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Respiratory failure because of neuromuscular disease.

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Abstract

PURPOSE OF REVIEW: Understanding the mechanisms and abnormalities of respiratory function in neuromuscular disease is critical to supporting the patient and maintaining ventilation in the face of acute or chronic progressive impairment.

RECENT FINDINGS: Retrospective clinical studies reviewing the care of patients with Guillain-Barré syndrome and myasthenia have shown a disturbingly high mortality following step-down from intensive care. This implies high dependency and rehabilitation management is failing despite evidence that delayed improvement can occur with long-term care. A variety of mechanisms of phrenic nerve impairment have been recognized with newer investigation techniques, including EMG and ultrasound. Specific treatment for progressive neuromuscular and muscle disease has been increasingly possible particularly for the treatment of myasthenia, metabolic myopathies, and Duchenne muscular dystrophy. For those conditions without specific treatment, it has been increasingly possible to support ventilation in the domiciliary setting with newer techniques of noninvasive ventilation and better airway clearance. There remained several areas of vigorous debates, including the role for tracheostomy care and the place of respiratory muscle training and phrenic nerve/diaphragm pacing.

SUMMARY: Recent studies and systematic reviews have defined criteria for anticipating, recognizing, and managing ventilatory failure because of acute neuromuscular disease. The care of patients requiring long-term noninvasive ventilatory support for chronic disorders has also evolved. This has resulted in significantly improved survival for patients requiring domiciliary ventilatory support.

Publication type, MeSH terms

LinkOut - more resources