are passed through the upper and lower limbs of the curvature. The angle inscribed by these two lines defines the sciotic angle.
lungs. A typical physical finding is limited expansion of the chest wall on inspiration, despite normal findings on auscultation and percussion of the chest and normal muscle strength. Nonetheless, the alteration in function is almost never severe enough to produce symptoms, and this deformity does not produce respiratory failure.

**Deformities of the Sternum**

Deformities of the sternum and costochondral articulations are potentially dramatic in radiographic and physical appearance and may induce psychological problems, but functional consequences are rare. There are two main varieties of deformity: pectus excavatum, an inward concavity of the lower sternum, and pectus carinatum, an outward protuberance of the upper, middle, or lower sternum.

*Respiratory Compromise*

Pectus deformity is present in fewer than 0.5% of the general population and appears to be more common in patients with other evidence of structural or connective tissue disease, such as scoliosis, Marfan syndrome, Poland syndrome, or Pierre Robin syndrome. In these circumstances, ventilatory function impairments may result from the underlying disease rather than the pectus deformity. For example, defects in bronchial cartilage development may lead to repeated pneumonia and result in bronchiectasis.

*Treatment*

In most cases of pectus deformity, no significant functional limitations are caused by the deformity. Lung volumes are preserved, and cardiovascular function is normal. Surgical correction is therefore generally restricted to patients who have severe deformity accompanied by evidence of lung restriction or cardiovascular dysfunction. Although the severity of the pectus deformity may be assessed by determining the ratio of the transverse diameter of the chest to the anteroposterior diameter as measured by computed tomography, it is not clear whether this index predicts improvement in lung function with surgery.13 A few patients with cardiac compression or with lung restriction from pectus excavatum experience functional improvement after surgical repair. Right and left ventricular end-diastolic volumes also may improve as cardiac compression is relieved.12

Surgical correction may result in modest improvements in lung volumes, ventilatory capacity, and exercise capacity in patients with severe pectus deformities but may worsen the condition of patients with good preoperative lung function.13,14

**Flail Chest**

Flail chest is an acute process that may lead to life-threatening abnormalities of gas exchange and mechanical function. Stability of the thoracic cage is necessary for the muscles of inspiration to inflate the lung. In flail chest, a locally compliant portion of the chest wall moves inward as the remainder of the thoracic cage expands during inhalation; the same portion then moves outward during exhalation. Consequently, tidal volume is diminished because the region of lung associated with the chest wall abnormality paradoxically increases its volume during exhalation and defflates during inhalation. The result is progressive hypoxemia and hypercapnia. Multiple rib fractures, particularly when they occur in a parallel vertical orientation, can produce a flail chest. The degree of dysfunction is directly proportional to the volume of lung involved in paradoxical motion. Patient management may be complicated by other manifestations of trauma to the chest, such as splintering of ventilation because of pain, contusion of underlying lung, or hemothorax or pneumothorax. Positive pressure inflation of the lung or negative pressure applied to the chest wall corrects the abnormality until more definitive stabilization procedures can be undertaken.

The pathophysiologic disturbances of flail chest may also result from nonclosure of the wound after median sternotomy is performed. Any dehiscence of the sternal wound will lead to separation, loss of stability, and prominent inward motion during inspiration. The magnitude of the inward motion is directly related to the extent of the sternal separation and to the degree of negativity of inspiratory intrathoracic pressures. This condition is often the cause of difficulty in weaning a patient from mechanical ventilatory support after major cardiac surgery.

Other, rare causes of localized chest wall instability include destruction of the ribs from malignant disease (e.g., multiple myeloma) or from metabolic disorders (e.g., osteitis fibrosa cystica).

**Neuromuscular Disorders That Affect Respiratory Function**

Processes that interfere with the transfer of central neural output to the muscles that expand the rib cage, such as abnormalities of the spinal cord, peripheral nerves, neuromuscular junctions, or muscles, can lead to ventilatory impairment. Whereas central control problems allow creation of adequate inspiratory pressures by voluntary efforts [see 14:VI Ventilatory Control During Wakefulness and Sleep], central neural output abnormalities are characterized by an inability to generate normal respiratory pressures, either automatically or intentionally. Some diseases, such as poliomyelitis, can involve both the central controller and the peripheral neuromuscular apparatus.

*Pathophysiology*

Several factors are common to the neuromuscular disorders of the thoracic cage. The respiratory midposition volume is maintained at near-normal levels, whereas total lung capacity decreases (because of inspiratory muscle weakness) and residual volume increases (because of expiratory muscle weakness). Vital capacity is diminished along with maximal static inspiratory pressure. Because muscle strength and vital capacity can be substantially diminished without causing respiratory failure, the presence of respiratory failure with hypoxemia and hypercapnia indicates either extreme progression of the primary process or the effects of complications such as atelectasis caused by retained secretions from ineffective cough, pneumonia, or pulmonary thromboembolism. Onset of hypoxemia, hypercapnia, or both in the presence of reasonable inspiratory muscle function suggests the presence of a complication rather than progression of the primary process. In the acute setting, the need to distinguish between a complication and progression makes monitoring of maximal static inspiratory pressures (which assesses muscle strength) superior to measuring serial vital capacity because vital capacity may be diminished by either a complication or progression.

Neuromuscular disorders that persist for months are associated with chronic decreases in compliance of both the chest wall and the lungs. It is unknown whether the decreases in lung
compliance are the result of microatelectasis, altered surfactant, or mild fibrotic changes resulting from recurrent infections. Ventilation-perfusion mismatch occurs in the lungs of patients with these disorders and may lead to hypoxemia that is disproportionate to the degree of hypoventilation. Decreases in chest cage compliance have been attributed to gradual stiffening of the costochondral and costovertebral articulations and to fibrotic changes or spasticity of the muscles of the rib cage.

The diminution in lung volume that occurs in chronic neuromuscular disorders is caused by the combined effects of muscle weakness and secondary alterations in the mechanical properties of the lung and chest wall. Hence, for patients with chronic disease, measurement of vital capacity is a more accurate indicator of the total impact of the disorder than is maximal inspiratory pressure. Attempts to improve lung compliance by periodic hyperinflations with intermittent positive pressure breathing have usually not proved successful.

In contrast to the mechanical disorders of the thoracic cage, which preserve an effective cough, expiratory muscle weakness in the neuromuscular disorders prevents generation of sufficient expiratory velocities for a forceful cough. The extreme example is cervical spinal cord injuries in which paralysis of the abdominal and intercostal muscles severely reduces but does not eliminate spontaneous cough. Ineffective or absent cough eliminates a first-line defense against respiratory tract infection and is particularly troublesome when combined with airway mucus hypersecretion, as occurs in asthma or chronic bronchitis. Pneumonia followed by respiratory failure is a common cause of death in patients with neuromuscular syndromes.

Respiratory Compromise

Patients with neuromuscular disorders must be awake to maintain ventilation. During sleep, hypoxemia and hypercapnia develop or worsen and may contribute to complications such as cor pulmonale. The degree of hypoxemia that develops with sleep is related to the severity of the abnormalities in lung mechanics and to the degree of derangement in gas exchange that is present while the patient is awake.15

In the absence of major complications, the patient with neuromuscular involvement is often disproportionately tachypneic in relation to the decrease in tidal volume. The resulting increase in minute ventilation more than offsets the increase in dead space ventilation. Thus, early in the course of the illness, \( P_{\text{CO}_2} \) is often low. The basis for the tachypnea may be microatelectasis, which also accounts for mild arterial hypoxemia. Microatelectasis probably develops because of the patient’s inability to take intermittent deep breaths or sighs, which results in changes in alveolar surface forces. As weakness progresses, tidal volume decreases, dead space ventilation increases, and alveolar hypoventilation with worsening hypoxemia ensues [see Table 1]. The decision whether to treat with mechanical ventilatory support must then be made. Long-term results depend on the nature and prognosis of the neuromuscular process and on the potential success of a specific therapy.

Treatment

Whether the primary disorder is acute (e.g., Guillain-Barré syndrome), intermittent (e.g., myasthenia gravis), progressive (e.g., amyotrophic lateral sclerosis), or chronic (e.g., quadriplegia), onset of a pulmonary complication and the accompanying increase in mechanical load and decrease in gas-exchanging ability may precipitate overt and life-threatening respiratory failure [see Table 3]. Because the patient is unable to produce an effective cough, even minor causes of increased airway secretions, such as a viral tracheobronchitis, may lead to major respiratory compromise. Maintenance of bronchopulmonary drainage and the early treatment of infections are essential for avoidance of complications. Acute episodes precipitated by such complications usually respond well to specific treatment plus supportive measures, including bronchopulmonary drainage and mechanical ventilation.

DIAPHRAGMATIC PARALYSIS

In the absence of respiratory complications, neuromuscular syndromes rarely progress to the point of hypercapnic respiratory failure unless diaphragmatic weakness or paralysis is present. Thus, quadriplegic patients who have a preserved phrenic nerve and diaphragmatic function (e.g., \( C7 \) spinal cord transection) almost never progress to hypercapnic respiratory failure unless a major pulmonary complication supervenes or CNS-depressant drugs are administered. Because diaphragmatic paralysis or paresis uniformly accompanies hypercapnic respiratory failure caused by any of the neuromuscular syndromes, it is not usually considered apart from these disorders. However, because certain forms of diaphragmatic paralysis have distinguishing clinical features, they are best considered as discrete entities.

Bilateral Diaphragmatic Paralysis

Respiratory compromise Bilateral phrenic nerve interruption or injury may result in an isolated partial or complete diaphragmatic paralysis. Causes include cervical and thoracic surgery, cold cardioplegia for cardiac surgery, trauma, multiple sclerosis, and neuralgic amyotrophy.15 Orthopnea may be a prominent symptom. With the patient supine, the hydrostatic force of the abdominal contents pushes the patient’s diaphragm into the thorax. Negative intrapleural pressures generated by the accessory muscles cause the diaphragm to be sucked further into the thorax during inspiration, producing a paradoxical inward motion of the upper abdomen as the thorax expands [see Figure 3]. As a result, mechanical and gas exchange abnormalities similar to those seen in flail chest develop. In the upright position, patients often experience a dramatic increase in vital capacity, improvement in gas exchange, and alleviation of symptoms because the weight of the abdominal contents offsets the negative intrapleural pressures and, therefore, the diaphragm no longer ascends with inspiration.

Table 3  Neuromuscular Syndromes Associated with Respiratory Failure

<table>
<thead>
<tr>
<th>Site of Lesion</th>
<th>Disorder</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spinal cord</td>
<td>Quadriplegia</td>
</tr>
<tr>
<td></td>
<td>Amyotrophic lateral sclerosis</td>
</tr>
<tr>
<td></td>
<td>Poliomyelitis</td>
</tr>
<tr>
<td></td>
<td>Spinal muscular atrophies</td>
</tr>
<tr>
<td>Peripheral nerves</td>
<td>Guillain-Barré syndrome</td>
</tr>
<tr>
<td></td>
<td>Diptheritic neuropathy</td>
</tr>
<tr>
<td>Neuromuscular junctions</td>
<td>Myasthenia gravis</td>
</tr>
<tr>
<td></td>
<td>Eaton-Lambert syndrome</td>
</tr>
<tr>
<td></td>
<td>Botulism</td>
</tr>
<tr>
<td></td>
<td>Drug-induced weakness</td>
</tr>
<tr>
<td>Muscles</td>
<td>Muscular dystrophies (e.g., Duchenne dystrophy, myotonic dystrophy)</td>
</tr>
</tbody>
</table>

[December 2001 Update] © 2001 WebMD Inc. All rights reserved.
Treatment Sleeping in an upright position or nocturnal use of a thoracoabdominal cuirass negative-pressure device can facilitate breathing during sleep. In a limited study, bilateral plication of the diaphragm resulted in improved lung function and allowed patients to sleep in a supine position. The bilateral diaphragmatic pacemaker, although expensive and time consuming to place and stabilize, is an alternative for selected patients.

Unilateral Diaphragmatic Paralysis

Unilateral diaphragmatic paralysis is most often detected as an asymptomatic radiographic finding. The paralyzed hemidiaphragm maintains its normal contour but is displaced cephalad. On fluoroscopy, the paralyzed hemidiaphragm may descend slightly on normal inspiration, mimicking normal contraction. However, with a sudden forced inspiration (so-called sniff test), the paralyzed portion of the diaphragm ascends further into the thorax, opposite to the direction of the normally functioning side. It is driven cephalad by the sudden increase in intra-abdominal pressure and the sudden fall in intrathoracic pressure occasioned by the sniff. Thus, a fluoroscopic sniff test helps confirm the diagnosis of unilateral diaphragmatic paralysis.

Most cases of unilateral diaphragmatic paralysis are the result of neoplastic invasion of the phrenic nerve. Compression or destruction of the phrenic nerve by surgery, trauma, or enlarging lymph nodes or aneurysmal vessels may also cause the condition. Idiopathic cases, which may stem from an isolated phrenic neuropathy or acute infectious neuritis, appear to be evenly divided between the right and left sides and are usually permanent. Reversible paralysis is a rare complication of acute pneumonia and more commonly follows cardiac surgery in which the phrenic nerve is transiently injured by the ice slurry used to achieve cardioplegia. A thorough history and a CT scan of the thorax usually suffice for the workup of patients with unilateral diaphragmatic paralysis.

Respiratory compromise Vital capacity and total lung capacity are reduced 15% to 20% below normal levels with the patient upright and are reduced significantly more with the patient supine. Nonetheless, in the absence of associated pleuropulmonary disease, most adult patients with unilateral diaphragmatic paralysis but without a coexisting pulmonary disease remain asymptomatic.

Treatment Infants are more dependent than adults on bilateral diaphragmatic function for adequate respiratory pump function because their more deformable chest wall moves inward with inspiration along with the paralyzed hemidiaphragm. Plication of the diaphragm may be necessary to prevent flail motion in infants with ventilatory failure associated with unilateral diaphragm paralysis. Plication may also improve the lung function of symptomatic adults with unilateral diaphragmatic paralysis.

Postoperative Diaphragmatic Dysfunction

A transient form of diaphragmatic dysfunction occurs after upper abdominal surgery. In the hours after surgery, lung volumes and maximal inspiratory pressures fall and A-aDO2 increases. At the same time, tidal volume decreases and respiratory frequency increases. These changes may be caused by irritation of the diaphragm that produces reflex inhibition of phrenic nerve function. As a result of postoperative diaphragmatic dysfunction, atelectasis and hypoxemia occur. Deep breathing that focuses on inspiratory effort, possibly facilitated by incentive spirometry or by direct encouragement, may alleviate these abnormalities.

Spinal Cord Syndromes

Functional Transection

Functional transection, most often caused by trauma from motor vehicle or diving accidents, must be at or below the level of the C4 cervical nerve segment if the patient is to survive. If transection occurs above this level, the diaphragm ceases to function and breathing stops. For the quadriplegic patient, breathing is maintained solely or predominantly by the diaphragm.

Respiratory compromise Several aspects of breathing are affected when respiration must depend solely on diaphragmatic activity. Because the diaphragm is active only during inspiration, cough—which requires activity by expiratory muscles, including those of the abdominal wall—is almost totally absent. Intercostal muscles are required to stabilize the upper rib cage against inward collapse when negative intrathoracic pressures are produced by descent of the diaphragm. Thus, with diaphragmatic breathing alone, there is a paradoxical inward motion of the upper thorax during inspiration [see Figure 3]. The result is a diminished tidal volume for any level of diaphragmatic activation. Any sparing of the lower cervical segments allows some activity by accessory muscles and diminishes paradoxical motion. Paradoxical motion also diminishes as the condition becomes chronic because of stiffening of the thorax.

Treatment When the quadriplegic patient is upright, the weight of the abdominal contents pulls on the diaphragm, and because abdominal muscle tone has been lost and cannot provide restraining action, diaphragmatic shortening ensues. Thus, the diaphragm is less effective with the patient in this position, and platypnea may result. Abdominal binders serve to replace lost abdominal muscle tone and should be used whenever tidal volume falls with the patient upright. An inflatable anterior air bladder in the binder may be used to assist ventilation in patients with marginal respiratory function. External abdominal compression may be used to help these patients cough. Quadriplegic patients have a mild degree of bronchial hyperresponsiveness caused by parasympathetic tone from the uninjured vagus nerve that is unopposed by sympathetic tone from the spinal cord. Use of an anticholinergic bronchodilating agent or a beta2-adrenergic agonist will reverse this abnormality. Inspiratory muscle training with a resistive device can improve lung function and reduce dyspnea in these patients.

Respiratory failure almost never occurs in quadriplegic patients in the absence of a complication such as pneumonia, atelectasis, septicemia, or pulmonary thromboemboli; therefore, mechanical ventilation is required only during such complications.

Anterior Horn Cell Disease

Amyotrophic lateral sclerosis Anterior horn cell disease is most commonly seen in patients with amyotrophic lateral sclerosis [see 11:IV Cerebrovascular Disorders]. Although the disease causes weakness of the muscles of breathing and an associated restrictive abnormality in most patients, only a minority of patients have respiratory symptoms at presentation. Respiratory
failure may not be seen until an episode of bronchitis or aspiration pneumonia produces an acute event. Supportive measures administered during the acute event often return the patient to a stable condition. In most patients, however, respiratory failure caused by diaphragmatic involvement proves fatal within 3 to 4 years of the onset of symptoms.

Poliomyelitis Sporadic cases of poliomyelitis still occur in the United States. When anterior horn cells involving innervation of the diaphragm are affected, the patient may require ventilatory support, either continuously or during acute pulmonary events.

Spinal muscular atrophy Spinal muscular atrophy represents a heterogeneous collection of heredofamilial disorders that primarily involve spinal motor neurons. Patients with the slowly progressive infantile form, Werdnig-Hoffmann disease, may survive until late childhood. With the onset of a rapid growth phase during puberty, kyphoscoliosis develops and complicates the muscle weakness associated with these disorders.

DISORDERS OF THE PERIPHERAL NERVES

Guillain-Barré Syndrome
Respiratory insufficiency that requires artificial ventilation develops in 20% to 25% of patients with Guillain-Barré syndrome23 [see 11:II Diseases of the Peripheral Nervous System]. Duration of ventilatory support averages 2 months, but periods of up to 30 months have been reported.23 Approximately 30% of patients, however, can be extubated within 2 weeks. Mortality is less than 5%; the majority of survivors recover completely. A small minority may have persistent weakness and may be susceptible to recurring episodes of respiratory failure in association with respiratory infections. The chronic form of demyelinating polyneuropathy may account for some of these persistent abnormalities [see 11:II Diseases of the Peripheral Nervous System].

Toxin-Induced Neural Dysfunction
Peripheral nerve function may also be impaired by toxins from algae or plankton. Certain fish (grouper and snapper) or shellfish consume and concentrate toxins that may in turn be consumed by humans. Although many of the shellfish-related toxins have been associated with respiratory failure, mortality has been highest in connection with the ingestion of puffer fish, a delicacy in Japan.

Diphtheria
Cardiac and neural toxicity occurs in approximately 20% of cases of diphtheria [see 7:IV Infections Due to Gram-Positive Bacilli]. Clinical features and management of the neurologic syndrome are similar to those of Guillain-Barré syndrome.

DISORDERS OF NEUROMUSCULAR TRANSMISSION

Myasthenia Gravis
Myasthenia gravis is the most common disorder affecting neuromuscular transmission. Respiratory failure may occur during myasthenic crisis (acute episode of the basic disease process) or cholinergic crisis (increased weakness caused by an excess of anticholinergic medication), or it may occasionally follow initiation of glucocorticoid therapy. In patients with long-standing myasthenia, myopathy with severe diaphragmatic paresis may develop and lead to chronic respiratory failure. The principles of respiratory monitoring and ventilatory support for myasthenia are similar to those for the other neuromuscular syndromes.24

Myasthenic Syndrome
The myasthenic (Eaton-Lambert) syndrome may be confused with myasthenia gravis. As with other paraneoplastic syndromes, there may be coexistent cerebellar ataxia or carcinomatous neuropathy. The prognosis is not good, and respiratory failure may occur as a terminal event.

Drug-Related Muscle Paralysis
Paralysis lasting several hours can occur after a single dose of succinylcholine in patients with reduced functional pseudocholinesterase, as seen in those with severe liver disease, myxedema, or malnutrition; pregnant patients; and patients with a genetic deficiency. A syndrome of prolonged muscle

Figure 3 In a supine patient with bilateral diaphragmatic paralysis (top), at end expiration the weight of the abdominal contents forces the paralyzed diaphragm cephalad. With inspiration, only the accessory muscles contract, elevating the anterior chest wall, creating a negative intrathoracic pressure, and forcing the diaphragm further cephalad. The result is a paradoxical inward motion of the abdominal wall with inspiration. The diaphragm of a supine patient with quadriplegia caused by a spinal cord injury at or below the level of C4 (bottom) contracts and descends normally on inspiration, causing the anterior abdominal wall to protrude. However, because the paralyzed intercostal muscles fail to stabilize the rib cage, the anterior chest wall is pulled paradoxically inward during inspiration by the negative intrathoracic pressure generated by the diaphragm.

failure may not be seen until an episode of bronchitis or aspiration pneumonia produces an acute event.

Supportive measures administered during the acute event often return the patient to a stable condition. In most patients, however, respiratory failure caused by diaphragmatic involvement proves fatal within 3 to 4 years of the onset of symptoms.

Poliomyelitis Sporadic cases of poliomyelitis still occur in the United States. When anterior horn cells involving innervation of the diaphragm are affected, the patient may require ventilatory support, either continuously or during acute pulmonary events.

Spinal muscular atrophy Spinal muscular atrophy represents a heterogeneous collection of heredofamilial disorders that primarily involve spinal motor neurons. Patients with the slowly progressive infantile form, Werdnig-Hoffmann disease, may survive until late childhood. With the onset of a rapid growth
paralysis or weakness can develop after the extended administration of postsynaptic paralytic agents to facilitate mechanical ventilation, especially in patients with renal failure.23 A number of other commonly used drugs can interfere with neuromuscular transmission. The mechanism can involve anesthetic-like action at the presynaptic level (e.g., with clindamycin or propranolol), postsynaptic curarlike action (e.g., with lincomycin, polymyxin B sulfate, chloroquine, or procainamide), or stabilization of postsynaptic membranes (e.g., with gentamicin, streptomycin, or neomycin).24 Delayed recovery from anesthesia and difficulty in withdrawing a patient from ventilatory support should suggest a possible drug effect. Patients affected by these drugs often have mild or latent myasthenia; occasionally, concurrent electrolyte disturbances, such as hypokalemia, hypocalcemia, or hypomagnesemia, combine with drug effects to produce contributory muscle weakness in myasthenic patients. In severe cases, especially those involving antibiotic-induced postoperative muscle weakness, respiratory failure can ensue; ventilatory support, correction of associated electrolyte disturbances, and withdrawal of the drug usually lead to recovery. Botulism is an uncommon condition caused by ingestion of a polypeptide toxin produced by Clostridium botulinum [see 7:V Anaerobic Infections]. In addition to specific therapy, careful monitoring and provision of ventilatory support in the event of respiratory failure are required.

DISORDERS OF THE MUSCLES

Patients with pseudohypertrophic (Duchenne) dystrophy, myotonic dystrophy, and other forms of muscular dystrophy are predisposed to pulmonary complications, and respiratory failure is a frequent cause of death.27 Also, a specific abnormality of the diaphragm thought to be myopathic can occur in patients with systemic lupus erythematosus.28 Chronic alveolar hypoventilation caused by inspiratory muscle weakness may develop late in the course of a disease. Expiratory muscle weakness impairs cough in some patients, and accompanying weakness of the muscles of deglutition often leads to aspiration of food, which may precipitate acute deterioration. Chronic alveolar hypoventilation may also develop in patients with adequate muscle strength, which suggests that their disease may involve a defect in central control mechanisms. As with all neuromuscular syndromes, CNS-depressant drugs should be avoided whenever possible or given in minimal doses when necessary. Nocturnal ventilation with noninvasive techniques, such as nasal intermittent positive pressure ventilation or external negative pressure ventilation, may be useful in the later stages of these diseases.29

The authors have no commercial relationships with manufacturers of products or providers of services discussed in this subsection.

Acknowledgment

Figure 1   Janet Brielts.