CLINICAL GUIDELINES

Pediatric PVD Imaging Policy

Version 20.0.2018

Effective May 17, 2018

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: General Guidelines

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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, minor 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

- 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-1.3: 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 for image acquisition and the need for, the patient to lie still, 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 minimizing anesthesia exposure adhering to the following considerations:
    - MRI should always be performed without and with contrast unless there is a specific contraindication to gadolinium use since the patient already has intravenous access for anesthesia.
      - Recent evidence based literature demonstrates the potential for gadolinium deposition in various organs including the brain after the use of MRI contrast.

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        - 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 requesting clinicians indicate that a non-contrast study is being requested with specific concern for gadolinium retention, the exam can be approved
    - 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.

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- 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® 78805, CPT® 78806, or CPT® 78807 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.

References

1. Muratore F, Pipitone N, Salvarani C, et al. Imaging of vasculitis: state of the art. Best Practice & Research Clinical Rheumatology. 2016;30:688-706
2. ACR–SPR Practice parameter for the performance and interpretation of pediatric magnetic resonance imaging (MRI) Amended 2014, available at: https://www.acr.org/~/media/ACR/Documents/PGTS/guidelines/MRI_Pediatric.pdf.
3. ACR–ASER–SCBT-MR–SPR Practice parameter for the performance of pediatric computed tomography (CT) Revised 2014, available at: https://www.acr.org/~/media/ACR/Documents/PGTS/guidelines/CT_Pediatric.pdf.
4. Ing C, DiMaggio C, Whitehouse A, et al. Long-term differences in language and cognitive function after childhood exposure to anesthesia. *Pediatrics.* 2012 Sep;130(3):e476-e485. Accessed October 23, 2017. http://pediatrics.aappublications.org/content/130/3/e476.
5. Monteleone M, Khandji A, Cappell J, et al. Anesthesia in children: perspectives from nonsurgical pediatric specialists. *J Neurosurg Anesthesiol*. 2014;26(4):396-398. Accessed October 23, 2017. https://www.medscape.com/medline/abstract/25191959.
6. DiMaggio C, Sun LS, and Li G. Early childhood exposure to anesthesia and risk of developmental and behavioral disorders in a sibling birth cohort. *Anesth Analg.* 2011 Nov;113(5):1143-1151. http://journals.lww.com/anesthesia-analgesia/Fulltext/2011/11000/Early_Childhood_Exposure_to_Anesthesia_and_Risk_of.31.aspx.
7. MacDonald A and Burrell S. Infrequently performed studies in nuclear medicine: part 2*. *J Nucl Med Technol*. 2009 Mar;37:1-13. Accessed October 23, 2017. http://interactive.snm.org/docs/Infrequently%20Performed%20Studies%20Part%202.pdf.
8. McNeill GC, Witte MH, Witte CL, et al. Whole-body lymphangioscintigraphy: preferred method for initial assessment of the peripheral lymphatic system. *Radiology*. 1989 Aug;172(2):495-502. Accessed October 23, 2017. http://pubs.rsna.org/doi/citedby/10.1148/radiology.172.2.2748831.
9. Palestro CJ, Brown ML, Forstrom LA, et al. Society of nuclear medicine procedure guideline for 111In-Leukocyte Scintigraphy for suspected infection inflammation. Version 3.0, approved June 2, 2004: Accessed October 23, 2017. http://interactive.snm.org/docs/Leukocyte_v3.pdf.
10. De Vries EFJ, Roca M, Jamar F, et al. Guidelines for the labelling of leucocytes with ^{99m}Tc-HMPAO. *Eur J Nucl Med Mol Imaging*. 2010 Apr;37(4):842-848. Accessed October 23, 2017. https://link.springer.com/article/10.1007/s00259-010-1394-4.
11. Fraum TJ, Ludwig DR, Bashir MR, et al. Gadolinium-based contrast agents: a comprehensive risk assessment. *J Magn Reson Imaging*. 2017;46(2): 338–353. Accessed October 23, 2017. https://www.medscape.com/medline/abstract/28083913.
12. FDA Medical Imaging Drug Advisory Committee meeting 9/8/17 Minutes available at: https://www.fda.gov/downloads/AdvisoryCommittees/CommitteesMeetingMaterials/Drugs/MedicalImagingDrugsAdvisoryCommittee/UCM574746.pdf.

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13. FDA Drug Safety Communication: FDA identifies no harmful effects to date with brain retention of gadolinium-based contrast agents for MRIs; review to continue. FDA Drug Safety Communication. May 22, 2017. https://www.fda.gov/Drugs/DrugSafety/ucm559007.htm.

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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.

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- 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: 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 imaging because the diagnosis is made clinically. However, MR imaging may be required to evaluate an 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.

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- 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 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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References

1. Restrepo R. Multimodality imaging of vascular anomalies. *Pediatr Radiol*. 2013;43 (suppl): S141-s154.
2. Krishnamurthy R, Daldrup-Link HE, Jones JY, et al. Imaging studies in the diagnosis and management of pediatric malignancies. *Principles and Practice of Pediatric Oncology*. eds. Pizzo PA & Poplack DG. 2017;7:185-234.
3. Martin KL. Vascular Disorders. *Nelson Textbook of Pediatrics, Chapter 650.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 3122-3128.
4. Marler JJ, and Mulliken JB. Current management of hemangiomas and vascular malformations. *Clin Plast Surg*. Jan 2005;32(1):99-116. Accessed October 23, 2017. http://europepmc.org/abstract/MED/15636768.
5. Armstrong DC, and terBrugge K. Selected interventional procedures for pediatric head and neck vascular lesions. *Neuroimaging Clin N Am*. Feb 2000;1091):271-92. Accessed October. https://www.ncbi.nlm.nih.gov/pubmed/10658165.
6. Blei F, and Guarini A. Current workup and therapy of infantile hemangiomas. *Clinics in Dermatology*. 2014 July-Aug;32(4):459-470. Accessed October 23, 2017. http://www.cidjournal.com/article/S0738-081X(14)00035-2/fulltext.
7. Cahill AM and Els LFN. Pediatric vascular malformations: pathophysiology, diagnosis, and the role of interventional radiology. *Cardiovasc Intervent Radiol*. 2011 Aug;34(4):691-704. Accessed October 23, 2017. https://link.springer.com/article/10.1007/s00270-011-0123-0?no-access=true.
8. Bagrodia N, Defnet AM, and Kandel JJ. Management of lymphatic malformations in children. *Curr Opin Pediatr.* 2015 June;27(3):356-363. Accessed October 23, 2017. https://www.medscape.com/medline/abstract/25888145.
9. Wassef M, Beli F, Adams D, et al, Vascular anomalies classification: Recommendations from the International Society for the Study of Vascular Anomalies. *Pediatrics.* 2015 July;136(1):e203-e214. Accessed October 23, 2017. http://pediatrics.aappublications.org/content/136/1/e203.
10. Kutz AM, Aranibar L, Lobos N, et al. Color Doppler ultrasound follow-up of infantile hemangiomas and peripheral vascularity in patients treated with propranolol. *Pediatr Dermatol.* 2015 July-Aug;32(4):468-475. Accessed October 23, 2017. http://onlinelibrary.wiley.com/doi/10.1111/pde.12596/abstract;jsessionid=5FAF75A71163C41281F00EE7FB4FC09F.f03t01.
11. Adams DM, Trenor CC, Hammill AM, et al. Efficacy and safety of sirolimus in the treatment of complicated vascular anomalies. *Pediatrics*. 2016 Feb;137:e20153257. Accessed October. http://pediatrics.aappublications.org/content/pediatrics/137/2/e20153257.full.pdf.

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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 are 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)
  - Patients 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:
  - 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)
- Patients 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.

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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:
  - 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)
  - Eosinophilic granulomatosis with polyangiitis (EGPA, formerly known as Churg-Strauss Syndrome)
    - CT Chest without contrast (CPT®71250) or with contrast (CPT®71260)
  - Immune complex associated small-vessel vasculitis [immunoglobulin A–associated vasculitis (IgAV)]
    - Doppler ultrasound of the affected body part (most commonly abdomen)
  - 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

1. Muratore F, Pipitone N, Salvarani C, et al. Imaging of vasculitis: state of the art. Best Practice & Research Clinical Rheumatology 2016;30:688-706
2. Lensen KD, Comans EF, Voskuyl AE, et al. Large-vessel vasculitis: interobserver agreement and diagnostic accuracy of 18F-FDG-PET/CT. *Biomed Res Int*. 2015;2015:914692.
3. Soussan M, Nicolas P, Schramm C, et al. Management of large-vessel vasculitis with FDG-PET: a systematic literature review and meta-analysis. Medicine (Baltimore) 2015;94:e622.
4. Besson FL, Parienti JJ, Bienvenu B, et al. Diagnostic performance of (1)(8)F-fluorodeoxyglucose positron emission tomography in giant cell arteritis: a systematic review and meta-analysis. *Eur J Nucl Med Mol Imaging*. 2011;38: 1764e72.
5. Ardoin SP and Fels E. Vasculitis syndromes. *Nelson Textbook of Pediatrics*, chapter 167. eds Kliegman RM, Stanton BF, St. Geme JW III, Schor NF. 20th edition 2016, pp 1215-1224.
6. Soliman M, Laxer R, Manson D, et al. Imaging of systemic vasculitis in childhood. *Pediatr Radiol*. 2015 July;45(8):1110-1125. Accessed October 23, 2017. https://www.medscape.com/medline/abstract/26198677.
7. Sharma AM, Singh S, and Lewis JE. Diagnostic approach in patients with suspected vasculitis. *Tech Vasc Interv Radiol.* 2014 Dec; 17(4):226-233. Accessed October 23, 2017. http://www.techvir.com/article/S1089-2516(14)00058-4/fulltext.
8. Ammirati E, Moroni F, Pedrotti P, et al. Non-invasive imaging of vascular inflammation. *Front Immunol.* 2014 Aug;5:1-15. Accessed October 23, 2017. https://www.frontiersin.org/articles/10.3389/fimmu.2014.00399/full.
9. Granata C, Damasio MB, Zaottini F, et al. Imaging of childhood vasculitis. *Radiologic clinics of North America*. 2017 Sep;55(5):1131. Accessed October 23, 2017. http://www.radiologic.theclinics.com/article/S0033-8389(17)30075-1/fulltext.

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PEDPVD-4: Disorders of the Aorta and Visceral Arteries

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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 patients with Loeys-Dietz syndrome, Marfan syndrome, coarctation of the aorta, Takayasu arteritis, neurofibromatosis, William syndrome, Ehler Danlos syndrome, 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: 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.

References

1. Hanneman K, Newman B, and Chan F. Congenital variants and anomalies of the aortic arch. RadioGraphics. 2017;37:32-51
2. Erbel R, Aboyans V, Boileau C, et al. 2014 ESC Guidelines on the diagnosis and treatment of aortic diseases: Document covering acute and chronic aortic diseases of the thoracic and abdominal aorta of the adult. The Task Force for the Diagnosis and Treatment of Aortic Diseases of the European Society of Cardiology (ESC). European Heart Journal. 2014 Nov;35(41):2873–2926
3. Chan, Francis P, et al. “Acquired diseases of the great vessels (Chapter 83)” in Caffey’s Pediatric Diagnostic Imaging. 12th edition Brian Coley editor, Elsevier Saunders, Philadelphia PA, 2013.
4. Chan, Francis P. et al “Congenital diseases of the thoracic great arteries (Chapter 77)” in Caffey’s Pediatric Diagnostic Imaging. 12th edition Brian Coley editor, Elsevier Saunders, Philadelphia PA, 2013

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