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, specialist and/or patient’s Primary Care Physician (PCP) may provide additional insight.

This version incorporates MSI accepted revisions prior to 11/30/06
ABBREVIATIONS for PERIPHERAL NERVE DISORDERS GUIDELINES

AIDS: Acquired Immunodeficiency Syndrome
CIDP: Chronic Inflammatory Demyelinating Polyneuropathy
CNS: central nervous system
CPK: creatine phosphokinase
CT: computed tomography
EMG: electromyogram
LEMS: Lambert-Eaton myasthenic syndrome
MG: myasthenia gravis
MRI: magnetic resonance imaging
MRN: magnetic resonance neurography
MRS: magnetic resonance spectroscopy
NCV: nerve conduction velocity
PET: positron emission tomography
PNS: peripheral nervous system
POEMS: Polyneuropathy, Organomegaly, Endocrinopathy, M-protein, Skin changes
TOS: Thoracic Outlet Syndrome
# Peripheral Nerve Imaging Guidelines

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PERIPHERAL NERVE DISORDERS IMAGING GUIDELINES

PN-1~ GENERAL GUIDELINES

• Advanced imaging plays a limited role in the diagnosis and management of disorders of peripheral nerves and muscles. The extent of that role is currently being defined.
  o However, 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).

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 obtainable 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
      Accessed November 22, 2006
    ➢ Cigna Healthcare coverage position 0316, Magnetic resonance neurography. Revised March 15, 2006
• The Peripheral Nerve Disorders guidelines are the same for both the pediatric population and the adult population, unless there are specific Pediatric guidelines (highlighted in yellow).
• Reference:
• **PN-2.1 Carpal tunnel syndrome:**
  o Also see MS-18 Wrist in the Musculoskeletal guidelines and SP-12 Carpal tunnel syndrome under Cervical Radiculopathy in the Spine guidelines.
  o 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).
  o Noncontrast MRI (CPT 73221) reveals median nerve compression fairly well, but is less sensitive than nerve conduction measurements.
  o At this time, advanced imaging has no established role in the evaluation of carpal tunnel syndrome.
  o References:
    ➢ *Neurology* 2002;58:1583-1584
    ➢ *Neurology* 2002;58:1597-1602

• **PN-2.2 Ulnar neuropathy:**
  o Non-contrast MRI of the elbow (CPT 73221) may be of value as a preoperative study in some patients with documented ulnar neuropathy (confirmed by electro-diagnostic studies) who have failed non-operative management.
  o MRI can identify bony or soft tissue structures compressing the nerve. Since MRI serves to facilitate preoperative planning, requests should originate with a surgical consultant and be referred to a Medical Director.
  o Reference:
    ➢ *Neurosurgery* 1996;38:458-465

• **PN-2.3 Tarsal tunnel syndrome:**
  o Also see MS-23.8 Tarsal Tunnel Syndrome and MS-23.3 Tarsal Coalition in the Musculoskeletal guidelines.
  o Uncommon neuropathy involving the ankle which is analogous to the more common carpal tunnel syndrome at the wrist.
  o Nerve conduction studies and clinical evaluation are indicated initially.
  o Noncontrast MRI of the ankle (CPT 73721) is acceptable as a preoperative study to evaluate for masses or associated coalition.
  o If the concern is for a tarsal coalition, noncontrast CT (CPT 73700) is an acceptable alternative.
  o References:
PN-2.4 Meralgia paresthetica:
- A common sensory neuropathy involving the lateral femoral cutaneous nerve as it exits the pelvis under the inguinal ligament.
- Patients have objective sensory loss in the region supplied by the nerve (lateral thigh and buttocks).
- Spinal imaging is not indicated.
- In cases recalcitrant to medical management, studies to exclude a pelvic mass may be appropriate (see Pelvis 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 and with contrast (CPT 72197) are 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-2.5 Femoral neuropathy:
- Can arise as a complication of pelvic surgery in women or, in patients on anticoagulants, as a complication of retroperitoneal bleeding.
- Pelvic CT can be performed either with (CPT 72193) or without (CPT 72192) contrast for evaluation in either setting.
- 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.

- 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 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 and EMG picture.
- 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-24 Multiple Myeloma and Plasmacytomatas in the Oncology guidelines.
- Advanced imaging is appropriate for the non-neurological entities of this rare osteosclerotic plasmacytoma syndrome.

**Subacute sensory neuronopathy and other paraneoplastic demyelinating neuropathies:**
- Also see ONC-28.3 Paraneoplastic Syndromes in the Oncology guidelines.
- 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.
- Advanced imaging should be performed as indicated for the associated malignancy or to search for malignancy (see Oncology guidelines).
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

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.

Reference:
- Ann Neurol 2000;48:105-108

Several collagen vascular diseases may present with a progressive polyneuropathy.
- Systemic lupus, Sjogren’s syndrome, Beçet’s disease, polyarteritis nodosa, Churg-Strauss syndrome, and Wegener’s granulomatosis can all present in this manner. (See HD-33 Cerebral Vasculitis in the Head guidelines).
- 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.

**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 helpful.
- 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.
  - Brachial plexus studies can be coded either as upper extremity other than joint MRI (CPT 73218) or as chest MRI (CPT 71550). Chest MRI will image both brachial plexi and is useful for comparing one plexus with the other.
  - MRI studies should be without and with contrast (CPT 73220 or 71552) when tumor is part of the differential diagnosis.
- Reference:
  - Radiographics 2000;20:1023-1032
  - 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 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 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 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-12 Cervical Radiculopathy in the Spine 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:
    - Am Fam Physician 2000;62:2067-2072
• **Trauma:** the cause and extent are generally obvious, but noncontrast MRI of the brachial plexus (CPT 73218 or 71550) is often useful, especially when surgical repair is being considered.

• **Birth trauma:** injury to the baby's upper (Erb's palsy) or lower (Klempke's palsy) plexus can occur during birth.
  - Noncontrast MRI of the brachial plexus (CPT 73218 or 71550) can be useful to define the defect.
  - If there is clinical suspicion for cervical nerve root avulsion, noncontrast cervical spine MRI (CPT 72141) may be useful.

• **Thoracic outlet syndrome (TOS):**
  - 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-18 Wrist in the Musculoskeletal guidelines, and SP-12 Carpal tunnel syndrome under Cervical Radiculopathy in the Spine 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.

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**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 only (CPT 74160 or 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 regardless of the presence of 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.

- **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)
  - In children with dermatomyositis, MRI is often used to confirm a clinical diagnosis and thus avoid a biopsy.
  - **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.

Dermatomyositis in children: an issue specific to this disorder is the presence of progressive calcification in muscles. Noncontrast CT of the thighs (CPT 73700) is the procedure of choice to follow this condition, but MRI (CPT 73718) is often used since it permits assessment of the primary muscle disease as well.

References:
- *Rheumatology* 2000;39:7-17

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) should be done initially.
- CT abdomen and pelvis with contrast (CPT 74160 and 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

PN-6.3 Gaucher Disease (storage disorders):
- 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 the results of 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 suffice.
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
  McGovern M. *Gaucher Disease*. Updated October 15, 2003
  Accessed November 22, 2006
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