Communications

Pulmonary Hypertension and Sickle Hemoglobinopathy

To the Editor:

We read with interest the article by Rubin on "Approach to the Diagnosis and Treatment of Pulmonary Hypertension" (Chest 1989; 96:659-64). The author based the classification of pulmonary hypertension on the primary site of involvement, namely precapillary and postcapillary causes of pulmonary hypertension.

The diagnostic approach to thromboembolic forms of precapillary pulmonary hypertension deserves further comment in view of our recent case of hereditary spherocytosis with pulmonary hypertension, and known literature data on sickle cell hemoglobinopathies and thalassemia.

The vast majority of pulmonary emboli are due to dislodgement of venous thrombi with subsequent impaction in the pulmonary circulation. However, it is necessary to distinguish in situ thrombosis in cases of sickle cell hemoglobinopathies SS, SC and SB thalassemia. In such cases, autopsy specimens may reveal asymmetric intimal expansion and widespread occlusions of small- and medium-sized arteries with old, organized, recanalized thrombi. In these hemoglobinopathies, red cell sequestration may occur in the lungs, resulting in increased viscosity, vascular stasis and even complete blockage of the microvasculature.

This in situ vaso-occlusion is precipitated by gross sickling of red blood cells, especially in hypoxic areas, leading to microvascular thrombosis and microinfarcts with necrosis. Bone marrow and fat emboli released from areas of ischemic bone necrosis can contribute to the vaso-occlusion. Extensive vascular narrowing and occlusions lead to increased pulmonary resistance, pulmonary hypertension and cor pulmonale.

The venous-thrombus pulmonary embolism pattern is of little importance in such cases since the incidence of pulmonary embolism is not increased in patients with sickle cell hemoglobinopathies in comparison to the general population.

Therefore, we would suggest adding peripheral blood film examination, sickle cell test and hemoglobin electrophoresis to the diagnostic evaluation of anemic patients with complaints suggestive of pulmonary hypertension; this hypertension may already be present before the diagnosis of hemoglobinopathy is made.

Even though the true incidence of pulmonary hypertension and sickle hemoglobinopathy is largely unknown, it is likely that the effects of repeated pulmonary vascular occlusions may become even more apparent in view of the improved life expectancy of patients with sickle cell hemoglobinopathy.

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To the Editor:

The comments made by Doctor Verresen and his colleagues are valid and ones with which I agree. Patients with sickle cell disease can develop pulmonary hypertension due to in situ thrombosis with microvascular sickling; furthermore, this process may be complicated by pulmonary venous hypertension, which can result from the cardiomyopathy associated with sickle cell anemia. Patients with unexplained pulmonary hypertension and anemia should undergo a thorough evaluation to exclude hereditary or acquired conditions (such as hemoglobinopathies, granulomatous diseases, vasculitis, and malignancies with tumor embolization to the lungs) which may be associated with these two processes.

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Pleural and Lung Cryobiopsies

To the Editor:

We have recently published a paper entitled, "Pleural and Lung Cryobiopsies During Thoracoscopy" (Chest 1989; 95:3). The number of patients in the series has now reached 35 and our conclusions are the same (ie, analgesic effect, low risk of hemorrhage, and low risk of air escape). Using cryotherapy in pleural and lung lesions, perfectly adequate samples can be obtained for histologic examination using a single cycle of freezing and immediate fixation. However, in the latest series of patients we have used electron microscopy of lung and pleura biopsies and found degenerative changes in type 2 pneumocytes with very condensed cytoplasm and cellular detachment from the basal membrane. The collagenous and elastic framework of the alveolar wall is distorted by marked

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interstitial edema, and the endothelial cells of the capillaries are turgid with swollen mitochondria. These changes are evidence of damage to the blood/air barrier and, if they are consistently encountered, would represent a considerable drawback to detailed studies of lung parenchyma and interstitial lesions using this technique.

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Phrenic Nerve Involvement in Charcot-Marie-Tooth Disease

To the Editor:

We read with interest Chan et al's report (Chest 1989; 96:1197-99) of a patient who died due to respiratory failure secondary to diaphragmatic weakness and a previous history of Charcot-Marie-Tooth disease (CMT). The association of phrenic nerve involvement and diaphragmatic dysfunction with CMT was first noted in 1987. Since that time, additional reports have emphasized the potential for severe respiratory impairment. Abnormal spirometry has suggested involvement in less severely impaired individuals.

We would like to emphasize the possible detection of phrenic nerve involvement earlier in the course of CMT, as well as the role of phrenic nerve conduction studies performed with transcutaneous monopolar 28G EMG needles placed into the sternal origin and lateral portion of the diaphragm to confirm the diagnosis of CMT involving the phrenic nerve.

A 31-year-old man with an 11-year history of CMT and progressive lower extremity weakness was evaluated for a non-productive cough, and pulmonary function studies revealed a mild reduction of FVC and MVV. Lung volumes (measured utilizing nitrogen wash-out) were found to be normal. Nerve conduction studies of the upper and lower extremities revealed marked abnormalities consistent with CMT. Phrenic nerve studies with transcutaneous diaphragmatic electrodes revealed bilateral abnormalities with both demyelination as well as axonal loss.

Since then the patient has been followed with spirometry, which has revealed a stable impairment for one year. The patient continues to have no symptoms of dyspnea, but his activities are limited.

It has been noted that respiratory muscle weakness occurring in patients with chronic stable neuromuscular disorders cannot be predicted from clinical assessment of general muscle strength or from the absence of respiratory complaints. Relatively minor added respiratory loads, however, may result in severe respiratory impairment. In view of the uncertainty regarding the frequency of phrenic nerve involvement with CMT, we would therefore like to suggest spirometry and, when appropriate, phrenic nerve stimulation studies to clarify the nature and extent of impairment.


REFERENCES


High Frequency Ventilation

To the Editor:

In their recent review of high frequency ventilation, Standiford and Morganroth discussed "other modes of ventilation." One of the modes discussed was continuous flow anepnic ventilation (CFMV). The author stated that "continuous-flow anepnic ventilation has not been used clinically as yet." To my knowledge there have been at least three reports of its clinical use.

The first clinical evaluation of CFMV was in five adult women undergoing elective gynecologic procedures under general anesthesia. An endobronchial catheter was inserted in each mainstem bronchus. One hundred percent oxygen was delivered at a flow of 0.6 to 0.7 L/kg/min down the two catheters in the anepnic patients for 30 min. After 30 min, the mean PaCO2 was 55.0 ± 4.0 mm Hg compared with the starting value of 37 ± 4 mm Hg. The mean PaO2 after 30 min was 99 ± 37 mm Hg. There were no complications in these patients.

In 1986, Breen et al used CFMV in five patients having non-thoracic surgery. They used 50% N2O and 50% O2 at a flow of 1 L/kg/min. After 30 min of anepnia, PaCO2 increased to a mean of 69 ± 14 mm Hg from a starting value of 36 ± 3 mm Hg.

CFMV has also been evaluated in seven brain dead patients by Ebato et al. These authors confirmed the findings of the other reports: CFMV was most effective in eliminating CO2 when O2 was insufflated into both bronchi at high flow rates.

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Lymphangiomyomatosis

To the Editor:

Dr. Eliasson and his colleagues (Chest 1989; 96:1352-55) stated, after a review of individual case reports, that "single therapy with tamoxifen and combined therapy with oophorectomy, progesterone, and tamoxifen were successful in approximately 30 percent of the cases" of lymphangiomyomatosis (LAM). Following meta-analysis, they concluded that oophorectomy and progesterone offered the greatest benefit.