

UNITEDHEALTHCARE® COMMUNITY PLAN: RADIOLOGY IMAGING COVERAGE DETERMINATION GUIDELINE

Pediatric Peripheral Vascular Disease (PVD) Imaging Guidelines (For Ohio Only)

V2.0.2024

Guideline Number: CSRAD024OH.C

Effective Date: November 15, 2024

Application (for Ohio Only)

This Medical Policy only applies to the state of Ohio. Any requests for services that are stated as unproven or services for which there is a coverage or quantity limit will be evaluated for medical necessity using Ohio Administrative Code 5160-1-01.

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# Related Community Plan Policies

#### Guideline

**Related Community Plan Policies** 

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# **Related Community Plan Policies**

## Related Community Plan Policies v2.0.2024

**General Policies** 

Cardiac Imaging Guidelines

**Pediatric Policies** 

- Pediatric Peripheral Vascular Disease (PVD) Imaging Guidelines
- Pediatric Cardiac Imaging Guidelines

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# Application (For Ohio Only)

#### Guideline

Application (For Ohio Only)

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# **Application (For Ohio Only)**

## **Application for Ohio OH UHC**

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# Guideline Development (Preface-1)

#### Guideline

Guideline Development (Preface-1.1)

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# **Guideline Development (Preface-1.1)**

## PRF.GG.0001.1.UOH

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- The UnitedHealthcare's evidence-based, proprietary clinical guidelines evaluate a range of advanced imaging and procedures, including NM, US, CT, MRI, PET, Radiation Oncology, Sleep Studies, as well as Cardiac, musculoskeletal and Spine interventions.
- UnitedHealthcare reserves the right to change and update the guidelines. The guidelines undergo a formal review annually. UnitedHealthcare's guidelines are based on current evidence supported by major national and international association and society guidelines and criteria, peer-reviewed literature, major treatises as well as, input from health plans, and practicing academic and community-based physicians.
- These guidelines are not intended to supersede or replace sound medical judgment, but instead, should facilitate the identification of the most appropriate imaging or other designated procedure given the individual's clinical condition. These guidelines are written to cover medical conditions as experienced by the majority of individuals. However, these guidelines may not be applicable in certain clinical circumstances, and physician judgment can override the guidelines.
- These guidelines provide evidence-based, clinical benefits with a focus on health care quality and patient safety.
- Clinical decisions, including treatment decisions, are the responsibility of the individual and his/her provider. Clinicians are expected to use independent medical judgment, which takes into account the clinical circumstances to determine individual management decisions.
- UnitedHealthcare supports the Choosing Wisely initiative (https:// www.choosingwisely.org/) by the American Board of Internal Medicine (ABIM) Foundation and many national physician organizations, to reduce the overuse of diagnostic tests that are low value, no value, or whose risks are greater than the benefits.

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# Benefits, Coverage Policies, and Eligibility Issues (Preface-2)

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Benefits, Coverage Policies, and Eligibility Issues (Preface-2.1) References (Preface-2)

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# Benefits, Coverage Policies, and Eligibility Issues (Preface-2.1)

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#### Investigational and Experimental Studies

- Certain studies, treatments, procedures, or devices may be considered experimental, investigational, or unproven for any condition, illness, disease, injury being treated if one of the following is present:
  - if there is a paucity of supporting evidence;
  - if the evidence has not matured to exhibit improved health parameters;
  - if clinical utility has not been demonstrated in any condition; OR
  - if the study, treatment, procedure, or device lacks a collective opinion of support
- Supporting evidence includes standards that are based on credible scientific evidence
  published in peer-reviewed medical literature (such as well conducted randomized
  clinical trials or cohort studies with a sample size of sufficient statistical power)
  generally recognized by the relevant medical community. Collective opinion of
  support includes physician specialty society recommendations and the views of
  physicians practicing in relevant clinical areas when physician specialty society
  recommendations are not available.

#### **Clinical and Research Trials**

- Similar to investigational and experimental studies, clinical trial imaging requests will be considered to determine whether they meet UnitedHealthcare's evidence-based guidelines.
- Imaging studies which are inconsistent with established clinical standards, or are requested for data collection and not used in direct clinical management are not supported.

#### **Legislative Mandate**

• State and federal legislations may need to be considered in the review of advanced imaging requests.

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# **References (Preface-2)**

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1. Coverage of Clinical Trials under the Patient Protection and Affordable Care Act; 42 U.S.C.A. § 300gg-8.

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# Clinical Information (Preface-3)

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Clinical Information (Preface-3.1) References (Preface-3)

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# **Clinical Information (Preface-3.1)**

## PRF.CL.0003.1.UOH v2.0.2024

#### **Clinical Documentation and Age Considerations**

- UnitedHealthcare's guidelines use an evidence-based approach to determine the most appropriate procedure for each individual, at the most appropriate time in the diagnostic and treatment cycle. UnitedHealthcare's guidelines are framed by:
  - · Clinical presentation of the individual, rather than the studies requested
  - Adequate clinical information that must be submitted to UnitedHealthcare in order to establish medical necessity for advanced imaging or other designated procedures includes, but is not limited to, the following:
    - Pertinent clinical evaluation should include a recent detailed history, physical examination<sup>20</sup> since the onset or change in symptoms, and/or laboratory and prior imaging studies.
      - Condition-specific guideline sections may describe additional clinical information which is required for a pertinent clinical evaluation.
      - The Spine and Musculoskeletal guidelines require x-ray studies from when the current episode of symptoms has started or changed; x-ray imaging does not have to be within the past 60 days.
      - Advanced imaging or other designated procedures should not be ordered prior to clinical evaluation of an individual by the physician treating the individual. This may include referral to a consultant specialist who will make further treatment decisions.
      - Other meaningful technological contact (telehealth visit, telephone or video call, electronic mail or messaging) since the onset or change in symptoms by an established individual can serve as a pertinent clinical evaluation.
        - Some conditions may require a face-to-face evaluation as discussed in the applicable condition-specific guideline sections.
    - A recent clinical evaluation may be unnecessary if the individual is undergoing a guideline-supported, scheduled follow-up imaging or other designated procedural evaluation. Exceptions due to routine surveillance indications are addressed in the applicable condition-specific guideline sections.
  - UnitedHealthcare's evidence-based approach to determine the most appropriate procedure for each individual requires submission of medical records pertinent to the requested imaging or other designated procedures.
- Many conditions affecting the pediatric population are different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and adult populations, minor differences may exist in management due to individual

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age, comorbidities, and differences in disease natural history between children and adults.

- Individuals who are 18 years old or younger<sup>19</sup> should be imaged according to the Pediatric Imaging Guidelines if discussed in the condition-specific guideline sections. Any conditions not specifically discussed in the Pediatric Imaging Guidelines should be imaged according to the General Imaging Guidelines. Individuals who are >18 years old should be imaged according to the General Imaging Guidelines, except where directed otherwise by a specific guideline section.
- The terms "male" and "female" used in these guidelines refer to anatomic-specific diseases and disease predispositions associated with the individual's sex assigned at birth rather than their gender identity. It should be noted that gender identity and anatomic-specific diseases as well as disease predispositions are not always linked. As such, these guidelines should be applied to the individual's corresponding known or suspected anatomic-specific disease or disease predisposition. At UnitedHealthcare, we believe that it is important to understand how all individuals, including those who are gender-diverse, choose to identify themselves. To ensure that gender-diverse individuals are treated with respect and that decisions impacting their healthcare are made correctly and with sensitivity, UnitedHealthcare recognizes all individuals with the following gender marker options: Male, Female, Transgender Male, Transgender Female, "X", and "Not Specified."

### **General Imaging Information**

- "Standard" or "conventional" imaging is most often performed in the initial and subsequent evaluations of malignancy. Standard or conventional imaging includes plain film, CT, MRI, or US.
  - Often, further advanced imaging is needed when initial imaging, such as ultrasound, CT, or MRI does not answer the clinical question. Uncertain, indeterminate, inconclusive, or equivocal may describe these situations.
- Appropriate use of contrast is a very important component of evidence-based advanced imaging use.
  - The appropriate levels of contrast for an examination (i.e., without contrast, with contrast, without and with contrast) is determined by the evidence-based guidance reflected in the condition-specific guideline sections.
  - If, during the performance of a non-contrast imaging study, there is the unexpected need to use contrast in order to evaluate a possible abnormality, then that is appropriate.<sup>1</sup>

### Ultrasound

- Diagnostic ultrasound uses high-frequency sound waves to evaluate soft tissue structures and vascular structures utilizing grey scale and Doppler techniques.
- Ultrasound allows for dynamic real-time imaging at the bedside.

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- Ultrasound is limited in areas where there is dense bone or other calcification.
- Ultrasound also has a relatively limited imaging window so may be of limited value in evaluating very large abnormalities.
- In general, ultrasound is highly operator-dependent, and proper training and experience are required to perform consistent, high-quality evaluations.
- Indications for ultrasound may include, but are not limited to, the following:
  - Obstetric and gynecologic imaging
  - Soft tissue and visceral imaging of the chest, abdomen, pelvis, and extremities
  - Brain and spine imaging when not obscured by dense bony structures
  - Vascular imaging when not obscured by dense bony structures
  - Procedural guidance when not obscured by dense bony structures
  - Initial evaluation of ill-defined soft tissue masses or fullness and differentiating adenopathy from mass or cyst. Prior to advanced imaging, ultrasound can be very beneficial in selecting the proper modality, body area, image sequences, and contrast level that will provide the most definitive information for the individual.
- More specific guidance for ultrasound usage, including exceptions to this general guidance, can be found throughout the condition-specific guidelines.

## **Computed Tomography (CT)**

- The AMA CPT<sup>®</sup> manual does not describe nor assign any minimum or maximum number of sequences for any CT study. CT imaging protocols are often influenced by the individual's clinical situation and additional sequences are not uncommon. There are numerous CT protocols that may be performed to evaluate specific clinical questions, and this technology is constantly undergoing development.
- CT utilizes ionizing radiation to create cross-sectional and volumetric images of the body.
  - Advantages over ultrasound include a much larger field of view and faster completion time in general. Disadvantages compared to ultrasound include lack of portability and exposure to ionizing radiation.
  - Advantages over MRI include faster imaging and a more spacious scanner area limiting claustrophobia. Disadvantages compared to MRI include decreased soft tissue definition, especially with non-contrast imaging, and exposure to ionizing radiation.
- CT can be performed without, with, or without and with intravenous (IV) contrast depending on the clinical indication and body area.
  - In general, non-contrast imaging is appropriate for evaluating structures with significant tissue density differences such as lung parenchyma and bony structures, or when there is a contraindication to contrast.
  - In general, CT with contrast is the most common level of contrast and can be used when there is need for improved vascular or soft tissue resolution, including better

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characterization of known or suspected malignancy, as well as infectious and inflammatory conditions.

- CT without and with contrast has a limited role as the risks of doubling the ionizing radiation exposure rarely outweigh the benefits of multiphasic imaging, though there are some exceptions which include, but are not limited to, the following:
  - Characterization of a mass
  - Characterization of arterial and venous anatomy
  - CT with contrast may be used to better characterize findings on a very recent (within two weeks) inconclusive non-contrast CT where the guidelines would support CT without and with contrast.
- More specific guidance for CT contrast usage, including exceptions to this general guidance, can be found throughout the condition-specific guidelines.
- Shellfish allergy:
  - It is commonly assumed that an allergy to shellfish indicates iodine allergy, and that this implies an allergy to iodinated contrast media used with CT. However, this is NOT true. Shellfish allergy is due to tropomyosins. Iodine plays no role in these allergic reactions. Allergies to shellfish do not increase the risk of reaction to iodinated contrast media any more than that of other allergens.<sup>1</sup>
- Enteric contrast (oral or rectal) is sometimes used in abdominal imaging. There is no specific CPT<sup>®</sup> code which refers to enteric contrast.
- The appropriate contrast level and anatomic region in CT imaging is specific to the clinical indication, as listed in the condition-specific guideline sections.
- CT should not be used to replace MRI in an attempt to avoid sedation unless it is listed as a recommended study the appropriate condition-specific guideline.
- There are significant potential adverse effects associated with the use of iodinated contrast media. These include hypersensitivity reactions, thyroid dysfunction, and contrast-induced nephropathy (CIN). Individuals with impaired renal function are at increased risk for CIN.<sup>2</sup>
- Both contrast CT and MRI may be considered to have the same risk profile with renal failure (GFR <30 mL/min).
- The use of CT contrast should proceed with caution in pregnant and breastfeeding individuals. There is a theoretical risk of contrast toxicity to the fetal and infant thyroid. The procedure can be performed if the specific need for that contrast-enhanced procedure outweighs risk to the fetus. Breastfeeding individuals may reduce this risk by choosing to pump and discard breast milk for 12-24 hours after the contrast injection.
- CT without contrast may be appropriate if clinical criteria for CT with contrast are met AND the individual has:
  - Elevated blood urea nitrogen (BUN) and/or creatinine
  - Renal insufficiency
  - Allergies to iodinated contrast

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- Thyroid disease which could be treated with I-131
- Diabetes
- Very elderly
- Urgent or emergent settings due to availability
- Trauma
- CT is superior to other imaging modalities in certain conditions including, but not limited to, the following:
  - Screening following trauma
  - Imaging pulmonary disease
  - Imaging abdominal and pelvic viscera
  - Imaging of complex fractures
  - Evaluation of inconclusive findings on Ultrasound or MRI, or if there is a contraindication to MRI
- More specific guidance for CT usage, including exceptions to this general guidance, can be found throughout the condition-specific guidelines.

#### Magnetic Resonance Imaging (MRI)

- The AMA CPT<sup>®</sup> manual does not describe nor assign any minimum or maximum number of sequences for any MRI study. MRI protocols are often influenced by the individual's clinical situation and additional sequences are not uncommon. There are numerous MRI sequences that may be performed to evaluate specific clinical questions, and this technology is constantly undergoing development.
- Magnetic Resonance Imaging (MRI) utilizes the interaction between the intrinsic radiofrequency of certain molecules in the body (hydrogen in most cases) and a strong external magnetic field.
  - MRI is often superior for advanced imaging of soft tissues and can also define physiological processes in some instances (e.g., edema, loss of circulation [AVN], and increased vascularity [tumors]).
  - MRI does not use ionizing radiation and even non-contrast images have much higher soft tissue definition than CT or Ultrasound.
  - MRI typically takes much longer than either CT or Ultrasound, and for some individuals may require sedation. It is also much more sensitive to individual motion that can degrade image quality than either CT or Ultrasound.
- MRI Breast and MRI Chest are not interchangeable, as they focus detailed sequences on different adjacent body parts.
- MRI may be utilized either as the primary advanced imaging modality, or when further definition is needed based on CT or ultrasound imaging.
- Most orthopedic and dental implants are not magnetic. These include hip and knee replacements; plates, screws, and rods used to treat fractures; and cavity fillings. Yet,

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all of these metal implants can distort the MRI image if near the part scanned.	of the body being
<ul> <li>Other implants, however, may have contraindications to MRI. The following:</li> </ul>	ese include the
<ul> <li>Pacemakers</li> </ul>	
<ul> <li>ICD or heart valves</li> </ul>	
<ul> <li>Metal implants in the brain</li> </ul>	
<ul> <li>Metal implants in the eyes or ears</li> </ul>	
<ul> <li>Infusion catheters and bullets or shrapnel</li> </ul>	
<ul> <li>CT can therefore be an alternative study to MRI in these scenario</li> </ul>	S.
<ul> <li>The contrast level and anatomic region in MRI imaging is specific to indication, as listed in the specific guideline sections.</li> </ul>	the clinical
<ul> <li>MRI utilizing Xenon Xe 129 for contrast is considered investigational</li> </ul>	and
experimental at this time. MRI with or with and without contrast in the	ese guidelines
refers to MRI utilizing gadolinium for contrast.	
<ul> <li>MRI is commonly performed without, without and with contrast.</li> </ul>	
<ul> <li>Non-contrast imaging offers excellent tissue definition.</li> </ul>	
<ul> <li>Imaging without and with contrast is commonly used when neede characterize tissue perfusion and vascularization.</li> </ul>	d to better
<ul> <li>Most contrast is gadolinium based and causes T2 brightening of and extracellular spaces.</li> </ul>	of the vascular
<ul> <li>Some specialized gadolinium and non-gadolinium contrast age and most commonly used for characterizing liver lesions.</li> </ul>	ents are available,
<ul> <li>MRI with contrast only is rarely appropriate and is usually used to</li> </ul>	
characterize findings on a recent inconclusive non-contrast MRI, on a completion study.	commonly called
<ul> <li>MRI contrast is contraindicated in pregnant individuals.</li> </ul>	
<ul> <li>More specific guidance for MRI contrast usage, including exception guidance, can be found throughout the condition-specific guideline</li> </ul>	•
<ul> <li>MRI may be preferred in individuals with renal failure and in individual intravenous CT contrast.</li> </ul>	als allergic to
<ul> <li>Both contrast CT and MRI may be considered to have the same r renal failure (GFR &lt;30 mL/min).<sup>2</sup></li> </ul>	isk profile with
<ul> <li>Gadolinium can cause Nephrogenic Systemic Fibrosis (NSF). The exposure to gadolinium in individuals with a low GFR (especially i greater the chance of individuals developing NSF.</li> </ul>	0
<ul> <li>Multiple studies have demonstrated potential for gadolinium depo the use of gadolinium-based contrast agents (GBCAs) for MRI stu</li> </ul>	sition following udies. <sup>3,4,5,6,7</sup>
	re is currently no
The U.S. Food and Drug Administration (FDA) has noted that the	

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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.<sup>8</sup>

- A CT may be approved in place of an MRI when clinical criteria are met for MRI AND there is a contraindication to having an MRI (pacemaker, ICD, insulin pump, neurostimulator, etc.).
  - When replacing MRI with CT, contrast level matching should occur as follows:
    - MRI without contrast  $\rightarrow$  CT without contrast
    - MRI without and with contrast  $\rightarrow$  CT with contrast or CT without and with contrast
- The following situations may impact the appropriateness for MRI and or MR contrast:
  - Caution should be taken in the use of gadolinium in individuals with renal failure.
  - The use of gadolinium contrast agents is contraindicated during pregnancy unless the specific need for that procedure outweighs risk to the fetus.
  - MRI can be performed for non-ferromagnetic body metals (i.e., titanium), although some imaging facilities will consider it contraindicated if recent surgery, regardless of the metal type.
- MRI should not be used as a replacement for CT for the sole reason of avoidance of ionizing radiation when MRI is not supported in the condition-based guidelines, since it does not solve the problem of overutilization.
- MRI is superior to other imaging modalities in certain conditions including, but not limited to, the following:
  - Imaging the brain and spinal cord
  - · Characterizing visceral and musculoskeletal soft tissue masses
  - · Evaluating musculoskeletal soft tissues including ligaments and tendons
  - Evaluating inconclusive findings on ultrasound or CT
  - Individuals who are pregnant or have high radiation sensitivity
  - Suspicion, diagnosis, or surveillance of infections
- More specific guidance for MRI usage, including exceptions to this general guidance, can be found throughout the condition-specific guidelines.

## Positron Emission Tomography (PET)

- PET is a nuclear medicine study that uses a positron emitting radiotracer to create cross-sectional and volumetric images based on tissue metabolism.
- Conventional imaging (frequently CT, sometimes MRI or bone scan) of the affected area(s) drives much of initial and restaging and surveillance imaging for malignancy and other chronic conditions. PET is not indicated for surveillance imaging unless specifically stated in the condition-specific guideline sections.
- PET/MRI is generally not supported, see **PET-MRI (Preface-5.3)**.

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- PET is rarely performed as a single modality, but is typically performed as a combined PET/CT.
  - The unbundling of PET/CT into separate PET and diagnostic CT CPT<sup>®</sup> codes is not supported, because PET/CT is done as a single study.
- PET/CT lacks the tissue definition of CT or MRI, but is fairly specific for metabolic activity based on the radiotracer used.
- Indications for PET/CT may include the following:
  - Oncologic Imaging for evaluation of tumor metabolic activity
  - Cardiac Imaging for evaluation of myocardial metabolic activity
  - Brain Imaging for evaluation of metabolic activity for procedural planning
- More specific guidance for PET usage, including exceptions to this general guidance, can be found throughout the condition-specific guidelines.

## **Overutilization of Advanced Imaging**

- A number of recent reports describe overutilization in many areas of advanced imaging and other procedures, which may include the following:
  - High-level testing without consideration of less invasive, lower cost options which may adequately address the clinical question at hand
  - Excessive radiation and costs with unnecessary testing
  - Defensive medical practice
  - CT without and with contrast (so called "double contrast studies") requests, which have few current indications
  - MRI requested in place of CT to avoid radiation without considering the primary indication for imaging
  - Adult CT settings and protocols used for smaller people and children
  - Unnecessary imaging procedures when the same or similar studies have already been conducted
- A review of the imaging or other relevant procedural histories of all individuals
  presenting for studies has been recognized as one of the more important processes
  that can be significantly improved. By recognizing that a duplicate or questionably
  indicated examination has been ordered for individuals, it may be possible to avoid
  exposing them to unnecessary risks.<sup>9,10</sup> To avoid these unnecessary risks, the
  precautions below should be considered:
  - The results of initial diagnostic tests or radiologic studies to narrow the differential diagnosis should be obtained prior to performing further tests or radiologic studies.
  - The clinical history should include a potential indication such as a known or suspected abnormality involving the body part for which the imaging study is being requested. These potential indications are addressed in greater detail within the applicable guidelines.

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- The results of the requested imaging procedures should be expected to have an impact on individual management or treatment decisions.
- Repeat imaging studies are not generally necessary unless there is evidence of disease progression, recurrence of disease, and/or the repeat imaging will affect an individual's clinical management.
- Pre-operative imaging/pre-surgical planning imaging/pre-procedure imaging is not indicated if the surgery/procedure is not indicated. Once the procedure has been approved or if the procedure does not require prior authorization, the appropriate preprocedural imaging may be approved.

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# **References (Preface-3)**

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# Coding Issues (Preface-4)

Guideline

3D Rendering (Preface-4.1) CT-, MR-, or Ultrasound-Guided Procedures (Preface-4.2) Unlisted Procedures/Therapy Treatment Planning (Preface-4.3) CPT<sup>®</sup> 76380 Limited or Follow-up CT (Preface-4.5) SPECT/CT Imaging (Preface-4.6) CPT<sup>®</sup> 76140 Interpretation of an Outside Study (Preface-4.7) Quantitative MR Analysis of Tissue Composition (Preface-4.8)

HCPCS Codes (Preface-4.9)

References (Preface-4)

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# 3D Rendering (Preface-4.1)

## PRF.CD.0004.1.UOH v2.0.2024

## **CPT<sup>®</sup> 76376 and CPT<sup>®</sup> 76377**

- Both codes require concurrent supervision of the image post-processing 3D manipulation of the volumetric data set and image rendering.
  - Concurrent supervision is defined as active physician participation in and monitoring of the reconstruction process including design of the anatomic region that is to be reconstructed; determination of the tissue types and actual structures to be displayed (e.g., bone, organs, and vessels); determination of the images or cine loops that are to be archived; and, monitoring and adjustment of the 3D work product. The American College of Radiology (ACR) recommends that it is best to document the physician's supervision or participation in the 3D reconstruction of images.
- These two codes differ in the need for and use of an independent workstation for post-processing.
  - CPT<sup>®</sup> 76376 reports procedures not requiring image post-processing on an independent workstation.
  - CPT<sup>®</sup> 76377 reports procedures that require image post-processing on an independent workstation.
- These 3D rendering codes should not be used for 2D reformatting.
- Two-dimensional reconstruction (e.g., reformatting an axial scan into the coronal plane) is now included in all cross-sectional imaging base codes and is not separately reimbursable.
- The codes used to report 3D rendering for ultrasound and echocardiography are also used to report the 3D post processing work on CT, MRI, and other tomographic modalities.
- Providers may be required to obtain prior authorization on these 3D codes even if prior authorization is not required for the echocardiography and/or ultrasound procedure codes. It may appear that UnitedHealthcare pre-authorizes echocardiography and/or ultrasound when, in fact, it may only be the 3D code that needs the prior authorization.
- CPT<sup>®</sup> codes for 3D rendering should not be billed in conjunction with computeraided detection (CAD), MRA, CTA, nuclear medicine SPECT studies, PET, PET/ CT, Mammogram, MRI Breast, US Breast, CT Colonography (virtual colonoscopy), Cardiac MRI, Cardiac CT, or Coronary CTA studies.

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- CPT<sup>®</sup> 76377 (3D rendering requiring image post-processing on an independent workstation) or CPT<sup>®</sup> 76376 (3D rendering not requiring image post-processing on an independent workstation) can be considered in the following clinical scenarios:
  - Bony conditions:
    - Evaluation of congenital skull abnormalities in newborns, infants, and toddlers (usually for pre-operative planning)
    - Complex fractures (comminuted or displaced)/dislocations of any joint (for preoperative planning when conventional imaging is insufficient)
    - Spine fractures, pelvic/acetabulum fractures, intra-articular fractures (for preoperative planning when conventional imaging is insufficient)
    - Pre-operative planning for other complex surgical cases
    - Complex facial fractures
  - Pre-operative planning for other complex surgical cases
    - Cerebral angiography
  - Pelvis conditions:
    - Uterine intra-cavitary lesion when initial US is equivocal: See <u>Abnormal Uterine</u> <u>Bleeding (AUB) (PV-2.1)</u> and <u>Leiomyoma/Uterine Fibroids (PV-12.1)</u> in the Pelvis Imaging Guidelines.
    - Hydrosalpinxes or peritoneal cysts when initial US is indeterminate: See <u>Complex Adnexal Masses (PV-5.3)</u> in the Pelvis Imaging Guidelines.
    - Lost IUD (inability to feel or see IUD string) with initial US: See <u>Intrauterine</u> <u>Device (PV-10.1)</u> in the Pelvis Imaging Guidelines.
    - Uterine anomalies with initial US: See <u>Uterine Anomalies (PV-14.1)</u> in the Pelvis Imaging Guidelines.
    - Infertility: See <u>Initial Infertility Evaluation, Female (PV-9.1)</u> in the Pelvis Imaging Guidelines.
  - Abdomen conditions:
    - CT Urogram: See <u>Hematuria and Hydronephrosis (AB-39)</u> in the Abdomen Imaging Guidelines.
    - MRCP: See <u>MR Cholangiopancreatography (MRCP) (AB-27)</u> in the Abdomen Imaging Guidelines.

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# CT-, MR-, or Ultrasound-Guided Procedures (Preface-4.2)

### PRF.CD.0004.2.A

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- CT-, MR-, and Ultrasound-guidance procedure codes contain all of the imaging necessary to guide a needle or catheter. It is inappropriate to routinely bill a diagnostic procedure code in conjunction with a guidance procedure code.
- Imaging studies performed as part of a CT-, MR-, or Ultrasound-guided procedure should be reported using the CPT<sup>®</sup> codes in the following table:

### **TABLE: Imaging Guidance Procedure Codes**

CPT®	Description
19085	Biopsy, breast, with placement of breast localization device(s), when performed, and imaging of the biopsy specimen, when performed, percutaneous; first lesion, including MR guidance
19086	Biopsy, breast, with placement of breast localization device(s), when performed, and imaging of the biopsy specimen, when performed, percutaneous; each additional lesion, including MR guidance
75989	Imaging guidance for percutaneous drainage with placement of catheter (all modalities)
76942	Ultrasonic guidance for needle placement
77011	CT guidance for stereotactic localization
77012	CT guidance for needle placement
77013	CT guidance for, and monitoring of parenchymal tissue ablation
77021	MR guidance for needle placement
77022	MR guidance for, and monitoring of parenchymal tissue ablation

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## CPT<sup>®</sup> 19085 and CPT<sup>®</sup> 19086

- The proper way to bill an MRI-guided breast biopsy is CPT<sup>®</sup> 19085 (Biopsy, breast, with placement of breast localization device(s), when performed, and imaging of the biopsy specimen, when performed, percutaneous; first lesion, including MR guidance). Additional lesions should be billed using CPT<sup>®</sup> 19086.
  - **CPT**<sup>®</sup> **77021** (MR guidance for needle placement) is not an appropriate code for a breast biopsy.

## **CPT<sup>®</sup> 75989**

- This code is used to report imaging guidance for a percutaneous drainage procedure in which a catheter is left in place.
- This code can be used to report whether the drainage catheter is placed under fluoroscopy, Ultrasound-, CT-, or MR-guidance modality.

## CPT<sup>®</sup> 77011

- A stereotactic CT localization scan is frequently obtained prior to sinus surgery. The dataset is then loaded into the navigational workstation in the operating room for use during the surgical procedure. The information provides exact positioning of surgical instruments with regard to the individual's 3D CT images.<sup>3</sup>
- In most cases, the pre-operative CT is a technical-only service that does not require interpretation by a radiologist.
  - The imaging facility should report CPT<sup>®</sup> 77011 when performing a scan not requiring interpretation by a radiologist.
  - If a diagnostic scan is performed and interpreted by a radiologist, the appropriate diagnostic CT code (e.g., CPT<sup>®</sup> 70486) should be used.
  - It is not appropriate to report both CPT<sup>®</sup> 70486 and CPT<sup>®</sup> 77011 for the same CT stereotactic localization imaging session.
  - 3D Rendering (CPT<sup>®</sup> 76376 or CPT<sup>®</sup> 76377) should not be reported in conjunction with CPT<sup>®</sup> 77011 (or CPT<sup>®</sup> 70486 if used). The procedure inherently generates a 3D dataset.

## $\mbox{CPT}^{\mbox{\tiny B}}$ 77012 (CT) and $\mbox{CPT}^{\mbox{\tiny B}}$ 77021 (MR)

- These codes are used to report imaging guidance for needle placement during biopsy, aspiration, and other percutaneous procedures.
- They represent the radiological supervision and interpretation of the procedure and are often billed in conjunction with surgical procedure codes.
  - For example, CPT<sup>®</sup> 77012 is reported when CT guidance is used to place the needle for a conventional arthrogram.
  - Only codes representing percutaneous surgical procedures should be billed with CPT<sup>®</sup> 77012 and CPT<sup>®</sup> 77021. It is inappropriate to use with surgical codes for open, excisional, or incisional procedures.

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- **CPT<sup>®</sup> 77021** (MR guidance for needle placement) is not an appropriate code for breast biopsy.
  - CPT<sup>®</sup> 19085 would be appropriate for the first breast biopsy site and CPT<sup>®</sup> 19086 would be appropriate for additional concurrent biopsies.

## CPT<sup>®</sup> 77013 (CT) and CPT<sup>®</sup> 77022 (MR)

- These codes include the initial guidance to direct a needle electrode to the tumor(s), monitoring for needle electrode repositioning within the lesion, and as necessary for multiple ablations to coagulate the lesion and confirmation of satisfactory coagulative necrosis of the lesion(s) and comparison to pre-ablation images.
  - **NOTE:** CPT<sup>®</sup> 77013 should only be used for non-bone ablation procedures.
    - CPT<sup>®</sup> 20982 includes CT guidance for bone tumor ablations.
  - Only codes representing percutaneous surgical procedures should be billed with CPT<sup>®</sup> 77013 and CPT<sup>®</sup> 77022. It is inappropriate to use with surgical codes for open, excisional, or incisional procedures.
- CPT<sup>®</sup> 77012 and CPT<sup>®</sup> 77021 (as well as guidance codes CPT<sup>®</sup> 76942 [US], and CPT<sup>®</sup> 77002 - CPT<sup>®</sup> 77003 [fluoroscopy]) describe radiologic guidance by different modalities.
  - Only one unit of any of these codes should be reported per individual encounter (date of service). The unit of service is considered to be the individual encounter, not the number of lesions, aspirations, biopsies, injections, or localizations.

# Unlisted Procedures/Therapy Treatment Planning (Preface-4.3)

## PRF.CD.0004.3.UOH

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CPT <sup>®</sup>	Description
76497	Unlisted CT procedure (e.g., diagnostic or interventional)
76498	Unlisted MR procedure (e.g., diagnostic or interventional)
78999	Unlisted procedure, diagnostic nuclear medicine

- These unlisted codes should be reported whenever a diagnostic or interventional CT or MR study is performed in which an appropriate anatomic site-specific code is not available.
  - A Category III code that describes the procedure performed must be reported rather than an unlisted code if one is available.
- CPT<sup>®</sup> 76497 or CPT<sup>®</sup> 76498 (Unlisted CT or MRI procedure) can be considered in the following clinical scenarios:
  - Studies done for navigation and planning for neurosurgical procedures (i.e., Stealth or Brain Lab Imaging)<sup>1,2</sup>
  - Custom joint arthroplasty planning (not as an alternative recommendation): See Osteoarthritis (MS-12.1) in the Musculoskeletal Imaging Guidelines.
  - Any procedure/surgical planning if thinner cuts or different positional acquisition (than those on the completed diagnostic study) are needed. These could include navigational bronchoscopy: See <u>Navigational Bronchoscopy (CH-1.7)</u> in the Chest Imaging Guidelines.

### Therapy Treatment Planning

 Radiation Therapy Treatment Planning: See <u>Unlisted Procedure Codes in</u> <u>Oncology (ONC-1.5)</u> in the Oncology Imaging Guidelines.

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# CPT<sup>®</sup> 76380 Limited or Follow-up CT (Preface-4.5)

## PRF.CD.0004.5.UOH

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- CPT<sup>®</sup> 76380 describes a limited or follow-up CT scan. The code is used to report any CT scan, for any given area of the body, in which the work of a full diagnostic code is not performed.
- Common examples include, but are not limited to, the following:
  - Limited sinus CT imaging protocol
  - Limited or follow-up slices through a known pulmonary nodule
  - · Limited slices to assess a non-healing fracture (such as the clavicle)
- Limited CT (CPT<sup>®</sup> 76380) is not indicated for treatment planning purposes. See <u>Unlisted Procedure Codes in Oncology (ONC-1.5)</u> in the Oncology Imaging Guidelines.
- It is inappropriate to report CPT<sup>®</sup> 76380, in conjunction with other diagnostic CT codes, to cover 'extra slices' in certain imaging protocols.
  - There is no specific number of sequences or slices defined in any CT CPT<sup>®</sup> code definition.
  - The AMA, in *CPT*<sup>®</sup> 2019, does not describe nor assign any minimum or maximum number of sequences or slices for any CT study.
    - A few additional slices or sequences are not uncommon.
    - CT imaging protocols are often influenced by the individual's clinical situation. Sometimes the protocols require more time and sometimes less.

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# SPECT/CT Imaging (Preface-4.6)

## PRF.CD.0004.6.A

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- SPECT/CT involves SPECT (Single Photon Emission Computed Tomography) nuclear medicine imaging and CT for optimizing location, accuracy, and attenuation correction and combines functional and anatomic information.
  - Common studies using this modality include <sup>123</sup>I- or <sup>131</sup>I-Metaiodobenzylguanidine (MIBG) and octreotide scintigraphy for neuroendocrine tumors.
- Hybrid Nuclear/CT scan can be reported as CPT<sup>®</sup> 78830 (single area and single day), CPT<sup>®</sup> 78831 (2 or more days), or CPT<sup>®</sup> 78832 (2 areas with one day and 2-day study).
- CPT<sup>®</sup> 78072 became effective January 1, 2013 for SPECT/CT parathyroid nuclear imaging.

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# **CPT<sup>®</sup> 76140 Interpretation of an Outside** Study (Preface-4.7)

### PRF.CD.0004.7.UOH

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- It is inappropriate to use diagnostic imaging codes for interpretation of a previously performed exam that was completed at another facility.
  - If the outside exam is being used for comparison with a current exam, the diagnostic code for the current examination includes comparison to the prior study.<sup>4</sup>
  - CPT<sup>®</sup> 76140 is the appropriate code to use for an exam which was completed elsewhere and a secondary interpretation of the images is requested.<sup>5</sup>

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# Quantitative MR Analysis of Tissue Composition (Preface-4.8)

## PRF.CD.0004.8.A

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- Category III CPT<sup>®</sup> codes for quantitative analysis of multiparametric-MR (mp-MRI) data with and without an associated diagnostic MRI have been established. Quantitative mp-MRI uses software to analyze tissue physiology of visceral organs and other anatomic structures non-invasively. At present, these procedures are primarily being used in clinical trials and there is no widely recommended indications in clinical practice. As such, these procedures are considered to be investigational and experimental for coverage purposes.
  - CPT<sup>®</sup> 0648T (without diagnostic MRI) and CPT<sup>®</sup> 0649T (with diagnostic MRI) refer to data analysis with and without associate imaging of a single organ, with its most common use being LiverMultiScan (LMS).
    - See <u>Fatty Liver (AB-29.2)</u> in the Abdomen Imaging Guidelines.
  - CPT<sup>®</sup> 0697T (without diagnostic MRI) and CPT<sup>®</sup> 0698T (with diagnostic MRI) refer to data analysis with and without associate imaging of a multiple organs, with its most common use being CoverScan.

# HCPCS Codes (Preface-4.9)

## PRF.CD.0004.9.UOH

#### v2.0.2024

- Healthcare Common Procedure Coding System (HCPCS) codes are utilized by some hospitals in favor of the typical Level-III CPT<sup>®</sup> codes. These codes are typically 4 digits preceded by a C or S.<sup>6</sup>
  - Many of these codes have similar code descriptions to Level-III CPT<sup>®</sup> codes (i.e., C8931 MRA with dye, Spinal Canal; and, CPT<sup>®</sup> 72159 MRA Spinal Canal).
  - If cases are submitted with HCPCS codes with similar code descriptions to the typical Level-III CPT<sup>®</sup> codes, those procedures should be managed in the same manner as the typical CPT<sup>®</sup> codes.
  - HCPCS code management is discussed further in the applicable guideline sections.
- Requests for many Healthcare Common Procedure Coding System (HCPCS) codes, including non-specific codes such as S8042 (Magnetic resonance imaging [MRI], lowfield), should be redirected to a more appropriate and specific CPT<sup>®</sup> code. Exceptions are noted in the applicable guideline sections.

# **References (Preface-4)**

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# Whole-Body Imaging (Preface-5)

#### Guideline

Whole-Body CT Imaging (Preface-5.1) Whole-Body MR Imaging (Preface-5.2) PET-MRI (Preface-5.3) References (Preface-5)

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# Whole-Body CT Imaging (Preface-5.1)

### PRF.WB.0005.1.UOH

### v2.0.2024

- Whole-body CT or LifeScan (CT Brain, Chest, Abdomen, and Pelvis) for screening of asymptomatic individuals is not indicated. The performance of whole-body screening CT examinations in healthy individuals does not meet any of the current validity criteria for screening studies and there is no clear documentation of benefit versus radiation risk.
- Whole-body low-dose CT is supported for oncologic staging in Multiple Myeloma. See <u>Multiple Myeloma and Plasmacytomas (ONC-25)</u> in the Oncology Imaging Guidelines.

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# Whole-Body MR Imaging (Preface-5.2)

### PRF.WB.0005.2.A

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- Whole-body MRI (WBMRI) is, with the exception of select cancer predisposition syndromes and autoimmune conditions discussed below, generally not supported at this time due to lack of standardization in imaging technique and lack of evidence that WBMRI improves outcome for any individual disease state.
  - While WBMRI has the benefit of whole-body imaging and lack of radiation exposure, substantial variation still exists in the number of images, type of sequences (STIR vs. diffusion weighting, for example), and contrast agent(s) used.
- Coding considerations:
  - There are no established CPT<sup>®</sup> or HCPCS codes for reporting WBMRI.
  - WBMRI is at present only reportable using CPT<sup>®</sup> 76498. All other methods of reporting whole-body MRI are inappropriate including the following:
    - Separate diagnostic MRI codes for multiple individual body parts
    - MRI Bone Marrow Supply (CPT<sup>®</sup> 77084)
- Disease-specific considerations:
  - Cancer screening:
    - Interval WBMRI is recommended for cancer screening in individuals with select cancer predisposition syndromes. Otherwise, WBMRI has not been shown to improve outcomes for cancer screening.
      - For additional information, see <u>Li-Fraumeni Syndrome (LFS)</u> (PEDONC-2.2), <u>Hereditary Paraganglioma-Pheochromocytoma (HPP)</u> Syndromes (PEDONC-2.13), or <u>Constitutional Mismatch Repair</u> <u>Deficiency (CMMRD or Turcot Syndrome) (PEDONC-2.15)</u> in the Pediatric Oncology Imaging Guidelines.
  - Cancer staging and restaging:
    - While the feasibility of WBMRI has been established, data remain conflicting on whether WBMRI is of equivalent diagnostic accuracy compared with standard imaging modalities such as CT, scintigraphy, and PET imaging.
    - Evidence has not been published establishing WBMRI as a standard evaluation for any type of cancer.
  - Autoimmune disease:
    - WBMRI can be approved in some situations for individuals with chronic recurrent multifocal osteomyelitis.
      - For additional information, see <u>Chronic Recurrent Multifocal</u> <u>Osteomyelitis (PEDMS-10.2)</u> in the Pediatric Musculoskeletal Imaging Guidelines.

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# **PET-MRI (Preface-5.3)**

## PRF.WB.0005.3.A

#### v2.0.2024

- PET-MRI is generally not supported for a vast majority of oncologic and neurologic conditions due to lack of standardization in imaging technique and interpretation. However, it may be appropriate in select circumstances when the following criteria are met:
  - The individual meets condition-specific guidelines for PET-MRI OR
  - The individual meets ALL of the following:
    - The individual is a pediatric patient or being treated under a pediatric guideline and treatment plan AND
    - The individual meets guideline criteria for PET-CT, AND
    - PET-CT is not available at the treating institution, AND
    - The provider requests PET-MRI in lieu of PET-CT
- When the above criteria are met, PET-MRI may be reported using the code combination of PET Whole-Body (CPT<sup>®</sup> 78813) and MRI Unlisted (CPT<sup>®</sup> 76498). All other methods of reporting PET-MRI are inappropriate.
  - When clinically appropriate, diagnostic MRI codes may be indicated at the same time as the PET-MRI code combination.
- For more information, see <u>PET Imaging in Pediatric Oncology (PEDONC-1.4)</u> in the Pediatric Oncology Imaging Guidelines, and <u>PET Brain Imaging (PEDHD-2.3)</u> and <u>Special Imaging Studies in Evaluation for Epilepsy Surgery (PEDHD-6.3)</u> in the Pediatric Head Imaging Guidelines.

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# References (Preface-6)

Guideline

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# **References (Preface-6.1)**

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- Complete reference citations for the journal articles are embedded within the body of the guidelines and/or may be found on the Reference pages at the end of some guideline sections.
- The website addresses for certain references are included in the body of the guidelines but are not hyperlinked to the actual website.
- The website address for the American College of Radiology (ACR) Appropriateness Criteria<sup>®</sup> is http://www.acr.org.

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# Copyright Information (Preface-7)

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# Trademarks (Preface-8)

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# General Guidelines (PEDPVD-1)

#### Guideline

Procedure Codes Associated with PVD Imaging (PEDPVD) General Guidelines (PEDPVD-1.0) Age Considerations (PEDPVD-1.1) Modality General Considerations (PEDPVD-1.3) References (PEDPVD-1)

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# Procedure Codes Associated with PVD Imaging (PEDPVD)

## PVDP.GG.0001.A

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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)	
Magnetic resonance angiography, lower extremity, with or without contrast material(s)	73725
Magnetic resonance angiography, abdomen, with or without contrast material(s)	74185

СТА	CPT <sup>®</sup>
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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СТА	CPT <sup>®</sup>
Computed tomographic angiography, chest (noncoronary), with contrast material(s), including noncontrast images, if performed, and image postprocessing	71275
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

Nuclear Medicine	CPT <sup>®</sup>
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

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Ultrasound	CPT <sup>®</sup>
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 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)**

### PVDP.GG.0001.0.A

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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 (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.

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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 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<sup>14</sup> 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 may be indicated 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 may be indicated for further evaluation of abnormalities suggested on prior US or MRI Procedures.
- CT may be indicated without prior MR or US, especially in the following (nonexhaustive 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 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<sup>®</sup> 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 <u>3D Rendering (Preface-4.1)</u> in the Preface Imaging Guidelines
- Nuclear Medicine
  - Nuclear medicine studies are rarely used in the evaluation of peripheral vascular disorders but are indicated in the following circumstances:
    - Lymphoscintigraphy (CPT<sup>®</sup> 78195) is indicated for evaluation of lower extremity lymphedema when a recent Doppler ultrasound is negative for valvular insufficiency.
    - Vascular flow imaging (CPT<sup>®</sup> 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<sup>®</sup> 78456, CPT<sup>®</sup> 78457, and CPT<sup>®</sup> 78458) 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<sup>®</sup> 78800, CPT<sup>®</sup> 78801, CPT<sup>®</sup> 78802 or CPT<sup>®</sup> 78803) can be approved for evaluation of the following:
      - Mycotic aneurysms
      - Vascular graft infection

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- 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 indicated and warranted for specific clinical situations.

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# Vascular Anomalies (PEDPVD-2)

#### Guideline

General Information (PEDPVD-2.1) Lymphatic Malformations (PEDPVD-2.2) Venous Malformations (PEDPVD-2.3) Capillary Malformations (PEDPVD-2.4) Arteriovenous Malformations (AVMs) and Fistulas (PEDPVD-2.5) Vascular Tumors (PEDPVD-2.6) References (PEDPVD-2)

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# **General Information (PEDPVD-2.1)**

### PVDP.AN.0002.1.A

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- 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).

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# Lymphatic Malformations (PEDPVD-2.2)

### PVDP.AN.0002.2.A

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- Ultrasound is indicated as an initial examination for superficial lesions.<sup>18</sup>
  - 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<sup>17</sup>
  - Inconclusive ultrasound findings
  - Preoperative planning
  - Post treatment evaluation<sup>18</sup>
- 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.

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# **Venous Malformations (PEDPVD-2.3)**

### PVDP.AN.0002.3.A

### v1.0.2024

- 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<sup>®</sup> 71260) or CTA Chest (CPT<sup>®</sup> 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.

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# **Capillary Malformations (PEDPVD-2.4)**

### PVDP.AN.0002.4.A

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 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.

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

### PVDP.AN.0002.5.A

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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 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.

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# Vascular Tumors (PEDPVD-2.6)

### PVDP.AN.0002.6.A

#### v1.0.2024

- 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.

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# Vasculitis (PEDPVD-3)

Guideline

General Information (PEDPVD-3.1) Large Vessel Vasculitis (PEDPVD-3.2) Medium Vessel Vasculitis (PEDPVD-3.3) Small Vessel Vasculitis (PEDPVD-3.4) References (PEDPVD-3)

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# **General Information (PEDPVD-3.1)**

## PVDP.VI.0003.1.A

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- PET/CT is **not** medically necessary 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.

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# Large Vessel Vasculitis (PEDPVD-3.2)

### PVDP.VI.0003.2.A

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- 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.

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## Medium Vessel Vasculitis (PEDPVD-3.3)

### PVDP.VI.0003.3.A

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- 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 <u>Multisystem</u> <u>inflammatory syndrome in children (MIS-C) (PEDCD-12)</u> in the pediatric cardiac imaging guidelines.
- Imaging guidelines for Kawasaki Disease- see <u>Kawasaki Disease (PEDCD-6)</u> 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.

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# Small Vessel Vasculitis (PEDPVD-3.4)

## PVDP.VI.0003.4.A

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- 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<sup>®</sup> 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<sup>®</sup> 70486) and/or CT Chest without contrast (CPT<sup>®</sup> 71250) or with contrast (CPT<sup>®</sup> 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<sup>®</sup> 71250) or with contrast (CPT<sup>®</sup> 71260) is indicated in the following circumstances:
      - New or worsening clinical symptoms affecting the body area requested
      - To assess response to medical therapy when a change in treatment regimen is being considered
      - Annually-to evaluate the extent of disease
  - Immune complex associated small-vessel vasculitis [immunoglobulin A–associated vasculitis (IgAV)]:
    - Doppler ultrasound of the affected body part (most commonly abdomen) is indicated in the following circumstances:
      - New or worsening clinical symptoms affecting the body area requested
      - To assess response to medical therapy when a change in treatment regimen is being considered
      - Annually-to evaluate the extent of disease

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

Guideline

Thoracic Aortic Disease (PEDPVD-4.1) Aortic Congenital Vascular Malformations (PEDPVD-4.2) Visceral Artery Aneurysms (PEDPVD-4.3) References (PEDPVD-4)

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### **Thoracic Aortic Disease (PEDPVD-4.1)**

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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 firstdegree 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
    - MRA or CTA of (any or all):
      - Head
      - Neck
      - Chest
      - Abdomen

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- 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)
        - 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

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Cardiovascular and Radiology Imaging Guidelines

- Individuals with Turner syndrome see section <u>Aortic disease in Turner Syndrome</u> (CD-11.2.10) in the Cardiac Imaging Guideline
- Williams syndrome See section <u>LVOT lesions (PEDCD-2.4.10)</u> in the Pediatric Cardiology Imaging Guideline
- Individuals with congenital heart disease would be managed based on <u>Imaging</u> and <u>Surveillance per Congenital lesion (PEDCD-2.4)</u> 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 <u>Aortic Coarctation and IAA (interrupted aortic</u> <u>arch) (PEDCD-2.4.11)</u> in the Pediatric Cardiac Imaging Guidelines
  - Congenital rubella syndrome- See <u>Imaging and Surveillance per Congenital</u> <u>lesion (PEDCD-2.4)</u> in the Pediatric Cardiac Imaging Guidelines
  - Kawasaki Syndrome- See Kawasaki Disease (PEDCD-6)
  - Neurofibromatosis- See <u>General Guidelines (PEDCD-1.0)</u> in the Pediatric Cardiac Imaging Guidelines

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### Aortic Congenital Vascular Malformations (PEDPVD-4.2)

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

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### **Visceral Artery Aneurysms (PEDPVD-4.3)**

### PVDP.AD.0004.3.A

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

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# Infantile Hemangiomas (PEDPVD-5)

Guideline

Infantile Hemangiomas – General Considerations (PEDPVD-5.1) Multiple Infantile Hemangiomas (PEDPVD-5.2) PHACE(S) Syndrome (PEDPVD-5.3) LUMBAR Syndrome (PEDPVD-5.4) References (PEDPVD-5)

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### Infantile Hemangiomas – General Considerations (PEDPVD-5.1)

### PVDP.IH.0005.1.A

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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 <u>Vascular Tumors (PEDPVD-2.6)</u>.

- Multiple (5 or more) infantile hemangiomas can be associated with hepatic hemangiomas with risk potential for high-output cardiac failure and other risks see <u>Multiple Infantile Hemangiomas (PEDPVD 5.2)</u>.
- 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<sup>®</sup> 93303 ordered with CPT<sup>®</sup> 93320 and CPT<sup>®</sup> 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<sup>®</sup> 72148) or MRI Lumbar Spine without and with contrast (CPT<sup>®</sup> 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 <u>PHACE(S) Syndrome</u> <u>PEDPVD-5.3</u>), and for "LUMBAR syndrome" See <u>LUMBAR Syndrome PEDPVD-5.4</u>)

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#### 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 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."

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### Multiple Infantile Hemangiomas (PEDPVD-5.2)

### PVDP.IH.0005.2.A

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- Multiple (5 or more) hemangiomas is an indication for Ultrasound with Doppler exam of the liver (CPT<sup>®</sup> 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.

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## PHACE(S) Syndrome (PEDPVD-5.3)

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"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<sup>®</sup> 70551) or MRI Brain without and with contrast (CPT<sup>®</sup> 70553)
  - MRI Orbits without contrast (CPT<sup>®</sup> 70540) or MRI Orbits without and with contrast (CPT<sup>®</sup> 70543)
  - MRA Head without contrast (CPT<sup>®</sup> 70544) or MRA Head without and with contrast, (CPT<sup>®</sup> 70546)
  - MRA Neck may be done either without contrast (CPT<sup>®</sup> 70547), with contrast (CPT<sup>®</sup> 70548), or without and with contrast (CPT<sup>®</sup> 70549)
  - MRA Chest (CPT<sup>®</sup> 71555).

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- A screening transthoracic echocardiogram, CPT<sup>®</sup> 93303 (CPT<sup>®</sup> 93320 and CPT<sup>®</sup> 93325 are also indicated if ordered with CPT<sup>®</sup> 93303). If abnormalities are identified on echocardiogram, a cardiac MRI (CPT<sup>®</sup> 75557 or CPT<sup>®</sup> 75561) is then indicated.
- If other clinical information or imaging shows involvement of the aorta, then MRI Chest without contrast (CPT<sup>®</sup> 71550) or MRI Chest without and with contrast (CPT<sup>®</sup> 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 <u>Imaging and Surveillance per Congenital lesion (PEDCD-2.4)</u> in the Pediatric Cardiac Imaging Guidelines

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## LUMBAR Syndrome (PEDPVD-5.4)

### PVDP.IH.0005.4.A

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- "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<sup>®</sup> 76800) in infants up to 6 months of age, abdomen (CPT<sup>®</sup> 76700), and pelvis (CPT<sup>®</sup> 76856), with color Doppler.
  - MRI Lumbar Spine without contrast (CPT<sup>®</sup> 72148) or without and with contrast (CPT<sup>®</sup> 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<sup>®</sup> 72195) or without and with contrast (CPT<sup>®</sup> 72197) and/or
    - MRI Abdomen without contrast (CPT<sup>®</sup> 74181) or without and with contrast (CPT<sup>®</sup> 74183).
  - MRA Abdomen CPT<sup>®</sup> 74185 and/or Pelvis CPT<sup>®</sup> 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<sup>®</sup> 73718) or lower extremity without and with contrast (CPT<sup>®</sup> 73720) and/or lower extremity joint without contrast (CPT<sup>®</sup> 73721) or lower extremity joint without and with contrast (CPT<sup>®</sup> 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
    - Note: this should be reported as CPT<sup>®</sup> 74185 and CPT<sup>®</sup> 73725; the CPT<sup>®</sup> code for MRA Pelvis (CPT<sup>®</sup> 72198) should not be included in this circumstance.

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### 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.

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## Policy History and Instructions for Use

#### Guideline

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### **Policy History and Instructions for Use**

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#### Instructions for Use

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#### **Policy History/Revision Information**

Date	Summary of Changes
02/01/2024	Annual evidence-based updates
07/01/2024	Interim evidence-based updates

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