CIGNA MEDICAL COVERAGE POLICIES- RADIOLOGY

Pediatric Peripheral Vascular Disease (PVD) Imaging Guidelines

Effective Date: September 1, 2026

Instructions for use

The following coverage policy applies to health benefit plans administered by Cigna. Coverage policies are intended to provide guidance in interpreting certain standard Cigna benefit plans and are used by medical directors and other health care professionals in making medical necessity and other coverage determinations. Please note the terms of a customer's particular benefit plan document may differ significantly from the standard benefit plans upon which these coverage policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a coverage policy.

In the event of a conflict, a customer's benefit plan document always supersedes the information in the coverage policy. In the absence of federal or state coverage mandates, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of:

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

Coverage policies relate exclusively to the administration of health benefit plans. Coverage policies are not recommendations for treatment and should never be used as treatment guidelines.

This evidence-based medical coverage policy has been developed by EviCore, Inc. Some information in this coverage policy may not apply to all benefit plans administered by Cigna.

These guidelines include procedures EviCore does not review for Cigna. Please refer to the **Cigna CPT code list** for the current list of high-tech imaging procedures that EviCore reviews for Cigna.

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General Guidelines (PEDPVD-1)

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Procedure Codes Associated with PVD Imaging (PEDPVD)

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Description	CPT®
MRA	
Magnetic resonance angiography, head; without contrast material(s), followed by contrast material(s) and further sequence	70546
Magnetic resonance angiography, neck; without contrast material(s), followed by contrast material(s) and further sequences	70549
Magnetic resonance angiography, chest (excluding myocardium), with or without contrast material(s)	71555
Magnetic resonance angiography, pelvis, with or without contrast material(s)	72198
Magnetic resonance angiography, upper extremity, with or without contrast material(s)	73225
Magnetic resonance angiography, lower extremity, with or without contrast material(s)	73725
Magnetic resonance angiography, abdomen, with or without contrast material(s)	74185
CTA	
Computed tomographic angiography, head, with contrast material(s), including noncontrast images, if performed, and image postprocessing	70496
Computed tomographic angiography, neck, with contrast material(s), including noncontrast images, if performed, and image postprocessing	70498

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Description	CPT®
Computed tomographic angiography, chest (noncoronary), with contrast material(s), including noncontrast images, if performed, and image postprocessing	71275
Computed tomographic angiography (CTA), head and neck, with contrast material(s), including noncontrast images, when performed, and image postprocessing	70471
Computed tomographic angiography, upper extremity, with contrast material(s), including noncontrast images, if performed, and image postprocessing	73206
Computed tomographic angiography, lower extremity, with contrast material(s), including noncontrast images, if performed, and image postprocessing	73706
Computed tomographic angiography, abdomen and pelvis, with contrast material(s), including noncontrast images, if performed, and image postprocessing	74174
Computed tomographic angiography, abdomen, with contrast material(s), including noncontrast images, if performed, and image postprocessing	74175
CTA Abdominal Aorta with Bilateral Iliofemoral Runoff	75635
Ultrasound	
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

Description	CPT®
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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General Guidelines (PEDPVD-1.0)

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- A pertinent clinical evaluation since the onset or change in symptoms, including a detailed history, physical examination, appropriate laboratory studies, and basic imaging such as plain radiography or ultrasound should be performed prior to considering advanced imaging unless the individual is undergoing guideline-supported scheduled imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) can serve as a pertinent clinical evaluation.
- The use of advanced imaging to screen asymptomatic individuals for disorders involving the peripheral vascular system is not considered medically necessary unless otherwise stated in a specific guideline section.
- Advanced imaging of the peripheral vascular system is considered medically necessary in individuals who have documented active clinical signs or symptoms of disease involving the peripheral vascular system.
- Repeat imaging studies of the peripheral vascular system are not medically necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect the individual's management or treatment decisions unless otherwise stated in a specific guideline section.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Age Considerations (PEDPVD-1.1)

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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 general populations, differences may exist in management due to the individual's age, comorbidities, and differences in disease natural history between children and adults.

- Individuals who are 18 years old and younger should be imaged according to the Pediatric Peripheral Vascular Disease imaging guidelines if discussed. Any conditions not specifically discussed in the pediatric peripheral vascular disease imaging guidelines should be imaged according to the general peripheral vascular disease imaging guidelines. Individuals who are >18 years old should be imaged according to the general Peripheral Vascular Disease imaging guidelines, except where directed otherwise by a specific guideline section.

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Modality General Considerations (PEDPVD-1.3)

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- 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 required for MRI acquisition and the need to minimize the individual's 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 population, MRI 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 is considered medically necessary 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 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

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- CT or CTA is considered medically necessary for further evaluation of abnormalities suggested on prior US or MRI procedures.
- CT is considered medically necessary without prior MRI or US, especially in the following (non-exhaustive list of) settings:
  - Lymphatic malformations
  - Vascular abnormalities (including vasculitis, thrombosis, narrowing, aneurysm, dissection, and varices)
  - For pre-operative 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 the 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's 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.
- 3D Rendering
  - 3D Rendering indications in pediatric imaging are identical to those in the general imaging guidelines. See **3D Rendering (Preface-4.1)** in the Preface Imaging Guidelines
- 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 indicated and warranted for specific clinical situations.

References (PEDPVD-1)

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13. Center for Drug Evaluation and Research. New warnings for gadolinium-based contrast agents (GBCAs) for MRI. U.S. Food and Drug Administration. Published May 16, 2018. https://www.fda.gov/drugs/drug-safety-and-availability/fda-drug-safety-communication-fda-warns-gadolinium-based-contrast-agents-gbcas-are-retained-body.
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Vascular Anomalies (PEDPVD-2)

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Lymphatic Malformations (PEDPVD-2.2)

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Initial imaging

- Ultrasound is medically necessary as an initial examination for superficial lesions.
- CT with contrast of the affected body part is medically necessary for lesions with acute enlargement and concerns for compression when MRI is contraindicated.
- MRI without contrast or without and with contrast of the affected body part is medically necessary for:
  - Lymphatic malformations involving deep tissues
  - Malformations too large to be completely imaged with ultrasound
  - Inconclusive ultrasound findings
  - Preoperative planning

Clinical changes and monitoring treatment

MRI without contrast or without and with contrast of the affected body part is medically necessary every 3 months during active treatment for individuals with aggressive lesions being treated with systemic therapy.

MRI without contrast or without and with contrast of the affected body part is medically necessary for:

- Preoperative planning
- Post-treatment evaluation

CT with contrast of the affected body part is considered medically necessary to evaluate lesions with acute enlargement and concerns for compression when MRI is contraindicated.

Surveillance

Annual surveillance imaging with MRI without contrast or without and with contrast is medically necessary when lymphatic malformations are located in body areas where growth could cause significant organ dysfunction or functional impairment (e.g. airway, intestine).

Evidence Discussion

Vascular and lymphatic malformations encompass a broad variety of conditions and have very heterogeneous natural history and treatment approaches. Lesions can be

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divided into low flow lesions (lymphatic, capillary and venous malformations), and high-flow lesions (arteriovenous malformations and fistulas).

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. MRI is often used for further characterization of lymphatic malformations, especially for deep or complex lesions, providing detailed images without ionizing radiation. Large lesion characterization may be limited by ultrasound imaging window. Ultrasound is the initial imaging of choice however it is limited in evaluating malformation relationship to airway or bony structures.[1]

[1] Snyder EJ, Sarma A, Borst AJ, Tekes A. Lymphatic Anomalies in Children: Update on Imaging Diagnosis, Genetics, and Treatment. *AJR Am J Roentgenol*. 2022;218(6):1089-1101. doi:10.2214/AJR.21.27200.

Venous Malformations (PEDPVD-2.3)

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Venous Malformation Imaging Indications

Initial imaging

- Ultrasound with Doppler is medically necessary as an initial examination for superficial lesions.
- MRI without contrast or without and with contrast of the affected body part is medically necessary 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 is considered medically necessary (contrast as requested of the affected body part) if MRI or CT is 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 is considered medically necessary when MRI is inconclusive or contraindicated.

Clinical changes and monitoring treatment

MRI without contrast or without and with contrast of the affected body part is medically necessary for preoperative assessment of venous malformations.

CT Chest with contrast with PE protocol (CPT® 71260) or CTA Chest (CPT® 71275) is medically necessary to evaluate for suspected pulmonary embolism in individuals with Klippel-Trenaunay syndrome and CLOVES syndrome.

Surveillance

Annual surveillance imaging is medically necessary for venous malformations located in body areas where growth could cause significant organ dysfunction or functional impairment.

Evidence Discussion

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. These lesions can enlarge over time and become painful when associated with thrombophlebitis.[2]

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. Both Klippel-Trenaunay syndrome and CLOVES syndrome have been found to have increased risk of venous thrombosis and pulmonary embolism, particularly after surgery or sclerotherapy.

[2] Bertino FJ, Hawkins CM. Contemporary management of extracranial vascular malformations. *Pediatr Radiol*. 2023;53:1600–1617. doi:10.1007/s00247-023-05670-1.

Capillary Malformations (PEDPVD-2.4)

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Imaging Indications for Capillary Malformations

Initial Imaging

- MRI (without contrast or without and with contrast) is considered medically necessary to evaluate occult underlying neurologic structures associated with encephalocele, spinal dysraphism, or Sturge-Weber syndrome.

Evidence Discussion

Capillary malformations including Nevus simplex (NS) and Port wine birthmarks (PWBs) 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, MRI is considered medically necessary to evaluate underlying neurologic structures in specific cases, such as suspected Sturge-Weber syndrome.[3] Additional imaging is not considered medically necessary in the absence of other complex associated clinical findings such as Sturge-Weber syndrome (SWS) and Klippel-Trenaunay syndrome (KTS). [4]

[3] Bertino FJ, Hawkins CM. Contemporary management of extracranial vascular malformations. *Pediatr Radiol*. 2023;53:1600–1617. doi:10.1007/s00247-023-05670-1

[4] Paradiso MM, Shah SD. Infantile Hemangiomas and Vascular Anomalies. *Pediatr Ann*. 2024;53(4):e129-e137. doi:10.3928/19382359-20240205-04.

Arteriovenous Malformations (AVMs) and Fistulas (PEDPVD-2.5)

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Imaging Indications for AVMs and Fistulas

Initial Imaging

- Ultrasound with Doppler is medically necessary as an initial examination for superficial lesions.
- MRI without contrast or without and with contrast of the affected body part is also medically necessary to evaluate the extent of AVMs and their relationship to normal structures.
- MRA (contrast as requested) of the affected body part is medically necessary for evaluation 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 are considered medically necessary 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 is considered medically necessary when MRI and/or MRA is inconclusive or contraindicated.

Clinical changes and monitoring treatment

MRI or MRA (contrast as requested) of the affected body part is medically necessary for evaluation of treatment response.

It is unusual for both MRI and MRA to be necessary for routine treatment response or surveillance imaging of AVMs, but both may be medically necessary for preoperative planning.

CT with contrast and/or CTA (contrast as requested) of the affected body part is medically necessary when MRI and/or MRA is inconclusive or contraindicated.

Surveillance

MRI or MRA (contrast as requested) of the affected body part is medically necessary for annual surveillance of known AVMs located in body areas where growth could cause significant organ dysfunction or functional impairment.

Note:

For imaging indications specific to pulmonary AVM see **Pulmonary Arteriovenous Malformations (PEDCH-14.2)** in the Pediatric Chest Imaging guidelines, and for imaging indications specific to cerebral AVM see **Pediatric Intracranial Arteriovenous Malformations (AVM) (PEDHD-10.2)** in the Pediatric Head Imaging guidelines.

Evidence Discussion

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.

Vascular Tumors (PEDPVD-2.6)

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Imaging Vascular Tumors

Initial imaging

Any or all of the following imaging of the affected body part is considered medically necessary for the initial imaging and characterization of vascular tumors:

- Ultrasound with Doppler an initial examination
- MRI without contrast or without and with contrast to determine the extent of arteriovenous malformations and their relationship to normal structures
- MRA (contrast as requested) of the affected body part is considered medically necessary for evaluation.

CT with contrast and/or CTA (contrast as requested) of the affected body part is considered medically necessary when MRI and/or MRA is inconclusive or contraindicated.

Clinical changes and monitoring treatment

For changes in clinical status (symptoms or exam findings) related to the vascular tumor either of the following imaging is considered medically necessary:

- MRI without contrast or without and with contrast of the affected body to evaluate treatment response
- MRA (contrast as requested) of the affected body part

For preoperative planning both MRI and MRA (as requested) of the affected body part are considered medically necessary.

CT with contrast and/or CTA (contrast as requested) of the affected body part is considered medically necessary when MRI and/or MRA is inconclusive or contraindicated.

Surveillance imaging

- MRA (contrast as requested) of the affected body part is medically necessary for the surveillance of known vascular tumors.
- Imaging of vascular tumors with both MRI and MRA for routine treatment response or surveillance is not considered medically necessary.

- CT with contrast and/or CTA (contrast as requested) of the affected body part is considered medically necessary when MRI and/or MRA is inconclusive or contraindicated.

Evidence Discussion

Vascular tumors include a variety of benign, borderline, and malignant tumors, which have variable clinical courses, including but not limited to Infantile hemangiomas see **Infantile Hemangiomas (PEDPVD-5)**, Epithelioid hemangioma, Kaposiform hemangioendothelioma, Kaposi sarcoma, Epithelioid hemangioendothelioma, and Angiosarcoma of soft tissue.

References (PEDPVD-2)

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Vasculitis (PEDPVD-3)

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Large Vessel Vasculitis (PEDPVD-3.2)

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Imaging Indications

Initial imaging

- ANY of the following modalities is considered medically necessary for the initial 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)

Clinical changes and monitoring treatment

- In individuals being treated with systemic therapy, imaging (MRA or CTA or US) of the affected areas is medically necessary every 3 months to monitor active treatment for response.

Surveillance

- Imaging with MRA or CTA or US of the affected areas is medically necessary annually for surveillance of known involved body areas to detect progressive vascular damage that may require intervention.

Evidence Discussion

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

Systemic vasculitis is much less common in pediatric individuals than in adults. The diagnostic pathways and treatment options are similar for both age groups. For additional information on vasculitis see **Large Vessel Vasculitis (PVD-6.9)** in the general Peripheral Vascular Disease Imaging Guidelines.

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Medium Vessel Vasculitis (PEDPVD-3.3)

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Imaging Indications

- Some children who have had COVID 19 develop a severe inflammatory disease that can present in a similar way to Kawasaki disease or toxic shock syndrome. This syndrome has been defined by the US Centers for Disease Control and Prevention as multisystem inflammatory syndrome in children (MIS-C). See **Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDCD-12)** in the Pediatric Cardiac Imaging Guideline.
- Imaging guidelines for Kawasaki Disease- see **Kawasaki Disease (PEDCD-6)** in the Pediatric Cardiac Imaging Guideline.

Initial Imaging for Polyarteritis Nodosa

- For evaluation of polyarteritis nodosa:
  - ANY of the following modalities are considered medically necessary for the initial evaluation:
    - 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)

Clinical Changes and Monitoring Treatment for Polyarteritis Nodosa

- For evaluation of polyarteritis nodosa:
  - MRA or CTA or US of the affected body part is medically necessary every 3 months during active treatment with systemic therapy for treatment response.

Surveillance Imaging for Polyarteritis Nodosa

- MRA or CTA or US of the affected body part is medically necessary annually for surveillance of known involved body areas to detect progressive vascular damage that may require intervention.

Evidence Discussion

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

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Small Vessel Vasculitis (PEDPVD-3.4)

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Imaging Indications Small Vessel Vasculitis

- Advanced imaging is not sensitive enough to detect changes in small vessels and is **not** considered medically necessary for primary assessment of any small vessel vasculitis.
- End-organ damage occurs with several of the small vessel vasculitides. Advanced imaging is considered medically necessary for the following:
  - Henoch-Schönlein Purpura (HSP) is the most common vasculitis of childhood, mainly involving small blood vessels. Ultrasound abdomen (CPT® 76700) is commonly used to evaluate possible gastrointestinal complications (including bowel wall edema and hemorrhage, and intussusception) in known or suspected HSP, and should be approved when requested for that indication.
  - 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 considered medically necessary 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
  - Eosinophilic granulomatosis with polyangiitis (EGPA, formerly known as Churg-Strauss Syndrome):
    - CT Chest without contrast (CPT® 71250) or with contrast (CPT® 71260) is considered medically necessary 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 considered medically necessary in the following circumstances:
      - New or worsening clinical symptoms affecting the body area requested

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- To assess response to medical therapy when a change in treatment regimen is being considered
- Annually to evaluate the extent of disease

Evidence Discussion

Henoch-Schonlein Purpura (HSP) is the most common vasculitis of childhood, mainly involving small blood vessels. Imaging modalities such as ultrasound, CT, and Doppler ultrasound are used to evaluate gastrointestinal complications and monitor treatment response.

References (PEDPVD-3)

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Disorders of the Aorta and Visceral Arteries (PEDPVD-4)

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Thoracic Aortic Disease (PEDPVD-4.1)

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Imaging Indications for Thoracic Aortic Disease

Familial Aortopathies

- For aortopathies such as the following:
  - Marfan
  - Ehlers-Danlos (EDS)- a genetic mutation known to predispose to aortic aneurysms/ dissections (TGFBR1, TGFBR2, FBN1, ACTA2, or MYH11)
  - Loeys-Dietz
  - Familial thoracic aneurysm and dissections
- Screening: for Family history with first-degree relative of aortopathy
  - Asymptomatic individuals with no signs or symptoms of disease, whose first-degree relative has no definitive gene defect, can have screening echo (TTE) annually.
- Initial workup: Individuals with suspected aortopathies (gene positive, physical exam positive, or other findings) or definite disease associated with aortopathy
  - Echocardiogram (TTE) at the time of evaluation.
  - If the consideration is for Loeys-Dietz any of the following are considered medically necessary in addition to the TTE at the time of work up:
    - MRA or CTA Head
    - MRA or CTA Neck
    - MRA or CTA Chest
    - MRA or CTA Abdomen and Pelvis
    - MRA or CTA of area of concern when there is an incidental finding on other imaging
- Surveillance: Suspected or known disease but **normal** aortic imaging:
  - Individuals with suspected genetic aortopathies but no disease can have an echocardiogram to assess for change:
    - At 6 months
    - Then annually
  - Individuals with Loeys-Dietz can be imaged with any of the following:
    - Echocardiogram
    - MRA or CTA of (any or all):
      - Head

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        - Neck
        - Chest
        - Abdomen
        - Pelvis
    - Individuals with Loeys-Dietz can be imaged with the above at the following intervals:
        - At 6 months
        - Then annually
- Surveillance: Suspected disease and **previous abnormal** imaging
    - Individuals with abnormal thoracic imaging can be imaged with (both):
        - Echocardiogram
        - CTA or MRA of (any):
            - Chest
            - Abdomen
            - Pelvis
            - Head (Loeys-Dietz)
            - Neck (Loeys-Dietz)
    - The above imaging is considered medically necessary as follows:
        - At the time of diagnosis
        - In 6 months after diagnosis (if older than 2 years)
        - Then as follows based on the individual's age:
            - Individual's age 0 to 2 years:
                - Every 3 months
            - Individual's age 3 to 12 years:
                - Every 6 months
            - Individual's age 13 years and older:
                - Every 12 months (if <4.5 or < 0.5 cm growth per year)
                - Every 6 months if ≥4.5 or ≥0.5 cm growth per year, or any Loeys-Dietz patient)
    - If the diameter z score is increased, then a repeat study can be done prior to the next allowed study, to assess for rate of change
- If there are symptoms of dissection, any or all of the following are considered medically necessary:
    - Echo
    - CTA or MRA of (any or all):
        - Chest
        - Abdomen
        - Pelvis

- For pediatric individual with dissection, imaging per vascular surgery and cardiology or any provider in consultation with vascular surgery at **any** interval.
- Miscellaneous syndromes with potential aortopathy as major feature of congenital heart disease
  - Individuals with Turner syndrome see section **Aortic disease in Turner Syndrome (CD-11.2.10)** in the Cardiac Imaging Guideline
  - Williams syndrome See section **LVOT lesions (PEDCD-2.4.10)** in the Pediatric Cardiology Imaging Guideline
  - Individuals with congenital heart disease would be managed based on **Imaging and Surveillance per Congenital lesion (PEDCD-2.4)** in the Pediatric Cardiology Imaging Guideline
- Miscellaneous disorders that can affect the aorta such as osteogenesis imperfecta, homocystinuria, polycystic kidney disease, pseudoxanthoma elasticum, and Hurler syndrome.
  - Screening echocardiogram yearly.
  - If positive findings, follow protocol for aortic root dilatation.
- Follow-up of thoracic aortic abnormalities related to the following conditions are addressed in other imaging guidelines:
  - Coarctation of the Aorta- See **Aortic Coarctation and IAA (interrupted aortic arch) (PEDCD-2.4.11)** in the Pediatric Cardiac Imaging Guidelines
  - Congenital rubella syndrome- See **Imaging and Surveillance per Congenital lesion (PEDCD-2.4)** in the Pediatric Cardiac Imaging Guidelines
  - Kawasaki Syndrome- See **Kawasaki Disease (PEDCD-6)**
  - Neurofibromatosis- See **General Guidelines (PEDCD-1.0)** in the Pediatric Cardiac Imaging Guidelines

Evidence Discussion

Familial aortopathies such as Marfan syndrome, Ehlers-Danlos syndrome, Loeys-Dietz syndrome, and familial thoracic aneurysm and dissections are included. Imaging modalities such as echocardiogram (TTE), magnetic resonance angiography (MRA), and computed tomography angiography (CTA) are used to evaluate the extent of aortic involvement and monitor disease progression.

Aortic Congenital Vascular Malformations (PEDPVD-4.2)

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Imaging Indications for Aortic Congenital Vascular Malformations

Imaging for Aortic Congenital Vascular Malformation

Description	CPT®
CT Chest with contrast	71260
CTA Chest	71275
MRA Chest	71555
Cardiac MRI without contrast	75557
Cardiac MRI without and with contrast	75561

Any of the imaging in the **above table** is medically necessary for the initial diagnosis of Congenital Aortic Vascular Malformation.

Vascular rings may impact both the esophagus and trachea. CTA or MRA of the Neck is considered medically necessary to evaluate the relationship of the aortic arch to the trachea and esophagus for surgical planning.

Evidence Discussion

Congenital Aortic Vascular Malformations can be asymptomatic or can present with impact on breathing or swallowing when there is associated compression from vascular rings. Vascular rings may impact both the esophagus and trachea. Imaging modalities such as cardiac MRI, MRA, CT, and CTA are used to evaluate these malformations and their impact on surrounding structures.[5]

[5] Priya S, Thomas R, Nagpal P, et al. Congenital anomalies of the aortic arch. *Cardiovasc Diagn Ther*. 2018;8(Suppl 1):S26-S44. doi:10.21037/cdt.2017.10.15.

Visceral Artery Aneurysms (PEDPVD-4.3)

PVDP.AD.0004.3.A
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Imaging Indications for Visceral Artery Aneurysms

- Visceral artery imaging indications in pediatric individuals are identical to those for adult individuals. See **Visceral Artery Aneurysm (PVD-6.5)** in the General Peripheral Vascular Disease Imaging Guidelines.

Treatment is generally indicated for visceral aneurysms ≥2cm. Imaging of visceral artery aneurysms for initial evaluation or for treatment planning is medically necessary as follows:

- Initial imaging of calcifications seen on plain film imaging suspicious for visceral artery aneurysms (spleen, kidney, liver or intestines) with ultrasound (CPT® 76700, 76705, 93975, 93976, 93978, or 93979), **or** CTA Abdomen (CPT® 74175), **or** CT Abdomen with contrast (CPT® 74160)
- MRA Abdomen (CPT® 74185) without contrast in place of CTA or CT Abdomen in pediatric individuals or when there is a contraindication to contrast materials (i.e., renal insufficiency, contrast allergy, pregnancy)
- Ultrasound (CPT® 76700,76705, 93975, 93976, 93978, or 93979) **or** CTA Abdomen (CPT® 74175) **or** CT Abdomen with contrast (CPT® 74160) is considered medically necessary for further monitoring based on the intervals below or as determined by a vascular specialist or any provider in consultation with a vascular specialist:
  - Splenic artery aneurysms:
    - <20mm can be imaged every three years
    - 20mm to 29mm can be imaged annually
    - If ≥30mm, they should be referred for treatment, either stent, excision or splenectomy
  - For all other visceral artery aneurysms:
    - Initial evaluation with six-month follow-up for one year
    - Further follow-up annually if no significant enlargement is seen
- CTA Abdomen (CPT® 74175), MRA Abdomen (CPT® 74185), or CT Abdomen with contrast (CPT® 74160) is considered medically necessary following stent placement at:
  - 1 month
  - 6 months
  - 12 months
  - Then every year

Evidence Discussion

Visceral artery aneurysms are rare but can be life-threatening if ruptured. Imaging indications in pediatric individuals are identical to those for adult individuals, with modalities such as MRA, CTA, and ultrasound being used for evaluation. Aneurysmal disease, besides primarily involving the large vessels, can also affect medium and smaller sized vessels. Visceral cases are uncommon and occasionally are associated with certain connective tissue and genetic disorders. They are often found incidentally on imaging. Ultrasound, CT, or CTA imaging may be indicated for surveillance in these cases. Due to the anatomic location of the visceral vessels, duplex ultrasound may have technical limitations. Consideration of best surveillance study should be decided on initial imaging and whether a certain modality is felt to provide diagnostic information. Example: some splenic arterial aneurysms may be diagnostic with US, but others may be obscured by bowel gas requiring CT/CTA.

References (PEDPVD-4)

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Infantile Hemangiomas (PEDPVD-5)

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General Considerations (PEDPVD-5.1)

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Most infantile hemangiomas do not require any imaging. Ultrasound with Doppler can be used when the diagnosis is uncertain, or with high-risk clinical considerations. Other general imaging considerations for other vascular neoplasms regarding MRI, MRA, CT, and CTA also apply to infantile hemangiomas. See **Imaging vascular tumors**.

- Multiple (5 or more) infantile hemangiomas can be associated with hepatic hemangiomas with risk potential for high-output cardiac failure and other risks see **Multiple Infantile Hemangiomas (PEDPVD-5.2)**.
- High-output cardiac failure can also be caused rarely by large cutaneous infantile hemangiomas. Affected infants may present with “failure-to-thrive”, a hyperdynamic precordium, tachycardia, bounding pulses with a widened pulse pressure, and a palpable thrill and/or audible bruit over the hemangioma. This is an indication for cardiac evaluation, including echocardiography (CPT® 93303 ordered with CPT® 93320 and CPT® 93325).
- Life threatening risk of airway obstruction is associated with infantile hemangiomas of the lower face (“beard distribution”), or of the anterior neck, or of the oral and/or pharyngeal mucosa.
- Location-associated functional impairment can be found with periocular infantile hemangiomas larger than 1 cm (impairing vision), or infantile hemangiomas involving lip(s) or oral cavity (impairing feeding).
- Ulceration can occur with profuse bleeding that can be life threatening.
- Disfigurement risk is increased with large (5 cm or larger) infantile hemangiomas, facial or scalp infantile hemangiomas, and breast infantile hemangiomas in female infants.
- An infantile hemangioma at least 2.5 cm in diameter overlying the lumbar spine or sacrum is an indication to do a spinal ultrasound (under 6 months of age) and/or MRI Lumbar Spine without contrast (CPT® 72148) or MRI Lumbar Spine without and with contrast (CPT® 72158).
- Infantile hemangiomas 5 cm or larger in size have an increased risk of extracutaneous structural abnormalities.
- Other high-risk indications include Syndromes or Associations with extracutaneous structural changes: for “PHACE(S) syndrome” See **Imaging Indications for PHACE(s) Syndrome**, and for “LUMBAR syndrome” See **Imaging Indications for LUMBAR Syndrome**.

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Evidence Discussion

Infantile Hemangiomas are the most common benign tumor of childhood, occurring in close to 5% of infants. Infantile Hemangiomas typically have a phase of rapid and significant growth between 1 month and 3 months of age; growth is usually completed by 5 months of age. Gradual involution then occurs, completed in 90% by age of 4 years but with residual skin changes frequently persisting. Though usually not needed for diagnosis, biopsy can be done when needed to identify unique markers not found on other vascular tumors.

When treatment is needed, imaging may be used to monitor response; consultation with a hemangioma specialist may be useful in guiding evaluation, treatment, and follow-up. The 2019 Clinical Practice Guideline of the American Academy of Pediatrics states "Unlike many diseases, management of IHs is not limited to 1 medical or surgical specialty. A hemangioma specialist may have expertise in dermatology, hematology-oncology, pediatrics, facial plastic and reconstructive surgery, ophthalmology, otolaryngology, pediatric surgery, and/or plastic surgery, and his or her practice is often focused primarily or exclusively on the pediatric age group."

Multiple Infantile Hemangiomas (PEDPVD-5.2)

PVDP.IH.0005.2.A
v2.0.2026

- Multiple (5 or more) hemangiomas is an indication for Ultrasound with Doppler exam of the liver (CPT® 76700):
  - Initial imaging to look for hepatic hemangiomas
  - Repeat doppler ultrasound abdomen:
    - Monitor hepatic hemangiomas for progression
    - Monitor response to treatment.

Evidence Discussion

Multiple (5 or more) hemangiomas- though hepatic hemangiomas can be asymptomatic, they rarely can cause a high flow rate that can cause high-output cardiac failure and can be potentially fatal. "Diffuse" hepatic infantile hemangiomas are a rare subset of hepatic hemangiomas at high-risk for morbidity and mortality; affected infants usually present before 4 months of age with severe hepatomegaly, which can lead to lethal abdominal compartment syndrome with compromised ventilation, renal failure caused by renal vein compression, or compromise of inferior vena cava blood flow to the heart. Hepatic hemangiomas can also inactivate (via deiodination) thyroid hormones, causing risk of severe hypothyroidism.

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PHACE(S) Syndrome (PEDPVD-5.3)

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Imaging Indications for PHACE(s) Syndrome

Initial Imaging

- Initial diagnostic imaging is medically necessary when PHACE(S) syndrome is suspected by clinical findings including **any** of the following:
  - Infantile hemangioma ≥5 cm diameter on the face, scalp, and/or neck.
  - Infantile hemangioma <5cm on face, scalp, or neck in the setting of one or more major anomalies associated with PHACE(S) syndrome (i.e., coarctation of the aorta or midline ventral defect).
  - Infantile hemangioma on upper chest or proximal upper extremity ≥5 cm with no visible facial infantile hemangioma with PHACE(S) syndrome in the setting of one or more major anomalies associated with PHACE(S) syndrome (i.e., coarctation of the aorta or midline ventral defect).
  - Large intraorbital infantile hemangioma.
- Initial imaging for suspected PHACE(S) syndrome includes **any** of the following:
  - MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553)
  - MRI Orbits without contrast (CPT® 70540) or MRI Orbits without and with contrast (CPT® 70543)
  - MRA Head without contrast (CPT® 70544) or MRA Head without and with contrast, (CPT® 70546)
  - MRA Neck may be done either without contrast (CPT® 70547), with contrast (CPT® 70548), or without and with contrast (CPT® 70549)
  - MRA Chest (CPT® 71555)
  - Transthoracic echocardiogram (CPT® 93303 with CPT® 93320 and CPT® 93325)
    - Cardiac MRI (CPT® 75557 or CPT® 75561) is medically necessary if abnormalities are identified on echocardiogram.
  - MRI Chest without contrast (CPT® 71550) or MRI Chest without and with contrast (CPT® 71552) is medically necessary if other clinical information or imaging shows involvement of the aorta.

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Surveillance

Repeat imaging is medically necessary when results will impact clinical management of the individual based on the results of the initial clinical and imaging assessment and any subsequent clinical changes, or when high-risk findings have been identified by clinical evaluation such as any of the following:

- Evidence of past arterial stroke
- Arterial stenosis or occlusions, with or without moyamoya-like vascular changes
- Structural brain changes, with neurosurgical evaluation clarifying the need for follow-up.
- Changes in the aortic arch, coarctation of the aorta, and congenital cardiac anomalies, with pediatric cardiology evaluation clarifying the need for follow-up see **Imaging and Surveillance per Congenital Lesion (PEDCD-2.4)** in the Pediatric Cardiac Imaging Guidelines

Evidence Discussion

"PHACE" (Posterior fossa malformations, Hemangiomas, Arterial anomalies, Coarctation of the aorta and Cardiac defects, and Eye abnormalities) syndrome or association (or "PHACE(S)" syndrome when also associated with sternal cleft and/or supraumbilical raphe) is frequently suspected when an infant has a large (5 cm in diameter or larger) infantile hemangioma of the face, scalp, or neck (risk of PHACE(S) Syndrome is then approximately 30%).

In rare cases, the face or scalp is not involved, with a large infantile hemangioma located on the torso and/or upper extremity instead. Cerebrovascular anomalies, present in more than 90% of individuals with PHACE(S) syndrome, are the most common extracutaneous feature of the syndrome, followed by cardiac anomalies (67%) and structural brain anomalies (about 50%).

LUMBAR Syndrome (PEDPVD-5.4)

PVDP.IH.0005.4.A
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Imaging Indications for LUMBAR Syndrome

- "LUMBAR syndrome" is reasonably suspected in a child with a large (5 or more cm in diameter) infantile hemangioma of any lumbosacral or perineal region or lower extremity. The following imaging is considered medically necessary:
  - Ultrasound spine (CPT® 76800) in infants up to 6 months of age, abdomen (CPT® 76700), and pelvis (CPT® 76856), with color Doppler.
  - MRI Lumbar Spine without contrast (CPT® 72148) or without and with contrast (CPT® 72158) at 3 to 6 months of age, or earlier when either findings on an Ultrasound exam are inadequate or when requested by a hemangioma specialist or any provider in consultation with a hemangioma specialist.
  - MRI of other relevant spinal level (relevance based on proximity of observed infantile hemangiomas larger than 5 cm) without contrast or MRI of the relevant spinal level without and with contrast.
  - When ultrasound findings are inadequate and/or when recommended by a hemangioma specialist or any provider in consultation with a hemangioma specialist:
    - MRI Pelvis without contrast (CPT® 72195) or without and with contrast (CPT® 72197) **and/or**
    - MRI Abdomen without contrast (CPT® 74181) or without and with contrast (CPT® 74183).
  - MRA Abdomen CPT® 74185 and/or Pelvis CPT® 72198, is considered medically necessary based on proximity of infantile hemangioma(s) at least 5 cm in diameter and/or other clinical evidence of vascular involvement, and/or when recommended by a hemangioma specialist or any provider in consultation with a hemangioma specialist.
  - Infantile hemangioma of the lower extremity that is at least 5 cm in diameter is an indication for MRI of the relevant portion of the lower extremity without contrast (CPT® 73718) or lower extremity without and with contrast (CPT® 73720) and/or lower extremity joint without contrast (CPT® 73721) or lower extremity joint without and with contrast (CPT® 73723).
  - When there is extensive lower extremity involvement with infantile hemangiomas the following are all considered medically necessary:
    - MRA (for both arterial and venous phase imaging) Abdomen
    - MRA Pelvis

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- MRA Lower extremities
- Note: this should be reported as CPT® 74185 and CPT® 73725; the CPT® code for MRA Pelvis (CPT® 72198) should not be included in this circumstance.

Evidence Discussion

The acronym "LUMBAR syndrome" refers to the association of lower body infantile hemangiomas of at least 5 cm in size (and other cutaneous defects), urogenital anomalies and ulceration, myelopathy (lipomyelocele/lipo-myelomeningocele and/or tethered spinal cord), bony deformities, anorectal malformations and arterial anomalies, and renal anomalies. Though not exclusively true, there is a general regional correlation between the location of the cutaneous large infantile hemangioma(s) with underlying structural anomalies.

References (PEDPVD-5)

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