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1. The terms of the applicable benefit plan document in effect on the date of service
2. Any applicable laws and regulations
3. Any relevant collateral source materials including coverage policies
4. The specific facts of the particular situation

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# PEDIATRIC PERIPHERAL VASCULAR DISORDER (PVD) IMAGING GUIDELINES

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### PEDPVD-1~General Guidelines

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PEDPVD-1~General Guidelines

PEDPVD-1.1 Pediatric Peripheral Vascular Disease Imaging 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, minor differences may exist in management due to individual age, comorbidities, and differences in disease natural history between children and adults.

- Individuals age < 18 years old should be imaged according to the Pediatric Peripheral Vascular Disease Imaging Guidelines, and individuals age ≥ 18 years should be imaged according to the Peripheral Vascular Disease Imaging Guidelines, except where directed otherwise by a specific guideline section.

PEDPVD-1.2 Pediatric Peripheral Vascular Disease Imaging Appropriate Clinical Evaluation

- 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, unless the individual is undergoing guideline-supported scheduled follow-up imaging evaluation.

- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic individuals for disorders involving the peripheral vascular system is not supported. Advanced imaging of the peripheral vascular system should only be approved in individuals 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 individual management or treatment decisions.

PEDPVD-1.3 Pediatric Peripheral Vascular Disease Imaging Modality General Considerations

- MRI
  - MRI is generally performed without and with contrast unless the individual has a documented contraindication to gadolinium or otherwise stated in a specific guideline section
Due to the length of time for image acquisition and the need for stillness, anesthesia is required for almost all infants and young children (age < 7 years), as well as older children with delays in development or maturity. In this patient population, MRI imaging sessions should be planned with a goal of avoiding a short-interval repeat anesthesia exposure due to insufficient information using the following considerations:

- MRI should always be performed without and with contrast unless there is a specific contraindication to gadolinium use, since the individual already has intravenous access for anesthesia.
- 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 individual 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.

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.
References

   https://www.acr.org/~/media/ACR/Documents/PGTS/guidelines/MRI.pdf
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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 and venous malformations), and high flow lesions (arteriovenous malformations).

✓ Individuals 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 filled with proteinaceous fluid and may 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.
  o Large lesion characterization may be limited by ultrasound imaging window.
  o 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:
  o Lymphatic malformations involving deep tissues
  o Malformations too large to be completely imaged with ultrasound
  o Inconclusive ultrasound findings
  o Preoperative planning

✓ CT is of limited value in evaluating lymphatic malformations
  o 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 soft, compressible, non-pulsatile lesions that are usually blue to deep purple in color. Lesions can range from very small to large and invasive. 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’s syndrome, Proteus syndrome, Bannayan-Riley-Ruvalcaba syndrome, and CLOVE/S 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 have 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~Arteriovenous Malformations (AVMs)**

Arteriovenous malformations may have an aggressive clinical course and are characterized by firm pink abnormalities which have 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-2.5~Vascular Tumors**

Vascular tumors include a variety of benign, borderline, and malignant tumors, which have variable clinical courses

- 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-3.1 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
  o 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 individual outcomes to date

PEDPVD-3.2 Large Vessel Vasculitis
Takayasu arteritis is the predominant large vessel vasculitis occurring in children.

✓ Any of the following are indicated for evaluation of Takayasu arteritis:
  o MRA of the affected body area(s) (contrast as requested)
  o CTA of the affected body area(s) (contrast as requested)
  o Ultrasound with Doppler of the affected body area(s)

✓ Individuals with aggressive disease 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 involved body areas can be approved 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. See PEDCD-6~Kawasaki Disease for imaging guidelines for that disease.

✓ Any of the following are indicated for evaluation of Polyarteritis nodosa:
  o MRA of the affected body area(s) (contrast as requested)
  o CTA of the affected body area(s) (contrast as requested)
  o Ultrasound with Doppler of the affected body area(s)

✓ Individuals with aggressive disease 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 involved body areas can be approved 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, and the following advanced imaging is indicated:
  o Granulomatosis with polyangiitis (GPA, formerly known as Wegener’s granulomatosis)
      ▪ CT Sinuses (CPT®70486)
      ▪ CT Chest without contrast (CPT®71250) or with contrast (CPT®71260)
  o Eosinophilic granulomatosis with polyangiitis (EGPA, formerly known as Churg-Strauss Syndrome)
      ▪ CT Chest without contrast (CPT®71250) or with contrast (CPT®71260)
  o Immune complex associated small-vessel vasculitis (IgAV)
      ▪ Doppler ultrasound of the affected body part (most commonly abdomen)
  o These imaging studies are indicated in the following circumstances:
      ▪ New or worsening clinical symptoms affecting the body area requested
      ▪ Assessment of response to medical therapy when a change in treatment regimen is being considered
      ▪ Annual imaging to evaluate extent of disease

References
PEDPVD-4~Disorders Of The Aorta And Visceral Arteries

**PEDPVD-4.1 Thoracic Aortic Disease**

✓ MRA (CPT®71555) or CTA (CPT®71275) Chest can be used for screening and follow-up of thoracic aortic abnormalities in individuals with Loeys-Dietz syndrome, Marfan syndrome, coarctation of the aorta, Takayasu arteritis, neurofibromatosis, congenital rubella syndrome, or Kawasaki syndrome.

**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~ESOPAHGUS and/or PEDNECK-8~TRACHEA for additional guidelines.

**PEDPVD-4.3 Visceral Artery Aneurysms**

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

**References**