**DISCUSSION**

Hemoptysis coincident with menstrual periods helps to differentiate catamenial hemoptysis from hemoptysis of other causes. When recurrent hemoptysis is associated with menstrual periods, pulmonary endometriosis is the likely cause in terms of the patient's history. A definitive diagnosis of pulmonary endometriosis may be difficult to make. Histologic confirmation was obtained in less than one third of these cases. Because the chest roentgenogram in pulmonary endometriosis is usually normal or demonstrated densities are presumed to be secondary to hemorrhage, careful history taking is an essential first step. Fiberoptic bronchoscopy and arteriography provide much information about the vascular disturbance causing hemoptysis. Though bronchoscopy should be performed during an episode of hemoptysis in an attempt to localize the site of bleeding, it is very difficult to detect the lesion by bronchoscopy. Elliot et al emphasize that CT scanning is the method of choice in confirming this diagnosis and its accurate pulmonary localization, provided that chest CT scanning is performed during the presence of catamenial hemoptysis. Jelihovsky and Grant reviewed 12 cases in which endometrial tissue was found on histologic examination of the lung. One half of these cases revealed the presence of large capillaries, thin-walled vessels or bronchial arteries in their endometrial tissues. Since the type of hemoptysis under discussion is caused by rupture of the pulmonary vessels, these vessels must be responsible for catamenial hemoptysis in pulmonary endometriosis. Thus, we reason that a diagnostic angiogram to detect the localization and therapeutic embolization is useful for hemoptysis in pulmonary endometriosis, in so far as angiograms reveal pathologic changes of bronchial and pulmonary arteries. Unfortunately, however, the bronchial and pulmonary arteriograms revealed no findings to help us detect the localization in our cases. We suggest that angiograms may have little value in the evaluation of patients with pulmonary endometriosis.

**REFERENCES**


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**Kyphoscoliosis and Respiratory Failure**

**A Patient Treated with Assisted Ventilation for 27 Years**

Colin R. Woolf, M.D., F.C.C.P.

A 62-year-old woman had had kyphoscoliosis since age 12 years. Respiratory failure developed at age 35 years following a respiratory infection. A tracheostomy was done and she required assisted ventilation using a respirator (Bird). Many attempts at weaning her from the ventilator were unsuccessful. She has remained dependent on assisted ventilation for more than 27 years but has had a relatively comfortable and meaningful life. In 1983, a left pleuroscopy was done related to recurrent pneumothorax and numerous small bullae were seen on the lung surface. Recent investigation using computed tomographic scanning has shown patchy areas of emphysema in both lungs. Emphysema is not a feature of kyphoscoliosis and it is suggested that intermittent positive pressure applied to the lung over long duration may cause the lung destruction of emphysema.

*(Chest 1990; 98:1297-98)*

Severe kyphoscoliosis often results in respiratory problems that begin in late teenage years and progress through early adulthood causing chronic respiratory failure. Acute respiratory failure may be precipitated by a relatively mild respiratory infection and death may occur unless assisted ventilation is provided. However, the patient may become ventilator dependent. The present report describes the situation in which a patient has continued to require assisted ventilation for 27 years. There is some evidence that, under these circumstances, emphysema may develop.

**Case Report**

The patient was a normal 10-year-old girl in 1937 when she developed poliomyelitis. This affected her right leg and left arm but there was no requirement for assisted ventilation. Two years later, at age 12 years, early kyphoscoliosis was noted. Subsequent to this, she noted some slow progressive shortness of breath. In 1962, at age 35 years, a respiratory infection developed and she became acutely short of breath. She was admitted to the emergency department at the Toronto General Hospital. She was unconscious and deeply cyanosed and had intermittent shallow gasping respirations. Arterial blood gases showed a pH of 7.02 and PCO₂ of 141 mm Hg. A tracheostomy was done on the evening of hospital admission. She did well and after a month, the tracheostomy was removed and she was discharged from the hospital. At that time, the arterial blood gases showed a pH of 7.43 and PCO₂ of 44 mm Hg.

Three weeks after hospital discharge, she was readmitted again with acute respiratory failure. Her course during this second hospital admission was similar to the first admission. However, many attempts to wean her from the ventilator were unsuccessful and the arterial blood PCO₂ might rise to 100 mm Hg. Trials off the ventilator were done for the next three years but these were stopped in 1965 and she became a permanent resident of the Respiratory Failure Unit.

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In 1981, a left-sided spontaneous pneumothorax developed requiring tube drainage. This recurred in 1983 and a pleuroscopy showed the surface of the left lung to be covered with numerous small bullae. A talc poudrage was done. In August 1989, a pneumothorax developed on the right side and this required a bullectomy and poudrage.

Over the years, she has adapted to life with a permanent tracheostomy and a need for continuous assisted ventilation. For many years (1969 to 1980), she was a part-time secretary at the hospital, typing discharge summaries. She is able to walk about the hospital using a wheeled support with a ventilator (Bird) attached. With assistance, she leaves the hospital to attend the theater or have dinner in a restaurant and she makes various trips to visit friends and relatives as far as 160 km away. Although she has a cuffed tracheostomy tube, she is able to speak in a voice that is peculiar but quite understandable. To do this she probably swallows air into the upper trachea and then expels the air through the vocal cords. As her talking is completely independent of her breathing, she can talk indefinitely or count indefinitely without pause.

Recently, there has been a complete review of her case to decide if there is any possibility of making a further attempt to wean her off the respirator. Vital capacity was 250 ml. Maximum inspiratory pressure − 15 cm H2O. MUCA scan showed a right ventricular ejection fraction of 20 percent and a left ventricular ejection fraction of 40 percent. There was prominence to the right ventricular outflow tract, the right ventricle was enlarged, and the distal free wall and apex were hypokinetic.

Ventilation-perfusion lung scan showed matched perfusion and ventilation defects affecting the left lower lung and right mid lung zone. Computed tomography (CT) of the thorax using a thin-section high-resolution technique showed patchy areas of arterial deficiency-type emphysema in both lungs.

There seemed very little chance of successfully weaning this patient where there was severe lung volume restriction, probable respiratory muscle weakness, emphysema of the lungs, and evidence of chronic cor pulmonale. Therefore, she will continue indefinitely in a single room in the intensive care unit.

**DISCUSSION**

Patients with severe kyphoscoliosis develop abnormalities of ventilation-perfusion relationships in addition to the restrictive ventilatory defect. In time, there is worsening of the hypoventilation and an increasing rise in arterial blood carbon dioxide tension probably due to fatigue and weakness of inspiratory muscles that are working at a mechanical disadvantage because of the chest deformity. There is also a low compliance of the chest wall that requires increased muscle effort for inspiration.

It has been suggested that resting the respiratory muscles during the night by using assisted respiration or nasal continuous positive pressure ventilation results in less fatigue and better function during the day. We considered all these techniques for the patient described in this report but she requires continuous intermittent positive pressure assistance throughout the 24 hours and the nocturnal portion alone was very unlikely to be of value at this stage. Also, many years ago, we had tried nocturnal assisted ventilation and this had been insufficient to allow weaning of the patient.

Further attempts at weaning were discussed with the patient and she expressed a very definite opinion that she did not wish to go through the discomfort involved. She felt that she now had established her life in a definite way and she was unwilling to make any changes. Even moving her from the intensive care unit area to an ordinary chest ward or considering some form of home ventilation was completely unacceptable to the patient. Therefore, it was decided to continue indefinitely with the present situation. The patient does not even remember clearly life in society outside the hospital. She is now so used to the situation that when she has a dream, she dreams with herself using assisted ventilation.

What happens to a lung where there has been intermittent positive pressure applied over 27 years? The pleuroscopy showed multiple small cystic areas on the surface of the lung and the presence of emphysema has been confirmed by the CT scan in this patient. To be sure that there was no genetic aspect, an α1-antitrypsin blood level was taken and this was within normal limits. Usually autopsies of patients with kyphoscoliosis who have died of respiratory failure show that the lung is collapsed and there is neither thinning of lung substance nor bullae. Possibly, intermittent positive pressure for many years can cause the lung destruction of emphysema.

There have been other reports of long-term assisted ventilation in patients with kyphoscoliosis, but the longest period reported has been 12 years. Therefore, to our knowledge, our patient has been receiving assisted ventilation because of kyphoscoliosis for the longest reported time.

The patient would certainly have died if she had not received assisted ventilation 27 years ago, but was it ethical to treat this patient in such a way that she would be ventilator-dependent for the rest of her life? At a recent interview for a television documentary, she stated that she had adapted to the situation and she had made a satisfactory and quite enjoyable life for herself and she has a role in inspiring others to adjust to a major disability. Our patient is a real example of courage and determination.

**REFERENCES**