



CLINICAL GUIDELINES

Pediatric Peripheral Vascular Disease (PVD) Imaging Guidelines

Version 1.1

Effective October 15, 2020



eviCore healthcare Clinical Decision Support Tool Diagnostic Strategies: This tool addresses common symptoms and symptom complexes. Imaging requests for individuals with atypical symptoms or clinical presentations that are not specifically addressed will require physician review. Consultation with the referring physician, specialist and/or individual's Primary Care Physician (PCP) may provide additional insight.

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Pediatric Peripheral Vascular Disease (PVD) Imaging Guidelines

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Procedure Codes Associated with PVD Imaging

MRA	CPT®
Upper Extremity MRA	73225
Lower Extremity MRA	73725
CTA	CPT®
CTA Abdominal Aorta with Bilateral Iliofemoral Runoff	75635
Upper Extremity CTA	73206
Lower Extremity CTA	73706
Nuclear Medicine	CPT®
PET Imaging; limited area (this code not used in pediatrics)	78811
PET Imaging; skull base to mid-thigh (this code not used in pediatrics)	78812
PET Imaging; whole body (this code not used in pediatrics)	78813
PET with concurrently acquired CT; limited area (this code rarely used in pediatrics)	78814
PET with concurrently acquired CT; skull base to mid-thigh	78815
PET with concurrently acquired CT; whole body	78816
Ultrasound	CPT®
Duplex scan of extracranial arteries; complete bilateral study	93880
Duplex scan of extracranial arteries; unilateral or limited study	93882
Non-invasive physiologic studies of extracranial arteries, complete bilateral study	93875
Limited bilateral noninvasive physiologic studies of upper or lower extremity arteries	93922
Complete bilateral noninvasive physiologic studies of upper or lower extremity arteries	93923
Duplex scan of upper extremity arteries or arterial bypass grafts; complete bilateral	93930
Duplex scan of upper extremity arteries or arterial bypass grafts; unilateral or limited	93931
Non-invasive physiologic studies of extremity veins, complete bilateral study	93965
Duplex scan of extremity veins including responses to compression and other maneuvers; complete bilateral study	93970
Duplex scan of extremity veins including responses to compression and other maneuvers; unilateral or limited study	93971
Duplex scan of hemodialysis access (including arterial inflow, body of access, and venous outflow)	93990

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PEDPVD-1.0: General Guidelines

- A recent (within 60 days) face to face evaluation including a detailed history, physical examination, and appropriate laboratory studies should be performed prior to considering advanced imaging (CT, MR, Nuclear Medicine), unless the patient is undergoing guideline-supported scheduled imaging evaluation.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic patients for disorders involving the peripheral vascular system is not supported. Advanced imaging of the peripheral vascular system should only be approved in patients who have documented active clinical signs or symptoms of disease involving the peripheral vascular system.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the peripheral vascular system are not necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect patient management or treatment decisions.

PEDPVD-1.1: Age Considerations

Many conditions affecting the peripheral vascular system in the pediatric population are different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and adult populations, differences may exist in management due to patient age, comorbidities, and differences in disease natural history between children and adults.

- Patients who are < 18 years old should be imaged according to the Pediatric peripheral vascular disease imaging guidelines, and patients who are ≥ 18 years old should be imaged according to the Adult peripheral vascular disease imaging guidelines, except where directed otherwise by a specific guideline section.

PEDPVD-1.2: Imaging Appropriate Clinical Evaluation

- See **PEDPVD-1.0: General Guidelines**

PEDPVD-1.3: Modality General Considerations

- MRI
 - ◆ MRI is generally performed without and with contrast unless the patient has a documented contraindication to gadolinium or otherwise stated in a specific guideline section.
 - ◆ Due to the length of time required for MRI acquisition and the need to minimize patient movement, anesthesia is usually required for almost all infants (except neonates) and young children (age < 7 years), as well as older children with delays in development or maturity. This anesthesia may be administered via oral or intravenous routes. In this patient population, MRI imaging sessions should be planned with a goal of minimizing anesthesia exposure adhering to the following considerations:
 - MRI procedures can be performed without and/or with contrast use as supported by these condition-based guidelines. If intravenous access will

- already be present for anesthesia administration and there is no contraindication for using contrast, imaging without and with contrast may be appropriate if requested. By doing so, the requesting provider may avoid repetitive anesthesia administration to perform an MRI with contrast if the initial study without contrast is inconclusive.
- Recent evidence-based literature demonstrates the potential for gadolinium deposition in various organs including the brain after the use of MRI contrast.
 - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
 - If multiple body areas are supported by eviCore guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session.
 - ◆ The presence of surgical hardware or implanted devices may preclude MRI.
 - ◆ The selection of best examination may require coordination between the provider and the imaging service.
- CT
- ◆ CT or CTA may be appropriate for further evaluation of abnormalities suggested on prior US or MRI Procedures.
 - ◆ CT may be appropriate without prior MR or US, especially in the following (non-exhaustive list of) settings:
 - Lymphatic malformations
 - Vascular abnormalities including vasculitis, thrombosis, narrowing, aneurysm, dissection, and varices.
 - For preoperative planning or assessment of post-operative complications.
 - ◆ In some cases, especially in follow-up of a known finding, it may be appropriate to limit the exam to the region of concern to reduce radiation exposure.
 - ◆ CT should not be used to replace MRI in an attempt to avoid sedation unless listed as a recommended study in a specific guideline section.
 - ◆ The selection of best examination may require coordination between the provider and the imaging service.
- Ultrasound
- ◆ Ultrasound can be helpful in evaluating arterial, venous, and lymphatic malformations.
 - ◆ Ultrasound can be limited by the imaging window and the patient body type.
 - ◆ CPT® codes vary by body area and presence or absence of Doppler imaging and are included in the table at the beginning of this guideline.
- Nuclear Medicine
- ◆ Nuclear medicine studies are rarely used in the evaluation of peripheral vascular disorders, but are indicated in the following circumstances:

- Lymphoscintigraphy (CPT® 78195) is indicated for evaluation of lower extremity lymphedema when a recent Doppler ultrasound is negative for valvular insufficiency.
 - Vascular flow imaging (CPT® 78445) is an obsolete study that has been replaced by MRA, CTA, or Duplex ultrasonography, and is not supported for any indication at this time.
 - Venous thrombosis imaging (CPT® 78456, CPT® 78457, and CPT® 75458) are obsolete studies that have been replaced by MRA, CTA, or Duplex ultrasonography, and are not supported for any indication at this time.
 - Radiopharmaceutical nuclear medicine studies (CPT® 78800, CPT® 78801, CPT® 78802 or CPT® 78803) can be approved for evaluation of the following:
 - Mycotic aneurysms
 - Vascular graft infection
 - Infection of central venous catheter or other indwelling device
- The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

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PEDPVD-2: Vascular Anomalies

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PEDPVD-2.1: General Information

Vascular and lymphatic malformations encompass a broad variety of conditions and have very heterogeneous natural history and treatment approaches. Lesions can be divided into low flow lesions (lymphatic, capillary and venous malformations), and high flow lesions (arteriovenous malformations and fistulas).

- Patients with aggressive lesions being treated with systemic therapy can have imaging (see specific sections for details regarding modality and contrast level) approved for treatment response every 3 months during active treatment.
- Annual surveillance imaging of known vascular or lymphatic malformations can be approved for body areas where growth could cause significant organ dysfunction or functional impairment.

PEDPVD-2.2: Lymphatic Malformations

Lymphatic malformations are composed of dilated lymphatic channels filled with proteinaceous fluid and do not connect to normal lymphatic channels. They are typically soft, non-pulsatile masses with normal overlying skin.

- Ultrasound is indicated as an initial examination for superficial lesions.
 - ◆ Large lesion characterization may be limited by ultrasound imaging window.
 - ◆ Ultrasound is also limited in evaluating malformation relationship to airway or bony structures.
- MRI without contrast or without and with contrast of the affected body part is indicated for:
 - ◆ Lymphatic malformations involving deep tissues
 - ◆ Malformations too large to be completely imaged with ultrasound
 - ◆ Inconclusive ultrasound findings
 - ◆ Preoperative planning
- CT is of limited value in evaluating lymphatic malformations
 - ◆ CT with contrast of the affected body part can be approved for lesions with acute enlargement and concerns for compression when MRI is contraindicated.

PEDPVD-2.3: Venous Malformations

Venous malformations are slow-flow lesions characterized by dilated venous spaces and a normal arterial component. They are soft, compressible, non-pulsatile lesions that are usually blue to deep purple in color. Lesions can range from very small to large infiltrating ones. Some may change size with Valsalva.

Venous malformations are usually isolated, but they may be seen in multiple syndromes including Klippel-Trenaunay (KT) syndrome, Blue Rubber Bleb Nevus syndrome (BRBN), Maffucci syndrome, Proteus syndrome, Bannayan-Riley-Ruvalcaba syndrome, Parkes-Weber syndrome and congenital lipomatous overgrowth, vascular malformations, epidermal nevi and scoliosis/skeletal/spinal anomalies (CLOVES) syndrome.

- Ultrasound with Doppler is indicated as an initial examination for superficial lesions.
 - ◆ Large lesion characterization may be limited by ultrasound imaging window.
 - ◆ Ultrasound is also limited in evaluating malformation relationship to airway or bony structures.
- MRI without contrast or without and with contrast of the affected body part can be approved for venous malformations for preoperative assessment to evaluate the extent of malformation and their relationship to normal structures.
- MRA or CTA has a limited role in evaluating most venous malformations, but may be approved (contrast as requested of the affected body part) if MRI or CT are equivocal and the results will impact acute management decisions.
- CT can also be used to characterize venous malformations and their relationship to normal structures but is generally not as accurate as MRI.
 - ◆ CT with contrast of the affected body part can be approved when MRI is inconclusive or contraindicated

PEDPVD-2.4: Capillary Malformations

- Capillary malformations also known as port wine stains are characterized by a collection of small vascular channels in the dermis and generally do not require advanced imaging because the diagnosis is made clinically. However, MR imaging (without contrast or without and with contrast) may be approved to evaluate occult underlying neurologic structures, since these malformations are associated with encephalocele, spinal dysraphism, or Sturge-Weber syndrome

PEDPVD-2.5: Arteriovenous Malformations (AVMs) and Fistulas

Arteriovenous malformations are characterized by a network of multiple abnormal vascular channels interposed between enlarged feeding arteries and draining veins. The arteriovenous fistula has a single communication interposed between a feeding artery and a draining vein. The normal capillary bed is absent in both lesions. Both lesions may have an aggressive clinical course and are characterized by a reddish pulsatile mass which has a thrill or bruit. Though often recognized at birth, these lesions may grow and present near adolescence.

- Ultrasound with Doppler is indicated as an initial examination for superficial lesions.
 - ◆ Large lesion characterization may be limited by ultrasound imaging window.
 - ◆ Ultrasound is also limited in evaluating AVM relationship to airway or bony structures.
- MRI without contrast or without and with contrast of the affected body part is also indicated for evaluation of AVMs, and is useful in evaluating the extent of AVMs and their relationship to normal structures.
- MRA (contrast as requested) of the affected body part can be approved for evaluation and surveillance of known AVMs.
- It is unusual for both MRI and MRA to be necessary for routine treatment response or surveillance imaging of AVMs, but both may be approved for preoperative planning.
- CT and CTA can also be used to characterize AVMs and their relationship to normal structures, but is generally not better than MRI and has associated radiation risks.
 - ◆ CT with contrast and/or CTA (contrast as requested) of the affected body part can be approved when MRI and/or MRA is inconclusive or contraindicated.

PEDPVD-2.6: Vascular Tumors

Vascular tumors include a variety of benign, borderline, and malignant tumors, which have variable clinical courses, including but not limited to Epithelioid hemangioma, Kaposiform hemangioendothelioma, Kaposi sarcoma, Epithelioid hemangioendothelioma, and Angiosarcoma of soft tissue.

- Ultrasound with Doppler is indicated as an initial examination for vascular tumors.
 - ◆ Large lesion characterization may be limited by ultrasound imaging window.
 - ◆ Ultrasound is also limited in evaluating malformation relationship to airway or bony structures.
- MRI without contrast or without and with contrast of the affected body part is also indicated for evaluation of vascular tumors, and is useful in evaluating the extent of arteriovenous malformations and their relationship to normal structures, as well as response to therapy.
- MRA (contrast as requested) of the affected body part can be approved for evaluation and surveillance of known vascular tumors.

- It is unusual for both MRI and MRA to be necessary for routine treatment response or surveillance imaging of vascular tumors, but both may be approved for preoperative planning.
- CT and CTA can also be used to characterize vascular tumors and their relationship to normal structures, but is generally not better than MRI and has associated radiation risks.
 - ◆ CT with contrast and/or CTA (contrast as requested) of the affected body part can be approved when MRI and/or MRA is inconclusive or contraindicated.

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PEDPVD-3: Vasculitis

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PEDPVD-3.1: General Information

Systemic vasculitis is much less common in children than in adults, although the diagnostic pathways and treatment options are similar.

- PET/CT is considered investigational for management of pediatric vasculitis at this time.
 - ◆ There are limited data suggesting PET may have similar accuracy to MRA in the initial diagnosis of Takayasu arteritis but is not helpful in assessing treatment response and has not been shown to improve patient outcomes to date.

PEDPVD-3.2: Large Vessel Vasculitis

Takayasu arteritis is the predominant large vessel vasculitis occurring in children.

- Any of the following modalities may be indicated for evaluation of Takayasu arteritis:
 - ◆ MRA of the affected body area(s) (contrast as requested)
 - ◆ CTA of the affected body area(s) (contrast as requested)
 - ◆ Ultrasound with Doppler of the affected body area(s)
- Imaging is indicated at the following intervals:
 - ◆ Every 3 months for treatment response during active treatment in patients being treated with systemic therapy.
 - See specific sections for details regarding modality and contrast level .
 - ◆ Annually for surveillance of known involved body areas to detect progressive vascular damage that may require intervention.

PEDPVD-3.3: Medium Vessel Vasculitis

Polyarteritis nodosa and Kawasaki Disease are the primary medium vessel vasculitides occurring in children.

- Imaging guidelines for Kawasaki Disease are in the pediatric cardiac imaging guideline, see **PEDCD-6: Kawasaki Disease**
- For evaluation of polyarteritis nodosa:
 - ◆ Any of the following modalities may be indicated:
 - MRA of the affected body area(s) (contrast as requested)
 - CTA of the affected body area(s) (contrast as requested)
 - Ultrasound with Doppler of the affected body area(s)
 - ◆ Imaging is indicated at the following intervals:
 - Every 3 months during active treatment with systemic therapy for treatment response.
 - For details regarding modality and contrast level see **PEDPVD-1.3: Modality General Considerations**
 - Annually for surveillance of known involved body areas to detect progressive vascular damage that may require intervention.

PEDPVD-3.4: Small Vessel Vasculitis

- Advanced imaging is not sensitive enough to detect changes in small vessels, and is not indicated for primary assessment of any small vessel vasculitis.
- End-organ damage occurs with several of the small vessel vasculitides. Advanced imaging is indicated for the following:
 - ◆ Granulomatosis with polyangiitis (GPA, formerly known as Wegener's granulomatosis):
 - CT Sinuses (CPT®70486) and/or CT Chest without contrast (CPT®71250) or with contrast (CPT®71260) is indicated for the following:
 - New or worsening clinical symptoms affecting the body area requested
 - To assess response to medical therapy when a change in treatment regimen is being considered
 - Annually-to evaluate the extent of disease
 - ◆ Eosinophilic granulomatosis with polyangiitis (EGPA, formerly known as Churg-Strauss Syndrome):
 - CT Chest without contrast (CPT®71250) or with contrast (CPT®71260) is indicated in the following circumstances:
 - New or worsening clinical symptoms affecting the body area requested
 - To assess response to medical therapy when a change in treatment regimen is being considered
 - Annually-to evaluate the extent of disease
 - ◆ Immune complex associated small-vessel vasculitis [immunoglobulin A-associated vasculitis (IgAV)]:
 - Doppler ultrasound of the affected body part (most commonly abdomen) is indicated in the following circumstances:
 - New or worsening clinical symptoms affecting the body area requested
 - To assess response to medical therapy when a change in treatment regimen is being considered
 - Annually-to evaluate the extent of disease

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PEDPVD-4: Disorders of the Aorta and Visceral Arteries

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PEDPVD-4.1: Thoracic Aortic Disease

- MRA Chest (CPT® 71555) or CTA Chest (CPT® 71275) can be used in patients with Takayasu arteritis, William syndrome, and Ehlers Danlos syndrome for both:
 - ◆ Screening
 - ◆ Follow-up of thoracic aortic abnormalities
- MRAs (preferred) or CTAs from the head through the pelvis may be performed in patients diagnosed with Loeys-Dietz syndrome for:
 - ◆ Screening-one time
 - ◆ Follow-up imaging of discovered aneurysms-no more frequently than annually as requested by a specialist
- For other conditions please see discussions indicated elsewhere in the guidelines
 - ◆ Marfan Syndrome- See **CH-29.3 Screening Guidelines for Familial Syndromes**
 - ◆ Coarctation of the Aorta- See **PEDCD-2.3 Congenital Heart Disease**
 - ◆ Congenital rubella syndrome- See **PEDCD-2.3 Congenital Heart Disease**
 - ◆ Kawasaki Syndrome- See **PEDCD-6 Kawasaki Disease**
 - ◆ Neurofibromatosis- See **PEDCD-1.2 Pediatric Cardiac Imaging Appropriate Clinical Evaluation**

PEDPVD-4.2: Aortic Congenital Vascular Malformations

- Cardiac MRI without contrast (CPT® 75557) or without and with contrast (CPT® 75561), MRA Chest (CPT® 71555), CT Chest with contrast (CPT® 71260), or CTA Chest (CPT® 71275) may be indicated for evaluation.
- Vascular rings may impact both the esophagus and trachea. See **PEDNECK-7: Esophagus** and/or **PEDNECK-8: Trachea** for additional guidelines.

PEDPVD-4.3: Visceral Artery Aneurysms

- Visceral artery imaging indications in pediatric patients are identical to those for adult patients. See **PVD-6: Aortic Disorders and Renal Vascular Disorders and Visceral Artery Aneurysms** for imaging guidelines.

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