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.


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High Frequency Ventilation

To the Editor:

In their recent review of high frequency ventilation, Standiford and Morganroth1 discussed "other modes of ventilation." One of the modes discussed was continuous flow apneic ventilation (CFV). The author stated that "continuous-flow apneic 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 CFV 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 apneic 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±3.1 mm Hg. The mean PaO₂ after 30 min was 99.0±37 mm Hg. There were no complications in these patients.

In 1986, Breen et al. used CFV 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 apnea, PaCO₂ increased to a mean of 69±14 mm Hg from a starting value of 36±3.3 mm Hg. CFV has also been evaluated in seven brain dead patients by Ebato et al. These authors confirmed the findings of the other reports: CFV 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.

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