

# Myopathies in the Intensive Care Unit

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**ABSTRACT:** Myopathies that occur in the intensive care unit can be divided into preexisting myopathies or newly acquired myopathies that develop in the intensive care unit. Myotonic dystrophy is an example of a preexisting myopathy that may render patients susceptible to acute respiratory failure following surgical procedures and anaesthesia. A group of myopathies that develop within the intensive care unit have been labelled acute necrotizing myopathy of intensive care, thick filament myopathy and acute steroid myopathy. Corticosteroids and nondepolarizing muscle blocking agents may play a role in their development.

**RÉSUMÉ:** Myopathies à l'unité de soins intensifs. Les myopathies rencontrées à l'unité de soins intensifs sont soit préexistantes, soit acquises au cours du séjour à l'unité de soins intensifs. La dystrophie myotonique est un exemple d'une myopathie préexistante qui peut précipiter une insuffisance respiratoire aiguë suite à une chirurgie et à une anesthésie. Un groupe de myopathies survenant à l'unité de soins intensifs pourraient être interreliées et ont été identifiées comme la myopathie nécrosante aiguë des soins intensifs, la myopathie à filaments larges et la myopathie aiguë aux stéroïdes. Les corticostéroïdes et les agents curarisants non dépolarisants pourraient jouer un rôle dans leur développement.

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## MYOPATHIES IN CRITICAL ILLNESS

This is a very brief review of both newly acquired and preexisting myopathies that may develop in adults patients admitted to the intensive care unit (Table). The reader is referred to more detailed reviews on this topic.<sup>1</sup>

**Table:** Myopathies in the Intensive Care Unit.

### Preexisting disorders:

- Inflammatory myopathies<sup>22,23</sup>
- Alcoholic rhabdomyolysis<sup>15</sup>
- Myotonic dystrophy<sup>2-5</sup>
- Periodic paralyses
- Malignant hyperthermia<sup>21</sup>
- Acquired hypokalemic weakness<sup>15</sup>
- Acid maltase deficiency<sup>6-14</sup>
- Mitochondrial myopathies<sup>17</sup>
- Limb girdle muscular dystrophy<sup>18</sup>
- HIV related myopathy<sup>19</sup>
- Trichinosis<sup>25</sup>
- Sarcoid myopathy<sup>20</sup>
- Carnitine palmitoyl transferase deficiency<sup>16</sup>
- Hypophosphatemic myopathy<sup>15,24</sup>

### Myopathies that develop in the intensive care unit:

- Necrotizing myopathy associated with non-depolarizing muscle blocking agents<sup>26,29</sup>
- Acute steroid myopathy<sup>27,28</sup>
- Septic myopathy<sup>39</sup>
- Myopathy of critical illness<sup>39</sup>
- Hypophosphatemia<sup>15,24</sup>

## General Features

It is uncommon, but not rare to make the diagnosis of a more longstanding myopathy in an adult for the first time in the intensive care unit. Other myopathies may be iatrogenic. Patients in the intensive care unit with myopathies may have varying degrees of bulbar involvement including ptosis and neck flexor weakness. Reflexes are lost in some patients but sensation is preserved. Electrophysiological features include reduced M potential amplitudes, normal motor conduction velocity and preserved sensory conduction. Needle electromyography may disclose abnormal spontaneous activity such as fibrillation potentials, positive sharp waves and myotonic discharges. Fibrillations are less numerous than those recorded after denervation and are probably associated with muscle fiber necrosis. If the patient can recruit motor units, they may be reduced in amplitude and duration.

## Patients With Preexisting Myopathies

Patients with myotonic dystrophy have an important but avoidable susceptibility to acute respiratory failure following surgical procedures requiring the use of anaesthesia, opioids and sedatives.<sup>2</sup> The cause of this susceptibility is uncertain but may be due to an abnormality of central respiratory drive linked to hypersomnia.<sup>3-5</sup> There is no evidence that specific treatment for myotonia facilitates more rapid weaning. Acid maltase deficiency, an autosomal recessive deficiency of glycogen degrading

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alpha-1, 4-glucosidase may be associated with a myopathy and respiratory insufficiency in adults.<sup>6-14</sup> Myotonia may be a prominent feature on EMG testing. Muscle biopsy may identify accumulation of PAS positive glycogen but specific enzyme assays are required for the diagnosis. Ventilatory support is important in these patients because spontaneous recovery may occur, even after prolonged ventilator dependence.

There are a variety of other myopathies that rarely might render enough weakness to place the patient in an intensive care unit. These might include: a rhabdomyolytic syndrome from hypokalemia,<sup>15</sup> myopathy associated with carnitine palmitoyl transferase deficiency,<sup>16</sup> certain mitochondrial myopathies,<sup>17</sup> limb girdle muscle dystrophy,<sup>18</sup> HIV myopathy<sup>19</sup> and sarcoid myopathy involving the diaphragm.<sup>20</sup> Patients with malignant hyperthermia may be admitted to an intensive care unit during crisis.<sup>21</sup> There are reports of patients with inflammatory myopathies requiring admission to the unit.<sup>22,23</sup> Finally, one might include acute alcoholic rhabdomyolysis<sup>15</sup> myopathy from hypophosphatemia<sup>15,24</sup> and myositis from trichinosis.<sup>25</sup>

### Myopathies That Develop in the Intensive Care Unit

The most important of these occurs in patients that have been exposed to high doses of glucocorticoids and infusions of non-depolarizing muscle blocking agents to treat acute pulmonary disorders, such as asthma. They have been labelled as an acute necrotizing myopathy of intensive care, floppy person syndrome, thick filament myopathy, acute quadriplegic myopathy, or acute steroid myopathy.<sup>26-29</sup> There may be a severe flaccid areflexic quadriplegia, sometimes with complete ophthalmoplegia. Electrophysiological studies may identify a persistent neuromuscular transmission deficit<sup>26,30</sup> in instances where vecuronium has been given. This is from the accumulation, and activity of a metabolite of vecuronium, 3 desacetyl-vecuronium.<sup>30</sup> The presence of myopathy is indicated by only partial recovery of the patient despite later (after 1 week) complete repair of the neuromuscular transmission deficit. At this time there may be persistently low amplitude motor M potentials with preserved motor conduction velocity and relatively intact sensory conduction. Needle electromyography may detect mild to moderate abnormal spontaneous activity consisting of fibrillation potentials and positive sharp waves. If the patient is able to recruit motor unit potentials, they may be reduced in amplitude and highly polyphasic suggesting a primary myopathy. Serum CK levels may rise very high, especially with vecuronium because of rhabdomyolysis rendering myoglobinuria, and acute renal failure. Muscle biopsy may identify milder changes such as fiber atrophy and myosin filament loss, or severe muscle fiber necrosis.<sup>29,31</sup> Necrosis may be panfascicular with fibers exhibiting vacuolation or hypereosinophilia.<sup>29</sup> If the patient is appropriately supported complete recovery can occur. Critical illness neuropathy may also occur in these patients.<sup>26</sup> Some instances, where the patient has been treated with high dose steroid alone for asthma, are probably examples of steroid myopathy alone. These patients may only have thick filament loss and Type II fiber atrophy. In other cases, where patients exposed to vecuronium develop rhabdomyolysis with less or no exposure to steroid, there may be a unique toxic action of vecuronium on muscle. Whether additional factors, such as age or preexisting nutrition influence the development of these myopathies is uncertain.

Patients with sepsis that have not been exposed to steroid and nondepolarizing muscle blocking agents can develop myofiber necrosis sometimes linked to specific organisms, such as *Escherichia coli* septicaemia,<sup>32</sup> *Leptospirosis*,<sup>33,34</sup> *Legionella* disease<sup>35</sup> and *staphylococcus aureus*.<sup>36-38</sup> Muscle biopsy may, on occasion identify multiple microabscesses. In previous work, we have suggested that sepsis may have a direct action on muscle resulting in a loss of phosphocreatine energy stores with resulting contractile failure.<sup>39</sup> Sepsis itself may also be associated with catabolic muscle proteolysis, but it is controversial whether this alone can account for a myopathy involving respiratory muscles. CK and electrophysiological studies are normal in catabolic myopathy.

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