This tool addresses common symptoms and symptom complexes. Imaging requests for patients with atypical symptoms or clinical presentations that are not specifically addressed will require physician review. Consultation with the referring physician, specialist and/or patient’s Primary Care Physician (PCP) may provide additional insight.

PERIPHERAL NERVE DISORDERS (PND)
IMAGING GUIDELINES
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MedSolutions, Inc. Clinical Decision Support Tool
for Advanced Diagnostic Imaging

Common symptoms and symptom complexes are addressed by this tool. Imaging requests for patients with atypical symptoms or clinical presentations that are not specifically addressed will require physician review. Consultation with the referring physician may provide additional insight.

This version incorporates MSI accepted revisions prior to 7/22/11
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<td>AIDS</td>
<td>Acquired Immunodeficiency Syndrome</td>
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PERIPHERAL NERVE DISORDERS (PND) IMAGING GUIDELINES

PN-1~GENERAL GUIDELINES

- The initial evaluation of a suspected peripheral nerve disorder should include a detailed neurological history and examination followed by electromyography and nerve conduction (EMG/NCV) studies.
  - EMG may not be helpful in the immediate time period when new signs or symptoms of a peripheral nerve disorder first appear. Therefore, for the acute presentation of a possible peripheral nerve disorder, EMG should not be performed until the problem is at least three weeks old.
- Advanced imaging plays a limited role in the diagnosis and management of disorders of peripheral nerves and muscles.
  - NOTE: many disorders of these structures are associated with systemic diseases in which there are well-established indications for advanced imaging.
- When imaging of peripheral nervous tissue or muscles is indicated, MRI is used. In general, CT is not an acceptable alternative (occasional exceptions will be mentioned below).
- MRI is sometimes useful as a preoperative procedure since surgical decisions often depend on the presence or absence of anatomic integrity of the nerves (EMG tests functional integrity).
- Reference:

PN-2~FOCAL NEUROPATHY

- PN-2.1 Carpal Tunnel Syndrome
  - Also see MS-22 Wrist in the Musculoskeletal Imaging Guidelines
  - Also see SP-11.5 Carpal Tunnel Syndrome under Cervical Radiculopathy in the Spine Imaging Guidelines.
  - Common clinical syndrome causing intermittent hand numbness usually worse at night and with some aching.
  - Diagnosis is by clinical evaluation and electro-diagnostic studies (EMG/NCV).
  - Noncontrast wrist MRI (CPT®73221) reveals median nerve compression fairly well, but is less sensitive than nerve conduction measurements.
  - At this time, advanced imaging has no established role in the evaluation of carpal tunnel syndrome.
  - References:
    - Neurology 2002;58:1583-1584
      Accessed November 22, 2006
    - Neurology 2002;58:1597-1602
• **PN-2.2 Ulnar Neuropathy**
  o The diagnosis of ulnar neuropathy should be made based upon symptoms, clinical examination, and NCV/EMG results.
  o Advanced imaging is not generally indicated.

• **PN-2.3 Radial Neuropathy**
  o An infrequent upper extremity neuropathy causing wrist drop. Common sites of entrapment include the inferior aspect of the humerus (Saturday night palsy) or the forearm (Posterior Interosseus Syndrome).
  o Trauma or fractures of the humerus, radius, or ulna can damage the radial nerve.
  o NCV/EMG should be performed initially.
  o Noncontrast MRI of the upper arm or forearm (CPT®73218) is indicated only in severe cases where surgery is considered.
  o **Reference:**
    > *Radiographics* 2006;26:1267-1287

• **PN-2.4 Sciatic Neuropathy**
  o Although the term sciatica is common, at least 98% of cases are due to lumbar radiculopathy.
  o Rarely, trauma to the gluteal area with hematoma, injection palsy, hip or pelvic fractures, or hip replacement (arthroplasty) can damage the proximal sciatic nerve.
  o A controversial disorder called Piriformis Syndrome involves entrapment of the sciatic nerve at the sciatic notch in the pelvis by a tight piriformis muscle band.
  o EMG/NCV should be performed initially to localize the problem.
  o Evaluation by a Neurology, Orthopedic, Physiatry (PM&R), or Pain Management specialist is helpful in determining the need for advanced imaging.
    > Rarely, CT pelvis without contrast (CPT®72192) or MRI pelvis without contrast (CPT®72195) may be performed in severe cases to evaluate sciatic neuropathy.
  o **Reference:**
    > *Neurologic Clinics* 1999 August;17(3):617-631

• **PN-2.5 Femoral Neuropathy**
  o Can arise as a complication of pelvic surgery in women or in patients on anticoagulants as a complication of retroperitoneal bleeding.
  o Pelvic CT can be performed either with (CPT®72193) or without (CPT®72192) contrast for evaluation in either setting.

• **PN-2.6 Meralgia Paresthetica**
  o A common sensory neuropathy involving the lateral femoral cutaneous nerve as it exits the pelvis under the inguinal ligament.
  o Patients have objective sensory loss in the region supplied by the nerve (lateral thigh without extension into lower leg).
  o Spinal imaging is not indicated.
  o In cases recalcitrant to medical management, studies to exclude a pelvic mass may be appropriate (see Pelvis Imaging Guidelines).
In women, pelvic ultrasound is recommended initially.
In men (and in women with nondiagnostic ultrasound), either pelvic CT with contrast (CPT® 72193) or pelvic MRI without contrast (CPT® 72195) is acceptable.
Abdominal imaging may be useful if the clinical picture suggests involvement in the upper lumbar plexus.
If imaging is being done as a preoperative study to evaluate for decompression of the nerve, MRI is preferred.

PN-2.7 Peroneal Neuropathy
A common neuropathy in the lower leg causing foot drop.
The most common site of entrapment is on the lateral aspect of the knee as the nerve wraps around the neck of the fibula.
Peroneal neuropathy usually resolves over time with no specific treatment.
Neurology, Orthopedic, or Pain Management consultation is helpful in distinguishing this disorder from an L5 radiculopathy causing foot drop.
EMG/NCV should be performed initially.
Rarely, noncontrast knee MRI (CPT® 73721) or noncontrast MRI lower extremity other than joint (CPT® 73718) may be performed in severe cases when surgery is considered.
Reference:
Neurology 2005;65:1829-1831

PN-2.8 Tarsal Tunnel Syndrome
See MS-30.9 Tarsal Tunnel Syndrome in the Musculoskeletal Imaging Guidelines.

PN-2.9 Other Peripheral Mononeuropathies
Advanced imaging is generally not useful in other peripheral mononeuropathies and should be regarded as largely investigational in those settings.
However, following major trauma, MRI (not CT) may have some role as a preoperative study to evaluate an injured peripheral nerve for anatomical integrity.
Only nerves greater than 2 mm in diameter can be visualized.
Note: For indications other than preoperative imaging of traumatized nerves, imaging without and with contrast is preferred, if imaging is indicated.
These cases should be sent for Medical Director review.
PN-3~POLYNEUROPATHY

- MRI (not CT) has very uncommon but distinct usefulness in a variety of peripheral neuropathies. Neurological consultation is helpful to clarify the diagnostic pathway in these unusual settings.
- NCV/EMG should be done initially in the evaluation of polyneuropathy.

**PNS/CNS Crossover Syndromes:**
- Occasional cases of Guillain-Barré syndrome and CIDP (Chronic Inflammatory Demyelinating Polyneuropathy) manifest signs of central nervous system involvement.
- Advanced neuroimaging (MRI without and with contrast) of brain or spinal cord may be appropriate if the clinical findings point to abnormalities in those areas.

**AIDS Related Cytomegaloviral Neuropathy/Radiculopathy:**
- This is a rapidly progressive but treatable disorder which may present with urinary retention and a clinically confusing picture in the legs.
- Lumbar spine MRI without and with contrast (CPT®72158) may be useful in suspected cases to identify swelling and enhancement of lumbar roots.
- **Reference:**
  - *Clin Infect Dis* 1998;27:345-352

**Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)**
- Treatable neuropathy usually diagnosed by clinical features, EMG, and nerve biopsy.
- If the diagnosis remains uncertain following EMG and careful clinical evaluation, lumbar spine MRI without and with contrast (CPT®72158) can be performed.
  - Approximately 70% of CIDP cases show enlarged, enhancing lumbar nerve roots on MRI.
- **Reference:**
  - *Brain* 1999;122:1383-1390

**Multifocal Motor Neuropathy:**
- Treatable neuropathy which can generally be diagnosed based upon the clinical examination and EMG.
- If the diagnosis remains uncertain after full evaluation, MRI of the brachial plexus (CPT®71552) may be useful. This is an uncommon situation in a very uncommon disorder.
- **Reference:**
  - *Muscle & Nerve* 2001;24:311-324

**POEMS** (Polyneuropathy, Organomegaly, Endocrinopathy, M-protein, Skin changes):
- Also see **ONC-26 Multiple Myeloma, Waldenstrom’s Macroglobulinemia, and Plasmacytomas** in the Oncology Imaging Guidelines.
- Advanced imaging is appropriate for the non-neurological entities of this rare osteosclerotic plasmacytoma syndrome.
• **Subacute Sensory Neuronopathy and Other Paraneoplastic Demyelinating Neuropathies:**
  o Also see **ONC-30.3 Paraneoplastic Syndromes** in the Oncology Imaging Guidelines.
  o Subacute sensory neuronopathy is a paraneoplastic syndrome associated most strongly with small cell lung cancer, but at times seen with lymphomas, adenocarcinoma of the lung, or tumors of breast or ovary.
  o Advanced imaging should be performed as indicated for the associated malignancy or to search for malignancy (see Oncology Guidelines).
  o Most of the syndromes have antibodies associated with different groups of tumor type, and evaluation should be guided by such titers when positive. *N Engl J Med* 2003;349:1543-1554
  o PET in occult tumor evaluation:
    ➢ Scattered case reports and small studies suggest that PET may be of value in detecting occult neoplasms in patients with paraneoplastic syndromes in whom other diagnostic studies have failed to diagnose a tumor.
    ➢ Currently, there is insufficient data to support the use of PET to find an occult malignancy in patients with paraneoplastic syndromes.
      ▪ **Exception:** PET can be used to characterize an abnormality seen on other imaging in a location difficult to biopsy.
  
  o Reference:
    ▪ *Ann Neurol* 2000;48:105-108

• Several collagen vascular diseases may present with a progressive polyneuropathy.
  o Systemic lupus, Sjogren’s syndrome, Bechet’s disease, polyarteritis nodosa, Churg-Strauss syndrome, and Wegener’s granulomatosis can all present in this manner. (See **HD-32 Cerebral Vasculitis** in the Head Imaging Guidelines).
  o Imaging studies will be those relevant to the diagnosis and treatment of the underlying disorder or to any central nervous features which may also be identified clinically.

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**PN-4~BRACHIAL PLEXUS**

• Disorders of the brachial plexus can generally be identified and distinguished from lesions in other locations by clinical and EMG examination. If the diagnosis remains unclear, advanced imaging can be useful.
• Advanced imaging can be helpful as a preoperative study to evaluate the anatomy of brachial plexus lesions which should have already been defined by clinical examination.
• MRI is the preferred modality. CT is not often useful and should generally not be used as a substitute for MRI to image the brachial plexus.
  o Brachial plexus studies can be coded either as upper extremity other than joint MRI (CPT®73218) or as chest MRI (CPT®71550).
    ➢ Occasionally, for upper trunk lesions, neck MRI (CPT®70540) may be requested.
Chest MRI will image both brachial plexi and is useful for comparing one plexus with the other.

Rarely, more than one CPT® code may be necessary to adequately image the brachial plexus area of interest.

- MRI studies should be without and with contrast (CPT®73220 or CPT®71552) when tumor is part of the differential diagnosis.
- **Reference:**
  - Radiographics 2000;20:1023-1032
  - ACR Appropriateness Criteria, Plexopathy, 2009
  - Eur Radiol 2001;11:325-336

The principal brachial plexus disorders include: malignant infiltration, radiation plexitis, Parsonage-Turner syndrome (so-called brachial plexitis), trauma, birth trauma, and the "neurogenic thoracic outlet syndrome."

**Malignant Infiltration:**
- Most often involves the lower plexus and may include Horner’s syndrome.
- Most cases arise in patients with lung or breast cancer.
- Pain is an early and very prominent symptom. Sensory loss and weakness follow.
- EMG will aid in localization, but will not reveal etiology.
- MRI without and with contrast (CPT®73220 or CPT®71552) is appropriate to aid in the differential diagnosis and localization of the tumor mass, although in cases with perineural spread, MRI may fail to show a mass.
- **Reference:**
  - Neurology 1981;31:45-50

**Radiation Plexitis:**
- Occurs several months to 1-2 years following radiation therapy.
- The upper plexus is usually most involved and pain is infrequent.
- EMG may show changes specific to radiation plexitis.
- MRI without and with contrast (CPT®73220 or CPT®71552) is often done for reassurance that there is no malignant infiltration, especially in the infrequent painful cases.
- An acute form of transient plexitis resembling the Parsonage-Turner syndrome (see Brachial Plexitis bullet below) can occur during radiation treatment. It clears with time even if radiation treatment is continued. MRI without and with contrast (CPT®73220 or CPT®71552) can be performed.
- **References:**
  - Neurology 1989;39:502-506
  - Radiology 2000;214:837-842

**“Brachial Plexitis”** (Parsonage-Turner Syndrome or painful brachial amyotrophy).
- A benign and largely self-limited syndrome characterized by initial shoulder region pain followed by weakness of specific muscles in a pattern which does not conform to involvement of a single root or distal peripheral nerve.
- Careful clinical examination should distinguish brachial plexitis from radiculopathy (see SP-11 Cervical Radiculopathy in the Spine Imaging Guidelines) and its temporal profile does not resemble that of radiation plexitis or malignant infiltration.
The value of imaging is very limited in these cases.
- MRI of the plexus is generally normal and MRI should be performed only in clinically confusing cases.
- MRI of the cervical spine is often requested in these cases. However, unless the clinical picture truly resembles radiculopathy, the results often lead to misdiagnosis, since asymptomatic findings are commonly seen on the cervical spine MRI scans of normal individuals.
- MRI of overtly weak muscles may show increased T2 signal, but this adds no important information.

Reference:

**Trauma:** the cause and extent are generally obvious, but noncontrast MRI of the brachial plexus (CPT®73218 or CPT®71550) is often useful, especially when surgical repair is being considered.

**Thoracic Outlet Syndrome (TOS)**
- Also see CH-34 Thoracic Outlet Syndrome in the Chest Imaging Guidelines
- This is a contentious diagnosis, and the much more common carpal tunnel syndrome should be excluded prior to considering a diagnosis of thoracic outlet syndrome. (See PN-2.1 Carpal Tunnel Syndrome, MS-22 Wrist in the Musculoskeletal Imaging Guidelines, and SP-11.5 Carpal Tunnel Syndrome under Cervical Radiculopathy in the Spine Imaging Guidelines).
- Diagnosis of neurogenic TOS is most reliably made by the electro-diagnostic studies (EMG/NCV) which will be done to exclude carpal tunnel syndrome.
- Brachial plexus imaging is appropriate only in patients in whom the diagnosis has been confirmed by EMG and who have failed a 2 to 3 month trial of conservative management and are being considered for surgical treatment.
- **Note:** "Adson’s sign" (transient radial pulse extinction by abduction and external rotation of the arm) is common in normal individuals and is not itself an indication for advanced imaging.

**PN-5~LUMBAR AND LUMBOSACRAL PLEXUS**

**Anatomy:**
- The upper lumbar plexus is located in the abdominal retroperitoneal space and gives rise to, among others, the femoral, lateral femoral cutaneous, and obturator nerves.
- The lumbosacral plexus lies in the pelvis and gives rise to the sciatic and gluteal nerves.
- Radiation plexopathy, malignant infiltration, and trauma can involve these structures.

**Malignant Infiltration:**
- As with brachial plexus infiltration, pain is early and severe, and the sensorimotor findings follow the onset of pain.
- Colon cancer, gynecological cancers, and genitourinary cancers are the most common primaries.
MRI of the abdomen (CPT®74183) or pelvis (CPT®72197) without and with contrast with fat suppression imaging is appropriate, and the imaging study chosen (abdomen vs pelvis) will depend on which plexus is involved.

- Tumors usually appear as soft tissue masses compressing the plexus.
- CT scan with contrast (CPT®74160 or CPT®72193) is inferior to MRI but can be performed if MRI is unavailable or contraindicated.

**Reference:**


**Radiation Plexopathy**

- Less common in the lumbar and lumbosacral plexi than in the brachial plexus.
- Imaging is similar to that performed for radiation plexitis of the brachial plexus (see PN-4 Brachial plexus).

**Trauma**

- These cases will involve either major abdominal trauma or local surgical procedures and initial management will be in an inpatient setting.
- If later surgical repair of a plexus injury is contemplated, noncontrast MRI of the relevant region with fat suppression may be appropriate. These cases should be sent for Medical Director review.

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**PN-6~MUSCLE DISORDERS**

- There are no established indications for advanced imaging in the muscular dystrophies. However, in neuromuscular disorders, the inflammatory myopathies, and Gaucher’s Disease, noncontrast MRI has its roles. CT is not useful for visualizing muscle disease.

**PN-6.1 Neuromuscular Disease:** Both myasthenia gravis (MG) and the less common Lambert-Eaton myasthenic syndrome (LEMS) are associated with chest neoplasms (thymoma and small cell lung cancer, respectively).

  - **Myasthenia Gravis (MG):**
    - Initial diagnosis of MG is by clinical presentation, electro-diagnostic studies, and antibody titers.
    - 15% of patients (mostly elderly) harbor a thymoma, but most others have thymic hypertrophy.
    - Thymectomy is widely used as a treatment of MG with or without thymoma.
    - Chest CT with contrast (CPT®71260) is part of the initial evaluation of confirmed myasthenia gravis.
    - In patients with initial chest CT scans that are negative and who have not undergone thymectomy, repeat chest CT is not appropriate without a specific indication (e.g. symptoms of chest mass, rising anti-striated muscle antibody titers, or need for preoperative evaluation).

  - **Lambert–Eaton myasthenic syndrome (LEMS):**
    - Oat cell lung tumors (small cell lung cancer) are found in about half of cases. The cancer often cannot be found initially but will surface a month to 1-2 years after diagnosis of the neuromuscular syndrome.
Initial diagnosis of LEMS is by clinical presentation, electro-diagnostic studies, and antibody titers (anti-voltage gated calcium channel).

Chest x-ray and chest CT with contrast (CPT®71260) are appropriate in confirmed cases.

If the initial tumor evaluation is negative, chest imaging can be repeated in 3 months or when symptoms of a chest mass are present.

Although no published consensus has been established regarding further follow-up imaging, if the second chest CT is negative, repeat chest imaging at 6, 12, and 24 months seems appropriate.

See PN-3 Polyneuropathy for use of PET in paraneoplastic syndromes.

Reference:
- N Engl J Med 2003;349:1543-1554 (Includes useful lists of the various syndromes and the antibodies associated with them)

- Stiff man syndrome
  - A rare presynaptic disorder which can be associated with small cell lung cancer and breast cancer in those with anti-amphiphysin antibodies.
  - Chest CT with contrast (CPT®71260) and, in women, mammography, are appropriate.

  Reference:

• PN-6.2 Inflammatory Muscle Diseases
  - Includes dermatomyositis, polymyositis, and sporadic inclusion body myositis.
  - Advanced imaging is used in these disorders for three purposes:
    1) Selection of biopsy site
    2) Treatment monitoring
    3) Detection of occult malignancy (for patients with dermatomyositis and polymyositis)

  - Initial evaluation:
    - Involvement of muscles is patchy, and noncontrast MRI can be useful to select biopsy sites.
    - Noncontrast MRI of one or both thighs is usual (CPT®73718).
    - Studies have shown both the diagnostic value and cost effectiveness of this approach.

  References:
  - AJR 1995;165:1469-1471
  - Radiographics 1995;15:563-574
  - Curr Rheumatol Reports 2001;3:334-345

  - Sporadic inclusion body myositis is seen in older adults. Involvement of the deep finger flexors is early and striking, and noncontrast MRI of the forearm (CPT®73218) can be useful to establish the diagnosis early in the course.
    - Neurology 1997;48:863-866

  - Management of Inflammatory Muscle Diseases:
    - Clinical evaluation of muscle strength and endurance along with assay of muscle enzyme levels (especially CPK) is the principal method of monitoring the results of therapy in all three disorders.
- Noncontrast MRI, including fat suppression techniques, can be useful, especially when enzyme and clinical function assessments differ.
- When available, P-31 MRS has also shown value.
- No data has emerged to support surveillance imaging in these patients.

**References:**

- *Rheumatology* 2000;39:7-17

1. **Search for occult neoplasm in adults with dermatomyositis and in all patients with polymyositis:**
   - Lung and ovarian tumors are the most common, but lymphomas and other carcinomas can also be found.
   - Chest CT with contrast (CPT®71260) and pelvic ultrasound (in women) (CPT®76856 or CPT®76857 and/or CPT®76830 [transvaginal]) should be done initially.
   - CT abdomen and pelvis with contrast (CPT®74160 and CPT®72193) are indicated if the above fail to make a diagnosis.
   - Tumors may remain occult for months to several years after the onset of the myositis.

**Reference:**

- *Lancet* 2001;357:96-100

2. **PN-6.3 Gaucher's Disease (storage disorders)**
   - See also AB-16 Gaucher’s Disease in the Abdomen Imaging Guidelines.
   - Gaucher’s disease is group of autosomal recessive inborn errors of metabolism characterized by lack of the enzyme acid β-glucuronidase with destructive ceramide storage in various tissues.
   - Gaucher’s disease is a treatable disorder (enzyme replacement) in which the liver, spleen, and bone marrow/bones are the most affected organs.
   - This guideline addresses Type I Gaucher’s disease, which is by far the most common type in North America.
   - MRI is used to follow progression of disease in order to make treatment decisions, to monitor response to treatment, and to evaluate complications as they occur.
   - Liver and spleen size are followed by annual noncontrast abdominal MRI (CPT®74181).
   - Annual noncontrast thigh MRI (CPT®73718) is used to follow marrow replacement by the disease and to monitor response to treatment. MRI of a single thigh should be sufficient.
   - These patients often develop avascular necrosis of the hips and compression fractures of the spine, and relevant noncontrast MRI scans are appropriate when the clinical setting suggests these complications.
   - In addition, many experts routinely perform MRI of the hips in untreated patients.

**References:**

- *BJR* 2002;75 suppl 1:A13-A24
- *Haematologica* 2000;85:792-799
PN-7~NEWER IMAGING TECHNIQUES

• PN-7.1 Magnetic Resonance Neurography (MRN)
  o MRI using a phased array of coils can be used to produce striking T2
    weighted images of Wallerian degeneration in larger peripheral nerves (>2
    mm diameter) involved in a variety of pathological processes.
  o At this time there is no compelling evidence indicating that the results of such
    studies add significant information to the knowledge that can be obtained by
    traditional clinical and electro-diagnostic studies.
  o MRN must be regarded as experimental at this time.
  o Current studies of the value of MRN are plagued by small sample size, limited
    clinical definition of the cases, and lack of longer term follow-up.
  o References:
     Neurology 2002;58:1597-1602
     Cigna Medical Coverage Policy 0316, Magnetic resonance neurography. March
      15, 2011
     Aetna Clinical Policy Bulletin 0387, Magnetic resonance neurography. July 27,
      2010

PN-8~AMYOTROPHIC LATERAL SCLEROSIS (ALS)

• Progressive disease of motor neurons (primarily in the spinal cord)
• Results in muscle weakness and is typically fatal due to respiratory failure or
  complications which occur an average of 3 years following diagnosis.
• Hallmark feature of ALS is a combination of upper motor and lower motor neuron
  findings (e.g. spasticity or hyperreflexia may exist in combination with muscle
  atrophy and fasciculations in the setting of subacute progressive weakness,
  usually starting in one limb or involving the muscles of swallowing or respiration
  and gradually spreading to other limbs).
• Diagnosis of ALS is made clinically by history and a detailed neurological
  examination, and confirmed by NCV/EMG.
• Imaging studies are appropriate initially to rule out combined brain and spine
  disorders such as spinal cord compression and nerve root pathology which can
  occasionally mimic ALS
  o MRI of the brain, cervical, thoracic, and lumbar spine (contrast as requested)
    can be performed in cases of known or suspected ALS.
  o Repeat spine imaging is usually not necessary once the diagnosis has been
    established. Requests for repeat imaging should be evaluated based on the
    appropriate Spine guidelines.
• Reference:
  o Semin Neurol 2008;28(2):205-211
PN-9—PERIPHERAL NERVE SHEATH TUMORS (PNST)

- **Tumors (Schwannomas or Neurofibromas)** that arise from Schwann cells or other connective tissue of the nerve
  - These tumors can be located anywhere in the body including intraspinal, along the course of the peripheral nerves, and in a cutaneous distribution.
  - Neurofibromas are the hallmark of the genetic disorder Neurofibromatosis type 1. (see HD-23.2 Neurofibromatosis, Type 1 in the Head Imaging Guidelines)
  - Acoustic neuromas or Schwannomas, when bilateral or associated with other intracranial tumors, are diagnostic of Neurofibromatosis type 2 (see HD-23.2 Neurofibromatosis, Type 1 in the Head Imaging Guidelines).

- **Initial imaging studies in the evaluation of PNST include:**
  - MRI brain without and with contrast (CPT® 70553)
  - If a paraspinal neurofibroma is found at any level of the spine or if the patient has multiple simplex perineural neurofibromas, MRI of the cervical, thoracic, and lumbar spine without and with contrast (CPT® 72156, CPT® 72157, and CPT® 72158) should be performed, since all dorsal roots may be involved.
  - Follow-up imaging of PNST is generally not indicated unless there are new symptoms or neurological findings.
  - Also see HD-23.2 Neurofibromatosis, Type 1 in the Head Imaging Guidelines.

- **Malignant potential of PNST**
  - Vast majority are benign but malignant transformation can occur in up to 5% of patients with Neurofibromatosis type 1
  - Malignant PNST’s are also called MPNST, malignant schwannomas, or neurofibrosarcomas and tend to be large (average size 8 cm) with poorly defined margins on MRI.
  - Plexiform neurofibromas also have a higher incidence of malignant transformation.
  - **Imaging**
    - Close observation and occasional follow-up imaging of malignant neurofibromas is necessary depending on the location.
    - Metastatic work-up is appropriate, including CT chest and abdomen with contrast (CPT® 71260 and CPT® 74160) in patients with suspected or biopsy proven malignant PNST’s.

- **References:**
  - Neurosurg Focus 2007 June;22(6):E3
  - Clin Imaging 2008 Mar-April;32(2):121-127
  - Neurosurg Focus 2006 Jan;20(1):E1
PERIPHERAL NERVE DISORDERS REFERENCES

PN-1~General Guidelines


PN-2~Focal Neuropathy

- Fleckenstein JL, Wolfe GI. MRI vs EMG Which has the upper hand in carpal tunnel syndrome? *Neurology* 2002;58:1583-1584.

PN-3~Polyneuropathy


PN-4~Brachial Plexus

PN-5~Lumbar and Lumbosacral Plexus

PN-6~Muscle Disorders

PN-7~Newer Imaging Techniques
- Cigna Medical Coverage Policy 0316, Magnetic resonance neurography. March 15, 2011

PN-8~Amyotrophic Lateral Sclerosis

PN-9~Peripheral Nerve Sheath Tumors (PNST)