Cervical Accessory Respiratory Muscle Function in a Patient with a High Cervical Cord Lesion*

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The function of the accessory respiratory muscles (ARM) of the neck were studied in a quadriplegic patient suffering from a C_{3-4} lesion of the cervical spinal cord. Sub-total expiratory paralysis resulted in an essentially complete loss of expiratory reserve volume such that residual volume and functional residual capacity were equal (RV = FRC). Tidal volume and vital capacity were severely reduced. Being extrinsic to the chest, the ARM of the neck functioned independently of changes in thoracic gas volume; however, their performance appeared posture-dependent, and was less efficient in the sitting position. Despite the fact that the ARM preferentially expanded the upper part of the ribcage, significant V/Q mismatch did not seem to occur. Voluntary use of glossoharyngeal breathing (GPB) greatly enhanced the patient’s ventilatory capability. Flow volume data during GPB documented the efficiency of the glossoharyngeal muscles, which function as a positive pressure pump to force air into the lungs.

Quadriplegia from cervical lesions above the C_{3} level is associated with severe loss of respiratory motor function, including that of the diaphragm, the expiratory muscles of the abdomen, and the musculature of the ribcage and the shoulder girdle. The only muscles that escape paralysis are those innervated by the cranial nerves and the first two spinal nerves. Traumatically-acquired high cervical cord lesions often prove fatal due to the inability of the accessory muscles of respiration to take over the work of breathing acutely; in contrast, paralysis of the respiratory muscles resulting from transverse myelitis, though rapidly progressive, often occurs in patients already hospitalized, patients who can be supported by mechanical ventilation during the acute phase of their disease. Furthermore, in this disease, some functional recovery can be expected in convalescence, usually from partial return of spinal cord function. In addition, adaptation of the accessory muscles of the neck may occur and may permit a level of ventilation adequate for gas exchange at rest.

Since the level of ventilation required at rest is modest, patients with vital capacities (VC) as low as 200 ml have been shown capable of breathing unassisted for periods up to 45 minutes.¹ Their voluntary ventilation can be increased further with the technique of glossoharyngeal breathing (GPB), a learnable process utilizing the tongue and other muscles of the bucco-laryngopharyngeal area for supplemental inspiratory volume.²³ These quadriplegic patients represent an unique pathophysiological condition wherein only the accessory respiratory muscles of the neck and head are functioning. The effect of the accessory musculature on the respiratory system is not masked by the diaphragm, the ribcage, or the abdominal musculature as it is in other disease states. We analyzed the function of the accessory respiratory muscles in a patient with quadriplegia secondary to transverse myelitis at the C_{3-4} level, who had been taught the technique of glossoharyngeal breathing. The purposes of the investigation were to determine the respiratory mechanics characteristic of ventilation by the ARM and to define the flow-volume relationships during GPB.

**Material and Methods**

The patient studied was a 30-year-old woman who, in 1958, at age 11, suffered an acute attack of idiopathic transverse myelitis at the C_{3-4} level of the spinal cord. Initially, she required continuous use of a Drinker respirator, but she gradually regained the ability to breathe unassisted via a permanent indwelling tracheostomy tube. In 1957, she learned the technique of glossoharyngeal breathing at the Rancho Los Amigos Center in Downey, California. She also required...
an abdominal corset to avoid dyspnea in the sitting position, but she reported no subjective benefit from the corset when prone or supine.

Over the following 19 years, she had numerous episodes of respiratory infections, despite physical therapy as an out-patient and a detailed pulmonary toilet regimen at home.

In November, 1974, she was admitted to the University Hospital in San Diego with complaints of increased shortness of breath, cough, and difficulty in handling secretions. Physical examination disclosed rapid, shallow respirations; blood pressure, pulse rate, and temperature were normal. The sternocleidomastoid muscles were hypertrophied bilaterally and contracted rhythmically with tidal breathing. An indwelling tracheostomy tube was functioning well. The chest was slightly asymmetrical with moderate thoracolumbar scoliosis. On inspiration, there was minimal chest wall expansion, which seemed limited to the upper rib cage; with glossoptharyngeal breathing, chest expansion was much greater. Diaphragmatic excursion was not detected by either inspection or careful percussion. Coarse ronchi were heard at both bases. Cardiac examination was normal. The abdominal musculature was flaccid; no contractions were detectable when the patient was instructed to perform a maximal inspiratory maneuver. An elastic abdominal corset with metallic stays was arranged around the waist to minimize her dyspnea in the sitting position. Neurologic examination disclosed motor and sensory losses corresponding to a complete functional interruption of the spinal cord at C7 level. Roentgenograms of the chest were normal aside from mild scoliosis of the thoracic spine.

The patient improved with physical therapy and postural drainage; her dyspnea and sputum production decreased, and she was discharged. Six weeks later she underwent pulmonary function testing, with her tracheostomy tube occluded. Static lung volumes and expiratory flow rates were measured by water-filled spirometer; functional residual capacity was estimated by helium dilution (the patient could not tolerate testing by body plethysmography). Static lung volumes were measured in sitting and supine positions, with and without the abdominal corset. Flow-volume loops were recorded with an Ohio 840 dry spirometer during "normal" breathing and during GPB. Static elastic recoil of the lung was measured with an anode* esophageal balloon via a Statham PM5 transducer connected to an Electronics-for-Medicine DR-8 oscilloscope. Lung volume was obtained by integration of the flow signal from a Fleisch pneumotachygraph. Arterial blood gas volumes were measured with an Instrumentation Laboratories IL-213 blood gas analyzer. Maximal inspiratory and expiratory pressures were measured according to the technique of Hyatt et al.** Electromyographic studies were done during spontaneous breathing for the sternocleidomastoid muscles, and during percutaneous stimulation of the phrenic nerves for the diaphragm, using a Meditron Model 312 electromyograph.**

RESULTS

Spirometric data and arterial blood gas levels are summarized in Table 1. During spontaneous breathing with room air, arterial blood gas levels were within normal limits except for mild hypocapnia. In the sitting position, with the abdominal corset fastened, all static lung volumes were well below normal values predicted for body size and age.7 The reduction was most pronounced for ERV, IC, VC (single effort) and TLC; decrease in RV was modest, resulting in an elevated RV/TLC ratio. When the patient lay supine, ERV remained extremely reduced; RV and FRC decreased. Removing the abdominal corset resulted in increased FRC and RV in both positions.

Glossoptharyngeal breathing allowed the patient to triple her VC and increase TLC by 65 percent. Expiratory flow rates were lower than predicted with FEV1.0 of 75 percent of VC and an MMEF25-75 of 0.7 L/sec.

At a TLC of only 1,571 ml BTPS (33 percent of predicted), esophageal pressure was —12.0 cmH2O. This was more than twice the negative pressure found by Turner et al in a group of normal subjects of the same age, at 50 percent of achievable TLC. Although the transpulmonary pressures at lung volumes as low as 33 percent of predicted TLC have not been reported in a large number of normal subjects, this finding suggests significantly increased static elastic recoil of the lungs.

Maximal inspiratory pressure was low; maximal expiratory pressure was normal when corrected for lung volume.9 There was no difference between the maximal expiratory pressures measured at two lung volumes (1,571 and 2,591 ml BTPS) achieved at "normal" TLC and at GPB-induced TLC, respectively.

Flow-volume curves during tidal breathing and during normal maximal inspiration, maximal expiration, and during GPB are shown in Figure 1. Although maximal expiratory flow was low, as expected from her small static lung volumes, there was no significant excavation of the effort-independent zone of the expiratory flow-volume curve from a TLC achieved by GPB, suggesting absence of significant airway obstruction.10 Though a small (less than 100 ml) ERV was observed by spirometry, none was recorded on the flow-volume loops.

During spontaneous breathing, EMG output recorded from the sternocleidomastoid muscles was typical of an inspiratory muscle, with bursts of action potentials synchronous with the inspiratory phase of each breath, arranged in a crescendo pattern before the following expiratory phase. The intercostal muscles were electromyographically silent during spontaneous breathing, as were the abdominal muscles, the muscles of the shoulder girdles, and the right hemidiaphragm. Electromyographic evaluation of the left side showed an essentially silent hemidiaphragm, although at intervals, intermittent action potentials of low amplitude were seen. Fluoroscopy was not performed.

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**Crescent Engineering and Research, El Monte, California.
Table 1—Pulmonary Function Data

<table>
<thead>
<tr>
<th></th>
<th>Predicted Sitting</th>
<th>Observed Sitting†</th>
<th>% predicted†</th>
<th>Observed Supine</th>
<th>Observed Sitting, Using GPB</th>
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<tbody>
<tr>
<td>PaO₂, mm Hg</td>
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<tr>
<td>PaCO₂, mm Hg</td>
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<td>pH, units</td>
<td></td>
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<tr>
<td>Tidal volume, ml</td>
<td>533</td>
<td>327 (299)</td>
<td>61 (42)</td>
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<tr>
<td>Vital capacity, ml</td>
<td>3,334</td>
<td>600</td>
<td>18</td>
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<tr>
<td>Functional residual capacity, ml*</td>
<td>2,528</td>
<td>1,020 (1,477)</td>
<td>40 (58)</td>
<td>536 (788)</td>
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<tr>
<td>Residual volume, ml</td>
<td>1,417</td>
<td>971 (1,379)</td>
<td>68 (97)</td>
<td>454 (706)</td>
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<tr>
<td>Total lung capacity, ml</td>
<td>4,760</td>
<td>1,571</td>
<td>33</td>
<td></td>
<td>2,591</td>
</tr>
<tr>
<td>RV/TLC, %</td>
<td>35</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Expiratory reserve volume, ml</td>
<td>1,111</td>
<td>49 (98)</td>
<td>4 (9)</td>
<td>82 (82)</td>
<td></td>
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<tr>
<td>Inspiratory capacity, ml</td>
<td>2,232</td>
<td>551</td>
<td>25</td>
<td></td>
<td>1,571</td>
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<tr>
<td>FEV₁, % of FEV</td>
<td>80</td>
<td></td>
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<tr>
<td>Maximum mid-expiratory flow (25-75%), L/sec</td>
<td>3.0</td>
<td>0.7</td>
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<td>VEMS, L/min</td>
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<td>7.6</td>
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<td>Transpulmonary pressure cm H₂O, at TLC***</td>
<td>−3.8 ± 5.3</td>
<td>−12.0</td>
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<tr>
<td>Maximum inspiratory pressure, cm H₂O**</td>
<td>89 ± 19</td>
<td>20</td>
<td></td>
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<tr>
<td>Maximum expiratory pressure, cm H₂O‡</td>
<td>48 ± 25</td>
<td>66</td>
<td></td>
<td>60</td>
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</tbody>
</table>

*Measured by helium dilution. †Values in parentheses were obtained without elastic abdominal corset; all other values were obtained with corset fastened. **Measured sitting at FRC (21.4% of predicted TLC); predicted value from Ref. 9 for females at normal FRC. ††Measured at TLC (33% of predicted TLC); predicted value from Ref. 9 for females at 40% of TLC. ***Predicted values derived from data on normal 30-year-old subjects at 50% of TLC (Table 3, Ref. 8).

**DISCUSSION**

The data indicate a marked restrictive defect secondary to paralysis of respiratory muscles, with a significant degree of lung stiffening; the latter may have resulted from years of inadequate inflation of the lungs, together with repeated pulmonary infections.

The decrease in FRC was consistent with a stiffening of the lung parenchyma. FRC represents an equilibrium between the inward recoil of the lung and the outward recoil of the relaxed chest;11,12 since the outward recoil of the relaxed chest wall is not thought to be significantly affected by muscular paralysis,12,17 reduction in FRC indicates an increase in lung recoil. The esophageal pressure measurements also indicated a decrease in lung compliance; however, the major cause for the severe restriction of her static lung volumes was neuromuscular, as indicated by the low maximum inspiratory pressure measurements. Also, patients with diseases associated exclusively with loss of compliance of the lung, such as sarcoidosis and diffuse interstitial fibrosis, usually exhibit proportional losses in TLC and in FRC; our patient had considerably greater loss of TLC (33 percent of predicted) than FRC (58 percent of predicted), indicating a loss of inspiratory capacity out of proportion to loss of compliance of the lung.

**Expiratory Functions of Cervical Accessory Muscles**

In the absence of contributions from the major expiratory muscles of the abdomen and rib cage, the observed expiratory pressure and volume changes were achieved by the accessory expiratory muscles of the neck. Under these circumstances, the expiratory reserve volumes measured in the sitting and supine positions were markedly reduced; when the abdominal corset was removed, the ERV in-
increased minimally. Thus, FRC approximated RV, and IC essentially equalled VC. This is clearly illustrated by the flow-volume curves during the tidal breathing and during the FVC maneuver (Fig 1A); these were performed with the abdominal corset fastened. The curves have a common end-expiratory point on the volume axis representing both FRC and RV. Such changes in the subdivisions of lung volume are similar to those reported in patients with poliomyelitis.12-17 The small (49 ml) ERV measured in the sitting position by spirometry (Table 1) was not seen on the flow-volume curve (Fig 1A); the reasons for this difference are not clear, but may be related to a small, variable volume contribution from the buccal cavity.

The maximal expiratory pressures of 60 and 66 cmH2O developed by the patient at lung volumes of 2,591 and 1,571 ml BTPS, respectively, apparently were not produced by the accessory respiratory muscles, the weakness of which was indicated in the measurements of static lung volumes. Instead, these pressures apparently were generated inside the buccal cavity with the cheek musculature against the closed glottis when the patient was told to expire forcefully into Hyatt's device.44 The maximal "expiratory" pressure was independent of lung volume, consistent with the fact that the extrathoracic buccal muscles do not depend upon lung volume for their length-tension relationship; changes in thoracic gas volume, therefore, should not condition the maximum pressure they can generate.

The decrease in ERV and the increase in RV/TLC ratio observed in the sitting position without the abdominal corset might be explained by collapse of small airways, with the resultant trapping of intrathoracic gas. This did not appear to be the case. Her RV was reduced in the sitting position by application of the abdominal corset; and declined further when she lay supine, with or without abdominal corset. Thus, the marked ERV reduction is attributable to the respiratory muscle paralysis.

**Inspiratory Function of Cervical Accessory Muscles**

In this patient, the cervical accessory muscles of the neck were completely responsible for her inspiratory effort. Such effort was sufficient to maintain normal gas exchange in the resting state; in fact, moderate chronic alveolar hyperventilation was present. However, this degree of ventilation was achieved with almost all of the respiratory capability of the accessory muscles, as any added mechanical challenge to the respiratory system, such as increased amount or viscosity of secretions, or any increase in CO2 production from an elevated metabolic rate, historically could not be met by an appropriate increase in alveolar ventilation and therefore resulted in CO2 retention.

Despite these limitations, inspiratory function was well-maintained, due to hypertrophy of the accessory respiratory muscles and to the ability of the patient to perform glosso-pharyngeal breathing to supplement ventilation.

Glosso-pharyngeal breathing was first discovered by a patient with poliomyelitis at the Ranch Los Amigos Hospital in Downey, California. It consists of using coordinated movements of the tongue, cheeks, and soft palate to pump a bolus of air into the lungs using the glottis as a one-way check valve.
Since the average volume of the mouth and throat is small, repeated GPB strokes are necessary to achieve an inspiratory volume of significant magnitude. Because of its resemblance to the respiratory movements of amphibians, GPB has been called “frog breathing.” When this peculiar breathing pattern was carefully analyzed, a standardized technique was developed and subsequently was taught to many patients. Our patient's success with GPB is illustrated by her ability to increase her VC three-fold and to support herself without assisted ventilation for long periods.

Analysis of the flow-volume relationships in our patient revealed interesting information concerning her GPB performance (Fig 1B). GPB increased the IC from 500 ml to almost 1,400 ml. This increase in inspiratory volume was achieved with 20 consecutive inspiratory strokes, requiring about 15 seconds. The first inspiratory stroke from the right (i.e., starting at FRC) probably resulted from a “tug” of spontaneous inspiration. The next 14 GPB maneuvers were similar, delivering a bolus equal to approximately 75 percent of the initial spontaneous inspiration, averaging 80 ml. The inspiratory flow of each bolus increased during the inspiratory stroke, then dropped abruptly to zero. Peak flow occurred at the end of the inspiratory stroke volume, at which point the glottis presumably closed and flow fell to zero. The characteristics of these 14 strokes indicate that the performance of the accessory muscles during GPB is independent of overall lung volume: they generate a uniform inspiratory volume during inflation of the lungs from FRC to approximately 1 liter above FRC. Such independence reflects the fact that they are extrinsic to the chest, and lung volume does not affect their length-tension relationships. In contrast, the function of inspiratory muscles intrinsic to the chest, such as the diaphragm, is influenced by thoracic volume.

The regularity of the 14 GPB curves also indicates that the inspiratory volume depends upon the volume of the buccopharyngeal cavity, which is constant despite a decreasing gradient across the glottic check-valve. With progressive filling of the thorax during GPB, intrathoracic pressure increases progressively, thus narrowing the pressure gradient between pharynx and lungs. The 90 cmH2O of positive pressure that the accessory muscles in this patient could generate in the buccal cavity was enough to maintain the pressure gradient across the glottis until the last five GPB strokes. During these last five maneuvers, the GPB mechanism reached its limits due to the pressure increase within the respiratory system. At this point, the inspiratory volume became markedly reduced and irregular, and expiratory flow occurred.

The variation in the peak flow of each of the 20 inspiratory strokes is also of interest. Inspiratory peak flow increases from the first to the tenth strokes when it reached its maximum; then it decreased progressively. The 20 individual inspiratory peak flow points describe a discontinuous curve which is similar to the maximal inspiratory flow-volume curve obtained in normal subjects, where maximal inspiratory flow also occurs near the middle of the vital capacity.

Posture-Dependence of Cervical Accessory Muscle Function

Our patient experienced dyspnea in the sitting position which was alleviated by fastening her abdominal corset or by laying supine. When supine, she experienced no benefit from the abdominal corset. This posture-related dyspnea may have been related to the weight of the abdominal contents; in the sitting quadriplegic patient, the flaccid sagging abdomen pulls on the rib cage and directly opposes the function of the cervical accessory muscles to lift the chest in inspiration. By supporting the weight of the upright abdomen, the corset decreases this opposing force. However, in the supine position, the weight of the abdominal contents no longer opposes the cephalocaudal force developed by the cervical accessory muscles and the corset cannot decrease the load applied to these muscles.

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REFERENCES
9. Cook CD, Mead J, Orzaless MM: Static volume-pressure

ANNOUNCEMENTS

Seventh Annual Fleischner Society Symposium

The Fleischner Society, a multidisciplinary international organization devoted to the study of the chest, will present its Seventh Annual Symposium on Diseases of the Chest in Miami Beach, February 27-

March 2. For information, contact Ms. Lucy Kelley, Department of Radiology, Jackson Memorial Hospital, Miami, Florida 33154.

Seventh Aspen Radiology Conference

The Seventh Annual Aspen Radiology Conference will be held February 28-March 4 at the Aspen Institute for Humanistic Studies. For information, write

Emanuel Salzman, M.D., Conference Chairman, Division of Radiology, Beth Israel Hospital, Denver 80204.