valve (Medtronic) and the mitral valve with a 25-mm St. Jude mitral prosthesis. The patient's postoperative course was uneventful, and she was discharged 12 days later.

DISCUSSION

Cardiac involvement in the mucopolysaccharidoses is common and has been reported in all forms. Echocardiographic abnormalities in these storage diseases have been reported, with patchy thickening of the valves and other associated echocardiographic abnormalities noted. Schieie's syndrome has been associated with mitral and aortic valve involvement, either stenotic, insufficient, or both.

The clinical manifestations of Schieie's disease include corneal clouding and mild to moderate skeletal malformations, with broad hands and short stubby feet common. Joint abnormalities with fixation of the phalangeal joints are seen. Mental retardation is uncommon, and high intelligence is not uncommon. Four of the ten patients initially described were adults; thus, survival beyond infancy is common, as opposed to Hurley's disease, in which this is uncommon. The diagnosis is confirmed by demonstration of high quantities of mucopolysaccharides in the urine and characteristic abnormalities on skin biopsy, also seen in Hurley's syndrome.

Aortic stenosis in a different type of mucopolysaccharidosis was recently described in a 43-year-old man with Maroteaux-Lamy syndrome who underwent aortic valve replacement and had a good long-term result. The mitral valve is the most frequently involved in this variant of Hurley's syndrome, and insufficiency is the more common manifestation, although mitral stenosis is seen. While aortic involvement is described in Schieie's syndrome, precise definition of which is more common is unknown.

Surgical treatment of the cardiac malformations in this syndrome have not been reported frequently, perhaps due to caution suggested by some for potential surgical difficulties. Our patient, as those previously described, had an uneventful postoperative course and recovery, and both annuluse were of sufficient integrity for good prosthetic valvular placement.

References


Reversibility of Upper Airway Obstruction after Levodopa Therapy in Parkinson's Disease

Walter G. Vincken, M.D., F.C.C.P.; Carmen M. Derouay, M.Sc.; and Manuel G. Costo, M.D.

Serial flow-volume loops obtained in a 66-year-old patient with Parkinson's disease and recurrent episodes of dyspnea revealed a pattern consistent with upper airway obstruction, reversible after oral intake of levodopa. This observation shows that extrapyramidal involvement of the striated upper airway musculature may limit airflow and cause respiratory symptoms. Persistence of flow oscillations on the flow-volume loop contour after reversal of upper airway obstruction and dyspnea should be considered to reflect upper airway dysfunction with possibly serious consequences. (Chest 1989; 96:10-12)

In patients with Parkinson's disease and other extrapyramidal disorders, pulmonary problems commonly contribute to morbidity and mortality. A variety of respiratory problems have been recognized, such as a "cogwheel" breathing pattern, respiratory dysrhythmias, whether or not associated with levodopa therapy, aspiration pneumonia and chronic or recurrent airflow limitation. Recently, we have demonstrated that in patients with extrapyramidal disorders, the configuration of the flow-volume loop is frequently abnormal even in the absence of respiratory symptoms. In particular, the flow-volume loop contour frequently reveals flow oscillations or more abrupt and irregular flow interruptions, which were shown to be due to involvement of the striated musculature surrounding the upper airway rather than to involvement of the respiratory pump muscles. These configurational changes, which subsequently have also been noticed by other authors, were associated with physiologic or clinical evidence of upper airway obstruction in one third of our patients. Based on these findings, we have suggested that upper airway dysfunction resulting from extrapyramidal involvement of the upper airway muscles is causally related to the recurrent or chronic airflow limitation noticed in these patients. We report an observation that adds further evidence to this hypothesis.

CASE REPORT

The patient was a 66-year-old man, a smoker until 1965, with recurrent episodes of shortness of breath, diagnosed as having asthma but not responsive to bronchodilators. He also had a longstanding history of moderately advanced idiopathic Parkinson's disease with pronounced tremor and occasional dyskinetic movements of the hands, mouth, and tongue, associated with dysarthria and dysphagia. All symptoms were worse during the night and on awakening. He was being treated with 150 mg levodopa/15 mg carbidopa (Sinemet) every 3 hs while awake and 2 mg trihexyphenidyl (Artane) three times a day.

After obtaining his informed consent, pulmonary function tests*

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during and without levodopa therapy were done. Spirometry was performed with the patient in the sitting position through a large, scuba-type mouthpiece providing a tight seal, using an 8-1. Stead-Wells spirometer. Flow was obtained by electronic differentiation of volume. Flow and volume were recorded in a fast response XY recorder (Hewlett Packard HP 7042B) to obtain the flow-volume loops.

The first flow-volume loop (Fig 1, left panel), was obtained at 9 AM, without levodopa therapy since the previous night. The patient had mild tremor and moderate shortness of breath. Reduced inspiratory and expiratory flow rates and an inspiratory plateau interrupted by flow oscillations were seen.

At 9:10 AM levodopa/carbidopa (150 mg/15 mg) was given to the patient because of worsening parkinsonian and respiratory symptoms. A second flow-volume loop (Fig 1, middle panel) obtained 10 min later showed further reduction of expiratory flow rates and of the forced vital capacity. The configuration of the expiratory loop showed a plateau with small flow oscillations. The inspiratory curve still had a plateau-like configuration.

In the following hour, both parkinsonian symptoms and dyspnea improved, and a third flow-volume loop obtained 90 min after the intake of levodopa showed significant improvement of inspiratory and expiratory flow rates, with disappearance of inspiratory and expiratory plateaus. Flow oscillations with a frequency corresponding to that of the peripheral tremor persisted on the inspiratory curve (Fig 1, right panel). A fourth flow-volume loop obtained 15 min after inhalation of a β2-sympathomimetic aerosol did not reveal further improvement of flow rates.

Pulmonary function test data are provided in Table 1.

**DISCUSSION**

The sequence of flow-volume loops obtained in this parkinsonian patient with recurrent episodes of dyspnea reveals a pattern of upper airway obstruction associated with a symptomatic episode and reversibility following oral intake of levodopa. This sequence of events strongly suggests that extrapyramidal involvement of the upper airway muscles was the mechanism of his upper airway obstruction and consequent dyspnea.

The baseline flow-volume loop, obtained without the levodopa therapy, not only documents airflow limitation, but its contour suggests that airflow limitation is due to a variable extrathoracic upper airway obstruction. Twenty minutes later, the second flow-volume loop, obtained at the height of parkinsonian and respiratory symptoms, reveals severe airflow limitation and a contour typically seen with fixed upper airway obstruction. Hence, inspection of flow-volume loop contours without levodopa therapy allows localization of the airflow limitation in the upper airway.

Following oral intake of levodopa, improvement of parkinsonian and respiratory symptoms is associated not only with increased flow rates but also with disappearance of the flow plateaus, indicating reversal of upper airway obstruction. The time course of these changes is consistent with an effect of levodopa, for which peak plasma concentrations and maximal clinical efficacy are obtained between 30 and 120 min after oral intake. The changing nature of the observed upper airway obstruction, from a variable extrathoracic to a fixed upper airway obstruction without levodopa therapy and its reversibility after levodopa administration is consistent with extrapyramidal involvement of the upper airway muscles as the cause of upper airway obstruction. That these muscles may be affected in the extrapyramidal disorders has been demonstrated previously by electromyography, showing abnormal electric activity of the intrinsic laryngeal muscles. Abnormal involuntary movement of the vocal cords and supraglottic structures can also be visualized by direct fiberoptic examination of the upper airway.

Flow oscillations on the flow-volume loop persisted after improvement of respiratory symptoms and flow rates. These flow oscillations, corresponding to involuntary movements of upper airway structures, reflect the sensitivity of the flow-volume loop to upper airway dysfunction associated with extrapyramidal and other neuromuscular disorders. When recognized, this oscillatory pattern should direct the attention to the upper airway as the possible site of recurrent upper airway obstruction.

**Figure 1.** Y-axis: expiratory (Ve) and inspiratory (Vi) flow, 1 mark along y-axis = 1 L/s; x-axis: volume (V), 1 mark along x-axis = 1 L; 9:00 AM: flow-volume loop "off-therapy"; 9:30 AM: flow-volume loop shortly after oral intake of levodopa (at 9:10 AM) at the peak of parkinsonian and respiratory symptoms. 10:40 AM: after improvement of parkinsonian symptoms and dyspnea as a result of levodopa ("on-therapy").

**Table 1—Pulmonary Function Data**

<table>
<thead>
<tr>
<th>Measurement</th>
<th>9:00</th>
<th>9:20</th>
<th>10:40</th>
</tr>
</thead>
<tbody>
<tr>
<td>FVC, L</td>
<td>1.90</td>
<td>1.60</td>
<td>3.65</td>
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<tr>
<td>FEV1</td>
<td>1.20</td>
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<td>2.01</td>
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<td>0.95</td>
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<tr>
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<td>0.95</td>
<td>1.30</td>
</tr>
<tr>
<td>FEF75, L/s</td>
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<td>0.55</td>
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<td>PIF, L/s</td>
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</tr>
<tr>
<td>FIF50, L/s</td>
<td>1.40</td>
<td>1.40</td>
<td>2.90</td>
</tr>
</tbody>
</table>

*Oral levodopa intake at 9:10 AM.
†FVC, forced vital capacity; FEV1, forced expired volume in 1 second; PEF, peak expiratory flow; FEF25, expiratory flow after exhalation of 25% of the FVC; PIF, peak inspiratory flow; FIF50, mid-vital capacity forced inspiratory flow.
episodes of respiratory distress. The involvement of the respiratory muscles by the extrapyramidal disorders as the cause of reduced inspiratory and expiratory flows and flow oscillations is unlikely. The diaphragm seems to be spared in extrapyramidal disorders, and we have been unable to show any changes in pleural pressure associated with rhythmic contractions of respiratory muscles.

Absence of a significant bronchodilator effect and the good response of this patient’s respiratory symptoms and flow rates to levodopa suggest that good control of parkinsonism may be all that is required to improve respiratory function in these patients.

Parkinsonian involvement of the upper airway can produce a dysfunctional upper airway obstruction of varying degree but severe enough to produce serious airflow limitation and dyspnea in these patients.

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Echocardiographic Detection of an Infected Superior Vena Caval Thrombus Presenting as a Right Atrial Mass*

Andrew E. Dick, M.D.;† Charles M. Gross, M.D.;‡ and Joseph W Rubin, M.D., F.C.C.P.§

A patient was found to have a large mobile right atrial mass by two-dimensional echocardiography after developing sepsis due to prolonged central hyperalimentation. Contrast echocardiography was helpful in localizing the origin of the mass. A large infected thrombus emanating from the superior vena cava was removed at operation. The discussion includes a review of the literature on the echocardiography of right atrial masses.

The ability of two-dimensional echocardiography to noninvasively examine the right atrium makes the differential diagnosis of right atrial masses an issue of considerable practical importance. There is an abundance of literature on right atrial thromboembolism, but the differential diagnostic spectrum includes other entities, from normal structures to primary or metastatic tumors. This report demonstrates the utility of echocardiography in characterizing a particular type of right atrial mass that developed due to prolonged central venous catheterization. To our knowledge, this entity has been unreported previously in the literature.

CASE REPORT

A 45-year-old man was transferred to our institution for further evaluation of persistent fever. He had presented to another hospital with several days of epigastric pain, nausea, abdominal distention, anorexia, and early satiety. He denied consumption of alcohol, although there was a history of alcohol abuse and recurrent pancreatitis, for which he had last been hospitalized three months earlier. At that time, a large pancreatic pseudocyst was demonstrated on computerized tomographic scan, and the patient’s condition improved with conservative management. Initial laboratory studies at the other hospital revealed blood cultures negative at 48 hours, with normal serum levels of amylase and lipase. There was improvement with nasogastric suction, intravenous fluids, and cepofaropenone intravenously. Past medical history was significant for an exploratory laparotomy four years previously, a right inguinal hernia repair five years earlier, and glucose intolerance.

On physical examination, the patient was a thin man in no apparent distress. His blood pressure was 110/70 mm Hg, the pulse rate was 120 beats per minute, the respiratory rate was 20/minute, and the oral temperature was 36.2°C (97.2°F). The abdomen was distended, and normoactive bowel sounds were present throughout. On palpation of the left upper quadrant, mild tenderness was elicited, and a cystic mass was palpable. The ECG showed sinus tachycardia at a rate of 100 beats per minute, early transition in

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Echo Detection of Infected SVC Thrombus (Dick, Gross, Rubin)