Cigna Medical Coverage Policies – Radiology
Pediatric Peripheral Vascular Disease (PVD) Imaging Guidelines

Effective April 01, 2023

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.

CPT® (Current Procedural Terminology) is a registered trademark of the American Medical Association (AMA). CPT® five digit codes, nomenclature and other data are copyright 2022 American Medical Association. All Rights Reserved. No fee schedules, basic units, relative values or related listings are included in the CPT® book. AMA does not directly or indirectly practice medicine or dispense medical services. AMA assumes no liability for the data contained herein or not contained herein.

Table of Contents

Guideline Page

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

General Guidelines (PEDPVD-1)

Procedure Codes Associated With PVD Imaging (PEDPVD)

PVDP.GG.0001.C
v1.0.2023

MRA	**CPT ®**
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	**CPT ®**
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
Computed tomographic angiography, chest (noncoronary), with contrast material(s), including noncontrast images,	71275

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

if performed, and image postprocessing	
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	**CPT ®**
Ultrasound, abdominal, real time with image documentation; complete	76700
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	93931

© 2023 eviCore healthcare. All Rights Reserved.

arterial bypass grafts; unilateral or limited	
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

© 2023 eviCore healthcare. All Rights Reserved.

General Guidelines (PEDPVD-1.0)

PVDP.GG.0001.0.A
v1.0.2023

- 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 (CT, MR, Nuclear Medicine), 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.
- 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 the individual's management or treatment decisions.

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Age Considerations (PEDPVD-1.1)

PVDP.GG.0001.1.A

v1.0.2023

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

- Individuals who are 18 years old and younger[14] 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.

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Modality General Considerations (PEDPVD-1.3)

PVDP.GG.0001.3.C
v1.0.2023

- 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 may be appropriate if requested. By doing so, the requesting provider may avoid repetitive anesthesia administration to perform an MRI with contrast if the initial study without contrast is inconclusive.
      - Recent evidence-based literature demonstrates the potential for gadolinium deposition in various organs including the brain after the use of MRI contrast.
      - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
    - If multiple body areas are supported by eviCore guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session.
  - The presence of surgical hardware or implanted devices may preclude MRI.
  - The selection of best examination may require coordination between the provider and the imaging service.
- CT

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

- 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'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 appropriate and warranted for specific clinical situations.

© 2023 eviCore healthcare. All Rights Reserved.

References (PEDPVD-1)

v1.0.2023

1. Muratore F, Pipitone N, Salvarani C, Schmidt WA. Imaging of vasculitis: State of the art. *Best Practice & Research Clinical Rheumatology*. 2016;30(4):688-706. doi:10.1016/j.berh.2016.09.010.

2. American College of Radiology. Practice parameter for performing and interpreting magnetic resonance imaging (MRI): Amended 2018 (Resolution 44). ACR.org. Published October 1, 2018. https://www.acr.org/-/media/ACR/Files/Practice-Parameters/mr-perf-interpret.pdf?la=en.

3. Faerber EN, Abramson SJ, Benator RM, et. al. Practice Parameters by Modality: ACR–ASER–SCBT-MR–SPR Practice parameter for the performance of pediatric computed tomography (CT). *American College of Radiology | American College of Radiology*. Published 2017. https://www.acr.org/Clinical-Resources/Practice-Parameters-and-Technical-Standards/Practice-Parameters-by-Modality.

4. Ing C, Dimaggio C, Whitehouse A, et al. Long-term Differences in Language and Cognitive Function After Childhood Exposure to Anesthesia. *Pediatrics*. 2012;130(3):e476-e485. doi:10.1542/peds.2011-3822.

5. Monteleone M, Khandji A, Cappell J, Lai WW, Biagas K, Schleien C. Anesthesia in Children. *Journal of Neurosurgical Anesthesiology*. 2014;26(4):396-398. doi:10.1097/ana.0000000000000124.

6. Dimaggio C, Sun LS, Li G. Early Childhood Exposure to Anesthesia and Risk of Developmental and Behavioral Disorders in a Sibling Birth Cohort. *Anesthesia & Analgesia*. 2011;113(5):1143-1151. doi:10.1213/ane.0b013e3182147f42.

7. Macdonald A, Burrell S. Infrequently Performed Studies in Nuclear Medicine: Part 2. *Journal of Nuclear Medicine Technology*. 2009;37(1):1-13. doi:10.2967/jnmt.108.057851.

8. Mcneill GC, Witte MH, Witte CL, et al. Whole-body lymphangioscintigraphy: preferred method for initial assessment of the peripheral lymphatic system. *Radiology*. 1989;172(2):495-502. doi:10.1148/radiology.172.2.2748831.

9. Palestro CJ, Brown ML, Forstrom LA, et. al. SNMMI Procedure Standard for 111In-Leukocyte Scintigraphy for Suspected Infection/Inflammation 3.0. SNMMI. http://www.snmmi.org/ClinicalPractice/content.aspx?ItemNumber=6414. Published June 2, 2004.

© 2023 eviCore healthcare. All Rights Reserved.

10. De Vries EFJ, Roca M, Jamar F, Israel O, Signore A. Guidelines for the labelling of leucocytes with 99mTc-HMPAO. *European Journal of Nuclear Medicine and Molecular Imaging*. 2010;37(4):842-848. doi:10.1007/s00259-010-1394-4.

11. Fraum TJ, Ludwig DR, Bashir MR, Fowler KJ. Gadolinium-based contrast agents: A comprehensive risk assessment. *Journal of Magnetic Resonance Imaging*. 2017;46(2):338-353. doi:10.1002/jmri.25625.

12. Center for Drug Evaluation and Research. Medical Imaging Drugs Advisory Committee. U.S. Food and Drug Administration. Published September 8, 2017. https://www.fda.gov/advisory-committees/human-drug-advisory-committees/medical-imaging-drugs-advisory-committee.

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.

14. Implementation Guide: Medicaid State Plan Eligibility Eligibility Groups Mandatory Coverage Infants and Children under Age 19 at https://www.hhs.gov/guidance/document/implementation-guide-medicaid-state-plan-eligibility-eligibility-groups-aeu-mandatory-2.

References (PEDPVD-1)

© 2023 eviCore healthcare. All Rights Reserved.

Vascular Anomalies (PEDPVD-2)

General Information (PEDPVD-2.1)

PVDP.AN.0002.1.A
v1.0.2023

- 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.

Background and Supporting 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).

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Lymphatic Malformations (PEDPVD-2.2)

PVDP.AN.0002.2.A
v1.0.2023

- 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 is indicated for lesions with acute enlargement and concerns for compression when MRI is contraindicated.

Background and Supporting Information

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.

© 2023 eviCore healthcare. All Rights Reserved.

Venous Malformations (PEDPVD-2.3)

PVDP.AN.0002.3.A
v1.0.2023

- 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 is indicated for venous malformations for preoperative assessment to evaluate the extent of malformation and their relationship to normal structures.
- MRA or CTA has a limited role in evaluating most venous malformations, but may be indicated (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 indicated when MRI is inconclusive or contraindicated
  - Both Klippel-Trénaunay syndrome and CLOVES syndrome have been found to have increased risk of venous thrombosis and pulmonary embolism, particularly after surgery or sclerotherapy. When pulmonary embolism is suspected in such individuals, CT Chest with contrast with PE protocol (CPT® 71260) or CTA Chest (CPT® 71275) is indicated.

Background and Supporting Information

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.

© 2023 eviCore healthcare. All Rights Reserved.

Capillary Malformations (PEDPVD-2.4)

PVDP.AN.0002.4.A
v1.0.2023

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

Background and Supporting Information

Capillary malformations also known as port wine stains are characterized by a collection of small vascular channels in the dermis and generally do not require advanced imaging because the diagnosis is made clinically.

© 2023 eviCore healthcare. All Rights Reserved.

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

PVDP.AN.0002.5.A
v1.0.2023

- 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 is indicated 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 indicated 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 indicated when MRI and/or MRA is inconclusive or contraindicated.

Background and Supporting Information

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.

© 2023 eviCore healthcare. All Rights Reserved.

Vascular Tumors (PEDPVD-2.6)

PVDP.AN.0002.6.A
v1.0.2023

- 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 is indicated 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 indicated 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 is indicated when MRI and/or MRA is inconclusive or contraindicated.

Background and Supporting Information

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.

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

References (PEDPVD-2)

v1.0.2023

1. Pizzo PA, Poplack DG, Krishnamurthy R, Daldrup-Link HE, Jones JY, et. al. Imaging studies in the diagnosis and management of pediatric malignancies. In: Principles and Practice of Pediatric Oncology. Vol 7. Philadelphia: Wolters Kluwer; 2016:185-234.

2. Martin KL. Vascular disorders. *Nelson Textbook of Pediatrics*, Chapter 669. eds Kliegman R, St. Geme JW III, Blum NJ, et al. 21st ed. Philadelphia, PA: Elsevier; 2020:3461-3469.

3. Blei F, Guarini A. Current workup and therapy of infantile hemangiomas. *Clinics in Dermatology*. 2014;32(4):459-470. doi:10.1016/j.clindermatol.2014.02.001.

4. Cahill AM, Nijs ELF. Pediatric Vascular Malformations: Pathophysiology, Diagnosis, and the Role of Interventional Radiology. *CardioVascular and Interventional Radiology*. 2011;34(4):691-704. doi:10.1007/s00270-011-0123-0.

5. Bagrodia N, Defnet AM, Kandel JJ. Management of lymphatic malformations in children. *Current Opinion in Pediatrics*. 2015;27(3):356-363. doi:10.1097/mop.0000000000000209.

6. Wassef M, Blei F, Adams D, et al. Vascular Anomalies Classification: Recommendations From the International Society for the Study of Vascular Anomalies. *Pediatrics*. 2015;136(1):e203-e214. doi:10.1542/peds.2014-3673.

7. Kutz AM, Aranibar L, Lobos N, Wortsman X. Color Doppler Ultrasound Follow-Up of Infantile Hemangiomas and Peripheral Vascularity in Patients Treated with Propranolol. *Pediatric Dermatology*. 2015;32(4):468-475. doi:10.1111/pde.12596.

8. Adams DM, Trenor CC, Hammill AM, et al. Efficacy and Safety of Sirolimus in the Treatment of Complicated Vascular Anomalies. *Pediatrics*. 2016;137(2). doi:10.1542/peds.2015-3257.

9. Snyder E, Puttgen K, Mitchell S, Ahlawat S, Tekes A. Magnetic Resonance Imaging of the Soft Tissue Vascular Anomalies in Torso and Extremities in Children: An Update With 2014 International Society for the Study of Vascular Anomalies Classification. *Journal of Computer Assisted Tomography*. 2017;42(2):167-177. doi:10.1097/rct.0000000000000675.

10. Merrow AC, Gupta A, Patel MN, Adams DM. 2014 Revised Classification of Vascular Lesions from the International Society for the Study of Vascular Anomalies: Radiologic-Pathologic Update. *RadioGraphics*. 2016;36(5):1494-1516. doi:10.1148/rg.2016150197.

© 2023 eviCore healthcare. All Rights Reserved.

11. Johnson CM, Navarro OM. Clinical and sonographic features of pediatric soft-tissue vascular anomalies part 1: classification, sonographic approach and vascular tumors. *Pediatric Radiology*. 2017;47(9):1184-1195. doi:10.1007/s00247-017-3885-y.

12. Johnson CM, Navarro OM. Clinical and sonographic features of pediatric soft-tissue vascular anomalies part 2: vascular malformations. *Pediatric Radiology*. 2017;47(9):1196-1208. doi:10.1007/s00247-017-3906-x.

13. Sadick M, Müller-Wille R, Wildgruber M, Wohlgemuth W. Vascular Anomalies (Part I): Classification and Diagnostics of Vascular Anomalies. RöFo - Fortschritte auf dem Gebiet der Röntgenstrahlen und der bildgebenden Verfahren. 2018;190(09):825-835. doi:10.1055/a-0620-8925.

14. Olivieri B, White CL, Restrepo R, et. al. Low-Flow Vascular Malformation Pitfalls: From Clinical Examination to Practical Imaging Evaluation—Part 2, Venous Malformation Mimickers. *American Journal of Roentgenology*. 2016;206(5):952-962. doi:10.2214/ajr.15.15794.

15. White CL, Olivieri B, Restrepo R, et.al. Low-Flow Vascular Malformation Pitfalls: From Clinical Examination to Practical Imaging Evaluation—Part 1, Lymphatic Malformation Mimickers. *American Journal of Roentgenology*. 2016;206(5):940-951. doi:10.2214/ajr.15.15793.

16. Kulungowski AM, Patel M. Lymphatic malformations. *Semin Pediatr Surg*. 2020;29(5):150971. doi:10.1016/j.sempedsurg.2020.150971.

© 2023 eviCore healthcare. All Rights Reserved.

Vasculitis (PEDPVD-3)

General Information (PEDPVD-3.1)

PVDP.VI.0003.1.A
v1.0.2023

- 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 individual outcomes to date.

Background and Supporting Information

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

© 2023 eviCore healthcare. All Rights Reserved.

Large Vessel Vasculitis (PEDPVD-3.2)

PVDP.VI.0003.2.A
v1.0.2023

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

Background and Supporting Information

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

© 2023 eviCore healthcare. All Rights Reserved.

Medium Vessel Vasculitis (PEDPVD-3.3)

PVDP.VI.0003.3.A
v1.0.2023

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

Background and Supporting Information

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

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Small Vessel Vasculitis (PEDPVD-3.4)

PVDP.VI.0003.4.A
v1.0.2023

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

© 2023 eviCore healthcare. All Rights Reserved.

- Annually-to evaluate the extent of disease

© 2023 eviCore healthcare. All Rights Reserved.

References (PEDPVD-3)

v1.0.2023

1. Muratore F, Pipitone N, Salvarani C, Schmidt WA. Imaging of vasculitis: State of the art. *Best Practice Research Clinical Rheumatology*. 2016;30(4):688-706. doi:10.1016/j.berh.2016.09.010.

2. Lensen KDF, Comans EFI, Voskuyl AE, et al. Large-Vessel Vasculitis: Interobserver Agreement and Diagnostic Accuracy of18F-FDG-PET/CT. *BioMed Research International*. 2015;2015:1-8. doi:10.1155/2015/914692. .

3. Soussan M, Nicolas P, Schramm C, et al. Management of Large-Vessel Vasculitis With FDG-PET. *Medicine*. 2015;94(14). doi:10.1097/md.0000000000000622.

4. Besson FL, Parienti J-J, Bienvenu B, et al. Diagnostic performance of 18F-fluorodeoxyglucose positron emission tomography in giant cell arteritis: a systematic review and meta-analysis. *European Journal of Nuclear Medicine and Molecular Imaging*. 2011;38(9):1764-1772. doi:10.1007/s00259-011-1830-0.

5. Sivaraman V, Fels EC, and Ardoin SP. Vasculitis syndromes. *Nelson Textbook of Pediatrics*, Chapter 192. eds Kliegman RM, St. Geme JW III, Blum NJ, et al. 21st ed. Philadelphia, PA: Elsevier; 2020:1317-1327.

6. Soliman M, Laxer R, Manson D, et. al. Imaging of systemic vasculitis in childhood. *Pediatric Radiology*. 2015;45(8):1110-1125. doi:10.1007/s00247-015-3339-3.

7. Sharma AM, Singh S, Lewis JE. Diagnostic Approach in Patients With Suspected Vasculitis. *Techniques in Vascular and Interventional Radiology*. 2014;17(4):226-233. doi:10.1053/j.tvir.2014.11.002.

8. Ammirati E, Moroni F, Pedrotti P, et al. Non-Invasive Imaging of Vascular Inflammation. *Frontiers in Immunology*. 2014;5:1-15. doi:10.3389/fimmu.2014.00399.

9. Granata C, Damasio MB, Zaottini F, et al. Imaging of Childhood Vasculitis. *Radiologic Clinics of North America*. 2017;55(5):1131-1143. doi:10.1016/j.rcl.2017.05.001.

10. Broncano J, Vargas D, Bhalla S, Cummings KW, Raptis CA, Luna A. CT and MR Imaging of Cardiothoracic Vasculitis. RadioGraphics. 2018;38(4):997-1021. doi:10.1148/rg.2018170136.

11. Jennette JC, Falk RJ, Bacon PA, et al. 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. *Arthritis & Rheumatism*. 2012;65(1):1-11. doi:10.1002/art.37715.

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

12. Ozen S, Pistorio A, Iusan SM, et al. EULAR/PRINTO/PRES criteria for Henoch-Schonlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Part II: Final classification criteria. *Annals of the Rheumatic Diseases*. 2010;69(5):798-806. doi:10.1136/ard.2009.116657.

© 2023 eviCore healthcare. All Rights Reserved.

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

Thoracic Aortic Disease (PEDPVD-4.1)

PVDP.AD.0004.1.A
v1.0.2023

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 may be indicated 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

© 2023 eviCore healthcare. All Rights Reserved.

        - MRA or CTA of (any or all):
            - Head
            - Neck
            - Chest
            - Abdomen
            - Pelvis
    - Individuals with Loyes-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 (Loyes-Dietz)
            - Neck (Loyes-Dietz)
    - The above imaging is indicated 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)

Thoracic Aortic Disease (PEDPVD-4.1)

© 2023 eviCore healthcare. All Rights Reserved.

        - Every 6 months if ≥4.5 or ≥0.5 cm growth per year, or any Loyes-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 indicated:
    - 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 aorta, Osteogenesis imperfecta, Homocystinuria, polycystic kidney disease, Pseudo xanthoma elasticum, Hurler syndrome.
    - Screening echocardiogram yearly.
    - If positive findings, follow protocol for aortic root dilatation.
- Follow-up of thoracic aortic abnormalities for other conditions please see discussions indicated elsewhere in the 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)**

© 2023 eviCore healthcare. All Rights Reserved.

- Neurofibromatosis- See **General Guidelines (PEDCD-1.0)** in the Pediatric Cardiac Imaging Guidelines

Thoracic Aortic Disease (PEDPVD-4.1)

© 2023 eviCore healthcare. All Rights Reserved.

Aortic Congenital Vascular Malformations (PEDPVD-4.2)

PVDP.PC.0004.2.A
v1.0.2023

- 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 **Esophagus (PEDNECK-7)** and/or **Trachea (PEDNECK-8)** in the Pediatric Neck Imaging Guidelines for additional guidelines.

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Visceral Artery Aneurysms (PEDPVD-4.3)

PVDP.AD.0004.3.A
v1.0.2023

- Visceral artery imaging indications in pediatric individuals are identical to those for adult individuals. See **Aortic Disorders and Renal Vascular Disorders and Visceral Artery Aneurysms (PVD-6)** in the Peripheral Vascular Disease Imaging Guidelines.

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

References (PEDPVD-4)

v1.0.2023

1. Byers PH, Belmont J, Black J, et al. Diagnosis, natural history, and management in vascular Ehlers-Danlos syndrome. *Am J Med Genet Part C Semin Med Genet*. 2017;175C:40-47. doi:10.1002/ajmg.c.31553.

2. Hanneman K, Newman B, Chan F. Congenital Variants and Anomalies of the Aortic Arch. *RadioGraphics*. 2017;37(1):32-51. doi:10.1148/rg.2017160033.

3. Erbel R, Aboyans V, Boileau C, et al. 2014 ESC Guidelines on the diagnosis and treatment of aortic diseases. *European Heart Journal*. 2014;35(41):2873-2926. doi:10.1093/eurheartj/ehu281.

4. Caglayan AO, Dundar M. Inherited diseases and syndromes leading to aortic aneurysms and dissections. *European Journal of Cardio-Thoracic Surgery*. 2009;35(6):931-940. doi:10.1016/j.ejcts.2009.01.006.

5. Coley BD, Chan FD. Acquired diseases of the great vessels. In: *Caffey's Pediatric Diagnostic Imaging*. Vol 1. 12th ed. Elsevier/Saunders; 2013:835.

6. Coley BD, Chan FD. Congenital diseases of the thoracic great arteries. In: *Caffey's Pediatric Diagnostic Imaging*. Vol 1. 12th ed. Elsevier/Saunders; 2013:772.

7. Collins RT. Cardiovascular Disease in Williams Syndrome. *Circulation*. 2013;127(21):2125-2134. doi:10.1161/circulationaha.112.000064.

8. D'hondt S, Damme TV, Malfait F. Vascular phenotypes in nonvascular subtypes of the Ehlers-Danlos syndrome: a systematic review. *Genetics in Medicine*. 2017;20(6):562-573. doi:10.1038/gim.2017.138.

9. Hiratzka LF, Creager MA, Isselbacher EM, et al. Surgery for Aortic Dilatation in Patients with Bicuspid Aortic Valves. *Journal of the American College of Cardiology*. 2016;67(6):724-731. doi:10.1016/j.jacc.2015.11.006.

10. Knadler JJ, Lemaire S, Mckenzie ED, et.al. Thoracic Aortic, Aortic Valve, and Mitral Valve Surgery in Pediatric and Young Adult Patients with Marfan Syndrome: Characteristics and Outcomes. *Seminars in Thoracic and Cardiovascular Surgery*. 2019;31(4):818-825. doi:10.1053/j.semtcvs.2019.06.005.

11. Landis BJ, Ware SM, James J, Shikany AR, Martin LJ, Hinton RB. Clinical Stratification of Pediatric Patients with Idiopathic Thoracic Aortic Aneurysm. *J Pediatr*. 2015;167(1):131-137. doi:10.1016/j.jpeds.2015.02.042.

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

12. Loughborough WW, Minhas KS, Rodrigues JCL, et al. Cardiovascular Manifestations and Complications of Loeys-Dietz Syndrome: CT and MR Imaging Findings. RadioGraphics. 2018;38(1):275-286. doi:10.1148/rg.2018170120.

13. Huang Y, Qu G. Faculty of 1000 evaluation for 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the diagnosis and management of patients with thoracic aortic disease. A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology Society of Thoracic Surgeons, and Society for Vascular Medicine.F1000 - Post-publication peer review of the biomedical literature. 2010. doi:10.3410/f.4998963.4932064.

14. MacCarrick G, Black JH 3rd, Bowdin S, et al. Loeys-Dietz syndrome: a primer for diagnosis and management. Genet Med. 2014;16(8):576–587. doi:10.1038/gim.2014.11.

15. Maron BJ, Zipes DP, Kovacs RJ. Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Preamble, Principles, and General Considerations. *Journal of the American College of Cardiology*. 2015;66(21):2343-2349. doi:10.1016/j.jacc.2015.09.032.

16. Meester JAN, Verstraeten A, Schepers D, et. al. Differences in manifestations of Marfan syndrome, Ehlers-Danlos syndrome, and Loeys-Dietz syndrome. *Annals of Cardiothoracic Surgery*. 2017;6(6):582-594. doi:10.21037/acs.2017.11.03.

17. Loeys BL, Dietz HC. Loeys-Dietz Syndrome. GeneReviews® [Internet]. https://www.ncbi.nlm.nih.gov/books/NBK1133/. Published March 1, 2018.

18. Oner T, Akgun G, Ergin SO, Karadag H, Yucel IK, Celebi A. Risk Factors Associated with Ascending Aortic Aneurysms and Aortic Elasticity Parameters in Children with a Bicuspid Aortic Valve. *Pediatric Cardiology*. 2019;40(5):980-986. doi:10.1007/s00246-019-02102-6.

19. Pierpont MEM, Lacro RV. Children with Thoracic Aortic Aneurysm: Challenges in Diagnosis and Therapy. *The Journal of Pediatrics*. 2015;167(1):14-16. doi:10.1016/j.jpeds.2015.03.056.

20. Sulli A, Talarico R, Scirè CA, et al. Ehlers-Danlos syndromes: state of the art on clinical practice guidelines. *RMD Open*. 2018;4(Suppl 1). doi:10.1136/rmdopen-2018-000790.

21. Williams JA, Loeys BL, Nwakanma LU, et al. Early Surgical Experience With Loeys-Dietz: A New Syndrome of Aggressive Thoracic Aortic Aneurysm Disease.

© 2023 eviCore healthcare. All Rights Reserved.

The Annals of Thoracic Surgery. 2007;83(2):s757-63. doi:10.1016/j.athoracsur.2006.10.091.

22. Zanotti G, Vricella L, Cameron D. Thoracic Aortic Aneurysm Syndrome in Children. *Seminars in Thoracic and Cardiovascular Surgery: Pediatric Cardiac Surgery Annual*. 2008;11(1):11-21. doi:10.1053/j.pcsu.2008.01.005.

© 2023 eviCore healthcare. All Rights Reserved.

Infantile Hemangiomas (PEDPVD-5)

Infantile Hemangiomas - General Considerations (PEDPVD-5.1)

PVDP.IH.0005.1.A

v1.0.2023

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 **Vascular Tumors (PEDPVD-2.6)** .

- 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 **PHACE(S) Syndrome PEDPVD-5.3)** , and for “LUMBAR syndrome” See **LUMBAR Syndrome PEDPVD-5.4)** .

© 2023 eviCore healthcare. All Rights Reserved.

Background and Supporting Information

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 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."

© 2023 eviCore healthcare. All Rights Reserved.

Multiple Infantile Hemangiomas (PEDPVD-5.2)

PVDP.IH.0005.2.A
v1.0.2023

- 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.

Background and Supporting Information

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 hemagniomas 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.

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

PHACE(S) Syndrome (PEDPVD-5.3)

PVDP.IH.0005.3.A
v1.0.2023

"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%).

- Indications for imaging a young child for suspected PHACE(S) syndrome include the following:
  - Large (5 or more cm in diameter) infantile hemangioma of the face, scalp, and/or neck.
  - Infantile hemangioma on face, scalp, or neck that is smaller than 5 cm in diameter, but with at least one major anomaly found in PHACE(S) syndrome, such as coarctation of the aorta or midline ventral defect.
  - Without any visible facial infantile hemangioma, PHACE(S) syndrome can also reasonably be suspected with the following:
    - Infantile hemangioma on upper chest or proximal upper extremity that is 5 cm or larger in size, with also major anomalies found in PHACE(S) syndrome
    - Large intraorbital infantile hemangioma.
- When PHACE(S) syndrome is reasonably suspected, initial imaging would include 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).

© 2023 eviCore healthcare. All Rights Reserved.

- A screening transthoracic echocardiogram, CPT® 93303 (CPT® 93320 and CPT® 93325 are also indicated if ordered with CPT® 93303). If abnormalities are identified on echocardiogram, a cardiac MRI (CPT® 75557 or CPT® 75561) is then indicated.
- If other clinical information or imaging shows involvement of the aorta, then MRI Chest without contrast (CPT® 71550) or MRI Chest without and with contrast (CPT® 71552) is also indicated.

- Need for follow up or surveillance imaging is dictated by the results of the initial clinical and imaging assessment, and any subsequent clinical change. The most frequent follow up will be needed for those deemed at highest risk, including when the following has been found:
  - 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

PHACE(S) Syndrome (PEDPVD-5.3)

© 2023 eviCore healthcare. All Rights Reserved.

Lumbar Syndrome (PEDPVD-5.4)

PVDP.IH.0005.4.A
v1.0.2023

- "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 then indicated:
  - 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 indicated 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 indicated:
    - MRA (for both arterial and venous phase imaging) Abdomen
    - MRA Pelvis
    - MRA Lower extremities

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

- 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.

Background and Supporting Information

The acronym "LUMBAR syndrome" refers to the association of Lower body infantile hemangiomas 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.

© 2023 eviCore healthcare. All Rights Reserved.

References (PEDPVD-5)

v1.0.2023

1. Blei F. Vascular anomalies: From bedside to bench and back again. *Current Problems in Pediatric and Adolescent Health Care*. 2002;32(3):72-93. doi:10.1067/mps.2002.125533.

2. Darrow DH, Greene AK, Mancini AJ, Nopper AJ. Diagnosis and Management of Infantile Hemangioma: Executive Summary. *Pediatrics*. 2015;136(4):786-791. doi:10.1542/peds.2015-2482.

3. Drolet BA, Chamlin SL, Garzon MC, et al. Prospective Study of Spinal Anomalies in Children with Infantile Hemangiomas of the Lumbosacral Skin. *The Journal of Pediatrics*. 2010;157(5):789-794. doi:10.1016/j.jpeds.2010.07.054.

4. Ferriero DM, Fullerton HJ, Bernard TJ, et al. Management of Stroke in Neonates and Children: A Scientific Statement from the American Heart Association/American Stroke Association. *Stroke*. 2019;50(3). doi:10.1161/str.0000000000000183.

5. Holland KE, Drolet BA. Approach to the Patient with an Infantile Hemangioma. *Dermatologic Clinics*. 2013;31(2):289-301. doi:10.1016/j.det.2012.12.006.

6. Iacobas I, Burrows PE, Frieden IJ, et al. LUMBAR: Association between Cutaneous Infantile Hemangiomas of the Lower Body and Regional Congenital Anomalies. *The Journal of Pediatrics*. 2010;157(5). doi:10.1016/j.jpeds.2010.05.027

7. Krowchuk DP, Frieden IJ, Mancini AJ, et al. Clinical Practice Guideline for the Management of Infantile Hemangiomas. *Pediatrics*. 2019;143(1). doi:10.1542/peds.2018-3475.

8. Léauté-Labrèze C, Harper JI, Hoeger PH. Infantile haemangioma. *The Lancet*. 2017;390(10089):85-94. doi:10.1016/s0140-6736(16)00645-0.

9. Menapace D, Mitkov M, Towbin R, Hogeling M. The changing face of complicated infantile hemangioma treatment. *Pediatric Radiology*. 2016;46(11):1494-1506.doi:10.1007/s00247-016-3643-6

10. Nelson WE, Kliegman R, St. Geme JW, et al. Chapter 669 Vascular disorders . In: Nelson Textbook of Pediatrics. 21st ed. Philadelphia, PA: Elsevier; 2020:3461-3469.

11. Obara P, Mccool J, Kalva SP, et al. ACR Appropriateness Criteria® Clinically Suspected Vascular Malformation of the Extremities. Journal of the American College of Radiology. 2019;16(11). doi:10.1016/j.jacr.2019.05.013.

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

12. Restrepo R, Francavilla ML, Mas R, Lee EY. Up-To-Date Practical Imaging Evaluation of Neonatal Soft-Tissue Tumors: What Radiologists Need to Know. *American Journal of Roentgenology*. 2017;209(1):195-204. doi:10.2214/ajr.16.17576.

13. Steiner JE, Mccoy GN, Hess CP, et al. Structural malformations of the brain, eye, and pituitary gland in PHACE syndrome. *American Journal of Medical Genetics Part A*. 2017;176(1):48-55. doi:10.1002/ajmg.a.38523.

14. Tuite GF, Thompson DN, Austin PF, Bauer SB. Evaluation and management of tethered cord syndrome in occult spinal dysraphism: Recommendations from the international children's continence society. *Neurourology and Urodynamics*. 2017;37(3):890-903. doi:10.1002/nau.23382.

© 2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com