11 The Muscular System

Objectives

In this chapter we will study
- methods used to diagnose muscle disorders; and
- some structural disorders of skeletal muscle—rhabdomyolysis, myositis, and the muscle tumor rhabdomyosarcoma.

Diagnosing Structural Disorders of the Muscular System

The body contains about 600 skeletal muscles, specialized organs that produce movement of body parts and perform several other essential and overlapping functions. Health-care professionals employ a knowledge of muscle form and function when they turn patients, give injections, or engage in the diagnosis and treatment of muscular system disorders (myopathies). Although muscle structure and function cannot be strictly separated, we deal with myopathies of a primarily structural nature in this chapter and with more functional disorders in chapter 12.

The symptoms of muscular disorders can be confusing because some of them, such as pain upon movement, are also produced by disorders of the nervous and skeletal systems. A skilled clinician must be able to determine which system is responsible for a given symptom so that proper treatment can be initiated.

The most common symptom of a muscular disorder is pain in the affected muscle or muscles; the second most prevalent symptom is weakness. Either of these symptoms may be due to nervous or skeletal system pathologies, trauma to the muscle itself, or muscle infections. Additional symptoms include muscle atrophy, tenderness, stiffness, and cramping. When a patient presents with these rather broad symptoms, how does a clinician complete the diagnostic process?

During the physical examination, observing the patient’s gait and posture can help detect disorders of muscles in the limbs or trunk, while assessing the patient’s speech and facial movements provides information about the musculature of the head. In addition, various diagnostic tests can be conducted to evaluate muscle strength and motion.

Often muscle strength is first tested manually. Conducting this exam requires the clinician to understand muscle anatomy and the functional relationships of the muscles in a given region. For example, to test the quadriceps femoris muscle, which extends the knee, the clinician may ask the patient to sit on an examination table and then lift and straighten his knee. This will show whether the patient can lift his leg against gravity. If he can do this, the clinician may next push down on the lower leg as the patient tries to straighten his knee against this resistance. This exercise tests the muscle’s ability to work against an externally applied force. The clinician judges the amount of force the patient can generate and assigns a grade according to the guidelines followed at the particular clinic. There are three commonly used scales: (1) a percentage scale, which rates muscle strength between 0% and 100%; (2) a terminology-based scale, in which muscle strength can range anywhere between “trace/zero” and “good/normal”; and (3) a numerical scale, which ranges from 1 for “no contraction” to 5 for “normal contraction strength.” To further differentiate muscle strength when using this scale, a plus (+) or minus (–) may be added to the number grade.

In clinical settings, muscle force is also frequently evaluated using various devices called myometers or dynamometers, which measure the extent and force of muscle contraction at individual joints. As noted in chapter 10, the goniometer measures the range of motion of a joint in degrees.

Other tests that may help diagnose muscular disorders include magnetic resonance imaging (MRI) and electromyography. An MRI can distinguish between soft tissues, making it extremely useful in muscle evaluations. Electromyography is the recording of the electrical activity of a muscle, using either a needle electrode inserted into the muscle or electrodes applied to the skin overlying the muscle.
Electromyography can help differentiate muscle diseases from diseases of the nervous system or of the neuromuscular junction.

Muscle biopsies are sometimes performed to diagnose muscle infections and diseases that alter muscle structure. In this procedure, a small sample of muscle tissue is removed and then examined with a light microscope to detect histological or histochemical abnormalities or with an electron microscope to assess changes in cell ultrastructure (the thick and thin filaments, organelles, and plasma membrane).

A battery of blood and urine tests can also help identify muscle dysfunctions. Important measurements include:

- muscle enzymes that have leaked into the blood, such as creatine kinase (CK), lactate dehydrogenase (LDH), aldolase, or aspartate transaminase;
- changes in the concentration of electrolytes (such as calcium and potassium), which are normally more concentrated in the skeletal muscle than in the blood; and
- the concentration of muscle myoglobin in the urine.

The normal ranges for these clinical tests are given in the Appendix of Normal Values at the end of this manual. The most useful of these tests measures serum CK concentration. Because this enzyme is found in high quantities in muscle fibers, muscle damage releases CK into the tissue fluid and consequently into the blood. Like CK, myoglobin is released from muscle cells by trauma, ischemia, extreme exertion, and certain genetic disorders; measurable amounts quickly appear in the urine. The serum aldolase level can help distinguish between muscular dystrophy, which raises the aldolase level, and such diseases as myasthenia gravis and multiple sclerosis, which do not.

Changes in serum electrolyte concentrations can result in muscle pathologies, so measuring the electrolytes provides information about the possible cause of a change in muscle function. For example, muscle weakness can stem from decreased serum potassium or elevated serum calcium concentrations, while muscle twitches and tetany can be induced by decreased serum calcium concentrations.

Finally, the forearm ischemic exercise test enables a clinician to test the enzymatic pathways used for energy production in the muscle. The patient performs a series of strenuous exercises employing a maximum grip strength and graded workloads. Blood is drawn for up to 20 minutes after the conclusion of the exercise and then analyzed for lactate and serum enzymes. The exercise induces lactic acid fermentation and raises the lactate level in the blood, but an excessively elevated lactate level may indicate mitochondrial disorders, and an abnormally low lactate level suggests disorders of the glycolytic pathway. Ammonia levels normally rise along with the lactate level, but if no increase in ammonia concentration is seen, this suggests a deficiency of the enzyme monoadenylate deaminase, which is important in making creatine phosphate and ATP. A deficiency of this enzyme can make a person unable to tolerate exercise.

**Structural Disorders of Skeletal Muscle**

**Rhabdomyolysis**

Rhabdomyolysis is a severe, potentially fatal destruction of muscle cells. The breakup of muscle cells releases myoglobin, which soon appears in the urine, a condition called myoglobinuria. In fact, myoglobinuria is such a common sign of rhabdomyolysis that the two terms are sometimes used synonymously.

The presence of myoglobin gives the urine a dark, reddish-brown color. Damage to as little as 200 grams of muscle is sufficient to induce a change in the appearance of the urine. Serum CK is also elevated in myoglobinuria, often to a concentration 100 times greater than normal.

The most severe form of rhabdomyolysis is crush syndrome—massive destruction of muscle tissue resulting from major trauma such as war injuries, getting a limb caught in farm or factory machinery, or being run over by a vehicle. Such serious injuries can release enough myoglobin to block kidney tubules and cause death from renal failure. Another form of rhabdomyolysis is compartment syndrome, which involves selective injury to the muscles within a particular compartment of the upper or lower limb. Rhabdomyolysis can also be caused by viral infections, bacterial tetanus, heatstroke, fractures, excessive muscular activity, or certain anesthetics.
When a limb is injured, the mere weight of the immobile body part can cause muscle ischemia. The resulting decrease in venous drainage allows fluid to accumulate in the limb, causing edema (swelling). This edema causes the pressure within the compartment to rise, and if not relieved, it may further compress the blood vessels in the compartment, reduce blood flow, and cause cells to die from a lack of oxygen or from accumulation of metabolic wastes. As muscular tissue dies, it is replaced by scar tissue. This can lead to an abnormal shortening of the muscle called contracture.

Management of rhabdomyolysis is aimed at diagnosing and treating the underlying cause. It is particularly urgent, however, to relieve the pressure in the muscle compartment and to prevent or reverse the myoglobinuria, which can cause renal failure if left untreated. Compartment pressure can be reduced by fasciotomy (cutting the muscle fascia) to relieve pressure and, if necessary, debridement (removing dead and nonviable tissue from the injury site).

Myositis

Myositis is any form of muscle inflammation. It may be caused by a viral, bacterial, or parasitic infection. Three types of myositis are described here: trichinosis, polymyositis, and dermatomyositis.

Trichinosis is caused by the larvae of several species of parasitic roundworms in the genus Trichinella. These larvae can be found encysted in the muscles of infected mammals, including humans, dogs, pigs, horses, and wild game. People are most commonly exposed when they eat undercooked meat, especially pork, but also wild game such as bears and seals. Thorough cooking or freezing destroys the larvae and eliminates the risk of trichinosis. After being ingested, the larvae migrate to the intestinal mucosa where they develop, mate, and produce new larvae. These new larvae then move from the intestine into the lymphatic and circulatory systems, which transport them to muscles throughout the body. Once there, the larvae penetrate individual muscle fibers and encyst.

Symptoms of trichinosis include severe muscle and joint pain, generalized muscle stiffness, rash, and edema. Diagnosis is often accomplished through muscle biopsy. It is estimated that up to 4% of the U.S. population is infected with Trichinella at any given point in time. Of the people who experience symptoms severe enough to require treatment, approximately 1% die. Death usually results from tissue damage caused by larvae migrating through the heart, kidneys, and respiratory muscles.

Polymyositis is an inflammation of multiple muscles at one time; it is sometimes accompanied by characteristic skin lesions and is then called dermatomyositis. Although these two conditions affect only 6 out of 1 million people, they are the most frequent types of myositis requiring chronic care. The causative agent has not been identified, but it is believed to affect the immune system as well as the muscular system.

Both polymyositis and dermatomyositis are characterized by inflammation of the muscle and associated connective tissue, which is often severe enough to induce necrosis and destroy muscle fibers. Both diseases produce a wide variety of symptoms, including swollen, tender, or painful muscles, fever, lethargy, and malaise. In addition to these symptoms, symmetrical muscle weakness occurs—that is, the corresponding muscles on both sides of the body are similarly weak. Dermatomyositis is characterized by a purple-colored rash on the face, chest, eyelids, and upper limbs.

Because the symptoms are so nonspecific, both diseases are difficult to diagnose, and the clinician must complete a thorough examination in order to rule out other diseases with similar symptoms. The most helpful diagnostic procedures are muscle biopsy, MRI, and clinical laboratory tests. Definitive signs of dermatomyositis revealed by a biopsy are atrophied muscle fascicles and a large number of inflammatory cells surrounding the blood vessels. MRI results support the biopsy findings, showing inflammation and muscle edema. The primary clinical laboratory finding is an elevated serum CK concentration.

Treatments for polymyositis and dermatomyositis center on reducing the inflammation (often by administering anti-inflammatory drugs) and prescribing physical therapy to minimize muscle atrophy and contractures.

Muscle Tumors

Muscle tumors occur most frequently in children. They may arise either from muscle tissue or from associated tissues such as adipose tissue, synovial membranes, or nervous tissue.

Rhabdomyoma is a rare, benign muscle tumor. It most often occurs in the tongue, neck, larynx, nasal
cavity, uvula, heart, and vulva. The treatment is simply to remove the tumor surgically; they usually do not recur.

Malignant skeletal muscle tumors, termed rhabdomyosarcomas (RMS), are seldom seen in adults. Even in children, RMS accounts for less than 3% of cancer cases, but it is the most common form of pediatric soft-tissue sarcoma. It usually appears at the age of 2 to 6 years or 15 to 19 years, with no difference between the sexes.

During fetal development, some embryonic myoblasts (the embryonic cells that give rise to skeletal muscle fibers) become cancerous rather than differentiating into normal striated muscle. These abnormal myoblasts are called rhabdomyoblasts. The cause of the transformation is as yet unknown, but one tumor suppressor gene (p53) and three oncogenes (c-myb, src, and H/K-ras) may be involved. Any skeletal muscle can develop a tumor, but RMS is most common in the head and neck, trunk, limbs, and urogenital tract. RMS metastasizes frequently and rapidly to the lymph nodes, bone, bone marrow, brain, liver, heart, and lungs.

The signs and symptoms of RMS are determined by tumor location. In most cases, there is no pain, and the tumor is first detected when a visible or palpable mass is noticed. If a tumor is suspected, imaging techniques are used to confirm its location and possible metastatic sites. Final diagnosis is achieved by biopsy. Metastasis and organ involvement are confirmed by blood tests and organ-specific evaluations, such as measuring liver function or obtaining bone marrow by aspiration (suction) for microscopic examination.

Treatment combines surgery, chemotherapy, and radiation therapy. If the tumor is localized (has not spread), the survival rate is high (70–80%). However, with widespread metastasis, the probability of survival drops to 20%

Case Study 11  The Pitcher with a Sore Arm

Jason, a 20-year-old college junior, is getting ready for the start of the baseball season by working out with some friends over the winter. Since he is hoping to be selected as one of the top two starting pitchers, he spends many hours in the weight room improving his strength and conditioning. In addition, he has a friend catch for him each day. Jason starts working out slowly and carefully because he is concerned about overtraining and incurring injury before he has gotten in shape.

As the start of the baseball season approaches, Jason does not feel he has progressed enough, so he increases the duration and intensity of his workouts. He also decides that his conditioning is sufficient to spend most of his time pitching. This approach seems to work because the coaches choose him as one of the starting pitchers for the team.

About five games into the season, Jason notices that his pitching arm tires more quickly than it did earlier. At the beginning of the season, he felt he could pitch an entire game, but now he is tiring by the sixth or seventh inning. He also notices that his fastball no longer has its “zip,” so he begins relying more on his curve ball. This tactic seems successful in the next two games, but by the eighth game of the season, Jason can’t get any of his pitches over the plate with enough velocity to keep the batters from hitting them. The coach removes him from the game in the second inning and sends him to the team physician for evaluation.

After obtaining a complete history, the physician conducts a physical examination and orders a series of tests, including evaluation of serum enzyme and electrolyte concentrations, an MRI of Jason’s pitching shoulder, and range-of-motion and strength tests on the joint. The results of these tests are presented here.

Visual examination: Right shoulder “droop” and lack of free movement of right arm during walking. Right shoulder and arm held more closely to body.

Muscle strength: All normal with the exception of the right arm rotator cuff muscles, which are given a grade of 4– in a manual test.

Range of motion: Normal for all areas tested except for the right shoulder, which is reduced and guarded.

MRI: Inflammation of muscles of the right shoulder and arm.
Laboratory tests:
- Serum creatine kinase (CK) = 200 IU/L
- Serum lactate dehydrogenase (LDH) = 190 IU/L
- Serum potassium = 4.2 mEq/L
- Serum calcium = 0.2 mEq/L

Based on the test results, the team physician diagnoses a strained rotator cuff. He tells Jason to rest his shoulder for the next 2 weeks and then come back for reevaluation. In the meantime, he recommends that Jason ice his shoulder, wrap it with an elastic bandage, and use a sling. The physician also advises Jason to take Motrin twice daily to reduce the pain and inflammation.

Upon reevaluation 2 weeks later, the physician allows Jason to return to practice but cautions him about overexertion. He warns Jason that since his rotator cuff has been inflamed, the chances of more serious damage, such as tearing, are greater for him than for other pitchers.

Based on this case study and other information in this chapter, answer the following questions.

1. Why is Jason given an MRI scan rather than an X-ray?
2. Why do Jason’s symptoms develop during the season and not during training?
3. Why does the team physician suspect a rotator cuff injury? How do the results of the physical examination support that diagnosis?
4. List the muscles of the rotator cuff.
5. If this injury affected the nerve supply to the muscles of the rotator cuff, which nerve(s) would you suspect, and why?
6. Joyce has broken her femur in a skiing accident and had her leg in a cast for several weeks. When the cast is removed, she is referred to a physical therapist. After having Joyce extend her leg at the knee from a sitting position, the therapist pushes down with her hands on Joyce’s shin while asking her to resist the downward force. Which of the following muscles is being tested?
   a. biceps femoris
   b. piriformis
   c. quadriceps femoris
   d. sartorius
   e. tensor fasciae latae

7. For some time, Adam has been experiencing fever, lethargy, muscle swelling and tenderness, and a rash on his face, chest, and eyelids. After a number of blood tests and treatments with various anti-inflammatory agents, the symptoms have not been alleviated—in fact, they seem to be worsening. The strength of Adam’s upper limb muscles is measured at significantly below normal (3–) on both sides of his body. Based on these signs and symptoms, his physician orders a muscle biopsy. Which muscle disorder does the physician suspect? What questions could the physician ask to aid his diagnosis, and how can the biopsy help identify the disease affecting Adam?

8. A patient has drooping upper eyelids and is unable to raise them. Determine the muscles affected and what nerve(s) may be involved.

9. John, a 25-year-old construction worker, is brought to the hospital emergency room with severe damage to his left forearm. The injury occurred when he caught his arm between a wall and some falling steel beams as part of a building collapsed; it took approximately 2 hours to free him. In the emergency room, his arm appears swollen, and laboratory tests reveal elevated concentrations of serum CK, LDH, and aldolase. As John’s condition is monitored, the hospital staff should be alert for
   a. the appearance of dark, reddish-brown urine.
   b. atrophy of the muscles of the left forearm.
   c. contractures in the muscles of the left forearm.
   d. all of the above.
   e. none of the above.

10. Why are muscle pain and stiffness characteristic of trichinosis?
Selected Clinical Terms

**compartment syndrome**  A disorder in which abnormally high pressure in any confined anatomical space impedes blood flow and thus leads to dysfunction of nerves and muscles within the space.

**contracture**  Abnormal shortening of a muscle due to fibrosis, spasm, or other causes, resulting in reduced joint mobility.

**crush syndrome**  A shocklike state occurring when muscles are crushed and the pressure is then relieved; characterized by reduced urine output resulting from damage to the kidney tubules by myoglobin released from the injured muscles.

** electromyography**  The diagnostic recording of electrical activity of the muscles, using either surface electrodes on the skin or needle electrodes inserted into the muscle.

**goniometer**  An instrument for measuring the angle through which an individual can move a joint.

**myometer**  An instrument for measuring the force and extent of muscle contraction.

**myopathy**  Any pathology involving any muscular tissue, but especially referring to the muscular system (skeletal muscle).

**myositis**  Inflammation of a skeletal muscle.

**polymyositis**  Chronic, progressive inflammation of multiple skeletal muscles, characterized by symmetric pain and weakness of the neck, pharynx, and limb muscles, and often accompanied by skin lesions.

**rhabdomyolysis**  An acute, potentially fatal breakdown of skeletal muscle cells characterized by myoglobin in the blood and urine.

**rhabdomyoma**  A benign tumor of either skeletal or cardiac muscle.

**rhabdomyosarcoma (RMS)**  A highly malignant tumor originating in skeletal muscle.

**trichinosis**  Infection with parasitic roundworms of the genus *Trichinella*, typically acquired by eating undercooked pork or other meat; causes myositis when worm larvae invade and encyst in the skeletal muscles.