Severe weakness with respiratory failure is a serious and common complication of critically ill patients in the intensive care unit setting. Recent studies have elucidated 2 entities characterized by severe weakness that occur as a result of these circumstances: critical illness neuropathy and critical illness myopathy. Both are the result of a serious illness, in contrast to weakness arising from a primary neurological disorder that results in admission to an intensive care unit (eg, myasthenic crisis, Guillain-Barré syndrome, rhabdomyolysis, or botulism). Critical illness neuropathy and myopathy are difficult to distinguish from each other on purely clinical grounds, although certain clues may make one more likely. Nerve conduction studies, needle electromyography, and muscle biopsy are often necessary to make a firm diagnosis.

CRITICAL ILLNESS NEUROPATHY

As initially described by Bolton et al in 1984, critical illness neuropathy is a sensorimotor polyneuropathy that is often a complication of sepsis and multiorgan failure, occurring in 70% of such patients. The severity of the underlying illness, the frequently associated encephalopathy, and the use of nondepolarizing neuromuscular blocking agents and ventilator support make recognition of the onset of the neuropathy difficult. It usually becomes apparent when the patient cannot be weaned from the ventilator. Flaccid weakness of the extremities, often severe, and loss of tendon reflexes are associated findings.

Critical illness polyneuropathy is primarily a distal axonopathy in which distal degeneration of both motor and sensory axons, without inflammation, occurs. The underlying cause of the axonal degeneration may relate to a lack of vascular auto-regulation and increased microvascular permeability resulting in endoneurial edema and capillary occlusion. Electrophysiological studies show reduction or absence of both compound muscle and sensory nerve action potentials, fibrillations, and loss of motor unit potentials with a maximal effort. Significant slowing of nerve conduction or nerve conduction blocks are not expected findings and, if present, would implicate other diagnostic possibilities, eg, Guillain-Barré syndrome. Reduction of diaphragmatic compound muscle action potentials and the presence of fibrillations of chest wall muscles reflect the weaning difficulties.

The differential diagnosis includes Guillain-Barré syndrome, acute porphyria, botulism, myasthenic crisis, prolonged effect of nondepolarizing neuromuscular blocking agents, and critical illness myopathy. Multiorgan failure, sepsis, and critical illness polyneuropathy have a mortality rate of 50%. The neuropathy shows spontaneous improvement, with resolution of the underlying illness, but recovery may be limited or absent when the neuropathy is severe. Physical therapy is the only effective rehabilitation therapy available.

CRITICAL ILLNESS MYOPATHY

Critical illness myopathy is underrecognized because it has a clinical appearance that is similar to critical illness polyneuropathy. It has been referred to by a number of different terms in the literature, including acute myopathy, acute quadriplegic myopathy, critical care myopathy, acute necrotizing myopathy, and acute myopathy with selective loss of myosin filaments. Muscle bi-
The prognosis of critical illness myopathy depends on the severity of the underlying illness and age of the patient. Young patients with status asthmatics may have complete recovery in 2 to 3 months.11

CONCLUSIONS

Critical illness neuropathy and critical illness myopathy, either singly or in combination, are a common complication of critical illnesses. Both disorders may lead to severe weakness and require mechanical ventilation. Multigain failure and sepsis predispose to the neuropathy, while a variety of serious problems (eg, pneumonia, severe asthma, and liver or lung transplantation) and the concomitant use of high-dose intravenous corticosteroids and nondepolarizing neuromuscular blocking agents predispose to the myopathy. Minimizing the use of corticosteroids and nondepolarizing neuromuscular blocking agents in the critical illness setting may prove helpful in preventing the occurrence of these disorders. The prognosis is directly related to the age of the patient and the seriousness of the underlying illness.

Accepted for publication May 1, 1998.

Corresponding author: Ludwig Gutmann, MD, Department of Neurology, Robert C. Byrd Health Sciences Center, Morgantown, WV 26506-9180.

REFERENCES

3. Bolton CF, Young GB, Zochodne DW. The pathological features of critical illness myopathy are somewhat complex. Light microscopy commonly shows angulated atrophic myofibers that are predominantly type 2, with basophilic cytoplasm on hematoxylin-eosin stain.5,9,11,13,15 Associated vesicular nuclei and positive staining with alkaline phosphatase suggest that regeneration is occurring. Decreased or absent myosin adenosine triphosphatase staining of atrophic fibers may spread loss of all filaments but often reveals selective staining with alkaline phosphatase suggest that regeneration is occurring. Decreased or absent myosin adenosine triphosphatase staining of atrophic fibers may spread loss of all filaments4,13 but often reveals selective loss of myosin filaments with relative sparing of actin filaments and Z discs.5,6,8,11-15 The electron microscopic findings seem to correlate with the altered staining of atrophic fibers with adenosine triphosphatase staining.

Selective myosin filament loss can be produced in the denervated soleus muscle of rats receiving simultaneous high-dose corticosteroids. The combination of a superimposed polynuropathy (occurring in a number of the patients) or the use of nondepolarizing neuromuscular blocking agents in conjunction with high-dose intravenous corticosteroid therapy may simulate the rat experimental model and result in the selective myosin filament loss.6 The expression of steroid receptors is enhanced in denervated muscle, making myofibers potentially more vulnerable to corticosteroid injury.17

The pathological features of critical illness myopathy are somewhat complex. Light microscopy commonly shows angulated atrophic myofibers that are predominantly type 2, with basophilic cytoplasm on hematoxylin-eosin stain. Associated vesicular nuclei and positive staining with alkaline phosphatase suggest that regeneration is occurring. Decreased or absent myosin adenosine triphosphatase staining of atrophic fibers may spread loss of all filaments but often reveals selective loss of myosin filaments with relative sparing of actin filaments and Z discs. The electron microscopic findings seem to correlate with the altered staining of atrophic fibers with adenosine triphosphatase staining.

The pathological features of critical illness myopathy are somewhat complex. Light microscopy commonly shows angulated atrophic myofibers that are predominantly type 2, with basophilic cytoplasm on hematoxylin-eosin stain. Associated vesicular nuclei and positive staining with alkaline phosphatase suggest that regeneration is occurring. Decreased or absent myosin adenosine triphosphatase staining of atrophic fibers may spread loss of all filaments but often reveals selective loss of myosin filaments with relative sparing of actin filaments and Z discs. The electron microscopic findings seem to correlate with the altered staining of atrophic fibers with adenosine triphosphatase staining.

The pathological features of critical illness myopathy are somewhat complex. Light microscopy commonly shows angulated atrophic myofibers that are predominantly type 2, with basophilic cytoplasm on hematoxylin-eosin stain. Associated vesicular nuclei and positive staining with alkaline phosphatase suggest that regeneration is occurring. Decreased or absent myosin adenosine triphosphatase staining of atrophic fibers may spread loss of all filaments but often reveals selective loss of myosin filaments with relative sparing of actin filaments and Z discs. The electron microscopic findings seem to correlate with the altered staining of atrophic fibers with adenosine triphosphatase staining.

The pathological features of critical illness myopathy are somewhat complex. Light microscopy commonly shows angulated atrophic myofibers that are predominantly type 2, with basophilic cytoplasm on hematoxylin-eosin stain. Associated vesicular nuclei and positive staining with alkaline phosphatase suggest that regeneration is occurring. Decreased or absent myosin adenosine triphosphatase staining of atrophic fibers may spread loss of all filaments but often reveals selective loss of myosin filaments with relative sparing of actin filaments and Z discs. The electron microscopic findings seem to correlate with the altered staining of atrophic fibers with adenosine triphosphatase staining.

The pathological features of critical illness myopathy are somewhat complex. Light microscopy commonly shows angulated atrophic myofibers that are predominantly type 2, with basophilic cytoplasm on hematoxylin-eosin stain. Associated vesicular nuclei and positive staining with alkaline phosphatase suggest that regeneration is occurring. Decreased or absent myosin adenosine triphosphatase staining of atrophic fibers may spread loss of all filaments but often reveals selective loss of myosin filaments with relative sparing of actin filaments and Z discs. The electron microscopic findings seem to correlate with the altered staining of atrophic fibers with adenosine triphosphatase staining.

The pathological features of critical illness myopathy are somewhat complex. Light microscopy commonly shows angulated atrophic myofibers that are predominantly type 2, with basophilic cytoplasm on hematoxylin-eosin stain. Associated vesicular nuclei and positive staining with alkaline phosphatase suggest that regeneration is occurring. Decreased or absent myosin adenosine triphosphatase staining of atrophic fibers may spread loss of all filaments but often reveals selective loss of myosin filaments with relative sparing of actin filaments and Z discs. The electron microscopic findings seem to correlate with the altered staining of atrophic fibers with adenosine triphosphatase staining.

The pathological features of critical illness myopathy are somewhat complex. Light microscopy commonly shows angulated atrophic myofibers that are predominantly type 2, with basophilic cytoplasm on hematoxylin-eosin stain. Associated vesicular nuclei and positive staining with alkaline phosphatase suggest that regeneration is occurring. Decreased or absent myosin adenosine triphosphatase staining of atrophic fibers may spread loss of all filaments but often reveals selective loss of myosin filaments with relative sparing of actin filaments and Z discs. The electron microscopic findings seem to correlate with the altered staining of atrophic fibers with adenosine triphosphatase staining.

The pathological features of critical illness myopathy are somewhat complex. Light microscopy commonly shows angulated atrophic myofibers that are predominantly type 2, with basophilic cytoplasm on hematoxylin-eosin stain. Associated vesicular nuclei and positive staining with alkaline phosphatase suggest that regeneration is occurring. Decreased or absent myosin adenosine triphosphatase staining of atrophic fibers may spread loss of all filaments but often reveals selective loss of myosin filaments with relative sparing of actin filaments and Z discs. The electron microscopic findings seem to correlate with the altered staining of atrophic fibers with adenosine triphosphatase staining.