the left ventricular lumen.  

Septal ruptures have been associated with inferior damage in less than 25 percent of acute myocardial infarctions, whereas it is usual for papillary involvement to occur with posterior or inferior locations. Furthermore, posterior papillary involvement is four times as frequent as anterior papillary muscle damage.  

Widening of the mediastinum led to speculation that an aortic dissection or rupture of the free ventricular wall had occurred. In retrospect, this widening may have been due to venous engorgement from acute tricuspid insufficiency caused by right ventricular overload from the ventricular septal defect. 

The sudden appearance of a loud systolic murmur and thrill favors the diagnosis of perforation. There has been at least one case report however, in which a thrill has been associated with papillary muscle involvement and incomplete rupture without septal perforation.  

The only published cases similar to ours were those observed by Skoulas and Beier in 1967, and by Rawlins et al in 1972. In the earlier of these, a small defect was found only after a careful anatomic search. Approximately half of the base of the posterior muscle was necrotic, without definite rupture. In the more recent case, both a septal defect and mitral regurgitation were documented by cineangiography. Their patient had been free of any cardiac murmurs prior to his acute ischemic episode. The autopsy showed that the posterior papillary muscle was fibrotic and distorted, but not ruptured. 

In our case it is impossible to assess exactly which of the lesions of septal defect and papillary muscle rupture came first. The early systolic ejection murmur heard initially was attributed to turbulence originating in the left ventricular outflow tract. The postmortem discovery of a calcified septal aortic leaflet is consistent with this finding. There was no thrill during the final event, which can be explained by the presence of severe hypotension. Furthermore, the cause of death was pulmonary edema, a characteristic finding in acute severe mitral insufficiency. Resuscitation was not attempted; thus, postmortem changes due to closed chest trauma could not have been responsible for any of the ruptures. 

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Prolonged Respirator Support for the Treatment of Intractable Myasthenia Gravis  

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The advent of modern respiratory care has provided the ability to maintain patients on full respirator support via a tracheostomy for prolonged periods, without serious sequelae. We recently had the opportunity to care for a patient who suffered from severe neuromuscular weakness secondary to myasthenia gravis. This necessitated continuous respirator support for a total of 661 days, a duration previously unreported.  

The availability of specialized team care in the modern hospital intensive care unit (ICU) has greatly improved the prognosis in certain disease states. We describe here a patient with a case of myasthenia gravis that was particularly intractable to many modes of therapy. The neuromuscular weakness was so severe that she required almost continuous respirator support via auffed tracheostomy tube for a total of 661 days, the longest period on record.  

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CASE REPORT

The patient is a 47-year-old woman who developed right facial weakness and progressive fatigue in May, 1970. An edrophonium chloride (Tensilon) test and an electromyogram were consistent with the diagnosis of myasthenia gravis. The patient initially responded to therapy until hospitalization was required on Aug. 10, 1970 because of respiratory distress. Tracheostomy was performed Aug. 16, 1970 after an episode of cardiorespiratory arrest. With improvement in the patient's strength, the tracheostomy was discontinued on Sept. 23, 1970, and the patient was subsequently discharged.

Because of progressive weakening, she was rehospitalized Oct. 30, 1970. Tracheostomy was again performed on Nov. 25, 1970, and following respiratory arrest while not using the respirator, she was transferred to Harbor General Hospital Dec. 18, 1970 for further management in the medical-respiratory intensive care unit.

At the time of admission she was on assisted ventilation by a pressure respirator. There was generalized muscle weakness, with ptosis and masklike facies. Results of initial laboratory tests were within normal limits, except for arterial blood gas values on the respirator, which revealed a slight respiratory alkalosis. Vital capacity was 400 ml.

A treatment program of pyridostigmine bromide (Mestinon), neostigmine methylsulfate (Prostigmine), aneomobnum chloride (Mytelase), neostigmine, ephedrine, oxtriphylline (Choledyl) and guanidine hydrochloride in varying dosages, combinations and durations was begun with no persistent benefits. ACTH and high-dose prednisone were also given without success.

Chest roentgenogram revealed a mediastinal mass, which was excised Jan. 18, 1971; it proved to be a benign thymoma. Subsequently, the mediastinum was irradiated with 4,000 rads during July, 1971. There was again no consistent response to these therapeutic maneuvers.

During her hospitalization, the patient's dependence on the respirator was almost always total. There was a third and a fourth cardiorespiratory arrest; both occurrences were attributed to respirator failure. Daily measurement of her vital capacity (2,800 ml predicted) ranged widely from 50-1,200 ml (Fig 1). Arterial blood gas measurements repeatedly revealed a mild, fully compensated respiratory alkalosis.

During the early part of her acute illness, the patient received respirator support from a pressure-cycled respirator (Bird, Mark 10, with heated Ohio nebulizer). Under this management, she had periodic occurrences of atelectatic infiltrates on chest roentgenogram associated with mild fever but no evidence of a respiratory pathogen. Close observation subsequently revealed that frequent lung inflations to her total lung capacity (TLC) prevented atelectasis. This was achieved by changing to a volume-cycled respirator (Bennett MA-1) with an automatic "sigh" mechanism that provided paired inflations to TLC 15 times/hour.

Inspired oxygen concentration was maintained at 21 percent during most of her hospital course. It was occasionally increased to 30 percent during transient episodes of plateau atelectasis, but never higher. The alveolar-arterial Po2 difference fluctuated between 30-35 mm Hg.

A Rusch red rubber double-cuffed tracheostomy tube was in place at all times. The cuffs were inflated alternately every hour during the early part of her hospitalization, but the tracheostomy stoma gradually enlarged, such that inflating the upper cuff caused it to bulge out into the stoma. Subsequently only the lower cuff was used. The details of her tracheostomy care are listed in Table 1.

In March, 1972, after 15 months of hospitalization, the patient was given a five-week course of antilymphocyte globulin (ALG). She began to show improvement, but in May, 1972 developed fever, with chest and epigastric pain. A diagnosis of polyserositis was made, with documented pancreatitis, pericarditis and effusion, and pleurisy with effusion on the left. Multiple studies of pleural fluid, including a pleural biopsy, were nondiagnostic. All skin tests except dinitrochlorobenzene (DNCB) were also negative. The findings gradually subsided on supportive management.

In July, 1972 a course of alternate-day, high-dose (100 mg) prednisone was repeated. Within several weeks, the patient began to demonstrate gradual but progressive improvement in muscle strength. By Sept. 28, 1972, her vital capacity increased to over 1,200 ml (Fig 1) and persisted as such throughout the day; she was then transferred to the ward, some 652 days after admission to the ICU (Table 2). Her tracheostomy was left in place, but the cuff could now remain deflated for the first time since it was installed Nov. 25, 1970.

At the time of discharge on Nov. 22, 1972, the patient was unable to completely appose her vocal cords. The tracheostomy care listed in Table 1.

Figure 1. Clinical course of patient over two-year hospitalization.
This case report illustrates that it is possible to successfully manage a totally dependent, tracheostomized patient on a respirator for an indefinite period. Careful attention to the routine care of the patient and respirator by an interested nursing team and inhalation therapy service can minimize complications. This requires constant monitoring of all parameters as listed in Table 1, in order to prevent many of the problems associated with chronic respirator use, such as oxygen toxicity, infection, "respirator lung," and simple mechanical failure.4

The use of a cuffed tracheostomy tube is subject to a myriad of complications,5 which are thought by many investigators to be prevented by the use of meticulous tracheostomy care.6 The fact that this patient survived such a long course without these complications is plainly a tribute to her over-all nursing care.

The modern concept of acute respiratory care includes the integration of multiple specialties, including nursing, inhalation therapy, physical and occupational therapy, as well as general psychologic support for the patient.7 The cost of such care is significant, but the total benefits achieved by this patient, as well as the many members of the health care team who participated in her care, readily underwrite such expenditures.8 Not all patients with respiratory failure will respond to prolonged ICU care. Careful selection of patients, however, such as those with reversible neuromuscular disease, can lead to gratifying results.

In summary, this case report illustrates that the team concept of acute respiratory care can be extended to the prolonged chronic care of the occasional patient who is in need of such attention. In addition, the prolonged course of myasthenia gravis is depicted, as is the importance of providing adequate respiratory support to the severely ill patient, while awaiting a remission or a response to therapy.

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