A common complaint of many patients in a primary care setting is “Doctor, I am weak.” One could accept this outright as a manifestation of loss of strength in some muscle groups but a wiser approach is to ask the patient to specifically define what he or she means by weakness. The answer is often surprising and may encompass complaints as varied as fatigability, apathy, loss of sensation, imbalance or excessive drowsiness. If, indeed, the complaint is actual loss of strength in muscle groups, then an accurate history directed to causes of weakness must be obtained.

Information important for the evaluation of weakness includes:

The temporal profile of weakness onset:
- Acute
- Subacute
- Chronic

The distribution of the weakness:
- Proximal
- Distal
- Symmetric
- Asymmetric
  - Hemiparesis (one-half of the body)
  - Paraparesis (lower extremities)
  - Monoparesis (one extremity)
  - Focal weakness (portion of an extremity or the face)

Once the history is obtained the neurological examination should confirm the presence of any weakness as well as its distribution. Important associated findings are muscle atrophy, pain and tenderness, swelling, fasciculations (a visible twitch under the skin caused by the spontaneous firing of a motor unit...
[one anterior horn cell and all the muscle fibers it innervates]), deep tendon reflex changes and sensory loss.

The following cases will illustrate different types of weakness, with their associated histories and physical findings.

**Case 1**

A 71-year-old male is eating breakfast and has abrupt onset of visual loss in the left eye and weakness in the right arm and leg. There is some tingling of the right hand and right corner of his mouth. He has difficulty standing and is taken to the emergency room where you are called to see him. On exam his vision has recovered but he has weakness of the right arm involving the triceps, wrist and finger extensors and the right leg involving the hamstrings (lower leg flexors) and anterior tibial (foot dorsiflexors). The deep tendon reflexes are more brisk on the right, and he has a right Babinski sign. Sensory exam reveals decreased sensation in the right hand and arm, and less on the right the face and leg. He has difficulty identifying objects placed in his hand by sensation alone. The patient has no difficulty understanding speech, but speaks infrequently and with a paucity of otherwise meaningful words.

General physical examination reveals a left carotid bruit and a normal sinus rhythm.

**Analysis.** The patient is elderly and had abrupt onset of weakness (acute), with a distribution best classified as a hemiparesis. Associated symptoms are ipsilateral sensory loss and contralateral monocular blindness. The acute onset suggests a vascular etiology and the clinical findings best fit the brain location supplied by the left internal carotid artery.

The first branch of the left internal carotid artery is the ophthalmic artery (OA). Decreased blood flow in this artery would produce transient monocular visual loss, known as amaurosis fugax (fleeting blindness). The patient usually experiences this as a curtain being pulled down over the eye, lasting several minutes, and then clearing. The retina often recovers due to collateral circulation from the external carotid artery (ECA), and vision returns.

The carotid artery then divides into the middle and anterior cerebral artery. The middle cerebral artery (MCA) supplies the lateral hemisphere (see homunculus, Chapter 1, Figure 2-22). The prefrontal motor area supplied by the MCA encompasses the face, hand, upper extremity and trunk. The motor cortex supplied by the anterior cerebral artery (ACA), encompasses the leg. The MCA and ACA supply analogous sensory areas (parietal).

The best diagnosis is stroke in the distribution of the left internal carotid artery (ICA), due to involvement of the ophthalmic, middle and anterior cerebral arteries. Another clue on examination is the left carotid bruit. Bruits are due to turbulent blood flow, which can in turn be caused by vessel stenosis. When a vessel completely occludes, the bruit disappears.

**Summary.** Abrupt onset (vascular) of left visual loss (left OA) and right hemiparesis (MCA and ACA = ICA) is suggestive of a stroke in the left internal carotid artery distribution. This could be embolic or thrombotic in etiology.

If the patient presents to an Emergency Department or is a hospital in-patient and is seen within three hours of symptom onset, he/she should be evaluated...
for possible treatment with tissue plasminogen activator.

Table 4-1: Characteristics of upper motor neuron weakness

<table>
<thead>
<tr>
<th>Distribution</th>
<th>Lower face, Extensor muscles in the arm Flexor muscles in the leg</th>
</tr>
</thead>
<tbody>
<tr>
<td>Muscle Tone&lt;</td>
<td>Spasticity, Increased resistance overcoming flexion in the arm and overcoming extension in the leg (Like opening a clasped knife)</td>
</tr>
<tr>
<td>Reflexes</td>
<td>Deep tendon reflexes are increased and clonus may be elicited</td>
</tr>
<tr>
<td>Pathological Reflexes</td>
<td>Clonus at the patella or ankle Babinski sign in the lower extremity</td>
</tr>
<tr>
<td>Atrophy</td>
<td>Does not occur with UMN weakness Mild disuse atrophy may be seen</td>
</tr>
<tr>
<td>Coordination</td>
<td>There is slowness or incoordination of fine motor movements, e.g., tapping fingers or wiggling toes</td>
</tr>
</tbody>
</table>

Case 2

A 33-year-old male construction worker has been in good health except for a recent bout of gastroenteritis. Initially he noted some paresthesias in his feet and less so in his fingers. The following day he noted difficulty climbing a ladder. He presented to the Emergency Department and was noted to have hypoactive deep tendon reflexes, no sensory loss and was able to walk unassisted. A diagnosis of “flu” was given and he was given symptomatic treatment. He spent the remainder of the day resting in bed. The following morning he fell while attempting to walk to his bathroom and at that time, was unable to stand without support. Re-evaluation, at the Emergency Department, revealed distal leg weakness and absent deep tendon reflexes. There was no objective sensory deficit.

Other studies included normal EKG, CPK and troponin and normal chest x-ray. Neurology consultation was obtained. Nerve conduction/EMG studies revealed prolonged distal motor latencies and slowed nerve conduction velocities. CSF was obtained and showed a protein level of 80 mg/dl with no cells and normal glucose. Treatment was started.

The following day the patient noted difficulty grasping and elevating his arms but then stabilized. After one week of treatment strength started to return in the upper, then lower extremities.

Discussion. This individual had a subacute process causing progressive weakness that started distally and progressed proximally over days, ultimately affecting the patient’s ability to walk. Over time, the deep tendon reflexes disappeared, suggesting a lower motor neuron or peripheral nerve lesion. Associated findings are elevated CSF protein with a normal cell count (albuminocytologic dissociation) and slowed motor nerve conduction velocities (demyelination). The disorder that is most likely to produce this profile is acute inflammatory demyelinating polyradiculoneuropathy (AIDP) also known as Guillain-Barré syndrome.

Current treatment involves alternate day plasmapheresis or daily administration of intravenous immune globulin (IVIG). These treatments have significantly improved the outcome of patients with this immune mediated disorder.

Failure to provide timely treatment could lead to progressive weakness, and respiratory failure, with need for assisted ventilation. Prolonged disease duration
could also produce axonal nerve damage with subsequent prolonged recovery times, muscle atrophy, contractures and motor disability. With severe untreated disease autonomic involvement could lead to hypo- or hypertension and life threatening cardiac arrhythmias.

Table 4-2: Characteristics of lower motor neuron weakness

<table>
<thead>
<tr>
<th>Distribution</th>
<th>Peripheral neuropathy, distal and symmetrical; or, follows the distribution of root, plexus or peripheral nerve</th>
</tr>
</thead>
<tbody>
<tr>
<td>Muscle Tone</td>
<td>Decreased in the affected muscles</td>
</tr>
<tr>
<td>Reflexes</td>
<td>Decreased or absent</td>
</tr>
<tr>
<td>Atrophy</td>
<td>Present in the affected muscles</td>
</tr>
</tbody>
</table>
| Fasciculations may be noted | Does not occur with UMN weakness                          
                                       Mild disuse atrophy may be seen                              |
| Coordination          | There is motor ataxia proportional to the degree of weakness                                                  |

There is current evidence that peripheral neuropathies whether generalized or focal may have an immune basis if no other etiology is found. If certain criteria are met these neuropathies may be classified as a form of chronic inflammatory demyelinating polyradiculoneuropathy and respond to treatment with IVIG or plasmapheresis. Chronic progressive or recurrent neuropathies without an apparent etiology should be referred for detailed neurological investigation.

Case 3

A 68-year-old male was diagnosed with prostate cancer two years earlier. His biopsy showed an undifferentiated carcinoma and he was treated with surgical excision and local radiation therapy. Several months later chemotherapy was started because of rising prostatic alkaline phosphatase and prostate specific antigen (PSA) levels. Three months later he noted increasing leg fatigue when he went for his daily walk. Two weeks later he fell while walking his dog but sustained no injuries. There were several subsequent falls followed by increasing complaints of low thoracic mid back pain. Finally one fall caused a right proximal humerus fracture and he was admitted to the hospital. Initial evaluation revealed mild to moderate proximal bilateral leg weakness and he was kept on bed rest. Spine x-rays revealed anterior compression fractures at several levels in the upper lumbar, lower and mid thoracic spine. A technetium bone scan showed uptake in the spine corresponding to the fracture sites and other areas.

Difficulty initiating urination and occasional small volume incontinence prompted insertion of an indwelling Foley catheter. On insertion the initial volume drained was 800 cc.

Physical therapy was ordered but discontinued on day 2 because of lack of cooperation. Oncology consultation was requested.

On day 4 after admission, nursing service informed the admitting physician that the patient could no longer move his legs to cooperate with bathing. Repeat examination disclosed that the patient had flaccid paraplegia of both lower extremities. Neurological consultation confirmed flaccid lower extremity paraplegia, a sensory level at T-10 and diminished rectal sphincter tone. Beevor’s sign, was present (upward motion of the umbilicus with neck flexion while lying supine, caused by weakness of abdominal muscles below T-10).

An emergency MRI scan showed some lysis of the T-10 vertebral body with a
posterior tumor mass compressing the conus medullaris and distal spinal cord. Emergency radiation therapy to the area was started. Six months later the patient had some return of antigravity leg movement, but was unable to ambulate independently.

Discussion. The above case illustrates how spinal cord lesions can progress, subacutely, to full paraplegia even when the patient is in a setting where he/she is observed. Once the patient is on bed rest, progressive leg weakness is harder to detect, by medical staff or the patient.

Numerous early warning signs could have alerted the physician to the presence of an early myelopathy in this patient:

- Multiple spine compression fractures; and an abnormal bone scan with a known malignancy (prostate carcinoma), that is known for bone metastasis.
- Increased frequency of falls most likely due to progressive leg weakness.
- Development of spine pain as part of the current symptomatology, secondary to metastases and compression fractures.
- Overflow urinary incontinence, with a large residual volume on catheterization, due to evolving weakness of the bladder detrusor muscles secondary to sacral outflow interruption.
- Daily neurological examination would to have led to earlier detection of evolving leg weakness, a rising sensory level, and the development of lower abdominal wall weakness (Beevor’s sign). The weakness would have had the characteristic upper motor neuron distribution with extensor muscle groups being stronger than the corresponding flexors. (Gluteus maximus stronger than iliopsoas, quadriceps stronger than hamstrings and gastrocnemius stronger than anterior tibial, posterior tibial stronger than peronei. It is this distribution of weakness that produces the classic leg posture in upper motor neuron weakness; leg extended at the hip and knee with the foot pointed down and inverted).

The importance of this case lies in the fact that, once paraplegia develops, and lasts for up to 24 hours, surgical or radiation intervention may not be as beneficial in terms of restoration of normal neurological function. Any spinal cord lesion, once suspected should be monitored closely for progression, while the work-up proceeds to determine the cause, and thus the treatment of the pathological process.

This particular patient had spinal metastases that produced spinal cord compression with clinical signs that evolved in a subacute fashion. Spinal cord metastases frequently involve the T-10 region because a main arterial feeder; the artery of Adamkiewicz, arises around the L1 level and supplies the distal spinal cord. The practitioner must beware of ordering just a “lumbar” MRI scan or “lumbar” myelogram on a patient with evolving leg weakness. This study, as ordered, will visualize the spine up to T12-L1 and potentially miss lower thoracic spine lesions.

A key clinical pearl is always to suspect an evolving spinal cord lesion in a patient with spine pain, progressive leg weakness, and urinary incontinence or retention. The presence of the above clinical findings in a known cancer
patient should be particularly alarming, especially primary cancer of the prostate, breast, colon, and kidney. Another malignancy that commonly causes spine involvement is multiple myeloma.

Early recognition can lead to prompt treatment, which will prolong survival and hopefully preserve the ability to walk and maintain bladder control.

<table>
<thead>
<tr>
<th>Intrinsic</th>
<th>Extrinsic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weakness</td>
<td>Upper motor neuron; below level of cord involvement.</td>
</tr>
<tr>
<td>Sensory</td>
<td>Sensory dissociation (Some pathways severely affected, others are normal)</td>
</tr>
<tr>
<td>Pain</td>
<td>Usually none</td>
</tr>
<tr>
<td>Root lesion</td>
<td>No</td>
</tr>
<tr>
<td>Reflexes</td>
<td>Brisk below level of lesion</td>
</tr>
<tr>
<td>Example</td>
<td>Multiple Sclerosis</td>
</tr>
</tbody>
</table>

**Case 4**

A 23-year-old female, first noted difficulty exercising on the stair-climber at her local health spa. She had been in top physical condition, engaging in daily aerobic workouts for the past 18 months. Her first symptoms were a feeling of heaviness in her legs after stair climbing for about 15 minutes. As she continued, the weakness would worsen. She sought the advice of her personal trainer who thought she should vary her routine and exercise her upper body more. She changed her routine so that once she tired on the stair climber she would switch to the rowing machine. This seemed to solve the problem for the next several weeks.

Over the ensuing weeks she noted increasing fatigue and actual weakness as she continued her workouts. Variation in her program and new routines seemed to give very transient relief. She stopped her exercise program on the advice of a friend and noted that she symptomatically improved. Things went along well for another few weeks as she substituted reading for exercise in her leisure hours. A visit to her physician was finally prompted by some new episodes consisting of diplopia and ptosis after about an hour of reading. She noted improvement when she rested, only to have the diplopia recur when she began reading again.

Examination by her primary care physician noted her to be in good health otherwise. Her peripheral strength appeared normal. Her reflexes were on the brisk side but normal and there were no sensory deficits. Coordination and cranial nerve examination was normal.

Laboratory studies revealed normal CBC, chemistry profile, sedimentation rate, ANA, thyroid function studies, CPK, urinalysis and RPR. Chest X-ray, and EKG. Pulmonary function studies were also normal.

She was referred for neurological consultation. Physical findings confirmed that the patient demonstrated weakness with sustained muscle activity. If she held her gaze upward, after two minutes some ptosis was noted in the right and then left eye. With sustained upward gaze she reported vertical diplopia. In a similar fashion she had difficulty doing deep knee bends after 8 repetitions and
then could not stand upright without assistance. After a brief period of rest the weakness resolved, only to recur with repeated exercise.

Exercise was then repeated to demonstrate observable ptosis, dysconjugate gaze and inability to stand up from a squatting position. At this point the patient was given 5 mg of edrophonium chloride (Tensilon®) intravenously, after a 1 mg test dose. This led to immediate resolution of weakness for several minutes.

A repetitive nerve stimulation test demonstrated a neuromuscular transmission defect and a blood test revealed a high titer of antibodies directed against acetylcholine receptor antigen. (ACh receptor Ag).

By now you have surmised that the patient has myasthenia gravis (MG), an autoimmune disease. The hallmark of this disease process is the development of muscle weakness with exercise. Patients are usually stronger in the mornings and after rest, and weaker after exercise and later in the day. The distribution of weakness can be ocular (ptosis and diplopia), bulbar (dysarthria, dysphagia), or generalized, (extremity weakness and difficulty breathing).

The diagnosis depends on demonstrating a defect in neuromuscular transmission by:

- Specialized nerve conduction studies such as repetitive stimulation and single fiber EMG.
- Transiently overcoming the neuromuscular blockage using edrophonium chloride, which is an acetylcholinesterase inhibitor.
- Demonstrating antibody directed against the ACh receptor antigen.
- Treatment of MG consists of two major components:
  - **Symptomatic:** Varying doses of pyridostigmine (Mestinon®), at 4-6 hour intervals provide improvement in muscle fatigability. This drug is a long acting acetylcholinesterase inhibitor.
  - **Immunosuppression:**
    - Prednisone initiated in small alternate day doses (12.5 mg) and slowly increased by 12.5 mg every other dose. Full benefit is usually obtained at doses of 60 to 100 mg every other day. Once improvement is achieved the dosage can be gradually reduced to the smallest one that maintains benefit.
    - Azathioprine may be a better agent if long-term use is contemplated due to fewer overall side effects than steroids. However, the potential toxic effect on bone marrow and liver function must be carefully monitored. Long-term use on a young patient may increase the risk of oncogenesis.
    - Plasmapheresis will provide short-term improvement in function by lowering antibody titer. It is a useful modality to prepare myasthenic patients for surgery (thymectomy) or as an adjunctive treatment in myasthenic crisis (situations of disease worsening when patients may be unable to swallow or breathe adequately without assistance).
    - IVIg is a newer form of therapy that may have fewer risks than plasmapheresis, e.g., central venous catheter infection or thrombosis, but is expensive. IVIg is currently achieving wider use in treating patients with MG that is more difficult to control with conventional...
therapy. IVlg has a duration of action that may last several weeks and sometimes several months in patients being treated with other immunosuppressive drugs. It is easier to administer than plasmapheresis especially in patients with poor venous access. Potential risks in adults with other health problems are congestive heart failure, acute renal failure and deep vein thrombosis.

• Thymectomy may be a curative procedure or result in clinical improvement with reduction of medication requirements. The thymus gland appears to have a major role in promulgating this disease process and removal may lead to disease remission in a significant number of patients. The patient should be at maximal functional capacity prior to surgery. This may require pretreatment with steroids and plasmapheresis. The thymus should be removed with a sternal splitting procedure and not through mediastinoscopy. Any remaining thymus tissue may interfere with attainment of remission and mediastinoscope procedures may miss rests of thymic tissues located deep in the mediastinal gutters.

Finally, the thymus gland should be evaluated in all patients diagnosed with myasthenia gravis. Typically, it is hypertrophied. However, about 15 percent of myasthenic patients have a malignant thymoma, which can be locally invasive and requires surgical as well as radiation therapy. MG in this setting is more difficult to treat and these myasthenics appear to be more resistant to conventional therapy.

Table 4-4. Characteristics of weakness in myasthenia gravis

<table>
<thead>
<tr>
<th>Distribution</th>
<th>Cranial nerves: diplopia, dysphagia, dysarthria Proximal or distal, usually symmetric</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reflexes</td>
<td>Normal or increased</td>
</tr>
<tr>
<td>Muscle tone</td>
<td>Normal</td>
</tr>
<tr>
<td>Coordination</td>
<td>Affected in proportion to degree and distribution of weakness</td>
</tr>
<tr>
<td>Unique features</td>
<td>Weakness increases with exercise and improves with rest</td>
</tr>
</tbody>
</table>

Case 5
A 28-year-old construction worker was injured on the job, twisting his back to the right. He initially complained of soreness but was able to continue working the remainder of the day if he avoided lifting. The following morning he noted increased low back pain, which radiated down the posterior aspect of his left leg. There was also paresthesia in the left first toe.

He reported the injury and was sent to the company physician who ordered plain LS spine X-rays. These showed some loss of the L4-5 disc height but were otherwise normal. Light duty, NSAIDs and muscle relaxants were prescribed, but to no avail. His symptoms were particularly bad at night, although sleeping in a reclining chair with his knees and back flexed helped.

He remained off work and attended physical therapy daily for the next three weeks. He noted no further improvement and towards the end of the three-week period noted he had to purposefully lift his left leg higher to prevent the toes from scraping the ground when he walked.

Neurologic examination at this time, revealed pain with straight leg raising at 50
degrees on the left and 80 degrees on the right. He was unable to heel walk on the left and there was specific weakness of the left anterior tibial, posterior tibial, peronei and toe extensor muscles. The left internal hamstring reflex was reduced and there was vague subjective sensory loss over the dorsum of the left foot.

This history and associated clinical findings strongly suggest the subacute development of a left L-5 radiculopathy. In this setting the most likely diagnosis is a herniated intervertebral disc, which could be demonstrated on MRI. Had the onset been acute, in a known diabetic with a negative MRI scan, consideration to nerve root infarction should be given. Chronic onset of similar symptoms in a patient with multiple myeloma or prostate cancer should make one investigate metastatic disease as a likely possibility.

He reported back pain radiating down the posterior leg, now with paresthesias on the sole of the foot, weakness with toe walking, and a diminished Achilles tendon reflex, makes S1 radiculopathy the underlying cause. Pain in the neck, which radiates to an upper extremity, is often due to cervical radiculopathy. The most common roots affected are C7 and C6. C7 root lesions are associated with pain, paresthesias in the middle fingers, usually weakness of the triceps, pronator teres and finger extensors. There is usually a diminished triceps reflex. C6 weakness involves the biceps, deltoid, brachioradialis and pronator teres, with associated paresthesias in the index finger and a diminished biceps and brachioradialis reflex.

If weakness is present with any radiculopathy, an imaging study should be done to identify the underlying cause. With cervical spondylosis or herniated disc, immobilization with a soft cervical collar for one to two weeks may be curative.

With lumbar radiculopathy, associated with weakness, conservative measures may be less successful and surgery may be indicated. Needless to say, if the patient has weakness, he should be monitored closely for progression while undergoing conservative management. If weakness is progressive during therapy, and a surgical lesion is responsible, neurosurgical consultation should be obtained.

Table 4-5. Characteristics of nerve root weakness

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Distribution</th>
<th>Muscle Tone</th>
<th>Reflexes</th>
<th>Atrophy</th>
<th>Coordination</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain in a radicular distribution</td>
<td>Cervical: upper extremity</td>
<td>Decreased in the affected muscles</td>
<td>Decreased or absent in the affected dermatome or myotome</td>
<td>Present in the affected muscles</td>
<td>Usually unaffected</td>
</tr>
<tr>
<td>Paresthesias in the sensory distribution on the affected nerve root</td>
<td>Thoracic: chest or abdominal wall</td>
<td></td>
<td></td>
<td>Fasciculations may be noted</td>
<td></td>
</tr>
<tr>
<td>Aggravated by activities that lengthen the involved nerve root, e.g., straight leg raising</td>
<td>Lumbar: lower extremity</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Weakness in the distribution of the involved nerve root</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Case 6

A 45-year-old male had been working late at home doing accounting deskwork. He had been to a social affair earlier and had consumed some alcohol. To unwind he had another drink while working. He next recalls awakening in a
leaned back position in his rigid-back wooden chair. He noted an aching pain in his right triceps area. He cleared his head and then attempted pick up his pen. He noted he could not open his right hand or elevate his wrist and the dorsal surface of the hand was tingling. He assumed he was having a stroke and had his wife take him to the nearest Emergency Room. Examination there revealed right fingers and wrist extensor weakness and a diminished right brachioradialis reflex. There was vague subjective sensory loss on the dorsal hand, but no other findings. A CT scan of the head was obtained which was normal. Routine laboratory studies were normal and the patient was normotensive. There were no other stroke risk factors, and a chest x-ray and EKG were normal. The local neurologist was consulted to determine if the patient was a candidate for treatment with tissue plasminogen activator (tPA).

The neurologist evaluated the patient and decided the weakness was fairly acute in onset and had occurred during sleep. The weakness, as previously described, involved wrist and finger extensors, with dorsal hand sensory loss and a diminished brachioradialis reflex. He concluded that the problem was an acute radial neuropathy, caused by prolonged pressure of the nerve at a vulnerable point (the radial groove of the humerus), while the patient slept. He told the patient to wear a neutral wrist splint to prevent a flexion contracture and to follow up in the ambulatory clinic. If the weakness persisted beyond three weeks, nerve conduction and EMG studies were planned.

This case illustrates that some cases of acute onset weakness can have etiologies other than stroke. The history and examination help establish the correct diagnosis. Inappropriate administration of tPA, in the above case, may have led to unnecessary complications. Fortunately, the patient would have been excluded from tPA treatment since the exact time of onset of weakness was not known.

Other peripheral nerve syndromes that may have acute or subacute onset include:

- **Peroneal neuropathy or “crossed leg palsy”:** In this condition there is inability to dorsiflex the foot and the person lifts the involved leg higher to enable the foot to clear the ground. Weakness is noted in the anterior tibial, peronei and toe extensor muscles. The posterior tibial muscle is spared and the deep tendon reflexes are normal. Usually the superficial peroneal nerve is spared and only mild sensory loss is noted in the webspace between the first and second toe. The condition is usually caused by pressure on the peroneal nerve as it goes across the fibular head. This is most frequently caused by crossing of the legs while sitting and is also called “crossed leg palsy.” Treatment is discontinuation of leg crossing or other activity that puts pressure on the peroneal nerve. Recovery develops over a few weeks if the lesion is only demyelinating.

- **Ulnar neuropathy:** This is one of the most common compression neuropathies. Normally the ulnar nerve is protected at the elbow because it lies in the ulnar groove. The olecranon usually rests on a hard surface when the elbow is leaned on, thereby protecting the ulnar nerve. As one ages, the ulnar groove may become shallower, which exposes the ulnar nerve. Common activities involving leaning on the elbows, may now compress the ulnar nerve leading to the following symptoms:
  - Numbness and sensory loss involving the fifth and lateral half of the
fourth finger and medial portion of the palm on the affected hand.

- Weakness of the interossei and fourth and fifth lumbrical muscles (claw hand deformity).
- Atrophy of the involved muscles.
- Tinel’s sign may be present at the ulnar groove.

See Table 4-2 for characteristics of lower motor neuron weakness.

Case 7

A 46-year-old female stock clerk noted increasing pain in her shoulders and thighs at the end of the workday. She was somewhat better in the mornings but worsened at work. Her workload had not changed and she had no work related injuries. She also noted difficulty elevating stock boxes to shelves above shoulder level and some difficulty stepping up onto high platforms. The pain and weakness slowly worsened and within six months she had difficulty keeping her arms up when setting her hair, climbing stairs at home, and getting out of the bathtub.

She sought medical attention and was initially diagnosed as having myalgia and musculoskeletal pain related to her job. She was treated with muscle relaxants and NSAIDs with only minimal relief of symptoms.

A second visit to her physician revealed some difficulty getting out of a chair. Laboratory values showed mildly elevated transaminases and a CPK of 12,000 U/L, which fractionated to predominantly MM bands. Neurological consultation was requested.

On neurologic examination she was found to have symmetric weakness of the neck flexors, shoulder and hip girdle muscles. She had difficulty getting out of a low chair and in attempting to get up from a prone position, she had to extend her trunk by pushing upward with her hands on her thighs or using furniture to pull her trunk erect with her arms (Gower’s Sign).

This type of weakness is illustrative of myopathy, which may have many causes. A common acquired form is polymyositis, an autoimmune inflammatory disorder. Metabolic myopathies can be due to hypothyroidism or Cushing’s disease among others. Congenital dystrophies can also present with proximal muscle weakness.

Once suspected, diagnosis is confirmed with NC/EMG studies and muscle biopsy. A muscle biopsy should only be performed and interpreted by someone with sufficient training and experience. A routine muscle biopsy, i.e., a sample of any muscle is placed in formalin and then sent to the pathology laboratory, is not sufficient. Formalin actually distorts muscle architecture, so the specimen must be frozen in liquid nitrogen before being processed. Normal values have been ascertained for only a few muscles such as the biceps and quadriceps femoris. This information should be obtained from the myologist before a muscle is selected for biopsy. Special staining and architecture measurement techniques performed by the myologist give useful information about the etiology of the myopathy.

Accurate diagnosis depends on family history, laboratory evaluation, muscle biopsy and sometimes, genetic studies. Consultation with a specialist familiar with these disorders is frequently required. Treatment is directed to the
underlying etiology.

Table 4-6. Characteristics of myopathic weakness

<table>
<thead>
<tr>
<th>Distribution</th>
<th>Proximal: involves neck flexors, shoulder and hip girdle muscles</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptoms</td>
<td>Problems climbing stairs, stepping up onto high steps, getting out of low places such as deep chairs, and holding arms over head for prolonged periods. Pain, tenderness, and sometimes swelling of involved muscles.</td>
</tr>
<tr>
<td>Muscle Tone</td>
<td>Normal or diminished. Severe weakness and contractures may develop.</td>
</tr>
<tr>
<td>Reflexes</td>
<td>Normal or diminished in proportion to weakness.</td>
</tr>
<tr>
<td>Atrophy</td>
<td>Present with moderate to severe involvement.</td>
</tr>
</tbody>
</table>

Summary. The preceding cases represent some of the more common presentations of weakness including some that require urgent treatment intervention. When evaluating weakness, important considerations are the temporal profile, distribution of weakness, historical progression of the weakness, and associated symptoms such as pain and sensory changes. A thorough clinical examination to determine whether the weakness involves the upper or lower motor neuron, the neuromuscular junction or muscle itself, is mandatory. Once the above features are categorized, one can then establish a differential diagnosis and proceed with a proper workup designed to establish the correct clinical diagnosis. The appropriate treatment can then be chosen.

Psychological Impact

Although weakness may be a sign of a serious neurological condition, many patients with no neurological disorder present with a complaint of weakness. As noted in the introduction to this chapter, “weakness” doesn’t always mean a true loss of motor function. Patients who are depressed often complain of weakness. Hypothyroidism may present this way, as can patients with anemia, coronary artery disease, and numerous other non-neurological conditions. Thus, it is critical that the physician considers not only the patient’s description of the problem and the physical examination, but also the context in which the patient presents.

Patients have a great fear of loss of neurological function. The resulting loss of control, independence, and dignity is often more frightening to patients than cancer or myocardial infarction. It is critical that the physician be willing and able to address these fears. Many of the conditions discussed in the chapter will lead to permanent and even progressive disability. The physician must strike a balance between truthfully sharing information and taking away all hope. In most cases, some positive information can be provided. For example, it may be comforting for a stroke patient to know that many similar patients do recover some or all of their function over time. Remember, also, that the shock of a new diagnosis may make it difficult for the patient and family to fully absorb your information. Be prepared to repeat and elaborate it over time.

Community Resources

Ongoing needs will vary tremendously from patient to patient. For example, the patient with a large CVA may require skilled care in an extended care facility, while patients with small strokes may recover complete function. Most patients, though, benefit from various community resources. Home health services are
a wonderful way to allow patients to remain in their home, at the same time receiving needed therapy. Physical therapy, speech therapy, and occupational therapy can all be given in the home, as well as in acute and extended care facilities. Family members can often be taught to provide these services. A growing array of supportive devices exists for helping patients compensate for their disabilities. Some “low tech” examples include eating utensils that can be strapped to the hand, splints, bathroom rails and elevated toilet seats. Sophisticated technology also comes into play, including motorized wheelchairs and scooters, seat lifters, symbol keyboards, text-to-speech systems, and speech recognition devices.

National organizations for specific conditions offer a number of services, including literature directed to the lay audience, lists of local support groups, and treatment advances.

**National Stroke Association**
96 Inverness Drive East, Suite I
Englewood, Colorado 80112-5112
800-787-6537
Website:
Review article “Evaluation of the patient with muscle weakness” in *American Family Physician*

**Myasthenia Gravis**

**Patient Support Group**

**Guillain-Barré Syndrome**

**Patient Support Group**

**Peripheral Neuropathy**

**Spondylotic myelopathy**

**Peripheral nerve injuries**

**Cervical radiculopathy**

**Lumbar radiculopathy**

**American Heart Association**
National Center
7272 Greenville Avenue
Dallas, Texas 75231
800-242-8721

**The ALS Association National Office**
27001 Agoura Road, Suite 150
Calabasas Hills, California 91301-5104
800-782-4747
Email: alsinfo@alsa-national.org

**Myasthenia Gravis Foundation of America**
123 W. Madison Street, Suite 800
Chicago, IL 60602
800-541-5454
E-Mail: myasthenia@myasthenia.org

**The Neuropathy Association**
Self-Assessment Questions

1. All of the following clinical findings are important to determine the etiology of muscle weakness EXCEPT:
   A. distribution of weakness
   B. presence of atrophy
   C. muscle tone
   D. deep tendon reflexes
   E. absence of muscle tenderness

2. A feature of upper motor neuron weakness is:
   A. muscle atrophy
   B. fasciculations
   C. fibrillations
   D. diminished muscle tone
   E. increased deep tendon reflexes

3. In weakness due to motor peripheral neuropathy, all of the following are true EXCEPT:
   A. the weakness is symmetric
   B. the weakness is distal.
   C. the weakness is proximal
   D. the weakness is usually greater in the lower extremities
   E. there may be associated muscle atrophy

4. Upper motor neuron weakness from a cortical lesion has the following distribution:
   A. upper, flexors; lower, flexors
   B. upper, extensors; lower, flexors
   C. >upper, flexors; lower, extensors
   D. upper, extensors; lower, extensors

5. Progressive weakness in the lower extremities associated with back pain and urinary retention can be seen with:
   A. myelopathy secondary to a herniated thoracic disc.
   B. myasthenia gravis
   C. acute stroke
   D. inflammatory myopathy.
E. all of the above

6. Acute or subacute onset of cervical pain with radiation to an upper extremity may be caused by:
   A. herniated cervical disc
   B. spondylosis of the cervical spine
   C. Herpes zoster
   D. nerve root infarction secondary to diabetes
   E. all of the above

7. Clinical findings associated with inflammatory myopathy include all EXCEPT:
   A. muscle pain
   B. elevated CPK level
   C. proximal symmetric muscle weakness
   D. Babinski’s sign
   E. dysphagia

8. Abnormal laboratory findings in myasthenia gravis include:
   A. elevated CPK level
   B. elevated transaminase levels.
   C. reduced acetylcholine antibody titer
   D. elevated smooth muscle antibody titer.
   E. none of the above

9. Characteristics of radiculopathy include:
   A. pain, weakness, diminished deep tendon reflex
   B. numbness, pain and increased muscle tone
   C. weakness, atrophy and increased deep tendon reflex
   D. spasticity, atrophy and diminished deep tendon reflex
   E. none of the above

10. Progressive bilateral, distal muscle weakness, distal paresthesias, and loss of deep tendon reflexes over one to two weeks is suggestive of:
    A. myasthenia gravis
    B. stroke in evolution
    C. polymyositis
    D. Guillain-Barré syndrome
    E. diabetic peripheral neuropathy
11. Proximal and bulbar muscle weakness that worsens with exercise and improves with rest may be associated with:
   A. lung cancer
   B. AIDS
   C. thymoma
   D. sepsis
   E. liver disease

12. All of the following may be seen with ulnar neuropathy  EXCEPT:
   A. numbness of the fourth and fifth fingers
   B. hypothenar atrophy
   C. interosseous atrophy
   D. thenar atrophy
   E. Tinel’s sign at the ulnar groove (elbow).

13. CPK elevations are most commonly associated with:
   A. stroke
   B. myelopathy
   C. inflammatory myopathy
   D. Guillain-Barré syndrome
   E. myasthenia gravis

14. Amaurosis fugax may be a premonitory symptom of:
   A. disc herniation
   B. respiratory arrest
   C. facial weakness
   D. stroke
   E. none of the above

**Answers:**
1. E
2. E
3. C
4. B
5. A
6. E
7. D
8. E
9. A
10. D
11. C
12. D
13. C
14. D