# Myopathies


- Author: Courtney A Bethel, MD, MPH; Chief Editor: Barry E Brenner, MD, PhD, FACEP more...


Updated: Dec 27, 2015

## Practice Essentials

Myopathy is a muscle disease unrelated to any disorder of innervation or neuromuscular junction. This condition has widely varying etiologies, including congenital or inherited, idiopathic, infectious, metabolic, inflammatory, endocrine, and drug-induced or toxic.

### History

Important information to obtain during the patient's history includes the following:

- Family history: Any periodic paralysis or muscular dystrophy?
- Personal history: Presence of autoimmune disease, endocrinopathy, renal insufficiency, and/or alcoholism? Previous episodes of severe weakness (eg, postexercise, after exposure to cold [possibly one of periodic paralyses]; post high-carbohydrate meals [familial hypokalemic periodic paralysis])
- Medications (eg, steroids, lipid lowering agents, retroviral agents, alcohol, colchicine, pentachlorophenol [PCP], heroin)
- Occupational and travel history (potential ingestion of barium chloride or carbonate [acute hypokalemic paralysis])

### Signs and symptoms

The common symptoms of myopathy are muscle weakness, impaired function in activities of daily life, and, rarely, muscle pain and tenderness. Significant muscle pain and tenderness without weakness should prompt consideration of other causes.

General signs and symptoms of myopathy include the following:

- Symmetric proximal muscle weakness
- Malaise, fatigue
- Dark colored urine (suggests myoglobinuria) and/or fever
- Absence of sensory complaints or paresthesias; however, deep tendon reflexes (DTRs) may be diminished/absent in hypokalemic paralysis
- Very late findings: Atrophy and hyporeflexia (early presence usually implicates neuropathies)
- Normal level of consciousness
- Gottron papules in dermatomyositis: Pink-to-violaceous scaly areas over knuckles, elbows, and knees

The acuity of symptom onset may aid in the diagnosis, as follows:

- Weakness progressing over hours: Possible toxic etiology or one of episodic paralyses
- Weakness developing over days: May be an acute dermatomyositis or rhabdomyolysis
- Symptom development over a period of weeks: May be polymyositis, steroid myopathy, or myopathy resulting from endocrine causes (eg, hyperthyroidism, hypothyroidism)

Indications of which muscle groups are involved include the following symptoms:

- Proximal muscle weakness: Difficulty rising from chairs, getting out of the bathtub, climbing stairs, and/or shaving or combing the hair
- Weakness of distal muscles: Weak grasp, handwriting problems, and walking difficulties, (eg, flapping gait)

See Clinical Presentation for more detail.

### Diagnosis

*Laboratory testing*

The following laboratory tests may be used to evaluate patients with myopathies:

- Creatine kinase (CK) levels with isoenzymes
- levels of electrolytes, calcium, and magnesium
- Serum myoglobin levels
- Serum creatinine and blood urea nitrogen levels
- Urinalysis: Myoglobinuria indicated by positive urinalysis with few red blood cells on microscopic evaluation
- Complete blood count
- Erythrocyte sedimentation rate
- Thyroid function tests
- Aspartate aminotransferase levels

Other studies may include the following:

- Electrocardiography
- Antinuclear antibody levels
- Genetic testing
- Electromyography
- Magnetic resonance imaging (to assess complications or rule out neurologic disease)

- Muscle biopsy

See Workup for more detail.

### Management

The treatment of a myopathy is dependent on its etiology and can range from supportive and symptomatic management to therapy for specific conditions. Such treatments may include the following:

- Supportive: Management of airway, breathing, circulation; hydration; intensive care management may be needed in some cases
- Drug therapy
- Physical therapy
- Bracing
- Surgery

See Treatment for more detail.

## Contributor Information and Disclosures

Author
**Courtney A Bethel, MD, MPH** Clinical Assistant Professor, Department of Emergency Medicine, Mercy Catholic Medical Center, Drexel University School of Medicine

Courtney A Bethel, MD, MPH is a member of the following medical societies: American Academy of Emergency Medicine, American College of Emergency Physicians

Disclosure: Nothing to disclose.

Specialty Editor Board
**Francisco Talavera, PharmD, PhD** Adjunct Assistant Professor, University of Nebraska Medical Center College of Pharmacy; Editor-in-Chief, Medscape Drug Reference

Disclosure: Received salary from Medscape for employment. for: Medscape.

**Paul Blackburn, DO, FACOEP, FACEP** Attending Physician, Department of Emergency Medicine, Maricopa Medical Center

Paul Blackburn, DO, FACOEP, FACEP is a member of the following medical societies: American College of Emergency Physicians, Arizona Medical Association, American College of Osteopathic Emergency Physicians, American Medical Association

Disclosure: Nothing to disclose.

Chief Editor
**Barry E Brenner, MD, PhD, FACEP** Professor of Emergency Medicine, Professor of Internal Medicine, Program Director for Emergency Medicine, Case Medical Center, University Hospitals, Case Western Reserve University School of Medicine

Barry E Brenner, MD, PhD, FACEP is a member of the following medical societies: Alpha Omega Alpha, American Heart Association, American Thoracic Society, Arkansas Medical Society, New York Academy of Medicine, New York Academy of Sciences, Society for Academic Emergency Medicine, American Academy of Emergency Medicine, American College of Chest Physicians, American College of Emergency Physicians, American College of Physicians

Disclosure: Nothing to disclose.

Additional Contributors
**Jerry R Balentine, DO, FACEP, FACOEP** Vice President, Medical Affairs and Global Health, New York Institute of Technology; Professor of Emergency Medicine, New York Institute of Technology College of Osteopathic Medicine

Jerry R Balentine, DO, FACEP, FACOEP is a member of the following medical societies: American College of Emergency Physicians, New York Academy of Medicine, American College of Osteopathic Emergency Physicians, American Association for Physician Leadership, American Osteopathic Association

Disclosure: Nothing to disclose.

## References

1. Nance JR, Dowling JJ, Gibbs EM, Bonnemann CG. Congenital myopathies: an update. *Curr Neurol Neurosci Rep*. 2012 Apr. 12(2):165-74. [Medline].
2. Valiyil R, Christopher-Stine L. Drug-related myopathies of which the clinician should be aware. *Curr Rheumatol Rep*. 2010 Jun. 12(3):213-20. [Medline]. [Full Text].
3. Elston MS, Orr-Walker BJ, Dissanayake AM, Conaglen JV. Thyrotoxic, hypokalaemic periodic paralysis: Polynesians, an ethnic group at risk. *Intern Med J*. 2007 May. 37(5):303-7. [Medline].
4. Kuncl RW. Agents and mechanisms of toxic myopathy. *Curr Opin Neurol*. 2009 Oct. 22(5):506-15. [Medline].
5. Carstens PO, Schmidt J. Diagnosis, pathogenesis and treatment of myositis: recent advances. *Clin Exp Immunol*. 2014 Mar. 175 (3):349-58. [Medline].
6. Ahlawat SK, Sachdev A. Hypokalaemic paralysis. *Postgrad Med J*. 1999 Apr. 75(882):193-7. [Medline].

7. Barclay L. Genetic marker may identify risk for statin-induced myopathy. Medscape Medical News from WebMD. August 31, 2013. Available at http://www.medscape.com/viewarticle/810297. Accessed: September 9, 2013.
8. Engel AG. Diseases of muscles (myopathies) and neuromuscular junction. Plum F, Bennett JC, eds. *Cecil Textbook of Medicine*. 20th ed. Philadelphia: WB Saunders Co; 1996. 1500-03, 2158-73.
9. Fauci AS, Braunwald E, Isselbacher KJ, et al, eds. *Harrison's Principles of Internal Medicine*. 14th ed. New York: McGraw-Hill; 1998. 2473-2483.
10. Griggs RC, Ptacek LJ. The periodic paralyses. *Hosp Pract (Off Ed)*. 1992 Nov 15. 27(11):123-6, 129-30, 136-7. [Medline].
11. Mangravite LM, Engelhardt BE, Medina MW, et al. A statin-dependent QTL for GATM expression is associated with statin-induced myopathy. *Nature*. 2013 Oct 17. 502 (7471):377-80. [Medline].
12. Plate AM, Boyle BA. Musculoskeletal manifestations of HIV infection. *AIDS Read*. 2003 Feb. 13(2):62, 69-70, 72, 76. [Medline].
13. Riggs JE, Schochet SS, Joynt RJ, Griggs RC, eds. Muscle disease. *Clinical Neurology*. 1997. Vol 4: 1-37.
14. Stedwell RE, Allen KM, Binder LS. Hypokalemic paralyses: a review of the etiologies, pathophysiology, presentation, and therapy. *Am J Emerg Med*. 1992 Mar. 10(2):143-8. [Medline].
15. Stobo JD, Hellman DB. Myopathies. *The Principles and Practice of Medicine*. 23rd ed. New York: McGraw-Hill; 1996. 898-904.
16. Tintinelli JE, Krome RL, Ruiz E. *Emergency Medicine: A Comprehensive Study Guide*. 4th ed. New York: McGraw-Hill; 1996. 1036.
17. Wortmann RL. Lipid-lowering agents and myopathy. *Curr Opin Rheumatol*. 2002 Nov. 14(6):643-7. [Medline].
18. Angelini C. Spectrum of metabolic myopathies. *Biochim Biophys Acta*. 2015 Apr. 1852 (4):615-21. [Medline].
19. Cherin P, Belizna C, Odile C, et al. Long-term subcutaneous immunoglobulin use in inflammatory myopathies: A retrospective review of 19 cases. *Autoimmun Rev*. 2015 Dec 11. [Medline].
20. Mathevon L, Michel F, Decavel P, Fernandez B, Parratte B, Calmels P. Muscle structure and stiffness assessment after botulinum toxin type A injection. A systematic review. *Ann Phys Rehabil Med*. 2015 Dec. 58 (6):343-50. [Medline].
21. Tansley SL, McHugh NJ. Serological subsets of Juvenile Idiopathic Inflammatory myopathies - an update. *Expert Rev Clin Immunol*. 2015 Dec 11. [Medline].
22. Allen DG, Whitehead NP, Froehner SC. Absence of dystrophin disrupts skeletal muscle signaling: roles of Ca2+, reactive oxygen species, and nitric oxide in the development of muscular dystrophy. *Physiol Rev*. 2016 Jan. 96 (1):253-305. [Medline].