Dyspnea after polio can occur for a variety of reasons, including neuromuscular disease and upper airway abnormalities resulting from prolonged intubation, including tracheal stenosis, tracheomalacia, and vocal cord paralysis. Routine studies such as spirometry and maximum voluntary ventilation (MVV) measurements can give similar results in these conditions. We present a 50-year-old woman who as a child developed poliomyelitis that required tracheostomy and negative pressure ventilation. Thirty-nine years later, she developed breathlessness with normal spirometry but decreased MVV. The flow volume loop showed flattening of the inspiratory and expiratory limbs, consistent with a fixed upper airway obstruction or neuromuscular weakness. Exercise testing with measurement of exercise flow volume loops and respiratory pressures was performed. The patient was ventilatory limited with increasing end-expiratory lung volume through exercise. Flow volume loops confirmed flow limitation. Respiratory pressures did not change after maximal exercise. Further evaluation confirmed left vocal cord paralysis and tracheomalacia. This patient demonstrates that the causes of dyspnea after poliomyelitis can be multifactorial, and that routine evaluation may fail to elucidate the limiting factor. In this case, exercise testing provided valuable insight into the limiting factor for this patient and provided useful data for counseling and for further management.

(Chest 1994; 105: 777-81)

CAO = chronic airflow obstruction; CPET = cardiopulmonary exercise testing; EELV = end-expiratory lung volume; HRR = heart rate response; IC = inspiratory capacity; MVV = maximum voluntary ventilation; Pdi = transdiaphragmatic pressure; Ppl = pleural pressure; UAO = upper airway obstruction

Neuromuscular disease or upper airway obstruction can present with breathlessness on exertion. Routine physiologic studies such as spirometry or maximum voluntary ventilation (MVV) may provide clues to their presence but unfortunately often give similar results making it difficult to separate the contribution of these diseases. Cardiopulmonary exercise testing (CPET) has been used in the evaluation of patients with dyspnea, but most of these studies have been in patients with cardiac disease, chronic airflow obstruction (CAO), or pulmonary parenchymal disorders. There are few reports of exercise studies in patients with tracheal stenosis or in patients with neuromuscular disease. The following case demonstrates the utility of CPET in discerning the cause of dyspnea in a patient who suffered from both neuromuscular respiratory disease and upper airway obstruction following remote poliomyelitis.

CASE REPORT

A 50-year-old white woman presented with acute poliomyelitis at age 10 years. Severe bulbar symptoms developed and required tracheostomy and negative pressure mechanical ventilation for approximately 6 months. The patient recovered with no discernible peripheral muscle weakness. She did note mild, subjective abdominal muscle weakness. She remained quite active with no limitation until age 34 years when she noted mild shortness of breath with exertion. Pulmonary function studies supported a central airway obstruction (Table 1). An otolaryngologist confirmed normal vocal cords but a tracheal abnormality. Tracheogram demonstrated narrowing of the trachea within 4 cm of the carina with "dynamic narrowing of the trachea to less than 50 percent of the normal tracheal lumen" consistent with tracheomalacia.

Six years later, the patient was noted to have "tracheal stenosis at the glottic chink" by bronchoscopy. However, she did well until 1 year prior to presentation when she again reported worsening dyspnea with exertion and was evaluated at the University of Michigan. She was a life-long nonsmoker and had no other medical problems or allergies. There was no history of major surgical procedures other than the prior tracheostomy. She complied with a regular exercise program (daily aerobic exercise, including walking) that was unchanged over the several weeks preceding this evaluation. She was receiving no medication at the time of the study. Review of systems identified no symptoms of sleep disturbance, daytime hypersomnia, or morning headaches.

The patient's weight (61.8 kg) was appropriate for her height (165 cm). Physical examination revealed no wheezing, stridor, peripheral muscle weakness, or fasciculation. No evidence of abnormal musculoskeletal alignment or limb length discrepancy was noted. Tracheal tomography demonstrated no fixed stenotic lesion. Laryngoscopy revealed paralysis of the left true vocal cord that was fixed in the midline position. There was also decreased abduction of the right true vocal cord with only a 3 to 4 mm glottic chink. A plot of maximal expiratory and inspiratory flow and volume (Med Science, St. Louis, Mo), created from multiple vital capacity maneuvers performed by the patient with varying effort, demonstrated flattening of both the inspiratory and expiratory limbs

*From the Division of Pulmonary and Critical Care Medicine (Drs. Becker and Martinez, Ms. Harper, and Mr. Graf), Department of Internal Medicine (Dr. Knobil), and the Department of Otolaryngology-Head and Neck Surgery (Dr. Wolf), University of Michigan Medical Center, Ann Arbor. Supported by grants from the NIH (P50-HL46487 and GCRC-MCAP 3 M01 RR 00042-3553)

Manuscript received February 26, 1993; revision accepted June 15

Reprint requests: Dr. Martinez, BHI 372, 1500 East Medical Center Drive, Ann Arbor, MI 48109-0026

CHEST / 105 / 3 / MARCH, 1994 777
Table 1—Serial Pulmonary Function Tests in a Patient With a History of Poliomyelitis

<table>
<thead>
<tr>
<th>Parameter</th>
<th>4/76</th>
<th>11/84</th>
<th>3/87</th>
<th>9/92</th>
</tr>
</thead>
<tbody>
<tr>
<td>FVC, % predicted</td>
<td>92</td>
<td>79</td>
<td>81</td>
<td>90</td>
</tr>
<tr>
<td>FEV1, % predicted</td>
<td>99</td>
<td>92</td>
<td>89</td>
<td>88</td>
</tr>
<tr>
<td>FEV1/FVC</td>
<td>85</td>
<td>92</td>
<td>84</td>
<td>74</td>
</tr>
<tr>
<td>TLC, % predicted</td>
<td></td>
<td></td>
<td>105</td>
<td></td>
</tr>
<tr>
<td>MVV, L/min</td>
<td></td>
<td></td>
<td>53</td>
<td>47</td>
</tr>
</tbody>
</table>

supporting a fixed or both variable intrathoracic and extrathoracic upper airway obstruction (Fig 1). Spirometric parameters were expressed according to the predicted values published by Morris et al.* Chest radiograph was normal.

Cardiopulmonary Exercise Testing

Cardiopulmonary exercise testing was a symptom-limited study performed on an electronically braked, calibrated cycle ergometer while breathing into a calibrated pneumotach and metabolic cart (Collins CPXII, Warren E. Collins, Inc, Braintree, Mass) (Table 2). After an initial baseline period of resting data collection and unloaded cycling, work rate on the ergometer was increased by 20-W increments every minute until exhaustion. The exercise test was terminated by the patient due to severe breathlessness (see below). Arterial oxygen saturation was monitored by finger and ear pulse oximetry (OXimeter, Radiometer, Copenhagen, Denmark). Twelve-lead electrocardiograms were recorded every minute, as was noninvasive measurement of blood pressure. The predicted values of Wasserman et al.8 were used for exercise oxygen consumption.

Heart rate response (HRR) was calculated using the following formula: HRR = (HRmax - HRrest)/Vo2max - Vo2rest).14

Anaerobic threshold was estimated noninvasively using the break point of ventilatory equivalents for VO2 and VCO2 and the V-slope method.12 Dyspnea was estimated using a Borg scale13 that ranges from 0 for no breathlessness to 10 corresponding to greatest breathlessness. This measurement was made at rest and at the end of each minute of exercise.

Functional residual capacity (FRC) at rest and during exercise was calculated using inspiratory capacity (IC) measurements at rest and during each workload. An impedance plethysmograph calibrated using the isovolume method14 was used to simplify the identification of the end-expiratory position prior to the performance of an IC. In addition, the ventilatory flow signal was displayed on a computer screen in real-time to better identify the moment of zero flow. This technique has been used extensively in the quantification of changes in end-expiratory lung volume (EELV) with exercise in patients with CAO.15 An adequate IC was confirmed by monitoring minimal pleural pressure (see below).16 The flow signal during exercise was stored on a personal computer (IBM PS/2) and integrated at a later point, allowing the construction of tidal flow-volume loops during the course of exercise. End-expiratory lung volume for correct placement of these loops was calculated from the IC maneuver.

Respiratory Muscle Testing

Pleural (Ppl) and gastric (Pg) pressures were measured using thin-walled balloons placed transnasally in the middle third of the esophagus and the stomach, respectively. A separate transducer (Validyne Co, Northridge, Calif) measured each pressure while a third transducer measured mouth pressure. The calibrated output of all pressure measurements was continuously displayed in real time on a computer screen. All measurements, including the vital capacity maneuvers, were made with the patient in the sitting position. Transdiaphragmatic pressure (Pdi) was electronically calculated as the difference between Pg and Ppl. DPdi was calculated as the difference between end-inspiratory and end-expiratory Pdi. End-inspiration and end-expiration were defined at the point of zero flow. Maximal transdiaphragmatic pressure (Pdimax) was measured from FBC with a maximal inspiratory effort against a partially closed shutter while the patient was at rest. The patient was asked to maximally expand the chest and abdomen and was coached in the performance of this maneuver. At least three measurements using previously published methods16 and the maximal DPdi were reported (Table 3). Transdiaphragmatic measurements were also recorded during sharp, maximal sniffs (sniff Pdi) as described by Mier-Jedrzejowicz et al.17 These measures were repeated immediately after the end of exercise.

RESULTS

The results of CPET are shown in Table 2. Peak VO2 was in the normal range as was anaerobic threshold (AT) and HRR. This suggests an appropriate cardiac response to exercise. Ventilatory response to exercise appeared to be mechanically limited with a maximum

Figure 1. Maximal flow-volume loop (solid line); resting, tidal flow-volume loop (dot-dash); and peak exercise, tidal flow-volume loop (dashed line).
Table 2 — Maximum Cardiopulmonary Exercise Testing Results*

<table>
<thead>
<tr>
<th></th>
<th>Peak</th>
<th>Predicted</th>
<th>% Predicted</th>
</tr>
</thead>
<tbody>
<tr>
<td>Time, min</td>
<td>8.00</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Power, W</td>
<td>120</td>
<td></td>
<td></td>
</tr>
<tr>
<td>VO2, ml/kg/min</td>
<td>20.7</td>
<td>24.1</td>
<td>86</td>
</tr>
<tr>
<td>AT, L/min</td>
<td>1.0</td>
<td>&gt; 0.6</td>
<td></td>
</tr>
<tr>
<td>HR, bpm</td>
<td>149</td>
<td>170</td>
<td>88</td>
</tr>
<tr>
<td>HRR</td>
<td>46.1</td>
<td>&lt; 50</td>
<td></td>
</tr>
<tr>
<td>O2 pulse, ml/beat</td>
<td>8.8</td>
<td>8.8</td>
<td>100</td>
</tr>
<tr>
<td>VE, L/min</td>
<td>42.1</td>
<td>47.3</td>
<td>89</td>
</tr>
<tr>
<td>VE/VO2</td>
<td>32.9</td>
<td>&lt; 40</td>
<td></td>
</tr>
<tr>
<td>VE/VCO2</td>
<td>29.6</td>
<td>&lt; 35</td>
<td></td>
</tr>
<tr>
<td>VT/VC, %</td>
<td>41</td>
<td>&lt; 55</td>
<td></td>
</tr>
<tr>
<td>f, br/min</td>
<td>30</td>
<td></td>
<td></td>
</tr>
<tr>
<td>SaO2, %</td>
<td>98</td>
<td></td>
<td></td>
</tr>
<tr>
<td>PetCO2, mm Hg</td>
<td>41</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*VO2 = oxygen consumption; AT = anaerobic threshold; HR = heart rate; HRR = heart rate response; O2 pulse = oxygen pulse; VE = minute ventilation; VCO2 = carbon dioxide production; f = respiratory rate; SaO2 = oxygen saturation; and PetCO2 = end-tidal carbon dioxide.

VE that approached the patient’s measured MVV. End-tidal O2, CO2, and oxygen saturation remained in the normal range throughout exercise. Figure 1 illustrates the composite maximal, resting tidal, and peak exercise tidal flow-volume loops. It is clear that EELV during peak exercise rose significantly. Furthermore, peak expiratory flow overlapped the maximal expiratory flows at this elevated lung volume. The peak inspiratory flows approached those of the maximal efforts. Although not shown graphically, transpulmonary pressure during exercise surpassed that required to achieve maximal expiratory flows during the graded maximal vital capacity efforts.

Table 3 demonstrates significant baseline respiratory muscle weakness. Both the Pdimax and sniff Pdi were well below the expected range. After maximal exercise there was only a minimal drop in sniff Pdi. Figure 2 presents the increase in dyspnea that occurred throughout the course of exercise (Fig 2, right). It is clear that breathlessness was severe at peak exercise and supports this as the major symptom-limiting exercise. Figure 2, left, shows a steady drop in Ppl throughout exercise with a rise in ΔPdi. These two did not reach the maximal values measured at rest.

**DISCUSSION**

Cardiopulmonary exercise testing has been used in the evaluation of patients with dyspnea, but there are few data in patients with upper airway obstruction or neuromuscular diseases. This patient illustrates the difficulties encountered in determining the cause of dyspnea using routine studies in a patient with multiple confounding factors. Post-poliomyelitis muscle weakness, tracheomalacia, and vocal cord paralysis may have contributed to dyspnea in our patient making the evaluation particularly difficult.

Postpoliomyelitis syndrome is an entity that can cause respiratory compromise later in life, even after full recovery from the acute illness. In the survey by Halstead and Rossi, the risk factors most strongly associated with developing the postpolio syndrome were hospitalization at the time of acute illness, onset beyond the age of 10 years, the need for mechanical ventilation, and the presence of quadriplegia at onset. The presence of any of these factors was associated with the onset of fatigue within 30 years of the acute illness. As many as 60 to 80 percent of the patients in the survey of Halstead and Rossi experienced symptoms later in life.

Pulmonary function in patients years after polio has been studied by various groups. Patients complaining of dyspnea in one study had a significantly lower FEV1 and FVC than those who did not. In addition, it was found that peak mouth inspiratory pressure (Pimax) was near normal while peak expiratory mouth pressure (Pemax) was reduced to approximately 40 percent of predicted regardless of symptoms, suggesting significant occult neuromuscular weakness.

Our patient showed only minimal decrements in FEV1 and stable FVC over the course of 16 years. However, the marked diminution in inspiratory pressures confirmed neuromuscular disease. Some authors have emphasized the utility of the vital capacity measurement in following neuromuscular function. However, there may be marked abnormality in respiratory muscle function before a significant decrease in vital capacity is seen. This was seen in our patient who had significant inspiratory muscle weakness with little alteration in spirometry.

The plateau of maximal flows seen in our patient suggests an upper airway lesion. Unfortunately, neuromuscular disease may also result in a similar abnormality, particularly during inspiration. Similarly, the MVV, which depends partially on muscle strength and endurance but which may be decreased to a larger extent in upper airway obstruction (UAO), was disproportionately decreased in our patient. This supports either neuromuscular disease or UAO.

Clearly in our patient, routine pulmonary function
testing could not reliably separate the contribution of UAO or neuromuscular disease to the patient's breathlessness on exertion. To better define this, we utilized CPET, which has been advocated in the evaluation of dyspnea, hypothesizing that flow limitation and hyperinflation with exercise would implicate UAO as the predominant factor. Unfortunately, little has been written regarding the response to exercise in UAO or neuromuscular disease. As a result, many of our comparisons of the responses during exercise have been made to normal subjects and to patients with CAO.

We demonstrate in Table 2 that a normal peak VO\textsubscript{2} was achieved with a normal cardiac response. Ventilatory limitation was demonstrated by a low ventilatory reserve and would be expected with both UAO and neuromuscular disease. Figure 1 shows the absence of flow limitation at rest but clear expiratory limitation over 26 percent of the peak tidal volume. This may occur in fit normal subjects during heavy exercise and patients with CAO, even in those with mild disease. However, the significant rise in EELV seen in our patient is more typical of patients with CAO. The response in patients with UAO, particularly that associated with expiratory obstruction, would be expected to be similar. The fact that transpulmonary pressure during peak exercise exceeded the value associated with peak expiratory flows during resting maximal maneuvers also supports the importance of the UAO in our patient. As such, it is apparent that tracheomalacia and the new vocal cord paralysis were instrumental in the symptoms noted by our patient.

The abnormal respiratory muscles in our patient were likely further disadvantaged during maximal exercise as result of the UAO. Respiratory muscle function may be impaired during exercise in patients with airflow obstruction by muscle shortening associated with the need for greater inspiratory flow and by an increase in EELV. This was particularly so in our patient. In patients with advanced amyotrophic lateral sclerosis, Kreitzer et al showed that even small added resistance markedly decreased vital capacity and flows and resulted in an increased residual volume. As a result of the increase in EELV seen in our patient, the end-inspiratory lung volume was 95 percent of total lung capacity which is increased from what is expected in normal subjects. This has been associated with a significant increase in elastic load to the respiratory muscles and an increase in the metabolic cost of breathing. The absence of significant change in sniff Pdi following exercise suggests significant muscle fatigue did not develop in our patient, however.

The timing of our patient's clinical deterioration remains speculative, as pulmonary function tests (Table 1) showed little change in forced volumes and MVV over 16 years. No prior respiratory muscle pressures were available for comparison. As discussed earlier, a significant inspiratory muscle reserve exists allowing a significant deterioration in muscle strength before decreases may be seen in forced expiratory volumes. It is our impression that the deterioration in our patient likely reflects a greater physiologic effect of UAO (tracheomalacia and new vocal cord paralysis) in the face of progressive weakness of the respiratory musculature. The latter is a well-described phenomenon as a late sequela of poliomyelitis. As such, it is expected that further progression will be seen over time. Strong consideration has been given to surgical correction of the upper airway lesions to minimize their deleterious effects.

In conclusion, postpoliomyelitis syndrome may be associated with significant neuromuscular respiratory impairment.
failure, which may contribute to dyspnea. Superimposed UAOs, such as vocal cord paralysis or tracheal disorders associated with prior tracheostomy, may also play a role. Maximal flow volume loops and MVV may be helpful in identifying an abnormality but may not be able to separate between these diagnostic possibilities. A CPET incorporating measurements of lung volumes, tidal flows, and respiratory muscle function may be instrumental in identifying the factors accounting for the exercise dysfunction.

REFERENCES

1 Acres JC, Kryger MH. Upper airway obstruction. Chest 1981; 80:207-11
4 Owens GR, Murphy DMF. Spirometric diagnosis of upper airway obstruction. Arch Intern Med 1983; 143:1331-34
19 Dean E, Ross J, Road JD, Courtenay L, Madill KJ. Pulmonary function in individuals with a history of poliomyelitis. Chest 1991; 100:118-23