Respiratory Dysfunction and Management in Spinal Cord Injury

Robert Brown MD, Anthony F DiMarco MD, Jeannette D Hoit PhD CCC-SLP,
and Eric Garshick MD MOH

Respiratory dysfunction is a major cause of morbidity and mortality in spinal cord injury (SCI), which causes impairment of respiratory muscles, reduced vital capacity, ineffective cough, reduction in lung and chest wall compliance, and excess oxygen cost of breathing due to distortion of the respiratory system. Severely affected individuals may require assisted ventilation, which can cause problems with speech production. Appropriate candidates can sometimes be liberated from mechanical ventilation by phrenic-nerve pacing and pacing of the external intercostal muscles. Partial recovery of respiratory-muscle performance occurs spontaneously. The eventual vital capacity depends on the extent of spontaneous recovery, years since injury, smoking, a history of chest injury or surgery, and maximum inspiratory pressure. Also, respiratory-muscle training and abdominal binders improve performance of the respiratory muscles. For patients on long-term ventilation, speech production is difficult. Often, practitioners are reluctant to deflate the tracheostomy tube cuff to allow speech production. Yet cuff-deflation can be done safely. Standard ventilator settings produce poor speech quality. Recent studies demonstrated vast improvement with long inspiratory time and positive end-expiratory pressure. Abdominal binders improve speech quality in patients

Robert Brown MD is affiliated with the Pulmonary and Critical Care Unit, Department of Medicine, Massachusetts General Hospital, and with Harvard Medical School, Boston, Massachusetts. Anthony DiMarco MD is affiliated with Rammelkamp Research Center, MetroHealth Medical Center, and with the Department of Physiology and Biophysics, Case Western Reserve University, Cleveland, Ohio. Jeannette D Hoit PhD CCC-SLP is affiliated with the Department of Speech, Language, and Hearing Sciences, University of Arizona, Tucson, Arizona. Eric Garshick MD MOH is affiliated with the Pulmonary and Critical Care Medicine Section, Medical Service, Veterans Affairs Boston Healthcare System, and with Channing Laboratory, Department of Medicine, Brigham and Women's Hospital, and with Harvard Medical School, Boston, Massachusetts.

with phrenic-nerve pacers. Recent data show that the level and completeness of injury and older age at the time of injury may not be related directly to mortality in SCI, which suggests that the care of SCI has improved. The data indicate that independent predictors of all-cause mortality include diabetes mellitus, heart disease, cigarette smoking, and percent-of-predicted forced expiratory volume in the first second. An important clinical problem in SCI is weak cough, which causes retention of secretions during infections. Methods for secretion clearance include chest physical therapy, spontaneous cough, suctioning, cough assistance by forced compression of the abdomen ("quad cough"), and mechanical insufflation-exsufflation. Recently described but not yet available for general use is activation of the abdominal muscles via an epidural electrode placed at spinal cord level T9-L1. *Key words: spinal cord injury, respiratory failure, diaphragm, rib cage, airway clearance.*
[Respir Care 2006;51(8):853–868. © 2006 Daedalus Enterprises]

Introduction

It is estimated that in the United States each year there are about 11,000 new cases of spinal cord injury (SCI)[1] and that there are currently about 250,000 persons alive with SCI. Because of improvements in medical care and survival, the prevalence of people living with SCI has increased, and it is predicted that there will be greater and greater numbers of older patients with SCI. Currently the average age at injury is 37.6 years, and about 80% of those affected are male. The racial distribution appears to be changing. Between 1973 and 1979, 76.9% were white and 14.2% were African American. Since 2000, 62.9% of those injured have been white and 22% have been African American. The cause of this apparent trend is unclear, but it may be due to actual race-specific incidence rates rather than changing location of centers collecting data or changing referral patterns to those centers.[1]

The American Spinal Injury Association has promulgated standards (revised in 2002) for the classification of level of injury and extent of impairment in SCI.[2] In tetraplegia there is injury to one of the 8 cervical segments of the spinal cord, whereas with paraplegia the lesions involve thoracic, lumbar, or sacral regions of the cord. The most caudal segment of the spinal cord with normal motor function defines the motor level of injury. In the thoracic region, where there are no key muscles to test, sensory level is used to estimate the extent of motor impairment. The higher and more complete the motor level of injury, the greater the respiratory muscle impairment.

Respiratory dysfunction and related diseases, such as pneumonia, which can be complicated by septicemia or pulmonary emboli, are common causes of death in SCI. From the point of view of respiratory dysfunction, it is instructive to consider SCI in 2 phases: (1) the initial phase immediately following the injury and the year thereafter and (2) the later, chronic phase during the rest of the life of the affected individual.

Physiologic Effects of Spinal-Cord Injury on the Respiratory System

Injuries above the level of the phrenic motoneurons (C3, 4, and 5) cause virtually complete paralysis of both the muscles of inhalation and exhalation and dependence on mechanical ventilation or phrenic-nerve stimulation. At lower levels of injury, the prospect of breathing without mechanical assistance is improved. Scanlon et al[3] observed that within a month of injury there appears to occur a reduction in lung compliance that does not change during the year thereafter. This observation casts doubt on the previously held notion that reduced lung compliance in SCI is due to chronic lung injury from repeated infections.[4–10] The cause of the early reduction in lung compliance is unclear but has been ascribed partially to reduced lung volume and partially to changes in the mechanical properties of the lung from alterations in surfactant, which can occur rapidly with ventilation at low lung volume.[3,9,11–13] Chest-wall compliance is also probably reduced in tetraplegia.[3,14] This issue is complex because the abdominal compartment of the chest wall is highly compliant in SCI, but the rib cage compartment may be stiff because of muscle spasticity or abnormalities that develop in rib articulations with the spine and sternum, due to poor inspiratory-muscle performance that prevents stretching to the predicted total lung capacity.[15–17]

In tetraplegia, distortion of the respiratory system causes inefficient ventilation (ie, energy cost that is high for the ventilation achieved).[18–22] This inefficiency contributes to the risk of respiratory muscle fatigue, in particular when loads are imposed on the muscles, as with pneumonia or airways obstruction. During spontaneous breathing, distortion occurs such that the upper anterior rib cage moves inward during inhalation.[18,21,23] This is caused by the lack of activity in the external intercostal muscles when the diaphragm contracts and by the high compliance of the abdominal wall, which diminishes the extent to which the diaphragm (acting through the zone of apposition and the

insertion of the diaphragm to the rib cage) can contribute to rib-cage expansion (see Appendix).[18]

During inspiratory resistive-loaded breathing in the able-bodied, the lower transverse dimension of the rib cage increases and functional residual capacity (FRC) decreases because of active expiration below the relaxed volume of the respiratory system. Since the optimal length of the diaphragm for tension development is below FRC, and since contractions at the optimal length of the diaphragm for tension development are most efficient,[24–26] the active expiration increases diaphragm length to one that is advantageous for tension development and efficiency. During inspiratory resistive-loaded inhalation in tetraplegia, there occurs a decrease in the lower transverse dimension of the rib cage.[27] This distortion has a related oxygen cost but does not contribute to minute ventilation, and thus causes inefficiency. In addition, there are 2 reasons for inefficiency of diaphragm performance during resistive-loaded breathing in tetraplegia. Unlike in the able-bodied, FRC does not decrease in tetraplegia[27] so the diaphragm does not operate at or near the optimal length of the diaphragm for tension development. Also, because of the highly compliant abdomen, the diaphragm shortens much more during inhalation in tetraplegia than in the able-bodied, which causes the diaphragm to operate over a disadvantageous length. It appears that the distortion is less severe in chronic than in acute SCI, but there have not been studies to compare the efficiency of ventilation in those 2 groups.

Because of the changes in compliance, chest-wall distortion, and impairment in both muscles of inhalation and exhalation in neurologically complete cervical SCI below C2, there occurs a reduction in vital capacity (VC) to 20–50% of predicted, inefficiency in ventilation, and markedly impaired cough. For those who require long-term assisted ventilation there is the additional problem of speech production.

Partial recovery of respiratory-muscle performance may occur over the year following injury. For example, Bluechardt et al[28] found improvement with time in forced expiratory volume in the first second (FEV_1) and FVC in 12 subjects with motor complete injuries (8 cervical, one C7-T1, and 3 thoracic). They did not measure the role of specific respiratory muscles or of changes in the compliance of the components of the respiratory system in the observed improvement. Between 90 days and 210 days, FEV_1 improved from 1.82 L to 2.54 L, and FVC increased from 2.03 L to 2.69 L. Such progress has been attributed to improvement in diaphragm performance,[28–31] reflex activity in the intercostal muscles,[32] and enhanced performance of the accessory muscles of the neck.[33] In that regard, Axen et al[34] followed 36 tetraplegic subjects for about 10 months following injury. VC improved from 45% of predicted to 58% of predicted, with the greatest rate of improvement occurring during the first 3 months. Corresponding to the improvement in VC was improvement in shoulder and upper-arm muscles, with some segmental innervation in common with the diaphragm (C3-C5). The data suggested that the spontaneous improvement in VC was mediated, at least in part, by corresponding improvement in the neural supply to the diaphragm. In a related study, Brown et al[35] made measurements of total lung capacity and its subdivisions and of respiratory-muscle performance directly (including transdiaphragmatic pressure) in 5 neurologically complete acute tetraplegics within 47 days of injury and during the year thereafter. Increases occurred in mean inspiratory capacity (1.84 L to 2.71 L) and expiratory reserve volume (0.11 L to 0.27 L). Expiratory and inspiratory muscle performance (including that of the diaphragm) also improved with time. The improvement in transdiaphragmatic pressure was consistent with the conclusion of Axen et al,[34] that after acute cervical SCI there occurs some spontaneous recovery of diaphragm innervation.

With regard to pulmonary function (such as FEV_1 and FVC), the eventual outcome depends on the level and completeness of the injury, the extent of the spontaneous recovery noted above, and other factors. The American Thoracic Society (ATS) has suggested testing standards for the able-bodied.[36] These standards require subjects to reproducibly inhale to total lung capacity and expire forcefully, completely, and without hesitation for at least 6 seconds.

To determine whether individuals with SCI could meet these standards, 278 individuals with chronic SCI were tested at least 1 year after injury.[37] It appeared that expiratory muscle weakness caused a relatively long transition time between maximum inhalation and maximum exhalation. Participants with the weakest respiratory muscles, who had the highest and more complete injuries, were unable to successfully meet ATS testing standards. Using 1994 ATS testing criteria,[36] 83% (230/278) had at least 3 acceptable expiratory efforts, and in 78% (217/278) FVC and FEV_1 met reproducibility standards (ie, the 2 highest values were within 200 mL of each other). In the 48 subjects who did not meet acceptability standards, the percent-of-predicted FVC, FEV_1, peak expiratory pressure, and peak inspiratory pressure were lower, and a greater proportion had complete cervical injuries.

The investigators examined the nature of the delay at the beginning of a forced expiratory maneuver in these subjects. The delay (referred to as excessive back-extrapolated volume by ATS[36]) was just slightly more than that observed in the able-bodied (7.4% of the FVC, versus ≤ 5% in the able-bodied), the expiratory efforts were otherwise technically acceptable, and the testing was reproducible. Individuals unable to sustain an expiratory effort for 6 seconds were able to achieve a flow plateau of at

least 0.5 seconds at residual volume. Only 65% of individuals with cervical motor complete SCI were able to produce spirometry values that were acceptable and reproducible by the ATS standards intended for the able-bodied. That value increased to 88% if the ATS standards were modified for SCI to allow excessive back-extrapolated volume and an effort less than 6 seconds if there was a plateau of at least 0.5 second at residual volume. Based on the data from the study, it was recommended that laboratories that test individuals with SCI should adopt the modified testing standards for persons with SCI, since the data obtained thereby are highly reproducible and since otherwise it might be considered erroneously that these persons cannot be tested reliably.

Factors other than level and completeness of injury and extent of spontaneous recovery may influence FVC and FEV_1 in SCI. Early assessments of pulmonary function in SCI included relatively few subjects and focused mainly on the relationship between level and completeness of injury and the resultant reduction in pulmonary function.[38–41] Studies that included fewer subjects whose smoking history was unknown and who often were from rehabilitation centers reported mean FEV_1 and FVC values of approximately 40–50% of predicted in complete cervical injury.[40,42,43]

More recently, investigators have begun to address factors in addition to SCI level in 3 cross-sectional studies of larger cohorts that included persons tested years after SCI, while otherwise healthy. These studies include 216 persons tested at the Bronx Veterans Affairs hospital,[44] 239 persons tested in Los Angeles,[45] and a cohort of 339 veterans and nonveterans tested at the Boston Veterans Affairs Medical Center.[46] In the latter study, after adjusting for age and stature, lower FEV_1 values were related to increasing years since injury, lifetime cigarette smoking (pack years), a history of chest injury or operation, a history of physician-diagnosed asthma, self-report of wheeze (using a respiratory symptom questionnaire), and maximum inspiratory pressure. In the study from the Bronx, smoking resulted in a reduction in FEV_1/FVC in both tetraplegics and paraplegics. In the Los Angeles cohort the effect of smoking was variable, but, as in the Boston cohort, decrements in FEV_1 were associated with greater years since injury. In the Boston study, the adjusted mean FEV_1 and FVC in C4-C5 complete motor SCI were 56% and 53% of predicted values, respectively, and, in C6-C8 complete motor SCI the values were 65% and 64% of predicted, respectively. The Bronx cohort had a mean FVC of 59% in nonsmoking tetraplegics. A mean FVC of 60% was observed in the Los Angeles SCI cohort. These more recent studies provide evidence that pulmonary function attributable to SCI level and extent of injury is better than recognized previously. When pulmonary function is less,

it is likely that factors other than the injury itself are responsible.

Mortality in Chronic Spinal-Cord Injury

Following SCI, the mortality rate is higher than in the able-bodied,[47–54] and the most common causes of death are related to respiratory illnesses.[1,48,50,51,53,54] Impaired muscles of inhalation prevent deep breaths, and some patients with cervical-level SCI may not sigh,[55] even to the extent that they can, leading to atelectasis and related gas-exchange and lung-compliance abnormalities. Dysfunctional muscles of exhalation cause impaired cough and secretion clearance (with associated atelectasis), increase in airways resistance, and persistence of infection when it occurs.

Retrospective assessments have indicated a correlation between excess mortality and neurological level of injury, older age at injury, and injury in earlier calendar years.[1,47–49,53,54] The latter has been thought to reflect recent improvement in medical care of SCI. A limitation of the earlier retrospective studies is that, other than those factors noted above, it was difficult to assess additional risk factors for mortality.

New information is available from a recent prospective study of the role of both respiratory and nonrespiratory factors on mortality in 361 men with chronic SCI.[56] Thirty-seven deaths occurred over a median of 55.6 months. After adjusting for age, the data identified potentially treatable factors related to SCI mortality. Independent predictors of all-cause mortality were diabetes mellitus, a history of heart disease, current or recent (within 7 years) cigarette smoking, and percent-of-predicted FEV_1 at entry into the study. Unlike in prior retrospective studies, level and completeness of SCI, older age at injury, and injury in earlier calendar years were not directly related to mortality. This suggests that care of those with high-level SCI has improved in recent decades. However, it is possible that these factors will emerge as significant as the cohort is followed longer and more deaths occur. The results indicate that, as in the able-bodied, as persons with SCI survive longer, comorbid diseases and personal behavior, such as smoking, are increasingly important in determining mortality. Compared to the United States rates, overall mortality was 47% greater. When both the underlying and contributing causes of death were considered, diseases of the respiratory and circulatory systems contributed to 24.3% and 40.5% of the deaths, respectively. Whether interventions such as smoking cessation, treatment of cardiovascular disease and diabetes mellitus and improvement in FEV_1 with respiratory-muscle training (see below) affect mortality in SCI has not been established.

Dyspnea

It is often assumed that the prevalence of breathlessness due to a specific task is greater at higher levels of SCI. However, this may not be the case, because the perception of breathlessness may be abnormal in tetraplegia. For example, in tetraplegia, steady-state hypercapnia produces less breathlessness than in able-bodied controls.[57]

There are now limited data available on the prevalence and predictors of breathlessness in neurologically complete SCI. In one study,[58] those with cervical and high thoracic SCI reported breathlessness with "activity" more frequently than those with low paraplegia (45% vs 25%, respectively). Yet, in that study, the need to "stop to catch their breath" was similar among those with cervical, high, and low paraplegia, which makes the impact of injury level on breathlessness unclear. Also, with the exception of a 12% prevalence of dyspnea when the subjects were dressing, the data obtained were not task-specific, so specific activities that cause shortness of breath were not evaluated. The issue is complex to study, because, in incomplete SCI, motor function may range from nearly normal (many such persons can walk without much difficulty) to nearly neurologically complete (with almost no function below the level of the lesion).

Ayas et al[59] provided additional information related to breathlessness, specifically during activities related to moving about. The subjects analyzed had neurologically complete SCI and, to get around, used hand-driven wheelchairs > 50% of the time. In that study, the prevalence of breathlessness was greater when the level of injury was higher. Obesity, cigarette-smoking status (current, former, never), age, and years since SCI were not factors related to breathlessness while moving about, so the finding was attributable only to SCI level.

Further information about breathlessness in SCI was provided by Grandas et al,[60] who studied dyspnea during activities of daily living in SCI, categorized by level of mobility. The activities of daily living included talking, eating, dressing or undressing, and leaving the house. Those who needed a motorized wheelchair to get around and who were thereby deemed to be most neurologically impaired were compared to others who used a hand-propelled wheelchair, walked with an aid such as a crutch or cane, or walked without assistance. Overall, dyspnea during activities of daily living was most common, among motorized-wheelchair users (23.5%). In them, 17.7% noted dyspnea while talking for more than a few minutes. In the others, dyspnea while talking was uncommon, and the prevalence of dyspnea varied with activity (range 3.4–14.3%). They more commonly experienced dyspnea while dressing or undressing or dyspnea limiting their ability to leave the house. Percent-of-predicted FEV$_1$ did not predict dyspnea in that study, so the higher prevalence of dyspnea in the motorized-wheelchair users was not due to worse pulmonary function.

The mechanisms for the increased breathlessness with a higher level of injury are not clear. In the able-bodied, for a given exercise task, those with the least exercise capacity have the greatest breathlessness. This may explain the effect of level of injury on breathlessness in SCI, since, in those with higher-level injury there is decreased exercise capacity, which is limited by muscle paralysis, decreased stroke volume, and chronotropic response to exercise, due to loss of supraspinal control over the sympathetic nervous system, and diminished cardiac preload from low plasma volume and peripheral venous pooling.[59,61–64] Afferent information from inflation of the lungs relieves breathlessness, even when mechanoreceptor activity from the chest wall is absent, such as in compete tetraplegia.[65] During exercise, tetraplegics have smaller tidal volume (V$_T$) than those with lower levels of injury.[66] The diminished lung inflation may thereby contribute to the increased breathlessness in tetraplegia. The sensation of breathlessness is also affected by mechanoreceptor information from the chest wall. For example, in heart-lung transplantation, although vagus-nerve innervation of the lung is interrupted, breathlessness from hypercapnia is reduced when V$_T$ is increased. This relief must be due to mechanoreceptor information from chest wall muscles, joints, and tendons. In neurologically complete tetraplegia, perception of such mechanoreceptor information is absent, so breathlessness may be greater.

In motorized-wheelchair users, the relatively high rate of breathlessness during talking may be related to difficulty interrupting breathing to manipulate phrasing and speech loudness, because their breathing is already greatly impaired.[60,67] The motorized-wheelchair users who did not experience dyspnea during talking probably used an adaptive breathing strategy. This has not been studied, but if a successful adaptive strategy is used, training those who do get breathless while talking could provide relief.

It appears that, as in the able-bodied and in COPD,[68] exercise diminishes breathlessness in SCI. Wien et al[69] studied breathlessness in 183 subjects with neurologically motor complete or incomplete cervical or high thoracic (T6 and above) SCI who, to get around, used hand-propelled wheelchairs more than 50% of the time. Spirometry and maximum inspiratory and expiratory pressure were measured. Those with complete injuries had a greater prevalence of breathlessness than the others. Wheelchair athletes were defined as those who responded "yes" to the question, "Are you a wheelchair athlete?" The distribution of level of SCI was not different between wheelchair athletes and nonathletes. Current cigarette smoking (> 20 per day) was a significant risk factor for breathlessness, independent of pulmonary function and SCI level and completeness. Athletes were significantly less likely to answer

"yes" to the questions about experiencing breathlessness, and they had significantly higher percent-of-predicted FEV_1 (82.7% [n = 47] vs 74.6% [n = 132], p = 0.001) and peak inspiratory pressure (104.2 ± 40 cm H_2O [n = 46] vs 87.2 ± 31.3 cm H_2O [n = 127], p = 0.01). Statistical analysis of the data showed that the contribution of FEV_1 to breathlessness was explained by smoking and a history of obstructive lung disease, and when maximum inspiratory pressure was included in the prediction model, the athletes still reported less breathlessness. Thus, the data showed that the mechanism of decreased breathlessness in wheelchair athletes is not better pulmonary function or respiratory-muscle performance. As in others who exercise regularly, the decrease in breathlessness is probably a fitness effect. Large-scale prospective studies to test whether smoking cessation and fitness programs reduce breathlessness in SCI have not been performed. It is not clear that exercise programs would be successful over the long-term, since few Americans are motivated to perform exercise regularly.

Interventions

Respiratory Muscle Training

The problems of poor respiratory-muscle performance, distortion of the respiratory system, impaired cough, and speech production in ventilated persons with SCI have at least partial solutions that will be addressed in the ensuing paragraphs.

In the able-bodied it has been demonstrated that, like other skeletal muscles, the muscles of ventilation can be trained for both strength and endurance.[70] Typically, vigorous and forceful efforts are required to produce a significant effect. Even marathon runners, who breathe vigorously but not forcefully, have respiratory muscle strength that is not enhanced.[71] Respiratory muscle weakness and inefficiency of breathing predispose tetraplegics to fatigue of the respiratory muscles,[27,72] but substantial training efforts can enhance performance.[72–75]

For example, the subjects studied by Gross et al[72] inspired for 30 min daily, 6 days a week, against a resistance just sufficient to produce electromyographic evidence of fatigue. Uijl et al[75] trained their subjects at 70% of the endurance capacity of their respiratory muscles twice daily for 6 weeks. Rutchik et al[74] trained their subjects over 8 weeks, by having them breathe for 15 min twice daily against progressively higher resistances, The subjects studied by Kogan et al[73] trained weekdays for 2.5–4.5 months by inhaling through a narrow orifice and developing with each breath a target of 80% of their maximum inspiratory muscle pressure. In each of the various studies cited, performance enhancement occurred. For example, in the study by Gross et al[72] there was improvement in both strength
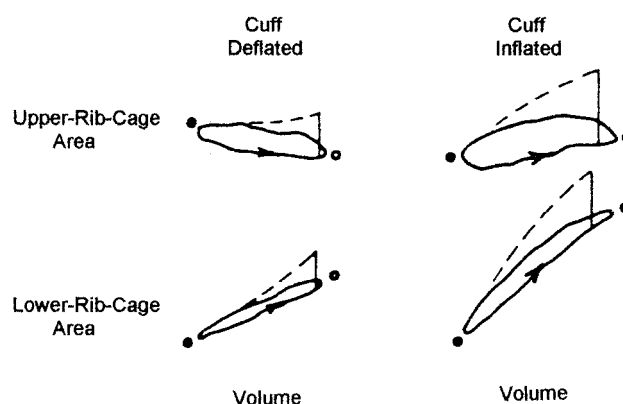

Fig. 1. Data obtained during tidal breathing (solid lines) and during relaxation (dashed lines) from a representative subject with abdominal cuff deflated versus inflated. The vertical axis represents the cross-sectional area of the (upper or lower) rib cage. The horizontal axis represents the change in lung volume. The vertical lines were obtained when, at or near end-tidal inspiration, the subject relaxed against an occluded valve. With the cuff deflated, the upper-rib-cage area was less at end-inspiration than at functional residual capacity. With the cuff inflated, the lower-rib-cage area increased more per unit change in lung volume than it did with the cuff deflated. This is indicated by the increase in slope of the plot of volume versus lower-rib-cage area. Also, as a result of cuff inflation, upper-rib-cage area increased during tidal inspiration, whereas it had decreased with the cuff deflated. (From Reference 18, with permission.)

and endurance. Uijl's subjects experienced an increase in respiratory-muscle endurance and peak oxygen consumption achieved during arm-crank exercise.[75] In the study by Rutchik et al[74] there were significant increases in peak inspiratory pressure (from 66.2 cm H_2O to 78.5 cm H_2O), in FVC (from 2.81 L to 3.07 L) and in total lung capacity (from 5.17 L to 5.71 L). The results from Kogan et al[73] were similar and, in addition, thickening of the diaphragm occurred, which indicates hypertrophy as a result of the training.

During the often-long period of liberating patients with SCI from assisted ventilation, the spontaneous recovery of respiratory muscle function noted above is likely to contribute to success. It is likely that simultaneous respiratory-muscle training would help, but the best training regimen has not been established. In principle, regimens that target both endurance and strength would seem most desirable. It is also not known whether trained subjects will continue the training, with the associated expectation of deterioration once the training is abandoned. Although the subjects studied by Kogan et al benefited from the training, none chose to continue training after the end of the study.

Abdominal Binder

Using inductance pneumography belts during spontaneous breathing in 6 tetraplegic subjects, Urmey et al[18] mon-

itored upper-rib-cage and lower-rib-cage displacements and compared them to the relaxation (undistorted) characteristic of the rib cage. Subjects were studied without and with extrinsic abdominal support, which covered the abdominal wall and did not overlie the lower lateral rib cage. (Note, however, that abdominal binders that do overlie the lower lateral rib cage are not as effective.) Extrinsic abdominal support increased abdominal pressure and, as confirmed by percussion, increased the zone of apposition of the diaphragm to the rib cage.

In addition, the results of axial forcing of the lower rib cage were studied by applying a 4.5-kg cephalad force to the costal margin to mimic the action of the muscular insertion of the diaphragm to the rib cage. Figure 1 shows a typical plot of the area of the upper-rib-cage and the area of the lower-rib-cage as functions of lung volume. During spontaneous breathing, upper-rib-cage area was smaller at end-tidal inhalation than at FRC in all the subjects who demonstrated distortion from the relaxed configuration. With support of the abdominal wall, the upper-rib-cage area increased in all but one subject during tidal inhalation, which shows that the support diminished distortion. In none of the subjects did upper-rib-cage area superimpose the relaxation characteristic, so some distortion remained. Cephalad forcing of the lower-rib-cage at the costal margin caused the upper rib cage to move outward, which shows that the lower rib cage and upper rib cage are coupled well. In summary, the study showed that, in tetraplegia, over the tidal-breathing range, increased abdominal pressure and decreased abdominal compliance due to the presence of extrinsic abdominal support diminishes rib-cage distortion. As inferred from the force-balance analysis by Loring and Mead,[76] the diminished distortion is due to (1) an increase in the area of diaphragmatic apposition to the rib cage and (2) an increase in the insertional component of diaphragm action on the rib cage (see Appendix).

McCool et al[77] subsequently showed that abdominal binding influences rib-cage motion over the entire range of inspiratory capacity in tetraplegia. In able-bodied controls, total lung capacity decreased because the binder impeded diaphragm decent. In tetraplegia, binding led to an increase in total lung capacity and in rib-cage dimensions, which showed that expansion of the paralyzed rib cage dominated the disadvantage of impeded diaphragm descent. Abdominal binders are in common use in tetraplegia, but the beneficial effects need to be weighed against the simultaneous reduction in FRC and the related potential effects on gas exchange.

Ventilator-Assisted Speech

Because the respiratory system provides the aeromechanical drive to the larynx and upper-airway structures

for speech production, SCI can impair speech. Speech is often affected by injuries to the cervical cord (and is typically normal with lower injuries). When cervical injury spares diaphragm function, common speech signs include low loudness (due to impaired alveolar pressure-generation capability) and short phrases (due to small V_T).[78] In such cases, an abdominal binder may improve speech.[79]

When cervical injury impairs diaphragm function, a ventilator is required and speech is substantially altered. The specific nature of ventilator-assisted speech depends on the type of ventilator used. Discussion here will focus on invasive positive-pressure ventilation and ventilation with a phrenic-nerve pacer. More detailed accounts of these forms of ventilator-assisted speech are available elsewhere.[80–82]

Speech produced with invasive positive-pressure ventilation delivered via tracheostomy is substantially different from normal speech, primarily because the ventilator-delivered air enters below the larynx. If the tracheostomy tube cuff is inflated (and the tube is unfenestrated), the patient will not be able to speak at all. Therefore, the first and most critical step toward allowing a patient to speak is to deflate the cuff. This can be done safely by increasing the ventilator-delivered V_T to compensate for air loss through the larynx. A typical error is to choose a V_T that is too small to maintain a constant P_{aCO_2}. Commonly, we have used V_T as high as 1.2 L. This is not as large as first appears. The inspiratory capacity of an adult of average size is about 3 L, so a 1.2-L breath does not bring the subject even close to total lung capacity. In addition, the deflated cuff allows gas to leak out of the mouth for speech production, so the effective V_T is much less than 1.2 L.

Deflation of the cuff makes it possible for the patient to communicate relatively easily with family, friends, and hospital staff. Nevertheless, tracheostomy-ventilation speech is usually abnormal and is characterized by short phrases, long pauses, poor voice quality, and variable loudness.[83] These abnormal speech characteristics are closely linked to the abnormal tracheal pressure associated with tracheostomy ventilation. As shown in Figure 2, tracheostomy-ventilation tracheal pressure (under a volume-controlled condition) rises quickly to a high peak, falls quickly, and remains below the minimum pressure required to vibrate the vocal folds until the next inspiration. This is in stark contrast to the steady and low tracheal pressure associated with normal speech production.

Fortunately, tracheal pressure can be modified with ventilator adjustments to improve speech. Perhaps the most simple and successful set of ventilator adjustments is the combination of prolonged inspiratory time and positive end-expiratory pressure (PEEP).[80,84] Prolonged inspiratory time (or decreased inspiratory flow) increases the time that speech can be produced during inspiration, and PEEP increases the time that speech can be produced during ex-
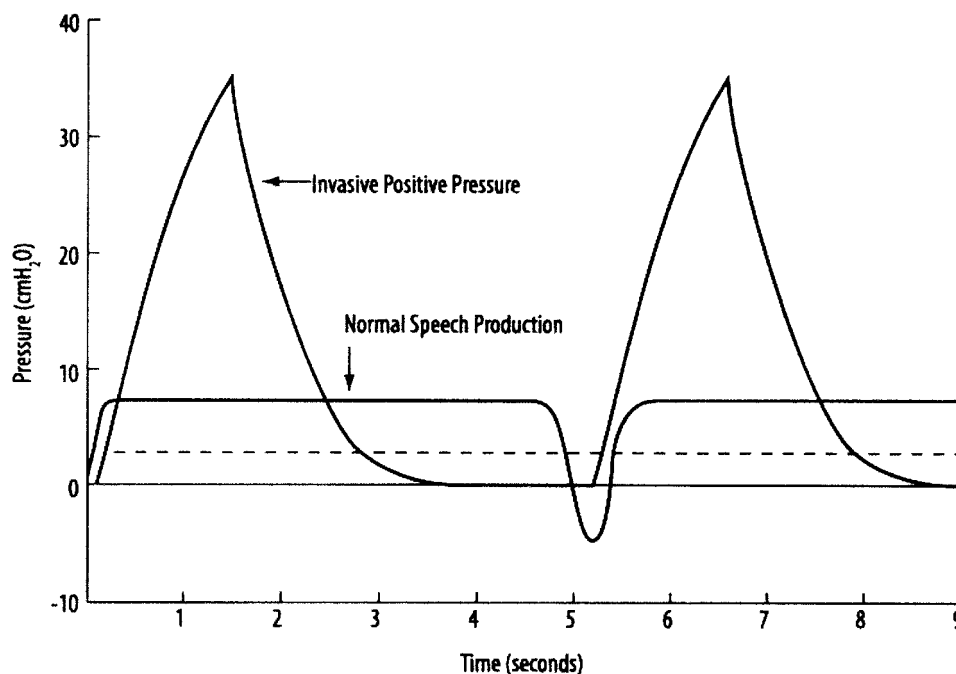
Fig. 2. Phonation relative to pressure and time. The dashed line represents the minimum pressure required for phonation. See text for details. (From Reference 81, with permission.)
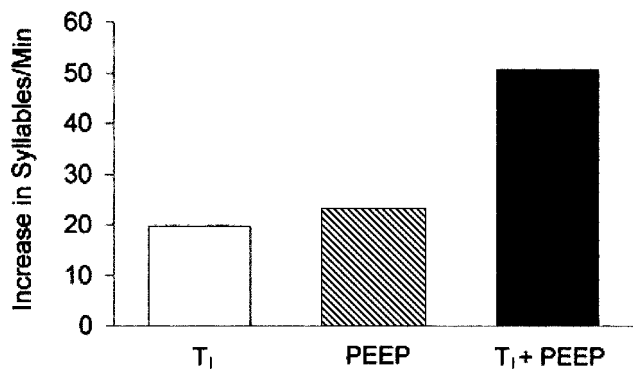

Fig. 3. Changes (from usual) in speaking rate with lengthened inspiratory time (T_I), positive end-expiratory pressure (PEEP), and lengthened T_I plus PEEP. (From Reference 84, with permission.)

Fig. 4. Tracheal pressure during speaking with a one-way valve (top panel) versus with positive end-expiratory pressure (PEEP) set to 15 cm H_2O (bottom panel). Note that the tracings are almost indistinguishable. See text for details. (From Reference 84, with permission.)

piration. Thus, as shown in Figure 3, more speech can be produced per breath with each of these adjustments individually, and their effects are additive when they are combined. Although other ventilator adjustments can also improve speech,[81,85] the effectiveness of prolonged inspiratory time and PEEP has the most empirical support. Whatever ventilator adjustments are implemented, speech may be further improved with behavioral therapy provided by a speech-language pathologist.

A common clinical approach to improving tracheostomy-ventilation speech following cuff deflation is the insertion of a one-way inspiratory valve (ie, a "speaking valve") between the tracheostomy tube and the ventilator
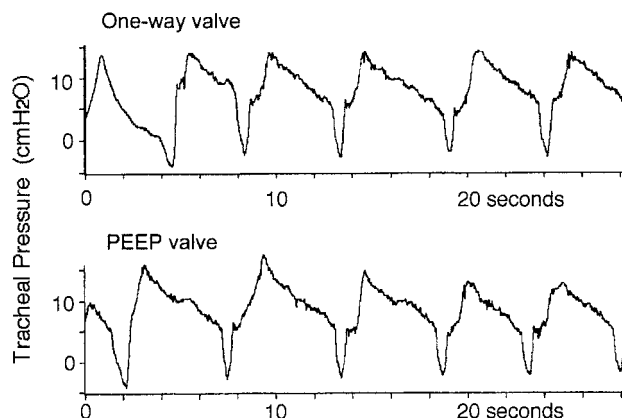
line. This approach carries high risks (including death) if the valve is inserted without the cuff being deflated. Ventilator adjustments such as those just described (prolonged inspiratory time and PEEP) are much safer than a one-way valve, and the speech can be just as good. For example, a comparison of tracheostomy ventilation with a one-way valve and with 15 cm H_2O PEEP was found to generate nearly identical tracheal-pressure waveforms (Fig. 4) and equally high-quality speech.[84]

Speech with phrenic-nerve-pacer ventilation also can be abnormal, and is usually characterized by short phrases, low auditory volume, and long inspiratory pauses.[86] As with tracheostomy-ventilation speech, adjustments to the pacer may improve speech, although this has not been proven. Speech improvements have been found with the use of an abdominal binder,[87] and include louder speech, longer phrases, and better voice quality. These improvements are probably linked to the V_T increase caused by the binder (ie, inward positioning of the abdominal wall by the binder stretches the diaphragm, increases its contractile force, and, as described above and in the Appendix, reduces distortion of the rib cage). Other means of increasing V_T, such as glossopharyngeal breathing, also hold potential for improving speech in patients with phrenic-nerve pacers.

Respiratory Muscle Pacing

New techniques have provided the capacity to restore inspiratory-muscle function in SCI. Conventional bilateral phrenic-nerve pacing can allow patients with ventilator-dependent tetraplegia complete freedom from mechanical ventilation. Compared to mechanical ventilation, phrenic-nerve pacing provides better patient mobility, better comfort level, and lower health-care costs.[88,89] Although this has not been studied formally, patients often claim that their breathing feels more natural with phrenic-nerve pacing than with mechanical ventilation. Why this should be so is not entirely clear. Perhaps it is because of elimination of the noise associated with the mechanical ventilator and the tubing that tugs on the neck. Also, with conventional mechanical ventilation via tracheostomy tube, the inspired gas does not pass through the nose, whereas during inhalation from phrenic-nerve stimulation, the negative pressure generated by diaphragm contraction causes inspired gas to pass through the nose and thereby restores the sense of smell. This may contribute substantially to the sense that the breath is more natural. Phrenic-nerve pacing usually does not allow removal of the tracheostomy tube, because most patients develop obstructive sleep apnea. This is because the pacing of the diaphragm is not synchronous with respirophasic activation of oropharyngeal muscles that prevent airway collapse during normal breathing during sleep. It would seem possible to avoid this problem by using CPAP during sleep in phrenic-nerve paced patients, but this has not been studied.

With any method of phrenic-nerve pacing, intact phrenic-nerve function must be assured. This is usually accomplished by electrical stimulation of the phrenic nerves in the neck and recording the resultant compound action potential on the surface of the rib cage overlying the diaphragm. An alternative method involves magnetic stimulation of the phrenic motoneurons. For electrode placement,
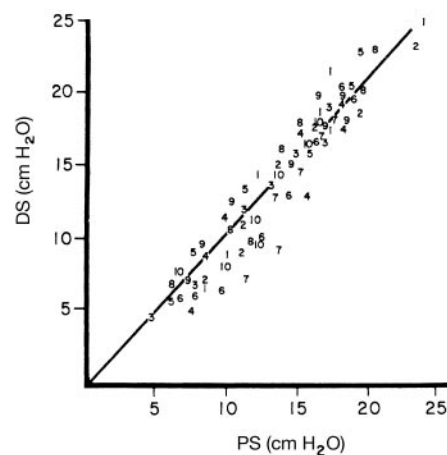

Fig. 5. Comparison of transdiaphragmatic pressure generated during phrenic-nerve stimulation (PS) and intramuscular diaphragm stimulation (DS) in anesthetized dogs. Supramaximal amplitudes were applied over a 10–40 Hz range during airway occlusion. Each number represents results from a separate animal. (From Reference 93, with permission.)

the technique of phrenic-nerve pacing generally requires a thoracotomy,[90–92] which is associated with substantial morbidity, high cost, and hospitalization. Also, the procedure carries a small risk of phrenic-nerve injury. Recent studies, however, suggest that phrenic-nerve stimulation can be achieved less invasively by placement of intramuscular electrodes directly into the diaphragm via laparoscopic surgery. Laparoscopy is less invasive, has cosmetic benefits, and can be performed as an out-patient procedure or with a single overnight observation stay, with lower overall cost. Also, laparoscopic placement of intramuscular electrodes does not require manipulation of the phrenic nerves and therefore carries less risk of phrenic-nerve injury.

Animal experiments first suggested that each hemidiaphragm could be activated with intramuscular diaphragm electrodes.[93] Electrodes were placed on the ventral surface of the diaphragm via a midline abdominal incision. To evaluate the efficacy of this method, transdiaphragmatic pressure generated by a single intramuscular electrode positioned near the point of entrance of the phrenic nerve (motor point) was compared to transdiaphragmatic pressure generated by direct phrenic-nerve stimulation with cuff electrodes. The study demonstrated that intramuscular electrodes positioned within 1–2 cm of the motor point resulted in transdiaphragmatic pressure similar to the pressure obtained with direct phrenic-nerve stimulation (Fig. 5).

Clinical trials of this technique were initially hindered by lack of the necessary tools to safely place the electrodes within the diaphragm and methods to accurately locate the diaphragm motor points. In early animal studies, laparoscopic placement of intramuscular diaphragm electrodes often resulted in the inadvertent placement of electrodes
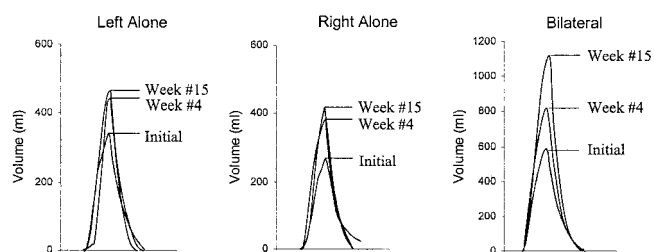
Fig. 6. Superimposed maximum changes in inspired volume from separate left, separate right, and bilateral diaphragm stimulation, over the course of the reconditioning period of one subject. Supramaximal stimulus variables (24 mA or 25 mA, 50 Hz) were employed. (From Reference 100, with permission.)
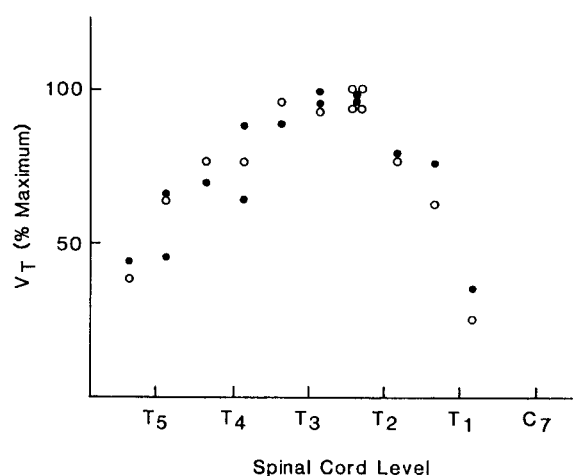

Fig. 7. Effects of electrical stimulation applied at different spinal cord regions before phrenicotomy (circles) and after phrenicotomy (dots) in a dog anesthetized with pentobarbital. Inspired volume is expressed as a percentage of maximum. Inspired-volume generation was greatest in the vicinity of the T2-T3 spinal-cord region and decreased progressively at spinal levels cephalad and caudad to that region. Stimulation was applied with supramaximal stimulus amplitude and stimulus frequency of 50 Hz. (From Reference 101, with permission.)

through the diaphragm and into the thoracic cavity, because the laparoscope delivering the electrode approached the diaphragm almost perpendicular to the plane of the diaphragm. Therefore, an insertion instrument was designed that allowed electrode placement in the same plane as the diaphragm.[94–96] With this instrument, both in animal and clinical studies, there were not any instances of electrodes traversing the diaphragm and entering the thoracic cavity.

Since uniform activation of the diaphragm requires placement of intramuscular electrodes near the motor points of the phrenic nerves into each hemidiaphragm, and the phrenic nerves are not visible on the abdominal surface of the diaphragm, a mapping procedure was developed to precisely determine the location of the motor points.[97,98] This procedure entails evaluating several test sites in the general region of the motor points (based on previous anatomic studies) with a suction electrode that can be reversibly applied to the diaphragm.

After extensive animal testing, studies were performed in an initial group of 5 ventilator-dependent tetraplegic subjects.[99,100] Two intramuscular electrodes were placed laparoscopically in each hemidiaphragm, to activate the diaphragm, with the goal of full-time ventilatory support. Two weeks following placement of the diaphragm electrodes, inspired-volume generation was evaluated with each of the 4 electrodes individually.[101] In each instance, maximum or near-maximum amplitude (24–25 mA) was required to achieve maximum inspired-volume-generation. Since patients on long-term mechanical ventilation suffer from diaphragm atrophy, a period of gradually increasing stimulation (reconditioning period) was necessary to restore diaphragm function. Stimulation of both electrodes within each hemidiaphragm generally resulted in greater inspired volumes than did stimulation of one electrode alone, so stimulation was applied with all 4 electrodes. During the reconditioning period, attempts were made to gradually reduce the stimulus frequency to the lowest value that resulted in adequate ventilation. Respiratory rate was set at 10–12 breaths/min and inspiratory time (train duration) at 1.1 s. Initially, pacing was provided for 5–10 min

each hour for 5–6 h/d, and gradually increased, as tolerated. When continuous pacing for 6–8 hours was achieved, the number of hours per day was gradually increased.

Figure 6 shows the maximum changes in inspired volume that resulted from separate left and right hemidiaphragm stimulation and bilateral stimulation, at various intervals during the reconditioning period, superimposed, from one subject. Similar results were observed in the other subjects.[100] The inspired volumes achieved during long-term pacing were in the range 800–950 mL. Each of the subjects achieved substantial independence from mechanical ventilatory support (ie, full-time ventilatory support in 3 subjects and up to 20 h/d in the remaining subject). End-tidal P_{CO_2} during long-term pacing was 30–32 mm Hg. Intramuscular diaphragm pacing has since been performed on 7 additional subjects, each of whom also achieved either full-time or near-full-time independence from mechanical ventilation.[100]

For patients who do not have intact phrenic nerves, conventional or intramuscular diaphragm pacing is not an option. However, studies of animals with surgical phrenicectomy found that the application of electrical current on the epidural surface of the spinal cord in the upper thoracic region results in substantial inspired-volume generation.[101] The maximum inspired volumes were generated by stimulation at the T2-T3 region on the ventral surface (Fig. 7). Stimulation of the intercostal muscles at this level in combination with bilateral phrenic-nerve stimulation resulted in inspired-volume generation that approximated the inspiratory capacity. These studies suggested
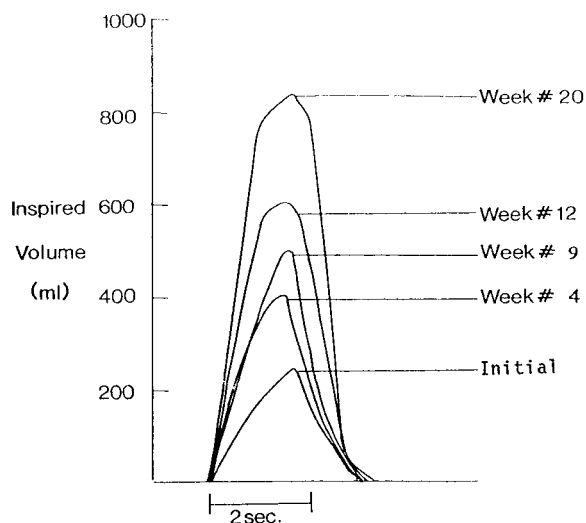
Fig. 8. Maximum inspired volumes with intercostal stimulation at various times during the reconditioning period of one patient. As a result of gradually increasing the pacing duration, there were progressive increments in inspired volume. Because inspiratory time was fixed, increases in inspired volume were achieved by increases in inspiratory flow. The improvement in inspired volume is attributable to the training effect of long-term stimulation and reversal of intercostal muscle atrophy. (From Reference 102, with permission.)

that near-maximal, coordinated activation of the inspiratory intercostal muscles could be accomplished by electrical stimulation with a single electrode.[101] Because the mechanism of intercostal activation involves direct stimulation of thoracic ventral roots, this technique is referred to as ventral root stimulation.

An initial clinical trial of ventral root stimulation was undertaken to provide ventilatory support in ventilator-dependent tetraplegic patients who were not candidates for phrenic-nerve pacing.[102] An epidural disc electrode was positioned on the ventral surface at the T2-T3 level of the spinal cord, through a cervical hemilaminectomy. Initial ventral root stimulation resulted in very small inspired volumes (150–240 mL), most likely secondary to muscle atrophy. Following a reconditioning program, there were substantial increases in maximum inspired volumes (to 470–850 mL). Figure 8 shows an example of the changes in inspired volume over the course of the conditioning program. The maximum time that patients could be maintained off mechanical ventilation, however, was a few hours. This study demonstrated that intercostal muscle pacing alone does not result in sufficient inspired-volume production to support ventilation for prolonged periods.

There are a number of factors that may have accounted for the different outcome of the clinical trial, compared to the previous animal studies.[103] An important factor that may have decreased inspired-volume production in the human trial is the reduction in rib-cage and lung compli-

ance in patients with tetraplegia.[3,11] It is also possible that the intercostal muscles were not activated to the same extent in humans. The differences in shape of the thorax between species may also be important. During inspiration, there is greater expansion of the anteroposterior diameter in the quadruped thorax. These differences may provide the intercostal muscles of the dog with a mechanical advantage over humans.

Though not successful in maintaining long-term ventilatory support, ventral root stimulation did result in substantial inspired-volume production. The method was applied in a second clinical trial of 4 patients with intact unilateral phrenic-nerve function.[104] Unilateral phrenic-nerve pacing does not provide V_T sufficient to support ventilation. It was thought that intercostal pacing combined with unilateral phrenic-nerve pacing could provide long-term ventilatory support. Spinal-cord electrodes were positioned at the T2-T3 spinal-cord level. A phrenic-nerve electrode was placed unilaterally, via the thoracic approach, to pace the diaphragm. With reconditioning, the maximum inspired volumes resulting from combined intercostal and diaphragm stimulation gradually increased to between 550 mL and 1,310 mL. With the lower stimulus frequencies employed with long-term pacing, inspired volumes during long-term pacing ranged between 350 mL and 850 mL. Each patient was able to achieve full-time or near-full-time ventilatory support. End-tidal P_{CO_2} was in the range of 33–35 mm Hg during long-term pacing. As with diaphragm pacing, the subjects reported improved sense of smell, mobility, and overall sense of well being. In fact, one patient achieved employment as a result. This technique was associated with some adverse effects, including mild hand flexion.[102] Also, with high stimulus currents, one of the subjects experienced symptoms suggestive of autonomic dysfunction, including headache, flushing, and diaphoresis. Reducing the stimulus current eliminated the symptoms.

Perhaps the single most important impairment of the respiratory system that occurs in SCI is ineffective cough due to dysfunction of the abdominal and internal intercostal muscles. To address this problem, the technique of direct stimulation of the spinal cord by epidural electrodes has now been extended to produce vigorous contraction of the abdominal muscles.[105] In one subject, during combined stimulation at the T9 and L1 levels, peak airway pressure and peak expiratory flow were 132 cm H_2O and 7.4 L/s, respectively. These are within the range of predicted values for an able-bodied person. The subject is able to trigger the stimulation on his own and, as a result, does not require assistance for airway clearance. The technique may have a substantial impact on the outcome of respiratory-tract infections in SCI.

In summary, the successful development of new techniques and approaches to restore inspiratory and expira-

tory muscle activation has expanded the options for patients with SCI. Whether these methods will reduce the need for mechanical ventilation, reduce the incidence of respiratory complications, and significantly impact the morbidity and mortality of high SCI remains to be determined.

Assisted Cough

Impaired cough leads to accumulation of secretions that arise from respiratory infections or that are due to the presence of a tracheostomy tube. Several methods are available to deal with the secretions. Each may be performed in association with conventional chest physical therapy.

Spinal-Cord Stimulation. The new technique described above, involving stimulation of the spinal cord at T9 and L1, is still being developed[105] but shows great promise.

Spontaneous Cough. For mild and nonviscous secretions, spontaneous coughs may suffice. Often, the effectiveness is enhanced if the seated subject lurches forward. This maneuver raises intra-abdominal pressure, which is transmitted to the thorax, enhancing expiratory flow.

Quad Cough. A related maneuver known as a "quad cough" is also often effective. The subject is placed in the supine or slightly upright posture and is straddled by the therapist whose hands are placed under the left and right costal margins. Then the subject inhales to total lung capacity and, upon a signal, begins to cough. The therapist enhances the cough effort by vigorous pressure applied to the abdomen, in the rhythm of a cough, and the process is continued to low lung volume. The forceful application of pressure to the abdomen greatly enhances expiratory flow and mobilization of secretions. Indeed, maximum expiratory flow and the phenomenon of expiratory flow limitation that is essential to an effective cough can be achieved when the maneuver is done properly (unpublished data from Robert Brown [author], and Steven M Scharf MD, University of Maryland, Baltimore, Maryland). Contraindications to performing quad coughs are the presence of an inferior vena cava filter or abdominal aortic aneurysm or prosthesis.

Suction Catheter. Other methods to enhance cough utilize negative pressure applied at the mouth or orifice of the endotracheal tube or tracheostomy tube. The most common device is a standard suction catheter, which is convenient when there is a tracheal tube in place. When there is not, nasotracheal or orotracheal suctioning can be done, but this is uncomfortable and can lead to trauma to the nose, pharynx, and larynx. The usefulness of the standard suction catheter is limited, because it barely reaches the right main bronchus and does not enter the left main bronchus. Versions of the catheter that can be turned to enter the proximal portion of the left main bronchus are not in common use and are of unproven efficacy. In addition, each use of a suction catheter introduces bacteria to the trachea and large bronchi, with the associated risk of infection.

Mechanical In-exsufflation. The latter limitations can be overcome with the use of a mechanical insufflation-exsufflation device (CoughAssist, JH Emerson, Cambridge, Massachusetts). This device is particularly effective when used attached to a tracheal tube. When applied to the mouth, the negative-pressure phase can be uncomfortable or cause collapse of the pharyngeal airway, particularly in patients with dysfunction of the oropharyngeal muscles, such as due to amyotrophic lateral sclerosis. The device can be used frequently. For secretion clearance in patients with neuromuscular disease, one of us (Brown) has used it as often as a patient with excessive secretions would ordinarily cough. For example, use every 5 minutes for an hour may be very effective in clearing retained secretions. The clinical effectiveness of the device has been demonstrated.[106,107]

Bronchoscopy. Perhaps the most-effective method of secretion clearance is bronchoscopy, and this should be used when other methods have been inadequate. The experience of one of us (Brown) has been that performing quad coughs during the bronchoscopy mobilizes secretions from airways that are not bronchoscopically visible and thereby enhances secretion clearance. During the quad cough, the bronchoscope must be poised to suction the secretions that are "milked" into the visible airways, lest those secretions migrate distally and out of vision during the subsequent inhalation.

Mechanical Ventilation

The care of patients with high-level SCI is made more complex by the need for mechanical ventilation. Although most patients are ventilated via tracheostomy tube, Bach and colleagues found that patients with SCI and other patients with neuromuscular disorders can be successfully decannulated and treated with noninvasive ventilation.[108–111] Patient-ventilator interfaces for such ventilation are reviewed elsewhere in this issue of RESPIRATORY CARE.[112] Patients seem to prefer noninvasive ventilation for reasons of appearance, comfort, swallowing, and speech.[113] Other advantages appear to be reduction in secretions because the tracheostomy tube is not present to irritate the trachea, fewer hospitalizations, and lower cost of care.[114] More attention should be paid to the option of noninvasive ventilation, especially given the considerable literature supporting this approach.

Mechanically ventilated patients typically prefer large V_T. The reason is not established, but it may be related to

relief of the dyspnea that is associated with small tidal breaths.[65] Large breaths also have the benefit of preventing small-airway narrowing or closure by stretching airway smooth muscle, and by reducing surface tension by expanding the surface area of pulmonary surfactant. Breath sizes as large as 1.0 L (often with PEEP of 5 cm H_2O) are common and do not cause ventilator-associated lung damage in the absence of acute lung injury from other causes. The common result is low P_{aCO_2}, but there are not any known deleterious long-term consequences of this.

Summary

Respiratory dysfunction is a major cause of morbidity and mortality in SCI. In the last several decades, much has been learned about the mechanisms of respiratory dysfunction, and this has led to useful interventions. Respiratory-muscle training improves VC. A multicenter effort to assess the effect of respiratory-muscle training on problems such as dyspnea and weak cough is warranted. Respiratory-muscle pacing has shown promise in liberating patients from mechanical ventilation and in improving cough. Several techniques are now available to clear secretions, including mechanical in-exsufflation, which appears to be underutilized in both the acute and chronic settings. For those who require assisted ventilation and have tracheotomies, the quality of speech production can be improved dramatically with simple adjustments of the ventilator settings. Noninvasive methods to provide assisted ventilation are also effective, yet underutilized. With further research, there will perhaps be enough progress to substantially reduce the morbidity and mortality from respiratory dysfunction in SCI.

REFERENCES

1. Spinal cord injury: facts and figures at a glance. National Spinal Cord Injury Center Statistical Center, Birmingham, Alabama. J Spinal Cord Med 2006;29(4):379–80.
2. ASIA impairment scale, clinical syndromes, and standard neurological classification of spinal cord injury. http://www.asia-spinalinjury.org/publications/index.html.
3. Scanlon PD, Loring SH, Pichurko BM, McCool FD, Slutsky AS, Sarkarati M, Brown R. Respiratory mechanics in acute quadriplegia: lung and chest wall compliance and dimensional changes during respiratory maneuvers. Am Rev Respir Dis 1989;139(3):615–620.
4. Sharp JT, Sweany SK, VanLith P. Physiologic observations in diffuse pulmonary fibrosis and granulomatosis. Am Rev Respir Dis 1966;94(3):316–331.
5. Gibson GJ, Pride NB. Lung distensibility: the static pressure-volume curve of the lungs and its use in clinical assessment. Br J Dis Chest 1976;70(3):143–184.
6. Gibson GJ, Pride NB, Davis JN, Loh LC. Pulmonary mechanics in patients with respiratory muscle weakness. Am Rev Respir Dis 1977;115(3):389–395.
7. Bergofsky EH. Respiratory insufficiency in mechanical and neuromuscular disorders of the thorax. In: Fishman AP, editor. Pulmonary diseases and disorders. New York: McGraw-Hill; 1980:1559.
8. Bergofsky EH. Respiratory failure in disorders of the thoracic cage. Am Rev Respir Dis 1979;119(4):643–669.
9. Bergofsky EH. Mechanism for respiratory insufficiency after cervical cord injury: a source of alveolar hypoventilation. Ann Intern Med 1964;61:435–447.
10. Stone DJ, Keltz H. The effect of respiratory muscle dysfunction on pulmonary function: studies in patients with spinal cord injuries. Am Rev Respir Dis 1963 Nov;88:621–629.
11. DeTroyer A, Heilporn A. Respiratory mechanics in quadriplegia: the respiratory function of the intercostal muscles. Am Rev Respir Dis 1980;122(4):591–600.
12. Young SL, Tierney DF, Clements JA. Mechanism of compliance changes in excised rat lungs at low transpulmonary pressure. J Appl Physiol 1970;29(6):780–785.
13. Grinton SF, Hyatt RE, Rohrback MS, Deschamps C. Causes of pressure volume curve shifts in chest restricted dogs. Fed Proc 1987;46:1108.
14. DeTroyer A, Pride NB. The respiratory system in neuromuscular disease. In: Roussos C, Macklem PT, editors. The thorax. (Lung biology in health and disease, vol 85). New York: Marcel Dekker; 1985:1089–1121.
15. Goldman JM, Williams SJ, Denison DM. The rib cage and abdominal components of respiratory system compliance in tetraplegic patients. Eur Respir J 1988;1(3):242–247.
16. Estenne M, DeTroyer A. The effect of tetraplegia on chest wall statics. Am Rev Respir Dis 1986;134(1):121–124.
17. Goldman JM, Rose LS, Morgan MD, Denison DM. Measurement of abdominal well compliance in normal subjects and tetraplegic patients. Thorax 1986;41(7):513–518.
18. Urmey W, Loring S, Mead J, Slutsky AS, Sarkarati M, Rossier A, Brown R. Upper and lower rib cage deformation during breathing in quadriplegics. J Appl Physiol 1986;60(2):618–622.
19. Derenne J, Macklem PT, Roussos C. The respiratory muscles: mechanics control and pathophysiology. Am Rev Respir Dis 1978;118(1):119–133.
20. Fugl-Meyer AR, Grimby G. Rib cage and abdominal volume ventilation partitioning in tetraplegic patients. Scand J Rehab 1971;3(4):161–167.
21. Mortola JP, Sant'Ambrogio G. Motion of the rib cage and the abdomen in tetraplegic patients. Clin Sci Mol Med 1978;54(1):25–32.
22. Moulton A, Silver JR. Chest movements in patients with traumatic injuries of the cervical cord. Clin Sci 1970;39(3):407–422.
23. Danon J, Druz WS, Goldberg NB, Sharp JT. Function of the isolated paced diaphragm and the cervical accessory muscles in C1 quadriplegics. Am Rev Respir Dis 1979;119(6):909–919.
24. Braun NM, Arora NS, Rochester DF. Force-length relationship of the normal human diaphragm. J Appl Physiol 1982;53(2):405–412.
25. Kim MJ, Druz WS, Danon J, Machnach W, Sharp JT. Mechanics of the canine diaphragm. J Appl Physiol 1976;41(3):369–382.
26. de Haan A, de Jong J, van Doorn JE, Huijing PA, Woittiez RD, Westra HG. Muscle economy of isometric contractions as function of stimulation time and relative muscle length. Pfluegers Arch 1986;407(4):445–450.
27. Manning H, McCool FD, Scharf SM, Garshick E, Brown R. Oxygen cost of resistive-loaded breathing in quadriplegia. J Appl Physiol 1992;73(3):825–831.
28. Bluechardt MH, Wiens M, Thomas SG, Plyley MJ. Repeated measurement of pulmonary function following spinal cord injury. Paraplegia 1992;30(11):768–774.
29. Oo T, Watt JW, Soni BM, Sett PK. Delayed diaphragm recovery in 12 patients after high cervical spinal cord injury: a retrospective review of the diaphragm status of 107 patients ventilated after acute spinal cord injury. Spinal Cord 1999;37(2):117–122.

30. McKinley WO. Late return of diaphragm function in a ventilator-dependent patient with a high cervical tetraplegia: case report, and interactive review. Spinal Cord 1996;34(10):626–629.

31. Lieberman J, Corkill G, Nayak NN, French BN, Taylor RG. Serial phrenic nerve conduction studies in candidates for diaphragmatic pacing. Arch Phys Med Rehab 1985;61(11):528–531.

32. Silver JR, Lehr RP. Electromyographic investigation of the diaphragm and intercostal muscles in tetraplegics. J Neurol Neurosurg Psychiatry 1981;44(9):837–842.

33. Frisbie JH, Brown R. Waist and neck enlargement after quadriplegia. J Am Paraplegia Soc 1994;17(4):177–178.

34. Axen K, Pineda H, Shunfenthal I, Haas F. Diaphragmatic function following cervical cord injury: neurally mediated improvement. Arch Phys Med Rehabil 1985;66(4):219–222.

35. Brown R, Loring SH, Pichurko BM, Scanlon PD, Slutsky AS, Sarkarati M, et al. The role of respiratory muscles in the recovery of respiratory function in acute tetraplegia. J Spinal Cord Med. In press 2006.

36. Standardization of spirometry, 1994 Update. American Thoracic Society Am J Respir Crit Care Med 1995;152(3):1107–1136.

37. Kelley A, Garshick E, Gross ER, Lieberman SL, Tun CG, Brown R. Spirometry testing standards in spinal cord injury. Chest 2003;123(3):725–730.

38. Fugl-Meyer AR. Effects of respiratory muscle paralysis in tetraplegic and paraplegic patients. Scand J Rehabil Med 1971;3(4):141–150.

39. Fugl-Meyer AR, Grimby G. Ventilatory function in tetraplegic patients. Scand J Rehabil Med 1971;3(4):151–160.

40. Kokkola K, Moller K, Lehtonen T. Pulmonary function in tetraplegic and paraplegic patients. Ann Clin Res 1975;7(2):76–79.

41. Ohry A, Molho M, Rozin R. Alterations of pulmonary function in spinal cord injured patients. Paraplegia 1975;13(2):101–108.

42. Forner JV. Lung volumes and mechanics of breathing in tetraplegics. Paraplegia 1980;18(4):258–266.

43. Haas F, Axen K, Pineda H, Gandino D, Haas A. Temporal pulmonary function changes in cervical cord injury. Arch Phys Med Rehab 1985;66(3):139–144.

44. Almenoff PL, Spungen AM, Lesser M, Bauman WA. Pulmonary function survey in spinal cord injury: influences of smoking and level and completeness of injury. Lung 1995;173(5):297–306.

45. Linn WS, Spungen AM, Gong H Jr, Bauman WA, Adkins RH, Waters RL. Smoking and obstructive lung dysfunction in persons with chronic spinal cord injury. J Spinal Cord Med 2003;26(1):28–35.

46. Jain NB, Brown R, Tun CG, Gagnon D, Garshick E. Determinants of FEV_1, FVC, and FEV_1/FVC in chronic spinal cord injury. Arch Phys Med Rehab. In press 2006.

47. Samsa GP, Patrick CH, Feussner JR. Long-term survival of veterans with traumatic spinal cord injury. Arch Neurol 1993;50(9):909–914.

48. DeVivo MJ, Stover SL. Long-term survival and causes of death. In: Stover SL, DeLisa JA, Whiteneck GG, editors. Spinal cord injury: clinical outcomes from the models systems. Gaithersburg MD: Aspen Publishers; 1995:289–316.

49. Mesard L, Carmody A, Mannarino E, Ruge D. Survival after spinal cord trauma: a life table analysis. Arch Neurol 1978;35(2):78–83.

50. Soden RJ, Walsh J, Middleton JW, Craven ML, Rutkawki SB, Yeo JD. Causes of death after spinal cord injury. Spinal Cord 2000;38(10):604–610.

51. Hartkopp A, Bronnum-Hansen H, Seidenschnur AM, Biering-Sorensen F. Survival and cause of death after traumatic spinal cord injury: a long-term epidemiological survey from Denmark. Spinal Cord 1997;35(2):76–85.

52. Yeo JD, Walsh J, Rutkowski S, Soden R, Craven M, Middleton J. Mortality following spinal cord injury. Spinal Cord 1998;36(5):329–336.

53. Frankel HL, Coll JR, Charlifue SW, Whiteneck GG, Gardner BP, Jamous MA, et al. Long-term survival in spinal cord injury; a fifty year investigation. Spinal Cord 1998;36(4):266–274.

54. Whiteneck GG, Charlifue SW, Frankel HL, Fraser MH, Gardner BP, Gerhart KA, et al. Mortality, morbidity, and psychosocial outcomes of persons spinal cord injured more than 20 years ago. Paraplegia 1992;30(9):617–630.

55. McKinley AC, Auchicloss JH Jr, Gilbert R, Nicholas JJ. Pulmonary function, ventilatory control, and respiratory complications in quadriplegic subjects. Am Rev Respir Dis 1969;100(4):526–532.

56. Garshick E, Kelley A, Cohen SA, Garrison A, Tung CG, Gagnon D, Brown R. A prospective assessment of mortality in chronic spinal cord injury. Spinal Cord 2005;43(7):408–416.

57. Lieberman SL, Mourad I, Brown R, Schwartzstein RM. Spinal cord injury diminishes both the ventilatory response and air hunger due to steady state hypercapnia (abstract). Am Rev Respir Dis 1993;147:A550.

58. Spungen AM, Grimm DR, Lesser M, Bauman WA, Almenoff PL. Self-reported prevalence of pulmonary symptoms in subjects with spinal cord injury. Spinal Cord 1997;35(10):652–657.

59. Ayas NT, Garshick E, Lieberman SL, Wien MF, Tun C, Brown R. Breathlessness in spinal cord injury depends on injury level. J Spinal Cord Med 1999;22(2):97–101.

60. Grandas NF, Jain NB, Denckla JB, Brown R, Tun CG, Gallagher ME, Garshick E. Dyspnea during daily activities in chronic spinal cord injury. Arch Phys Med Rehab 2005;86(8):1631–1635.

61. Burkett LN, Chisum J, Stone W, Fernhall B. Exercise capacity of untrained spinal cord injured individuals and the relationship of peak oxygen uptake to the level of injury. Paraplegia 1990;28(8):512–521.

62. Irizawa M, Yamasaki M, Muraki S, Komura T, Seki K, Kikuchi K. Relationship between heart rate and oxygen uptake during submaximal arm cranking in paraplegics and quadriplegics. Ann Physio Anthrop 1994;13(5):275–280.

63. Coutts KD, Rhodes EC, McKenzie DC. Maximal exercise responses of tetraplegics and paraplegics. J Appl Physiol 1983;55(2):479–482.

64. Figoni S. Exercise responses and quadriplegia. Med Sci Sports Exerc 1993;25(4):433–441.

65. Manning HL, Shea SA, Schwartzstein RM, Lansing RW, Brown R, Banzett RB. Reduced tidal volume increases "air hunger" at fixed P_{CO_2} in ventilated quadriplegics. Respir Physiol 1992;90(1):19–30.

66. Van loan MD, McCluer S, Loftin JM, Boileau RA. Comparison of physiological responses to maximal arm exercise among able-bodied, paraplegics and quadriplegics. Paraplegia 1987;25(5):397–405.

67. Bailey ER, Hoit JD. Speaking and breathing in high respiratory drive. J Speech Lang Hear Res 2002;45(1):89–99.

68. Carrieri-Kohlman V, Gormley JM, Douglas MK, Paul SM, Stulbarg MS. Exercise training reduces dyspnea and the distress and anxiety associated with it: monitoring alone may be as effective as coaching. Chest 1996;110(6):1526–1535.

69. Wien MF, Garshick E, Tun CG, Lieberman SL, Kelley A, Brown R. Breathlessness and exercise in spinal cord injury. J Spinal Cord Med 1999;22(4):297–302.

70. Leith DR, Bradley M. Ventilatory muscle strength and endurance training. J Appl Physiol 1976;41(4):508–516.

71. Loke J, Mahler DA, Virgulto JA. Respiratory muscle fatigue after marathon running. J Appl Physiol 1982;52(4):821–824.

72. Gross D, Ladd HW, Riley EJ, Macklem PT, Grassino A. The effect of training on strength and endurance of the diaphragm in quadriplegia. Am J Med 1980;68(1):27–35.

73. Kogan I, McCool FD, Liberman SL, Garshick E, Shannon K, Frisbee JH, Brown R. Diaphragm hypertrophy during inspiratory muscle training in tetraplegia (abstract). Am J Respir Crit Care Med. 1996;153(4):A25.

74. Rutchik A, Weissman AR, Almenoff PL, Spungen AM, Bauman WA, Grimm DR. Resistive inspiratory muscle training in subjects with chronic cervical spinal cord injury. Arch Phys Med Rehab 1998;79(3):293–297.

75. Uijl SG, Houtman S, Folgering HT, Hopman MT. Training of the respiratory muscles in individuals with tetraplegia. Spinal Cord 1999;37(8):575–579.

76. Loring SH, Mead J. Action of the diaphragm on the rib cage inferred from a force balance analysis. J Appl Physiol 1982;53(3):756–760.

77. McCool FD, Pichurko BM, Slutsky AS, Sarkarati M, Rossier A, Brown R. Changes in lung volume and rib cage configuration with abdominal binding in quadriplegia. J Appl Physiol 1986;60(4):1198–1202.

78. Hoit JD, Banzett RB, Brown R, Loring SH. Speech breathing in cervical spinal cord injury. J Speech Hear Res 1990;33(4):798–807.

79. Watson PJ, Hixon TJ. Effects of abdominal trussing on breathing and speech in men with cervical spinal cord injury. J Speech Lang Hear Res 2001;44(4):751–762.

80. Hoit JD, Banzett RB. Simple adjustments can improve ventilator-supported speech. Am J Speech Lang Pathol 1997;6(1):87–96.

81. Hixon TJ, Hoit JD. Evaluation and management of speech breathing disorders: principles and methods. Tucson AZ: Redington Brown; 2005.

82. Hoit, JD, Banzett, RB, Brown, R. Ventilator-supported speech. In: Tobin M, editor. Principles and practice of mechanical ventilation, 2nd ed. New York: McGraw-Hill; 2006.

83. Hoit JD, Shea SA, Banzett RB. Speech production during mechanical ventilation in tracheostomized individuals. J Speech Hear Res 1994;37(1):53–63.

84. Hoit JD, Banzett RB, Lohmeier HL, Hixon TJ, Brown R. Clinical ventilator adjustments that improve speech. Chest 2003;124(4):1512–1521.

85. Prigent H, Samuel C, Louis B, Abinun MF, Zerah-Lancner F, Lejaille M. Comparative effects of two ventilatory modes on speech in tracheostomized patients with neuromuscular disease. Am J Respir Crit Care Med 2003;167(2):114–119.

86. Hoit JD, Shea SA. Speech production and speech with a phrenic nerve pacer. Am J Speech-Lang Pathol 1996;5(2):53–60.

87. Hoit JD, Banzett RB, Brown R. Binding the abdomen can improve speech in men with phrenic nerve pacers. Am J Speech Lang Pathol 2002;11(1):71–76.

88. Esclarin A, Bravo P, Arroyo O, Mazaira J, Garrido H, Alcaraz MA. Tracheostomy ventilation versus diaphragmatic pacemaker ventilation in high spinal cord injury. Paraplegia 1994;32(10):687–693.

89. Dobelle MH, D'Angelo MS, Goetz BF, Kiefer DG, Lallier TJ, Lamb JI, Yazwinsky JS. 200 cases with a new breathing pacemaker dispel myths about diaphragm pacing. ASAIO J 1994;40(3):M244–M252.

90. Glenn WW, Phelps ML. Diaphragm pacing by electrical stimulation of the phrenic nerve. Neurosurgery 1985;17(6):974–984.

91. Glenn WW, Sairenji H. Diaphragm pacing in the treatment of chronic ventilatory insufficiency. In: Roussos C, Macklem PT, editors. The thorax (Lung biology in health and disease, vol 85). New York: Marcel Dekker 1985:1407–1440.

92. Glenn WW, Phelps ML, Elefteriades JA, Dentz B, Hogan JF. Twenty years of experience in phrenic nerve stimulation to pace the diaphragm. Pacing Clin Electrophysiol 1986;9(6 Pt 1):780–784.

93. Nochomovitz ML, DiMarco AF, Mortimer JT, Cherniack NS. Diaphragm activation with intramuscular stimulation in dogs. Am Rev Respir Dis 1983;127(3):325–329.

94. Stellato TA, Peterson DK, Buehner P, Nochomovitz ML, Mortimer JT. Taking the laparoscope to the laboratory for ventilatory research. Am Surg 1990;56(3):131–133.

95. Aiyar H, Stellato TA, Onders RP, Mortimer JT. Laparoscopic implant instrument for the placement of intramuscular electrodes in the diaphragm. IEEE Trans Rehabil Eng 1999;7(3):360–371.

96. Onders RP, DiMarco AF, Ignagni AR, Aiyar H, Mortimer JT. Mapping the phrenic nerve motor point: the key to a successful laparoscopic diaphragm pacing system in the first human series. Surgery 2004;136(4):819–826.

97. Schmit BD, Stellato TA, Miller ME, Mortimer JM. Laparoscopic placement of electrodes for diaphragm pacing using stimulation to locate the phrenic nerve motor points. IEEE Trans Rehabil Eng 1998;6(4):382–390.

98. Schmit BD, Mortimer JT. The effects of epimysial electrode location on phrenic nerve recruitment and the relation between tidal volume and interpulse interval. IEEE Trans Rehabil Eng 1999;7(2):150–158.

99. DiMarco AF, Onders RP, Kowalski KE, Miller ME, Ferek S, Mortimer JT. Phrenic nerve pacing in a tetraplegic patient via intramuscular diaphragm electrodes. Am J Respir Crit Care Med 2002;166(12 Pt 1):1604–1606.

100. DiMarco AF, Onders RP, Ignagni A, Kowalski KE, Stefan SL, Mortimer JT. Phrenic nerve pacing via intramuscular diaphragm electrodes in tetraplegic subjects. Chest 2005;127(2):671–678.

101. DiMarco AF, Altose MD, Cropp A, Durand D. Activation of intercostal muscles by electrical stimulation of the spinal cord. Am Rev Respir Dis 1987;136(6):1385–1390.

102. DiMarco AF, Supinski GS, Petro JA, Takaoka Y. Evaluation of intercostal pacing to provide artificial ventilation in quadriplegics. Am J Respir Crit Care Med 1994;150(4):934–940.

103. DiMarco AF, Budzinska K, Supinski GS. Artificial ventilation by means of electrical activation of intercostal/accessory muscles alone in anesthetized dogs. Am Rev Respir Dis 1989;139(4):961–967.

104. DiMarco AF, Takaoka Y, Kowalski KE. Combined intercostals and diaphragm pacing to provide artificial ventilation in tetraplegics. Arch Phys Med Rehabil 2005;86(6):1200–1207.

105. DiMarco AF, Kowalski KE, Geertman RT, Hromyak DR. Spinal cord stimulation: a new method to produce an effective cough in patients with spinal cord injury. Am J Respir Crit Care Med 2006;173(12):1386–1389.

106. Tzeng AC, Bach JR. Prevention of pulmonary morbidity for patients with neuromuscular disease. Chest 2000;118(5):1390–1396.

107. Winck JC, Goncalves MR, Lourenco C, Viana P, Almeida J, Bach JR. Effects of mechanical insufflation-exsufflation on respiratory parameters for patients with chronic airway secretion encumbrance. Chest 2004;126(3):774–780.

108. Bach JR. Prevention of respiratory complications of spinal cord injury: a challenge to model spinal cord injury units. J Spinal Cord Med 2006;29(1):3–4.

109. Bach JR. Alternative methods of ventilatory support for the patient with ventilatory failure due to spinal cord injury. J Am Paraplegia Soc 1991;14(4):158–174.

110. Bach JR. Noninvasive alternatives to tracheostomy for managing respiratory muscle dysfunction in spinal cord injury. Top Spinal Cord Injury Rehabil 1997;2:49–58.

111. Bach JR. Continuous noninvasive ventilation for patients with neuromuscular disease and spinal cord injury. Semin Respir Crit Care Med 2002;23(3):16–22.

112. Hess DR. Noninvasive ventilation in neuromuscular disease: equipment and application. Respir Care 2006;51(8):896–911.

113. Bach JR. A comparison of long-term ventilatory support alternatives from the perspective of the patient and care giver. Chest 1993;104(6):1702–1706.

114. Bach JR, Intintola P, Alba AS, Holland I. The ventilator-assisted individual: cost analysis of institutionalization versus rehabilitation and in-home management. Chest 1992;101(1):26–30.

115. Estenne M, DeTroyer A. The effects of tetraplegia on chest wall statics. Am Rev Respir Dis 1986;134(1):121–124.

The Interaction Between the Diaphragm and the Rib Cage

When muscles that inflate the rib cage contract (such as the external intercostal and neck accessory muscles), the negative pressure that is generated in the chest tends to pull the diaphragm cephalad (as seen in diaphragm paralysis). In that sense, the muscles cited are "deflationary" to the diaphragm. When the diaphragm contracts and moves caudad, the negative pressure that is generated in the thorax tends to pull the rib cage inward. In that sense, the diaphragm is "deflationary" to the rib cage. In other respects, as inferred from the force-balance analysis by Loring and Mead,[1] the diaphragm can act to elevate the rib cage, which is hinged at the sternum and spine.
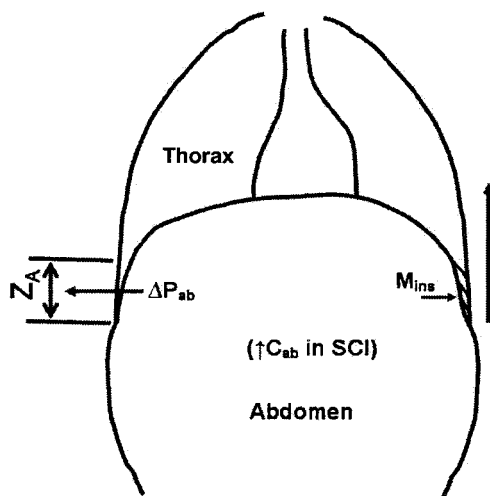
Principles related to the interaction between the diaphragm and the rib cage.

Z_A = zone of apposition of the diaphragm to the rib cage

ΔP_{ab} = change in abdominal pressure during inhalation

M_{ins} = muscular insertion of the diaphragm to the rib cage

C_{ab} = compliance of the abdomen

SCI = spinal-cord injury

At the lower level of the thorax, the diaphragm is in direct apposition to the rib cage through the "zone of apposition." When the diaphragm contracts, intra-abdominal pressure rises and elevates the rib cage through the zone of apposition. The zone of apposition is small in circumstances such as hyperinflation from emphysema or spinal-cord injury, in which the highly compliant abdomen results in low intra-abdominal pressure *at the onset* of inhalation.[2] This diminishes the effectiveness of the elevating action of the diaphragm on the rib cage. The magnitude of the increase in intra-abdominal pressure during inhalation also affects the extent to which the diaphragm elevates the rib cage. In spinal-cord injury, the abdomen is highly compliant.[2] This leads to a smaller-than-normal increase in intra-abdominal pressure *during* inhalation, which reduces the effectiveness of the action of the diaphragm on the rib cage through the zone of apposition.

The orientation of the muscular insertion of the diaphragm to the rib cage provides another mechanism by which diaphragm contraction contributes to rib-cage expansion. If abdominal pressure did not change at all during inhalation, the diaphragm would move caudad and it would not exert any inflationary action on the rib cage. To the extent that abdominal pressure rises as the diaphragm contracts, the muscle fibers of the diaphragm pull cephalad on and thereby expand the rib cage. Since the compliance of the abdomen is high in spinal-cord injury, abdominal-pressure changes are small during inhalation and the magnitude of the cephalad force on the rib cage is diminished. In summary, intra-abdominal pressure and changes in it can be viewed as the fulcrum for the elevating action of the diaphragm on the rib cage. The high compliance of the abdomen in spinal-cord injury diminishes the effectiveness with which the diaphragm elevates the rib cage, because both the zone of apposition and intra-abdominal pressure changes are small. An elastic abdominal binder tends to improve the action of the diaphragm on the rib cage. With the binder in place, intra-abdominal pressure is greater at the onset of diaphragm contraction, so the zone of apposition is larger. As well, with the binder in place, intra-abdominal pressure increases more during inhalation. This improves the action of the diaphragm on the rib cage, both via the zone of apposition and the muscle insertions (see text for details).

1. Loring SH, Mead J. Action of the diaphragm on the rib cage inferred from a force balance analysis. J Appl Physiol 1982;53(3):756-760.

2. Urmey W, Loring S, Mead J, Slutsky AS, Sarkarati M, Rossier A, Brown R. Upper and lower rib cage deformation during breathing in quadriplegics. J Appl Physiol 1986;60(2):618-622.

Discussion

Jubran: Congratulations to your group for highlighting the importance of the patient's being able to talk while on the ventilator. Based on your data, should we use a PEEP valve more often, in particular in patients who require prolonged mechanical ventilation for reasons other than neuromuscular disease? What do you recommend as a reasonable PEEP level? How long should we keep the patient on the PEEP valve?

Brown: The PEEP valve functions as an impedance to exhalation through the expiratory line of the ventilator. We recommend its use, although I have to acknowledge that we don't do it all that often in our unit, because there's someone around all the time. We recommend its use for safety purposes.

If you have a speaking valve in place and someone inflates the cuff of the tracheostomy tube, there will be a disaster. Or if phlegm accumulates around the deflated cuff or between the larynx and the tracheostomy tube and impedes gas going out through the larynx, then you can have a high-pressure problem in the respiratory system. A PEEP valve provides safety.

Let's say the PEEP is 10 cm H_2O. As soon as the pressure gets above 10 cm H_2O, gas can get out through the exhalation line of the ventilator and thereby protect the patient. Dean Hess told me that a speaking valve costs $65 at Massachusetts General Hospital, whereas a PEEP valve costs very little. How much PEEP to use is determined by how much gas you want to allow to go out through the ventilator, as opposed to through the larynx. We found, empirically, that 5 to 12 cm H_2O creates a really substantial improvement in speech, so that's what we use.

How long do we have the PEEP valve on? In patients I've managed we have it on all the time during the day. In patients with high-level spinal-cord injury we often take it off at night. And we partially inflate the cuff while the patient is sleeping, so that they get sufficient ventilation, but if something happens to them during the night, they'll still be able to talk and call for help if necessary.

Benditt: I would agree. I have a question about the mortality statistics that you quoted. You said that vascular disease is a very common cause of death. Is that also true in the younger population—the 20–30-year-olds? Or is that dominated by the older patients?

Brown: It is dominated by the older patients. Bauman and his colleagues at the Bronx Veterans Affairs Hospital have some interesting data with regard to that. Atherosclerosis seems to proceed more rapidly in people with spinal-cord injury than in others. And they are trying to figure out the mechanisms of that.[1,2] One possible mechanism is inactivity and lack of exercise. About 35% of patients with neurologically complete cervical-spinal-cord injury have diabetic glucose-tolerance test results. The mechanism of that is unclear. It's thought to be related to the autonomic nervous system. At the Bronx they found abnormalities in serum lipids in individuals with spinal-cord injury. Cardiovascular death is distinctly common in these individuals years after the injury has occurred.

REFERENCES

1. Bauman WA, Adkins RH, Spungen AM, Waters RL. The effect of residual neurological deficit on oral glucose tolerance in persons with chronic spinal-cord injury. Spinal Cord 1999;37(11):765–771.
2. Bauman WA, Spungen AM. Disorders of carbohydrate and lipid metabolism in veterans with paraplegia or quadriplegia: a model of premature aging. Metabolism 1994;43(6):749–756.

Hill: My question has to do with dyspnea. I was intrigued by the observation about wheelchair users and their greater dyspnea, because that's the opposite of what I've seen in other quad-riplegic states. I've always been struck by how minor a complaint dyspnea is among patients with, say, Duchenne muscular dystrophy, who have no more or less capacity than the ones you're describing, and yet these patients, who are often on noninvasive ventilation, almost invariably deny dyspnea. They say, "No, my breathing is fine. I just might have a little trouble getting out as many words as I'd like." And some of them are big talkers, too. They love to talk. And they don't complain of dyspnea when they're talking.

So I'm wondering what's different about them. I suppose one possibility is that their P_{aCO_2} is higher. If you're talking about patients using wheelchairs and ventilators, whose P_{CO_2} is maintained at a normal or even low level, small changes in their breathing patterns might affect their sensation. I know that Bob Banzett has shown that very minute changes in P_{aCO_2} can bring about big changes in the sensation of dyspnea.[1,2] On the other hand, most of these patients using noninvasive ventilation for chronic respiratory failure sustain higher P_{CO_2}, which might blunt their sensitivity. So that's one possibility. What are your thoughts? Why the difference? It has been pretty striking in my experience.

REFERENCES

1. Banzett RB, Lansing RW, Brown R, Topulos GP, Yager D, Steele SM, et al. Air hunger from increased P_{CO_2} persists after complete neuromuscular block in humans. Respir Physiol 1990;81(1):1–17.
2. Bloch-Salisbury E, Shea SA, Brown R, Evans K, Banzett RB. Air hunger induced by acute increase in P_{CO_2} adapts to chronic elevation of P_{CO_2} in ventilated humans. J Appl Physiol 1996;81(2):949–956.

Brown: Well, it's intriguing, but let me make sure that I'm clear. We studied *motorized*-wheelchair users, not all wheelchair users. These people were so impaired that they couldn't propel a wheelchair on their own. They were severely impaired. Among those individuals, 18% complained of short-

ness of breath while talking. They may be more impaired than some of your subjects.

Hill: But they're not. I'm talking about quadriplegics. They're advanced neuromuscular patients who are in wheelchairs and can't move much of anything.

Brown: Well, it would be fascinating to try to work out that puzzle.

Hill: In fact, it's the less disabled ones who tend to complain of dyspnea. For example, in the ALS [amyotrophic lateral sclerosis] group, you hear people complain of dyspnea if they can still walk around, but the respiratory muscles are impaired; but they *don't* complain of dyspnea if their limb muscles are paralyzed.

Brown: I don't think I have the answer. I think, though, that to study it you have to give them a questionnaire in which they give you specific answers to specific questions. It would be interesting to contrast the 2 groups.

Lechtzin: I want to add to the debate on dyspnea in these patients. I agree with Nick, because I've seen muscular dystrophy patients who have almost no respiratory complaints and no dyspnea but are wheelchair-bound,

completely paralyzed for all intents and purposes, and have very low lung capacities and elevated CO_2. Though this is completely speculation, I've always attributed it to the time frame that this is developed over. I see adults with Duchenne muscular dystrophy who have developed respiratory-muscle weakness over decades, whereas somebody with a spinal-cord injury developed diaphragm paralysis very abruptly, and now you're following them, even if it's just a year or 2 later. I wonder if that has a role?

Hill: Of course, you have the ALS group, too.

Lechtzin: Which I have found to be somewhere intermediate. Some ALS patients have dyspnea, some don't; and they tend to be more dyspneic than the muscular dystrophy patients.

Brown: The patients we studied were 1 to 20 years after injuries, so it was hardly acute. And 83% of them were *not* dyspneic while they were talking. We wondered whether it was some adaptive strategy they had figured out. We have yet to study that.

Benditt: One of the things I've noticed is that some of my patients, in attempting to prolong their speech, use

a higher-pitched voice. I assume that it relates to narrowing the orifice in the larynx and thus prolonging the breath. Is that something that you've noticed or studied?

Brown: I have not noticed higher-pitched speech in these patients, but I will look for it in the future. It's an interesting idea.

Upinder Dhand: In tracheostomized patients who have not spinal-cord injury but other neuromuscular disorders such as spinal-muscle atrophy, will the same principle of modification of the inspiratory pressure and PEEP apply for improving their speech?

Brown: Yes, but problems arise in disorders that affect oropharyngeal or laryngeal function. In such cases, even though you change the ventilator settings, they don't have the muscular or neurocontrol with which to phonate. On the other hand, I've had numerous patients with ALS who could barely speak without a ventilator, and then, following the tracheostomy and adjustment of the ventilator, they could speak much better, until they deteriorated further.