Orthopnea in a 75-Year-Old Man After Cardiac Catheterization*

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A 75-year-old man was evaluated in consultation for dyspnea. He was admitted to the hospital 1 day earlier with chest pain and progressive dyspnea over 12 months, which was mild with exertion but worse when he lay flat. He noted increasing daytime fatigue over the same time period. He had been able to work until recently as a building maintenance man. After hospital admission, he underwent cardiac catheterization, which showed no evidence of coronary artery disease. A sudden worsening of dyspnea developed, however, while he was lying supine at the end of the procedure.

Physical Examination

On physical examination immediately after cardiac catheterization, the patient had a respiratory rate of 12 breaths/min and an oxygen saturation of 90% on 2 L of oxygen via nasal cannula. He had inward movement of his abdomen during inspiration with tidal breathing in the supine position. Auscultation of his chest revealed decreased air entry. The cardiac examination was unremarkable, and there was no neck vein distension or peripheral edema. His neurologic examination revealed fasciculations in his extremities, but normal muscle tone and strength.

Laboratory and Radiographic Findings

Admission laboratory study findings included a normal blood count, electrolytes, and renal function indexes. Samples for arterial blood gas studies were not obtained. A hospital admission portable chest radiograph demonstrated lower lobe atelectasis (Fig 1).

After cardiac catheterization, arterial blood gases on room air showed a pH 7.29; Pco$_2$, 70 mm Hg; and Pco$_2$, 54 mm Hg, with a calculated bicarbonate of 35 mEq/L. A repeat chest radiograph and CT scan (Fig 2) of the chest showed low lung volumes and bilateral lower lobe atelectasis.

The patient remained in a supine position for 12 h after cardiac catheterization. A repeat arterial blood gas analysis showed pH 7.25; Pco$_2$, 83 mm Hg; and Pco$_2$, 111 mm Hg, with a calculated bicarbonate of 37 mEq/L on 2 L of oxygen via nasal cannula.

What is the likely diagnosis and how should it be confirmed?
Figure 1. Chest radiograph demonstrating low lung volumes and lower lobe atelectasis.

Figure 2. CT scan demonstrating bilateral lower lobe atelectasis.
Answer: Amyotrophic lateral sclerosis with diaphragmatic paralysis

Amyotrophic lateral sclerosis (ALS) is a fatal degenerative disorder of the CNS of unknown etiology that progressively affects the upper and lower motor neurons at the spinal or bulbar level. It usually affects patients > 50 years old. The presenting symptoms typically include weakness and clumsiness of the hands. Muscle cramps and fasciculations commonly occur early in the course of the disease, with progressive involvement of the muscles of the lower extremities, trunk, and neck. Weakness of the diaphragm subsequently ensues. This pattern of muscle weakness results in a clinical presentation characterized by limb muscle weakness followed by respiratory symptoms late in the clinical course of the disease. The presence of upper and lower motor neuron involvement with progressive weakness strongly suggests ALS. Such patients should undergo nerve conduction studies with repetitive stimulation and electromyography to confirm lower motor neuron degeneration and exclude disorders of peripheral nerves and muscles in addition to disorders of the neuromuscular junction, such as myasthenia gravis. Routine clinical laboratory tests are necessary to exclude other neurologic disorders caused by metabolic and endocrine conditions, plasma cell dyscrasias, and heavy metal poisoning. These tests should include blood glucose, thyroid function tests, parathyroid hormone level, calcium and phosphate, vitamin B₁₂ and folate levels, serum protein electrophoresis, and a urine collection for heavy metals.

Median survival times in patients with ALS vary from 36 to 48 months from the time of initial diagnosis. Involvement of respiratory muscle groups leads to a restrictive ventilatory defect and ultimately to hypercapnic respiratory failure. In most cases, death occurs from respiratory complications. The presence of diaphragmatic dysfunction is associated with a marked reduction in survival.

Although respiratory symptoms represent an unusual presenting manifestation of ALS, a few patients may present with acute respiratory failure. Patients with bilateral diaphragm weakness experience dyspnea sometimes associated with orthopnea and hypercapnea. They may also acquire respiratory failure requiring intubation and mechanical ventilation. Such patients are usually difficult to wean.

The physical examination can suggest the presence of bilateral diaphragmatic paralysis as a contributing cause of orthopnea. Most patients with severe diaphragmatic paralysis demonstrate paradoxical inward movement of the abdomen (abdominal paradox). This physical finding may not be present in patients with mild diaphragmatic weakness.

When diaphragmatic paralysis is suspected, spirometry should be performed in the supine and erect positions. Supine FVC is a sensitive measure of diaphragmatic weakness in ALS and correlates with transdiaphragmatic pressure. Simple measures of respiratory muscle strength such as maximal inspiratory pressure and maximal voluntary ventilation may be within normal limits early in the course of the disease because of preservation of the intercostal muscles. Evaluation of diaphragmatic involvement in ALS by fluoroscopy can be misleading. Paradoxic movement of the diaphragm with inspiratory "sniffing" is a characteristic feature of unilateral diaphragmatic paralysis being found in 90% of patients. This finding is rarely observed, however, in bilateral paralysis because cephalad movement of the ribs from accessory muscle contraction simulates the appearance of caudad displacement of the diaphragm. Although measurement of transdiaphragmatic pressure is the "gold standard" for confirming bilateral diaphragmatic function, the diagnosis is adequately supported by electromyographic findings in the appropriate clinical setting, obviating the need for transdiaphragmatic pressure monitoring.

Patients with ALS complicated by respiratory insufficiency benefit from noninvasive positive pressure ventilation (NPPV). Intermittent use of nocturnal NPPV can correct orthopnea at night, and prevent the development of atelectasis, which can contribute to hypoxemia. Nocturnal NPPV improves quality of life and survival in patients with ALS and respiratory muscle involvement. In the present patient, the presentation of orthopnea with abdominal paradox associated with bilateral lower lobe atelectasis suggested diaphragmatic weakness. The patient’s age and fasciculations in his extremities further suggested ALS as the underlying disease. Spirometry demonstrated a decrease in

Table 1—Pulmonary Function Test Results in Supine and Erect Positions*

<table>
<thead>
<tr>
<th>Variables</th>
<th>Erect</th>
<th>Supine</th>
<th>% Change</th>
</tr>
</thead>
<tbody>
<tr>
<td>FVC, L</td>
<td>1.58</td>
<td>1.09</td>
<td>31</td>
</tr>
<tr>
<td>FEV₁, L</td>
<td>1.21</td>
<td>0.82</td>
<td>32</td>
</tr>
<tr>
<td>FEV₁/FVC, %</td>
<td>76</td>
<td>76</td>
<td></td>
</tr>
<tr>
<td>Maximum inspiratory</td>
<td></td>
<td></td>
<td></td>
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<td>pressure, cm H₂O</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Maximum voluntary</td>
<td>50</td>
<td></td>
<td></td>
</tr>
<tr>
<td>ventilation, L/min</td>
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<td></td>
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</tbody>
</table>

*Pulmonary function testing was performed after the patient recovered from an acute episode of respiratory failure. Spirometry showed a significant decrease in flow rates in the supine position. Simple indices of respiratory muscle strength, maximal inspiratory pressure, and maximal voluntary ventilation were well preserved.
FEV$_1$ and FVC in the supine as compared with the erect position (Table 1). Electromyography showed denervation and reinervation of diaphragmatic and thoracoabdominal muscles. Further evaluation of the patient’s neurologic condition did not suggest alternative diagnoses. These findings indicated that the patient had ALS with diaphragmatic involvement. He was treated with bilevel pressure ventilation continuously for 24 h, until he regained an ability to ambulate. The patient’s condition continued to improve with resolution of lower lobe atelectasis. He continued at home with nocturnal bilevel pressure ventilation, and returned to all of his usual activities.

**Clinical Pearls**

1. Patients with ALS may occasionally present with isolated weakness of respiratory muscles. Such patients experience dyspnea without associated weakness of limb muscles.
2. Orthopnea may be a presenting manifestation of diaphragmatic muscle weakness.
3. Inward movement of the abdomen on inspiration during tidal breathing suggests diaphragmatic weakness. This finding is most notable in the supine position.
4. A fall in FEV$_1$ and FVC of $>20\%$ in the supine position compared to a baseline in the upright position suggests diaphragmatic weakness.

**Suggested Readings**

Meyrignac C, Poirier J, Degos JD. Amyotrophic lateral sclerosis presenting with respiratory insufficiency as the primary complaint; clinicopathological study of a case. Eur Neurol 1985; 24:115–120