CIGNA MEDICAL COVERAGE POLICIES– RADIOLOGY

Pediatric and Special Populations

Oncology Imaging Guidelines

Effective Date: February 02, 2027

Instructions for use

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

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

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

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

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

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

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Abbreviations for Pediatric and Special Populations Oncology Imaging Guidelines

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Abbreviations for Pediatric and Special Populations Oncology Imaging Guidelines	
AFP	alpha-fetoprotein (tumor marker)
ALCL	anaplastic large cell lymphoma
ALL	acute lymphoblastic leukemia
AML	acute myelogenous leukemia
ß-hCG	human chorionic gonadotropin beta-subunit (tumor marker)
BKL	Burkitt's lymphoma
BWT	bilateral Wilms tumor
CCSK	clear cell sarcoma of the kidney
CNS	central nervous system
COG	Children's Oncology group
CPT®	current procedural terminology; trademark of the American Medical Association
CSF	cerebrospinal fluid
CT	computed tomography
CXR	chest x-ray

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Abbreviations for Pediatric and Special Populations Oncology Imaging Guidelines	
DAWT	diffuse anaplasia Wilms tumor
ESFT	Ewing sarcoma family of tumors
FAWT	focal anaplasia Wilms tumor
FHWT	favorable histology Wilms tumor
HL	Hodgkin lymphoma
HSCT	hematopoietic stem cell transplant (bone marrow or peripheral blood)
HVA	homovanillic acid
LL	lymphoblastic lymphoma
MIBG	metaiodobenzylguanidine (nuclear scan using ^{123}i or ^{131}i)
MPNST	malignant peripheral nerve sheath tumor
MRI	magnetic resonance imaging
NBL	neuroblastoma
NED	no evidence of disease
NHL	non-Hodgkin lymphoma
NPC	nasopharyngeal carcinoma
NRSTS	nonrhabdomyosarcomatous soft tissue sarcomas
OS	osteosarcoma
PET	positron emission tomography
PMBCL	primary mediastinal b-cell lymphoma

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Abbreviations for Pediatric and Special Populations Oncology Imaging Guidelines	
PNET	primitive neuroectodermal tumor
RCC	renal cell carcinoma
RMS	rhabdomyosarcoma
US	ultrasound
VMA	vannilylmandelic acid
WBC	white blood cell count
XRT	radiation therapy

Pediatric and Special Populations Oncology

General Criteria (PEDONC-1.0)

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Criteria

- A relevant clinical evaluation or meaningful contact (telephone call, electronic mail or messaging) should be performed prior to considering advanced imaging, unless the individual is undergoing guideline-supported scheduled off therapy surveillance evaluation or cancer screening. The clinical evaluation may include a relevant history and physical examination, including biopsy, appropriate laboratory studies, and results of non-advanced or advanced imaging modalities.
  - Because of the relatively small number of childhood cancer treatment centers, it is common to combine off-therapy visits with imaging and other sub-specialist visits to accommodate families traveling long distances for their child's care.
- Unless otherwise stated in the disease-specific guideline, a histological confirmation of malignancy (or recurrence) and the stage of disease is required to perform a medical necessity review of the requested imaging.
- Unless otherwise stated in the disease-specific guideline, advanced imaging of asymptomatic individuals is not routinely considered medically necessary without signs or symptoms of systemic involvement of cancer.
- Conventional imaging performed prior to diagnosis should not be repeated unless there is a delay of at least 6 weeks since previous imaging and treatment initiation or there are new or significantly worsening clinical signs or symptoms
- Generally, the studies listed in the disease-specific sections reflect the studies supported by current literature and research for that condition. If a study is not listed, then it is not considered medically necessary.
- Routine imaging of brain, spine, neck, chest, abdomen, pelvis, bones, or other body areas is not considered medically necessary except where explicitly stated in a diagnosis-specific guideline section, or if one of the following applies:
  - Known prior disease involving the requested body area
  - New or worsening symptoms or physical exam findings involving the requested body area (including non-specific findings such as ascites or pleural effusion)
  - New finding on basic imaging study such as plain x-ray or ultrasound
  - New finding on adjacent body area CT/MRI study (i.e., pleural effusion observed on CT abdomen)
  - Unless otherwise stated in the disease-specific guideline, advanced imaging of asymptomatic individuals is not considered medically necessary without signs or symptoms of systemic involvement of cancer.

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  - Repeat imaging studies are not generally considered medically necessary unless there is evidence of disease progression, recurrence of disease, and/or the repeat imaging will affect an individual's clinical management.
- Unless otherwise stated in the diagnosis-specific guidelines, imaging for treatment response is considered medically necessary after every 2 cycles, which is usually ~6 weeks of therapy for solid tumors and usually ~8 to 12 weeks for CNS tumors
- Unless otherwise specified for a specific cancer type, once PET has been documented to be negative for a given individual's cancer or all PET-avid disease has been surgically resected, PET is not considered medically necessary for continued disease monitoring or surveillance unless one of the following applies:
  - Conventional imaging (CT, MRI, US, plain film) reveals findings that are inconclusive or suspicious for recurrence
  - Residual mass that has not changed in size since the last conventional imaging does not justify PET imaging
  - PET avidity in a residual mass at the end of planned therapy is not an indication for PET imaging during surveillance

Clarification of Phases of Therapy

Phases of Oncology Imaging	Definition
Screening	• Imaging requested for individuals at increased risk for a particular cancer in the absence of known clinical signs or symptoms • Screening using advancing imaging is only considered medically necessary for conditions listed in **Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2)**

Suspected/Initial Staging	• All imaging studies requested from the time cancer is first clinically suspected until the initiation of specific treatment
	  ◦ CT Chest prior to anesthesia for biopsy or resection of solid tumors and CTs of other involved body areas are generally considered medically necessary and should be performed concurrently
	  ◦ Metastatic CNS imaging and nuclear medicine imaging are generally deferred until after a histologic diagnosis is made, unless otherwise indicated by diagnosis-specific guideline

Phases of Oncology Imaging	Definition
Treatment Response	• Imaging performed during any type of active treatment (chemotherapy or other medications, radiation therapy, or surgery), including evaluation at the end of planned active treatment ◦ Unless otherwise stated in the diagnosis-specific guidelines, imaging for treatment response is considered medically necessary after every 2 cycles, which is usually ~6 weeks of therapy for solid tumors and usually ~8 to 12 weeks for CNS tumors
Surveillance	• Imaging performed in individuals who are asymptomatic or have chronic stable symptoms and not receiving any active treatment, even if residual imaging abnormalities are present ◦ PET imaging is not considered medically necessary for surveillance imaging unless specifically stated in elsewhere in the diagnosis-specific guideline sections
Recurrence	• All imaging studies completed at the time a recurrence or progression of a known cancer is strongly suspected or documented based on clinical signs or symptoms, laboratory findings, or results of basic imaging studies such as plain radiography or ultrasound • Following documented recurrence of childhood cancer, any studies recommended for initial staging of that cancer type in the diagnosis-specific imaging guideline section are considered medically necessary ◦ During active treatment for recurrent pediatric cancer, conventional imaging evaluation (CT or MRI, should use the same modality for ongoing monitoring as much as possible) of previously involved areas is considered medically necessary according to the treatment response imaging in the diagnosis-specific guideline section ▪ Always refer to the diagnosis specific guideline for PET indications in recurrence.

Criteria

- Brain imaging is performed for signs or symptoms of brain disease.

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  - MRI Brain without and with contrast (CPT® 70553) is the medically necessary study for evaluation of suspected or known brain metastases. If a non-contrast CT head shows suspicious lesion, MRI Brain is considered medically necessary to further characterize the lesion.
  - CT Head without and with contrast (CPT® 70470) is considered medically necessary when MRI is contraindicated or not available, or if there is skull bone involvement.
  - Certain malignancies including, but not limited to, melanoma, lung cancer, and renal cell cancer have indications for brain imaging for asymptomatic individuals.
  - If stage IV disease is demonstrated elsewhere or if systemic disease progression is noted, refer to disease specific guidelines.
  - Initiation of angiogenesis therapy is not an indication for advanced imaging of the brain in asymptomatic individuals (Avastin/Bevacizumab; <3% risk of bleeding and <1% risk of serious bleeding).

- Bone Scan:
  - Primarily used for evaluation of bone metastases in individuals with solid malignancies.
  - Indications for bone scan in individuals with history of malignancy include – bone pain, rising tumor markers, elevated alkaline phosphatase, or in individuals with primary bone tumor.
  - For evaluation of suspected or known bony metastases, CPT® 78306 (Nuclear bone scan whole-body) is considered medically necessary.
  - Radiopharmaceutical Localization scan SPECT (CPT® 78803 or CPT® 78831) or SPECT/CT (CPT® 78830 or CPT® 78832) is considered medically necessary as an add-on test for further evaluation of a specific area of interest.
- Bone scan supplemented by plain x-rays are the initial imaging modalities for suspected malignant bone pain. For specific imaging indications, see also:
  - **Bone (Non-Vertebral) Metastases (ONC-31.5)**
  - **Spinal/Vertebral Metastases (ONC-31.6)**
  - **Carcinoma of Unknown Primary Site (ONC-31.7)**
- Delay PET/CT for at least 12 weeks after completion of radiation treatment, unless required sooner for imminent surgical resection.
- PET/CT may be considered medically necessaryprior to biopsy in order to determine a more favorable site for biopsy when a prior biopsy was nondiagnostic or a relatively inaccessible site is contemplated which would require invasive surgical intervention for biopsy attempt.
- Unless otherwise specified in diagnosis-specific guideline section, PET/CT may be considered medically necessary if:

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  - Conventional imaging (CT, MRI, or bone scan) reveals findings that are inconclusive (including signs or symptoms that are highly suspicious for recurrence with no source found on imaging), with continued suspicion for active disease
- **Unless specified in diagnosis-specific guideline section PET/CT Imaging is NOT considered medically necessary for**:
  - Infection, inflammation, trauma, post-operative healing, granulomatous disease, rheumatological conditions
  - Concomitantly with separate diagnostic CT studies
  - Conclusive evidence of distant or diffuse metastatic disease on recent conventional imaging studies
  - Metastatic disease in the central nervous system (CNS)
  - Lesions less than 8mm in size
  - Follow up after localized therapy (i.e., radiofrequency ablation, embolization, stereotactic radiation, etc.)
  - Rare malignancies, due to lack of available evidence regarding the diagnostic accuracy of PET in rare cancers
  - Surveillance
    - Serial monitoring of individuals who are not currently receiving anti-tumor treatment or are receiving maintenance treatment
    - Serial monitoring of FDG avidity until resolution.
    - PET/CT avidity in a residual mass at the end of planned therapy is not an indication for PET/CT imaging during surveillance.
    - Residual mass that has not changed in size since the last conventional imaging does not justify PET imaging
- Please refer to general guidelines section **PET Imaging in Oncology (ONC-1.4)** and **PET Imaging in Pediatric Oncology (PEDONC-1.4)** for further guidance regarding PET. Those guidelines should be applied with regard to radiotracer coverage.
- Please refer to general guidelines in **Unlisted Procedure Codes in Oncology (ONC-1.5)** for unlisted procedures in pediatric oncology.

Clinical Trials

- Similar to investigational and experimental studies, clinical trial imaging requests will be considered to determine whether they meet these 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 considered medically necessary.

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Health Equity Considerations

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

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Age Considerations (PEDONC-1.1)

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- The majority of malignancies occurring in the pediatric population are different diagnoses than those occurring in the adult population.
- For those diseases which occur in both pediatric and adult populations, minor differences may exist in management between pediatric and adult medical oncologists due to the following:
  - Age of the individual
  - Comorbidities
  - Differences in disease natural history between children and adults

AGE APPROPRIATE GUIDELINES	
Age of Individual	**Appropriate Imaging Guidelines**
19 years of age or older at initial diagnosis	• General Oncology Imaging Guidelines, except where directed otherwise by a specific guideline section
Under 19 years of age at initial diagnosis	• Pediatric and Special Populations Oncology Imaging Guidelines, except where directed otherwise by a specific guideline section
15 to 39 years of age at initial diagnosis (defined as Adolescent and Young Adult (AYA) oncology individuals)	• When unique guidelines for a specific cancer type exist only in either Oncology or Pediatric and Special Populations Oncology, AYA individuals should be imaged according to the guideline section for their specific cancer type, regardless of the individual's age • When unique guidelines for a specific cancer type exist in both Oncology and Pediatric and Special Populations Oncology, AYA individuals should be imaged according to the age rule in the previous bullet

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Appropriate Clinical Evaluations (PEDONC-1.2)

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Criteria

- The majority of pediatric oncology imaging indications are listed in the diagnosis-specific guideline sections
  - Rare malignancies and other circumstances not specifically addressed elsewhere in the Pediatric Oncology guidelines, the following general principles apply:
    - Routine imaging of brain, spine, neck, chest, abdomen, pelvis, bones, or other body areas is not considered medically necessary in the absence of:
      - Localizing symptoms or
      - Abnormalities on plain radiography or ultrasound
- The overwhelming majority of pediatric oncology individuals treated in the United States will be enrolled on or treated according to recent Children's Oncology Group (COG) protocols
  - COG is a National Cancer Institute (NCI) supported clinical trials group
  - These imaging guidelines are generally consistent with evaluations recommended by COG protocols commonly used for direct individual care (whether formally enrolled on study or not)
    - Requests for exception to guidelines based on COG protocol should be accompanied by the COG road map or COG details in the medical record for case-by-case consideration.
- For individuals enrolled in a COG study, imaging recommended by COG protocols is generally considered medically necessary unless the imaging is being performed solely to address a study objective and would not be considered medically necessary in usual clinical care
  - Requests for exception to guidelines based on COG protocol should be accompanied by the COG road map or COG details in the medical record for case-by-case consideration.

Phases of Pediatric Oncology Imaging:

- Pediatric malignancies, in general, behave more aggressively than adult cancers, and the time from initial suspicion of cancer to specific therapy initiation can be measured in hours to days for most pediatric cancers
- It is recommended that children with pediatric solid tumors undergo CT evaluation of the chest prior to general anesthesia for biopsy or resection due to the risk of post-

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operative atelectasis mimicking pulmonary metastasis resulting in inaccurate staging and/or delay in therapy initiation
- Unlike adult cancers, in most pediatric cancers surveillance does not begin until all planned multi-modal therapy is completed
- Pediatric cancers where surgical resection is considered curative are listed in the diagnosis-specific guideline sections
- Certain tumor types do not require surveillance with advanced imaging as individual outcomes following relapse are not improved by surveillance imaging. See diagnosis-specific guideline sections for details
- Imaging may be considered medically necessary more frequently than recommended by guidelines with clinical documentation that the imaging results are likely to result in a treatment change for the individual, including a change from active treatment to surveillance

Phases of Oncology Imaging	Definition
Screening	• Imaging requested for individuals at increased risk for a particular cancer in the absence of known clinical signs or symptoms • Screening using advancing imaging is only considered medically necessary for conditions listed in **Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2)**
Suspected/Initial Staging	• All imaging studies requested from the time cancer is first clinically suspected until the initiation of specific treatment ◦ CT Chest prior to anesthesia for biopsy or resection of solid tumors and CTs of other involved body areas are generally considered medically necessary and should be performed concurrently ◦ Metastatic CNS imaging and nuclear medicine imaging are generally deferred until after a histologic diagnosis is made, unless otherwise indicated by diagnosis-specific guideline

Phases of Oncology Imaging	Definition
Treatment Response	• Imaging performed during any type of active treatment (chemotherapy or other medications, radiation therapy, or surgery), including evaluation at the end of planned active treatment ◦ Unless otherwise stated in the diagnosis-specific guidelines, imaging for treatment response is considered medically necessary after every 2 cycles, which is usually ~6 weeks of therapy for solid tumors and usually ~8 to 12 weeks for CNS tumors
Surveillance	• Imaging performed in individuals who are asymptomatic or have chronic stable symptoms and not receiving any active treatment, even if residual imaging abnormalities are present ◦ PET imaging is not considered medically necessary for surveillance imaging unless specifically stated in elsewhere in the diagnosis-specific guideline sections
Recurrence	• All imaging studies completed at the time a recurrence or progression of a known cancer is strongly suspected or documented based on clinical signs or symptoms, laboratory findings, or results of basic imaging studies such as plain radiography or ultrasound • Following documented recurrence of childhood cancer, any studies recommended for initial staging of that cancer type in the diagnosis-specific imaging guideline section are considered medically necessary ◦ During active treatment for recurrent pediatric cancer, conventional imaging evaluation (CT or MRI, should use the same modality for ongoing monitoring as much as possible) of previously involved areas is considered medically necessary according to the treatment response imaging in the diagnosis-specific guideline section ▪ Always refer to the diagnosis specific guideline for PET indications in recurrence.

Cardiac Function Assessment Criteria in Pediatric Oncology During Active Treatment:

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Indication	Medically Necessary Imaging Study
• Evaluation of cardiac function prior to cardiotoxic chemotherapy, and for monitoring while on active therapy or at end of therapy at the discretion of the treating pediatric oncologist based on: ◦ Cumulative cardiotoxic therapy received to date ◦ Individual's age and gender ◦ Most recent echocardiogram results ◦ New or worsening cardiac symptoms	• Echocardiography (CPT® 93306, CPT® 93307, or CPT® 93308)
• For either of the following: ◦ Echocardiography yielded a borderline shortening fraction (<30%) and additional left ventricular function data are necessary to make a chemotherapy decision OR ◦ Echocardiography windowing is suboptimal due to body habitus or tumor location	• Multi-gated acquisition (MUGA, CPT® 78472) blood pool nuclear medicine scanning

Immunosuppression Criteria During Pediatric Cancer Therapy and Imaging Ramifications:

- Individuals may be severely immunocompromised during active chemotherapy treatment and any conventional imaging request to evaluate for infectious complications during this time frame is considered medically necessary
  - Imaging requests for infectious disease concerns for all individuals with absolute neutrophil count (ANC) <500 **or**
  - Inconclusive findings on chest x-ray or US at any ANC during active treatment are considered medically necessary as requested
  - Individuals may have therapy-induced hypogammaglobulinemia which requires supplemental intravenous immune globulin (IVIG) during maintenance therapy. These individuals receiving supplemental IVIG should be treated similarly to individuals with ANC <500 with regards to imaging for infectious disease

- Some individuals are treated with very intensive chemotherapy regimens (including autologous stem cell transplantation) and spend the majority of their chemotherapy treatment phase in the hospital. See: **Hematopoietic Stem Cell Transplantation (HSCT) (PEDONC-20)**.
- Due to the high risk of invasive infections, frequent CT may be considered medically necessary to evaluate known sites of invasive fungal infection, and in general these are considered medically necessary as requested
  - Surveillance imaging of asymptomatic individuals to detect invasive fungal infection has not been shown to impact individual outcomes
    - Imaging requests are considered medically necessary when acute clinical decisions will be made based on the imaging

Hematopoietic Stem Cell Transplant (HSCT) Criteria in Pediatric Oncology:

- Transplantation of hematopoietic stem cells from bone marrow, peripheral blood, or cord blood is commonly used in the following clinical situations:
  - High-risk or recurrent leukemia (allogeneic)
  - Recurrent lymphoma (allogeneic or autologous)
  - Hemophagocytic lymphohistiocytosis (allogeneic)
  - High-risk sickle cell disease (allogeneic)
  - High-risk neuroblastoma (autologous)
  - High-risk CNS tumors (autologous)
  - Recurrent Ewing sarcoma family of tumors (autologous, rarely allogeneic)
- Imaging considerations for HSCT should follow guidelines in: **Hematopoietic Stem Cell Transplantation (HSCT) (PEDONC-20)**.

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Modality General Considerations (PEDONC-1.3)

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Criteria

- Plain radiography
  - Chest x-ray (CXR) can provide a prompt means to evaluate primary intrathoracic tumors and continues to be the initial imaging study recommended to detect complications, such as suspected infection, in symptomatic individuals undergoing treatment.
  - CXR continues to be the initial imaging study recommended for pulmonary surveillance for some pediatric cancers. See diagnosis-specific guideline sections for details.
  - Plain radiography continues to be the initial imaging study recommended for evaluation of lesions involving the appendicular skeleton, both during and after completion of treatment. See diagnosis-specific guideline sections for details.
  - Plain abdominal radiographs have largely been replaced by ultrasound, CT, or MRI.
- Ultrasound
  - Ultrasound is not widely used in pediatric oncology for staging but is frequently used for surveillance in individuals who have successfully treated (primarily abdominal or pelvic) tumors with little or no residual disease.
  - See diagnosis-specific guideline sections for details.
- CT
  - CT with contrast is the imaging study of choice in pediatric individuals with lymphomas or solid tumors of the neck, thorax, abdomen, and/or pelvis
    - If CT contrast use is contraindicated due to allergy or impaired renal function, either CT without contrast or MRI without and with contrast may be substituted at the discretion of the ordering physician
- MRI
  - MRI without and with contrast is the study of choice for CNS tumors and musculoskeletal tumors
    - If MRI contrast use is contraindicated due to allergy or impaired renal function, MRI without contrast may be substituted at the discretion of the ordering physician
  - Due to the length of time for image acquisition and the need for stillness, anesthesia is required for almost all infants and young children (age <7 years), as

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well as older children with delays in development or maturity. In this population, MRI imaging sessions should be planned with a goal of avoiding a short-interval repeat anesthesia exposure due to insufficient information using the following considerations:

- MRI should generally be performed without and with contrast unless there is a specific contraindication to gadolinium use since the individual already has intravenous access for anesthesia
  - 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 requesting clinicians indicate that a non-contrast study is being requested due to concerns regarding the use of gadolinium, the exam is considered medically necessary

◦ In some instances, to reduce time under anesthesia for MRI in pediatric individuals, surveillance or restaging studies may be requested with contrast only when it is determined that repeat non-contrast imaging does not add to an individual clinical case. These are considered medically necessary on a case-by-case basis.
  - If multiple body areas are supported by these guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session

• Nuclear medicine
◦ General PET imaging consideration can be found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**.
◦ Bone scan is frequently used for evaluation of bone metastases in pediatric oncology during initial treatment, treatment response, and surveillance
  - CPT® 78315 has no specific indications for evaluation of malignant disease
◦ ^{123}I-metaiodobenzylguanidine (MIBG) scintigraphy is the preferred metabolic imaging for neuroblastoma and is positive in 90% to 95% of neuroblastomas. MIBG is also used for evaluation of pheochromocytomas, paragangliomas, ganglioneuromas, and ganglioneuroblatomas, PET/CT indications are provided in the relevant sections.

Study Type	Coding
Bone scan	• ANY of the following codes can be approved: ◦ CPT® 78300 ◦ CPT® 78305 ◦ CPT® 78306 ◦ CPT® 78803, 78830, or 78832 ▪ May be approved alone or in combination with: - CPT® 78305 - CPT® 78306
^{123}I-metaiodobenzylguanidine (MIBG) scintigraphy	• ANY one of the following codes can be approved: ◦ CPT® 78801 ◦ CPT® 78802 ◦ CPT® 78804 • In addition to the above imaging, **ONE** of the following codes is considered medically necessary: ◦ CPT® 78803 ◦ CPT® 78830 ◦ CPT® 78831 ◦ CPT® 78832 **OR** ◦ CPT® 78800 may be approved for KNOWN neuroblastoma when only a single site follow up is desired but is not sufficient for the initial workup of suspected disease
Octreotide scan	Same coding as MIBG
Gallium scan	Same coding as MIBG

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PET Imaging in Pediatric Oncology (PEDONC-1.4)

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Throughout these guidelines, unless otherwise specified, the term "PET" refers specifically to ^{18}F-FDG-PET imaging and also applies to PET/CT fusion studies.

Criteria

- PET imaging in pediatric oncology should use PET/CT fusion imaging (CPT® 78815 or CPT® 78816)
- The decision whether to use skull base to mid-femur ("eyes to thighs") procedure code for PET (CPT® 78815) or Whole-body PET (CPT® 78816) is addressed in the diagnosis-specific guideline sections
- PET imaging in oncology should use PET/CT fusion imaging (CPT® 78815 or CPT® 78816)
- PET/MRI is generally not considered medically necessary for a vast majority of oncologic conditions due to lack of standardization in imaging technique and interpretation. However, it is considered medically necessary in select circumstances when all of the following criteria are met:
  - The individual meets condition-specific guidelines for PET/MRI OR
  - The individual meets ALL of the following:
    - 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 is considered medically necessary using the code combination of PET Whole-Body (CPT® 78813) and MRI Unlisted (CPT® 76498). All other methods of reporting PET/MRI are inappropriate.
    - When clinically appropriate, diagnostic MRI codes may be considered medically necessary at the same time as the PET/MRI code combination.
- Unbundling PET/CT imaging into separate PET (such as CPT® 78812 or CPT® 78813) and diagnostic CT codes is otherwise not considered medically necessary.
- PET imaging is not reliable for the detection of anatomic lesions smaller than 8mm in size.
- Delay PET/CT for at least 12 weeks after completion of radiation treatment, unless required sooner for imminent surgical resection.
- PET imaging using isotopes other than ^{18}F-FDG, ^{68}Ga-DOTATATE, or ^{68}Ga-DOTATOC is considered not medically necessary at this time. Please see the table below for the most commonly used isotopes and their corresponding codes

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- Covered:
  - 18F-FDG
  - 68Ga-DOTATATE (NETSPOT®) for low grade neuroendocrine tumors
  - 68Ga-DOTATOC for low grade neuroendocrine tumors Not covered:
  - PET/CT imaging using isotopes other than those specified above

CPT/ HCPCS Code	Code Description	Brand or common name	FDA-approved?	Code reviewed by EviCore for Cigna?
A9552	fluorine-18 (F-18) fluorodeoxyglucose (FDG), diagnostic, per study dose, up to 45 millicuries	FDG	Yes, to assess abnormal glucose metabolism	No
A9587	Gallium GA-68, dotatate, diagnostic, 0.1 millicurie	NETSPOT®	Yes, for localization of somatostatin receptor positive neuroendocrine tumors in adult and pediatric population	No
C9067	^{68}Ga Gallium-DOTA-TOC	N/A	Yes, for somatostatin receptor (SSTR) positive gastroentero-pancreatic neuroendocrine tumors	No

- PET has not been shown to be diagnostically useful in all forms of childhood cancer. PET is considered medically necessary for pediatric malignancies with significant published evidence regarding its diagnostic accuracy and importance in accurately

directing individual care decisions. See diagnosis-specific guideline sections for details.

- PET imaging is not specific to cancer, and has a high rate of false positivity. Inflammation, infection (especially granulomatous), trauma, and post-operative healing may show high levels of FDG uptake and be false-positive for malignant lesions.
- PET for rare malignancies not specifically addressed by these guidelines is generally not considered medically necessary due to lack of available evidence regarding diagnostic accuracy of PET in the majority of rare cancers. Conventional imaging studies should be used for initial staging and treatment response for these diagnoses.
- Unless otherwise specified in diagnosis-specific guideline section, PET is considered medically necessary if **all** of the following apply:
  - Conventional imaging (CT, MRI, US, plain film) reveals findings that are inconclusive (including signs or symptoms that are highly suspicious for recurrence with no source found on imaging), with continued suspicion for active disease
  - No other specific metabolic imaging (MIBG, octreotide, technetium, etc.) is considered medically necessary for the cancer type
  - The submitted clinical information describes a specific decision regarding the individual's care that will be made based on the PET results
- PET imaging for surveillance imaging **only** when specifically stated elsewhere in the diagnosis-specific guideline sections
- Unless otherwise specified for a specific cancer type, once PET has been documented to be negative for a given individual's cancer or all PET-avid disease has been surgically resected, PET is not considered medically necessary for continued disease monitoring or surveillance unless **one** of the following applies:
  - Conventional imaging (CT, MRI, US, plain film) reveals findings that are inconclusive or suspicious for recurrence
    - Residual mass that has not changed in size since the last conventional imaging does **not** justify PET imaging
    - PET avidity in a residual mass at the end of planned therapy is **not** an indication for PET imaging during surveillance.
  - Very rare circumstances where tumor markers or obvious clinical symptoms show strong evidence suggesting recurrence and PET would replace conventional imaging modalities
  - The individual is undergoing salvage treatment for a recurrent solid tumor with residual measurable disease on conventional imaging and confirmed repeat negative PET imaging will allow the individual to transition from active treatment to surveillance

Diagnostic Radiation Exposure in Pediatric Oncology (PEDONC-1.5)

ONCP.GG.0001.5.A
v1.0.2027

Criteria

- Young children are presumed to be at increased risk for malignancy from diagnostic radiation exposure, most commonly from CT and nuclear medicine imaging. They are more sensitive to radiation than adults and generally live longer after receiving radiation doses from medical procedures, resulting in a larger number of years during which to manifest a cancer.
- Because of this presumed increased risk in young children, requests to substitute MRI without and with contrast for CT with contrast to avoid radiation exposure are considered medically necessary if ALL of the following criteria apply:
  - The individual is presently a young child and the ordering physician has documented the reason for MRI, rather than CT, is to avoid radiation exposure.
  - The disease-specific guidelines do not list CT as superior to MRI for the current disease and time point, meaning the MRI will provide equivalent or superior information relative to CT.
  - The request is for a body area other than Chest as MRI is substantially inferior to CT for detection of small pulmonary metastases.
- The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

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Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2)

Guideline Page

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Screening Imaging in Cancer Predisposition Syndromes – General Considerations (PEDONC-2.1)

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Criteria

- This section is intended to give guidance for screening imaging prior to diagnosis with a specific malignancy. Once an individual with a cancer predisposition syndrome has been diagnosed with a malignant disease, future imaging decisions should be guided by the appropriate disease-specific guidelines except as explicitly stated elsewhere in this section.
- This section's guidelines are limited to cancer predisposition syndromes with screening imaging considerations. Syndromes requiring only clinical or laboratory screening are not discussed here.
- In general, a relevant detailed history and physical examination and appropriate laboratory studies should be performed prior to considering advanced imaging, unless the individual is undergoing guideline-supported scheduled screening evaluation identified in this section.
- Where MRI is indicated in these guidelines, CT is considered medically necessary only if MRI is contraindicated, given the risk of radiation exposure in these syndromes.
- Many of these cancer predisposition syndromes also affect adults as survival continues to improve for these individuals.
    - Adults with syndromes covered in this section may follow these imaging guidelines except where contradicted by specific statements in the general imaging guidelines.
- In some instances, to reduce time under anesthesia for MRI in pediatric individuals, surveillance or restaging studies may be requested with contrast only when it is determined that repeat non-contrast imaging does not add to an individual clinical case. These are considered medically necessary on a case-by-case basis.
- If a given genetic syndrome does not have documented evidence-based criteria for whole-body MRI as noted in these guidelines, then whole-body MRI is not considered medically necessary.

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Li-Fraumeni Syndrome (LFS) (PEDONC-2.2)

ONCP.SC.0002.2.A
v1.0.2027

Criteria

When a specific malignancy is suspected, the individuals should be imaged according to the imaging guideline specific to the suspected cancer type.

The following imaging studies should be considered medically necessary in individuals with LFS:

Indication	Medically Necessary Imaging Study
All individuals with LFS	• BOTH of the following, annually: ◦ MRI Brain without and with contrast (CPT® 70553) ◦ Whole-body MRI (WBMRI, CPT® 76498) ▪ CPT® 76498 is the only code for whole-body MRI at this time. • Every 3 months from birth: ◦ Abdominal (CPT® 76700) and pelvic (CPT® 76856) ultrasound • Beginning at age 20: ◦ Annual Breast MRI (CPT® 77049), alternating every 6 months with breast ultrasound, in addition to the previously noted annual MRI studies.
Individuals with LFS with documented signs or symptoms suggestive of possible malignancy	• Targeted MRI without and with contrast of the involved body area(s)

Background and Supporting Information

- LFS is a hereditary cancer predisposition syndrome characterized by autosomal dominant inheritance, with a 50% risk of transmission to offspring, caused by germline pathogenic variants in the TP53 tumor suppressor gene (Schneider et al., 2025).
- LFS is associated with markedly increased lifetime cancer risk across multiple organ systems, with early onset of malignancy common. Core tumor types include

adrenocortical carcinoma, central nervous system tumors, osteosarcoma, soft tissue sarcomas, and early onset breast cancer (Schneider et al., 2025).
  - Additional malignancies relevant to imaging surveillance include leukemia, colorectal cancer, lung cancer, melanoma, pancreatic cancer, and prostate cancer, with affected individuals at increased risk for developing multiple primary cancers over their lifetime (Schneider et al., 2025).
- The majority of individuals with LFS have germline TP53 pathogenic variants. In contrast, somatic (tumor-specific) TP53 variants are common in sporadic malignancies and do not confer inherited cancer risk or eligibility for LFS-specific imaging surveillance protocols (Schneider et al., 2025).
  - Accurate distinction between germline and somatic TP53 alterations is necessary to determine appropriateness of screening and surveillance imaging strategies (Schneider et al., 2025).
- In individuals who meet clinical diagnostic criteria for LFS but do not have an identifiable germline TP53 pathogenic variant, formal diagnosis should be assigned by a physician with significant training and experience in LFS, typically a geneticist or oncologist, prior to initiation of a screening imaging program (Schneider et al., 2025).

Evidence Discussion

- Individuals with LFS have increased susceptibility to radiation-induced malignancies due to defective TP53 mediated DNA damage response, and exposure to ionizing radiation may increase the risk of secondary cancers. As a result, routine surveillance strategies that rely on ionizing radiation, including CT and most nuclear medicine studies, are not considered medically necessary for ongoing screening in LFS (Schneider et al., 2025).
- Whole body MRI and brain MRI provide broad, multi-organ cancer detection without ionizing radiation and are therefore preferred surveillance modalities in individuals with LFS. MRI-based surveillance enables detection of a wide spectrum of tumor types characteristic of LFS and aligns with the need for lifelong, repeat imaging in this population. Prospective cohort studies of individuals with LFS undergoing MRI-based surveillance have demonstrated earlier cancer detection and improved survival outcomes compared with historical controls (Schneider et al., 2025).
  - Recognized limitations of MRI-based surveillance include false positive findings, increased follow up imaging or procedures, and the need for sedation in younger children; however, these risks are generally outweighed by the benefit of early detection in this high risk population (Schneider et al., 2025).

Health Equity Considerations

LFS is a rare hereditary cancer predisposition syndrome caused by pathogenic germline TP53 variants and is associated with an exceptionally high lifetime risk of developing multiple, early onset malignancies, often beginning in childhood. Families affected by

LFS face disproportionate challenges related to access to timely genetic diagnosis, specialized surveillance programs, and pediatric-specific expertise. Comprehensive surveillance protocols, often initiated from birth and including frequent whole body and brain MRI, have demonstrated survival benefit but require advanced imaging infrastructure, pediatric sedation services, and multidisciplinary coordination that may not be uniformly available across care settings (Kiermeier et al., 2025). Delays in diagnosis or limited access to recommended surveillance can exacerbate disparities, particularly for families in rural areas, lower resource health systems, or those with limited health literacy navigating complex hereditary cancer care pathways. These structural barriers may result in missed early cancers or inconsistent longitudinal imaging, contributing to unequal outcomes despite shared genetic risk.

In addition to structural access issues, psychosocial burden represents a critical and often underrecognized equity consideration in pediatric and special populations with LFS. Evidence indicates that individuals with LFS, including adolescents, young adults, parents, and "previvors" without a prior cancer diagnosis, experience high levels of distress, fear of progression, and impaired health-related quality of life associated with lifelong cancer risk and intensive surveillance regimens (Kiermeier et al., 2025). These psychological stressors may influence adherence to imaging surveillance and follow up recommendations, particularly in families facing cumulative socioeconomic stress or limited access to integrated psychosocial support. Consistent application of evidence based, radiation-sparing imaging strategies supports equity by standardizing high value surveillance while minimizing harm. Integrating guideline directed imaging with coordinated genetic counseling and psychosocial resources promotes equitable care delivery for children and families living with LFS, ensuring that survival benefits of early detection are not undermined by disparities in access, support, or continuity of care (Kiermeier et al., 2025).

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Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)

ONCP.SC.0002.3.C
v1.0.2027

Criteria

- Considerations for PET/CT coding and indications/coding for PET/MRI are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

The following imaging studies should be considered medically necessary in individuals with NF1:

Indication	Medically Necessary Imaging Study
• Clarification of the diagnosis of NF1 if evaluation by a physician with significant training and/or experience in neurofibromatosis is inconclusive ◦ Most commonly a neurologist, geneticist, ophthalmologist, or oncologist	• One-time MRI Brain (CPT® 70553) and Orbits (CPT® 70543) without and with contrast
• New or worsening neurological or visual symptoms in individuals with NF1	• MRI Brain (CPT® 70553) and Orbits (CPT® 70543) without and with contrast
• Clinical symptoms suggestive of change in a known plexiform neurofibroma in individuals with NF1 ◦ Examples include: pain, rapid growth, and neurologic dysfunction	• MRI without and with contrast
• Clinical symptoms concerning for malignant transformation of a known plexiform neurofibroma in individuals with NF1, and ALL of the following are met: ◦ Recent MRI is inconclusive regarding transformation or progression ◦ Negative PET will result in a decision to avoid biopsy in a difficult or morbid location	• PET/CT (CPT® 78815 or CPT® 78816)

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Indication	Medically Necessary Imaging Study
• Baseline tumor burden in individuals with NF1 at age 16 or older (one-time imaging)	• WBMBRI (CPT® 76498)
• New soft tissue mass(es) in individuals with NF1	• See: Soft tissue mass information in **Soft Tissue Mass and Morton's Neuroma (MS 10.3)** in the Musculoskeletal Imaging Guidelines or **Soft Tissue and Bone Masses - General Considerations (PEDMS 3.1)** in the Pediatric Musculoskeletal Imaging Guidelines depending on the individual's age at the time the mass is discovered. Plain x-ray or ultrasound is not required prior to advanced imaging in these individuals.
• New bone mass(es) in individuals with NF1	• See: **Bone Tumors - General Considerations (PEDONC-9.1)**
• Documented optic pathway gliomas in individuals with NF1	• See: **Intracranial Low Grade Gliomas (PEDONC-4.2)**
• Known plexiform neurofibromas in individuals with NF1	• MRI without and with contrast of a known body area containing a neurofibroma is indicated for any of the following: ◦ Every 3 months for treatment response in individuals receiving active treatment ◦ New or worsening clinical symptoms suggesting progression ◦ Preoperative planning
• Biopsy-proven MPNST in individuals with known NF1 or NF2	• For adult individuals, see: **Sarcomas - Bone, Soft Tissue, and GIST (ONC-12)** • For pediatric individuals, see: **Non-Rhabdomyosarcoma Soft Tissue Sarcomas (NRSTS) (PEDONC-8.3)**

- Annual ophthalmology evaluation is strongly recommended beginning at the time of diagnosis of NF1 to evaluate for optic pathway abnormalities:

- Screening MRIs Brain (CPT® 70553) and Orbits (CPT® 70543) for asymptomatic individuals with NF1 are not generally recommended due to the ~60% rate of unidentified bright objects (UBOs, T2-weighted signal abnormalities) which mostly disappear by age 30
  - Routine follow up imaging of UBOs is not considered medically necessary in the absence of acute symptoms suggesting new or worsening intracranial disease in individuals with NF1

Background and Supporting Information

- NF1 is a common hereditary multisystem tumor predisposition syndrome inherited in an autosomal dominant manner, with a 50% risk of transmission to offspring. Approximately 50% of affected individuals have de novo pathogenic variants. Penetrance is close to 100%, though clinical severity is highly variable (Friedman, 2025).
- NF1 affects approximately 1 in 2,000–2,500 individuals, making it one of the most common autosomal dominant genetic disorders (Friedman, 2025).
- NF1 is associated with an increased risk of benign and malignant tumors, many of which may be internal or asymptomatic (Friedman, 2025), including:
  - Plexiform neurofibromas, which are frequently internal and best characterized by MRI
  - Optic pathway gliomas, most commonly occurring in early childhood
  - Non optic central nervous system gliomas
  - MPNST, which typically arise from pre existing plexiform neurofibromas and may demonstrate imaging features suggestive of malignant transformation
- Individuals with NF1 are at increased risk for developing multiple tumors over time (Friedman, 2025).
- Although many NF1 associated tumors are benign, malignant transformation can occur, particularly to MPNST, which represents a major cause of morbidity and mortality in this population (Friedman, 2025).

Evidence Discussion

- A substantial proportion of NF1-associated tumors are not apparent on physical examination and may be detected incidentally or through targeted imaging performed for clinical indications (Friedman, 2025).
- Individuals with NF1 have increased sensitivity to ionizing radiation, with elevated risks of radiation associated malignancies and vasculopathy. Therefore, CT and nuclear medicine studies are not considered medically necessary for routine screening or surveillance, and MRI is preferred when imaging is clinically indicated (Friedman, 2025).

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- Individuals with NF1 are at increased risk for plexiform neurofibromas and MPNST; however, screening imaging of asymptomatic individuals is not considered medically necessary, as malignant transformation is typically signaled by clinical changes such as persistent pain, rapid growth, or new neurologic deficits rather than imaging findings alone (Friedman, 2025).
- FDG-PET imaging may be useful when malignant transformation to MPNST is clinically suspected, as the negative predictive value is high (approximately 96%–99%), although the positive predictive value is limited (approximately 60%–65%), even in symptomatic patients (Friedman, 2025).
  - PET imaging is not considered medically necessary for routine surveillance of plexiform neurofibromas (Friedman, 2025).
  - Inconclusive PET findings should prompt tissue biopsy; repeat PET imaging is not considered medically necessary due to limited additional diagnostic yield (Friedman, 2025).
- AACR recommendations support a single baseline whole body MRI in late adolescence or young adulthood to establish tumor burden. Subsequent imaging should be targeted and symptom driven; routine serial whole body MRI surveillance is not considered medically necessary (Evans et al., 2017).

Health Equity Considerations

NF1 is a common inherited cancer predisposition syndrome affecting approximately 1 in 2,000–3,000 individuals, with clinical manifestations that often begin in early childhood and evolve over time. Children with NF1 face disproportionate challenges related to access to specialized care, timely imaging, and consistent long term surveillance for tumor related complications. NF1 management frequently requires advanced imaging, including MRI for evaluation of optic pathway gliomas, plexiform neurofibromas, and other central or peripheral nervous system tumors. However, access to specialty neurofibromatosis clinics and receipt of guideline-recommended surveillance is uneven, with documented disparities by geography, socioeconomic status, insurance type, and race/ethnicity, particularly in the United States (Merker et al., 2025). Children who do not receive care through specialized NF clinics are less likely to complete recommended surveillance evaluations, increasing the risk of delayed diagnosis and potentially preventable morbidity (Merker et al., 2025).

Beyond access to specialty services, the clinical heterogeneity of NF1 contributes to inequities in recognition, imaging utilization, and care coordination. Pediatric patients with plexiform neurofibromas experience substantial clinical and humanistic burden, including chronic pain, disfigurement, functional impairment, and reduced health-related quality of life across physical, emotional, social, and school domains (Yang et al., 2022; Iheanacho et al., 2022). Disease severity and visibility are associated with worse quality of life outcomes and increased psychosocial stress, particularly in children with more extensive or symptomatic disease (Cavallo et al., 2024). Families with

fewer resources may face additional barriers navigating repeated imaging, prolonged surveillance, surgical consultations, and emerging targeted therapies, compounding disparities over time. Standardized, evidence-based imaging recommendations support equity by promoting appropriate, risk aligned surveillance while avoiding unnecessary imaging in lower risk scenarios. Consistent application of these guidelines, coupled with improved access to specialty expertise and psychosocial support, is essential to ensure that children with NF1 receive equitable, high quality care regardless of disease visibility, geographic location, or socioeconomic context (Merker et al., 2025; Cavallo et al., 2024).

Criteria

The following imaging studies should be considered medically necessary in individuals with NF2:

Indication	Medically Necessary Imaging Study
• All individuals with NF2, beginning at age 10 years	• Annual MRI Brain without and with contrast (CPT® 70553)
• All individuals with NF2 without a history of spinal tumors, beginning at age 10 years	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) annually for 2 years, then every 3 years
• All individuals with NF2 with a history of spinal tumors	• Annual MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
• Clinical symptoms of intracranial mass or vestibular disease in individuals with NF2	• MRI Brain without and with contrast (CPT® 70553)
• Any of the following in individuals with NF2: ◦ Clinical symptoms suggestive of new or progressive spinal or paraspinal tumors, including uncomplicated back pain or radiculopathy ◦ Recent diagnosis with a meningioma or vestibular schwannoma	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
• Surveillance for progression of unresected tumors in individuals with NF2 with known vestibular schwannoma	• Annual MRI Brain without and with contrast (CPT® 70553)

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Indication	Medically Necessary Imaging Study
• Known vestibular schwannomas and any of the following in individuals with NF2: ◦ New or worsening clinical symptoms including hearing loss ◦ Preoperative planning	• MRI Brain without and with contrast (CPT® 70553)
• Known meningioma in individuals with NF2	• See: **Meningiomas (ONC-2.8)** in the Oncology Imaging Guidelines
• Known ependymoma in individuals with NF2	• See: **Ependymoma (PEDONC-4.8)**

Background and Supporting Information

- NF2 is a hereditary tumor predisposition syndrome distinct from NF1 and substantially less common. It is inherited in an autosomal dominant manner, with up to a 50% risk of transmission to offspring. Approximately 50% of individuals have de novo pathogenic variants, and somatic mosaicism is common. Penetrance is close to 100% over a lifetime (Evans, 2023).
- The estimated prevalence of NF2 is approximately 1 in 50,000, with a birth incidence of about 1 in 28,000 (Evans, 2023).
- NF2 is characterized by a high burden of intracranial and spinal tumors, many of which are progressive and require longitudinal imaging assessment (Evans, 2023), including:
  - Bilateral vestibular schwannomas, present in approximately 85%–90% of affected individuals and nearly universal by age 30
  - Intracranial meningiomas, occurring in approximately 50% of individuals, with higher lifetime risk
  - Spinal tumors, present in at least two thirds of individuals, including intradural extramedullary schwannomas and intramedullary tumors
  - Ependymomas, occurring in approximately 25% of individuals
- NF2-associated tumors are often multiple, bilateral, and slow growing, and may be asymptomatic at the time of detection (Evans, 2023).
- Tumor burden and distribution vary widely among individuals, and multiple tumor types may coexist, supporting the need for comprehensive neuroaxis evaluation when NF2 is present or suspected (Evans, 2023).

Evidence Discussion

- MRI is the preferred imaging modality for evaluation and surveillance in NF2 due to the high lifetime risk of central nervous system tumors and the need for repeated longitudinal imaging (Evans, 2023).
- Annual brain MRI beginning in late childhood (approximately age 10 years) and continuing through at least early adulthood is considered medically necessary for affected and at risk individuals, as tumors are frequently asymptomatic in early stages and optimal outcomes depend on detection prior to neurologic compromise (Evans, 2023).
- Spinal MRI is considered medically necessary when clinically indicated (e.g., new neurologic deficits, pain, or pre procedural planning). Routine whole spine screening in asymptomatic individuals is not universally considered medically necessary, as many spinal tumors remain indolent and clinically silent (Evans, 2023).
- CT and other ionizing radiation-based imaging should be minimized, particularly in children and young adults with NF2, due to increased risks of radiation-induced malignant transformation and secondary central nervous system malignancies. Radiation-based therapies and repeated radiation imaging are associated with elevated long term risks of malignant progression in NF2-associated tumors and should be avoided for surveillance when MRI provides adequate diagnostic information (Evans, 2023).
    - MRI is preferred over CT for pre-operative planning and longitudinal assessment of vestibular schwannomas, meningiomas, and spinal tumors, given superior soft tissue characterization and avoidance of cumulative radiation exposure (Evans, 2023).

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Beckwith-Wiedemann Syndrome (BWS) (PEDONC-2.4)

ONCP.SC.0002.4.A
v1.0.2027

Criteria

Individuals with isolated hemihypertrophy are also imaged according to this guideline.

The following imaging studies should be considered medically necessary in individuals with BWS:

Indication	Medically Necessary Imaging Study
• All individuals with BWS, from birth to the 8th birthday	• Abdominal ultrasound (CPT® 76700) every 3 months
• Purely cystic adrenal mass found on screening ultrasound in individuals with BWS	• Continue screening ultrasound (CPT® 76700 or 76705) every 3 months without additional imaging (i.e., advanced imaging is not considered medically necessary for a purely cystic adrenal mass)
• Solid or mixed adrenal mass found on screening ultrasound in individuals with BWS AND: ◦ Individual age 0 months to 5 months, and ◦ Mass 0cm to 3cm in diameter	• MIBG imaging (See: **PEDONC-1.3** for coding) and either CT or MRI Abdomen (contrast as requested)
• Solid or mixed adrenal mass found on screening ultrasound in individuals with BWS AND: ◦ Individual age 0 to 5 months, and ◦ Mass >3cm in diameter	• MIBG imaging (See: **PEDONC-1.3** for coding) and MRI Abdomen (contrast as requested)
• Solid or mixed adrenal masses on screening ultrasound in individuals with BWS AND: ◦ Individual age 6 months or greater	• MIBG imaging (See: **PEDONC-1.3** for coding) prior to biopsy or resection ◦ If no evidence of malignancy on biopsy or resection, resume screening abdominal ultrasound every 3 months

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Indication	Medically Necessary Imaging Study
• Solid or mixed adrenal masses on screening ultrasound in individuals with BWS AND: ◦ No evidence of malignancy based on MIBG, CT or MRI, Urine HVA/ VMA, and serum ACTH	• Repeat abdominal ultrasound (CPT® 76700 or 76705) every 6 weeks for 2 years
• Known renal tumors in individuals with BWS	• See: **Pediatric Renal Tumors (PEDONC-7)**
• Known hepatoblastoma in individuals with BWS	• See: **Hepatoblastoma (PEDONC-11.2)**
• Known neuroblastoma in individuals with BWS	• See: **Neuroblastoma (PEDONC-6)**
• Known adrenocortical carcinoma in individuals with BWS	• See: **Pediatric Adrenocortical Carcinoma (PEDONC-14)**
• Known pheochromocytoma in individuals with BWS	• See: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines

Background and Supporting Information

- BWS, also referred to as Beckwith Wiedemann spectrum, is a congenital overgrowth disorder characterized by variable features including macroglossia, macrosomia, hemihyperplasia (lateralized overgrowth), visceromegaly, omphalocele, and neonatal hypoglycemia. Clinical expression is heterogeneous, and affected individuals may demonstrate only one or a subset of characteristic findings (Shuman et al., 2023).
- BWS is caused by epigenetic and/or genomic alterations involving imprinted genes on chromosome 11p15.5, leading to dysregulated growth factor expression. The condition is most commonly sporadic and frequently associated with somatic mosaicism, although rare familial cases occur depending on the underlying molecular mechanism (Shuman et al., 2023).
- Children with BWS are at increased risk for embryonal tumors, particularly Wilms tumor and hepatoblastoma, and less commonly neuroblastoma, rhabdomyosarcoma, and adrenocortical carcinoma. Tumor risk is concentrated in early childhood and varies by molecular subtype (Shuman et al., 2023).
- Risk for Wilms tumor is highest in the first seven years of life, while risk for hepatoblastoma is concentrated in the first three to four years of life (Shuman et al., 2023).

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- Many BWS-associated tumors arise intra-abdominally and may be clinically occult at early stages, supporting guideline-directed imaging surveillance during defined risk periods (Shuman et al., 2023).

Evidence Discussion

- Ultrasound is the preferred imaging modality for tumor surveillance in individuals with BWS due to its effectiveness in detecting renal, hepatic, adrenal, and other intra-abdominal tumors and its lack of ionizing radiation exposure (Shuman et al., 2023).
- Periodic tumor surveillance imaging is considered medically necessary in childhood for individuals with BWS, including those with mild clinical manifestations or nondiagnostic genetic testing results, because somatic mosaicism may limit molecular risk stratification and tumor development may occur in tissues not represented in tested samples (Shuman et al., 2023).
- CT and other ionizing radiation-based imaging modalities are not considered medically necessary for routine tumor surveillance in BWS and should be avoided when effective non-ionizing alternatives are available (Shuman et al., 2023).
- MRI is not routinely considered medically necessary for standard surveillance in asymptomatic individuals with BWS in the absence of concerning findings (Shuman et al., 2023).
- Imaging may be performed to evaluate non-neoplastic manifestations of BWS, such as nephromegaly, nephrocalcinosis, or structural renal anomalies, when clinically indicated. Routine cross-sectional imaging beyond guideline-directed tumor surveillance is not considered medically necessary in asymptomatic individuals and should not be performed solely on the basis of a BWS diagnosis (Shuman et al., 2023).

Health Equity Considerations

BWS is a rare imprinting disorder characterized by clinical heterogeneity, variable tumor risk, and the need for structured imaging surveillance throughout infancy and early childhood. Disparities in recognition, diagnosis, and follow-up care are well documented and have direct implications for timely oncologic imaging. Although BWS occurs across all racial and ethnic groups, differences in age at diagnosis, presenting features, and molecular subtypes have been observed among diverse populations. In particular, non-Caucasian and mixed race patients are more likely to be diagnosed later and to present with less visually apparent features such as hyperinsulinemic hypoglycemia or nephromegaly, rather than classic external findings like macroglossia or omphalocele, which may delay recognition and initiation of tumor surveillance imaging (Duffy et al., 2020). Delayed diagnosis can result in missed or inconsistent abdominal ultrasound screening for embryonal tumors, increasing the risk of later stage detection and worse outcomes.

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Health equity challenges in BWS also extend beyond diagnostic delay to the cumulative burden of surveillance and psychosocial stressors faced by affected families. Tumor screening protocols require frequent, age-specific imaging, which can impose logistical and financial strain, particularly for families with limited resources or reduced access to specialized centers. Parental anxiety related to cancer risk and repeated imaging is substantial and may influence adherence to surveillance recommendations, especially in populations already experiencing barriers to longitudinal care (Butti et al., 2024). Additionally, children and adolescents with BWS show increased rates of internalizing symptoms, including anxiety and social withdrawal, which may be amplified by frequent medical encounters and perceived stigma associated with physical differences or chronic disease (Butti et al., 2024). Ensuring equitable access to guideline-concordant imaging therefore requires not only standardized surveillance recommendations but also attention to culturally sensitive counseling, care coordination, and psychosocial support. Standardized, risk-stratified imaging recommendations for BWS support equity by reducing variability driven by clinician experience, implicit bias, or access to subspecialty expertise. Consistent application of evidence-based surveillance pathways helps mitigate disparities related to race, ethnicity, and socioeconomic status by ensuring that all children with BWS, regardless of phenotypic presentation or background, receive appropriate tumor screening at recommended intervals. Incorporating awareness of diagnostic bias, facilitating access to genetic evaluation, and supporting families through longitudinal imaging programs are critical components of equitable, high-quality care for this population (Duffy et al., 2020; Butti et al., 2024).

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Denys-Drash Syndrome (DDS) (PEDONC-2.5)

ONCP.SC.0002.5.A
v1.0.2027

Criteria

The following imaging studies should be considered medically necessary in individuals with DDS:

Indication	Medically Necessary Imaging Study
• All individuals with DDS, from birth to the 8th birthday	• Abdominal ultrasound (CPT® 76700) every 3 months
• Known renal tumors in individuals with DDS	• See: **Pediatric Renal Tumors (PEDONC-7)**

Background and Supporting Information

- DDS is a rare WT1-associated disorder characterized by the classic triad of disorders of sex development (including undervirilized or ambiguous genitalia), early onset progressive renal disease (most commonly diffuse mesangial sclerosis), and an exceptionally high risk of Wilms tumor. Renal disease typically presents in infancy or early childhood and frequently progresses to end-stage kidney disease (Turner, Brzezinski, & Dome, 2022).
- DDS is caused by heterozygous pathogenic variants in the WT1 gene on chromosome 11p13, most commonly missense variants involving exons 8 or 9. These variants disrupt WT1 transcriptional activity essential for normal renal and genitourinary development (Turner, Brzezinski, & Dome, 2022).
- Individuals with DDS have a markedly elevated risk of Wilms tumor, often cited as exceeding 90% lifetime risk, with frequent bilateral, multifocal, or early onset disease. Tumors may develop in infancy or early childhood and can arise concurrently with, or shortly after, recognition of renal dysfunction (Turner, Brzezinski, & Dome, 2022).
- Progressive renal failure is common in DDS, and preservation of renal function is a critical management goal. Early identification of Wilms tumor through structured imaging surveillance may facilitate nephron-sparing surgical approaches, potentially preserving residual renal function and delaying the need for renal replacement therapy (Turner, Brzezinski, & Dome, 2022).

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Evidence Discussion

- Abdominal ultrasound is the preferred imaging modality for Wilms tumor surveillance in children with DDS. Ultrasound is non-ionizing, well-tolerated in infants and young children, and effective for detecting renal masses and nephrogenic rests, which are common precursor lesions in WT1-associated disease (Turner, Brzezinski, & Dome, 2022).
- Routine surveillance with abdominal ultrasound is considered medically necessary in children with DDS due to the exceptionally high risk of Wilms tumor, the tendency toward early onset and bilateral disease, and the importance of early detection to support nephron-sparing management strategies. Surveillance imaging with abdominal ultrasound every three months is recommended in individuals with WT1-associated Wilms tumor predisposition, including DDS, to allow detection of rapidly growing tumors at an early and potentially more treatable stage (Turner, Brzezinski, & Dome, 2022).
  - Surveillance is generally continued through early childhood, encompassing the age range during which the vast majority of WT1-associated Wilms tumors are diagnosed; the duration of surveillance may be individualized based on genotype, clinical course, renal status, and treating specialist judgment (Turner, Brzezinski, & Dome, 2022).
- MRI is not considered medically necessary for routine Wilms tumor surveillance and should not replace ultrasound as the first line screening modality in asymptomatic individuals with DDS (Turner, Brzezinski, & Dome, 2022).
- CT imaging should be avoided for routine surveillance in DDS due to cumulative exposure to ionizing radiation in a population requiring frequent and repeated imaging (Turner, Brzezinski, & Dome, 2022).

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Wilms Tumor-Aniridia-Growth Retardation (WAGR) (PEDONC-2.6)

ONCP.SC.0002.6.A
v1.0.2027

Criteria

The following imaging studies should be considered medically necessary in individuals with WAGR:

Indication	Medically Necessary Imaging Study
• All individuals with WAGR, from birth to the 8th birthday	• Abdominal ultrasound (CPT® 76700) every 3 months
• Known renal tumors in individuals with WAGR	• See: **Pediatric Renal Tumors (PEDONC-7)**

Background and Supporting Information

- WAGR is a contiguous gene deletion disorder named for its cardinal features: Wilms tumor, aniridia, genitourinary anomalies, and growth retardation, with variable degrees of intellectual disability. The phenotype results from haploinsufficiency of multiple genes critical for renal, ocular, and genitourinary development (Turner, Brzezinski, & Dome, 2022).
- WAGR is caused by a heterozygous deletion at chromosome 11p13 encompassing the WT1 gene and the adjacent PAX6 gene. Loss of WT1 confers a high risk of Wilms tumor, while deletion of PAX6 causes congenital aniridia. Most cases arise de novo (Turner, Brzezinski, & Dome, 2022).
- Individuals with WAGR have a substantially increased risk of Wilms tumor, reported in approximately 45–60% of affected individuals. Tumors typically arise in early childhood, and bilateral or multifocal disease may occur (Turner, Brzezinski, & Dome, 2022).
- Children with WAGR are at increased risk for progressive renal dysfunction due to congenital kidney abnormalities and, in those who develop Wilms tumor, the cumulative effects of tumor-directed therapy. Among affected individuals who develop symptomatic Wilms tumor, the risk of subsequent renal failure is significant. Early tumor detection via imaging surveillance may facilitate nephron-sparing surgical approaches, with the goal of preserving renal function and delaying progression to end-stage kidney disease (Turner, Brzezinski, & Dome, 2022).

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Evidence Discussion

- Imaging surveillance is considered medically necessary in children with WAGR to enable early detection of Wilms tumor at a low stage, thereby reducing treatment intensity, treatment-related morbidity, and risk of renal failure. Given the rapid growth kinetics of Wilms tumor, regular imaging at defined intervals is required for effective early detection (Turner, Brzezinski, & Dome, 2022).
- Abdominal ultrasound is the preferred modality for routine Wilms tumor surveillance in WAGR. Ultrasound is non-ionizing, widely available, and effective for detection of renal masses and nephrogenic rests, which are recognized precursors to Wilms tumor in WT1-associated conditions. Surveillance with abdominal ultrasound every three months is recommended, beginning in infancy or at the time of diagnosis. Surveillance is generally continued until at least age eight years, encompassing the period during which the vast majority of Wilms tumors associated with WAGR occur (Turner, Brzezinski, & Dome, 2022).
- MRI is not considered medically necessary for routine surveillance and should not replace ultrasound as first line screening, particularly given the need for sedation in young children and limited incremental benefit for screening purposes (Turner, Brzezinski, & Dome, 2022).
- Routine cross-sectional imaging beyond established Wilms tumor surveillance protocols is not considered medically necessary in asymptomatic individuals with WAGR syndrome and should be reserved for specific clinical indications (Turner, Brzezinski, & Dome, 2022).

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Familial Adenomatous Polyposis (FAP) and Related Conditions (PEDONC-2.7)

ONCP.SC.0002.7.A
v1.0.2027

Criteria

- Individuals with Lynch, Gardner, and Turcot syndromes should also be imaged according to these guidelines.

The following imaging studies should be considered medically necessary in individuals with FAP and related conditions:

- For colonoscopy and endoscopy guidance for those with FAP and related conditions, See: *EGD-1.16*, *CAPEND-5*, or *COLON-17: Genetic Syndromes* for the endoscopic management of polyposis syndromes.

Indication	Medically Necessary Imaging Study
• All individuals with FAP and related conditions, from birth to the 6th birthday	• Abdominal ultrasound (CPT® 76700) every 3 months
• All individuals with FAP and related conditions, beginning at age 12 years	• Annual thyroid ultrasound (CPT® 76536)
• All individuals with FAP and related conditions, beginning at age 30 years	• Annual pelvic ultrasound (CPT® 76856)
• Family history of desmoid tumors in individuals with FAP and related conditions	• Abdominal ultrasound (CPT® 76700) annually for life after age 6
• Individuals with Spigelman Stage III or IV or if duodenectomy is being planned in individuals with FAP and related conditions • See: *Genetic Syndromes (EGD-1.16)* for additional information regarding Spigelman staging.	• MR Enterography (MRI Abdomen without and with contrast, CPT® 74183 and MRI Pelvis without and with contrast, CPT® 72197)

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Indication	Medically Necessary Imaging Study
• Known colorectal tumors in individuals with FAP and related conditions	• See: **Colorectal Cancer (ONC-16)** in the Oncology Imaging Guidelines
• Known desmoid tumors in individuals with FAP and related conditions	• See: **Sarcomas – Bone, Soft Tissue, and GIST (ONC-12)**

Background and Supporting Information

- FAP is an autosomal dominant cancer predisposition syndrome caused by pathogenic variants in the APC gene. Affected individuals have a 50% risk of transmission to offspring. Classic FAP is characterized by the development of hundreds to thousands of adenomatous colorectal polyps, typically beginning in childhood or adolescence (Yen et al., 2022).
- Without intervention, individuals with classic FAP have a greater than 90% lifetime risk of colorectal carcinoma, with cancer often developing by early adulthood. As a result, prophylactic colectomy is generally recommended once polyp burden becomes significant, often by late adolescence or early adulthood (Yen et al., 2022).
- In addition to colorectal neoplasia, FAP is associated with an increased risk of several extracolonic malignancies, including hepatoblastoma, duodenal and small bowel adenocarcinoma, thyroid cancer (particularly papillary carcinoma), pancreatic cancer, and central nervous system tumors such as medulloblastoma (Yen et al., 2022).
- The risk of hepatoblastoma is concentrated in early childhood, making targeted abdominal surveillance relevant in affected children and genetically at-risk infants. Other associated malignancies typically occur later in life but may present earlier than their sporadic counterparts, underscoring the importance of risk-appropriate, condition-specific surveillance strategies (Yen et al., 2022).

Evidence Discussion

- Imaging is not the primary modality for colorectal or duodenal surveillance in FAP, as endoscopy is required for detection, risk stratification, and management of adenomatous polyps. Imaging plays a supportive and adjunctive role in the evaluation of selected extracolonic malignancies and non-malignant complications associated with APC-associated polyposis conditions (Yen et al., 2022).
- Abdominal ultrasound is the recommended imaging modality for hepatoblastoma surveillance in infants and young children with FAP or in children known to carry a pathogenic APC variant. Surveillance is performed in conjunction with serial measurement of serum alpha fetoprotein. Ultrasound is non-ionizing and effective for

detection of hepatic masses during the high-risk early childhood period (Yen et al., 2022).

- Thyroid ultrasound is used for surveillance of papillary thyroid carcinoma, which occurs at increased frequency in individuals with FAP, particularly adolescent and young adult females (Yen et al., 2022).
- Routine cross-sectional imaging (CT or MRI) is not considered medically necessary for cancer screening in asymptomatic individuals with FAP and is not recommended for routine surveillance due to limited benefit and potential cumulative exposure risks (Yen et al., 2022).
- Adrenal masses are more prevalent in individuals with FAP than in the general population and are most often identified incidentally. There is no evidence to support routine imaging-based screening of the adrenal glands in asymptomatic individuals with FAP; evaluation is generally symptom driven or prompted by incidental findings (Yen et al., 2022).
- In individuals with advanced duodenal polyposis, CT or MR enterography or video capsule endoscopy may be considered selectively to assess the extent of small bowel involvement. These imaging studies are adjunctive and do not replace endoscopic surveillance; use is based on disease severity and clinical judgment rather than routine screening indications (Yen et al., 2022).

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Multiple Endocrine Neoplasias (MEN) (PEDONC-2.8)

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v1.0.2027

Criteria

The following imaging studies should be considered medically necessary in individuals with MEN1:

Indication	Medically Necessary Imaging Study
All individuals with MEN1, beginning at age 8 years	• MRI Brain without and with contrast (CPT® 70553) every 3 years AND • Annual MRI Abdomen without and with MRI Pelvis without and with contrast (CPT® 74183 +/- 72197), OR CT Abdomen with contrast (CPT® 74160) OR CT Abdomen/Pelvis with contrast (CPT® 74177) AND • Annual MRI Chest without and with contrast (CPT® 71552) or CT Chest with contrast (CPT® 71260)
Known thyroid cancer in individuals with MEN1	• See: **Thyroid Cancer (ONC-6)** in the Oncology Imaging Guidelines
Known pheochromocytoma in individuals with MEN1	• See: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines

The following imaging studies should be considered medically necessary in individuals with MEN2a and MEN2b:

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Indication	Medically Necessary Imaging Study
• One of the following in individuals with MEN2a and MEN2b: ◦ Elevated catecholamines ◦ Inconclusive adrenal mass on MRI	• See: **Adrenal Lesions (PEDAB-17)**
• Known pheochromocytoma in individuals with MEN2a and MEN2b	• See: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)**

Background and Supporting Information

- MEN syndromes are autosomal dominant cancer predisposition disorders characterized by the development of multiple endocrine and neuroendocrine tumors, often at younger ages than sporadic counterparts. Affected individuals have a 50% risk of transmission to offspring, necessitating early, genotype- and phenotype-specific surveillance strategies (Eng & Plitt, 2023; Giusti, Marini, & Brandi, 2022).
- MEN syndromes also include additional hereditary endocrine tumor syndromes and phenocopies beyond MEN1 and MEN2, which can present with overlapping tumor types and influence surveillance strategies (Brandi et al., 2025; Del Rivero et al., 2025; Johnston et al., 2001).
- MEN1 is caused by pathogenic variants in the MEN1 gene and is classically characterized by parathyroid tumors, pancreatic neuroendocrine tumors (PNETs), and anterior pituitary adenomas ("the 3 P's"). Additional associated manifestations include thymic, bronchial, and gastric carcinoid tumors, adrenal lesions, and other endocrine and non-endocrine neoplasms (Giusti, Marini, & Brandi, 2022).
  - MEN1 is associated with significant morbidity and reduced life expectancy, with pancreatic neuroendocrine tumors representing the leading cause of disease-related mortality (Brandi et al., 2025; Del Rivero et al., 2025).
  - Tumor penetrance increases with age, with more than half of affected individuals developing manifestations by age 20 and over 90% by age 40. Many MEN1-associated tumors are clinically silent in early stages, supporting the use of targeted biochemical testing complemented by selective imaging for early identification (Giusti, Marini, & Brandi, 2022).
  - Earlier identification through genetic testing has led to detection of smaller, often indolent tumors, reinforcing the need for balanced surveillance strategies (Brandi et al., 2025; Del Rivero et al., 2025).

- MEN2 is caused by activating pathogenic variants in the RET proto-oncogene and includes MEN2A, MEN2B, and familial medullary thyroid carcinoma (FMTC). All MEN2 subtypes are characterized by a high risk of medullary thyroid carcinoma (MTC), with age of onset varying by genotype and subtype (Eng & Plitt, 2023).
  - MEN2A is characterized by MTC, pheochromocytomas (often bilateral), and parathyrod adenomas or hyperplasia (Eng & Plitt, 2023).
  - MEN2B is associated with early-onset, aggressive MTC, mucosal neuromas, diffuse gastrointestinal ganglioneuromatosis, and a marfanoid habitus, frequently presenting in infancy or early childhood (Eng & Plitt, 2023).
  - Tumors in MEN2 are frequently multifocal, bilateral, and hormonally active, with potentially life threatening complications if unrecognized. Early identification supports timely surgical intervention, including genotype directed prophylactic thyroidectomy, and reduces morbidity related to metastatic or hormonally active disease (Eng & Plitt, 2023).
  - MEN2-associated pheochromocytomas are typically adrenal and adrenergic (epinephrine-producing), allowing for high sensitivity of biochemical screening. In contrast, SDHx-related tumors are often noradrenergic or dopaminergic and may include extra-adrenal paragangliomas, which are less reliably detected with biochemical testing alone and may require broader imaging approaches (Del Rivero et al., 2025; Geurts et al., 2020; Johnston et al., 2001).

Evidence Discussion

- In MEN syndromes, biochemical surveillance is the primary screening modality for most tumor types. Imaging is used selectively to localize disease following abnormal biochemical findings, evaluate extent of disease, or guide surgical planning, rather than for indiscriminate whole body screening (Eng & Plitt, 2023; Giusti, Marini, & Brandi, 2022). This selective approach reduces harms including radiation exposure, overdiagnosis of indolent tumors, and unnecessary procedures associated with lifelong surveillance (Brandi et al., 2025).
- MEN1
  - Imaging is not used for routine screening of MEN1-associated hyperparathyroidism, as diagnosis is based on biochemical hypercalcemia and elevated parathyroid hormone levels. Localization studies are reserved for pre-operative planning or recurrent disease (Giusti, Marini, & Brandi, 2022). This avoids unnecessary imaging in multiglandular disease where localization studies have limited diagnostic value outside of surgical planning (Del Rivero et al., 2025).
  - MRI of the brain is used at defined intervals for detection of pituitary adenomas. Cross-sectional abdominal imaging (CT, MRI, or endoscopic ultrasound) may be used periodically to evaluate for pancreatic neuroendocrine tumors in conjunction with biochemical surveillance but is not a substitute for laboratory monitoring (Giusti, Marini, & Brandi, 2022). MRI is often preferred in MEN1 surveillance due to

its sensitivity and avoidance of ionizing radiation, which is important in individuals requiring lifelong imaging beginning in childhood (Brandi et al., 2025; Del Rivero et al., 2025).
    - Chest CT or MRI may be considered selectively for detection of thymic or bronchial carcinoid tumors in higher risk individuals (Giusti, Marini, & Brandi, 2022). Selective thoracic imaging balances early detection of higher-risk tumors with the risks of cumulative radiation exposure, as only a subset of individuals develop these malignancies (Brandi et al., 2025).
- MEN2
    - For individuals with MEN2A or MEN2B, recommended screening includes annual biochemical testing for pheochromocytoma using plasma free metanephrines or 24 hour urine fractionated metanephrines, beginning at an age determined by the specific RET pathogenic variant risk category (Eng & Plitt, 2023). This biochemical-first approach is supported by the high sensitivity of testing for adrenergic tumors in MEN2 (Del Rivero et al., 2025).
    - MRI of the abdomen is the preferred first-line imaging modality for localization of pheochromocytoma when biochemical results are abnormal, due to high sensitivity and avoidance of ionizing radiation (Eng & Plitt, 2023).
    - Advanced nuclear imaging (e.g., MIBG or PET based techniques) is not considered medically necessary for routine screening (Eng & Plitt, 2023). These modalities are reserved for select cases due to limited incremental benefit and potential harms from radiation exposure and false positives (Del Rivero et al., 2025).
    - Imaging for hyperparathyroidism in MEN2A is not used for screening purposes. Localization studies are performed only after biochemical hyperparathyroidism is confirmed, typically following management of thyroid disease, and are used to guide surgical planning (Eng & Plitt, 2023).
    - Differences between MEN2-associated and SDHx-related tumors further support selective imaging strategies, as MEN2 lesions are typically identified biochemically, whereas non-adrenergic or extra-adrenal tumors in other hereditary syndromes may require imaging for detection (Geurts et al., 2020; Johnston et al., 2001).

Health Equity Considerations

MEN, including MEN1 and MEN2, is a rare hereditary cancer syndrome that requires lifelong, protocolized imaging surveillance beginning in childhood or adolescence. Disparities in access to specialized endocrine and oncologic care, genetic testing, and longitudinal imaging follow-up can significantly affect outcomes. Because MEN-associated tumors often develop silently over time, delayed diagnosis or inconsistent surveillance disproportionately increases the risk of advanced disease at presentation. Structural barriers such as limited access to high-volume specialty centers, insurance instability, and geographic distance to imaging facilities are especially relevant

for patients from lower socioeconomic backgrounds and medically underserved communities, potentially leading to gaps in guideline-concordant tumor screening (Diaz-Thomas et al., 2023; van Leeuwaarde et al., 2020).

Financial toxicity and employment disruption are key equity concerns in MEN, particularly MEN1, where patients frequently undergo repeated imaging, biochemical testing, and surgical interventions over decades. In a large U.S. cohort, the majority of adults with MEN1 reported substantial financial burden related to their condition, including unemployment, high out-of-pocket medication costs, and adverse financial events such as debt or bankruptcy. These financial stressors were strongly associated with worse health-related quality of life across multiple physical and psychosocial domains, underscoring how economic instability can impair adherence to surveillance recommendations and ongoing care (Peipert et al., 2017). Imaging guidelines that minimize unnecessary variation while maintaining adequate surveillance may help reduce cumulative cost burden and support more equitable long-term management.

Psychosocial and educational factors further intersect with health equity in MEN. Employment status has consistently emerged as a major determinant of quality of life in MEN1, with unemployed individuals experiencing significantly worse outcomes regardless of tumor burden. Additionally, discrepancies between individuals' understanding of their disease status and documented tumor findings have been observed, suggesting gaps in health literacy and communication that may be exacerbated in populations facing language barriers or limited access to culturally responsive care (van Leeuwaarde et al., 2020). Standardized, evidence-based imaging pathways for MEN can help mitigate disparities by promoting consistent surveillance independent of socioeconomic status, while reinforcing the importance of multidisciplinary care models, patient education, and structural supports that address financial and psychosocial barriers (Diaz-Thomas et al., 2023).

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Tuberous Sclerosis Complex (TSC) (PEDONC-2.9)

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Criteria

- Abdominal MRI (CPT® 74183) is considered medically necessary for women of childbearing age planning pregnancy.

The following imaging studies should be considered medically necessary in individuals with TSC:

Indication	Medically Necessary Imaging Study
• All individuals with TSC, at the time of suspected diagnosis until age 25 years	• Annual Brain MRI without and with contrast (CPT® 70553)
• All individuals at diagnosis of TSC	• Single baseline MRI Abdomen without and with contrast (CPT® 74183)
• If no renal lesions seen on baseline MRI Abdomen in individuals with TSC	• From diagnosis through age 11: ◦ Annual Renal US (CPT® 76770) • Age 12 (or 10 years earlier than the youngest family member with renal cell carcinoma, whichever comes earlier): ◦ MRI Abdomen without and with contrast (CPT® 74183) annually in lieu of ultrasound
• For documented renal lesions on baseline MRI Abdomen or any ultrasound in individuals with TSC	• MRI Abdomen without and with contrast (CPT® 74183) annually
• All individuals with TSC, beginning at age 18 years	• CT Chest without contrast (CPT® 71250) every 5 years
• All individuals with TSC, for cardiac screening or follow up of known cardiac disease	• See: **Initial Transthoracic Echocardiography (TTE) Indications (PEDCD-8.2)** and **Repeat Transthoracic Echocardiography Indications (PEDCD-8.3)** in the Pediatric Cardiology Imaging Guidelines

Indication	Medically Necessary Imaging Study
• Individuals with TSC with documented abnormalities on baseline CT Chest	• Additional CT Chest without contrast or with contrast (CPT® 71250 or 71260) every 1 year
• Any of the following in individuals with TSC: ◦ New pulmonary symptoms ◦ Worsening pulmonary function testing	• CT Chest without contrast (CPT® 71250)
• Known subependymal giant cell astrocytoma (SEGA) tumors in individuals with TSC	• See: **Intracranial Low Grade Gliomas (LGG) (PEDONC-4.2)**
• Known renal cell carcinoma in individuals with TSC	• See: **Pediatric Renal Cell Carcinoma (RCC) (PEDONC-7.4)**

Background and Supporting Information

- TSC is a multisystem genetic disorder caused by pathogenic variants in TSC1 or TSC2 and is inherited in an autosomal dominant manner with a 50% risk to offspring; disease penetrance is high but clinical expression is variable (Northrup et al., 2024).
- TSC affects approximately 1 in 6,000 individuals and commonly presents in childhood, necessitating long term surveillance strategies beginning early in life (Northrup et al., 2024).
- Individuals with TSC are predisposed to benign and malignant tumors involving the brain, kidneys, lungs, and heart, including SEGAs, renal angiomyolipomas, renal cell carcinoma, pulmonary lymphangioleiomyomatosis (LAM), and cardiac rhabdomyomas (Northrup et al., 2024).
  - SEGAs arise from subependymal nodules and may cause obstructive hydrocephalus; these tumors often grow silently, making scheduled neuroimaging an integral part of management (Northrup et al., 2024).
  - Renal angiomyolipomas and cystic disease are common and progressive, while renal cell carcinoma, though uncommon, occurs at younger ages than sporadic RCC, supporting renal imaging surveillance across the lifespan (Northrup et al., 2024).
  - Pulmonary LAM primarily affects females and may remain asymptomatic until advanced disease develops, justifying periodic pulmonary imaging in adulthood rather than symptom only evaluation (Northrup et al., 2024).

Evidence Discussion

- Longitudinal brain MRI enables early detection of SEGA growth prior to hydrocephalus or neurologic deterioration, allowing timely initiation of medical therapy or surgical intervention and reducing associated morbidity (Northrup et al., 2024).
- Serial renal imaging identifies angiomyolipoma growth and early malignant transformation, facilitating nephron-sparing interventions and mitigation of hemorrhage and renal insufficiency risks (Northrup et al., 2024).
- Periodic chest imaging in individuals at risk for LAM allows recognition of cystic lung disease before development of pneumothorax or respiratory failure, conditions that may otherwise present abruptly (Northrup et al., 2024).
- TSC-associated lesions demonstrate age-specific patterns of onset and progression, supporting tailored imaging strategies rather than indiscriminate or whole body screening imaging (Northrup et al., 2024).
- The clinical benefit of early tumor detection and prevention of irreversible organ damage outweighs the potential harms of repeated imaging when surveillance is targeted, modality appropriate, and performed at defined intervals (Northrup et al., 2024).

Health Equity Considerations

TSC is a rare, lifelong, multisystem genetic condition that imposes a disproportionate medical, psychosocial, and economic burden on affected children and their families, with important implications for health equity. Individuals with TSC require frequent surveillance imaging, specialty care across multiple organ systems, and ongoing neurodevelopmental and psychiatric assessment, resulting in high healthcare utilization and complexity of care (Rentz et al., 2015; Jansen et al., 2020). Access to comprehensive, multidisciplinary TSC clinics is uneven, with many families living significant distances from specialized centers, contributing to delays in diagnosis, inconsistent follow-up imaging, and fragmented care, particularly for patients reliant on public insurance or residing in rural or underserved areas (Rentz et al., 2015; Jansen et al., 2020). These structural barriers may lead to under-surveillance of clinically significant complications, including renal, neurologic, and pulmonary manifestations, thereby exacerbating health disparities.

Social determinants of health further compound inequities in this population. High rates of TSC associated neuropsychiatric disorders, epilepsy, intellectual disability, and autism spectrum disorder significantly disrupt educational attainment, employment, and caregiver workforce participation, disproportionately affecting families with fewer socioeconomic resources (Jansen et al., 2020; Sforza et al., 2026). Caregivers of children with TSC frequently report substantial out of pocket expenses, lost workdays, and unmet needs for psychological and social support, with greater burden observed in families managing epilepsy and neurodevelopmental comorbidities (Rentz et al.,

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2015; Sforza et al., 2026). Despite the high prevalence of psychiatric symptoms, mental health evaluation and treatment remain underutilized, reflecting disparities in access to behavioral health services and limited integration of psychosocial screening into routine TSC care (Rai & Sandefur, 2024; Jansen et al., 2020). Equity-focused imaging and surveillance strategies for TSC should therefore emphasize guideline-concordant, age-appropriate monitoring that accounts for barriers to specialty access, supports coordinated multidisciplinary care, and minimizes avoidable delays or disparities in diagnosis and ongoing disease management.

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Von Hippel-Lindau Syndrome (VHL) (PEDONC-2.10)

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Criteria

The following imaging studies are considered medically necessary in individuals with VHL:

Indication	Medically Necessary Imaging Study
• All individuals with VHL, beginning at age 11 years	• MRI Brain without and with contrast (CPT® 70553) and MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, and Lumbar-CPT® 72158) every 2 years
• Individuals with VHL with frequent ear infections, prior to age 8	• One-time MRI Brain without and with contrast (CPT® 70553)
• All individuals with VHL, beginning at age 5 years	• Annual Abdominal US (CPT® 76700)
• All individuals with VHL, beginning at age 15 years	• MRI Abdomen without and with contrast (CPT® 74183) every 2 years
• Individuals with VHL with suspected pheochromocytoma as indicated by **ONE** of the following: ◦ Elevated catecholamines ◦ Inconclusive adrenal mass on MRI	• Octreotide study (CPT® 78800, CPT® 78801, CPT® 78802, CPT® 78803, or CPT® 78804) OR • Adrenal Nuclear imaging study (CPT® 78075) OR • Dotatate PET (CPT® 78815 or CPT® 78816)
• Individuals with VHL with known hemangioblastoma anywhere in the body (based on imaging) that has not been resected	• Both of the following, every 1 year: ◦ MRI Brain without and with contrast (CPT® 70553) ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, and Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
• Known hemangioblastoma that has not been resected and new or worsening symptoms in individuals with VHL	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, and Lumbar-CPT® 72158)
• Known (based on imaging) CNS hemangioblastoma in individuals with VHL	• See: **Intracranial Low Grade Gliomas (LGG) (PEDONC-4.2)**
• Known renal cell carcinoma in individuals with VHL	• See: **Pediatric Renal Cell Carcinoma (RCC) (PEDONC-7.4)**
• Known pheochromocytoma or other neuroendocrine tumor in individuals with VHL	• See: **Neuroendocrine Cancers And Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines
• New abnormality is detected on MRI Brain in individuals with VHL	• A repeat MRI Brain without and with contrast (CPT® 70553) should be done in 6 months. • If stable, imaging should return to every 2 years starting 2 years from the prior study
• New abnormality is found in the pancreas on a screening MRI Abdomen in individuals with VHL	• Repeat MRI Abdomen without and with contrast (CPT® 74183) in 6 months then annually from initial scan to observe for development of pNET

Background and Supporting Information

- VHL is an autosomal dominant cancer predisposition syndrome caused by pathogenic variants in the VHL tumor suppressor gene, conferring a 50% risk of inheritance and high lifetime penetrance (van Léeuwaarde et al., 2025).
- VHL is characterized by multisystem tumor development, including central nervous system (CNS) hemangioblastomas, retinal hemangioblastomas, clear cell renal cell carcinoma (RCC), pheochromocytomas and paragangliomas, pancreatic cysts and neuroendocrine tumors, and endolymphatic sac tumors (van Léeuwaarde et al., 2025).
- CNS hemangioblastomas arise in the cerebellum, brainstem, and spinal cord and may remain asymptomatic until causing mass effect or neurologic compromise; retinal hemangioblastomas may present in childhood and can cause irreversible vision loss if untreated (van Léeuwaarde et al., 2025).
- Clear cell RCC develops in approximately 70% of individuals with VHL and is the leading cause of disease related mortality, often occurring at younger ages than

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sporadic RCC and frequently in a multifocal or bilateral pattern (van Léeuwaarde et al., 2025).
- Pheochromocytomas may be clinically silent or present with episodic hypertension, while endolymphatic sac tumors may cause sudden or progressive hearing loss, underscoring the role of biochemical and imaging surveillance prior to symptom onset (van Léeuwaarde et al., 2025).
- Implementation of systematic surveillance protocols for VHL has been associated with significant improvements in life expectancy and reductions in morbidity compared with historical cohorts lacking routine screening (van Léeuwaarde et al., 2025).

Evidence Discussion

- Periodic MRI of the brain and spine facilitates early detection of hemangioblastomas before neurologic deterioration, enabling timely surgical or medical intervention and reducing irreversible neurologic morbidity (van Léeuwaarde et al., 2025).
- Regular ophthalmologic evaluation allows identification of retinal hemangioblastomas at early stages, when treatment can preserve vision and prevent blindness, outcomes not achievable once advanced complications occur (van Léeuwaarde et al., 2025).
- Longitudinal abdominal imaging supports early recognition and size-based management of RCC, which correlates with improved survival and nephron preservation in individuals with hereditary VHL-associated renal tumors (van Léeuwaarde et al., 2025).
- Scheduled biochemical testing identifies pheochromocytomas prior to clinical presentation, reducing the risk of hypertensive crises and perioperative complications through planned surgical management (van Léeuwaarde et al., 2025).
- Serial audiologic monitoring detects early hearing changes related to endolymphatic sac tumors, allowing targeted imaging and intervention before permanent hearing loss develops (van Léeuwaarde et al., 2025).
- Evidence supports that structured, organ-specific surveillance in VHL yields substantial clinical benefit by preventing advanced disease, while limiting harm through targeted imaging rather than indiscriminate or symptom-only evaluation (van Léeuwaarde et al., 2025).

Health Equity Considerations

VHL syndrome is a rare, inherited cancer predisposition disorder that requires lifelong, intensive imaging surveillance and repeated specialty interventions across multiple organ systems, creating significant equity concerns. Because VHL is uncommon and lacks a dedicated ICD diagnosis code, delays in recognition and diagnosis are common, particularly outside of specialized hereditary cancer or academic centers, which may disproportionately affect individuals from rural, underserved, or lower resource settings (Jonasch et al., 2024). Management typically involves frequent MRI-based surveillance of the brain, spine, abdomen, and other organs, as well as repeated surgical procedures

over an individual's lifetime. Claims based data demonstrate markedly higher healthcare utilization and annual costs for individuals with VHL-associated central nervous system hemangioblastomas and pancreatic neuroendocrine tumors compared with matched controls, reflecting intensive inpatient, outpatient, and procedural care requirements (Jonasch et al., 2024). Limited access to specialty imaging facilities and multidisciplinary expertise may therefore result in delayed detection of tumors, inconsistent surveillance, or increased morbidity in socioeconomically disadvantaged populations.

Financial- and insurance-related barriers further exacerbate disparities in VHL care. Individuals with VHL-associated tumors incur substantial excess annual healthcare costs driven largely by imaging, hospitalizations, and tumor related surgeries, with surgical admissions and treatment-related complications representing particularly high-cost events (Jonasch et al., 2024). These burdens may be especially challenging for families with high deductible plans, limited insurance coverage, or reliance on public insurance programs, potentially leading to deferred imaging or gaps in recommended surveillance. Although emerging systemic therapies, such as HIF 2α inhibition, may reduce the frequency of surgeries for select individuals, equitable access to advanced imaging and novel treatments remains uneven (Jonasch et al., 2024). Equity-focused imaging strategies for VHL should therefore prioritize guideline-concordant surveillance across the lifespan, consider cumulative financial and logistical burdens, and support coordinated, multidisciplinary care pathways to reduce disparities in early tumor detection and long term outcomes.

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Rhabdoid Tumor Predisposition Syndrome (PEDONC-2.11)

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Criteria

The following imaging studies should be considered medically necessary in individuals with Rhabdoid Tumor Predisposition Syndrome:

Indication	Medically Necessary Imaging Study
• All individuals with rhabdoid tumor predisposition syndrome, at diagnosis, as early as birth if requested	• Whole-body MRI (WBMRI) (CPT® 76498)
• All individuals with rhabdoid tumor predisposition syndrome, from birth to 6 months of age	• ALL of the following, monthly: ◦ US Head (CPT® 76506) ◦ US Abdomen (CPT® 76700) ◦ US Pelvis (CPT® 76856) ◦ US Neck (CPT® 76536) ◦ MRI without and with contrast of areas of concern found on baseline WBMRI
• All individuals with rhabdoid tumor predisposition syndrome, from age 7 months to 5 years	• ALL of the following, every 3 months: ◦ US Abdomen (CPT® 76700) ◦ US Pelvis (CPT® 76856) ◦ US Neck (CPT® 76536) ◦ MRI Brain (CPT® 70553) ◦ MRI Spine (CPT® 72156, CPT® 72157, and CPT® 72158) without and with contrast ◦ MRI without and with contrast of areas of concern found on baseline WBMRI
• All individuals with rhabdoid tumor predisposition syndrome, after age 5 years	• Annual WBMRI (CPT® 76498)

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Indication	Medically Necessary Imaging Study
• Clinical symptoms or WBMRI findings suggesting malignancy in individuals with rhabdoid tumor predisposition syndrome	• Targeted advanced imaging of the suspected disease site (CT with or without contrast, or MRI without and with contrast)
• Inconclusive findings on ultrasound in individuals with rhabdoid tumor predisposition syndrome	• MRI without and with contrast of the inconclusive US site ◦ MRI is then considered medically necessary in place of ultrasound for remainder of planned screening

Background and Supporting Information

- Rhabdoid Tumor Predisposition Syndrome (RTPS) is a hereditary cancer predisposition syndrome caused by heterozygous germline pathogenic variants in SMARCB1 (RTPS1) or SMARCA4 (RTPS2), affecting members of the SWI/SNF chromatin remodeling complex (Nemes et al., 2022).
- RTPS is inherited in an autosomal dominant manner with a 50% risk to offspring; penetrance is extremely high for SMARCB1-related RTPS in early childhood and incomplete for SMARCA4-related RTPS, contributing to variable family histories (Nemes et al., 2022).
- RTPS is associated with highly aggressive malignant rhabdoid tumors arising in the central nervous system as atypical teratoid/rhabdoid tumors (ATRT), the kidney as rhabdoid tumor of the kidney, and multiple extracranial soft tissue sites; small cell carcinoma of the ovary, hypercalcemic type (SCCOHT) is specifically associated with SMARCA4 variants (Nemes et al., 2022).
- Most affected individuals present in infancy, with more than 70% developing tumors before 12 months of age, often with synchronous or multifocal tumors that demonstrate rapid growth and aggressive clinical behavior (Nemes et al., 2022).
- Rhabdoid tumors can occur in nearly any anatomic location, including brain, spine, kidney, liver, mediastinum, pelvis, and soft tissues; tumors may be clinically silent until advanced, supporting comprehensive imaging rather than symptom-based evaluation alone (Nemes et al., 2022).
- Although overall prognosis remains poor, early tumor detection and timely initiation of therapy have been associated with improved survival in select individuals with RTPS, particularly those identified through predisposition surveillance (Nemes et al., 2022).

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Evidence Discussion

- The extremely early age of onset and high risk of synchronous tumors support frequent, structured imaging in infancy and early childhood, allowing detection before clinical deterioration and metastatic spread (Nemes et al., 2022).
- Whole body MRI enables assessment of both CNS and extracranial tumor sites in a single session, improving detection of multifocal disease that may be missed with region-limited imaging strategies (Nemes et al., 2022).
- Regular brain and spine MRI permits early identification of ATRT, which often arises in the cerebellum and spinal axis and may progress rapidly, enabling earlier treatment planning and improved neurologic outcomes (Nemes et al., 2022).
- Abdominal and neck ultrasound provides a radiation-free modality for identifying extracranial tumors in young children and serves as an appropriate first line imaging tool for frequent surveillance in high risk infants (Nemes et al., 2022).
- Because affected individuals are typically infants and young children with extreme cancer susceptibility, limiting exposure to ionizing radiation is critical; MRI- and ultrasound- based surveillance strategies reduce long-term risks associated with cumulative radiation exposure (Nemes et al., 2022).
- Although surveillance requires frequent imaging and may involve anesthesia in young children, the substantial benefit of detecting aggressive tumors at a treatable stage outweighs these risks in individuals with RTPS (Nemes et al., 2022).

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Familial Retinoblastoma Syndrome (PEDONC-2.12)

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Criteria

- Regular physical and ophthalmologic evaluations under anesthesia (EUA) are the hallmark of surveillance strategies for these individuals, and asymptomatic screening imaging does not have a defined role at this time.

The following imaging studies should be considered medically necessary in individuals with Familial Retinoblastoma Syndrome:

Indication	Medically Necessary Imaging Study
• Individuals with familial retinoblastoma syndrome with retinomas (premalignant retinal lesions)	• Annual MRI Orbits (CPT® 70543) without and with contrast
• Individuals with familial retinoblastoma syndrome and either of the following: ◦ Inconclusive EUA findings ◦ New symptoms	• US Orbits (CPT® 76512, 76510, or 76511) or MRI Orbits (CPT® 70543) ◦ These studies should be used if at all possible in lieu of CT or nuclear imaging to avoid radiation exposure

Background and Supporting Information

- Familial (heritable) retinoblastoma syndrome is an autosomal dominant cancer predisposition syndrome caused by a heterozygous germline pathogenic variant in RB1, conferring a 50% risk of transmission to offspring (Lohmann & Gallie, 2023).
- Retinoblastoma is a malignant tumor of the developing retina that typically presents in early childhood, most often before age five years, with bilateral and multifocal disease strongly associated with germline RB1 pathogenic variants (Lohmann & Gallie, 2023).
- Tumors arise from retinal cone precursor cells and may be unilateral, bilateral, or multifocal; intracranial midline tumors of the pineal region (trilateral retinoblastoma) represent an important anatomic extension relevant to neuroimaging surveillance (Lohmann & Gallie, 2023).
- Individuals with heritable retinoblastoma have a markedly increased lifetime risk of subsequent malignant neoplasms, particularly osteosarcoma, soft tissue sarcomas, and melanoma, often presenting in adolescence or adulthood (Lohmann & Gallie, 2023).

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- Germline RB1 pathogenic variants are associated with significantly increased susceptibility to radiation-induced malignancies, particularly following external beam radiation therapy, supporting avoidance of ionizing radiation when possible (Lohmann & Gallie, 2023).
- Genetic risk categories based on germline RB1 status (e.g., confirmed germline variant vs. reduced residual risk due to mosaicism) inform intensity and duration of ophthalmologic and imaging surveillance without relying on symptoms alone (Lohmann & Gallie, 2023).

Evidence Discussion

- Frequent ocular evaluation in early childhood facilitates detection of small, asymptomatic retinal tumors, enabling vision sparing treatment and reducing the risk of metastatic spread (Lohmann & Gallie, 2023).
- MRI is effective for evaluating intraocular tumor extent, optic nerve involvement, and intracranial disease, including pinealoblastoma, while avoiding ionizing radiation exposure associated with CT (Lohmann & Gallie, 2023).
- Neuroimaging surveillance allows early identification of pineal or suprasellar tumors, which are often clinically occult and carry a poor prognosis if diagnosed late (Lohmann & Gallie, 2023).
- Limiting diagnostic imaging that uses ionizing radiation reduces cumulative exposure and mitigates the substantially elevated risk of second primary malignancies in individuals with heritable retinoblastoma (Lohmann & Gallie, 2023).
- Imaging and examination strategies tailored to genetic risk balance early cancer detection with minimization of sedation, radiation exposure, and unnecessary testing, aligning benefit with disease biology (Lohmann & Gallie, 2023).
- Although surveillance in young children may require anesthesia and repeated examinations, the benefit of preventing vision loss, detecting life threatening tumors early, and reducing overall cancer mortality outweighs these risks in high-risk individuals (Lohmann & Gallie, 2023).

Health Equity Considerations

Familial retinoblastoma syndrome is a rare, inherited pediatric cancer predisposition disorder that requires early detection, intensive ophthalmic imaging, and longitudinal surveillance, raising important health equity considerations. Timely diagnosis is critical, as disease severity at presentation strongly influences eye salvage, visual outcomes, and treatment intensity. Evidence indicates that children with paternally inherited RB1 mutations present at significantly older ages, with larger tumors and more advanced International Classification of Retinoblastoma groups, and are more likely to require enucleation compared with those with maternally inherited disease (Eiger Moscovich et al., 2024). While parental origin reflects underlying epigenetic and imprinting effects, delayed presentation may also intersect with socioeconomic and access related factors.

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Notably, paternal inheritance cases in the cohort were more frequently observed among African American individuals, suggesting that structural barriers to early ophthalmic evaluation, genetic counseling, or specialty referral may contribute to inequities in presentation severity despite adjustment for race in multivariable analyses (Eiger Moscovich et al., 2024).

Access to specialized care further influences equity in familial retinoblastoma. Optimal management relies on repeated examinations under anesthesia, high resolution ocular imaging, and coordinated care within tertiary ocular oncology centers. Families with limited geographic access, financial constraints, or inadequate insurance coverage may face delays in screening of at risk infants, inconsistent follow up, or limited access to globe sparing therapies. The study highlights that some families declined or were unable to obtain genetic testing due to financial limitations, underscoring disparities in access to molecular diagnostics that are essential for risk stratification, surveillance planning, and counseling of unaffected relatives (Eiger Moscovich et al., 2024). Equity focused imaging guidelines for familial retinoblastoma should therefore emphasize early, guideline-concordant screening of genetically at-risk children, reduce barriers to specialty imaging and genetic services, and support consistent surveillance across socioeconomic and racial groups to prevent avoidable vision loss and morbidity.

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Hereditary Paraganglioma-Pheochromocytoma (HPP) Syndromes (PEDONC-2.13)

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Criteria

- Individuals with multiple endocrine neoplasias should not use this guideline and should be imaged according to **Multiple Endocrine Neoplasias (MEN) (PEDONC-2.8)**.

The following imaging studies should be considered medically necessary in individuals with HPP Syndromes:

Indication	Medically Necessary Imaging Study
• All individuals with HPP syndromes with SDHx (SDHA, SDHB, SDHC, SDHD), MAX, TMEM127 mutations, beginning at age 6	• Every 2 years, ONE of the following sets of imaging: ◦ Whole-body MRI (CPT® 76498) ▪ CPT® 76498 is the only code for whole-body MRI at this time OR ◦ MRI Neck without and with contrast (CPT® 70543), MRI Chest without and with contrast (CPT® 71552), MRI Abdomen without and with contrast (CPT® 74183), and MRI Pelvis without and with contrast (CPT® 72197) ▪ If MRI cannot be performed: CT Neck with contrast (CPT® 70491), CT Chest with contrast (CPT® 71260), and CT Abdomen and Pelvis with contrast (CPT® 74177) ▪ MRI Neck imaging should include skull base, thus separate MRI Brain (CPT® 70553) without and with contrast is only considered medically necessary if there are CNS-specific symptoms.
• Initial screening for individuals diagnosed with HPP syndromes at age 18 or older	• One-time PET/CT (CPT® 78815 or 78816), with 68Ga-DOTA-SSAs ◦ FDG may be substituted if Dotatate radiotracers are not available

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Indication	Medically Necessary Imaging Study
• Known pheochromocytoma or other neuroendocrine tumors in individuals with HPP syndromes	• See: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines

Background and Supporting Information

- HPP syndromes are autosomal dominant cancer predisposition syndromes caused by germline pathogenic variants in SDHA, SDHB, SDHC, SDHD, SDHAF2, MAX, or TMEM127, resulting in increased risk for paragangliomas and pheochromocytomas arising from neural crest–derived paraganglial tissue (Else et al., 2023).
- HPP syndromes are inherited in an autosomal dominant manner with a 50% risk of transmission to offspring, with notable parent of origin effects for SDHD, SDHAF2, and possibly MAX, which influence disease penetrance and phenotype (Else et al., 2023).
- Paragangliomas may arise anywhere along the paravertebral axis from the skull base to the pelvis, while pheochromocytomas are confined to the adrenal medulla; head and neck paragangliomas are typically parasympathetic and non-secretory, whereas sympathetic paragangliomas and pheochromocytomas often secrete catecholamines (Else et al., 2023).
- In addition to paragangliomas and pheochromocytomas, HPP syndromes are associated with increased risk for metastatic disease (particularly in SDHB carriers), gastrointestinal stromal tumors, clear cell renal cell carcinoma, and less commonly pulmonary chondromas (Else et al., 2023).
- Tumors in HPP syndromes may present in childhood or adulthood, with variable penetrance and risk of metachronous or multifocal disease, necessitating lifelong biochemical and imaging surveillance (Else et al., 2023).
- Multiple expert consensus groups recommend genetic evaluation and ongoing surveillance for individuals with HPP syndromes due to the high prevalence of occult, multifocal, or recurrent disease and the inability of pathology alone to distinguish localized from metastatic tumors (Else et al., 2023).

Evidence Discussion

- Paragangliomas may arise at multiple anatomic sites and remain clinically silent for prolonged periods; cross-sectional imaging enables early detection of occult tumors before development of symptoms, mass effect, or metastatic spread (Else et al., 2023).
- MRI is favored over CT for routine surveillance, particularly in pediatric and young adult populations, to minimize cumulative radiation exposure given the need for

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repeated, lifelong imaging while maintaining high sensitivity for soft tissue tumors (Else et al., 2023).

- Regular measurement of plasma-free or urine fractionated metanephrines complements imaging by identifying catecholamine secreting tumors that may be radiographically occult or newly developing, enhancing overall surveillance effectiveness (Else et al., 2023).
- Functional imaging using somatostatin receptor-based PET (e.g., ^{68}Ga DOTATATE) provides high sensitivity for lesion detection and disease staging, particularly in metastatic or multifocal disease, and is supported by international expert consensus when clinically indicated (Else et al., 2023).
- Although surveillance involves repeated imaging and laboratory evaluation, early tumor detection enables curative surgical resection, reduces morbidity from catecholamine excess, and may prevent progression to metastatic disease, outweighing the potential harms of screening (Else et al., 2023).
- Imaging frequency and modality selection are supported by evidence demonstrating variable metastatic risk by genotype, particularly the higher malignancy risk associated with SDHB, justifying tailored imaging strategies rather than symptom-based evaluation alone (Else et al., 2023).

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Costello Syndrome (PEDONC-2.14)

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Criteria

The following imaging studies should be considered medically necessary in individuals with Costello Syndrome:

Indication	Medically Necessary Imaging Study
• Following confirmation of gene mutation in individuals with Costello syndrome	• ANY or ALL of the following: ◦ Echocardiogram ▪ CPT® 93306 or ▪ CPT® 93308 with 93321 and 93325 ◦ MRI Brain (CPT® 70553) without and with contrast ◦ MRI Cervical (CPT® 72156) and Thoracic Spine (CPT® 72157) without and with contrast
• All individuals with Costello syndrome, from birth to 10th birthday	• Every 3 months: ◦ US Abdomen (CPT® 76700) and Pelvis (CPT® 76856)
• Known cardiac disease in individuals with Costello syndrome	• See: **Initial Transthoracic Echocardiography (TTE) Indications (PEDCD-8.2)** and **Repeat Transthoracic Echocardiography Indications (PEDCD-8.3)** in the Pediatric Cardiology Guidelines
• Known rhabdomyosarcoma in individuals with Costello syndrome	• See: **Rhabdomyosarcoma (RMS) (PEDONC-8.2)**
• Known neuroblastoma in individuals with Costello syndrome	• See: **Neuroblastoma (PEDONC-6)**

Background and Supporting Information

- Costello syndrome is a multisystem RASopathy caused by activating heterozygous pathogenic variants in the HRAS gene, leading to dysregulation of the Ras/MAPK signaling pathway and abnormal cellular growth, differentiation, and oncogenic risk (Gripp & Weaver, 2023).

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- Costello syndrome is inherited in an autosomal dominant manner and is most commonly due to a de novo HRAS pathogenic variant; affected individuals with classic phenotypes rarely reproduce, though vertical transmission has been reported in rare attenuated cases (Gripp & Weaver, 2023).
- Individuals with Costello syndrome have an estimated 15% lifetime risk of malignancy, with embryonal rhabdomyosarcoma and neuroblastoma predominating in infancy and early childhood, and transitional cell carcinoma of the bladder occurring in adolescence and adulthood (Gripp & Weaver, 2023).
- Central nervous system abnormalities are common and may include progressive postnatal cerebellar overgrowth, Chiari I malformation, hydrocephalus, and syringomyelia; genitourinary tumors and abdominal soft tissue malignancies represent key extracranial imaging targets (Gripp & Weaver, 2023).
- The phenotypic range extends from mild or attenuated forms to severe presentations with early lethal cardiomyopathy or neurologic complications, supporting risk-based surveillance rather than symptom-driven evaluation alone (Gripp & Weaver, 2023).
- Published clinical management guidelines for Costello syndrome recommend structured surveillance for malignancy and neurologic complications due to the recognized tumor risk and progressive anatomic abnormalities that may be clinically occult (Gripp & Weaver, 2023).

Evidence Discussion

- Many malignancies associated with Costello syndrome, particularly rhabdomyosarcoma and neuroblastoma, present early in life and may be asymptomatic; periodic abdominal and pelvic imaging enables earlier detection that may improve treatment options and outcomes (Gripp & Weaver, 2023).
- Progressive cerebellar overgrowth and posterior fossa crowding can lead to Chiari I malformation, hydrocephalus, or syringomyelia, which may evolve over time; neuroimaging allows detection prior to irreversible neurologic compromise (Gripp & Weaver, 2023).
- Transitional cell carcinoma of the bladder occurs at a markedly younger age in individuals with Costello syndrome than in the general population, justifying surveillance strategies that include imaging or urinalysis triggered evaluation rather than symptom based detection alone (Gripp & Weaver, 2023).
- Biochemical screening for catecholamine metabolites has not demonstrated utility for detecting neuroblastoma in Costello syndrome due to frequent false elevations in the absence of tumor, supporting imaging-based approaches when surveillance is pursued (Gripp & Weaver, 2023).
- Although repeated imaging introduces considerations related to sedation and cumulative burden, the relatively high cancer risk, early age of onset, and potential for clinically silent progression support the net benefit of structured, age appropriate imaging surveillance (Gripp & Weaver, 2023).

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- Imaging recommendations are supported by genotype-driven penetrance data and observed cancer incidence, reinforcing a surveillance approach that balances early detection with minimization of unnecessary studies (Gripp & Weaver, 2023).

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Constitutional Mismatch Repair Deficiency (CMMRD or Turcot Syndrome) (PEDONC-2.15)

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Criteria

The following imaging studies should be considered medically necessary in individuals with CMMRD/Turcot Syndrome:

Indication	Medically Necessary Imaging Study
• All individuals, after CMMRD diagnosis is confirmed by genetic mutation	• MRI Brain without and with contrast (CPT® 70553) every 6 months
• All individuals with CMMRD, beginning at age 4 years	• Annual esophagogastroduodenoscopy and colonoscopy
• All individuals with CMMRD, beginning at age 6 years	• Annual whole-body MRI (CPT® 76498) ◦ 76498 is the only code for whole-body MRI at this time

Background and Supporting Information

- CMMRD is a highly penetrant pediatric cancer predisposition syndrome caused by autosomal recessive inheritance of biallelic pathogenic variants in mismatch repair genes (MSH2, MSH6, MLH1, or PMS2), leading to genomic instability and early, aggressive malignancies (Tabori et al., 2017).
- CMMRD is associated with a broad and severe malignancy spectrum, most commonly high-grade central nervous system tumors (malignant gliomas, embryonal tumors, medulloblastoma), hematologic malignancies (T cell lymphoblastic lymphoma and leukemia), and gastrointestinal cancers, often occurring in early childhood (Tabori et al., 2017).
- The median age at first cancer diagnosis in individuals with CMMRD is approximately 7–8 years, with many individuals developing multiple synchronous or metachronous malignancies during childhood and a median survival of less than three years after initial diagnosis (Tabori et al., 2017).
- Individuals with CMMRD universally develop gastrointestinal polyposis over time, frequently mimicking attenuated familial adenomatous polyposis, with progression to colorectal carcinoma often during childhood or adolescence (Tabori et al., 2017).

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- Central nervous system involvement is common and includes high-grade gliomas and embryonal tumors that may not be detected clinically until advanced; extracranial malignancies may arise in the abdomen, pelvis, bone, and soft tissues (Tabori et al., 2017).
- International expert groups recommend intensive, early, and multimodality imaging surveillance due to the extreme cancer risk and early onset in affected children (Tabori et al., 2017).

Evidence Discussion

- Central nervous system tumors are the most common malignancies in CMMRD and may occur during infancy; early and repeated brain MRI improves detection of tumors at a potentially treatable stage and outperforms ultrasound in sensitivity, even in young children (Tabori et al., 2017).
- The occurrence of diverse, non canonical tumors, including sarcomas and genitourinary malignancies, supports the use of whole body MRI as part of surveillance to detect clinically silent cancers across multiple organ systems (Tabori et al., 2017).
- Imaging and endoscopic surveillance allow early identification of polyps and carcinomas, enabling timely intervention such as polypectomy or surgery and reducing the likelihood of advanced colorectal cancer (Tabori et al., 2017).
- Hematologic malignancies lack highly sensitive imaging or laboratory screening tools; therefore, imaging strategies combined with clinical vigilance provide a pragmatic approach for early detection when routine laboratory testing is insufficient (Tabori et al., 2017).
- Although surveillance requires frequent imaging and may involve sedation in young children, the exceptionally high cancer penetrance, early age of onset, and fatal natural history of untreated disease strongly favor intensive imaging strategies (Tabori et al., 2017).
- Evidence from retrospective cohorts and expert consensus demonstrates that structured imaging surveillance, including brain MRI and whole body MRI, can identify tumors earlier and is justified by the severity and breadth of cancer risk in CMMRD (Tabori et al., 2017).

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Hereditary Leiomyomatosis and Renal Cell Cancer (HLRCC) (PEDONC-2.16)

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Criteria

The following imaging studies should be considered medically necessary in individuals with HLRCC:

Indication	Medically Necessary Imaging Study
• Beginning at age 8 years, individuals with HLRCC with at least ONE of the following: ◦ Documented fumarate hydratase (FH) gene mutation/variant consistent with HLRCC ◦ Histologically confirmed multiple cutaneous piloleiomyomas ◦ At least TWO of the following manifestations: ▪ Surgical treatment for symptomatic uterine leiomyomas before age 40 ▪ Type 2 papillary renal cell carcinoma or collecting duct renal carcinoma before age 40 ▪ A first-degree family member with either of the two above criteria	• MRI Abdomen without and with contrast (CPT® 74183), annually

Background and Supporting Information

- HLRCC is an autosomal dominant cancer predisposition syndrome caused by heterozygous pathogenic variants in the FH gene, which encodes fumarate hydratase, a key enzyme of the tricarboxylic acid cycle; loss of FH function results in tumorigenesis through metabolic dysregulation and pseudohypoxia signaling (PDQ Cancer Genetics Editorial Board, 2024).
- HLRCC is characterized by cutaneous leiomyomas, early-onset and often severe uterine leiomyomas in females, and a distinct form of renal cell carcinoma that is biologically aggressive and considered a unique renal tumor subtype (PDQ Cancer Genetics Editorial Board, 2024).

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- The lifetime risk of renal cell carcinoma (RCC) in individuals with HLRCC is generally estimated between 15% and 30%, with reported cases occurring from childhood through adulthood; renal cancers have been documented in individuals as young as 10–11 years of age (PDQ Cancer Genetics Editorial Board, 2024; Forde et al., 2020).
- HLRCC-associated RCCs are typically unilateral and solitary, often high grade at presentation, and demonstrate aggressive behavior with early metastatic potential, distinguishing them from other hereditary RCC syndromes (PDQ Cancer Genetics Editorial Board, 2024).
- Although not all mutation carriers develop RCC, the penetrance of non-renal manifestations is high and there is substantial intrafamilial and interfamilial variability, limiting the ability to identify individuals at highest risk without imaging surveillance (PDQ Cancer Genetics Editorial Board, 2024).
- Due to the aggressive growth pattern of HLRCC-associated RCC and the frequent presentation at advanced stages, expert groups and specialty societies recommend routine renal imaging surveillance, favoring modalities that allow early lesion detection while minimizing cumulative radiation exposure (PDQ Cancer Genetics Editorial Board, 2024).

Evidence Discussion

- Symptomatic HLRCC-associated RCCs frequently present at advanced stage (stage III-IV), at which point prognosis is poor; therefore, routine imaging surveillance is aimed at detecting small, localized tumors amenable to curative surgical management (Forde et al., 2020).
- Prospective data demonstrate that MRI screening identifies early stage RCCs in asymptomatic individuals with HLRCC, enabling intervention at stages associated with significantly improved survival compared with symptomatic detection (Forde et al., 2020).
- In a multicenter cohort, individuals diagnosed with stage I or II RCC had markedly longer survival than those diagnosed at stage III or IV, with all individuals presenting with advanced disease dying within three years, supporting early imaging-based detection strategies (Forde et al., 2020).
- Modeling based on observed outcomes suggests that approximately five individuals with HLRCC need to undergo renal MRI surveillance to prevent one RCC related death, reflecting a favorable benefit to burden ratio for imaging in this population (Forde et al., 2020).
- MRI is preferred for lifelong surveillance because it avoids ionizing radiation while maintaining sensitivity for small or subtle renal lesions, an important consideration given the early onset and need for repeated imaging in HLRCC (PDQ Cancer Genetics Editorial Board, 2024).
- Although routine imaging increases resource utilization and may detect incidental findings, the aggressive and often fatal natural history of untreated HLRCC-

associated RCC supports a lower threshold for surveillance imaging compared with other hereditary renal cancer syndromes (PDQ Cancer Genetics Editorial Board, 2024; Forde et al., 2020).

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Other Renal Cell Cancer Predisposition Syndromes (PEDONC-2.17)

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Birt-Hogg-Dube Syndrome (BHDS)

Criteria

The following imaging studies should be considered medically necessary in individuals with BHDS:

Indication	Medically Necessary Imaging Study
• All individuals with BHDS, beginning at age 20 years	• MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) annually ◦ If no family history of renal tumors AND after 2 consecutive normal MRIs, MRI is considered medically necessary every 2 years

Background and Supporting Information

- BHDS is an autosomal dominant disorder caused by heterozygous pathogenic variants in the FLCN gene, which encodes folliculin, a tumor suppressor protein involved in cellular metabolism and growth signaling pathways (Sattler & Steinlein, 2024).
- BHDS is characterized by a triad of cutaneous fibrofolliculomas and related skin lesions, pulmonary cysts with increased risk for spontaneous pneumothorax, and renal tumors of varied histology, with wide intrafamilial and interfamilial variability in disease expression (Sattler & Steinlein, 2024).
- Characteristic skin findings include fibrofolliculomas, trichodiscomas, acrochordons, angiofibromas, oral papules, collagenomas, and epidermal cysts, which typically develop in adulthood and may increase in number over time but are usually absent in childhood (Sattler & Steinlein, 2024).
- Pulmonary cysts, predominantly located in the basal lung regions, are common in BHDS and confer a markedly increased lifetime risk of spontaneous pneumothorax, often presenting as the first clinical manifestation in adolescents or young adults (Sattler & Steinlein, 2024).

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- Individuals with BHDS have an estimated 15%–30% lifetime risk of renal cell carcinoma, representing approximately a seven fold increase over the general population; tumors are frequently bilateral and multifocal (Sattler & Steinlein, 2024).
- The most common BHDS-associated renal tumors are oncocytic hybrid tumors (chromophobe/oncocytoma), followed by chromophobe RCC, oncocytoma, and clear cell carcinoma; the median age of renal tumor diagnosis is approximately 50–55 years, though adolescent onset has been reported (Sattler & Steinlein, 2024).
- Because renal tumors may be clinically silent until advanced and cumulative radiation exposure is a concern, expert consensus guidelines recommend periodic cross-sectional renal imaging, favoring MRI for long-term surveillance (Sattler & Steinlein, 2024).

Evidence Discussion

- Renal tumors in BHDS can be bilateral, multifocal, and slow growing but unpredictably aggressive in some cases; routine imaging allows detection at a size amenable to nephron-sparing intervention and reduces the likelihood of advanced disease presentation (Sattler & Steinlein, 2024).
- MRI is preferred for renal surveillance due to its sensitivity for small renal lesions and avoidance of ionizing radiation, an important consideration given the need for lifelong imaging beginning in early adulthood (Sattler & Steinlein, 2024).
- Evidence supports monitoring renal tumors smaller than 3cm with imaging, as early detection enables timely nephron-sparing surgery once tumors exceed this threshold, preserving renal function while maintaining oncologic safety (Sattler & Steinlein, 2024).
- While lung cysts are common, routine serial chest CT in asymptomatic individuals is not recommended because of cumulative radiation risks (Sattler & Steinlein, 2024).
- The benefits of periodic renal imaging in BHDS outweigh potential harms, as MRI-based screening minimizes radiation exposure while addressing a substantially increased renal cancer risk that cannot be predicted by phenotype or genotype alone (Sattler & Steinlein, 2024).
- The clinical evidence underlying BHDS demonstrates that organ specific, risk adapted imaging, particularly for renal manifestations, provides a rational balance between early detection, avoidance of unnecessary procedures, and long-term safety (Sattler & Steinlein, 2024).

BAP1 Tumor Predisposition Syndrome

Criteria

The following imaging studies should be considered medically necessary in individuals with BAP1:

Indication	Medically Necessary Imaging Study
• All individuals with BAP1, beginning at age 30 years	• Abdominal MRI without and with contrast (CPT® 74183) every 2 years
• All individuals with BAP1, beginning at age 18 years	The following studies are considered medically necessary every 2 years: • MRI Brain without and with contrast (CPT® 70553) • MRI Whole Spine without and with contrast (Cervical- CPT® 72156, Thoracic- CPT® 72157, Lumbar- CPT® 72158)

Background and Supporting Information

- BAP1 is an autosomal dominant cancer predisposition syndrome caused by heterozygous germline pathogenic variants in the BAP1 gene, which encodes a tumor suppressor involved in chromatin regulation, DNA damage response, and cell cycle control (Pilarski et al., 2024).
- BAP1 is associated with an increased risk of BAP1-inactivated melanocytic tumors, uveal melanoma, malignant mesothelioma, cutaneous melanoma, renal cell carcinoma, basal cell carcinoma, meningioma, chromophobe renal cancer, and cholangiocarcinoma, with affected individuals often developing multiple primary malignancies over their lifetime (Pilarski et al., 2024).
- Tumors associated with BAP1 generally occur at younger ages than their sporadic counterparts, and certain malignancies, particularly uveal melanoma and renal cell carcinoma, tend to demonstrate more aggressive biological behavior (Pilarski et al., 2024).
- Uveal melanoma is the most common malignancy reported in BAP1 and is typically characterized by high risk molecular features associated with increased metastatic potential and reduced survival compared with uveal melanoma in the general population (Pilarski et al., 2024).
- Malignant mesothelioma in BAP1 often presents earlier than sporadic disease and shows a higher proportion of peritoneal involvement; despite its severity, BAP1 associated mesothelioma may demonstrate improved survival and treatment responsiveness compared with non hereditary cases (Pilarski et al., 2024).
- Renal cell carcinoma in BAP1 most commonly exhibits clear cell histology, occurs at a younger median age than sporadic RCC, and is associated with higher tumor grade and worse prognosis, underscoring the importance of early detection (Pilarski et al., 2024).

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- Multiple expert groups have proposed structured surveillance strategies emphasizing dermatologic, ophthalmologic, abdominal, and thoracic evaluation, with a preference for imaging modalities that balance early tumor detection against cumulative radiation exposure (Pilarski et al., 2024).

Evidence Discussion

- The broad tumor spectrum and high penetrance of malignancies in BAP1 support proactive surveillance imaging to enable identification of clinically occult tumors at potentially curable stages (Pilarski et al., 2024).
- Regular ophthalmologic surveillance facilitates early identification of uveal melanoma, which is particularly critical given the aggressive phenotype and elevated metastatic risk associated with BAP1-related tumors (Pilarski et al., 2024).
- Surveillance imaging of the abdomen allows for early detection of renal cell carcinoma, which in BAP1 occurs at younger ages and is associated with poorer outcomes when diagnosed at advanced stages (Pilarski et al., 2024).
- Routine chest CT screening is not universally recommended due to radiation risk and uncertain benefit; instead, clinical evaluation is favored given the lack of consensus on optimal mesothelioma screening modalities (Pilarski et al., 2024).
- Surveillance strategies emphasize minimizing cumulative radiation exposure by prioritizing MRI and ultrasound where feasible, reflecting the need for lifelong monitoring in genetically predisposed individuals (Pilarski et al., 2024).
- While surveillance imaging may identify incidental findings or increase health care utilization, the high morbidity and mortality associated with advanced BAP1-related cancers support an imaging approach tailored to tumor risk, age, and organ system involvement (Pilarski et al., 2024).

Health Equity Considerations

BAP1 is a rare inherited cancer syndrome associated with increased risk of uveal melanoma, cutaneous melanoma, malignant mesothelioma, and renal cell carcinoma, requiring lifelong surveillance imaging across multiple organ systems. Health equity concerns arise from both the rarity of the syndrome and emerging evidence that biologic, social, and structural factors intersect to influence cancer presentation and outcomes in BAP1-associated malignancies. Data from population-level cancer genomics demonstrate that disparities in renal cell carcinoma outcomes between Black and White individuals have historically been attributed to differences in access to high-quality care, rates of surgical intervention, and broader social determinants of health rather than differences in tumor detection or imaging frequency (Paulucci et al., 2016). These findings underscore the importance of equitable access to specialty imaging, genetic evaluation, and multidisciplinary care for individuals with suspected or confirmed BAP1, particularly as cancers in this syndrome may present at younger ages and with aggressive clinical behavior.

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Notably, genomic analyses reveal that BAP1 expression and its relationship to tumor stage and pathologic features vary by race, with Black patients showing higher BAP1 expression and more localized disease despite historically worse overall survival outcomes (Paulucci et al., 2016). This paradox highlights the limitations of relying solely on biologic markers without addressing structural inequities, including delayed diagnosis, barriers to nephrectomy or tumor-directed therapies, and inconsistent follow up imaging. For individuals with BAP1, inequitable access to genetic testing may further delay identification of at risk family members and initiation of guideline concordant surveillance. Equity-focused imaging guidelines should therefore prioritize early genetic confirmation, consistent cross-sectional and organ-specific imaging surveillance, and care coordination across socioeconomic and racial groups to reduce preventable disparities in cancer stage at diagnosis and long term outcomes in this high risk population (Paulucci et al., 2016).

Hereditary Papillary Renal Carcinoma (HPRC)

Criteria

The following imaging studies should be considered medically necessary in individuals with HPRC:

Indication	Medically Necessary Imaging Study
• All individuals with HPRC, beginning at age 30 years	• Abdominal MRI without and with contrast (CPT® 74183) annually

Background and Supporting Information

- HPRC is an autosomal dominant cancer predisposition syndrome caused by activating germline pathogenic variants in the tyrosine kinase domain of the MET proto-oncogene, leading to constitutive MET signaling (PDQ Cancer Genetics Editorial Board, 2024).
- HPRC predisposes affected individuals to bilateral and multifocal papillary renal cell carcinoma (RCC), previously designated as type 1 papillary RCC, with no consistent extrarenal manifestations identified (PDQ Cancer Genetics Editorial Board, 2024).
- Germline MET pathogenic variants demonstrate very high penetrance, approaching 100%, with renal tumors typically developing in adulthood but reported across a wide age range (approximately 19–66 years) (PDQ Cancer Genetics Editorial Board, 2024).
- HPRC-associated tumors have a characteristic papillary morphology consisting of small basophilic cells arranged in single layers on thin papillae and are often low grade, although high grade and metastatic tumors can occur (PDQ Cancer Genetics Editorial Board, 2024).

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- Renal tumors in HPRC are frequently bilateral, multifocal, and numerous, with microscopic papillary adenomas and incipient lesions commonly present throughout the renal parenchyma (PDQ Cancer Genetics Editorial Board, 2024).
- Given the near universal penetrance and lifelong risk of tumor development, expert consensus supports ongoing cross-sectional renal imaging surveillance in individuals with HPRC to support early detection and renal preservation (PDQ Cancer Genetics Editorial Board, 2024).

Evidence Discussion

- Individuals with HPRC develop multiple bilateral renal tumors over time, often asymptomatic at small sizes; routine surveillance imaging enables identification of tumors before they progress to metastatic or symptomatic disease (PDQ Cancer Genetics Editorial Board, 2024).
- Papillary RCCs associated with HPRC are characteristically hypovascular and may demonstrate minimal enhancement, making them difficult to detect by ultrasound alone (PDQ Cancer Genetics Editorial Board, 2024).
- Because imaging surveillance must continue throughout life and many individuals will undergo repeated studies, MRI is generally preferred to limit cumulative radiation exposure while maintaining sensitivity for small renal tumors (PDQ Cancer Genetics Editorial Board, 2024).
- Prospective observational data support radiologic surveillance of HPRC associated renal tumors until the largest lesion approaches approximately 3cm, at which point nephron-sparing surgery can be performed to balance oncologic control with renal function preservation (PDQ Cancer Genetics Editorial Board, 2024).
- Although frequent imaging increases healthcare utilization and may identify incidental findings, the exceptionally high penetrance of renal tumors and effectiveness of early surgical intervention support the benefit of structured imaging surveillance in HPRC (PDQ Cancer Genetics Editorial Board, 2024).
- The natural history of HPRC, characterized by predictable, kidney-limited tumor development with prolonged asymptomatic growth, supports a tailored imaging approach that differs from sporadic RCC management and aligns with specialized surveillance criteria (PDQ Cancer Genetics Editorial Board, 2024).

Health Equity Considerations

HPRC is a rare autosomal dominant cancer predisposition syndrome characterized by bilateral, multifocal papillary renal cell carcinomas driven by germline MET activation. Although HPRC accounts for only a small subset of papillary renal cell carcinoma (PRCC), broader population level data on PRCC highlight important equity related factors that are directly relevant to imaging surveillance and outcomes in hereditary disease. Large registry based analyses demonstrate that Black race is independently associated with increased mortality in PRCC, even after adjustment for age, tumor grade, stage, and size, suggesting that social and structural determinants of health

contribute meaningfully to observed outcome disparities (Ullah et al., 2023). Delayed diagnosis, barriers to specialty referral, and inconsistent access to high quality imaging may disproportionately affect populations already shown to have worse prognoses, which is particularly consequential in HPRC where early detection of small, asymptomatic renal tumors is central to nephron-sparing management strategies (Ullah et al., 2023).

Equitable access to guideline-concordant imaging surveillance is therefore critical for individuals with HPRC and at-risk family members. Cross-sectional abdominal imaging with contrast enhanced MRI or CT is essential for identifying multifocal and bilateral renal tumors before they exceed size thresholds associated with metastasis and worse survival (Ullah et al., 2023). However, disparities in access to advanced imaging, genetic counseling, and longitudinal follow-up may lead to later detection with larger tumors, a factor shown to independently predict mortality and to be more prevalent among Black patients with PRCC (Ullah et al., 2023). Health equity-focused imaging guidelines for HPRC should prioritize early genetic identification, consistent high-quality renal imaging across the lifespan, and coordinated multidisciplinary care, with particular attention to populations historically shown to experience worse outcomes despite similar disease biology.

DICER1

Criteria

The following imaging studies should be considered medically necessary in individuals with DICER1:

Indication	Medically Necessary Imaging Study
• DICER1 mutation asymptomatic lung surveillance	• Once at age 3-6 months of age and again at 2.5-3 years of age: ◦ CT Chest (contrast as requested) • In addition, chest x-ray at birth and every 6 months until 8 years of age and annually until age 12

Indication	Medically Necessary Imaging Study
• DICER1 mutation asymptomatic abdominal and pelvic surveillance	• Every 3 months, from birth to the 8th birthday, then every 12 months until 12 years of age: ◦ Abdominal US (CPT® 76700) • After age 12, it is considered medically necessary for females with DICER1 mutation to continue abdominal and pelvic ultrasounds (CPT® 76700 and CPT® 76856 or CPT® 76857) annually until age 40
• DICER1 mutation thyroid surveillance	• Baseline thyroid US (CPT® 76536) by 8 years of age then every 3 years
• Individuals with DICER1 mutation and new CNS symptoms	• MRI Brain without and with contrast (CPT® 70553)

Background and Supporting Information

- DICER1-related tumor predisposition is an autosomal dominant hereditary cancer predisposition syndrome caused by heterozygous germline loss of function pathogenic variants in the DICER1 gene, which encodes an RNase III endoribonuclease essential for microRNA biogenesis and gene regulation (Schultz et al., 2026).
- DICER1 demonstrates reduced, age-dependent penetrance, with many individuals carrying a germline DICER1 pathogenic variant remaining clinically unaffected throughout life, while others develop one or more benign or malignant tumors, often during childhood or early adulthood (Schultz et al., 2026).
- DICER1 is associated with a broad spectrum of neoplasms affecting multiple organ systems, including lung (pleuropulmonary blastoma and lung cysts), thyroid (multinodular goiter and differentiated thyroid carcinoma), kidney (pediatric cystic nephroma, anaplastic sarcoma of the kidney, and Wilms tumor), female reproductive tract (ovarian sex cord stromal tumors), and central nervous system (pituitary blastoma, pineoblastoma, and DICER1-associated CNS sarcoma) (Schultz et al., 2026).
- Distinct tumors in DICER1 have characteristic age distributions, with pleuropulmonary blastoma and lung cysts primarily occurring in infancy and early childhood, pediatric cystic nephroma presenting most often before age four years, and ovarian and thyroid tumors typically arising in adolescence or young adulthood (Schultz et al., 2026).
- Pleuropulmonary blastoma (PPB) exists along a continuum from cystic (type I or regressed type Ir) to aggressive solid sarcomatous forms (types II-III), with early

cystic lesions representing a precursor state that may progress rapidly, underscoring the importance of early thoracic evaluation (Schultz et al., 2026).
- Thyroid involvement is common in DICER1, with a markedly increased lifetime risk of multinodular goiter and a substantially elevated risk of differentiated thyroid carcinoma, particularly follicular patterned tumors, which are often intrathyroidal and indolent but may be multifocal (Schultz et al., 2026).
- Multidisciplinary expert groups have established structured, age-specific surveillance strategies emphasizing symptom awareness, targeted imaging, and minimization of cumulative radiation exposure (Schultz et al., 2026).

Evidence Discussion

- The wide anatomic distribution, early age of onset, and potential for malignant progression of DICER1-associated tumors provide strong evidence for systematic, imaging-based surveillance to identify lesions at an early, more treatable stage (Schultz et al., 2026).
- Early chest imaging in infancy and young childhood facilitates detection of lung cysts and early-stage pleuropulmonary blastoma, enabling intervention before progression to aggressive solid tumors with significantly worse survival outcomes (Schultz et al., 2026).
- Abdominal ultrasound-based surveillance in childhood supports early identification of pediatric cystic nephroma and related renal tumors, which may otherwise grow rapidly or undergo sarcomatous transformation if not detected and managed promptly (Schultz et al., 2026).
- Periodic thyroid ultrasound allows detection of multinodular goiter and differentiated thyroid carcinoma; however, given the generally indolent biology of many DICER1-associated thyroid cancers, surveillance strategies emphasize judicious imaging to balance early detection against overdiagnosis (Schultz et al., 2026).
- Pelvic ultrasound surveillance in females permits early detection of ovarian sex cord stromal tumors, which are frequently diagnosed at early stage and have favorable outcomes when managed promptly (Schultz et al., 2026).
- Evidence does not support routine screening brain MRI in asymptomatic individuals; instead, symptom directed MRI is recommended to avoid unnecessary sedation and imaging burden while still enabling timely diagnosis of rare but serious CNS malignancies (Schultz et al., 2026).
- Surveillance protocols prioritize ultrasound and MRI over CT whenever feasible, reflecting evidence that individuals with DICER1 require repeated imaging across childhood and young adulthood, making radiation minimization a key consideration (Schultz et al., 2026).
- Although surveillance imaging may lead to incidental findings, increased healthcare utilization, and patient anxiety, available evidence supports that the benefits of early

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tumor detection and improved outcomes outweigh these potential harms in individuals with DICER1 related tumor predisposition (Schultz et al., 2026).

PTEN Hamartoma Tumor Syndrome (PHTS)/Cowden Syndrome (CS)

Criteria

The following imaging studies should be considered medically necessary in individuals with PHTS/CS:

Indication	Medically Necessary Imaging Study
All individuals with PHTS/CS	• Baseline MRI Abdomen without and with contrast (CPT® 74183) or CT Abdomen without and with contrast (CPT® 74170) at age 30, followed by renal US (CPT® 76770) every 1-2 years

Background and Supporting Information

- PTEN hamartoma tumor syndrome (PHTS), which includes Cowden syndrome (CS), is an autosomal dominant cancer predisposition syndrome caused by pathogenic variants in the PTEN tumor suppressor gene. Major manifestations include macrocephaly, mucocutaneous lesions, breast cancer, endometrial cancer, and follicular thyroid cancer. Renal cell carcinoma (RCC) is recognized as a syndrome-associated malignancy and is included as a minor diagnostic feature (NCCN, 2026).
- Renal neoplasms associated with PHTS/CS most commonly demonstrate chromophobe and papillary histology, although other RCC subtypes have been reported (NCCN, 2026).

Evidence Discussion

- Individuals with PHTS/CS have an increased lifetime risk of RCC relative to the general population, supporting the use of dedicated kidney cancer surveillance strategies rather than symptom-driven evaluation alone (NCCN, 2026).
- Surveillance recommendations include baseline abdominal imaging with contrast-enhanced MRI (preferred) or CT beginning at age 30 years, reflecting the documented risk of renal malignancy and the potential for clinically occult disease. (NCCN, 2026).
- Following baseline cross-sectional imaging, renal ultrasound at 1- to 2-year intervals is recommended as an imaging surveillance strategy that balances early detection of RCC with minimization of cumulative radiation exposure in a hereditary cancer population requiring lifelong monitoring (NCCN, 2026).
- CT should be limited when possible for surveillance of hereditary RCC syndromes because of lifetime radiation exposure concerns, supporting MRI as the preferred baseline cross-sectional modality in PHTS/CS (NCCN, 2026).

- Surveillance imaging may permit identification of renal tumors at an earlier stage, when nephron-sparing approaches can be considered, thereby preserving renal function while maintaining oncologic control. This consideration is particularly important for hereditary cancer syndromes in which lifelong cancer risk exists (NCCN, 2026).

Other Renal Predisposition Syndromes

Criteria

- The list of syndromes and congenital anomalies associated with Wilms Tumor specifically is ever growing. The following genetic variants and syndromes, not otherwise listed in **PEDONC-2** for more specific imaging, may have imaging as shown below.

The following imaging studies should be considered medically necessary in individuals with other renal predisposition syndromes:

Indication	Medically Necessary Imaging Study
• Individuals with any of the following genetic variants or syndromes: ◦ REST, TRIM28, FBXW7, NYNRIN, KDM3B, XPO5, CHECK2, PALB2, CTNNB1, DROSHA, WT1 and 2, WTX, DGCR8, SIC1 and 2, BCORL1, MLLT1, MYCN, DIS3L2 ◦ Bloom Syndrome/BLM mutations ◦ Frasier Syndrome ◦ Trisomy 18 ◦ Perlman Syndrome ◦ Bohring-Opitz Syndrome (ASXL1) ◦ MULIBREY and Nanism Syndrome ◦ Congenital anomalies associated with Wilms Tumor ▪ Horseshoe kidney ▪ Renal ectopia, hypoplasia, or renal/ureteral duplication ▪ Congenital mesoblastic nephroma	• Every 3 months, from birth to the 8th birthday: ◦ Abdominal US (CPT® 76700)

Infantile Myofibromatosis (PEDONC-2.18)

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Criteria

The following imaging studies should be considered medically necessary in individuals with Infantile Myofibromatosis:

Indication	Medically Necessary Imaging Study
• Assess for the presence of multifocal disease in children under two years of age with infantile myofibromatosis	• WBMRI (CPT® 76498) once within 6 months of diagnosis
• Pre-operative planning for infantile myofibromatosis or • Signs or symptoms suggesting progression of infantile myofibromatosis that may require treatment with surgery or chemotherapy	• Imaging of cutaneous/muscular sites with MRI with and without contrast of symptomatic sites or sites requiring treatment.
• Unresected, known visceral sites of disease in individuals with infantile myofibromatosis	• MRI without and with contrast of unresected visceral involved sites, every 6 months until the age of two years
• Post-operative evaluation and both of the following in individuals with infantile myofibromatosis: ◦ Adequacy of resection is unclear and ◦ Re-excision is being considered	• One-time MRI without and with contrast

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Indication	Medically Necessary Imaging Study
• Surveillance for emergence of visceral disease in individuals with infantile myofibromatosis	• Every 6 months, until the age of two years: ◦ CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) • CT should not duplicate simultaneous MRI imaging of involved sites
• Individuals with infantile myofibromatosis requiring chemotherapy	• See: **PEDONC-8.3**

Background and Supporting Information

- Infantile myofibromatosis (IM) is a rare benign mesenchymal tumor disorder characterized by the development of myofibroblastic tumors involving skin, subcutaneous tissue, muscle, bone, and, less commonly, visceral organs; lesions are non-metastatic but may cause morbidity through local growth and organ compression (NCI, 2020).
- IM occurs predominantly in infancy and early childhood, with more than 90% of cases diagnosed before two years of age, and an estimated incidence of approximately 1 in 150,000 live births, though underdiagnosis is suspected (NCI, 2020).
- IM presents in distinct clinical forms, including solitary lesions (most common), multicentric disease limited to skin and muscle, multicentric disease with limited visceral involvement, and diffuse multicentric disease with extensive visceral involvement, which carries the highest risk for morbidity and mortality (NCI, 2020).
- Solitary and multicentric non-visceral lesions most commonly affect the skin and muscles of the head, neck, and trunk, while visceral lesions may involve the lungs, heart, gastrointestinal tract, or abdominal organs, where mass effect may impair function (NCI, 2020).
- Most cases of IM are sporadic, but rare familial cases have been associated with pathogenic variants in PDGFRB and NOTCH3, supporting a genetic contribution in select patients (NCI, 2020).
- IM lesions confined to the skin, muscle, or bone frequently demonstrate spontaneous stabilization or regression without intervention, supporting conservative management in the absence of organ compromise (NCI, 2020).
- Prognosis is strongly influenced by the presence or absence of visceral involvement; non-visceral IM has an excellent prognosis, whereas untreated visceral disease is

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associated with markedly reduced survival due to organ compression, particularly cardiopulmonary compromise (NCI, 2020).

Evidence Discussion

- Imaging plays a critical role in determining lesion location, size, and extent, particularly to identify occult visceral involvement that may not be clinically apparent in infants and young children (NCI, 2020).
- Infants may present initially with a single lesion and subsequently develop additional lesions within the first two years of life, and their limited ability to express symptoms supports targeted imaging to evaluate for clinically significant internal disease (NCI, 2020).
- Routine serial imaging of cutaneous or muscular lesions is not necessary in asymptomatic infants, as spontaneous regression or stabilization is common and unnecessary imaging may increase burden without improving outcomes (NCI, 2020).
- Imaging is justified when symptoms, lesion burden, or clinical findings raise concern for visceral involvement, as unmanaged internal tumors are associated with substantial morbidity and poor survival (NCI, 2020).
- Surveillance beyond early childhood is generally not warranted, as the vast majority of IM lesions present before age two years and the risk of new lesion development declines significantly thereafter (NCI, 2020).
- Imaging strategies emphasizing symptom-directed and risk-based evaluation optimize early detection of life threatening visceral disease while minimizing unnecessary imaging, sedation, and radiation exposure in a population with high rates of spontaneous lesion regression (NCI, 2020).

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Bloom Syndrome (PEDONC-2.19)

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Criteria

The following imaging studies should be considered medically necessary in individuals with Bloom Syndrome:

Indication	Medically Necessary Imaging Study
Malignancy screening, particularly lymphomas, in individuals with Bloom syndrome	• Every 2 years, starting at age 13: ◦ Whole-Body MRI (CPT® 76498)
Wilms tumor screening in individuals with Bloom syndrome	• See: **Other Renal Cell Cancer Predisposition Syndromes (PEDONC-2.17)**
Gastrointestinal tumor screening in individuals with Bloom syndrome	• There is no strong data to support endoscopic screening
Breast cancer screening in individuals with Bloom syndrome	• There is no strong data to support Breast MRI. See: **Breast MRI Indications (BR-5.1)** for any updates to imaging

Background and Supporting Information

- Bloom syndrome is a rare autosomal recessive chromosomal instability disorder caused by biallelic pathogenic variants in the BLM gene, resulting in defective DNA repair, genomic instability, and markedly increased cancer susceptibility beginning in childhood or early adulthood (Langer et al., 2023).
- Bloom syndrome is characterized by severe pre- and post-natal growth deficiency, microcephaly, sun sensitive facial telangiectatic erythema, immunodeficiency, insulin resistance, and multisystem involvement, with normal or near normal intellectual development in most affected individuals (Langer et al., 2023).
- Individuals with Bloom syndrome have a substantially elevated lifetime risk of malignancy involving a wide range of tissues, including hematologic malignancies, gastrointestinal cancers, breast cancer, skin cancers, and Wilms tumor, often presenting decades earlier than in the general population (Langer et al., 2023).

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- Malignancies in Bloom synrome can arise in virtually any organ system, with particularly increased frequencies of leukemia, lymphoma, colorectal cancer, Wilms tumor, breast cancer, and head and neck tumors, reflecting the underlying defect in genome maintenance (Langer et al., 2023).
- Cancer risk in Bloom syndrome begins in childhood, with Wilms tumor and leukemia presenting in early childhood, followed by increasing rates of solid tumors, including colorectal and breast cancer, during adolescence and young adulthood (Langer et al., 2023).
- Individuals with Bloom syndrome demonstrate hypersensitivity to ionizing radiation and DNA-damaging agents, increasing the risk of treatment-related toxicity and secondary malignancies and influencing diagnostic and surveillance imaging strategies (Langer et al., 2023).
- Expert-based health supervision recommendations emphasize intensive clinical monitoring and targeted imaging for early tumor detection while minimizing radiation exposure, given the exceptionally high cancer risk and radiosensitivity in Bloom syndrome (Langer et al., 2023).

Evidence Discussion

- The markedly increased and early-onset cancer risk in Bloom syndrome supports the use of structured imaging surveillance to facilitate early tumor detection, when treatment may be more effective and less morbid (Langer et al., 2023).
- MRI is the preferred imaging modality in Bloom syndrome, as avoidance of ionizing radiation reduces long term cancer risk and treatment-related complications in individuals with impaired DNA repair mechanisms (Langer et al., 2023).
- Intensified surveillance may lead to false positive findings, incidentalomas, increased healthcare utilization, and anxiety; however, these risks are considered outweighed by the benefit of early malignancy detection in a high risk population (Langer et al., 2023).
- Imaging surveillance strategies in Bloom syndrome are tailored to age-specific cancer risks, focusing on childhood renal tumors and hematologic malignancies early in life and transitioning to surveillance for solid tumors during adolescence and adulthood (Langer et al., 2023).
- Available registry data and expert consensus support targeted, low-radiation imaging surveillance as a reasonable balance between early cancer detection and minimization of iatrogenic risk in individuals with Bloom syndrome, despite the absence of large prospective trials (Langer et al., 2023).

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Pediatric Leukemias (PEDONC-3)

Guideline Page

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Pediatric Leukemia General Considerations (PEDONC-3.1)

ONCP.LE.0003.1.A
v1.0.2027

Criteria

- MRI Brain without and with contrast (CPT® 70553) is considered medically necessary in individuals exhibiting CNS symptoms
  - CT Head without or with contrast (CPT® 70450 or CPT® 70460) is considered medically necessary for urgent concerns where MRI would delay care (i.e., suspected CNS bleeding)
  - Imaging due to CSF tumor burden has not been shown to improve the detection of CNS involvement compared with CSF alone
- See: **Hematopoietic Stem Cell Transplantation (HSCT) (PEDONC-20)** for imaging guidelines related to transplant
- Routine advanced imaging is not considered medically necessary in the evaluation and management of chronic myeloid leukemia in the absence of specific localizing clinical symptoms or clearance for hematopoietic stem cell transplantation.

Background and Supporting Information

- The vast majority of leukemias in children are acute, with acute lymphoblastic leukemia (ALL) accounting for approximately 75–80% of cases and acute myeloid leukemia (AML) representing 15–20%; chronic leukemias such as chronic myelogenous leukemia (CML) are rare in pediatric populations (NCCN, 2025).
- Pediatric leukemias originate in the bone marrow with early hematogenous dissemination; imaging is therefore not routinely required for diagnosis but may be used to evaluate suspected complications such as infection, hemorrhage, leukostasis, or extramedullary disease (NCCN, 2025).
- Prognosis and treatment intensity are determined by age, presenting white blood cell count, cytogenetic and molecular features, and minimal residual disease status, rather than radiographic tumor burden (NCCN, 2025).
- Central nervous system, testicular, and other extramedullary sites may be involved at presentation or relapse, with imaging reserved for symptomatic patients or targeted evaluation of suspected sites of disease (NCCN, 2025).
- Profound immunosuppression, cytopenias, and therapy-related toxicities increase the risk of infection and hemorrhage, supporting selective imaging when clinical findings suggest complications rather than routine surveillance (NCCN, 2025).

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Evidence Discussion

- Because pediatric leukemias are primarily marrow-based and monitored through laboratory and bone marrow assessment, routine advanced imaging does not improve outcomes and is not necessary in asymptomatic patients (NCCN, 2025).
- Evidence supports imaging when there is clinical suspicion for complications such as invasive infection, CNS hemorrhage, leukostasis-related organ dysfunction, or extramedullary relapse, where imaging findings may directly alter management (NCCN, 2025).
- Avoidance of unnecessary imaging reduces radiation exposure and sedation risk in children, while symptom-directed imaging maximizes diagnostic yield in a medically fragile population (NCCN, 2025).

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Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)

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v1.0.2027

Criteria

- Individuals with B-precursor or T-cell lymphoblastic lymphoma without bone marrow involvement are treated similarly to individuals with leukemia of the same cell type and should be imaged according to this guideline section
- This section does not apply to individuals with mature B-cell histology (primarily Burkitt's in children). Please refer to **Pediatric Aggressive Mature B-Cell Non-Hodgkin Lymphomas (NHL) (PEDONC-5.3)** for guidelines for these individuals

The following imaging studies should be considered medically necessary in individuals with ALL:

- Chest x-ray should be performed to evaluate for mediastinal mass in suspected cases of ALL or upon initial diagnosis.

Indication	Medically Necessary Imaging Study
Suspected testicular involvement in individuals with ALL	Scrotal Ultrasound CPT® 76870 and/or doppler ultrasound of the scrotum CPT® 93975 or 93976
Mediastinal widening is seen on chest x-ray in individuals with ALL	CT Chest (contrast as requested) is considered medically necessary to immediately evaluate for airway compression and anesthesia safety prior to attempting histologic diagnosis

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Indication	Medically Necessary Imaging Study
Individuals with ALL with known or strongly suspected T-cell histology or other suspected lymphoblastic lymphoma involvement	EITHER of the following: ◦ CT Neck (CPT® 70491), CT Chest (CPT® 71260), and CT Abdomen and Pelvis (CPT® 74177) with contrast OR ◦ PET/CT (CPT® 78816) ◦ Indications and coding for rare circumstances where PET/MRI is considered medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**
Individuals with ALL exhibiting CNS symptoms	MRI Brain without and with contrast (CPT® 70553)
Urgent concerns where MRI would delay care (i.e., suspected CNS bleeding) in individuals with ALL	CT Head without or with contrast (CPT® 70450 or CPT® 70460)

Additional imaging criteria in lymphoblastic lymphoma/lymphomatous extramedullary disease:

Indication	Medically Necessary Imaging Study
• To assess response to therapy **only** for individuals with ALL with known bulky nodal disease (usually with T-cell histology) at the end of induction (4 to 6 weeks) • Individuals with residual masses every new therapy phase (Consolidation, Interim maintenance, etc., generally every 8 to 12 weeks) until disease resolution is seen	CT (contrast as requested)

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Indication	Medically Necessary Imaging Study
Individuals with ALL when residual mass ≥8mm in diameter is present on recent CT imaging and immediate radiation or chemotherapy plan will be based on results • Residual mass of any size with no PET-avidity is considered a complete response at the extranodal/ lymphomatous site	PET/CT (CPT® 78815)
Further surveillance in individuals with ALL, once CT imaging shows no evidence of disease	Chest x-ray or Abdominal ultrasound (CPT® 76700) **only**, as indicated by site(s) of bulky disease present at diagnosis
Individuals with ALL with persistent residual masses performed as part of an end of therapy evaluation	CT of all involved bulky nodal areas (contrast as requested)

Immunosuppression criteria during ALL therapy:

- Individuals with ALL are severely immunocompromised during the first 4 to 6 weeks of treatment (induction) and any conventional imaging request to evaluate for infectious complications during this time frame is considered medically necessary

Indication	Medically Necessary Imaging Study
For infectious disease concerns for individuals with ALL with: • Absolute neutrophil count (ANC) <500 or • Inconclusive findings on chest x-ray or ultrasound at any ANC during active treatment	CT or MRI (contrast as requested)
Individuals with ALL with neurologic symptoms with a history of treatment with methotrexate	MRI Brain without and with contrast (CPT® 70553)
To rule out bleeding associated with sinus venous thrombosis in individuals with ALL treated with asparaginase	MRA/MRV of the head (CPT® 70544, 70545, or 70546)

Imaging criteria during therapy for relapsed ALL:

- Frequent CT or MRI imaging is considered medically necessary in individuals with ALL to evaluate known or suspected new sites of invasive fungal or other aggressive infections
- Surveillance imaging of asymptomatic individuals with ALL to detect invasive fungal infection is considered medically necessary **only** when acute clinical decisions will be made based on the imaging

Imaging criteria of known or suspected osteonecrosis in ALL:

- See: **Osteonecrosis In Long Term Cancer Survivors (PEDONC-19.4)** for information on osteonecrosis in ALL individuals who have completed therapy

Indication	Medically Necessary Imaging Study
Symptoms suggesting osteonecrosis in individuals with ALL	MRI without contrast or without and with contrast of the affected joint(s) • CT without contrast is considered medically necessary when MRI is contraindicated or unavailable, or for diagnosis of suspected subchondral fracture
Once at 6 to 9 months after diagnosis for individuals with ALL age ≥11 years of age or older	MRI Bilateral Hips (CPT® 73721 or CPT® 73723 with modifier -50)
Reintroduction of corticosteroids is being considered in individuals with ALL whose symptoms have resolved and are still receiving active treatment	Repeat MRI without contrast of the affected joint(s) every 2 cycles of maintenance (every 6 months)
Preoperative planning for individuals with ALL undergoing core decompression	MRI without contrast of the affected joint(s)

Background and Supporting Information

- ALL is the most common pediatric malignancy and is categorized into B-cell and T-cell subtypes, each with distinct patterns of risk stratification, treatment response, and complication profiles (NCCN, 2025).
- Most children with ALL have diffuse marrow disease without bulky masses; mediastinal involvement is more common in T-cell ALL, while CNS and testicular disease represent classic extramedullary sites (NCCN, 2025).
- Corticosteroid-based therapy is associated with osteonecrosis, particularly in adolescents, with peak incidence approximately one year after diagnosis (Expert Panel on Musculoskeletal Imaging, 2022).

- Treatment-related hypogammaglobulinemia may occur, necessitating intravenous immunoglobulin replacement and heightened vigilance for infectious complications (NCCN, 2025).

Evidence Discussion

- Because most patients have B-precursor ALL without bulky disease, routine cross-sectional imaging for staging or surveillance is not necessary and does not alter outcomes (NCCN, 2025).
- Patients receiving intravenous immunoglobulin replacement and heightened vigilance or those with severe neutropenia should be managed similarly with respect to infection-directed imaging, supporting targeted studies when clinical signs of infection are present (NCCN, 2025).
- Screening MRI for asymptomatic children less than 11 years of age has not been shown to improve functional outcomes or alter management and is therefore not necessary (Expert Panel on Musculoskeletal Imaging, 2022).
- When osteonecrosis is identified on clinically indicated MRI, modification of corticosteroid exposure, particularly during maintenance phases, may occur, demonstrating a role for imaging once symptoms develop (Expert Panel on Musculoskeletal Imaging, 2022).
- MRI of the brain can allow for discrimination between methotrexate neurotoxicity and posterior reversible encephalopathy syndrome (NCCN, 2025).
- CNS-directed imaging can detect clinically significant complications such as leukemic meningeal involvement or intracranial hemorrhage, which directly impact urgent management decisions; NCCN recommends neuroimaging in the presence of neurologic signs or symptoms to evaluate for CNS disease or bleeding, emphasizing a symptom-driven approach that avoids routine imaging in asymptomatic patients while ensuring timely diagnosis of serious complications (NCCN, 2025).
- Evidence supports symptom-driven imaging strategies that identify clinically meaningful complications while avoiding unnecessary sedation, cost, and radiation exposure in children with ALL (NCCN, 2025; Expert Panel on Musculoskeletal Imaging, 2022).

Health Equity Considerations

ALL is the most common pediatric malignancy in the United States, and despite major therapeutic advances with overall survival now exceeding 90%, substantial disparities in outcomes persist across racial, ethnic, and socioeconomic groups. Large population-based analyses demonstrate that Black and Hispanic children experience significantly lower overall survival and higher mortality risk compared with non-Hispanic White and Asian children, even after adjustment for insurance status, income, and comorbidities (Mishra et al., 2024; Winestone & Aplenc, 2019). Socioeconomic disadvantage is a powerful and independent driver of inequity; meta-analytic data show that children with

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ALL from lower socioeconomic status (SES) households have nearly double the risk of death compared with higher SES peers, with particularly pronounced gaps in the United States (Petridou et al., 2015). More recent registry studies further demonstrate that neighborhood-level deprivation remains associated with excess mortality even among children who survive the first 2 and 5 years after diagnosis, underscoring that equity gaps extend beyond induction therapy into long-term survivorship (Hoppmann et al., 2025).

Disparities in ALL outcomes reflect the intersection of structural, social, and biological factors that have direct implications for imaging and surveillance strategies. Limited access to high-quality comprehensive cancer centers, inconsistent follow-up, barriers to advanced diagnostic imaging, and challenges with adherence during prolonged therapy disproportionately affect children from low SES neighborhoods and marginalized racial and ethnic groups (Winestone & Aplenc, 2019; Burke & Thummel, 2020). At the same time, emerging evidence indicates that genetic ancestry and disease biology contribute to inequities; Latino children are more likely to harbor unfavorable cytogenetic features and to live in economically disadvantaged neighborhoods, compounding risk from both biological and social determinants (Kore et al., 2026). Equitable access to diagnostic and surveillance imaging, care coordination across treatment and survivorship, and integration of social context and ancestry informed risk stratification are needed to reduce preventable disparities in outcomes across diverse pediatric populations.

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Acute Myeloid Leukemia (AML) (PEDONC-3.3)

ONCP.LE.0003.3.A
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Criteria

- Frequent CT or MRI imaging is considered medically necessary to evaluate known sites of invasive fungal infection in individuals with AML
- Surveillance imaging of asymptomatic individuals with AML is considered medically necessary to detect invasive fungal infection **only** when acute clinical decisions will be made based on the imaging
- Advanced imaging may be approved on a case-by-case basis for rare individuals with AML with bulky tumor masses (commonly referred to as chloromas, leukemic sarcomas, or myeloid sarcomas) noted on physical examination or other imaging such as plain film or ultrasound

Background and Supporting Information

- AML represents a biologically heterogeneous group of leukemias with aggressive clinical behavior and a higher likelihood of systemic complications at presentation compared with ALL (NCCN, 2025).
- Myeloid sarcomas and CNS involvement can occur but are uncommon overall; AML typically presents without bulky nodal disease (NCCN, 2025).
- Cytogenetic and molecular abnormalities define AML risk groups and therapeutic approaches, reducing reliance on imaging-based disease assessment (NCCN, 2025).

Evidence Discussion

- The majority of pediatric individuals with AML do not have bulky or anatomically localized disease, and routine advanced imaging has not been shown to improve diagnostic accuracy or outcomes (NCCN, 2025).
- Imaging is appropriate when symptoms suggest leukostasis, extramedullary involvement, CNS disease, infection, or hemorrhage, as these findings may affect urgent management decisions (NCCN, 2025).
- Restricting imaging to clinically indicated scenarios minimizes radiation exposure and procedural risk while preserving the ability to detect life threatening complications in AML (NCCN, 2025).

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Health Equity Considerations

AML is a rare but highly aggressive pediatric malignancy associated with substantial treatment-related morbidity and mortality, and despite advances in risk stratification and supportive care, marked disparities in outcomes persist across racial, ethnic, and socioeconomic groups. Large institutional and population-based analyses demonstrate that Black children with AML are significantly more likely to present with high leukemic burden, metabolic derangements, and laboratory evidence of multiorgan dysfunction at diagnosis, factors that are strongly associated with increased induction-related complications and early mortality (Winestone et al., 2024). Survival inequities extend beyond initial presentation; analyses of cooperative group and registry data show that Black and Hispanic children experience inferior event-free and overall survival compared with non-Hispanic White children, including within cytogenetic subgroups traditionally considered favorable, such as core binding factor AML (Conneely et al., 2021). At a global level, children in lower sociodemographic index regions continue to bear a disproportionate burden of AML-related mortality and disability, reflecting persistent gaps in early diagnosis, access to advanced diagnostics, and availability of definitive therapy despite overall improvements in incidence and survival (Jia et al., 2025).

Disparities in AML outcomes reflect a complex interplay of structural, social, and biological factors with direct implications for diagnostic imaging and disease monitoring. Barriers to timely evaluation, limited access to high volume pediatric oncology centers, and inconsistent follow-up disproportionately affect children from socioeconomically disadvantaged communities and historically marginalized racial and ethnic groups, contributing to later presentation and higher acuity at diagnosis (Winestone et al., 2024). Emerging evidence further indicates that biologic differences contribute to inequity; pharmacogenomic variants influencing cytarabine metabolism and treatment response are more prevalent among Black pediatric patients, and standard intensity induction therapy is associated with inferior outcomes unless treatment is appropriately intensified (Lamba et al., 2024). Equitable access to diagnostic and surveillance imaging, timely identification of treatment-related complications, and integration of biological risk with social context are needed to support earlier intervention and reduce preventable disparities across diverse pediatric populations (Conneely et al., 2021; Jia et al., 2025).

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CNS Tumors (PEDONC-4)

Guideline Page

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CNS Tumors General Considerations (PEDONC-4.1)

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v1.0.2027

Criteria

- The classification of pediatric central nervous tumors has recently been revised to incorporate molecular biomarkers in addition to histology, immunohistochemical results, and ultrastructure characteristics. The changes in nomenclature are incorporated in these guidelines.
- Central nervous system tumors are the second most common form of childhood cancer, accounting for ~20% of all pediatric malignancies.

Red Flag Symptoms Raising Suspicion for CNS Tumors Include:
Any headache complaint from a child age ≤5 years
Headaches awakening from sleep
Focal findings on neurologic exam
Clumsiness (common description of gait or coordination problems in young children)
Headaches associated with morning nausea/vomiting
New onset of seizure activity with focal features
Papilledema on physical exam
Loss of developmental milestones (infants and young children)

MRI Considerations

- MRI is the preferred imaging modality for all pediatric CNS tumors
  - MRI Brain without and with contrast (CPT® 70553) is the primary imaging study for pediatric brain tumors
  - MRI Brain without contrast (CPT® 70551) if requested for initial evaluation of suspected CNS tumor for children able to undergo MRI without sedation

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  - Initial MRI should be performed without and with contrast in order to avoid a second anesthesia exposure in younger children requiring sedation for MRI
- MRI Spine with contrast only (Cervical-CPT® 72142, Thoracic-CPT® 72147, Lumbar-CPT® 72149) can be substituted where MRI Spine without and with contrast is considered medically necessary, if being performed immediately following a contrast-enhanced MRI Brain
- Functional MRI (fMRI) (CPT® 70555 or CPT® 70554) is considered medically necessary to depict spatial relationships between eloquent cortex and neoplasms for preoperative planning and to promote safe resections (following baseline MRI Brain).
- Refer to **Neuronavigation (HD-28.2)** for additional guidance on perioperative imaging.
- MRI Orbits without and with contrast (CPT® 70543) is considered medically necessary in individuals who present with papilledema, altered vision, strabismus, nystagmus, anisocoria, proptosis, ocular cranial nerve palsies, coloboma, or leukocoria.

CT Considerations

- CT is considered medically necessary for evaluation of ventriculomegaly or other operative considerations, or for children who cannot undergo MRI safely
  - CT is considered medically necessary for evaluation of headaches related to head trauma or evaluation of skull or facial bone abnormalities
  - Post-contrast CT is generally not considered medically necessary
- CT is not a recommended study for evaluation of pediatric headache when brain tumor is clinically suspected because of its limited diagnostic accuracy in this area. MRI should be used as first line imaging in these cases
- CT should not be used in place of MRI to avoid sedation in young children when red flag symptoms for CNS tumors are present

MRA/CTA and Perfusion Studies

- MRA or CTA is considered medically necessary **only** for preoperative planning or to clarify inconclusive findings on MRI or CT
- CT and MRI Perfusion
  - See: **CT or MRI Perfusion (HD-24.5)** in the Head Imaging guidelines

MR Spectroscopy (MRS, CPT® 76390)

- MRS is **only** considered medically necessary for use in brain tumors of specified histologies where diagnostic accuracy has been established in peer-reviewed literature
  - See diagnosis-specific guidelines for MRS indications

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- MRS is considered not considered medically necessary for all other histologies and indications not listed in a diagnosis-specific guideline section
- MR spectroscopy is not considered medically necessary for routine surveillance

PET Brain Imaging (CPT® 78608 and CPT® 78609)

- PET Brain Metabolic imaging (CPT® 78608) is **only** considered medically necessary for use in brain tumors of specified histologies where diagnostic accuracy has been established in peer-reviewed literature
  - See diagnosis-specific guidelines for PET indications
- PET Brain Metabolic is not considered medically necessary for routine surveillance
- PET Brain Metabolic imaging is considered not considered medically necessary for all other histologies and indications not listed in a diagnosis-specific guideline section
- PET Brain Perfusion imaging (CPT® 78609) is not considered medically necessary in the evaluation or management of primary CNS tumors
- Fusion PET/CT studies (CPT® 78814, CPT® 78815, or CPT® 78816) are not considered medically necessary in the evaluation or management of primary CNS tumors

Timing and Frequency of Imaging

- Definitive imaging should be completed prior to considering biopsy given the high degree of morbidity associated with operating on the CNS
  - Occasionally biopsy is not necessary because the imaging findings provide a definitive diagnosis
    - Examples include diffuse intrinsic pontine glioma and optic pathway gliomas in an individual with known neurofibromatosis
- Perioperative imaging frequency
  - Children may undergo very frequent imaging in the immediate perioperative period around resection or debulking of a CNS tumor due to the small anatomic spaces involved
    - Requests for imaging during this time period to specifically evaluate postoperative course or ventriculoperitoneal shunt functioning are considered medically necessary
  - MRI Brain without and with contrast (CPT® 70553) is considered medically necessary one time in the immediate preoperative period (even if another study has already been completed) to gain additional information which can be important in optimizing individual outcomes, such as:
    - Completion of additional specialized MRI sequences such as diffusion-tensor imaging
    - Perfusion imaging (CPT® 70553, 70551, OR 70552)
    - Tractography

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    - Other sequences not reported under a separate CPT® code but not part of a routine MRI Brain series
  - Repeat MRI Brain that is being requested solely for loading into operative navigation software should not be requested as a diagnostic code but is considered medically necessary under a treatment planning code (CPT® 76498).
  - **Clinical note**: for all pediatric CNS tumors, cerebrospinal fluid pathology positive for malignancy is considered leptomeningeal/spinal disease.

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CNS Low Grade Gliomas (LGG) (PEDONC-4.2)

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Criteria

- MR Spectroscopy and PET Brain Metabolic are not considered medically necessary for routine surveillance of CNS LGG.
- MRI is generally superior to CT for staging and restaging CNS malignancies, but CT is considered medically necessary in accordance with these guidelines where MRI is contraindicated.
- CNS LGG includes the following tumors:
  - Pediatric-type diffuse low-grade gliomas
    - Diffuse astrocytoma, MYB- or MYBL1-altered
    - Angiocentric glioma
    - Polymorphus low-grade neuroepithelial tumor of the young
    - Diffuse low-grade glioma, MAPK pathway-altered
  - Circumscribed astrocytic gliomas
    - Pilocytic astrocytoma
    - High-grade astrocystoma with piloid features
    - Pleomorphic xanthoastrocytoma
    - Subependymal giant cell astrocystoma (SEGA)
    - Choroid glioma
    - Astroblastoma, MN
  - Glioneuronal and neuronal tumors
    - Ganglioglioneuroma
    - Desmoplastic infantile ganglioglioneuroma/desmoplastic infantile astrocystoma
    - Dysembryoplastic neuroepithelial tumor
    - Diffuse glioneuronal tumor with oligodendroglioma-like features and nuclear clusters
    - Papillary glioneuronal tumor
    - Rosette-forming glioneuronal tumor
    - Myxoid glioneuronal tumor
    - Diffuse leptomeningeal glioneuronal tumor
    - Gangliocytoma Multinodular and vacuolating neuronal tumor
    - Dysplastic cerebellar gangliocytoma (Lhermitte-Duclos disease)
    - Central neurocytoma

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        - Extraventricular neurocytoma
        - Cerebellar liponeurocytoma
    - Any other glial tumor with a WHO grade I or II classification

Indication	Medically Necessary Imaging Study
Initial staging of all LGG	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • MRI Orbits without and with contrast (CPT® 70543) in addition to the above studies for individuals presenting with visual signs or symptoms as listed in **PEDONC-4.1**
In addition to the above imaging, for preoperative planning for individuals with LGG	• Functional MRI (fMRI) (CPT® 70555 or 70554) • For further perioperative indications, please refer to **Neuronavigation (HD-28.2).**
• At any time, for ANY of the following in individuals with LGG: ◦ For rapid assessment in the acute setting ◦ Evaluation of acute intracranial hemorrhage ◦ Evaluation of ventriculomegaly ◦ Evaluation of shunt-related issues	• CT Head without contrast (CPT® 70450)
• ANY of the following for individuals with LGG: ◦ Determining the need for biopsy when transformation to high grade glioma is suspected based on clinical symptoms or recent MRI findings ◦ Evaluation of a brain lesion of indeterminate nature when the PET findings will be used to determine whether biopsy/resection can be safely postponed	• PET Brain Metabolic imaging (CPT® 78608)

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Indication	Medically Necessary Imaging Study
• ANY of the following for individuals with LGG: ◦ Distinguish low grade from high grade gliomas ◦ Evaluate a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed ◦ Distinguish radiation-induced tumor necrosis from progressive disease within 18 months of completing radiotherapy	• MR Spectroscopy (MRS, CPT® 76390)
Baseline imaging after resection for individuals with LGG, to assess degree of resection	• MRI Brain without and with contrast (CPT® 70553) • MRI without and with contrast at level of resected spinal site • If orbital resection, MRI Orbits without and with contrast (CPT® 70543)
Treatment response at the completion of radiotherapy for individuals with LGG	• MRI Brain without and with contrast (CPT® 70553) • MRI without and with contrast at level of irradiated spinal site • If prior orbital involvement, MRI Orbits without and with contrast (CPT® 70543)
Treatment response on chemotherapy for individuals with LGG	• MRI Brain without and with contrast (CPT® 70553) approved every 2 cycles during active treatment and at the end of planned chemotherapy • If prior orbital involvement, MRI Orbits without and with contrast (CPT® 70543)
Additional treatment response imaging during induction chemotherapy for individuals with LGG with measurable spinal cord disease on MRI	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 2 cycles

Indication	Medically Necessary Imaging Study
Surveillance, including individuals with LGG with unresected disease on observation only	• For individuals with intracranial primary: ◦ MRI Brain without and with contrast (CPT® 70553) every 3 months for 2 years, then every 6 months for 3 years, then annually thereafter. ◦ If prior orbital involvement, MRI Orbits without and with contrast (CPT® 70543) ◦ MRI Spine is not considered medically necessary during surveillance in individuals without prior history of spinal involvement except to evaluate symptoms suspicious for spinal cord recurrence • For individuals with a history of spine primary tumor or metastatic spinal involvement: ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 3 months for 2 years, then every 6 months for 3 years, then annually thereafter • For individuals with NF1 **ANY** of the following: ◦ MRI Brain without and with contrast (CPT® 70553 every 3 months for 2 years, then every 6 months for 3 years, then annually thereafter ◦ MRI Orbits without and with contrast (CPT® 70543) every 3 months for 2 years, then every 6 months for 3 years, then annually thereafter
Suspected intracranial or intraspinal recurrence for individuals with LGG	• All imaging supported in initial staging criteria

Background and Supporting Information

- CNS LGGs are World Health Organization (WHO) grade I–II primary brain tumors and account for approximately 40%–60% of pediatric CNS tumors, making them the most common pediatric brain neoplasms (NCCN, 2025).
- These tumors may arise anywhere in the CNS, including the cerebral hemispheres, optic pathways, brainstem, cerebellum, and spinal cord, with clinical behavior influenced by location and resectability (PDQ® Pediatric Treatment Editorial Board, 2025).
- Prognosis is generally favorable, with long term survival exceeding 90% in many subgroups, particularly when gross total resection is achievable (NCCN, 2025).
- Optic pathway gliomas associated with NF1 may be diagnosed radiographically without biopsy due to characteristic imaging features and predictable clinical behavior (PDQ® Pediatric Treatment Editorial Board, 2025).

Evidence Discussion

- Imaging-based diagnosis and surveillance strategies balance early detection of progression with avoidance of unnecessary anesthesia and radiation exposure, particularly in tumors with indolent behavior (NCCN, 2025).
- Evidence supports observation with serial MRI in asymptomatic or completely resected tumors, as early imaging changes do not consistently correlate with clinically meaningful progression (PDQ® Pediatric Treatment Editorial Board, 2025).
- Restricting advanced imaging to situations of clinical concern reduces potential harms without compromising oncologic outcomes (Expert Panel on Neurological Imaging, 2019).

Health Equity Considerations

Pediatric LGGs are the most common childhood central nervous system tumors and are associated with excellent overall survival; however, they carry a substantial and often lifelong burden of disease- and treatment-related morbidity. Despite favorable survival, profound disparities exist in access to timely diagnosis, comprehensive neurosurgical care, molecular diagnostics, and long-term management across socioeconomic and geographic contexts. Global and population-based analyses demonstrate wide variability in incidence, diagnostic delay, and outcomes for pediatric LGGs, with children in low and middle income settings disproportionately affected by underdiagnosis, delayed presentation, and limited access to specialized care (Moreira et al., 2023). Even in high income countries, LGGs frequently present insidiously with nonspecific symptoms, leading to diagnostic delays that exacerbate neurologic morbidity and functional impairment, particularly among children facing barriers to specialty referral and advanced imaging (Moreira et al., 2023).

Disparities in LGG outcomes reflect an intersection of structural resource limitations and chronic disease burden with direct implications for imaging access and surveillance.

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Children with LGGs experience high and persistent healthcare utilization over multiple years, including repeated neuroimaging, specialty consultations, emergency care, and hospitalizations, underscoring the importance of consistent access to diagnostic and follow up imaging to monitor disease progression and treatment-related complications (Zelt et al., 2024). Socioeconomic disadvantage and insurance status influence access to these services and to multidisciplinary care, including neurosurgery, neuro-oncology, endocrinology, and rehabilitation, which are essential for optimizing long-term outcomes in a condition that often becomes chronic (Zelt et al., 2024). Equitable access to timely diagnostic imaging, standardized surveillance strategies across the disease continuum, and coordination with specialized centers are needed to reduce preventable neurologic morbidity and long-term functional disparities among diverse pediatric populations (Moreira et al., 2023; Zelt et al., 2024).

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CNS High Grade Gliomas (HGG) (PEDONC-4.3)

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Criteria

- Individuals with HGG who undergo complete resection should be imaged according to surveillance guidelines after post-resection imaging.
- CNS HGG includes the following tumors:
  - Diffuse midline glioma, H3 K27-altered
  - Diffuse hemispheric glioma, H3 G34-mutant
  - Diffuse pediatric-type high-grade glioma, H3-wildtype and IDH-wildtype
  - Infant-type hemispheric glioma
  - Any other glial tumor with a WHO grade of III or IV classification

Indication	Medically Necessary Imaging Study
Initial staging of all HGG	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
In addition to the above imaging, for preoperative planning for individuals with HGG	• Functional MRI (fMRI) (CPT® 70555 or 70554) • For further perioperative indications, please refer to **Neuronavigation (HD-28.2).**
• At any time, for ANY of the following for individuals with HGG: ◦ For rapid assessment in the acute setting ◦ Evaluation of acute intracranial hemorrhage ◦ Evaluation of ventriculomegaly ◦ Evaluation of shunt-related issues	• CT Head without contrast (CPT® 70450)

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Indication	Medically Necessary Imaging Study
• ANY of the following for individuals with HGG: ◦ Distinguishing radiation-induced tumor necrosis from progressive disease within 18 months of completing radiotherapy ◦ Evaluating inconclusive MRI findings when the PET findings will be used to determine need for biopsy or change in therapy, including a change from active therapy to surveillance ◦ Evaluation of a brain lesion of indeterminate nature when the PET findings will be used to determine whether biopsy/resection can be safely postponed	• PET Brain Metabolic Imaging (CPT® 78608)
• ANY of the following for individuals with HGG: ◦ To distinguish low grade from high grade gliomas ◦ To evaluate a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed ◦ To distinguish radiation-induced tumor necrosis from progressive disease within 18 months of completing radiotherapy	• MR Spectroscopy (MRS, CPT® 76390)
• To depict spatial relationships between eloquent cortex and tumor prior to resection for individuals with HGG	• Functional MRI (fMRI) (CPT® 70544 or CPT® 70555)

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Indication	Medically Necessary Imaging Study
Baseline imaging following resection, to assess degree of resection for individuals with HGG	• MRI Brain without and with contrast (CPT® 70553) AND any of the following • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Treatment response at the completion of radiotherapy for individuals with HGG	• MRI Brain without and with contrast (CPT® 70553) AND any of the following: • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Treatment response on chemotherapy for individuals with HGG	• MRI Brain without and with contrast (CPT® 70553) every 2 cycles during active treatment and at the end of planned chemotherapy AND • For those with known spinal involvement, ANY of the following: ◦ MRI Spine without and with contrast of any and all levels requested every 2 cycles (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • For those with no known spinal involvement, ANY of the following: ◦ MRI Spine without and with contrast of any and all levels requested at end of therapy (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Signs or symptoms of recurrence or progression in individuals with HGG	• MRI Brain without and with contrast (CPT® 70553) AND any of the following • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

Indication	Medically Necessary Imaging Study
Surveillance for individuals with HGG	• For individuals with intracranial primary: ◦ MRI Brain without and with contrast (CPT® 70553) every 2 months for 1 year, then every 3 months thereafter ◦ MRI Spine is not considered medically necessary during surveillance in individuals without prior history of spinal involvement except to evaluate symptoms suspicious for spinal cord recurrence • For individuals with a history of spine primary tumor or metastatic spinal involvement: ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 2 months for 1 year, then every 3 months thereafter • For both brain and spine disease, imaging above may be complemented by MR perfusion and/or MR spectroscopy, and these studies are considered medically necessary if requested.

Background and Supporting Information

- Pediatric CNS HGGs are WHO grade III–IV tumors and comprise approximately 10%–20% of pediatric CNS malignancies (NCCN, 2025).
- These tumors are biologically distinct from adult gliomas, frequently harboring pediatric-specific molecular alterations such as H3 K27 altered or H3 G34 mutant profiles (NCCN, 2025).
- Prognosis remains poor, with 5-year overall survival typically less than 20%, despite aggressive multimodality therapy (PDQ® Pediatric Treatment Editorial Board, 2025).
- Tumors may arise throughout the brain and less commonly in the spinal cord, with location influencing surgical and imaging approaches (PDQ® Pediatric Treatment Editorial Board, 2025).

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Evidence Discussion

- Baseline and post-treatment MRI of the brain (and spine when clinically indicated) is supported by evidence demonstrating its importance in assessing tumor burden, treatment response, and progression (NCCN, 2025).
- Routine surveillance imaging allows early identification of recurrence, which may inform eligibility for clinical trials or salvage therapy, despite limited impact on overall survival (PDQ® Pediatric Treatment Editorial Board, 2025).
- Limiting imaging frequency to periods of highest recurrence risk reduces cumulative anesthesia exposure without compromising clinical decision making (Expert Panel on Neurological Imaging, 2019).

Health Equity Considerations

HGGs in children and adolescents are rare but devastating central nervous system malignancies with poor overall prognosis, and substantial disparities in access to care and treatment persist across racial, ethnic, and socioeconomic groups. National database analyses in the United States demonstrate significant racial differences in insurance status, household income, educational attainment, and access to surgical intervention for pediatric malignant gliomas, with Black children significantly less likely to undergo surgical resection compared with White children (Lu & Niazi, 2022). These disparities are not fully explained by tumor location alone and reflect broader structural inequities that may influence diagnostic evaluation, referral patterns, and treatment planning. In low and middle income settings, emerging data suggest rising incidence of HGGs among younger populations accompanied by limited access to comprehensive molecular profiling and advanced therapies, with poor survival outcomes underscoring global inequities in neuro-oncologic care delivery (Rehman et al., 2026).

Disparities in HGG outcomes reflect the intersection of aggressive tumor biology and structural barriers with direct implications for imaging and disease management. Limited access to high quality diagnostic imaging, advanced neurosurgical services, and molecular characterization can delay accurate diagnosis, limit eligibility for maximal safe resection, and constrain tailoring of adjuvant therapy, disproportionately affecting socioeconomically disadvantaged and historically marginalized populations (Lu & Niazi, 2022). In resource constrained settings, incomplete availability of molecular testing further compounds inequity by limiting prognostication and treatment stratification, despite evidence that molecular features such as IDH mutation status and MGMT promoter methylation have important implications for management and outcomes (Rehman et al., 2026). Standardized access to timely diagnostic and follow-up imaging, equitable referral to specialized neuro-oncology centers, and integration of imaging with molecular and clinical risk assessment are needed to reduce preventable disparities in care and outcomes across diverse pediatric populations (Lu & Niazi, 2022; Rehman et al., 2026).

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Medulloblastoma (MDB), Other CNS Embryonal Tumors, and Pineoblastoma (PEDONC-4.4)

ONCP.CT.0004.4.A
v1.0.2027

Criteria

- Includes the following tumors:
  - Medulloblastoma
    - Molecularly-defined Medulloblastoma
      - WNT-activated Medulloblastoma
      - SHH-activated and TP53-wildtype Medulloblastoma
      - SHH-activated and TP53-mutant Medulloblastoma
      - Non-WNT/Non-SHH
    - Medulloblastoma, histologically defined
  - Other CNS Embryonal Tumors (previously supratentorial primitive neuro-ectodermal tumors)
    - CNS embryonal tumor
    - Cribriform neuroepithelial tumor
    - Embryonal tumor with multilayered rosettes
    - CNS neuroblastoma
    - FOXR2-activated CNS tumor with BCOR internal tandem duplication
  - Pineoblastoma
- Risk assessment is important in determining optimal treatment. High-risk features include the following:
  - Spinal metastasis (including cytology positive only)
  - Multifocal intracranial tumors
  - Anaplastic histology
  - All other CNS embryonal tumors and pineoblastomas
  - >1.5cm^2 residual tumor area on postoperative MRI and age <3 years
- Individuals without any high-risk features are considered "average risk"
- MR Spectroscopy and PET Brain Metabolic are not considered medically necessary for routine surveillance for individuals with MDB, other CNS embryonal tumors, and pineoblastoma.

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Indication	Medically Necessary Imaging Study
Initial staging for all individuals with MDB, other CNS embryonal tumors, and pineoblastoma, preoperatively	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
In addition to the above imaging, for preoperative planning for individuals with MDB, other CNS embryonal tumors, and pineoblastoma	• Functional MRI (fMRI) (CPT®) 70555 or 70554) • For further perioperative indications, please refer to **Neuronavigation (HD-28.2)**
• At any time, for ANY of the following for individuals with MDB, other CNS embryonal tumors, and pineoblastoma: ◦ For rapid assessment in the acute setting ◦ Evaluation of acute intracranial hemorrhage ◦ Evaluation of ventriculomegaly ◦ Evaluation of shunt-related issues	• CT Head without contrast (CPT® 70450)
• ANY of the following for individuals with MDB, other CNS embryonal tumors, and pineoblastoma: ◦ To distinguish radiation-induced tumor necrosis from progressive disease within 18 months of completing radiotherapy ◦ To evaluate inconclusive MRI findings when the PET findings will be used to determine need for biopsy or change in therapy, including a change from active therapy to surveillance ◦ To evaluate a brain lesion of indeterminate nature when the PET findings will be used to determine whether biopsy/resection can be safely postponed	• PET Brain Metabolic Imaging (CPT® 78608)

Indication	Medically Necessary Imaging Study
• ANY of the following for individuals with MDB, other CNS embryonal tumors, and pineoblastoma: ◦ To evaluate a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed	• MR Spectroscopy (CPT® 76390)
Postoperative (preferably within 48 hours of surgery) to quantify residual tumor volume for individuals with MDB, other CNS embryonal tumors, and pineoblastoma	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic (CPT® 72157, Lumbar-CPT® 72158) if spinal disease was resected
Within 28 days post-op, if spinal imaging was not performed preoperatively for individuals with MDB, other CNS embryonal tumors, and pineoblastoma	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic (CPT® 72157, Lumbar-CPT® 72158)
Treatment response for individuals with MDB, other CNS embryonal tumors, and pineoblastoma	• At the start of adjuvant chemotherapy and every 2 cycles until therapy is completed: ◦ MRI Brain without and with contrast (CPT® 70553) ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic (CPT® 72157, Lumbar-CPT® 72158)
Children age <3 years with MDB, other CNS embryonal tumors, and pineoblastoma treated with multiple cycles of high dose chemotherapy with autologous stem cell rescue in lieu of radiotherapy	• Disease evaluations (imaging per treatment response guidelines) prior to each cycle (every 4 to 6 weeks) if needed for response determination.

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Indication	Medically Necessary Imaging Study
End of treatment evaluation for individuals with MDB, other CNS embryonal tumors, and pineoblastoma	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine with contrast (Cervical-CPT® 72142, Thoracic-CPT® 72147, Lumbar-CPT® 72149) or MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Signs or symptoms of recurrence or progression in individuals with MDB, other CNS embryonal tumors, and pineoblastoma	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Surveillance for individuals with MDB, other CNS embryonal tumors, and pineoblastoma	For low- or average-risk individuals: • Every 3 months for 2 years, then every 6 months for 3 years: ◦ MRI Brain without and with contrast (CPT® 70553) • Every 6 months for 2 years: ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) For high- or very high-risk individuals: • Every 3 months for 2 years, then every 6 months for 3 years: ◦ MRI Brain without and with contrast (CPT® 70553) • Every 3 months for 2 years, then every year for 3 years: ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • For additional imaging guidelines for individuals in long-term follow-up after CNS tumor treatment that included radiation therapy, see:

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Background and Supporting Information

- Medulloblastoma and other CNS embryonal tumors collectively account for approximately 15%–25% of pediatric CNS tumors and are characterized by a high propensity for cerebrospinal fluid dissemination (PDQ® Pediatric Treatment Editorial Board, 2025).
- Medulloblastomas are molecularly classified into WNT-activated, SHH-activated (TP53 wildtype or mutant), and non-WNT/non-SHH subgroups, which guide risk stratification and therapy (NCCN, 2025).
- Pineoblastomas share treatment paradigms with embryonal tumors due to similar biological behavior (NCI, 2025).
- Risk stratification incorporates age, metastatic status, extent of resection, and molecular features (NCCN, 2025).

Evidence Discussion

- Whole neuroaxis MRI is supported by evidence demonstrating frequent leptomeningeal spread at diagnosis and recurrence, influencing staging and treatment planning (PDQ® Pediatric Treatment Editorial Board, 2025).
- Surveillance imaging detects asymptomatic recurrence but has limited impact on long term survival, supporting guideline-directed imaging intervals rather than intensive scanning (NCCN, 2025; Shepherd, 2026).
- Avoidance of unnecessary imaging reduces sedation-related risks and healthcare burden in long term survivors (Expert Panel on Neurological Imaging, 2019).

Health Equity Considerations

MDB, other central nervous system embryonal tumors, and pineoblastoma are rare, biologically heterogeneous pediatric malignancies that require timely diagnosis, advanced imaging, and risk-adapted multimodal therapy to optimize outcomes. Despite substantial advances in molecular classification and survival for some subgroups, significant disparities persist across racial, ethnic, socioeconomic, and geographic contexts. Population-based and multicenter studies demonstrate that children from socioeconomically disadvantaged backgrounds and those treated outside high volume or specialized centers experience delays in diagnosis, reduced access to molecular subgrouping, and inferior survival outcomes (Dressler et al., 2018; Kopecký et al., 2017). International analyses further highlight pronounced survival gaps between high income countries and low and middle income settings, driven by limited access to neuroimaging, neurosurgical expertise, craniospinal irradiation, and comprehensive supportive care (Greppin et al., 2022; Díaz Coronado et al., 2025).

Disparities in outcomes for embryonal CNS tumors reflect the interaction of aggressive disease biology and structural barriers with direct implications for imaging and disease monitoring. Accurate risk stratification for MDB and related tumors increasingly depends

on advanced imaging and molecular diagnostics, yet access to these resources remains uneven, particularly for historically marginalized populations and patients treated in under-resourced health systems (Hansford et al., 2022; Peterson et al., 2025). Limited availability of serial surveillance imaging and specialized follow-up further compounds inequity, as survivors of these tumors face high risks of relapse and long-term neurologic and neurocognitive morbidity (Ronsley et al., 2024; Dressler et al., 2018). Standardized access to timely diagnostic and surveillance imaging, referral to specialized pediatric neuro-oncology centers, and integration of imaging with molecular and clinical risk assessment are needed to reduce preventable disparities in outcomes across diverse pediatric populations (Greppin et al., 2022; Peterson et al., 2025).

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Atypical Teratoid/Rhabdoid Tumors (ATRT) (PEDONC-4.5)

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Criteria

- PET Brain Metabolic does not have a defined role in the evaluation of ATRT at this time and is, therefore, not considered medically necessary.
- MR Spectroscopy is not considered medically necessary for routine surveillance of ATRT.
- Rhabdoid malignancies occurring outside the CNS should be imaged according to **Malignant Rhabdoid Tumor of the Kidney (MRT) and Other Extracranial Sites (PEDONC-7.6)**.
- MRI performed at the end of consolidation therapy should serve as the diagnostic MRI prior to radiotherapy in individuals with ATRT.

Indication	Medically Necessary Imaging Study
Initial staging for all individuals with ATRT, preoperatively	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • Renal US (CPT® 76770) ◦ If renal US is abnormal, refer to: **Malignant Rhabdoid Tumor of the Kidney (MRT) and Other Extracranial Sites (PEDONC-7.6)**
In addition to the above imaging, for preoperative planning in individuals with ATRT	• Functional MRI (fMRI) (CPT® 70555 or 70554) • For further perioperative indications, please refer to **Neuronavigation (HD-28.2)**

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Indication	Medically Necessary Imaging Study
• At any time, for ANY of the following in individuals with ATRT: ◦ For rapid assessment in the acute setting ◦ Evaluation of acute intracranial hemorrhage ◦ Evaluation of ventriculomegaly ◦ Evaluation of shunt-related issues	• CT Head without contrast (CPT® 70450)
Evaluation of a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed in individuals with ATRT	• MR Spectroscopy (CPT® 76390)
Postoperative (preferably within 48 hours of surgery) to quantify residual tumor volume in individuals with ATRT	• MRI Brain without and with contrast (CPT® 70553)
Within 28 days post-op, if spinal imaging was not performed preoperatively in individuals with ATRT	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Treatment response to induction chemotherapy in individuals with ATRT	• After every 2 cycles: ◦ MRI Brain without and with contrast (CPT® 70553) ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Individuals treated with consolidation chemotherapy and autologous stem cell rescue in individuals with ATRT	• Disease evaluation is considered medically necessary following the end of the planned stem cell rescues but may occur prior to each cycle (every 4 to 6 weeks) if needed for response determination

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Indication	Medically Necessary Imaging Study
End of treatment evaluation in individuals with ATRT	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Signs or symptoms of recurrence or progression in individuals with ATRT	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Surveillance in individuals with ATRT	• Every 3 months for 2 years, then every 6 months for 3 years, then annually for 10 years: ◦ MRI Brain without and with contrast (CPT® 70553) ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see: **Second Malignant Neoplasms (SMN) (PEDONC-19.3)**

Background and Supporting Information:

- ATRT is a rare but highly aggressive embryonal tumor occurring primarily in infants and young children, often associated with loss of SMARCB1 (INI1) expression (PDQ® Pediatric Treatment Editorial Board, 2025).
- These tumors commonly demonstrate early leptomeningeal dissemination and may coexist with extracranial rhabdoid tumors, particularly in the kidney (NCCN, 2025).
- Overall prognosis is poor, with survival rates generally below 30% despite intensive multimodal therapy (PDQ® Pediatric Treatment Editorial Board, 2025).

Evidence Discussion

- MRI of the brain and spine is essential for staging and response assessment due to the high incidence of multifocal and metastatic disease (NCCN, 2025).

- Imaging following completion of systemic therapy guides radiation planning but should be limited thereafter to clinically meaningful intervals to avoid cumulative harm (Expert Panel on Neurological Imaging, 2019).

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Pineocytomas and Pineal Parenchymal Tumors (PEDONC-4.6)

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Criteria

- PET Brain Metabolic imaging and MR Spectroscopy do not have a defined role in the evaluation of pineocytoma.
- Individuals with a complete resection for pineocytomas and pineal parenchymal tumors should be imaged according to surveillance guidelines
- After end of radiotherapy imaging, individuals with pineocytomas and pineal parenchymal tumors should be imaged according to surveillance guidelines

Indication	Medically Necessary Imaging Study
Initial staging for all individuals with pineocytomas and pineal parenchymal tumors	• MRI Brain without and with contrast (CPT® 70553)
In addition to the above imaging, for preoperative planning in individuals with pineocytomas and pineal parenchymal tumors	• Functional MRI (fMRI) (CPT® 70555 or 70554) • For further perioperative indications, please refer to **Neuronavigation (HD-28.2)**
• Additional initial staging imaging for individuals with pineocytomas and pineal parenchymal tumors with: ◦ Multicentric tumors ◦ Atypical histology including pineoblastoma-like elements (grade 2 or 3 pineal parenchymal tumor which have not been considered a pineoblastoma) ◦ Clinical signs or symptoms suggesting spinal cord involvement	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
Baseline imaging following resection in individuals with pineocytomas and pineal parenchymal tumors	• MRI Brain without and with contrast (CPT® 70553)
End of radiotherapy in individuals with pineocytomas and pineal parenchymal tumors	• MRI Brain without and with contrast (CPT® 70553)
Additional imaging at end of radiotherapy for individuals individuals with pineocytomas and pineal parenchymal tumors with measurable spinal cord disease on MRI	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Surveillance in individuals with pineocytomas and pineal parenchymal tumors	• MRI Brain without and with contrast (CPT® 70553) every 3 months for 1 year, then every 4 months for 1 year, then every 6 months for 1 year, then annually thereafter
Additional surveillance imaging for individuals with pineocytomas and pineal parenchymal tumors with cord involvement at diagnosis	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) after completion of therapy every 3 months for 1 year, then every 4 months for 1 year, then every 6 months for 1 year, then annually thereafter
Signs or symptoms of recurrence or progression in individuals with pineocytomas and pineal parenchymal tumors	• MRI Brain without and with contrast (CPT® 70553)
Suspected spinal cord recurrence or progression in individuals with pineocytomas and pineal parenchymal tumors	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

Background and Supporting Information

- Pineocytomas and related pineal parenchymal tumors are generally low grade neoplasms with favorable outcomes following surgical resection (PDQ® Pediatric Treatment Editorial Board, 2025).
- These tumors may present with hydrocephalus due to obstruction of the aqueduct of Sylvius (NCCN, 2025).

Evidence Discussion

- Postoperative MRI establishes a baseline for surveillance, while long-term imaging is reserved for incomplete resection or adjuvant therapy scenarios (NCCN, 2025).
- Limiting advanced imaging in asymptomatic individuals avoids unnecessary interventions without impacting cure rates (Expert Panel on Neurological Imaging, 2019).

Health Equity Considerations

Pineocytomas and other pineal parenchymal tumors are rare pediatric central nervous system neoplasms with heterogeneous clinical behavior and outcomes that vary substantially by age at diagnosis, histologic subtype, and disease stage. Population-based analyses demonstrate that younger children, particularly those under 5 years of age, and patients with more aggressive subtypes, such as pineoblastoma, experience significantly worse survival outcomes compared with older children and those with less aggressive histologies (Shen et al., 2025). Although racial differences in survival were not statistically significant in large registry data, notable variation exists in tumor stage at diagnosis and use of radiation therapy, highlighting potential inequities in early detection, referral patterns, and access to specialized oncologic care for children with pineal parenchymal tumors (Shen et al., 2025).

Disparities in outcomes for pineocytomas and pineal parenchymal tumors reflect the interaction of rare tumor biology with structural differences in access to diagnostic imaging and definitive therapy. Advanced neuroimaging is critical for accurate staging and treatment planning in pineal region tumors, yet delays in diagnosis can result in more advanced disease at presentation, which is independently associated with inferior survival (Shen et al., 2025). Radiation therapy has been shown to significantly improve survival for pediatric patients with malignant pineal region tumors, underscoring the importance of equitable access to radiation oncology services and longitudinal imaging surveillance to guide therapy and monitor response (Shen et al., 2025). Timely diagnostic imaging, standardized staging and follow-up protocols, and referral to specialized pediatric neuro-oncology centers are needed to reduce preventable disparities in outcomes for children with pineocytomas and other pineal parenchymal tumors.

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CNS Germinomas and Non-Germinomatous Germ Cell Tumors (NGGCT) (PEDONC-4.7)

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v1.0.2027

Criteria

- PET Metabolic Brain imaging does not have a defined role in the evaluation of CNS Germinomas and NGGCT and is, therefore, considered not medically necessary.
- Includes the following tumors:
  - CNS Germinoma
  - Non-Germinomatous Germ Cell Tumors (NGGCT)
    - Embryonal carcinoma
    - Yolk sac tumor
    - Choriocarcinoma
    - Immature Teratoma
    - Mature Teratoma
    - Mixed germ cell tumor

Indication	Medically Necessary Imaging Study
Initial staging for all individuals with germinomas and NGGCT	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
In addition to the above imaging, for preoperative planning in individuals with germinomas and NGGCT	• Functional MRI (fMRI) (CPT® 70555 or 70554) • For further perioperative indications, please refer to **Neuronavigation (HD-28.2)**

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Indication	Medically Necessary Imaging Study
• At any time, for ANY of the following in individuals with germinomas and NGGCT: ◦ For rapid assessment in the acute setting ◦ Evaluation of acute intracranial hemorrhage ◦ Evaluation of ventriculomegaly ◦ Evaluation of shunt-related issues	• CT Head without contrast (CPT® 70450)
Evaluation of a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed in individuals with germinomas and NGGCT	• MR Spectroscopy (CPT® 76390)
Treatment response to induction chemotherapy in individuals with germinomas and NGGCT	• MRI Brain without and with contrast (CPT® 70553) every 2 cycles
Additional treatment response to induction chemotherapy for individuals with germinomas and NGGCTwith measurable spinal cord disease on MRI	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 2 cycles
End of induction chemotherapy for individuals with germinomas and NGGCT with localized intracranial tumors	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Prior to second-look surgery in individuals with germinomas and NGGCT	• MRI of all known sites of measurable disease
Prior to radiotherapy in individuals with germinomas and NGGCT	• MRI of all known sites of measurable disease
At the end of all planned therapy in individuals with germinomas and NGGCT	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine (with or without and with contrast)

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Indication	Medically Necessary Imaging Study
Signs or symptoms of recurrence or progression in individuals with germinomas and NGGCT	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Surveillance in individuals with germinomas and NGGCT	• Every 3 months for 1 year, then every 4 months for 1 year, then every 6 months for 1 year, then annually until 5 years after completion of therapy: ◦ MRI Brain without and with contrast (CPT® 70553) ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see:
Suspected recurrence - new or worsening neurologic symptoms (including worsening of diabetes insipidus) in individuals with germinomas and NGGCT	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

Background and Supporting Information

- Germinomas and NGGCT most frequently affect older school aged children and adolescents, though they can occur across the pediatric age spectrum (PDQ® Pediatric Treatment Editorial Board, 2025).
- Germinomas are highly sensitive to chemotherapy and radiotherapy and are associated with excellent long-term survival rates, often exceeding 90%, whereas NGGCTs have a less favorable prognosis and require more intensive therapy (NCCN, 2025).
- Leptomeningeal dissemination is common at diagnosis, supporting the need for neuraxis evaluation during staging (PDQ® Pediatric Treatment Editorial Board, 2025).

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Evidence Discussion

- Brain and spine MRI at diagnosis is supported by evidence demonstrating frequent multifocal and metastatic disease that alters staging and treatment planning (NCCN, 2025).
- Surveillance imaging following chemotherapy and/or radiotherapy enables detection of residual or recurrent disease while avoiding unnecessary imaging once disease stability is established (PDQ® Pediatric Treatment Editorial Board, 2025).
- Guideline-directed imaging intervals help minimize anesthesia exposure and cumulative imaging burden without compromising excellent outcomes in germinoma patients (Expert Panel on Neurological Imaging, 2019).

Health Equity Considerations

Germinomas and NGGCTs are rare pediatric malignancies with distinct biology, treatment intensity, and long term outcomes, yet substantial disparities persist in presentation patterns, survival, and survivorship burden across sex, age, and resource settings. Population-based analyses demonstrate marked sex-based disparities, with males experiencing higher incidence and, particularly for NGGCTs, higher early mortality risk compared with females (Lin et al., 2025). Disease stage at diagnosis is a critical determinant of outcomes; patients presenting with distant or metastatic disease experience significantly worse survival, highlighting the impact of delayed diagnosis and barriers to timely neuroimaging and comprehensive staging (Lin et al., 2025). Global and regional cohort studies further demonstrate that children treated in low and middle income or upper middle income settings face challenges related to access to standardized molecular diagnostics, advanced imaging, and treatment intensification strategies, contributing to outcome variability, especially for NGGCTs (Cappellano et al., 2024).

Equity considerations extend beyond initial cure to long-term survivorship, with direct implications for imaging surveillance and follow-up care. Despite excellent short term survival for germinomas, long term survivors experience substantially increased risks of late mortality, including recurrent disease, secondary malignancies, and cerebrovascular events, underscoring the importance of consistent access to longitudinal imaging and specialized survivorship monitoring across all populations (Acharya et al., 2014). Treatment paradigms increasingly rely on response-adapted strategies that integrate chemotherapy with reduced field or reduced dose radiation, placing added importance on high-quality imaging to guide risk stratification and minimize late toxicities; however, access to these approaches is uneven globally and within health systems (Hill Kayser et al., 2020; Cappellano et al., 2024). Timely diagnostic and staging imaging, equitable access to response-based surveillance strategies, and coordinated long term follow-up are necessary to reduce preventable disparities in survival and late effects among diverse pediatric and adolescent populations (Acharya et al., 2014; Lin et al., 2025).

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Ependymal Tumors (Ependymoma) (PEDONC-4.8)

ONCP.CT.0004.8.A
v1.0.2027

Criteria

- PET Brain Metabolic imaging does not have a defined role in the evaluation of ependymal tumors and therefore is considered not medically necessary.
- MR Spectroscopy is not considered medically necessary for routine surveillance of ependymal tumors.
- Individuals with ependymal tumors with a complete resection should be imaged according to surveillance guidelines.
- Individuals with ependymal tumors with incomplete resection or high-risk histology that receive adjuvant radiation therapy should be imaged according to surveillance guidelines after end of radiotherapy imaging.
- RELA-fusion supratentorial ependymoma did not portend worse overall survival outcomes in recent Clinical Oncology Group studies, and is not considered an indication for more frequent surveillance imaging.
- Includes the following tumors:
  - Ependymal tumors
    - Supratentorial ependymoma
    - ZFTA fusion-positive Supratentorial ependymoma
    - YAP1 fusion-positive Posterior fossa ependymoma
    - Posterior fossa ependymoma, group
    - PFA Posterior fossa ependymoma, group PFB

Indication	Medically Necessary Imaging Study
Initial staging for all individuals with ependymal tumors	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
In addition to the above imaging, for preoperative planning in individuals with ependymal tumors	• Functional MRI (fMRI) (CPT® 70555 or 70554) • For further perioperative indications, please refer to **Neuronavigation (HD-28.2)**

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Indication	Medically Necessary Imaging Study
Evaluation of a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed in individuals with ependymal tumors	• MR Spectroscopy (CPT® 76390)
Baseline imaging following resection in individuals with ependymal tumors	• MRI Brain without and with contrast (CPT® 70553) or MRI without and with contrast of involved spinal level(s)
Completion of radiotherapy in individuals with ependymal tumors	• MRI Brain without and with contrast (CPT® 70553) or MRI without and with contrast of involved spinal level(s)
Prior to radiotherapy in individuals with ependymal tumors	• MRI of all known sites of measurable disease
Treatment response to induction chemotherapy in individuals with ependymal tumors	• MRI Brain without and with contrast (CPT® 70553) or MRI without and with contrast of involved spinal level(s) every 2 cycles
End of induction chemotherapy and again at end of all therapy in individuals with ependymal tumors	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Prior to second-look surgery in individuals with ependymal tumors	• MRI of all known sites of measurable disease
Signs or symptoms of recurrence, progression, or neurologic emergency in individuals with ependymal tumors	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
Surveillance, primary intra**cranial** ependymal tumor and NO history of spinal cord involvement	• MRI Brain without and with contrast (CPT® 70553) every 3 months for 2 years, then every 4 months for 1 year, then every 6 months in years 4 and 5, then annually to 10 years post treatment ◦ For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see: **Second Malignant Neoplasms (SMN) (PEDONC-19.3)** • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) can be approved annually for 2 years
Surveillance, primary intra**cranial** ependymal tumor AND metastatic cord involvement at diagnosis	• MRI Brain without and with contrast (CPT® 70553) every 3 months for 2 years, then every 4 months for 1 year, then every 6 months in years 4 and 5, then annually to 10 years post treatment ◦ For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see: **Second Malignant Neoplasms (SMN) (PEDONC-19.3)** • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 3 months for 2 years, then every 4 months for 1 year, then every 6 months in years 4 and 5, then annually to 10 years post treatment
Surveillance, primary intra**spinal** ependymal tumor and NO history of intracranial involvement	• MRI without and with contrast of the involved spinal level(s) every 3 months for 2 years, then every 4 months for 1 year, then every 6 months in years 4 and 5, then annually to 10 years post treatment

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Indication	Medically Necessary Imaging Study
Surveillance, primary intra**spinal** ependymal tumor AND metastatic intracranial involvement at diagnosis	• MRI of the involved spinal level(s) without and with contrast every 3 months for 2 years, then every 4 months for 1 year, then every 6 months in years 4 and 5, then annually to 10 years post treatment • MRI Brain without and with contrast (CPT® 70553) every 3 months for 2 years, then every 4 months for 1 year, then every 6 months in years 4 and 5, then annually to 10 years post treatment

Background and Supporting Information

- Ependymal tumors arise from ependymal cells lining the ventricular system and spinal canal and include multiple molecularly defined entities, with approximately two-thirds occurring intracranially in the posterior fossa (NCCN, 2025).
- Prognosis is generally favorable, particularly for supratentorial tumors and those undergoing gross total resection, though molecular features such as chromosome 1q gain are associated with worse outcomes (NCCN, 2025).
- Primary spinal ependymomas occur less frequently in children than adults but follow similar treatment principles (PDQ® Pediatric Treatment Editorial Board, 2025).
- Surgical resection is the primary treatment, with adjuvant radiation therapy used for incompletely resected or high risk tumors (NCCN, 2025).

Evidence Discussion

- Postoperative MRI is critical for determining extent of resection, which is the most important prognostic factor in pediatric ependymoma (NCCN, 2025).
- Surveillance imaging following surgery or radiotherapy facilitates early identification of recurrence, which may be amenable to re-resection or salvage therapy (PDQ® Pediatric Treatment Editorial Board, 2025).
- Restricting imaging to guideline-supported intervals balances the benefit of early detection against the risks of repeated sedation and imaging in long-term survivors (Expert Panel on Neurological Imaging, 2019).

Health Equity Considerations

Ependymomas are rare pediatric central nervous system tumors with heterogeneous clinical behavior and long survivorship, yet substantial disparities exist in access to care, treatment delivery, and long-term outcomes across socioeconomic and demographic

groups. Population-based analyses demonstrate that uninsured and socioeconomically disadvantaged children experience worse perioperative outcomes, including significantly prolonged hospital length of stay, which can reflect delayed presentation, barriers to timely postoperative care, and limited access to supportive services (Jagtiani et al., 2024). Racial and ethnic disparities in care have also been identified, with Black children exhibiting lower odds of undergoing tumor resection and Hispanic patients experiencing longer postoperative hospitalizations, suggesting the influence of structural inequities, differences in access to high volume surgical centers, and downstream barriers to specialty referral (Jagtiani et al., 2024). These disparities are particularly consequential in ependymoma, where extent of safe surgical resection and timely radiotherapy are critical determinants of survival.

Equity considerations extend well beyond initial treatment, as survivors of childhood ependymoma face a substantial and lifelong burden of medical and functional morbidity with important implications for imaging surveillance and long-term follow up. Population-based survivorship studies demonstrate markedly increased risks of stroke, severe hearing loss, subsequent malignant neoplasms, disability, mental health service utilization, and premature mortality among ependymoma survivors compared with the general population, even in cohorts with limited exposure to chemotherapy and predominantly focal radiation (Coltin et al., 2024). These long-term risks underscore the need for consistent access to longitudinal neuroimaging, multidisciplinary survivorship care, and supportive services - resources that may be unevenly available across healthcare systems and communities. Timely diagnostic imaging, referral to experienced neurosurgical and radiation centers, and standardized long-term surveillance strategies are necessary to reduce preventable disparities in treatment outcomes and survivorship burden across diverse pediatric populations (Coltin et al., 2024; Jagtiani et al., 2024).

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Malignant Tumors of the Spinal Cord (PEDONC-4.9)

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Criteria

- If a disease-specific guideline exists, image according to the guidance found in that disease-specific guideline section.
- Treatment principles for malignant tumors of the spinal cord are the same as tumors of the brain, and should follow imaging guidelines according to the specific histologic type.
- Asymptomatic surveillance imaging for tumors of the spinal cord should generally end at the time point appropriate for the specific tumor type.
- Common histologies of primary spinal cord tumor in children include:
  - Low-Grade Glioma, see: **CNS Low Grade Glioma (LGG) (PEDONC-4.2)** for guidelines
  - High-Grade Glioma, see: **CNS High-Grade Glioma (HGG) (PEDONC-4.3)** for guidelines
  - Ependymoma, see: **Ependymal Tumors (Ependymoma) (PEDONC-4.8)** for guidelines
  - NF 1 or 2, see **Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)** for guidelines
  - Any type of malignant spinal cord tumor can occur, but other histologies are rare.

For rare histologies that do not have a disease-specific guideline section, follow the imaging outlined in the table below.

Indications	Medically Necessary Imaging Study
Initial staging for all individuals with tumors of the spinal cord	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
In addition to the above imaging, for preoperative planning in individuals with tumors of the spinal cord	• Functional MRI (fMRI) (CPT® 70555 or 70554) • For further perioperative indications, please refer to **Neuronavigation (HD-28.2)**

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Indications	Medically Necessary Imaging Study
Treatment response in individuals with tumors of the spinal cord, every 2-3 cycles	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • Additionally, MRI Brain without and with contrast (CPT® 70553) for known intracranial disease
Signs or symptoms of recurrence or progression in individuals with tumors of the spinal cord	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • Additionally, MRI Brain without and with contrast (CPT® 70553) for known intracranial disease
Surveillance of rare histologies is highly individualized. Surveillance imaging after recent evaluation by a physician with significant training and/or experience in pediatric spinal cord tumors (most commonly a pediatric neurosurgeon or pediatric oncologist) may be considered medically necessary in these rare individuals	• MRI Brain and/or MRI Spine may be considered

Background and Supporting Information

- Malignant tumors of the spinal cord in children are rare and include astrocytomas, ependymomas, and other histologies that share management principles with their intracranial counterparts (PDQ® Pediatric Treatment Editorial Board, 2025).
- Multiple spinal cord tumors raise suspicion for underlying genetic syndromes such as neurofibromatosis (NCCN, 2025).
- Treatment and imaging strategies are dictated by histology, extent of disease, and neurological status (NCCN, 2025).

Evidence Discussion

- MRI of the spine is the preferred imaging modality for diagnosis, staging, and follow-up due to superior soft tissue contrast and lack of ionizing radiation (NCCN, 2025).
- Surveillance imaging is supported during periods of highest recurrence risk and should generally conclude according to timeframes established for the specific tumor type once stability is achieved (Expert Panel on Neurological Imaging, 2019).

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Health Equity Considerations

Malignant tumors of the spinal cord represent a rare and clinically complex subset of pediatric CNS tumors, and children with these diagnoses face unique equity related challenges across diagnosis, treatment, and research participation. Despite accounting for approximately 10% of pediatric CNS tumors, spinal cord tumors remain markedly underrepresented in clinical trials, with only a small fraction of CNS trials explicitly including spinal tumors and virtually none dedicated specifically to primary pediatric spinal cord malignancies (Villanueva et al., 2024). This underrepresentation limits the development of evidence-based treatment pathways and contributes to variability in clinical management, as pediatric patients are often treated using protocols extrapolated from adult spinal or intracranial tumor studies (Villanueva et al., 2024). Geographic concentration of trials in high income regions, particularly North America, further restricts equitable access to investigational therapies for children treated outside major academic centers or in resource limited settings (Villanueva et al., 2024).

Health equity considerations for malignant spinal cord tumors also extend to diagnostic imaging, surgical decision making, and long-term functional outcomes. Maximal safe surgical resection is the primary treatment modality for many pediatric spinal cord malignancies, yet tumor infiltration of critical neural tracts frequently limits resectability and increases the risk of permanent neurologic morbidity, underscoring the need for high-quality imaging and specialized surgical expertise that may not be uniformly accessible across health systems (Villanueva et al., 2024). The absence of standardized, tumor-specific surveillance and imaging guidelines, coupled with limited data on the efficacy of adjuvant radiation or systemic therapy, disproportionately affects patients treated outside specialized centers with multidisciplinary pediatric neuro-oncology expertise (Villanueva et al., 2024). Timely access to high resolution diagnostic and surveillance imaging, referral to experienced pediatric spine oncology centers, and inclusion of spinal cord tumor-specific cohorts within CNS clinical trials are necessary to reduce preventable disparities in outcomes for children with malignant spinal cord tumors (Villanueva et al., 2024).

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Craniopharyngioma and Other Tumors of the Sellar Region (PEDONC-4.10)

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v1.0.2027

Criteria

- Individuals of all ages should be imaged according to these guidelines.
- PET Brain Metabolic Imaging and MR Spectroscopy do not have a defined role in the evaluation of craniopharyngioma and are, therefore, considered not medically necessary.
- Includes the following tumors:
  - Craniopharyngioma
  - Papillary craniopharyngioma
  - Pituicytoma, granular cell tumor of the sellar region, and spindle cell oncocytoma
  - PitNET
  - Pituitary blastoma
- Other less common tumors in the optic chiasm, sella, and suprasella region may include Germ Cell Tumors (GCT, see: PEDONC-4.7) and Langerhans Cell Histiocytosis (LCH, see: PEDONC-18).
- Imaging guidelines and treatment approaches for pediatric pituitary tumors other than those listed below are consistent with those used for adults with pituitary tumors.
  - For these tumors, follow guidelines in Pituitary, Sella, Hypothalamus (HD-19) in the Head Imaging Guidelines.
- Several key imaging findings can be used to differentiate the tumors in this region, including the presence of calcifications, cysts, and T1/T2 enhancement patterns in craniopharyngiomas.
  - These are best evaluated using a COMBINATION of both MRI and CT modalities. Preoperative prediction is much more successful when BOTH modalities are obtained prior to biopsy.
  - Those with craniopharyngioma with a complete resection should be imaged according to surveillance guidelines after post-resection imaging is completed.
- Individuals with craniopharyngioma with incomplete resection and receiving adjuvant radiation therapy can have a single MRI Brain (CPT® 70553) approved at completion of radiotherapy and should then be imaged according to surveillance guidelines.

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Indications	Medically Necessary Imaging Study
Initial staging for all individuals with craniopharyngioma and other tumors of the sellar region	• MRI Brain without and with contrast (CPT® 70553) ◦ Concurrent CT Head without contrast (CPT® 70450) in addition to MRI if craniopharyngioma is suspected
In addition to the above imaging, for preoperative planning in individuals with craniopharyngioma and other tumors of the sellar region	• Functional MRI (fMRI) (CPT® 70555 or 70554) • For further perioperative indications, please refer to **Neuronavigation (HD-28.2)**
• Additional initial staging for individuals with craniopharyngioma and other tumors of the sellar region with: ◦ Multicentric tumors ◦ Clinical signs or symptoms suggesting spinal cord involvement	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Operative planning or image guidance in individuals with craniopharyngioma and other tumors of the sellar region	• MRA Head (CPT® 70544, 70545, or 70546) OR • CTA Head (CPT® 70496)
Baseline imaging following resection in individuals with craniopharyngioma and other tumors of the sellar region	• MRI Brain without and with contrast (CPT® 70553)
Completion of radiotherapy in individuals with craniopharyngioma and other tumors of the sellar region	• MRI Brain without and with contrast (CPT® 70553)
Treatment response to chemotherapy in individuals with craniopharyngioma and other tumors of the sellar region	• MRI Brain without and with contrast (CPT® 70553) approved every 2 cycles during active treatment and at the end of planned chemotherapy
Additional treatment response imaging during induction chemotherapy for individuals with craniopharyngioma and other tumors of the sellar region measurable spinal cord disease on MRI	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 2 cycles

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Indications	Medically Necessary Imaging Study
Signs or symptoms of recurrence or progression in individuals with craniopharyngioma and other tumors of the sellar region	• MRI Brain without and with contrast (CPT® 70553) ◦ Concurrent CT Head without contrast (CPT® 70450) in addition to MRI for craniopharyngioma • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) for signs or symptoms suggesting spinal cord involvement
Surveillance in individuals with craniopharyngioma and other tumors of the sellar region	• MRI Brain without and with contrast (CPT® 70553) can be approved every 3 months for 1 year, then every 4 months for 1 year, then every 6 months for 1 year, then annually until 10 years after completion of therapy as late progressions can occur ◦ For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see .
Suspected spinal cord recurrence in individuals with craniopharyngioma and other tumors of the sellar region	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

Background and Supporting Information

- Craniopharyngiomas account for approximately 6%–8% of pediatric CNS tumors and most commonly present in pre-adolescent children (PDQ® Pediatric Treatment Editorial Board, 2025).
- These tumors arise in the sellar and suprasellar region and are frequently cystic and calcified, often causing endocrine dysfunction and visual impairment (NCCN, 2025).
- MRI provides optimal soft tissue characterization, while CT is particularly useful for identifying calcifications, supporting the use of complementary modalities in preoperative assessment (Expert Panel on Neurological Imaging, 2019).
- Other rare sellar region tumors include pituitary blastoma, pituicytoma, and pituitary neuroendocrine tumors (PitNETs) (PDQ® Pediatric Treatment Editorial Board, 2025).

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Evidence Discussion

- Combined MRI and CT improves diagnostic accuracy and surgical planning compared with either modality alone (Expert Panel on Neurological Imaging, 2019).
- Surveillance imaging following resection or radiotherapy detects cystic recurrence, which is common and may be asymptomatic initially (NCCN, 2025).
- Guideline-directed imaging minimizes unnecessary repeat studies while preserving the ability to detect clinically relevant disease progression (PDQ® Pediatric Treatment Editorial Board, 2025).

Health Equity Considerations

Craniopharyngioma and other tumors of the sellar region require highly specialized, multidisciplinary care, and substantial health equity challenges exist across diagnosis, treatment, imaging surveillance, and long-term follow-up. Delayed diagnosis is common due to the insidious onset of symptoms such as growth failure, visual disturbance, or endocrine dysfunction, with prolonged symptom duration disproportionately reported in low and middle income countries (LMICs) and underserved populations, contributing to worse functional outcomes at presentation (Baqai et al., 2024; Memon et al., 2023; Tenorio et al., 2025). Access to advanced neuroimaging, experienced neurosurgical teams, and hypothalamus sparing surgical approaches varies widely, often resulting in greater hypothalamic and pituitary injury in settings with limited subspecialty resources (Beckhaus et al., 2023; Şıklar et al., 2024). International and cross-border data further demonstrate that disparities in postoperative endocrine care and treatment adherence, rather than tumor recurrence alone, are strongly associated with reduced overall survival, underscoring the critical role of longitudinal care access beyond initial surgical management (Tenorio et al., 2025).

Long-term survivorship following sellar region tumors is characterized by high rates of permanent endocrine deficiencies, hypothalamic obesity, visual impairment, and neuropsychological morbidity, all of which necessitate ongoing imaging, endocrine surveillance, and coordinated multidisciplinary follow-up that may not be equitably available (Beckhaus et al., 2023; Şıklar et al., 2024). Studies from LMIC settings highlight significant loss to follow-up, limited access to routine MRI surveillance, and socioeconomic barriers that impede timely monitoring for recurrence and management of late effects, thereby amplifying disparities in quality of life and functional outcomes (Baqai et al., 2024; Memon et al., 2023). Caregiver burden and long-term dependency, particularly in patients with hypothalamic damage, disproportionately affect families with fewer financial and social supports, further reinforcing inequities in survivorship outcomes (Beckhaus et al., 2023). Timely diagnostic imaging, standardized surveillance strategies, and referral to experienced multidisciplinary centers, with particular attention to improving access to long-term endocrine and neuroimaging follow-up for individuals treated in resource-limited and marginalized

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settings are needed to reduce disparities in this population (Şıklar et al., 2024; Tenorio et al., 2025).

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Primary CNS Lymphoma (PEDONC-4.11)

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Criteria

- Primary CNS lymphoma imaging indications in pediatric individuals are identical to those in the general imaging guidelines. See: **CNS Lymphoma (ONC-2.7)** in the Oncology Imaging Guidelines.
  - Usually associated with immunodeficiency, this is a very rare entity in pediatrics accounting for <0.1% of pediatric malignancies, so age-specific guidelines have not been established.
- CNS lymphomas also involving bone marrow and/or lymph nodes should be imaged according to: **Pediatric Aggressive Mature B-Cell Non-Hodgkin Lymphomas (NHL) (PEDONC-5.3)**.

Background and Supporting Information

- Primary CNS lymphoma is defined as lymphoma confined to the brain, leptomeninges, spinal cord, or eyes without systemic involvement at diagnosis (NCCN, 2025).
- This entity is rare in children and is most often of diffuse large B-cell histology (PDQ® Pediatric Treatment Editorial Board, 2025).
- Lesions may be solitary or multifocal and frequently involve deep brain structures (NCCN, 2025).

Evidence Discussion

- MRI with contrast is essential for diagnosis and response assessment, as lesions demonstrate characteristic enhancement patterns (NCCN, 2025).
- Surveillance imaging supports early detection of relapse, which is common despite initial response to therapy (PDQ® Pediatric Treatment Editorial Board, 2025).
- Avoidance of unnecessary imaging outside recommended intervals reduces exposure to sedation and contrast related risks (Expert Panel on Neurological Imaging, 2019).

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Meningiomas (PEDONC-4.12)

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Criteria

- Meningioma imaging indications in pediatric individuals are identical to those in the general imaging guidelines
  - See: **Meningiomas (Intracranial and Intraspinal) (ONC-2.8)** in the Oncology Imaging Guidelines

Background and Supporting Information

- Pediatric meningiomas account for approximately 1%–3% of pediatric CNS tumors and are more frequently associated with NF2 or prior cranial radiotherapy (PDQ® Pediatric Treatment Editorial Board, 2025).
- Radiation-induced meningiomas may occur decades after exposure, underscoring the importance of long-term follow-up in survivors (NCCN, 2025).
- Most pediatric meningiomas are slow growing and amenable to surgical cure (PDQ® Pediatric Treatment Editorial Board, 2025).

Evidence Discussion

- MRI is the preferred modality for diagnosis and surveillance, enabling assessment of tumor growth and involvement of adjacent structures (NCCN, 2025).
- Surveillance imaging is generally prolonged in individuals with incomplete resection or genetic predisposition due to higher recurrence risk (PDQ® Pediatric Treatment Editorial Board, 2025).
- Imaging strategies guided by clinical risk factors optimize early detection while limiting unnecessary studies (Expert Panel on Neurological Imaging, 2019).

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Choroid Plexus Tumors (PEDONC-4.13)

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Criteria

- PET Metabolic Brain imaging does not have a defined role in the evaluation of choroid plexus tumors and is, therefore, considered not medically necessary.
- Spinal imaging may be approved for Choroid Plexus Adenoma or Atypical Choroid Plexus Papilloma if requested at initial diagnosis.
- For choiroid plexus carcinoma:
  - Individuals with confirmed gross total resection should be imaged according to surveillance guidelines.
  - Individuals with incomplete resection who receive adjuvant radiation therapy should be imaged according to surveillance guidelines after end of radiotherapy imaging.
  - MR Spectroscopy is not considered medically necessary for routine surveillance.

Choroid Plexus Papilloma:

Indication	Medically Necessary Imaging Study
Suspected/Diagnosis of choroid plexus papilloma	• MRI Brain without and with contrast (CPT® 70553)
In addition to the above imaging, for preoperative planning in individuals with choroid plexus papilloma	• Functional MRI (fMRI) (CPT® 70555 or 70554) • For further perioperative indications, please refer to **Neuronavigation (HD-28.2)**
Evaluation of a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed in individuals with choroid plexus papilloma	• MR Spectroscopy (CPT® 76390)
Suspected return of hydrocephalus, or return of hydrocephalus seen on CT imaging in individuals with choroid plexus papilloma	• MRI Brain without and with contrast (CPT® 70553)

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Choroid Plexus Adenoma or Atypical Choroid Plexus Papilloma:

Indication	Medically Necessary Imaging Study
Suspected/Diagnosis of choroid plexus adenoma or atypical choroid plexus papilloma	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
In addition to the above imaging, for preoperative planning in individuals with choroid plexus adenoma or atypical choroid plexus papilloma	• Functional MRI (fMRI) (CPT® 70555 or 70554) • For further perioperative indications, please refer to **Neuronavigation (HD-28.2)**
Evaluation of a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed in individuals with choroid plexus adenoma or atypical choroid plexus papilloma	• MR Spectroscopy (CPT® 76390)
Suspected return of hydrocephalus, or return of hydrocephalus seen on CT imaging in individuals with choroid plexus adenoma or atypical choroid plexus papilloma	• MRI Brain without and with contrast (CPT® 70553)

Choroid Plexus Carcinoma

Indication	Medically Necessary Imaging Study
Initial staging of all individuals with choroid plexus carcinoma	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
In addition to the above imaging, for preoperative planning in individuals with choroid plexus carcinoma	• Functional MRI (fMRI) (CPT® 70555 or 70554) • For further perioperative indications, please refer to

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Indication	Medically Necessary Imaging Study
Evaluation of a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed in individuals with choroid plexus carcinoma	• MR Spectroscopy (CPT® 76390)
Baseline imaging following resection in individuals with choroid plexus carcinoma	• MRI Brain without and with contrast (CPT® 70553)
Prior to radiotherapy in individuals with choroid plexus carcinoma	• MRI Brain without and with contrast (CPT® 70553) and MRI with and without contrast of all known sites with measurable disease prior to radiotherapy
Completion of radiotherapy in individuals with choroid plexus carcinoma	• MRI Brain without and with contrast (CPT® 70553)
Treatment response to chemotherapy in individuals with choroid plexus carcinoma	• MRI Brain without and with contrast (CPT® 70553) every 2 cycles during active treatment
Additional treatment response to chemotherapy for individuals with choroid plexus carcinoma with measurable spinal cord disease on MRI	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 2 cycles during active treatment
Prior to second-look surgery in individuals with choroid plexus carcinoma	• MRI of all known sites of measurable disease
End of all planned therapy in individuals with choroid plexus carcinoma	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Signs or symptoms of recurrence or progression in individuals with choroid plexus carcinoma	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

Indication	Medically Necessary Imaging Study
Surveillance, no history of spinal cord involvement in individuals with choroid plexus carcinoma	• MRI Brain without and with contrast (CPT® 70553) every 4 months for 3 years, then every 6 months for 2 years after completion of therapy, then annually to 10 years after treatment ◦ For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see **Second Malignant Neoplasms (SMN) (PEDONC-19.3)**. • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) at 12 and 24 months after completion of therapy
Surveillance, individuals with choroid plexus carcinoma with cord involvement at diagnosis	• MRI Brain without and with contrast (CPT® 70553) every 4 months for 3 years, then every 6 months for 2 years after completion of therapy, then annually to 10 years after treatment ◦ For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see **Second Malignant Neoplasms (SMN) (PEDONC-19.3).** • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 4 months for 3 years, then every 6 months for 2 years after completion of therapy, then annually to 10 years after treatment

Background and Supporting Information

- Choroid plexus tumors, including papilloma, atypical papilloma, adenoma, and carcinoma, account for approximately 1%–4% of pediatric CNS tumors and most commonly occur in infants under 2 years of age (PDQ® Pediatric Treatment Editorial Board, 2025).

- Papillomas and atypical papillomas are typically benign or intermediate grade lesions with excellent prognosis following surgical resection (NCCN, 2025).
- Choroid plexus carcinomas are highly aggressive, with frequent metastasis and poorer survival outcomes, particularly in association with TP53 mutations (PDQ® Pediatric Treatment Editorial Board, 2025).

Evidence Discussion

- MRI provides characteristic imaging appearances that often allow confident diagnosis and surgical planning (NCCN, 2025).
- Spinal imaging is necessary at diagnosis for carcinoma due to high metastatic risk but is not routinely necessary for benign subtypes (PDQ® Pediatric Treatment Editorial Board, 2025).
- Surveillance imaging tailored to tumor histology and extent of resection balances recurrence detection against the risks of repeated imaging and anesthesia (Expert Panel on Neurological Imaging, 2019).

Health Equity Considerations

Choroid plexus tumors, including choroid plexus papilloma and carcinoma, are rare pediatric central nervous system tumors that disproportionately affect very young children, with the majority of malignant cases diagnosed before five years of age (Dudley et al., 2015). This early age of onset poses significant equity challenges, as infants and toddlers depend on timely access to advanced neuroimaging, neurosurgical expertise, and specialized perioperative care, which may not be uniformly available across healthcare systems. Population-based data demonstrate high overall surgical intervention rates for both benign and malignant choroid plexus tumors, yet survival outcomes for choroid plexus carcinoma remain poor despite aggressive treatment, underscoring the importance of early diagnosis and access to high volume pediatric neurosurgical centers with experience managing highly vascular intraventricular tumors (Dudley et al., 2015). Limitations in imaging availability, delayed diagnosis of hydrocephalus, and variability in referral pathways may disproportionately affect children from underserved or rural communities, potentially contributing to worse neurologic morbidity and survival outcomes.

Health equity considerations for choroid plexus tumors also extend to the availability of adjuvant therapies, longitudinal imaging surveillance, and supportive care resources. National registry data indicate variation in the use of radiation therapy by age, with substantially lower utilization in children younger than five years due to concerns regarding neurodevelopmental toxicity, highlighting the need for individualized imaging and treatment strategies tailored to developmental stage and resource availability (Dudley et al., 2015). Importantly, large administrative datasets lack information on socioeconomic status, chemotherapy use, hydrocephalus management, and long-term

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functional outcomes, limiting the ability to fully characterize disparities in survivorship and quality of life (Dudley et al., 2015). Timely diagnostic neuroimaging for infants presenting with signs of hydrocephalus, coordinated referral to specialized pediatric centers, and consistent post-treatment surveillance imaging are necessary to monitor for recurrence and treatment related complications, particularly for individuals treated outside major tertiary care settings.

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Pediatric Lymphomas (PEDONC-5)

Guideline Page

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Pediatric Lymphoma – General Considerations (PEDONC-5.1)

ONCP.HL.0005.1.A
v1.0.2027

Criteria

- Lymphoma most commonly involves the lymph nodes (LNs). However, lymphoma can also arise from primary lymphoid tissues (bone marrow or thymus) or various secondary lymphoid tissues (spleen, mucosa-associated lymphoid tissue) or non-lymphoid organs (skin, bone, brain, lungs, liver, salivary glands, etc.).
- Pediatric lymphomas are generally Hodgkin Lymphomas, Aggressive B-Cell Non-Hodgkin Lymphomas, Lymphoblastic Lymphomas, or Anaplastic Large Cell Lymphomas
- Individuals with Lymphoblastic Lymphoma (even those with bulky nodal disease) are treated using the leukemia treatment plan appropriate to the cell type (B or T cell).
  - These individuals should be imaged using guidelines in **Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)**
- Other histologies are rare in pediatric individuals, and should be imaged according to the following guidelines:
  - Follicular lymphoma: **Follicular Lymphoma (ONC-27.3)** in the Oncology Imaging Guidelines
  - Marginal zone or MALT lymphomas: **Marginal Zone Lymphomas (ONC-27.4)** in the Oncology Imaging Guidelines
  - Mantle cell lymphomas: **Mantle Cell Lymphoma (ONC-27.5)** in the Oncology Imaging Guidelines
  - Cutaneous lymphomas: **Cutaneous Lymphomas and T Cell Lymphomas (ONC-27.8)** in the Oncology Imaging Guidelines
    - **Exception:** Cutaneous B-Lymphoblastic Lymphoma should be imaged using guidelines in **Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)**
  - Castleman's Disease: **Castleman's Disease (Unicentric and Multicentric) (ONC-31.11)** in the Oncology Imaging Guidelines
- All CT imaging recommended in this section refers to CT with contrast only.
  - Non-contrast CT imaging has not been shown to be beneficial in the management of pediatric lymphomas
  - Given the limited utility of non-contrast CT imaging in pediatric lymphomas, MRI without **or** without and with contrast is considered medically necessary in place of CT for individuals who cannot tolerate CT contrast due to allergy or impaired renal function.

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- MRI without and with contrast of symptomatic or previously involved bony areas is considered medically necessary in individuals with known lymphoma without prior plain x-ray or bone scan evaluation.
  - Bone scan is inferior to MRI for evaluation of known or suspected bone metastases in lymphoma.
- MRI Brain without and with contrast (CPT® 70553) is considered medically necessary as it is the preferred study for evaluation of suspected brain metastases in pediatric lymphoma.
  - CT Head with (CPT® 70460) or without and with contrast (CPT® 70470) is considered medically necessary when MRI is contraindicated.

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Pediatric Hodgkin Lymphoma (HL) (PEDONC-5.2)

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v1.0.2027

Criteria

- Indications and coding for rare circumstances where PET/MRI may be approved are found in **PET imaging in Pediatric Oncology (PEDONC-1.4)**

Indication	Medically Necessary Imaging Study
Initial staging of HL	ANY or ALL of the following: • CT Neck with contrast (CPT® 70491) ◦ MRI Neck without and with contrast (CPT® 70543) as a substitute for CT to limit radiation exposure if requested • CT Chest with contrast (CPT® 71260) ◦ MRI is not a substitution for CT Chest • CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast as substitute for CT to limit radiation exposure if requested • MRI Brain without and with contrast (CPT® 70553) for known CNS involvement or new signs or symptoms suggesting intracranial disease • PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) ◦ Can be performed prior to biopsy if necessary for individual scheduling • CT or MRI of other body areas may be indicated for rare individuals based on physical findings or PET/CT results

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Indication	Medically Necessary Imaging Study
Restaging of HL	• ALL of the following, as often as every 2 cycles of chemotherapy: ◦ CT of previously involved visceral areas ▪ In individuals on treatment for recurrent or refractory Hodgkin Lymphoma, all of the following: - CT Neck with contrast (CPT® 70491) - CT Chest with contrast (CPT® 71260) - CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Brain without and with contrast (CPT® 70553) for known CNS involvement AND • After cycles 2 and 4* of chemotherapy and at end of therapy: ◦ PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (only using combination of CPT® 78813 and 76498) ▪ *For individuals with low-risk stage IA or IIA mixed cellularity Hodgkin lymphoma, this restaging PET/CT is considered medically necessary after cycles 1 and 3 (instead of cycles 2 and 4) if requested. End of therapy PET/CT or PET/MRI is considered medically necessary for all individuals with Hodgkin lymphoma. ◦ Diagnostic CT or MRI of previously involved areas may be combined with PET at end of therapy. ◦ For individuals with Waldeyer's Ring involvement, CT Neck with contrast (CPT® 70491) or MRI Orbit/Face/Neck without and with contrast (CPT® 70543) may be done in addition to the above studies after cycles 2 and 4 and at end of therapy • In addition to the above studies: ◦ If end of therapy PET/CT or PET/MRI done prior to radiation therapy documents Deauville 4 or 5 FDG avidity, one follow-up PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI is considered medically necessary >12 weeks after radiation therapy to confirm complete response.

Indication	Medically Necessary Imaging Study
Surveillance for individuals with no history of recurrent HL	• Imaging studies are only considered medically necessary when relapse is clinically suspected, because most individuals will clinically declare themselves and there is no survival advantage in pre-emptive imaging. • **Routine surveillance imaging is not considered medically necessary in individuals without a prior history of recurrent disease**
Surveillance, individuals with **recurrent** HL and no evidence of disease following successful treatment	• ALL of the following, every 3 months for 2 years after completing therapy for recurrence: ◦ CT Neck with contrast (CPT® 70491) ◦ CT Chest with contrast (CPT® 71260) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177)
Clarify inconclusive findings on conventional imaging suspicious for recurrence AND considering biopsy to establish recurrence in individuals with HL	• PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498)

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Indication	Medically Necessary Imaging Study
Clinical symptoms suggesting recurrence of HL	• MRI Brain without and with contrast (CPT® 70553) for known CNS involvement or new signs or symptoms suggesting intracranial disease AND either of the following sets of imaging: • Conventional imaging: ◦ CT Neck with contrast (CPT® 70491) OR MRI Neck without and with (CPT® 70491) AND ◦ CT Chest with contrast (CPT® 71260) AND ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) OR MRI Abdomen and MRI Pelvis without and with contrast (CPT® 74183 and CPT® 72197) AND ◦ CT of other previously involved visceral areas or currently symptomatic areas OR • PET/CT (CPT® 78815 or 78816) or PET/MRI (CPT® 78813 and CPT® 76498)

Background and Supporting Information

- Pediatric HL is a malignant lymphoma arising from the lymphatic system and is classified into classic HL and nodular lymphocyte predominant HL according to contemporary classification systems, with management guided by pediatric cooperative group protocols (NCCN, 2025).
- HL in pediatric individuals is primarily a nodal disease, with frequent involvement of cervical, supraclavicular, and mediastinal lymph nodes; extranodal involvement is less common and typically associated with more advanced disease (Mhlanga et al., 2023).
- Staging is performed using the Ann Arbor staging system with Cotswolds modifications, incorporating disease distribution, bulky disease, and clinical symptoms, which collectively inform risk stratification and treatment planning (Mhlanga et al., 2023; NCCN, 2025).
- Contemporary pediatric HL treatment relies on response-adapted therapy, where early treatment response assessed by both anatomic and metabolic imaging is used to modify chemotherapy intensity and determine the need for radiotherapy (NCCN, 2025).

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Evidence Discussion

- Evidence from large pediatric cohorts demonstrates that most HL relapses are identified by clinical findings, including symptoms, laboratory abnormalities, or physical examination, rather than by routine surveillance imaging (Voss et al., 2012).
- Surveillance CT imaging after completion of therapy has not been shown to improve post recurrence overall survival, as the time to relapse, rather than the method of detection, is the primary determinant of outcome (Voss et al., 2012).
- NCCN pediatric HL guidelines recommend against routine pre-emptive surveillance imaging, supporting imaging only when relapse is clinically suspected (NCCN, 2025).
- FDG-PET combined with CT plays an essential role during initial staging and early response assessment, as treatment decisions frequently depend on both metabolic response and anatomic tumor regression (Mhlanga et al., 2023; NCCN, 2025).

Health Equity Considerations

Pediatric HL is highly curable, yet marked inequities persist in stage at diagnosis, treatment delivery, and outcomes across racial, ethnic, and socioeconomic groups. Multiple population-based and cooperative group studies demonstrate that Black and Hispanic children and adolescents are more likely to present with advanced stage HL, which is associated with greater treatment intensity, increased imaging burden for staging and response assessment, and worse survival (Kahn & Beauchemin, 2021; Kahn et al., 2022; Ou et al., 2024). These disparities are driven by multilevel social determinants of health, including insurance instability, structural racism, language barriers, and limited access to timely primary and specialty care, all of which may delay diagnostic evaluation and imaging (Ou et al., 2024). Community-level factors, such as residence in historically redlined counties, racial residential segregation, and low Child Opportunity Index neighborhoods, have also been associated with later stage at diagnosis for pediatric HL, emphasizing that structural and environmental disadvantages influence when children enter the diagnostic imaging and treatment pathway (Ou et al., 2024). Although national incidence trends for pediatric HL have remained relatively stable, racial and ethnic differences in incidence and age-specific patterns persist, reinforcing the need for equity-focused diagnostic and staging strategies (Cheng et al., 2025; Mousavi et al., 2025).

Disparities continue beyond diagnosis and affect treatment response assessment, surveillance imaging, and long-term follow-up. Children and adolescents who are publicly insured, from low income households, or treated outside National Cancer Institute or Children's Oncology Group affiliated centers are less likely to receive guideline-concordant, risk-adapted care, including standardized imaging protocols that inform treatment modification and survivorship planning (Kahn & Beauchemin, 2021; Kahn et al., 2022). Importantly, studies show that racial and ethnic disparities in outcomes are substantially reduced when patients are treated on cooperative group clinical trials, highlighting the role of standardized imaging and treatment guidelines

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in mitigating inequity (Kahn & Beauchemin, 2021). However, disparities re-emerge in relapse management and survivorship, areas that rely heavily on longitudinal imaging to detect recurrence and late effects such as cardiopulmonary toxicity and secondary malignancies, where minority and socioeconomically disadvantaged survivors are less likely to receive guideline-recommended surveillance (Kahn & Beauchemin, 2021; Cheng et al., 2025). Timely access to diagnostic and response-adapted imaging, referral to specialized pediatric cancer centers when feasible, and consistent survivorship imaging across care settings are necessary to reduce avoidable variation and promote equitable outcomes for all children with pediatric HL.

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Pediatric Aggressive Mature B-Cell Non-Hodgkin Lymphomas (NHL) (PEDONC-5.3)

ONCP.HL.0005.3.A
v1.0.2027

Criteria

- Indications and coding for rare circumstances where PET/MRI may be approved are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**.

Indication	Medically Necessary Imaging Study
Initial staging for individuals with aggressive mature B-cell NHL	ANY or ALL of the following: • CT Neck with contrast (CPT® 70491) ◦ MRI Neck without and with contrast (CPT® 70543) in place of CT Neck, if requested • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) in place of CT Abdomen and Pelvis, if requested. ◦ Abdominal ultrasound (CPT® 76700 or 76705) at initial presentation if CT/MRI not available. • CT with contrast or MRI without and with contrast any other symptomatic body area • PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498 [may be approved in addition to diagnostic CTs])
Additional initial staging for individuals with aggressive mature B-cell NHL if symptoms or extent of disease suggest intracranial extension or metastasis	• MRI Brain without and with contrast (CPT® 70553)

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Indication	Medically Necessary Imaging Study
Additional initial staging for individuals with aggressive mature B-cell NHL if symptoms or extent of disease suggest intraspinal extension or metastasis (back pain)	• MRI of whole spine without and with contrast if there is suspected leptomeningeal disease or if sedation will be required
Restaging for treatment response for individuals with aggressive mature B-cell NHL	• **ANY** of the following, as often as every cycle of chemotherapy (~every 3 weeks): ◦ CT with contrast or MRI without and with contrast (should be same modality as initial diagnosis if possible) of previously involved areas ◦ PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) until a negative PET is obtained ▪ Whole-body PET/CT (CPT® 78816) may be approved if there is clinical suspicion of, or known, skull or distal lower extremity involvement. ▪ PET/CT may be approved in conjunction with diagnostic CTs/MRIs, PET/CT should not replace imaging with contrast-enhanced diagnostic-quality CT or MRI.
Restaging for all subsequent treatment response, including end of therapy evaluation, after negative PET/CT (either Deauville or Lugano 1, 2 or 3 as reported in formal radiology interpretation) for individuals with aggressive mature B-cell NHL	• CT with contrast or MRI without and with contrast (should be same modality as initial diagnosis if possible) of previously involved areas, as often as every 2 cycles of chemotherapy, and at the end of therapy

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Indication	Medically Necessary Imaging Study
End of therapy PET with Deauville 4-5 avidity for individuals with aggressive mature B-cell NHL	ONCE, at least 6-8 weeks after end of therapy PET: • PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) OR • CT Neck, Chest, Abdomen, and Pelvis with contrast (CPT® 70491, CPT® 71260, CPT® 74177)
Assessment of disease activity in inconclusive residual masses seen on conventional imaging for individuals with aggressive mature B-cell NHL	• PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498)
Individuals with aggressive mature B-cell NHL being treated with Rituximab who present with abdominal pain, due to risk of bowel perforation and obstruction	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ US, x-ray, or other red flags are not required prior to CT
Surveillance of asymptomatic individuals with aggressive mature B-cell NHL with residual masses in the chest or abdomen and pelvis	• Chest x-ray and Abdominal (CPT® 76700) and Pelvic (CPT® 76856) ultrasound 3 months after completion of therapy. If stable, no further imaging. • In asymptomatic individuals without residual masses, surveillance imaging is not considered medically necessary.

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Indication	Medically Necessary Imaging Study
Clinical symptoms or laboratory findings suggesting recurrence for individuals with aggressive mature B-cell NHL	• MRI Brain without and with (CPT® 70553) for history of CNS involvement or new signs or symptoms suggesting intracranial disease In addition to the above imaging, **ANY** of the following: • CT Neck with contrast (CPT® 70491) **and** CT Chest with contrast (CPT® 71260) **and** CT Abdomen and Pelvis with contrast (CPT® 74177) **and** CT with contrast of other previously involved visceral areas or currently symptomatic areas (MRI with and without contrast in place of CT for all areas other than the chest if requested) **and** • PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498)
Suspected PTLD recurrence with documentation of new palpable nodes, rising LDH, or rising quantitative EBV PCR for individuals with aggressive mature B-cell NHL	• PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) • CT Neck with contrast (CPT® 70491) and CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177)

Background and Supporting Information

- Pediatric aggressive mature B-cell NHL includes Burkitt lymphoma/leukemia, diffuse large B-cell lymphoma, primary mediastinal large B cell lymphoma, post-transplant lymphoproliferative disorder, and viral associated lymphoproliferative disorders, all of which are addressed within a single imaging framework (NCCN, 2026).
- These malignancies are typically high grade and rapidly progressive, with frequent extranodal involvement of the abdomen, mediastinum, bone marrow, or central nervous system at presentation (Mhlanga et al., 2023).
- Staging is performed using the International Pediatric Non-Hodgkin Lymphoma Staging System, which emphasizes the presence of extranodal disease, bone marrow involvement, and CNS involvement (Mhlanga et al., 2023; NCCN, 2026).
- Initial management commonly includes a brief low intensity cytoreduction phase, followed by early treatment response assessment to determine subsequent therapy intensity (NCCN, 2026).

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Evidence Discussion

- Early response imaging performed during initial therapy is supported by pediatric treatment protocols, as it directly informs risk-adapted treatment escalation or de-escalation (NCCN, 2026).
- Due to the aggressive clinical behavior of these lymphomas, relapse most often presents with clear clinical symptoms, and routine asymptomatic surveillance imaging has not been shown to improve outcomes (Mhlanga et al., 2023).
- FDG-PET/CT is valuable for staging and response assessment when disease is FDG avid; however, its use in routine post-therapy surveillance is discouraged because of false positive findings and lack of outcome benefit (Mhlanga et al., 2023; NCCN, 2026).
- Imaging strategies focused on diagnosis, staging, and treatment response optimize disease control while limiting unnecessary radiation exposure in pediatric patients (NCCN, 2026).

Health Equity Considerations

Pediatric aggressive mature B-cell NHL, including Burkitt lymphoma and diffuse large B-cell lymphoma, are highly aggressive malignancies that require rapid diagnosis, comprehensive staging, and timely initiation of intensive, risk adapted therapy. Children frequently present with advanced disease involving extranodal sites such as the abdomen, bone marrow, or central nervous system, necessitating access to advanced imaging modalities, including cross-sectional imaging and functional imaging for accurate staging and response assessment (Jeon et al., 2022). Health equity challenges arise when timely access to specialized pediatric oncology services and advanced imaging is limited, as delays in diagnosis and staging may disproportionately affect children from underserved communities and contribute to higher disease burden at presentation. Because outcomes are closely linked to stage and risk group classification, inequities in early diagnostic pathways and imaging availability may translate into disparities in treatment intensity, toxicity exposure, and survival (Jeon et al., 2022).

Equity considerations extend beyond diagnosis to include treatment monitoring, toxicity surveillance, and long-term follow-up imaging. Intensive multi-agent chemotherapy regimens, such as LMB-based protocols and rituximab-containing therapies for high risk disease, are associated with substantial acute toxicities and risk of late effects, including secondary malignancies, underscoring the need for consistent imaging based surveillance across all patient populations (Jeon et al., 2022). Patients with advanced stage or high risk disease experience worse outcomes and higher rates of complications, highlighting the importance of standardized imaging guidelines to ensure uniform assessment of treatment response and complications regardless of care setting. Standardized staging, response evaluation, and survivorship imaging protocols are needed to reduce variability in care delivery, support early detection of treatment-related

complications, and promote equitable outcomes for children with aggressive mature B-cell NHL across healthcare systems (Jeon et al., 2022).

Pediatric and Special Populations Oncology

Anaplastic Large Cell Lymphoma (ALCL) (PEDONC-5.4)

ONCP.HL.0005.4.A
v1.0.2027

Criteria

- Indications and coding for rare circumstances where PET/MRI is considered medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

Indication	Medically Necessary Imaging Study
Initial staging for individuals with ALCL	• ANY OR ALL of the following: ◦ CT Neck with contrast (CPT® 70491), CT Chest with contrast (CPT® 71260), and CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI without and with contrast of affected area ▪ Substituted for CT in cases of paraspinal or soft tissue extremity primary tumors ◦ CT with contrast or MRI without and with contrast any other symptomatic body area ◦ PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and 76498)
Additional initial staging for individuals with ALCL with bony primary tumors or metastatic disease	• Bone scan (See: **Modality General Considerations (PEDONC-1.3)**)
Restaging at the end of induction chemotherapy (commonly 4 to 6 weeks) for individuals with ALCL	• CT with contrast or MRI without and with contrast of previously involved areas (should be same modality as initial diagnosis if possible)

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Indication	Medically Necessary Imaging Study
Treatment response in individuals with ALCL treated with cytotoxic chemotherapy	• Every 2 cycles: ◦ CT of previously involved areas ▪ If CT is performed for primary treatment response, PET/CT (CPT® 78815 or CPT® 78816) to clarify inconclusive findings detected on conventional imaging OR ◦ PET/CT or PET/MRI (CPT® 78813 and CPT® 76498) until a negative PET is obtained ▪ If PET/CT (CPT® 78815 or CPT® 78816) is performed for primary treatment response, CT or MRI to clarify inconclusive findings detected on PET imaging
Restaging after negative PET/CT (either Deauville or Lugano 1, 2 or 3 as reported in formal radiology interpretation) for individuals with ALCL	• CT with contrast of previously involved areas
Surveillance for individuals with ALCL	• CT with contrast or MRI without and with contrast of all previously involved areas every 6 months for 2 years after completion of therapy.
Additional surveillance for individuals with ALCL with bony primary tumors or metastatic disease	• Bone scan (See: **Modality General Considerations (PEDONC-1.3)** for coding) at 3, 6, 12, and 18 months after therapy is completed
Clinical symptoms suggesting recurrence for individuals with ALCL	• CT Neck with contrast (CPT® 70491), CT Chest with contrast (CPT® 71260), CT Abdomen and Pelvis with contrast (CPT® 74177), and CT with contrast of other previously involved or currently symptomatic areas

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Indication	Medically Necessary Imaging Study
Clarify inconclusive findings on conventional imaging to evaluate the need for biopsy to establish recurrence for individuals with ALCL	• PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498)

Background and Supporting Information

- ALCL is a distinct subtype of pediatric non-Hodgkin lymphoma and may have a clinical presentation similar to Hodgkin lymphoma until immunophenotypic and molecular studies establish the diagnosis (Mhlanga et al., 2023).
- ALCL frequently involves lymph nodes and has a higher incidence of extranodal disease, including skin, lung, bone, and soft tissues, compared with Hodgkin lymphoma (Mhlanga et al., 2023).
- Staging follows the International Pediatric Non-Hodgkin Lymphoma Staging System, consistent with other aggressive pediatric NHL subtypes (Mhlanga et al., 2023).
- Imaging is used to define disease extent and evaluate treatment response rather than to establish histologic diagnosis (Mhlanga et al., 2023).

Evidence Discussion

- FDG-PET/CT is effective for initial staging and response assessment in pediatric ALCL, as disease is typically FDG avid and metabolic response correlates with clinical outcomes (Mhlanga et al., 2023).
- Similar to other aggressive pediatric NHLs, routine surveillance imaging after completion of therapy has not demonstrated a survival benefit, as relapse is usually clinically apparent (Mhlanga et al., 2023).
- Restricting follow-up imaging to clinically indicated scenarios balances early detection of meaningful relapse with avoidance of unnecessary radiation exposure (NCCN, 2026).
- Imaging aligned with treatment milestones and symptom-driven evaluation supports appropriate disease monitoring without increasing false positive findings or unnecessary interventions (Mhlanga et al., 2023).

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Neuroblastoma (PEDONC-6)

Guideline Page

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Neuroblastoma – General Considerations (PEDONC-6.1)

ONCP.NP.0006.1.A
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Criteria

- PEDONC-6 should be used to review neuroblastoma, ganglioneuroblastoma, and ganglioneuroma in individuals of all ages, with the exception of esthesioneuroblastoma in individuals of all ages, which should be reviewed using **Squamous Cell Carcinomas of the Head and Neck (ONC-3)** in the Oncology Imaging Guidelines.
- Neuroblastoma is divided into very low, low, intermediate, and high-risk disease based on International Neuroblastoma Risk Group (INRG) Staging System (see: **Staging and Risk Grouping (PEDONC-6.2)**). The treatment approaches for each risk group vary widely and have distinct imaging strategies. The risk group for a given individual should be provided by the ordering provider in the clinical information provided for review.
- For metabolic imaging in individuals who are MIBG positive at diagnosis and then become MIBG negative in response to treatment:
  - Continue to use MIBG (see table below and **PEDONC-1.3** for coding)

Study Type	Coding
Bone scan	▪ **ANY** of the following codes are considered medically necessary: - CPT® 78300 - CPT® 78305 - CPT® 78306 - CPT® 78803, 78830, or 78832 • In addition to the above imaging, **ANY** of the following are considered medically necessary : ◦ CPT® 78305 ◦ CPT® 78306

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Study Type	Coding
^{123}I-metaiodobenzylguanidine (MIBG) scintigraphy	▪ **ONE** of the following codes is considered medically necessary: - CPT® 78801 - CPT® 78802 - CPT® 78804 ▪ In addition to the above imaging, **ONE** of the following is considered medically necessary: - CPT® 78803 - CPT® 78830 - CPT® 78831 - CPT® 78832 **OR** ▪ CPT® 78800 is considered medically necessary for KNOWN neuroblastoma when only a single-site follow up is desired but is not sufficient for the initial workup of suspected disease.
Octreotide scan	▪ Same coding as MIBG
Gallium scan	▪ Same coding as MIBG

- MIBG remains the standard of care metabolic imaging in neuroblastoma. 18F-FDG-PET-CT is not considered medically necessary unless one of the exceptions below is present. **All PET imaging in PEDONC-6 refers to 18F-FDG radiotracer.** All other radiotracers are considered not considered medically necessary at this time:

Indication	Medically Necessary Imaging Study
◦ Any of the following: ▪ individuals with MIBG-negativity documented at initial diagnosis ▪ individuals with discordant findings on MIBG and conventional imaging (i.e., it is suspected there is more active disease than is visible on MIBG) ▪ at major decision points (such as hematopoietic stem cell transplant or surgery), if MIBG and CT/MRI findings are inconclusive	◦ Whole-body 18F-FDG PET/CT (CPT® 78816) ◦ In scenarios where PET/CT is considered medically necessary, it does not preclude the other diagnostic imaging studies considered medically necessary throughout **PEDONC-6**. PET/CT is viewed as replacing MIBG in these scenarios. Indications and coding for rare circumstances where PET/MRI is considered medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

Indication	Medically Necessary Imaging Study
◦ Individuals currently receiving medications that may interfere with MIBG uptake that cannot be safely discontinued prior to imaging, including: ▪ tricyclic antidepressants (amitriptyline, imipramine, etc.) ▪ selective serotonin reuptake inhibitors (SSRI's, sertraline, paroxetine, escitolapram, etc.) ▪ neuroleptics (risperidone, haloperidol, etc.) ▪ antihypertensive drugs (alpha or beta blockers, calcium channel blockers) ▪ decongestants (phenylephrine, ephedrine, pseudoephedrine) ▪ stimulants (methylphenidate, dextroamphetamine, etc.)	◦ 18F-FDG PET/CT (CPT® 78816) is only considered medically necessary when specific documentation of the medication interaction is included with the current PET imaging request. ◦ Indications and coding for rare circumstances where PET/MRI is considered medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

Background and Supporting Information

- Neuroblastoma is the most common extracranial solid tumor of childhood and arises from the developing sympathetic nervous system, most frequently from the adrenal gland or paraspinal sympathetic chain (NCCN, 2026).
- Neuroblastoma demonstrates marked biologic heterogeneity, ranging from tumors that spontaneously regress to aggressive metastatic disease requiring multimodality therapy (NCCN, 2026).
- The majority of neuroblastomas produce catecholamines, and elevated urinary homovanillic acid (HVA) and vanillylmandelic acid (VMA) are commonly identified at diagnosis and may assist in disease evaluation (NCCN, 2026).
- Imaging plays a central role in diagnosis, staging, surgical planning, treatment response assessment, and post-treatment surveillance for neuroblastoma (NCCN, 2026).

Evidence Discussion

- Cross-sectional imaging and functional nuclear imaging provide complementary information that is essential for accurate disease characterization and risk stratification (NCCN, 2026).

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- Technetium 99m bone scintigraphy has limited sensitivity for neuroblastoma skeletal metastases and does not provide added value when MIBG scintigraphy is available, supporting exclusion from routine evaluation (NCCN, 2026).
- Imaging strategies that limit unnecessary radiation exposure while preserving diagnostic accuracy are particularly important in pediatric populations with favorable long term survival (NCCN, 2026).

Health Equity Considerations

Neuroblastoma is marked by substantial inequities in incidence, stage at diagnosis, treatment delivery, and outcomes that reflect both global and U.S.-based disparities in socioeconomic resources, race and ethnicity, and access to specialized pediatric oncology care. Global burden analyses demonstrate that children in low and low middle sociodemographic index (SDI) regions experience disproportionate increases in incidence, mortality, and disability-adjusted life years (DALYs), largely driven by delayed diagnosis, limited access to advanced imaging, and constrained availability of risk-adapted multimodal therapy (Nong et al., 2025; Zhang et al., 2025). In contrast, high SDI regions have seen declining disease burden, attributable to earlier detection, widespread availability of diagnostic imaging, and integration of advanced therapies such as immunotherapy (Zhang et al., 2025). Within the United States, population-based studies consistently show that Black and Hispanic children are more likely to present with high-risk disease and advanced stage, which necessitates more intensive imaging for staging, response assessment, and surveillance, and is associated with worse survival (Campbell et al., 2025; Chennakesavalu et al., 2023). These disparities persist across eras of modern therapy and reflect the cumulative impact of structural racism, socioeconomic disadvantage, and inequitable access to high quality diagnostic pathways and imaging resources (Chennakesavalu et al., 2023; Umaretiya et al., 2025).

Disparities also extend into treatment monitoring, toxicity management, and survivorship, all of which rely heavily on longitudinal imaging. Even within highly standardized Children's Oncology Group (COG) clinical trials, Black and Hispanic children with high-risk neuroblastoma experience inferior overall survival compared with non-Hispanic White children, indicating that uniform treatment protocols alone do not eliminate inequity (Umaretiya et al., 2025). Evidence suggests that differential treatment-related toxicity and healthcare utilization contribute to these outcomes; for example, Hispanic children undergoing post-consolidation immunotherapy have markedly higher rates of ICU-level care, reflecting potentially preventable differences in supportive care and toxicity monitoring (Shoag et al., 2025). Socioeconomic disadvantage, including public insurance and neighborhood level poverty, further compounds these risks and may limit consistent access to surveillance imaging for relapse detection and late effect monitoring (Campbell et al., 2025; Shoag et al., 2025). Timely and equitable access to diagnostic and functional imaging, standardized response adapted and survivorship imaging across care settings, and coordination with supportive care resources are

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needed. Such an approach can help reduce avoidable variation in imaging utilization, support early identification of complications or relapse, and promote more equitable outcomes for children with neuroblastoma across diverse populations (Chennakesavalu et al., 2023; Umaretiyia et al., 2025).

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Staging and Risk Grouping – Neuroblastoma (PEDONC-6.2)

ONCP.NP.0006.2.A
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Criteria

- Most recent treatment protocols are using the updated International Neuroblastoma Risk Group (INRG) staging system
  - L1: Localized tumor not involving vital structures as defined by the list of image-defined risk factors and confined to one body compartment
    - Image-defined risk factors include a list of specific imaging findings defining individuals less likely to be candidates for complete surgical resection
    - These risk factors involve the encasement of major blood vessels, airway, skull base, costovertebral junction, brachial plexus, spinal canal, or major organs or structures
  - L2: Locoregional tumor with presence of one or more image-defined risk factors
  - M: Distant metastatic disease (except stage MS)
  - MS: Metastatic disease in children younger than 18 months with metastases confined to skin, liver, and/or bone marrow with <10% involvement (MIBG must be negative in bone and bone marrow)
- The risk group for any given individual should be provided or documented by the requesting provider.

Background and Supporting Information

- Tumors are classified as L1, L2, M, or MS based on anatomic extent and the presence of image defined risk factors (IDRFs) that indicate local tumor invasiveness (NCCN, 2026).
- Risk stratification incorporates staging along with age, MYCN amplification status, histopathology, segmental chromosomal aberrations, and tumor ploidy to determine low, intermediate, or high-risk disease (NCCN, 2026).

Evidence Discussion

- Use of an imaging-based staging system improves consistency of disease classification and enables pretreatment surgical risk assessment (NCCN, 2026).
- Identification of IDRFs on imaging correlates with surgical complexity and informs treatment planning without delaying therapy initiation (NCCN, 2026).

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- Risk-adapted imaging recommendations support optimized outcomes while avoiding unnecessary diagnostic testing across distinct prognostic groups (NCCN, 2026).

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Neuroblastoma – Initial Staging (PEDONC-6.3)

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Criteria

The following imaging studies are considered medically necessary in the initial staging of individuals with neuroblastoma, ganglioneuroblastoma, or ganglioneuroma:

Indication	Medically Necessary Imaging Study
Initial staging for all individuals with neuroblastoma, ganglioneuroblastoma, or ganglioneuroma	• ^{123}I-metaiodobenzylguanidine scintigraphy (see table in **Neuroblastoma – General Considerations (PEDONC-6.1)** for MIBG coding details) AND • ONE of the following sets of imaging: ◦ CT Neck with contrast (CPT® 70491), CT Chest with contrast (CPT® 71260), and CT Abdomen and Pelvis with contrast (CPT® 74177) OR • MRI Neck without and with contrast (CPT® 70543), MRI Chest without and with contrast (CPT® 71552), and MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)
• All individuals with neuroblastoma, ganglioneuroblastoma, or ganglioneuroma with paraspinal tumors • Individuals with neuroblastoma, ganglioneuroblastoma, or ganglioneuroma with back pain or cord compression symptoms	In addition to the above imaging: • MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), and Lumbar (CPT® 72158) spine without and with contrast

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Indication	Medically Necessary Imaging Study
Evaluation of suspected adrenal neuroblastoma, ganglioneuroblastoma, or ganglioneuroma when CT or MRI is inconclusive for an adrenal lesion in individuals with neuroblastoma, ganglioneuroblastoma, or ganglioneuroma	• Adrenal nuclear imaging (CPT® 78075)
Clinical signs/symptoms suggest brain and/or orbital involvement in individuals with neuroblastoma, ganglioneuroblastoma, or ganglioneuroma	**ANY**of the following: • MRI Brain without and with contrast (CPT® 70553) • CT Head with contrast (CPT® 70470) • CT Orbits/Maxillofacial with contrast (CPT® 70487)
Pediatric Horner's syndrome with suspicion for neuroblastoma	ANY of the following: • MRI Brain without contrast or MRI Brain without and with contrast (CPT® 70551 or 70553) • MRI Orbits (CPT® 70543) or CT Neck with contrast (CPT® 70491) +/- CT Orbits/Temporal Bone with contrast (CPT® 70481) • MRI Chest without and with contrast (CPT® 71552) or CT Chest with contrast (CPT® 71260) If suspected spinal cord lesions, MRI Cervical Spine without contrast (CPT® 72141) or without and with contrast (CPT® 72156) If abdominal symptoms or elevated catecholemines (HVA and HMA), MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) For other etiologies of Horner's syndrome, see **Pupillary Abnormalities Including Horner's Syndrome (HD-32.2)**

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- PET should not be used unless one of the exceptions stated in section **Neuroblastoma – General Considerations (PEDONC-6.1)** is present.

Evidence Discussion

- Initial staging requires contrast-enhanced CT or MRI to evaluate the primary tumor, assess regional extension, and identify IDRFs (NCCN, 2026).
- I-123 metaiodobenzylguanidine (MIBG) scintigraphy, with SPECT or SPECT/CT when available, is the preferred modality for detecting metastatic disease due to high tumor avidity (NCCN, 2026).
- MIBG scintigraphy demonstrates high sensitivity and specificity for osseous and soft tissue metastases and is superior to conventional bone scintigraphy for neuroblastoma evaluation (NCCN, 2026).
- SPECT or SPECT/CT improves lesion localization and increases diagnostic confidence without requiring separate diagnostic CT imaging (NCCN, 2026).
- Delay of emergent therapy to complete imaging studies may be harmful in select cases, supporting timely but medically appropriate completion of staging evaluations (NCCN, 2026).

Neuroblastoma – Treatment Response Imaging (Risk Group Dependent) (PEDONC-6.4)

ONCP.NP.0006.4.A
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Criteria

- Risk Grouping will not be known at the time of initial staging, but is critical for all imaging decisions after initial staging is complete. **The treating oncologist should always know the individual's risk grouping**. It is not possible to establish the appropriate imaging plan for a neuroblastoma individual without knowing his/her risk group.

All Very Low Risk and Low Risk Neuroblastoma Not Receiving Chemotherapy:

Indication	Medically Necessary Imaging Study
All individuals with very low risk and low risk neuroblastoma not receiving chemotherapy, no sooner than 4 weeks after diagnosis to determine if additional treatment is necessary	• CT with contrast or MRI without and with contrast of the primary tumor site. If primary tumor site is abdomen or pelvis, imaging of both sites is considered medically necessary. ◦ Ultrasound is considered medically necessary if requested in place of CT or MRI to avoid radiation and/ or anesthesia exposure in low risk individuals

- Many individuals will be treated with surgical resection only without adjuvant therapy, and these individuals enter immediately into surveillance.

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All Intermediate Risk Neuroblastoma and Very Low Risk or Low Risk Neuroblastoma Receiving Chemotherapy:

Indication	Medically Necessary Imaging Study
Prior to surgical resection in individuals with intermediate risk neuroblastoma and individuals with very low risk or low risk neuroblastoma receiving chemotherapy	• Restaging imaging (MIBG and CT or MRI, as performed at initial diagnosis) • PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met. • Additional imaging for individuals with paraspinal disease: ◦ MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158)
Treatment response, as often as every 2 cycles of chemotherapy (~every 6 weeks and at the end of planned treatment) in individuals with intermediate risk neuroblastoma and individuals with very low risk or low risk neuroblastoma receiving chemotherapy	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) OR • MRI Neck (CPT® 70543) without and with contrast, MRI Chest without and with contrast (CPT® 71552), and MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) AND • CT or MRI of other sites with prior measurable disease • Additional imaging for individuals with paraspinal disease: ◦ MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158)
In addition to treatment response listed above, every 4 cycles, and at the end of planned chemotherapy treatment in individuals with intermediate risk neuroblastoma and individuals with very low risk or low risk neuroblastoma receiving chemotherapy	• MIBG scan (see table in **Neuroblastoma - General Considerations (PEDONC-6.1)** for MIBG coding details) • 18F-FDG PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met

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High-Risk Neuroblastoma:

Indication	Medically Necessary Imaging Study
• ANY of the following in individuals with high-risk neuroblastoma: ◦ Treatment response ▪ As often as every 2 cycles of chemotherapy, mAb, or biologic therapy (~every 6 weeks) ◦ Change in modality ▪ Prior to surgery, HSCT, XRT, or mAb therapy ◦ End of therapy	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) OR • MRI Neck (CPT® 70543) without and with contrast, MRI Chest without and with contrast (CPT® 71552), and MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) AND • CT or MRI of other sites with prior measurable disease • Additional imaging for individuals with paraspinal disease: ◦ MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158) • MIBG scan (see table in **Neuroblastoma - General consideration (PEDONC-6.1)** for MIBG coding details)
At completion of ^{131}I-MIBG therapy in individuals with high-risk neuroblastoma	• ^{123}I-MIBG scan ◦ FDG-PET cannot be used after ^{131}I-MIBG therapy
Preoperative planning in individuals with high-risk neuroblastoma	• More frequent imaging with any of the above modalities can be approved around the time of surgery if needed

Background and Supporting Information

- Treatment response assessment is guided by the International Neuroblastoma Response Criteria (INRC), which combine anatomic and functional imaging parameters (NCCN, 2026).
- Response evaluation incorporates RECIST-based measurements for soft tissue disease and semiquantitative scoring of MIBG avid skeletal disease (NCCN, 2026).
- The timing and frequency of response imaging vary by risk group and treatment phase, particularly for high-risk neuroblastoma receiving multimodal therapy (NCCN, 2026).

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Evidence Discussion

- Functional imaging changes often precede anatomic tumor regression, supporting continued use of MIBG-based response assessment during therapy (NCCN, 2026).
- Standardized response criteria reduce interobserver variability and support consistent interpretation across clinical trials and treatment centers (NCCN, 2026).
- Avoidance of non-informative imaging modalities reduces cumulative radiation exposure without compromising assessment of treatment efficacy (NCCN, 2026).

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Neuroblastoma – Surveillance Imaging (Risk Group Dependent) (PEDONC-6.5)

ONCP.NP.0006.5.A
v1.0.2027

Criteria

Very Low Risk and Low Risk Neuroblastoma:

Indication	Medically Necessary Imaging Study
All individuals with very low risk and low risk neuroblastoma	• At 3, 6, 9, 12, 18, 24, 30 and 36 months after surgery: ◦ Ultrasound of involved areas OR • 3, 6, 9, 12, 18, 24, 30 and 36 months after surgery or to clarify unclear findings on ultrasound: ◦ CT with contrast or MRI without and with contrast of the primary tumor site
Clarification of findings on CT or MRI suspicious for disease recurrence in individuals with very low risk and low risk neuroblastoma	• MIBG (see table in **Neuroblastoma - General Considerations (PEDONC-6.1)** for MIBG coding details) • 18F-FDG PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met.

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Intermediate Risk Neuroblastoma:

Indication	Medically Necessary Imaging Study
All individuals with intermediate risk neuroblastoma	• Every 3 months for 1 year, then every 6 months for 1 year, and then at 36 months after surgery: ◦ CT with contrast or MRI without and with contrast of the primary tumor ▪ If the primary tumor is paraspinal: - MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158) ◦ If primary site is abdomen or pelvis, both body sites are considered medically necessary (CPT® 74177 or the combination of CPT® 71297 and CPT® 74183) ◦ If negative at 36 months, ultrasound at 48 and 60 months after surgery to complete 5 years surveillance.
• BOTH of the following in individuals with intermediate risk neuroblastoma: ◦ Individuals with stage 4, or M disease, or 4S, or MS disease AND ◦ Positive MIBG at completion of therapy	• Until a negative scan is achieved, every 3 months in year 1, then once in year 2 and year 3: ◦ MIBG scan (see table in **Neuroblastoma – General Considerations (PEDONC-6.1)** for MIBG coding details) ◦ Once a negative MIBG is achieved, no further MIBG imaging is necessary. • MIBG is not considered medically necessary for all other intermediate risk individuals. • 18F-FDG PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met.

High Risk Neuroblastoma:

Indication	Medically Necessary Imaging Study
All individuals with high-risk neuroblastoma	• Every 3 months for 1 year, then every 6 months for 1 year, and then annually to complete 6 years surveillance: ◦ CT with contrast or MRI without and with contrast of the primary tumor site AND ◦ MIBG scan (see table in **Neuroblastoma – General Considerations (PEDONC-6.1)** for MIBG coding details) ▪ 18F-FDG PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met ◦ If primary site is abdomen or pelvis, both body sites are considered medically necessary (CPT® 74177 or the combination of CPT® 71297 and CPT® 74183) ◦ For history of paraspinal disease, MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158) ◦ If negative at 6 years, no further advanced imaging.
Suspected recurrence in individuals with high-risk neuroblastoma	• CT Chest/Abdomen/Pelvis with contrast (CPT® 71260 and CPT® 74177) or MRI Chest/Abdomen/Pelvis without and with contrast, (CPT® 71552, CPT® 74183, and CPT® 72197) and other sites of prior measurable disease or current symptoms • MRI Brain without and with contrast (CPT® 70553) for signs or symptoms of brain involvement • MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158) for history of paraspinal disease, back pain, or cord compression symptoms. • MIBG scan (see table in **Neuroblastoma - General Considerations (PEDONC-6.1)** for MIBG coding details) • 18F-FDG PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met.

Background and Supporting Information

- Post-therapy surveillance imaging strategies are risk-adapted and tailored to disease biology, stage at diagnosis, and treatment intensity (NCCN, 2026).
- Low-risk individuals typically undergo limited imaging, often ultrasound or targeted cross-sectional imaging of the primary site (NCCN, 2026).

- High-risk individuals require more intensive surveillance, including periodic MIBG or FDG PET imaging during the early post-treatment period (NCCN, 2026).

Evidence Discussion

- Risk stratified surveillance balances early detection of clinically significant relapse with minimization of radiation exposure and false positive findings (NCCN, 2026).
- Routine chest CT is not necessary in asymptomatic patients without prior thoracic involvement, supporting selective site directed imaging (NCCN, 2026).
- In high-risk neuroblastoma, early detection of recurrence using MIBG imaging has been associated with improved post-relapse outcomes, supporting continued use in this population (NCCN, 2026).

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Pediatric Renal Tumors (PEDONC-7)

Guideline Page

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Pediatric Renal Tumors – General Considerations (PEDONC-7.1)

ONCP.RC.0007.1.C
v1.0.2027

Criteria

- A variety of tumors can occur in the pediatric kidney, and include the following:
  - Wilms Tumor
    - Favorable histology (FHWT)
    - Focal anaplasia (FAWT)
    - Diffuse anaplasia (DAWT)
    - Bilateral Wilms Tumor (BWT)
  - Renal Cell Carcinoma (RCC)
  - Clear Cell Sarcoma of the Kidney (CCSK)
  - Malignant Rhabdoid Tumor of the Kidney (MRT)
  - Congenital Mesoblastic Nephroma (CMN)
  - Other Cancers occurring in the Kidney:
    - Neuroblastoma
    - Primitive Neuroectodermal Tumor
    - Rhabdomyosarcoma
    - Non-Rhabdomyosarcoma Soft Tissue Sarcomas
    - These and other rare tumors have been reported occurring primarily in the kidney and should be imaged according to the guidelines for the specific histologic diagnosis.
- For suspected renal tumor, cell type unknown, image according to **Pediatric Renal Cell Carcinoma (RCC) (PEDONC-7.4)**
- PET is not routinely considered medically necessary for initial staging, treatment response, or surveillance of any pediatric renal tumor – rare circumstances for exceptions are listed in the relevant guideline sections.

Background and Supporting Information

- Pediatric renal tumors most commonly present as an asymptomatic abdominal mass and are initially evaluated with ultrasound due to the absence of ionizing radiation and high sensitivity for renal origin (NCCN, 2025).
- Cross-sectional imaging with contrast-enhanced CT or MRI is used to define tumor extent, assess contralateral kidney involvement, evaluate vascular invasion, and identify metastatic disease, most commonly to the lungs (NCCN, 2025).

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Evidence Discussion

- Imaging strategies emphasizing ultrasound followed by targeted CT or MRI provide adequate staging information while minimizing radiation exposure in children, supporting guideline-directed modality selection (NCCN, 2025).
- PET imaging is not routinely considered medically necessary in the initial evaluation of pediatric renal tumors due to limited incremental diagnostic value over established anatomic imaging modalities (NCCN, 2025).
- Avoidance of unnecessary advanced imaging reduces cumulative radiation dose and procedural sedation risks without compromising oncologic outcomes (Mullen et al., 2018).

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Unilateral Wilms Tumor (UWT) (PEDONC-7.2)

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Criteria

Indication	Medically Necessary Imaging Study
Initial Staging for individuals with UWT	ONE of the following: • MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) OR • CT Abdomen with contrast (CPT® 74160) or CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ Doppler ultrasound to evaluate for tumor thrombus is not considered medically necessary unless CT findings are inconclusive AND • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ Should be completed prior to anesthesia exposure if possible • PET is not routinely considered medically necessary in the initial staging of any pediatric renal tumor
Bilateral renal lesions noted on ultrasound or CT for individuals with UWT	• MRI Abdomen (CPT® 74183) and Pelvis (CPT® 72197) without and with contrast
Additional initial staging imaging for any individual with UWT with neurologic signs or symptoms raising suspicion of CNS metastases	• MRI Brain without and with contrast (CPT® 70553)

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Indication	Medically Necessary Imaging Study
Additional initial staging imaging for any individual with UWT with signs or symptoms raising suspicion of bony metastases	• Bone scan (see: **PEDONC-1.3** for coding)
Treatment response ~every 2 cycles during treatment and at the end of planned therapy for individuals with UWT	• CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) • CT Abdomen with contrast (CPT® 74160) or CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen (CPT® 74183) and Pelvis (CPT® 72197) without and with contrast
Rare circumstances to establish the presence of active disease only when a major therapeutic decision depends on PET avidity for individuals with UWT	• PET/CT (CPT® 78815)
Surveillance for individuals with UWT	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250), +/- chest x-ray AND CT Abdomen with contrast (CPT® 74160) or Abdominal US (CPT® 76700) ◦ Every 3 months for 2 years, then every 6 months for 2 additional years, to complete 4 years surveillance • In addition, pelvic imaging (CT or ultrasound) for individuals with stage II or higher disease, for individuals treated with nephrectomy only, or for individuals with a history of tumor rupture, known pelvic involvement, diffuse anaplastic histology, or a history of disease recurrence.
• There are no data to support the use of PET imaging for routine surveillance in any individual with Wilms tumor. • A very low risk subset of stage I FHWT will be observed after nephrectomy, and enter directly into surveillance.	

Background and Supporting Information

- Most individuals with unilateral Wilms tumor present with an asymptomatic abdominal mass and undergo ultrasound as the initial diagnostic study (NCCN, 2026).
- Brain metastases are rare in Wilms tumor, occurring in approximately 0.5% of cases (NCCN, 2026).

Evidence Discussion

- Surveillance studies demonstrate that routine CT imaging does not improve overall survival after relapse compared with chest radiograph and abdominal ultrasound in favorable-histology unilateral Wilms tumor (Mullen et al., 2018).
- Elimination of CT from routine surveillance significantly reduces radiation exposure and cost without adversely affecting survival outcomes, supporting restricted imaging approaches (Mullen et al., 2018).
- Most recurrences are detected within the first two years after treatment, supporting time-limited and modality-selective surveillance strategies (Mullen et al., 2018; NCCN, 2026).

Health Equity Considerations

Children with unilateral Wilms tumor experience measurable disparities in incidence, treatment, and survival that are closely linked to social determinants of health (SDOH), including race, ethnicity, socioeconomic status (SES), and geography. Population-based analyses demonstrate higher incidence and lower survival among non-Hispanic Black children and those residing in non-metropolitan or socioeconomically disadvantaged counties, even in the context of modern, protocol-driven therapy (Espinoza et al., 2025; Chalfant et al., 2023). County- and neighborhood-level deprivation indices show a graded association with outcomes, with children from more deprived areas experiencing worse long-term overall survival compared with peers from less deprived areas, despite similar stage at presentation (Chalfant et al., 2023). These findings suggest that disparities are not explained solely by tumor biology but also by differences in access to care, resources supporting treatment adherence, and capacity for long-term surveillance.

Socioeconomic disadvantage has also been associated with differences in treatment delivery, including reduced likelihood of receiving recommended chemotherapy in lower-SES populations, which may contribute to observed survival gaps over time (Bhambhvani et al., 2022). Recent national database studies further demonstrate that higher neighborhood-level socioeconomic disadvantage independently predicts worse survival in Wilms tumor, even after adjusting for age, tumor size, bilateral or metastatic disease, and comorbidities, highlighting the cumulative impact of social risk factors on outcomes (Nofi et al., 2025). Structural barriers such as transportation challenges, insurance instability, distance from pediatric specialty centers, and reduced access to

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supportive services may delay diagnosis, complicate imaging follow-up, or limit timely detection of relapse. Awareness of these inequities is essential when applying imaging guidelines, as children from underserved communities may require additional care coordination and support to ensure equitable access to staging, surveillance imaging, and long-term follow-up (Espinoza et al., 2025; Nofi et al., 2025).

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Bilateral Wilms Tumor (BWT) (PEDONC-7.3)

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Criteria

Indication	Medically Necessary Imaging Study
Initial Staging for individuals with BWT	ONE of the following: • MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) OR • CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) is often performed prior to discovery of bilateral lesions and should not prevent MRI from being approved if requested ▪ Doppler ultrasound to evaluate for tumor thrombus is not considered medically necessary unless CT findings are inconclusive AND • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ Should be completed prior to anesthesia exposure if possible • PET is not routinely considered medically necessary in the initial staging of any pediatric renal tumor
Additional initial staging imaging for any individual with BWT with neurologic signs or symptoms raising suspicion of CNS metastases	• MRI Brain without and with contrast (CPT® 70553)

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Indication	Medically Necessary Imaging Study
Additional initial staging imaging for any individual with BWT with signs or symptoms raising suspicion of bony metastases	• Bone scan (see: **PEDONC-1.3** for coding)
Treatment response ~every 2 cycles during treatment and at the end of planned therapy for individuals with BWT	• MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) for individuals with a contraindication to MRI • CT Chest with (CPT® 71260) or without contrast (CPT® 71250)
Rare circumstances to establish the presence of active disease only when a major therapeutic decision depends on PET avidity for individuals with BWT	• PET/CT (CPT® 78815)

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Indication	Medically Necessary Imaging Study
Surveillance for individuals with BWT	• Every 3 months for 2 years, then every 6 months for 2 additional years, to complete 4 years surveillance: ◦ CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) +/- chest x-ray AND ◦ CT Abdomen with contrast (CPT® 74160) or Abdominal US (CPT® 76700) • Pelvic imaging (CT or ultrasound) for individuals with stage II or higher disease, for individuals treated with nephrectomy only, or individuals with a history of tumor rupture, diffuse anaplastic histology, known pelvic involvement, or a history of disease recurrence.
• PET is not routinely considered medically necessary to assess treatment response or for surveillance in Wilms tumor.	

Background and Supporting Information

- Individuals with bilateral Wilms tumor often present at a younger age and may initiate therapy without tissue diagnosis to preserve renal parenchyma and prevent disease upstaging (NCCN, 2025).
- Neoadjuvant chemotherapy is commonly used to reduce tumor burden prior to nephron-sparing surgical approaches (NCCN, 2025).
- Brain metastases remain rare in bilateral disease, similar to unilateral Wilms tumor (NCCN, 2025).

Evidence Discussion

- Risk-adapted imaging during neoadjuvant therapy allows assessment of tumor response and surgical feasibility while minimizing unnecessary repeat imaging (NCCN, 2025).
- Limiting surveillance imaging to clinically meaningful intervals reduces long-term radiation exposure in a population at elevated risk of chronic kidney disease and treatment-related morbidity (NCCN, 2025).

Health Equity Considerations

Bilateral Wilms tumor represents a more complex clinical entity than unilateral disease and amplifies the impact of health inequities related to social determinants of health (SDOH). Although bilateral tumors account for a minority of Wilms tumor cases, they require prolonged, resource-intensive care, including specialized imaging for staging and treatment response, nephron-sparing surgical expertise, and long-term

surveillance to preserve renal function. Population-based data demonstrate that children from socioeconomically disadvantaged or socially deprived communities experience worse overall survival in Wilms tumor, independent of tumor stage, suggesting that children with higher-risk and more complex presentations, such as bilateral disease, may be disproportionately affected by barriers to timely diagnosis, subspecialty referral, and longitudinal follow-up (Chalfant et al., 2023; Nofi et al., 2025). Racial and ethnic disparities persist as well, with non-Hispanic Black children experiencing higher incidence rates and lower survival compared with non-Hispanic White children, underscoring the intersection of biological risk and structural inequities in access to comprehensive oncology care (Espinoza et al., 2025).

Children with bilateral Wilms tumor are particularly vulnerable to inequities related to treatment delivery and imaging adherence, as optimal management often relies on serial imaging to guide preoperative chemotherapy, assess renal preservation strategies, and monitor for relapse. Socioeconomic disadvantage has been associated with reduced likelihood of receiving recommended chemotherapy in pediatric Wilms tumor populations, which may further exacerbate outcome disparities in bilateral disease, where protocol adherence is critical to balancing oncologic control and renal preservation (Bhambhvani et al., 2022). Geographic barriers, insurance instability, transportation limitations, and reduced access to pediatric centers with nephron-sparing surgical expertise may limit consistent imaging surveillance and coordinated multidisciplinary care. Incorporating health equity considerations into imaging guidelines for bilateral Wilms tumor is essential to support consistent staging and surveillance across populations and to recognize that children from underserved communities may require additional care coordination and support to ensure equitable access to imaging, treatment decision-making, and long-term follow-up (Espinoza et al., 2025; Nofi et al., 2025).

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Pediatric Renal Cell Carcinoma (RCC) (PEDONC-7.4)

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Criteria

- Individuals of any age with TFE3 or TFEB translocated RCC should be imaged according to this guideline section.
- Individuals with all other subtypes of RCC should be imaged according to **Renal Cell Cancer (RCC) (ONC-17)** in the Oncology Imaging Guidelines.

Indication	Medically Necessary Imaging Study
Initial Staging for individuals with RCC	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ If bilateral renal lesions are noted on ultrasound or CT, MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) are considered medically necessary • CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) ◦ Should be completed prior to anesthesia exposure if possible • PET scan is not routinely considered medically necessary in the initial staging of any pediatric renal tumor
Additional initial staging for any individual with RCC with neurologic signs or symptoms raising suspicion of CNS metastases	• MRI Brain without and with contrast (CPT® 70553)
Additional initial staging for any individual with RCC with signs or symptoms raising suspicion of bony metastases	• Bone scan (See: **PEDONC-1.3** for coding)

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Indication	Medically Necessary Imaging Study
Treatment response in individuals with RCC with residual measurable disease after initial surgery and receiving adjuvant medical therapy	• Every 2 cycles during active treatment: ◦ CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) and ◦ CT Abdomen with contrast (CPT® 74160) ▪ Pelvic imaging is not considered medically necessary unless prior pelvic involvement has been documented
Rare circumstances to establish the presence of active disease only when a major therapeutic decision depends on PET avidity for individuals with RCC	• PET/CT (CPT® 78815) • Indications and coding for rare circumstances where PET/MRI is considered medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**
Surveillance in individuals with RCC with documented CNS metastases	• Every 6 months for 2 years after completion of all therapy: ◦ MRI Brain without and with contrast (CPT® 70553)
Surveillance in individuals with RCC with TFE3 or TFEB subtype	• Every 3 months for 2 years, then every 6 months for 2 years after completion of all therapy: ◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183) ▪ Pelvic imaging is not considered medically necessary for surveillance unless prior pelvic involvement has been documented
Surveillance in all other histologies for individuals with RCC	• See: **Renal Cell Cancer (RCC) – Surveillance (ONC-17.4)**
New signs/symptoms suggestive of CNS recurrence for individuals with RCC	• MRI Brain without and with contrast (CPT® 70553)
• PET is not routinely utilized to assess treatment response in pediatric RCC. •	

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Background and Supporting Information

- Pediatric RCC is biologically distinct from adult RCC, with the majority of cases involving TFE3 or TFEB translocations, while 40%–45% demonstrate adult-type histologies (Geller et al., 2020).
- Most patients undergo complete surgical resection at diagnosis and enter surveillance without adjuvant therapy when disease is localized (Geller et al., 2020).

Evidence Discussion

- Doppler ultrasound for tumor thrombus evaluation is not routinely considered medically necessary when CT adequately assesses venous involvement (NCCN, 2025).
- Prospective COG data demonstrate excellent outcomes for completely resected pediatric RCC, including node-positive disease, supporting surveillance-based follow-up without routine adjuvant imaging escalation (Geller et al., 2020).
- Imaging decisions similar to adult RCC guidelines are appropriate only for pediatric cases with adult-type histology, supporting histology-driven imaging strategies (Geller et al., 2020).
- Limiting surveillance to clinically indicated imaging avoids unnecessary radiation exposure in a population with favorable long-term survival (Geller et al., 2020).

Health Equity Considerations

Pediatric RCC is a rare but aggressive malignancy that demonstrates pronounced health inequities related to race, socioeconomic conditions, and geographic context. Population-based and state-level analyses show that Black children are disproportionately affected by advanced-stage presentation and markedly worse survival compared with White children, particularly for non-Wilms renal tumors such as RCC (Neuzil et al., 2020; Holmes et al., 2024). National SEER data indicate that Black children with RCC experience a two- to four-fold higher risk of mortality than their White peers, even after adjustment for age, sex, insurance status, and selected socioeconomic variables, highlighting the persistent influence of structural and environmental inequities on outcomes (Holmes et al., 2024). These disparities are compounded by the fact that RCC often presents in adolescence, when symptoms may be nonspecific and access to timely diagnostic imaging and subspecialty evaluation may be uneven across populations and care settings (Neuzil et al., 2020).

Geographic and environmental factors further modify inequities in RCC outcomes. Children residing in urban or non-metropolitan areas, as opposed to metropolitan regions with access to high-volume pediatric oncology centers, demonstrate worse survival, and this effect is most pronounced among Black children, suggesting an interaction between race and place of residence (Holmes et al., 2024). Importantly, evidence from comprehensive pediatric cancer centers indicates that racial survival

disparities for pediatric renal tumors, including RCC, may be attenuated when children receive standardized, multidisciplinary care with consistent access to advanced imaging, surgery, and long-term follow-up (Neuzil et al., 2020). Imaging guidelines for pediatric RCC should therefore be applied with awareness that children from historically underserved communities may face barriers to timely diagnostic and surveillance imaging. Proactive care coordination and attention to social risk factors are essential to promote equitable staging, treatment planning, and outcome assessment for pediatric patients with RCC (Holmes et al., 2024; Neuzil et al., 2020).

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Clear Cell Sarcoma of the Kidney (CCSK) (PEDONC-7.5)

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v1.0.2027

Criteria

Be careful not to confuse the diagnosis with clear cell RCC. See: Renal Cell Cancer (RCC) (ONC-17) for imaging guidelines.

Indication	Medically Necessary Imaging Study
Initial Staging for individuals with CCSK	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ Doppler ultrasound to evaluate for tumor thrombus is not considered medically necessary unless CT findings are inconclusive • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ Should be completed prior to anesthesia exposure if possible • Bone scan (see: **PEDONC-1.3** for coding) • MRI Brain without and with contrast (CPT® 70553) • PET is not routinely considered medically necessary in the initial staging of any pediatric renal tumor
Bilateral renal lesions are noted on ultrasound or CT in initial staging for individuals with CCSK	• MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)
Treatment response ~every 2 cycles during treatment and at the end of planned therapy for individuals with CCSK	• CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) • CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) • Additionally, for individuals with CNS metastases at initial staging: ◦ MRI Brain without and with contrast (CPT® 70553) Bone scan (see: **PEDONC-1.3** for coding) at the end of planned therapy

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Indication	Medically Necessary Imaging Study
Rare circumstances to establish the presence of active disease only when a major therapeutic decision depends on PET avidity for individuals with CCSK	• PET/CT (CPT® 78815) • Indications and coding for rare circumstances where PET/MRI is considered medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**
Surveillance for individuals with CCSK	• Every 3 months for 2 years after completion of all therapy: ◦ CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) • Every 6 months for 3 years after completion of all therapy: ◦ MRI Brain without and with contrast (CPT® 70553) • Every 3 months for 1 year, then every 6 months for 2 years after completion of all therapy: ◦ Bone scan (see: **PEDONC-1.3** for coding) ◦ If negative at 36 months, no further advanced imaging. • Other surveillance imaging should be by Abdominal US (CPT® 76700) and chest x-ray

- Doppler ultrasound to evaluate for tumor thrombus is no longer necessary unless CT findings are inconclusive, and should not be performed if CT is already completed.

Background and Supporting Information

- CCSK typically presents as an asymptomatic abdominal mass and is initially evaluated with ultrasound, followed by cross-sectional imaging (NCCN, 2025).

Evidence Discussion

- CCSK demonstrates a higher propensity for osseous metastases than Wilms tumor, necessitating targeted staging imaging rather than generalized surveillance (NCCN, 2025).

- Imaging approaches focused on staging and treatment response assessment support early detection of clinically significant disease while limiting excessive surveillance imaging (NCCN, 2025).

Health Equity Considerations

CCSK is a rare pediatric renal malignancy that disproportionately affects very young children, particularly boys under the age of four, and often presents with aggressive clinical features that necessitate timely diagnosis and specialized care (Gao et al., 2021). Although population-based data suggest that the overall incidence of CCSK is low and relatively stable over time, its rarity poses inherent equity challenges, as children from underserved communities may experience delays in diagnosis due to limited access to advanced imaging, pediatric oncology expertise, and subspecialty pathology services required for accurate tumor identification. CCSK is frequently misdiagnosed as Wilms tumor without access to molecular or immunohistochemical confirmation, which can delay appropriate therapy and adversely affect outcomes, particularly in settings with fewer diagnostic resources (Gao et al., 2021).

While contemporary multimodal treatment has substantially improved survival for children with CCSK, outcomes remain inferior to Wilms tumor when comparisons are adjusted for disease characteristics, underscoring the aggressive nature of CCSK and the importance of consistent, protocol-driven care (Gao et al., 2021). Survival disparities may emerge when children lack access to comprehensive pediatric cancer centers capable of delivering intensive chemotherapy, specialized surgical management, and longitudinal surveillance imaging. In addition, CCSK has a predilection for hematogenous spread, particularly to bone and, at relapse, the brain, which increases reliance on high-quality staging and follow-up imaging that may be less accessible to populations facing socioeconomic or geographic barriers (Gao et al., 2021). Incorporation of health equity considerations is essential to support early detection of metastatic disease and recurrence and to ensure that children with CCSK receive equitable access to staging, surveillance, and survivorship care regardless of social or environmental context.

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Malignant Rhabdoid Tumor of the Kidney (MRT) and Other Extracranial Sites (PEDONC-7.6)

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Criteria

- Be careful not to confuse the diagnosis with rhabdomyosarcoma. See: **Rhabdomyosarcoma (RMS) (PEDONC-8.2)** for Imaging Guidelines
- Primary CNS rhabdoid malignancies should be imaged according to **Atypical Teratoid/Rhabdoid Tumors (ATRT) PEDONC-4.5)**
- MRT is a highly aggressive histologic variant that can also occur in other locations and all non-CNS sites should follow these guidelines.

Indication	Medically Necessary Imaging Study
Initial Staging for individuals with MRT of the kidney and other extracranial sites	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ Doppler ultrasound to evaluate for tumor thrombus is not considered medically necessary unless CT findings are inconclusive • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ Should be completed prior to anesthesia exposure if possible • Bone scan (see: **PEDONC-1.3** for coding) • MRI Brain without and with contrast (CPT® 70553) • PET is not routinely considered medically necessary in the initial staging of any pediatric renal tumor
Bilateral renal lesions are noted on ultrasound or CT in initial staging for individuals with MRT of the kidney and other extracranial sites	• MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)

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Indication	Medically Necessary Imaging Study
Treatment response ~every 2 cycles during treatment and at the end of planned therapy for individuals with MRT of the kidney and other extracranial sites	• CT Chest with (CPT® 71260) *or* CT Chest without contrast (CPT® 71250) • CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) ◦ If primary site other than kidney, perform CT with contrast or MRI without and with contrast of primary site in place of abdominal and pelvic imaging • MRI Brain without and with contrast (CPT® 70553): ◦ Every 2 cycles during treatment for individuals with CNS metastases at initial staging ◦ At the end of planned therapy for all individuals • Bone scan (see: **PEDONC-1.3** for coding) at the end of planned therapy only if positive at initial diagnosis
Rare circumstances to establish the presence of active disease only when a major therapeutic decision depends on PET avidity for individuals with MRT of the kidney and other extracranial sites	• PET/CT (CPT® 78815) • Indications and coding for rare circumstances where PET/MRI is considered medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

Indication	Medically Necessary Imaging Study
Surveillance for individuals with MRT of the kidney and other extracranial sites	• Every 3 months for 2 years after completion of all therapy: ◦ CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) • Every 3 months for 3 years after completion of all therapy: ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) ▪ If primary site other than kidney, perform CT with contrast or MRI without and with contrast of primary site in place of abdominal imaging • Every 3 months for 1 year, then every 6 months for 1 year after completion of all therapy: ◦ MRI Brain without and with contrast (CPT® 70553) • If bone scan positive at initial diagnosis: ◦ Every 3 months for 1 year, then every 6 months for 2 years: ▪ Bone scan (see: **Modality General Considerations (PEDONC-1.3)** for coding) ◦ If negative at 36 months, no further advanced imaging • The role of surveillance imaging beyond these timeframes in unclear. Abdominal US (CPT® 76700) and chest x-ray may be considered.
Continued surveillance of individuals with rhabdoid tumor predisposition syndrome	• See: **Rhabdoid Tumor Predisposition Syndrome (PEDONC-2.11)**

Background and Supporting Information

- Malignant rhabdoid tumors are rare, aggressive malignancies that may present as renal or extrarenal masses, often identified on ultrasound during evaluation of abdominal distension (NCCN, 2025).

Evidence Discussion

- Given the aggressive biology and early dissemination associated with MRT, imaging is primarily directed toward accurate staging and response assessment rather than prolonged surveillance (NCCN, 2025).
- Avoidance of routine PET imaging is supported by limited evidence of benefit over standard anatomic imaging modalities in pediatric renal tumors (NCCN, 2025).

Health Equity Considerations

MRT of the kidney and other extracranial sites represent some of the most aggressive pediatric malignancies and are diagnosed predominantly in infancy and early childhood. Population-based data demonstrate that MRTs disproportionately affect very young children, often within the first year of life, and are frequently associated with perinatal risk factors such as low birth weight, preterm delivery, and multiple gestation pregnancies (Heck et al., 2014). These associations suggest that structural and biologic vulnerabilities beginning in the prenatal and perinatal periods may intersect with social determinants of health, including access to high quality prenatal care and neonatal services. Given the rapid progression and poor prognosis of MRTs, delays in diagnosis or referral, more likely in resource limited or underserved settings, may exacerbate already poor outcomes for affected children (Heck et al., 2014).

Health inequities may also arise from variability in access to specialized pediatric oncology centers capable of providing timely diagnostic imaging, molecular testing for SMARCB1 loss, and intensive multimodal therapy. Outcomes for extracranial MRT remain poor globally, with survival strongly associated with younger age at diagnosis and advanced stage, both of which necessitate early and comprehensive staging and surveillance imaging (Shan et al., 2021). Differences in treatment completion rates, availability of standardized protocols, and access to advanced therapies such as radiation or investigational targeted agents may further contribute to outcome disparities, particularly in low resource or geographically isolated settings (Shan et al., 2021). Providers should emphasize the need for prompt, comprehensive imaging across disease sites and ongoing surveillance to support timely intervention and reduce disparities in survival for this highly vulnerable pediatric population (Heck et al., 2014; Shan et al., 2021).

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Congenital Mesoblastic Nephroma (CMN) (PEDONC-7.7)

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Criteria

Initial Staging

Indication	Medically Necessary Imaging Study
All individuals with CMN	CT Abdomen and Pelvis with contrast (CPT® 74177)
Inconclusive findings on chest x-ray in individuals with CMN	CT Chest with contrast (CPT® 71260)

Treatment Response

Indication	Medically Necessary Imaging Study
Individuals with CMN receiving preoperative chemotherapy every 2 cycles of therapy until surgery	CT Abdomen and Pelvis with contrast (CPT® 74177)
Baseline imaging following resection in individuals with CMN	CT Abdomen and Pelvis with contrast (CPT® 74177)
Individuals with a complete resection (i.e., no residual disease on post-resection imaging)	See: Surveillance Imaging

Surveillance Imaging

Indication	Medically Necessary Imaging Study
All individuals with CMN	Abdominal ultrasound (CPT® 76700) is the preferred surveillance imaging modality to avoid radiation and anesthesia exposures

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Indication	Medically Necessary Imaging Study
Individuals with CMN with residual abnormalities present on post-operative imaging or inconclusive findings on ultrasound	CT Abdomen and Pelvis with contrast (CPT® 74177) **or** MRI Abdomen and Pelvis without and with contrast (CPT® 74183 or 72197) every 3 months for 1 year after completion of all therapy

Background and Supporting Information

- CMN is the most common renal tumor in neonates and young infants and frequently presents at birth or on prenatal ultrasound (NCCN, 2026).
- Complete surgical resection is curative in most cases, and bilateral disease or histologically confirmed metastatic disease has not been reported (NCCN, 2026).

Evidence Discussion

- Recurrence after complete resection is rare, with most events occurring within the first year following diagnosis, supporting limited surveillance imaging (NCCN, 2026).
- Restricting follow-up imaging to symptom-directed evaluation minimizes radiation exposure in infants while maintaining excellent oncologic outcomes (NCCN, 2026).

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Pediatric Soft Tissue Sarcomas (PEDONC-8)

Guideline Page

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Pediatric Soft Tissue Sarcomas – General Considerations (PEDONC-8.1)

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Criteria

- Unless specified below, individuals under 18 years of age should be imaged according to this guideline section. Exceptions include:
  - Rhabdomyosarcoma in individuals (except uterine rhabdomyosarcoma) of all ages should be imaged according to guidelines in **Rhabdomyosarcoma (RMS) (PEDONC-8.2)**
  - Uterine rhabdomyosarcoma in individuals of all ages should be imaged according to guidelines in Uterine Cancer (ONC-22). See: **Uterine Cancer - General Considerations (ONC-22.0)** in the Oncology Imaging Guidelines
  - Kaposi's sarcoma in individuals of all ages should be imaged according to guidelines in **Kaposi's Sarcoma (ONC-31.10)** in the Oncology Imaging Guidelines
- Evaluation of soft tissue masses of uncertain nature prior to biopsy should follow general imaging guidelines:
  - Individuals 0 to 17 years of age, see: **Soft Tissue and Bone Masses (PEDMS-3)** in the Pediatric Musculoskeletal Imaging Guidelines
  - Individuals 18 years of age or older, see: **Bone Lesion (MS-10.2)** in the Musculoskeletal Imaging Guidelines
- Where there are indications for PET/CT in these guidelines, note that indications and coding for rare circumstances where PET/MRI is considered medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

Background and Supporting Information

- Pediatric soft tissue sarcomas are a heterogeneous group of malignant mesenchymal tumors that differ from adult soft tissue sarcomas in histologic distribution, biologic behavior, metastatic patterns, and treatment approaches (Acord et al., 2023; NCCN, 2026).
- Rhabdomyosarcoma (RMS) represents the most common pediatric soft tissue sarcoma, while non-rhabdomyosarcoma soft tissue sarcomas (NRSTS) comprise a diverse group of less common histologies (Acord et al., 2023).
- The lungs are the most common site of metastatic disease across pediatric soft tissue sarcomas, supporting routine chest imaging for initial staging and surveillance (Expert Panel on Musculoskeletal Imaging, 2022; NCCN, 2026).

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Evidence Discussion

- Imaging is central to diagnosis, staging, treatment planning, and surveillance, with modality selection dependent on tumor location, histology, and risk of metastatic disease (Acord et al., 2023; Expert Panel on Musculoskeletal Imaging, 2022).
- MRI with and without contrast is the preferred modality for local evaluation of most pediatric soft tissue sarcomas due to superior soft-tissue contrast and lack of ionizing radiation (Acord et al., 2023; Expert Panel on Musculoskeletal Imaging, 2022).
- Use of MRI for primary tumor evaluation improves assessment of local tumor extent, neurovascular involvement, and surgical planning while minimizing radiation exposure in children (Acord et al., 2023).
- Chest CT has greater sensitivity than chest radiography for detection of pulmonary metastases and directly impacts staging and treatment decisions in pediatric sarcoma (Expert Panel on Musculoskeletal Imaging, 2022).
- Routine use of extensive whole-body or abdominal CT imaging in asymptomatic children with soft tissue sarcoma has not demonstrated improved outcomes and increases cumulative radiation exposure (Acord et al., 2023; Expert Panel on Musculoskeletal Imaging, 2022).
- Restricting advanced imaging to scenarios with higher diagnostic yield balances early detection of clinically relevant disease with reduction of unnecessary radiation, false positives, and downstream interventions (Expert Panel on Musculoskeletal Imaging, 2022).

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Rhabdomyosarcoma (RMS) (PEDONC-8.2)

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Criteria

- Individuals of all ages are imaged according to this guideline, with exception of uterine rhabdomyosarcoma, which is imaged according to Uterine Cancer (ONC-22). See: **Uterine Cancer – General Considerations (ONC-22.0)** in the Oncology Imaging Guidelines.

Indication	Medically Necessary Imaging Study
Initial staging for all individuals with RMS	ALL of the following: • MRI without and with contrast of primary site ◦ CT with contrast if MRI is contraindicated • CT Chest with contrast or CT Chest without contrast ◦ should be completed prior to anesthesia exposure if possible • Whole-Body PET/CT (CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) ◦ Bone scan substituted for PET, if PET not available
Additional initial staging for individuals with RMS with ANY of the following: • evaluation of inconclusive PET findings in the abdomen or pelvis • primary site of abdomen or pelvis • lower extremity primary site	• CT Abdomen and Pelvis with contrast (CPT® 74177)

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Indication	Medically Necessary Imaging Study
Additional initial staging for individuals with RMS with ANY of the following: • Primary site involving the paraspinal or paravertebral region • PET or bone scan-avid lesions in skull, neck, vertebrae • Neurologic signs or symptoms raising suspicion of CNS metastases	ALL of the following: • MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Treatment response, every 2 cycles during treatment, prior to local control surgery, and at the end of planned therapy in individuals with RMS	ALL of the following: • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) • MRI without and with contrast of primary site ◦ CT if used at initial imaging for MRI contraindication • CT with contrast or MRI without and with contrast of all known metastatic sites using the same conventional imaging modality as per initial staging
ANY of the following in individuals with RMS: • Response assessment prior to local control surgery or radiation therapy • Evaluation of residual mass visible on conventional imaging as part of end of therapy evaluation • Response assessment of disease visible on PET but not conventional imaging • PET results are likely to result in a treatment change for the individual, including a change from active treatment to surveillance.	• Whole-body PET/CT (CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) ◦ Once PET has been documented to be negative for a given individual's cancer or all PET-avid disease has been surgically resected, PET is not considered medically necessary for continued disease monitoring or surveillance unless one of the exceptions in section **General Criteria (PEDONC-1.0)** applies.

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Indication	Medically Necessary Imaging Study
Restaging following local control surgery in individuals with RMS	ALL of the following: • MRI without and with contrast of primary site ◦ CT if used at initial imaging for MRI contraindication • CT with contrast or MRI without and with contrast of all known metastatic sites using the same conventional imaging modality as per initial staging
Surveillance in individuals with RMS	ALL of the following. every 3 months for 3 years, then every 6 months in years 4-5, then annually: • MRI without or without and with contrast of primary tumor site ◦ CT if MRI contraindicated • Chest x-ray ◦ CT Chest with (CPT® 71260) **or** CT Chest without contrast (CPT® 71250) for new or worsening clinical symptoms of chest disease or new findings on chest x-ray
Suspected recurrence in individuals with RMS	• Repeat conventional imaging as per initial staging for all individuals
ONE of the following in individuals with RMS: • Biopsy-proven recurrence • Conventional imaging (CT, MRI, US, plain film) reveals findings that are inconclusive or suspicious for recurrence, and PET avidity will determine whether biopsy or continued observation is appropriate • Rare circumstances where obvious clinical symptoms show strong evidence suggesting recurrence, and PET would replace conventional imaging modalities.	• Whole-body PET/CT (CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498)

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Background and Supporting Information

- RMS accounts for approximately 60% of pediatric soft tissue sarcomas and demonstrates distinct embryonal and alveolar subtypes with differing metastatic behavior and risk stratification (Acord et al., 2023).
- RMS can arise from skeletal muscle precursors anywhere in the body, resulting in wide variability in presenting symptoms and primary tumor locations (Acord et al., 2023; NCCN, 2026).

Evidence Discussion

- MRI without and with contrast is preferred for primary site imaging in most anatomic locations, including the head and neck, extremities, pelvis, and chest wall (Acord et al., 2023).
- CT with contrast is preferred for RMS arising within the thoracic cavity (excluding the chest wall) due to the need for detailed evaluation of intrathoracic structures (Acord et al., 2023).
- FDG-PET imaging is used in initial staging to evaluate nodal and osseous metastatic disease, which may be occult on conventional imaging (Acord et al., 2023; NCCN, 2026).
- FDG-PET imaging demonstrates higher sensitivity for detection of nodal and skeletal metastases in pediatric RMS compared with conventional staging methods and can alter initial staging and management (Acord et al., 2023).
- PET imaging has not shown consistent benefit for assessing treatment response in RMS, as changes in metabolic activity do not reliably correlate with prognosis or outcome (Acord et al., 2023).
- Persistent FDG uptake in residual masses at the end of therapy is common and does not necessarily indicate viable tumor, limiting the value of PET for routine surveillance (Acord et al., 2023).
- Avoidance of routine CT Abdomen and Pelvis imaging in initial RMS staging is supported by low incidence of clinically relevant abdominal metastases and helps reduce unnecessary radiation exposure (Acord et al., 2023).
- Restricting PET use during treatment and surveillance reduces false-positive findings, radiation exposure, and downstream testing without compromising detection of clinically meaningful recurrence (Acord et al., 2023).

Health Equity Considerations

RMS is the most common soft tissue sarcoma of childhood and adolescence, with incidence and outcomes influenced by biologic, sociodemographic, and health system factors. Population-based analyses demonstrate that while overall RMS incidence is relatively consistent across racial and ethnic groups, important disparities emerge in disease presentation and survival. National registry data show that non-Hispanic

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Black children have significantly worse survival compared with non Hispanic White children, even after adjustment for age, stage, histology, and socioeconomic status, suggesting that inequities in access to timely diagnosis, comprehensive staging, and high quality treatment may contribute to outcome differences (McEvoy et al., 2023). These findings contrast with results from Children's Oncology Group clinical trial cohorts, where non-Hispanic Black and Hispanic patients often present with higher risk features but experience comparable survival when treated on standardized protocols, underscoring the potential equity enhancing effect of protocol driven care (Munnikhuysen et al., 2023).

Disparities in RMS outcomes may also reflect differences in early recognition, diagnostic imaging access, and referral patterns. Higher risk features at presentation, including invasive tumors, alveolar histology, and metastatic disease, are more common among Hispanic and non-Hispanic Black patients, raising concern for delays in diagnosis that may be influenced by structural barriers, limited access to pediatric subspecialists, or implicit bias within health systems (Munnikhuysen et al., 2023). Socioeconomic factors further intersect with race and geography; population-based studies demonstrate trends toward poorer survival among children living in lower income counties, highlighting the importance of equitable access to imaging, multidisciplinary evaluation, and longitudinal surveillance regardless of insurance status or community resources (McEvoy et al., 2023).

Broader epidemiologic and etiologic data also emphasize that RMS disproportionately affects very young children and may be influenced by prenatal, perinatal, and early life factors. Associations with birth characteristics, parental exposures, and genetic cancer predisposition syndromes suggest that underlying vulnerability may be compounded in populations facing disparities in prenatal care, environmental exposures, and access to genetic evaluation (Martin Giacalone et al., 2021). Explicit attention should be paid to health equity, promoting consistent staging, response assessment, and follow-up imaging across care settings. Ensuring timely, guideline-concordant imaging for all children with suspected or confirmed RMS may help reduce disparities in stage at diagnosis and outcomes, particularly for patients from historically underserved or underrepresented populations (Martin Giacalone et al., 2021; McEvoy et al., 2023; Munnikhuysen et al., 2023).

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Non-Rhabdomyosarcoma Soft Tissue Sarcomas (NRSTS) (PEDONC-8.3)

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Criteria

Indication	Medically Necessary Imaging Study
Initial staging of all individuals with NRSTS	ALL of the following: • MRI without and with contrast of primary site ◦ CT with contrast if contraindication to MRI • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) ◦ Should be completed prior to anesthesia if possible • For primary tumor of the chest wall, both MRI Chest wall without and with contrast (CPT® 71552) and CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250) to assess for pulmonary metastatic disease (indicated simultaneously) • PET/CT (CPT® 78815) ◦ Whole-body PET/CT (CPT® 78816) if there is clinical suspicion of skull or distal lower extremity involvement • Nuclear bone scan (see: **PEDONC-1.3** for coding) if PET is not available

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Indication	Medically Necessary Imaging Study
ANY of the following in individuals with NRSTS: • Additional initial staging for individuals with disease in the abdomen, pelvis, or lower extremities • Angiosarcoma, alveolar soft part sarcoma, clear cell sarcoma, epithelioid sarcoma, hemangiopericytoma, leiomyosarcoma, liposarcoma, retroperitoneal or intra-abdominal primary site (including pelvic primary site), other histologies documented to have propensity for lymphatic spread and deep-seated tumors • Inconclusive findings in the abdomen or pelvis on PET/CT	In addition to the above, one of the following combinations: • CT Abdomen and Pelvis with contrast (CPT® 74177) if not already performed • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast • MRI Abdomen (CPT® 74183) with and without contrast and CT Pelvis (CPT® 72193) with contrast • CT Abdomen (CPT® 74160) with contrast and MRI Pelvis (CPT® 72197) without and with contrast
Further evaluation of ANY of the following in individuals with NRSTS: • PET or nuclear bone scan-avid lesions in skull, neck, or vertebrae seen on initial imaging • Neurologic signs or symptoms raising suspicion of CNS metastases • Myxoid round cell liposarcoma, angiosarcoma, alveolar soft part sarcoma, cardiac sarcoma • Additional staging for individuals with primary site arising in the paraspinal or paravertebral region	ALL of the following: • MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
Treatment response, every 2 cycles of treatment, prior to local control, and at the end of planned therapy in individuals with NRSTS	Either of the following: • CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250), and • MRI without and with contrast of primary site ◦ CT if contraindication to MRI and • CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/pelvic imaging for initial staging • Imaging of all known metastatic sites using the same conventional imaging modality as per initial staging OR • PET/CT (CPT® 78815) if disease visible on initial staging PET but not on conventional imaging ◦ Whole-body PET/CT (CPT® 78816) if there is clinical suspicion of skull or distal lower extremity involvement
ANY of the following, if PET positive at initial diagnosis in individuals with NRSTS: • Assessment prior to local control surgery or radiation therapy • Evaluation of residual mass visible on conventional imaging as part of end of therapy evaluation • Rare circumstances when PET results are likely to result in a treatment change for the individual, including a change from active treatment to surveillance	• PET/CT (78815) ◦ Whole-body PET/CT (78816) if there is clinical suspicion of skull or distal lower extremity involvement ◦ Once PET has been documented to be negative for a given individual's cancer or all PET-avid disease has been surgically resected, PET is not considered medically necessary for continued disease monitoring or surveillance unless one of the exceptions in section **General Criteria (PEDONC-1.0)** applies.

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Indication	Medically Necessary Imaging Study
Restaging following local control surgery in individuals with NRSTS	ALL of the following: • MRI without and with contrast of the primary site ◦ CT if contraindication to MRI • Imaging of all known metastatic sites using the same conventional imaging modality as per initial staging • CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/pelvic imaging for initial staging
Surveillance, low grade, localized NRSTS of extremity or trunk treated with resection (with or without radiation) or chemotherapy in individuals with NRSTS	Every 3 months for the first 12 months, then every 4 months in years 2 and 3, and every 6 months in years 4 and 5, then annually thereafter: • MRI without and with contrast of the primary site • CT if MRI contraindicated AND Every 6 months for 3 years, then annually thereafter: • CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/pelvic imaging for initial staging • CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250)

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Indication	Medically Necessary Imaging Study
Surveillance for all other NRSTS	Every 3 months for the first 2 years, then every 6 months in years 3 and 4, and then annually: • MRI without and with contrast of the primary site ◦ CT if MRI contraindicated ◦ Nuclear bone scan (see: **PEDONC-1.3** for coding) for surveillance of known bony metastases • CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250) • CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/pelvic imaging for initial staging
ANY of the following in individuals with NRSTS: • Angiosarcoma • Alveolar soft part sarcoma • Cardiac sarcoma	• In addition to the above studies: ◦ MRI Brain without and with contrast (CPT® 70553) annually For surveillance of individuals with known brain metastases, see: **Brain Metastases (ONC-31.3)**
Surveillance for recurrent NRSTS	ALL of the following after successful treatment for recurrent disease, every 3 months for 1 year (surveillance after year 1 should follow the standard timing listed in the appropriate surveillance section above): • CT Chest (CPT® 71260) with contrast • CT with contrast or MRI without and with contrast of the primary site • CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/pelvic imaging for initial staging

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Indication	Medically Necessary Imaging Study
ANY of the following in individuals with NRSTS: • Conventional imaging (CT, MRI, US, plain film) during surveillance reveals findings that are inconclusive or suspicious for recurrence, and PET avidity will determine whether biopsy or continued observation is appropriate • Obvious clinical symptoms show strong evidence suggesting recurrent and PET would replace conventional imaging modalities	• PET/CT (CPT® 78815) ◦ Whole-body PET/CT (CPT® 78816) if there is clinical suspicion of skull or distal lower extremity involvement

Background and Supporting Information

- NRSTS include all pediatric soft tissue sarcomas other than RMS and encompass a wide range of histologic subtypes with variable aggressiveness and metastatic potential (Acord et al., 2023; NCCN, 2026).
- Because NRSTS can arise from any soft tissue, presenting symptoms and primary tumor locations vary widely and influence imaging selection (Acord et al., 2023).
- Many pediatric patients with localized NRSTS are treated with surgical resection alone and transition directly into surveillance imaging (Acord et al., 2023).
- ue to limited pediatric-specific surveillance data, imaging recommendations incorporate pediatric literature and NCCN guidance for similar histologies (NCCN, 2026).

Evidence Discussion

- Extrapulmonary metastatic disease is uncommon at initial presentation in most NRSTS, supporting omission of routine CT Abdomen and Pelvis imaging in asymptomatic pediatric patients (Acord et al., 2023; Expert Panel on Musculoskeletal Imaging, 2022).
- MRI with and without contrast remains the preferred modality for evaluation and surveillance of the primary tumor site (Acord et al., 2023; Expert Panel on Musculoskeletal Imaging, 2022).
- FDG-PET imaging has not demonstrated consistent benefit for treatment response assessment or routine surveillance in NRSTS and may increase false-positive findings (Acord et al., 2023).

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- Stable residual masses without interval growth on conventional imaging do not justify PET imaging, even when residual FDG avidity is present at therapy completion (Acord et al., 2023).
- Surveillance strategies emphasizing MRI of the primary site and chest imaging appropriately balance early detection of recurrence with minimization of radiation exposure and unnecessary testing (Expert Panel on Musculoskeletal Imaging, 2022; NCCN, 2026).

Health Equity Considerations

NRSTS comprise a heterogeneous group of rare malignancies affecting children, adolescents, and young adults, with outcomes influenced not only by tumor biology and stage but also by socioeconomic factors. Population-based evidence demonstrates that insurance status, a proxy for income and access to care, is strongly associated with survival in pediatric and young adult patients with bone and soft tissue sarcomas, including NRSTS. Patients with low income public insurance or no insurance experience significantly worse overall survival compared with privately insured patients, a disparity that persists across localized, regional, and metastatic disease stages (Penumarthy et al., 2020). Importantly, low income patients are nearly twice as likely to present with metastatic disease at diagnosis, indicating that structural barriers such as delays in evaluation, limited access to specialized care, and reduced availability of timely diagnostic imaging may contribute to more advanced disease at presentation (Penumarthy et al., 2020).

Survival disparities associated with insurance status are only partially explained by stage at diagnosis, suggesting that inequities extend beyond initial presentation to include differences in treatment delivery, adherence, and follow-up. Patients with public insurance often face challenges related to transportation, health literacy, financial toxicity, and access to multidisciplinary sarcoma centers, all of which may affect receipt of optimal local control, surveillance imaging, and management of treatment-related complications (Penumarthy et al., 2020). Adolescents and young adults represent a particularly vulnerable subgroup, with inferior outcomes that may be exacerbated by gaps in insurance coverage and transitions between pediatric and adult health systems. There should be an explicit focus on health equity, supporting consistent staging, response assessment, and surveillance regardless of insurance status or socioeconomic background. Ensuring equitable access to high-quality imaging across disease stages may help mitigate delays in diagnosis and reduce outcome disparities for historically underserved pediatric and young adult populations with NRSTS (Penumarthy et al., 2020).

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Bone Tumors (PEDONC-9)

Guideline Page

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Bone Tumors – General Considerations (PEDONC-9.1)

ONCP.BT.0009.1.C
v1.0.2027

Criteria

These guidelines include both benign and malignant lesions.

- Unless specified below, individuals who are <18 years old should be imaged according to this guideline section. Exceptions include:
  - Osteogenic sarcoma in individuals of all ages should be imaged according to guidelines in **Osteogenic Sarcoma (OS) (PEDONC-9.3)**
  - Ewing Sarcoma and Primitive Neuroectodermal Tumor in individuals of all ages should be imaged according to guidelines in **Ewing Sarcoma and Primitive Neuroectodermal Tumors (ESFT) (PEDONC-9.4)**
  - Chondrosarcoma in individuals of all ages should be imaged according to guidelines in **Bone Sarcomas – Initial Work-up/Staging (ONC-12.6)**
  - Chordoma in individuals of all ages should be imaged according to guidelines in **Bone Sarcomas – Initial Work-up/Staging (ONC-12.6)**
  - Giant cell tumor of bone and enchondroma in individuals of all ages should be imaged according to guidelines in **Benign Bone Tumors – General Considerations (ONC-12.9)**
  - Other benign bone tumor individuals of all ages should be imaged according to guidelines in **Benign Bone Tumors (PEDONC-9.2)**
- Prosthetic devices for children after surgery for bony tumors are nearly all customized.
  - CT, contrast as requested in alignment with prosthetic manufacturer specifications, is considered medically necessary when requested by the operating surgeon for planning for customized-to-individual joint replacement or prosthetic surgery
- All bone tumors should be evaluated by plain x-ray prior to any advanced imaging
- Where indications for PET/CT are noted, please note that indications and coding for rare circumstances where PET/MRI is considered medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**
- CT Chest is superior to PET/CT for the detection of pulmonary metastases, and is considered medically necessary in the initial workup of all suspected malignant bone tumors.
  - CT Chest should be completed prior to anesthesia exposure, if possible.

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Background and Supporting Information

- Pediatric bone tumors encompass a spectrum of benign and malignant neoplasms, with osteosarcoma and Ewing sarcoma representing the most common primary malignant bone tumors in children and adolescents (NCCN, 2026).

Evidence Discussion

- Chest imaging is essential in malignant pediatric bone tumors, as the lungs are the most common site of metastatic disease (Expert Panel on Musculoskeletal Imaging, 2022; NCCN, 2026).
- Plain radiography is the cornerstone of initial evaluation for suspected bone tumors and frequently provides sufficient information to stratify lesions as benign, indeterminate, or aggressive (Expert Panel on Musculoskeletal Imaging, 2020).
- Initial evaluation with plain radiographs allows accurate biologic stratification in the majority of bone lesions and minimizes unnecessary advanced imaging and radiation exposure in children (Expert Panel on Musculoskeletal Imaging, 2020).
- MRI with and without contrast is the preferred modality for local staging of malignant bone tumors, providing superior assessment of intramedullary extent, soft-tissue involvement, and neurovascular relationships (Expert Panel on Musculoskeletal Imaging, 2020; NCCN, 2026).
- MRI enhances surgical planning and local staging accuracy while avoiding ionizing radiation, supporting its role as the preferred cross-sectional modality in pediatric bone malignancies (Expert Panel on Musculoskeletal Imaging, 2020; NCCN, 2026).
- Limiting advanced imaging to lesions with aggressive or indeterminate radiographic features balances diagnostic benefit with reduced radiation burden and cost (Expert Panel on Musculoskeletal Imaging, 2020).

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Benign Bone Tumors (PEDONC-9.2)

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Criteria

- Osteochondroma

Indication	Medically Necessary Imaging Study
Preoperative planning for individuals with osteochondroma	• MRI without and with contrast after evaluation by the operating surgeon • See: **General Considerations (PEDONC-9.1)** for requests related to prosthetic planning
• ANY of the following for individuals with osteochondroma: ◦ Concern for malignant transformation ◦ New or worsening pain/symptoms ◦ Change on a recent plain x-ray	• MRI without contrast or without and with contrast

- Osteoid osteoma

Indication	Medically Necessary Imaging Study
Suspected osteoid osteoma, based on clinical history and plain film findings	• CT without contrast
• ANY of the following in individuals wih osteois osteoma: ◦ CTs are not characteristic for diagnosis ◦ Individual has bone pain not localized to the area of findings on CT or x-ray	• ONE of the following: ◦ Triple phase bone scan (CPT® 78315) ◦ SPECT (CPT® 78803) ◦ Hybrid SPECT/CT (CPT® 78830)
• Individuals with osteoid osteoma with new pain who have been previously treated with radiofrequency ablation or curettage	• Bone scan (CPT® code 78830, 78315, or 78803 - as requested)

- Other benign tumors

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- Refer to:

Background and Supporting Information

- Many benign bone tumors demonstrate characteristic appearances on plain radiographs, allowing confident diagnosis without additional imaging (Expert Panel on Musculoskeletal Imaging, 2020).
- Osteochondroma is the most common benign bone tumor in pediatrics and is typically diagnostic on radiographs alone (Expert Panel on Musculoskeletal Imaging, 2020).

Evidence Discussion

- Reliance on radiographic diagnosis for benign-appearing bone lesions avoids unnecessary advanced imaging, sedation, radiation exposure, and downstream testing in pediatric patients (Expert Panel on Musculoskeletal Imaging, 2020).
- Advanced imaging (MRI or CT) is generally reserved for atypical appearances, symptomatic lesions, or preoperative planning (Expert Panel on Musculoskeletal Imaging, 2020).
- MRI or CT provides benefit primarily when complications, malignant transformation, or surgical planning must be assessed, supporting selective rather than routine use (Expert Panel on Musculoskeletal Imaging, 2020).
- This approach maintains diagnostic accuracy while minimizing harm and aligns with established pediatric imaging stewardship principles (Expert Panel on Musculoskeletal Imaging, 2020).

Health Equity Considerations

Benign bone tumors in children are frequently identified incidentally through radiographic imaging or after presentation with pain, swelling, or pathologic fracture, and timely access to diagnostic imaging plays a central role in preventing complications and unnecessary morbidity. Although these lesions are non-malignant, delays in evaluation or limited access to advanced imaging modalities can result in progression to structural instability, deformity, or fracture, disproportionately affecting children with barriers to specialty orthopedic or radiologic care. The large institutional series by Gozar et al. (2025) illustrates that a substantial proportion of benign bone tumors are discovered incidentally or at advanced stages requiring surgical intervention, emphasizing the importance of equitable access to early imaging and follow up across pediatric populations.

Health equity concerns are particularly relevant given the reliance on imaging modalities with differing availability and cost. Plain radiography is typically the first-line diagnostic tool and is widely accessible, while CT and MRI, often necessary for lesion characterization, surgical planning, or exclusion of malignancy, may be less accessible

in low resource or underserved settings due to cost, geographic limitations, or radiation concerns in children. Disparities in access to MRI may contribute to variability in diagnostic certainty, biopsy decisions, and surgical planning, potentially leading to either overtreatment or delayed intervention for children from underserved communities (Gozar et al., 2025). Ensuring guideline-concordant use of imaging that balances diagnostic accuracy with resource stewardship is essential to minimizing inequities in care.

Additionally, benign bone tumors that cause pain, fractures, or functional limitation often require longitudinal surveillance or surgical management, which may be disrupted by socioeconomic barriers such as transportation challenges, limited health literacy, or inconsistent follow-up. The observation that pathologic fractures were associated with longer hospital stays highlights the downstream impact of delayed detection and underscores the importance of early, equitable imaging evaluation to prevent avoidable complications (Gozar et al., 2025). Imaging guidelines for benign bone tumors should therefore be applied with an equity lens, promoting timely access to appropriate imaging, consistent follow up, and multidisciplinary care coordination to reduce disparities in outcomes for all pediatric patients.

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Osteogenic Sarcoma (OS) (PEDONC-9.3)

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Criteria

Indication	Medically Necessary Imaging Study
Suspected or Biopsy Proven Diagnosis of OS	• MRI without and with contrast of the primary site OR • CT, contrast as request, of the primary site if there is a contraindication to MRI AND • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Chest is considered medically necessary in initial workup of all suspected malignant bone tumors in children and should be completed prior to anesthesia exposure if possible • These studies are considered medically necessary even when PET/CT has already been authorized or performed

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Indication	Medically Necessary Imaging Study
Initial Staging of Biopsy Proven OS Disease	• 18F-FDG PET/CT whole-body (CPT® 78816) ◦ Nuclear bone scan (see: **(PEDONC-1.3)** for coding) if PET not available ◦ PET/CT is considered medically necessary in addition to the conventional imaging listed in the suspected/diagnosis sections • CT Abdomen and Pelvis with contrast (CPT® 74177) is not routinely considered medically necessary in the initial metastatic staging of pediatric OS but is considered medically necessary in the following situations: ◦ Evaluation of inconclusive PET findings ◦ Primary site of abdomen or pelvis
Suspected bony metastatic sites noted on PET or bone scan in individuals with OS	• MRI without and with contrast of the suspected metastatic site OR • CT with contrast of the suspected metastatic site
Restaging after 10 to 12 weeks of neoadjuvant chemotherapy prior to local control surgery in individuals with OS	• MRI without and with contrast of primary site • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) • 18F-FDG PET/CT whole-body (CPT® 78816) or bone scan (see: **(PEDONC-1.3)** for coding)

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Indication	Medically Necessary Imaging Study
Individuals with OS with metastatic disease undergoing current chemotherapy	• CT Chest with (CPT® 71260)or without contrast (CPT® 71250) can be performed every 2 cycles during treatment and at the end of planned chemotherapy • MRI without and with contrast of primary site can be performed every 2 cycles during treatment and at the end of planned chemotherapy • If previously positive for bony metastases, 18F-FDG PET/CT whole body (CPT® 78816) or bone scan (see: **(PEDONC-1.3)** for coding) every 2 cycles during treatment and at the end of planned chemotherapy • Imaging may be considered medically necessary more frequently around the time of surgical resection of primary or metastatic lesions to assess for resectability
Preoperative planning for local control surgery in individuals with OS	• CT, contrast as requested • MRA and/or CTA may rarely be considered medically necessary for complicated surgical resections, and is considered medically necessary after evaluation by the operating surgeon to clarify inconclusive MRI findings for preoperative planning • See: **General Considerations (PEDONC-9.1)** for advancing imaging requests related to prosthetic planning

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Indication	Medically Necessary Imaging Study
Following local control surgery, until the end of planned adjuvant chemotherapy in individuals with OS	• MRI without and with contrast of primary site ~6 weeks after surgical procedure, and at the end of planned chemotherapy • Plain x-rays of the primary site and chest immediately after local control and then every 2 months between the medically necessary, listed advanced imaging studies. • CT Chest with (CPT® 71260) or without contrast (CPT® 71250): ◦ Measurable pulmonary metastases: every 6 weeks, and at the end of planned chemotherapy ◦ No measurable pulmonary metastases: every 4 months, and at the end of planned chemotherapy • Bone scan (see: **(PEDONC-1.3)** for coding) every 4 months, and at the end of planned chemotherapy ◦ 18F-FDG PET/CT whole-body (CPT® 78816) in place of bone scan, if positive for distant bone metastases at initial diagnosis

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Indication	Medically Necessary Imaging Study
Recurrent metastatic or recurrent unresectable disease on treatment in individuals with OS	• The following every 2 cycles of treatment, and at the end of planned chemotherapy: ◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ MRI without and with contrast of primary site ◦ PET is generally not considered medically necessary during active treatment for recurrent pediatric cancer. In rare circumstances, 18F-FDG PET/CT imaging with CPT® 78816 may be considered medically necessary when results are likely to result in a treatment change for the individual, including a change from active treatment to surveillance.
Surveillance (all individuals regardless of primary site in individuals with OS	• Any or all of the following every 3 months for year 1 and 2, then every 4 months in year 3, then every 6 months in year 4 and 5 after completion of all therapy ◦ CT or MRI of primary site as performed during suspected or initial disease workup (provided the individual does not have an endoprosthesis that will cause MRI or CT artifact): ▪ CT Chest with (CPT® 71260) or without contrast (CPT® 71250) • Bone scan (see: **(PEDONC-1.3)** for coding) every 3 months for 1 year, then every 4 months for 1 year, then every 6 months for 1 year, then annually for 2 years after completion of all therapy

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Indication	Medically Necessary Imaging Study
Suspected recurrence in individuals with OS	• MRI without and with contrast of the primary site and /or site of suspected recurrence based on symptoms or other imaging OR • CT, contrast as request, of the primary site and/or site of suspected recurrence based on symptoms or other imaging if there is a contraindication to MRI AND • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Chest is considered medically necessary in initial workup of all suspected malignant bone tumors in children and should be completed prior to anesthesia exposure if possible • CT Abdomen and Pelvis with contrast (CPT® 74177) is not routinely considered medically necessary in the metastatic staging of pediatric OS, but is considered medically necessary in the following situations: ◦ Evaluation of inconclusive PET findings ◦ Primary site of abdomen or pelvis • These studies are considered medically necessary even when PET/CT has already been authorized or performed

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Indication	Medically Necessary Imaging Study
• For suspected recurrence in individuals with OS, ANY of the following: ◦ Conventional imaging reveals findings that are inconclusive or suspicious for recurrence and PET avidity will determine whether biopsy or continued observation is appropriate ◦ Rare circumstances where obvious clinical symptoms show strong evidence suggesting recurrence and PET would replace conventional imaging modalities	• 18F-FDG PET/CT (CPT® 78816)
• Biopsy-proven recurrence in individuals with OS	• PET/CT whole-body (78816) for biopsy-proven recurrence ◦ If disease is considered potentially resectable, follow osteosarcoma treatment response as previous

Background and Supporting Information

- Osteosarcoma is the most common primary malignant bone tumor in children and adolescents, typically arising in the metaphyses of long bones (NCCN, 2026).

Evidence Discussion

- MRI with and without contrast is essential for local staging, including evaluation of intramedullary tumor extent and skip lesions. MRI-based local staging improves surgical planning and limb-salvage outcomes while avoiding ionizing radiation exposure (Expert Panel on Musculoskeletal Imaging, 2020; NCCN, 2026).
- Chest CT is required for metastatic evaluation, as pulmonary metastases are the most frequent site of disease spread. Chest CT improves detection of small pulmonary metastases that influence staging and treatment decisions compared with chest radiography alone (Expert Panel on Musculoskeletal Imaging, 2022).
- FDG-PET/CT or bone scan may be used for whole-body evaluation, but findings require correlation with conventional imaging (NCCN, 2026).
- Whole-body PET/CT can identify metastatic disease but carries higher radiation exposure and potential false positives; selective use mitigates unnecessary downstream interventions (NCCN, 2026).

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- Surveillance strategies focusing on the primary site and chest balance early recurrence detection with cumulative radiation risk in long-term survivors (NCCN, 2026).

Health Equity Considerations

OS (osteosarcoma) is a rare but highly aggressive bone malignancy in children, adolescents, and young adults, and substantial evidence demonstrates that social and demographic factors influence disease presentation and care processes even when overall survival differences are attenuated after adjustment for disease severity. Population-based analyses show that Hispanic children are more likely to present with metastatic disease and larger primary tumors at diagnosis, while patients from communities with high language isolation are also at increased risk of advanced presentation, suggesting barriers to early evaluation and diagnostic imaging (Fritz et al., 2025; Goulding et al., 2024). Delays between diagnosis and initiation of treatment are more common among non Hispanic Black patients and adolescents and young adults, underscoring the role of structural barriers such as healthcare access, communication challenges, and care coordination during transitions from pediatric to adult systems (Fritz et al., 2025).

Access to high-quality imaging is a critical equity issue in OS, as accurate staging, assessment of local extent, and detection of metastases directly inform surgical planning and prognosis. Disparities in insurance coverage and county-level socioeconomic indicators have been associated with higher odds of amputation and increased mortality in population studies, particularly among publicly insured patients, even when controlling for metastatic status (Goulding et al., 2024). These findings suggest that inequitable access to specialized imaging, sarcoma-experienced multidisciplinary teams, and limb salvage-oriented care pathways may influence treatment intensity and long term outcomes. Consistent, guideline-based imaging is therefore essential to reduce variation in staging accuracy and treatment decision making across populations.

Importantly, evidence from Children's Oncology Group clinical trial cohorts demonstrates that when children with non-metastatic OS receive standardized, protocol-driven care, overall and event-free survival do not differ by race, ethnicity, or proxied poverty exposures, highlighting the capacity of equitable care delivery to mitigate disparities (Ilcisin et al., 2024). However, stark differences in post-relapse survival persist, with non-Hispanic Black children experiencing substantially worse outcomes after disease recurrence, likely reflecting inequities in access to aggressive surveillance imaging, timely relapse detection, and specialized post-relapse interventions (Ilcisin et al., 2024). Imaging guidelines for OS should therefore be applied with an explicit health equity lens, emphasizing timely, comprehensive imaging at diagnosis, during therapy, and in surveillance to support early detection, coordinated care, and equitable outcomes across diverse pediatric populations.

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Ewing Sarcoma Family of Tumors (ESFT), Including Primitive Neuroectodermal Tumors (PNET) (PEDONC-9.4)

ONCP.BT.0009.4.A
v1.0.2027

Criteria

Indication	Medically Necessary Imaging Study
Suspected/Diagnosis of ESFT or PNET – ill-defined or non-discrete soft tissue mass without bony involvement	• US (CPT® 76881 or 76882) in addition to plain x-ray
Suspected or Biopsy Proven Diagnosis of ESFT or PNET – Primary site	• MRI without and with contrast ◦ CT, contrast as requested, if there is a contraindication to MRI • MRA and/or CTA may rarely be considered medically necessary for complicated surgical resections, and is considered medically necessary after evaluation by the operating surgeon to clarify inconclusive MRI findings for preoperative planning • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Chest is considered medically necessary in initial workup of all suspected or confirmed malignant bone tumors in children and should be completed prior to anesthesia exposure if possible • These studies are considered medically necessary even when PET/CT has already been authorized or performed

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Indication	Medically Necessary Imaging Study
Suspected or Biopsy Proven Diagnosis of ESFT or PNET – Chest wall primary	• MRI Chest without and with contrast AND • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Chest is considered medically necessary in initial workup of all suspected or confirmed malignant bone tumors in children and should be completed prior to anesthesia exposure if possible • These studies are considered medically necessary even when PET/CT has already been authorized or performed
Initial Staging (additional imaging after biopsy confirmed disease) of ESFT or PNET	• PET/CT whole-body (CPT® 78816) ◦ Bone scan (see **(PEDONC-1.3)** for coding) substituted for PET imaging if PET not available ◦ PET/CT is considered medically necessary in addition to the conventional imaging listed in the suspected/diagnosis sections
• For ANY of the following in individuals with ESFT or PNET: ◦ Evaluation of inconclusive PET findings ◦ Primary site involving the abdomen or pelvis	• CT Abdomen and Pelvis with contrast (CPT® 74177)

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Indication	Medically Necessary Imaging Study
Restaging after 10 to 12 weeks of neoadjuvant chemotherapy prior to local control surgery in individuals with ESFT or PNET	Imaging modality should be the same as used for initial staging, any or all from the list below as described: • MRI without and with contrast of the primary site • CT (contrast as requested) if requested per valuation by the operating surgeon for pre-operative planning ◦ See **General Considerations (PEDONC-9.1)** for imaging requests related to prosthetic planning. • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) • ONE of the following: ◦ PET/CT whole-body (CPT® 78816) ◦ Whole-body bone scan (see **PEDONC-1.3** for coding) ◦ MRI bone marrow blood supply/diffusion-weighted MRI with ADC (CPT® 77084)
Treatment response following local control surgery in individuals with ESFT or PNET	• MRI without and with contrast of primary site 3 months after surgical procedure and at the end of planned chemotherapy • Plain x-rays of the primary site and chest immediately after local control then every 3 months • CT Chest with (CPT® 71260) or without contrast (CPT® 71250): ◦ Measurable pulmonary metastases: every 6 weeks and at the end of planned chemotherapy ◦ No measurable pulmonary metastases: every 3 months and at the end of planned chemotherapy • Whole-body bone scan (see **PEDONC-1.3** for coding) at the end of planned chemotherapy • PET/CT whole-body (CPT® 78816) for clinical or imaging findings suggestive of local recurrence

Indication	Medically Necessary Imaging Study
Individuals with metastatic disease undergoing current chemotherapy in individuals with ESFT or PNET	• CT Chest with (CPT® 71260) or without contrast (CPT® 71250) every 2 cycles during treatment and at the end of planned chemotherapy • MRI without and with contrast of primary site every 2 cycles during treatment and at the end of planned chemotherapy • If previously positive for bony metastases, PET/CT whole-body (CPT® 78816) or bone scan (see **PEDONC-1.3** for coding) every 2 cycles during treatment and at the end of planned chemotherapy • Imaging may be considered medically necessary more frequently around the time of surgical resection of primary or metastatic lesions to assess for resectability • CT with contrast of other sites of involvement
Recurrent metastatic or recurrent unresectable disease on treatment in individuals with ESFT or PNET	• Every 2 cycles of treatment, and at the end of planned chemotherapy: ◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ MRI without and with contrast of primary site ◦ PET imaging is considered medically necessary when results are likely to result in a treatment change for the individual, including a change from active treatment to surveillance.

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Indication	Medically Necessary Imaging Study
Surveillance for all disease **other than** low grade, stage I disease in individuals with ESFT or PNET	• Every 3 months for year 1 and 2, then every 4 months in year 3, then every 6 months in year 4 and 5, and annually for 5 years after completion of all therapy to complete 10 years of surveillance: ◦ CT or MRI of primary site as performed during suspected or initial disease workup (provided the individual does not have an endoprosthesis that will cause MRI or CT artifact): ◦ Bone scan (see **PEDONC-1.3** for coding) • Every 3 months for year 1 and 2, then every 4 months in year 3, then every 6 months in year 4 and 5: ◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ After year 5, CXR should be used for surveillance
Surveillance for low grade stage I disease only in individuals with ESFT or PNET	• Every 3 months for year 1 and 2, then every 4 months in year 3, then every 6 months in year 4 and 5, and annually for 5 years after completion of all therapy to complete 10 years of surveillance: ◦ CT or MRI of primary site as performed during suspected or initial disease workup (provided the individual does not have an endoprosthesis that will cause MRI or CT artifact) ◦ Bone scan (see **PEDONC-1.3** for coding) • Every 3 months for year 1 and 2 ◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ After 24 months off therapy, CXR should be used for surveillance with CT considered medically necessary for new or inconclusive findings on CXR

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Indication	Medically Necessary Imaging Study
Suspected recurrence in individuals with ESFT or PNET	• MRI without and with contrast of the primary site and/or site of suspected recurrence based on symptoms or other imaging, OR • CT, contrast as request, of the primary site and/or site of suspected recurrence based on symptoms or other imaging if there is a contraindication to MRI AND • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Chest is considered medically necessary in initial workup of all suspected malignant bone tumors in children and should be completed prior to anesthesia exposure if possible • CT Abdomen and Pelvis with contrast (CPT® 74177) is not considered medically necessary in the metastatic staging of pediatric EWS but is considered medically necessary in the following situations only: ◦ Evaluation of inconclusive PET findings ◦ Primary site of abdomen or pelvis • The studies above are considered medically necessary even when PET/CT has already been authorized or performed

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Indication	Medically Necessary Imaging Study
• For suspected recurrence in individuals with ESFT or PNET, any of the following: ◦ Conventional imaging reveals findings that are inconclusive or suspicious for recurrence and PET avidity will determine whether biopsy or continued observation is appropriate ◦ Rare circumstances where obvious clinical symptoms show strong evidence suggesting recurrence and PET would replace conventional imaging modalities	PET/CT whole-body (CPT® 78816)
• Biopsy proven recurrence in individuals with ESFT or PNET	• Refer to ESFT initial imaging for studies for suspected recurrence. • PET/CT whole-body (CPT® 78816) • If disease is considered potentially resectable, follow ESFT treatment response as previously noted

- 18F-FDG PET/CT can replace bone scan and bone marrow biopsy in ESFT individuals and is considered medically necessary in the initial staging of all ESFT individuals after histologic diagnosis is established.

Background and Supporting Information

- ESFT includes Ewing sarcoma of bone, extraosseous Ewing sarcoma, Askin tumor, and PNET, characterized by aggressive behavior and a high propensity for metastatic spread (NCCN, 2026).
- Bone and bone marrow metastases occur with sufficient frequency in ESFT to significantly alter staging, prognosis, and therapy (NCCN, 2026).

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Evidence Discussion

- Identification of bone and bone marrow metastases at diagnosis markedly changes treatment approach and prognosis, justifying comprehensive staging in ESFT (NCCN, 2026).
- MRI of the primary site and chest CT are integral components of initial staging (NCCN, 2026).
- FDG-PET/CT is preferred over conventional bone scan for detection of skeletal and nodal metastatic disease. FDG-PET/CT demonstrates high sensitivity and specificity for metastatic disease and can detect lesions occult on conventional imaging (NCCN, 2026).
- Routine use of MRI and PET/CT at defined disease milestones maximizes diagnostic yield while avoiding repetitive or low-value imaging during therapy (NCCN, 2026).
- Structured surveillance that emphasizes clinically impactful sites reduces radiation exposure and false-positive findings without compromising outcome assessment (NCCN, 2026).

Health Equity Considerations

ESFT, including PNET, disproportionately affects children, adolescents, and young adults, and a growing body of evidence demonstrates that sociodemographic factors shape disease presentation and early care. Population-based analyses show that Hispanic patients are more likely than non-Hispanic White patients to present with larger tumors and metastatic disease at diagnosis, while adolescents and young adults more frequently present with axial or pelvic tumors and disseminated disease compared with younger children (Fritz et al., 2024; Goulding et al., 2024). These patterns are clinically meaningful, as tumor size, metastatic burden, and axial location are among the strongest predictors of survival in ESFT (Wells et al., 2024). Barriers such as language isolation, underinsurance, and limited access to specialty care likely contribute to diagnostic delay and more advanced presentation in historically marginalized populations, reinforcing the need for early, accessible diagnostic evaluation (Fritz et al., 2024; Goulding et al., 2024).

Equitable access to timely, high-quality imaging is therefore essential to improving outcomes in ESFT, given the disease's propensity for early spread to lung, bone, and bone marrow and the central role of imaging in staging, surgical planning, and treatment selection. Adolescents and young adults are more likely to experience delays between diagnosis and treatment initiation, potentially reflecting challenges during transitions in care and gaps in insurance coverage, which may be exacerbated by delayed or incomplete imaging (Fritz et al., 2024). Disparities in treatment delivery further compound these risks, as non Hispanic-Black patients have been shown to be less likely to receive chemotherapy even after accounting for disease stage, and rural

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or socioeconomically disadvantaged communities face barriers to sarcoma experienced multidisciplinary care (Fritz et al., 2024; Goulding et al., 2024).

Diagnostic challenges in rare PNET subtypes, including CNS PNETs, further illustrate how limited access to advanced imaging and biopsy can lead to underdiagnosis and delayed therapy (Chamdine et al., 2018). Applying ESFT imaging guidelines with an explicit health equity lens, emphasizing prompt staging, consistent surveillance, and referral to specialized centers, may help reduce diagnostic delay, support timely multimodality treatment, and promote more equitable outcomes across pediatric and young adult populations (Wells et al., 2024).

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Pediatric Germ Cell Tumors (PEDONC-10)

Guideline Page

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Pediatric Germ Cell Tumors (PEDONC-10)

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Criteria

- Sex cord stromal tumors (granulosa cell, theca, sertoli, and leydig tumors) are rare in pediatrics and should be imaged according to general guidelines in: **Testicular, Ovarian and Extragonadal Germ Cell Tumors (ONC-20)** in the Oncology Imaging Guidelines
- This section applies to primary germ cell tumors occurring outside the central nervous system in children who are ≤15 years old at the time of initial diagnosis.
  - For individuals who are >15 years old at diagnosis, the overall prognosis is inferior and these individuals should be imaged according to general guidelines in: **Testicular, Ovarian and Extragonadal Germ Cell Tumors (ONC-20)** in the Oncology Imaging Guidelines.
- For CNS germ cell tumors, use the imaging guidelines in: **CNS Germinomas and Non-Germinomatous Germ Cell Tumors (NGGCT) (PEDONC-4.7)**.

Indication	Medically Necessary Imaging Study
Initial staging for all individuals with germ cell tumors	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ For testicular masses, MRI Abdomen without and with contrast (CPT® 74183) and CT Pelvis with contrast (CPT® 72193) are considered medically necessary instead, if requested • CT Chest with contrast (CPT® 71260) ◦ Should be completed prior to anesthesia exposure if possible

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Indication	Medically Necessary Imaging Study
• Initial staging in individuals with germ cell tumors, in lieu of CT Abdomen and Pelvis with contrast (above), for ANY of the following: ◦ Proven or highly suspected ovarian neoplasm that was already imaged with ultrasound ◦ Immature sacrococcygeal teratoma suspected on other imaging ◦ Contraindication to CT contrast OR • To clarify inconclusive findings on initial staging CT	• MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)
• Ovarian masses that are <10cm in size, have minimal or no visible solid component on ultrasound, and have normal tumor markers	• These are almost universally benign teratomas or functional cysts; thus, advanced imaging is not considered medically necessary
• Additional initial evaluation for individuals with germ cell tumors with ANY of the following: ◦ Symptoms suggesting CNS metastases ◦ Choriocarcinoma syndrome (hemorrhagic metastatic disease to lung with extremely elevated HCG) HCG >10,000miU/ml	• MRI Brain without and with contrast (CPT® 70553)

Indication	Medically Necessary Imaging Study
Additional initial evaluation for individuals with germ cell tumors with systemic symptoms or bone pain	• Nuclear bone scan (See: **PEDONC-1.3** for coding)
Restaging in individuals with germ cell tumors with disease not completely resected at initial diagnosis	• Every 2 cycles (~every 6 weeks) and at the end of planned therapy: ◦ CT Chest/Abdomen/Pelvis (CPT® 71260 and CPT® 74177) with contrast ▪ CT imaging may be considered medically necessary more frequently to assess for surgical resectability in individuals who have received more than 4 cycles of chemotherapy ▪ MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) in lieu of CT abdomen and pelvis for ovarian neoplasms if this modality was used for initial staging or for contraindication to CT contrast ◦ Imaging of any metastatic sites with the same modality used during initial staging
• Suspicious lesion seen on CT and both of the following in individuals with germ cell tumors: ◦ Relapse risk is determined to be low by the treating physician ◦ Biopsy would cause unnecessary morbidity for the individual	• Short-interval CT study of the involved area

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Indication	Medically Necessary Imaging Study
Stage I individuals with germ cell tumors 0-10 years of age treated with surgery only	• Chest x-ray should be completed every 3 months for 1 year, then every 6 months in year two, to complete two years surveillance after completion of all therapy ◦ For those with primary mediastinal tumors at diagnosis, CT Chest with contrast (CPT® 71260) in lieu of CXR according to the above schedule. • Every 3 months for 1 year, then every 6 months in year two, to complete two years surveillance after completion of all therapy: ◦ CT Abdomen/Pelvis with contrast (CPT® 74177) ▪ MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) substituted for surveillance for ovarian neoplasm if this modality was used for initial staging
Stage I individuals with germ cell tumors 11 years of age or older treated with surgery only	• Every 4 months for 2 years, then every 6 months for 1 year, then every 12 months for 2 years to complete 5 years surveillance imaging after completion of all therapy: ◦ Chest x-ray ◦ For individuals with primary mediastinal tumors at diagnosis, CT Chest with contrast (CPT® 71260) in lieu of chest x-ray on the above schedule. • Every 4 months for 2 years, then every 6 months for 1 year, then every 12 months for two years to complete 5 years surveillance ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) ▪ MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) substituted for surveillance for ovarian neoplasm if this modality was used for initial staging

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Indication	Medically Necessary Imaging Study
Stage II-IV individuals with germ cell tumors	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) and CT Abdomen and Pelvis with contrast (CPT® 74177) every 3 months for 1 year then every 6 months in year 2, then annually in years 3-5 after completion of all therapy. ◦ MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) substituted for surveillance for ovarian neoplasm if this modality was used for initial staging • Individuals with brain or bone metastases should have surveillance imaging of those areas on the same schedule as the primary site imaging with the same modality used during initial staging
• Suspected Recurrence in individuals with germ cell tumors: ◦ Any clinically significant rise in tumor markers ◦ Symptoms suggesting recurrent disease ◦ Abnormal chest x-ray	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) in lieu of CT for suspected recurrence of immature sacrococcygeal tumor or ovarian neoplasm or for contraindication to CT contrast • Whole-body bone scan (CPT® 78306) for individuals with a history of bone involvement or with bone pain
Clarify inconclusive findings on conventional imaging in individuals with germ cell tumors	PET/CT (CPT® 78815)

Background and Supporting Information

- Pediatric germ cell tumors are a heterogeneous group of malignant neoplasms arising from primordial germ cells and most commonly occur in the gonads (testes or ovaries) or extragonadal sites along the midline, including the abdomen, pelvis, mediastinum, and sacrococcygeal region (NCCN, 2026).
- Malignant pediatric germ cell tumors commonly include yolk sac tumor, embryonal carcinoma, choriocarcinoma, and mixed germ cell tumors (which may include immature teratomatous elements). The majority of pediatric germ cell tumors are non-seminomatous (NCCN, 2026).

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- Pediatric germ cell tumors may arise in testicular, ovarian, or extragonadal locations. Tumor location influences initial imaging modality, staging evaluation, and surgical planning (NCCN, 2026).
- Overall treatment strategies for malignant germ cell tumors are similar regardless of primary site. Localized disease is often treated with surgery alone, while advanced or high-risk disease is treated with multimodality therapy including combination chemotherapy. Imaging is used for staging, response assessment, and surveillance and must account for long-term survivorship (NCCN, 2026).
- Serum tumor markers, including alpha-fetoprotein (AFP) and beta-human chorionic gonadotropin (β-hCG), are integral to diagnosis, staging, response assessment, and surveillance, and often reduce reliance on serial imaging studies (NCCN, 2026).

Evidence Discussion

- Ultrasound, combined with serum tumor markers, is supported as the primary initial imaging modality for suspected ovarian, testicular, and abdominal extragonadal pediatric germ cell tumors, providing high diagnostic yield without radiation exposure. This approach minimizes harm while preserving sensitivity for initial disease detection (NCCN, 2026).
- Cross-sectional imaging of the chest, abdomen, and pelvis is considered medically necessary for initial staging when a suspected germ cell tumor is identified, as imaging findings guide surgical approach and assess for metastatic disease. This targeted use provides clinical benefit when performed prior to histologic confirmation (NCCN, 2026).
- Decline and normalization of AFP and β-hCG are reliable indicators of treatment response in pediatric germ cell tumors. Evidence supports prioritizing tumor marker trends over routine imaging for response assessment, reducing unnecessary radiation exposure and false-positive findings (NCCN, 2026).
- Surveillance strategies emphasizing frequent tumor marker assessment with limited imaging are supported by high cure rates and early biochemical detection of recurrence. Restricting routine imaging mitigates cumulative radiation risks, anesthesia exposure, and downstream testing in long-term pediatric survivors (NCCN, 2026).
- Chest radiography is the preferred surveillance imaging modality for thoracic evaluation in stages I-IV disease, balancing adequate detection of pulmonary relapse with lower radiation dose compared with CT imaging (NCCN, 2026).
- There is no evidence supporting routine use of PET or PET/CT in pediatric germ cell tumor evaluation. Data demonstrate that PET does not reliably predict treatment response or individual outcomes and offers no added benefit over conventional imaging in this population (NCCN, 2026).
- Suspicious lesions identified on conventional imaging require tissue confirmation, as imaging alone cannot reliably distinguish viable malignancy from post-treatment

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changes, residual teratoma, or necrosis. This approach prevents inappropriate escalation of therapy (NCCN, 2026).
- Given excellent long-term survival outcomes for pediatric germ cell tumors, imaging strategies emphasize diagnostic sufficiency while minimizing late effects, aligning clinical benefit with harm reduction principles central to pediatric imaging guidelines (NCCN, 2026).

Health Equity Considerations

Pediatric germ cell tumors (GCTs) demonstrate important disparities in incidence, stage at diagnosis, and survival across sex and race/ethnicity, with implications for timely imaging and risk stratification. Population-based analyses show that survival differences are most pronounced among males, with Hispanic and Asian/Pacific Islander boys experiencing significantly higher mortality compared with non-Hispanic White males, whereas no consistent survival differences are observed among females (Williams et al., 2021). In Hispanic males with gonadal tumors, more advanced stage at diagnosis partially mediates this disparity, suggesting that delayed detection and diagnosis contribute to worse outcomes, while persistent mortality differences in other subgroups point to additional factors such as tumor biology or treatment response (Williams et al., 2021). Sex-based differences are also evident in intracranial GCTs, where males consistently demonstrate higher long-term mortality across histologic subtypes and disease stages, particularly in distant disease, despite overall improvements in early detection over time (Lin et al., 2025). These findings underscore the prognostic importance of cancer stage and sex in pediatric GCTs and highlight populations at higher risk for adverse outcomes.

Equitable access to timely, high-quality imaging is central to addressing these disparities, as imaging plays a critical role in tumor localization, staging, and treatment planning for both gonadal and intracranial GCTs. Advances in neuroimaging and surveillance have contributed to a shift toward increased detection of localized disease, which is associated with substantially lower mortality than regional or distant disease (Lin et al., 2025). However, persistent excess mortality in males and in children presenting with advanced-stage tumors suggests that improvements in imaging are not uniformly realized across populations. Delays in diagnosis, differences in referral patterns to specialty centers, and potential barriers related to socioeconomic factors or healthcare access may disproportionately affect certain racial and ethnic groups, particularly Hispanic males (Williams et al., 2021). Applying pediatric GCT imaging guidelines with a health equity lens, emphasizing early evaluation, comprehensive staging at presentation, and consistent follow-up imaging, may help reduce diagnostic delay, support timely initiation of guideline concordant therapy, and promote more equitable outcomes across pediatric populations (Lin et al., 2025).

References (PEDONC-10)

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Pediatric Liver and Pancreatic Tumors (PEDONC-11)

Guideline Page

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Pediatric Liver and Pancreatic Tumors – General Considerations (PEDONC-11.1)

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Criteria

- Primary hepatic germ cell tumors should follow imaging guidelines in: **Pediatric Germ Cell Tumors (PEDONC-10)**.
- Primary hepatic sarcomas should follow imaging guidelines in: **Non-Rhabdomyosarcoma Soft Tissue Sarcomas (NRSTS) (PEDONC-8.3)**.
- Imaging requests relating to liver transplant surgery and surveillance should follow guidelines in section **Transplant (AB-42)** in the Abdomen Imaging Guidelines.

Background and Supporting Information

- Pediatric liver and pancreatic tumors are rare and biologically distinct from adult malignancies, requiring tailored imaging approaches that minimize radiation exposure while supporting accurate diagnosis, staging, and treatment planning (NCCN, 2026).
- Pediatric hepatic malignancies are dominated by hepatoblastoma and hepatocellular carcinoma, while pancreatic tumors are uncommon and most often low-grade epithelial neoplasms or embryonal tumors rather than invasive ductal adenocarcinoma (NCCN, 2026).
- Serum markers (e.g., AFP) are integral to diagnosis, response assessment, and surveillance in pediatric liver tumors, often reducing the need for repeated imaging studies (NCCN, 2026).

Evidence Discussion

- Evidence supports ultrasound as the preferred initial imaging modality for pediatric hepatobiliary and pancreatic tumors, offering high diagnostic utility while avoiding radiation exposure (NCCN, 2026).
- Cross-sectional imaging improves staging accuracy and surgical planning when disease is suspected or confirmed but does not demonstrate benefit as routine surveillance in asymptomatic, marker-negative pediatric patients (NCCN, 2026).
- Limiting serial CT imaging aligns with evidence demonstrating excellent cure rates in pediatric tumors and heightened lifetime radiation sensitivity in children, supporting restrictive imaging criteria (NCCN, 2026).
- PET/CT has not demonstrated sufficient sensitivity, predictive value, or outcome benefit in pediatric liver or pancreatic tumors to justify routine use (NCCN, 2026).

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Hepatoblastoma (PEDONC-11.2)

ONCP.LT.0011.2.A
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Criteria

Initial Staging

Indication	Medically Necessary Imaging Study
All individuals with suspected liver tumors	Abdominal ultrasound (CPT® 76700) is considered medically necessary even after MRI or CT imaging in order to allow evaluation for tumor thrombus
Individuals with a liver mass that is a biopsy proven primary liver malignancy or a liver mass that is clinically consistent with a primary liver mass	ANY of the following: • MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) for evaluating tumor margins and vascular anatomy ◦ Hepatobiliary-specific contrast agents (gadoxetate, gadobenate [MultiHance]) are preferred if available and are considered medically necessary if requested, whether or not a prior gadolinium-enhanced MRI has been previously performed ◦ CT Abdomen and Pelvis with (CPT® 74177) is inferior to MRI for pediatric liver malignancies and is only considered medically necessary if MRI is not available, contraindicated, or sedation is required but not available, or to clarify specific inconclusive areas on MRI • MRA Abdomen (CPT® 74185) or CTA Abdomen (CPT® 74175) to evaluate vascular invasion

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Indication	Medically Necessary Imaging Study
Initial work-up for all pediatric liver tumors	CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250) • Should be completed prior to anesthesia exposure if possible
Initial evaluation of bony metastases in individuals with hepatoblastoma only with systemic symptoms or bone pain	Bone scan (See: **Modality General Considerations (PEDONC-1.3)**)
Symptoms suggesting CNS metastases in individuals with hepatoblastoma	MRI Brain without and with contrast (CPT® 70553)
Rare circumstances for preoperative planning when MRI and CT scans are insufficient for surgical decision-making in individuals with hepatoblastoma	PET/CT (CPT® 78815) • Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

Treatment Response

- While the majority of individuals with hepatoblastoma will require abdomen and pelvis imaging at all time points, the pelvis imaging may be omitted at the discretion of the ordering physician

Indication	Medically Necessary Imaging Study
Individuals with hepatoblastoma with incomplete resection	Every 2 cycles **and** at the end of planned therapy • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) • MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) or CT Abdomen and Pelvis without and with contrast (CPT® 74178) is inferior to MRI for pediatric liver malignancies and is only considered medically necessary if MRI is not available, contraindicated, or sedation is required but not available • Imaging of any metastatic sites with the same modality used during initial staging
To evaluate vascular invasion in individuals with hepatoblastoma	MRA Abdomen (CPT® 74185) or CTA Abdomen (CPT® 74175)
Tumor thrombus was detected at initial diagnosis in individuals with hepatoblastoma	Abdominal ultrasound (CPT® 76700) • If no tumor thrombus was present, continued ultrasound evaluations are not considered medically necessary without a specific reason documented in the clinical records
Rare circumstances for preoperative planning when MRI and CT scans are insufficient for surgical decision-making in individuals with hepatoblastoma	PET/CT (CPT® 78815) • Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

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Surveillance Imaging

Indication	Medically Necessary Imaging Study
Surveillance in individuals with hepatoblastoma with an AFP of >100ng/mL	CT Chest with contrast (CPT® 71260) and MRI Abdomen without and with contrast (CPT® 74183) for any clinically significant rise in tumor markers or symptoms suggesting recurrent disease • CT Abdomen and Pelvis with contrast (CPT® 74177) is inferior to MRI for pediatric liver malignancies and is only considered medically necessary if MRI is not available, contraindicated, or sedation is required but not available, or to clarify specific inconclusive areas on MRI
Surveillance in individuals with hepatoblastoma with AFP ≤100ng/mL at diagnosis or recurrence	• MRI Abdomen with and without contrast (CPT® 74183) every 3 months for 2 years, then every 4 months for 2 years after completion of all therapy • Chest x-ray or CT Chest with contrast (CPT® 71260) every 3 months for 2 years, then every 4 months for 2 years after completion of all therapy • Individuals with brain or bone metastases should have surveillance imaging on the same schedule as the primary site imaging with the same modality used during initial staging

Background and Supporting Information

- Hepatoblastoma is the most common primary malignant liver tumor in children, typically diagnosed before 3 years of age, with a median age of approximately 19 months (NCCN, 2026).
- While most cases are sporadic, associations include Beckwith-Wiedemann syndrome, familial adenomatous polyposis, and trisomy 18 (NCCN, 2026).
- AFP is elevated in the majority of cases and is essential for diagnosis, treatment response assessment, and surveillance (NCCN, 2026).
- Localized tumors of pure fetal histology may be cured with surgery alone, while more advanced disease requires multimodality therapy, including chemotherapy and, in select cases, liver transplantation (NCCN, 2026).

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Evidence Discussion

- AFP monitoring reliably detects recurrence in hepatoblastoma, with evidence supporting biochemical monitoring as the primary surveillance method, reducing reliance on routine imaging (NCCN, 2026).
- Cross-sectional imaging remains necessary for unresected tumors or when AFP levels do not adequately reflect disease burden, supporting selective rather than routine imaging use (NCCN, 2026).
- PET/CT has not demonstrated clinical utility during initial staging, response assessment, or surveillance in hepatoblastoma and should not substitute for biopsy of suspicious lesions (NCCN, 2026).
- Restricting surveillance imaging in AFP-responsive patients minimizes cumulative radiation exposure without compromising outcomes (NCCN, 2026).

Health Equity Considerations

Hepatoblastoma, the most common primary liver malignancy in children, is characterized by marked disparities in incidence, stage at presentation, and outcomes across demographic and socioeconomic groups. Population-based studies demonstrate that hepatoblastoma incidence continues to rise in the United States, with disproportionately higher incidence among male infants and Hispanic children, as well as elevated risk among certain racial and ethnic minority populations (Kahla et al., 2022; Espinoza et al., 2023). Latino children are not only more likely to be diagnosed with hepatoblastoma but also have more than twice the risk of presenting with metastatic disease at diagnosis compared with non-Hispanic White children, a finding with direct prognostic significance given the strong association between metastatic disease and poorer survival (Espinoza et al., 2023). National data further show that non-Hispanic Black children experience significantly worse survival outcomes, even after adjusting for stage and age, suggesting that disparities are not fully explained by disease biology alone and may reflect structural inequities in access to timely diagnosis and high quality care (Kahla et al., 2022). Globally, although overall hepatoblastoma mortality has declined, persistent differences by sex, age, and geographic region underscore continuing inequities in disease burden and early detection, particularly in low resource settings (Chen et al., 2025).

Equitable access to timely, high-quality imaging is central to addressing these disparities, as imaging underpins accurate staging, risk stratification, surgical planning, and assessment of metastatic disease in hepatoblastoma. Delays in diagnosis or incomplete imaging may contribute to advanced stage at presentation, particularly among populations facing barriers related to insurance status, socioeconomic disadvantage, or limited access to specialized pediatric oncology centers (Kahla et al., 2022; Espinoza et al., 2023). Disparities also extend beyond diagnosis to treatment delivery; children who are Black or publicly insured are significantly less likely to undergo potentially curative surgical resection or liver transplantation, independent

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of disease related factors, highlighting inequities in access to definitive therapy that may be compounded by delayed or fragmented diagnostic evaluation (Ziogas et al., 2024). While recent evidence suggests relatively equitable enrollment in Children's Oncology Group clinical trials for hepatoblastoma during periods when trials are widely available, gaps in access to specialty care and advanced imaging may still limit timely referral and optimal management outside of clinical trial settings (Monterroso et al., 2025). Emphasizing prompt diagnostic workup, standardized staging imaging, and early referral to multidisciplinary liver tumor centers may help reduce delays, improve access to curative treatment, and support more equitable outcomes for infants and children with hepatoblastoma across diverse populations (Kahla et al., 2022; Ziogas et al., 2024).

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Pediatric Hepatocellular Carcinoma (HCC) (PEDONC-11.3)

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v1.0.2027

Criteria

Initial Staging

Indication	Medically Necessary Imaging Study
All individuals with suspected liver tumors	Abdominal ultrasound (CPT® 76700) is considered medically necessary even after MRI or CT imaging in order to allow evaluation for tumor thrombus
Individuals with a liver mass that is a biopsy proven primary liver malignancy or a liver mass that is clinically consistent with a primary liver mass	ANY of the following: • MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) ◦ Hepatobiliary specific contrast agents (gadoxetate, gadobenate [MultiHance]) are preferred if available and are considered medically necessary if requested, whether or not a prior gadolinium-enhanced MRI has been previously performed ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) is inferior to MRI for pediatric liver malignancies and is only considered medically necessary if MRI is not available, contraindicated, or sedation is required but not available, or to clarify specific inconclusive areas on MRI • MRA Abdomen (CPT® 74185) or CTA Abdomen (CPT® 74175) to evaluate vascular invasion

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Indication	Medically Necessary Imaging Study
Initial work-up for all pediatric liver tumors	CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250) • Should be completed prior to anesthesia exposure if possible
Symptoms suggesting CNS metastases in individuals with HCC	MRI Brain without and with contrast (CPT® 70553)
Initial evaluation of bony metastases in individuals with HCC only with systemic symptoms or bone pain	Nuclear bone scan (See: **Modality General Considerations (PEDONC-1.3)**)
Rare circumstances for preoperative planning when MRI and CT scans are insufficient for surgical decision-making in individuals with HCC	PET/CT (CPT® 78815) • Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

Treatment Response

- While the majority of individuals will require abdomen and pelvis imaging at all time points, the pelvis imaging may be omitted at the discretion of the ordering physician

Indication	Medically Necessary Imaging Study
All individuals with HCC	Imaging of any metastatic sites with the same modality used during initial staging

Indication	Medically Necessary Imaging Study
Individuals with HCC with disease not completely resected at initial diagnosis	Every 2 cycles (~6 weeks) **and** at the end of planned therapy: • CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250) • MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) is inferior to MRI for pediatric liver malignancies and is only considered medically necessary if MRI is not available, contraindicated, or sedation is required but not available, or to clarify specific inconclusive areas on MRI • MRA Abdomen (CPT® 74185) or CTA Abdomen (CPT® 74175) to evaluate vascular invasion
Individuals with HCC if tumor thrombus was detected at initial diagnosis	Abdominal ultrasound (CPT® 76700) • If no tumor thrombus was present, continued ultrasound evaluations are not considered medically necessary without a specific reason documented in the clinical records
Rare circumstances for preoperative planning when MRI and CT scans are insufficient for surgical decision-making in individuals with HCC	PET/CT (CPT® 78815) • Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

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Surveillance Imaging

Indication	Medically Necessary Imaging Study
All individuals with HCC	Every 3 months for 1 year, then every 6 months for 1 year, then annually for 3 years after completion of all therapy: • MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) • Chest x-ray or CT Chest with contrast (CPT® 71260)

Background and Supporting Information

- Pediatric HCC, including fibrolamellar HCC, primarily affects older children and adolescents and has substantially poorer outcomes than hepatoblastoma, particularly in advanced stages (NCCN, 2026).
- Ultrasound and tumor markers (including AFP) are used for initial evaluation, followed by cross-sectional imaging to assess resectability and metastatic disease (NCCN, 2026).
- Surgical resection remains the primary curative approach when feasible; most pediatric patients do not receive adjuvant therapy after complete resection (NCCN, 2026).

Evidence Discussion

- CT and MRI provide essential anatomic detail for staging and surgical candidacy but do not support routine high-frequency surveillance beyond guideline recommendations (NCCN, 2026).
- Surveillance imaging following complete resection is supported to detect recurrence, but intensity should balance recurrence risk with radiation exposure (NCCN, 2026).
- PET/CT lacks evidence of benefit for surveillance or routine evaluation in pediatric HCC and should not replace histologic confirmation of suspicious findings (NCCN, 2026).
- When brain or skeletal metastases are present, imaging follow-up should mirror the primary site's schedule and modality, ensuring consistency without over-imaging (NCCN, 2026).

Health Equity Considerations

Pediatric HCC is rare but associated with meaningful disparities in outcomes that mirror, and in some cases diverge from, those observed in adults. Population-based analyses demonstrate that while overall pediatric HCC incidence in the United States

has remained stable, children and adolescents frequently present with advanced disease due to the absence of routine surveillance in most pediatric populations, in contrast to adults with cirrhosis who may undergo screening (Arnett et al., 2025; Guo et al., 2021). Notably, non-Hispanic Black children with HCC experience a significantly higher risk of death compared with non-Hispanic White children, even after adjustment for stage and socioeconomic factors, suggesting that disparities extend beyond tumor burden alone and may reflect inequities in access to specialized care, timely diagnosis, and treatment delivery (Arnett et al., 2025). Geographic inequities are also evident, as pediatric patients diagnosed in smaller metropolitan or non-metropolitan counties have worse survival outcomes than those treated in large urban centers, underscoring the importance of access to high volume, multidisciplinary liver tumor programs (Arnett et al., 2025; Zheng et al., 2026).

Disparities in access to definitive therapy further compound outcome differences in pediatric HCC. Surgical resection or liver transplantation remains the cornerstone of curative treatment, yet national data show that children from racially marginalized groups and those with public or no insurance are less likely to undergo potentially curative liver surgery, even after accounting for disease-related factors (Ziogas et al., 2024). Structural barriers, such as limited access to transplant centers, delays in referral, and differences in institutional resources, may disproportionately affect these children and contribute to worse survival (Guo et al., 2021; Zheng et al., 2026). Although clinical trial participation does not appear to differ substantially by race or socioeconomic status in pediatric liver tumors overall, inequities in diagnostic imaging, staging evaluation, and referral pathways may still limit equitable access to advanced therapies outside of trial settings (Ziogas et al., 2024). Prioritizing timely cross-sectional imaging for accurate staging, early multidisciplinary consultation, and referral to specialized centers may help reduce disparities in treatment access and improve outcomes for children and adolescents with hepatocellular carcinoma (Arnett et al., 2025; Zheng et al., 2026).

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Pediatric Pancreatic Carcinoma (PEDONC-11.4)

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v1.0.2027

Criteria

- This guideline applies to suspected or diagnosed pancreatic neoplasms in children.
- Beckwith-Wiedemann syndrome increases pancreatoblastoma risk. MEN1, VHL, neurofibromatosis, and tuberous sclerosis are also risk factors for pancreatic endocrine neoplasms.
  - Screening studies in these conditions are found in **Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2)**.

Indication	Medically Necessary Imaging Study
Initial staging in individuals with pancreatic carcinoma	▪ CT Abdomen and Pelvis with or without and with contrast (CPT® 74177 or CPT® 74178) OR MRI Abdomen without and with contrast plus MRI Pelvis with and without contrast (CPT® 74183 and CPT® 72197) ▪ CT Chest with (CPT® 71260) or without (CPT® 71250) contrast ▪ ^{68}Ga-Dotatate PET/CT whole-body (CPT® 78816) - Exception: for pediatric pancreatic tumor of non-neuroendocrine origin (SPN, pancreatoblastoma and other exocrine origin) with equivocal conventional imaging, FDG PET/CT whole-body or skull to thighs (CPT® 78816 or 78815)
Treatment response ~every 2 cycles during treatment and at the end of planned therapy in individuals with pancreatic carcinoma	▪ CT or MRI as used at time of initial imaging

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Indication	Medically Necessary Imaging Study
If conventional imaging for treatment response is equivocal in individuals with pancreatic carcinoma	▪ Neuroendocrine tumors: ^{68}Ga Dotatate PET/CT skull to thighs or whole-body (CPT® 78815 or 78816) ▪ Non-neuroendocrine tumors: FDG PET/CT skull to thighs or whole-body (CPT® 78815 or 78816)
Assess candidacy for PRRT therapy in individuals with pancreatic carcinoma	▪ ^{68}Ga Dotatate PET/CT whole body CPT® 78816 to assess candidacy for PRRT therapy
Surveillance in individuals with pancreatic carcinoma	▪ MRI or CT, modality and contrast as used in initial imaging, every 3 months for 2 years, then every 4 months for 1 year, then every 6 months for 1 year, then in 12 months to complete 5 years surveillance
Suspected recurrence in individuals with pancreatic carcinoma	▪ Repeat all imaging as noted in initial staging section

Background and Supporting Information

- Pancreatic tumors in children are exceedingly rare and differ biologically from adult pancreatic ductal adenocarcinoma (NCCN, 2026).
- The most frequent pediatric pancreatic tumor is solid pseudopapillary neoplasm, a low-grade epithelial malignancy, followed by pancreatoblastoma (NCCN, 2026).
- Surgical resection is the primary treatment modality, with favorable outcomes for low-grade tumors (NCCN, 2026).

Evidence Discussion

- Cross-sectional imaging is valuable for diagnosis and operative planning but does not support intensive post-treatment surveillance in asymptomatic patients (NCCN, 2026).
- Given low recurrence risk and favorable prognosis for most pediatric pancreatic tumors, limiting surveillance imaging reduces unnecessary radiation and downstream testing (NCCN, 2026).
- There is no evidence supporting PET/CT use in pediatric pancreatic tumors, and its routine use may increase false-positive findings without clinical benefit (NCCN, 2026).

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Health Equity Considerations

Pediatric pancreatic carcinoma is exceedingly rare, and evidence specific to children and adolescents is limited; however, available data suggest that disparities affecting adults with pancreatic cancer likely extend to pediatric populations through shared structural and contextual factors. Pancreatic malignancies are often diagnosed at advanced stages due to nonspecific symptoms and lack of effective screening, making timely access to high quality diagnostic imaging critical for staging and treatment planning (Patel et al., 2025). Social determinants of health including socioeconomic status (SES), insurance coverage, geographic location, race, educational attainment, and health literacy have been consistently associated with delays in diagnosis, lower likelihood of receiving guideline-concordant care, and worse outcomes in pancreatic cancer overall (Patel et al., 2025). In pediatric patients, who already face diagnostic delays due to disease rarity, these structural barriers may further exacerbate inequities in access to timely cross-sectional imaging, subspecialty consultation, and referral to high volume multidisciplinary centers.

Socioeconomic disadvantage and insurance status are particularly influential across the pancreatic cancer care continuum. Lower SES and lack of private insurance are associated with more advanced stage at presentation, reduced access to multimodal therapy, and lower rates of surgical resection, the only potentially curative option for pancreatic carcinoma (Patel et al., 2025). Families experiencing financial insecurity may encounter additional barriers to completing recommended imaging studies, traveling to specialty centers, or maintaining follow up, challenges that can be magnified in pediatric care where caregiver availability and resources are essential. Geographic disparities compound these effects, as patients in rural or underserved areas have less access to high volume centers with pediatric surgical and oncology expertise, which are associated with improved outcomes and more consistent delivery of guideline-based imaging and treatment (Patel et al., 2025).

Racial and ethnic disparities documented in pancreatic cancer also warrant consideration in pediatric populations. In adults, Black and Hispanic patients are more likely to present with advanced disease, experience treatment delays, and receive care at lower volume institutions, even after adjustment for SES and insurance status (Patel et al., 2025). Language barriers and limited health literacy further impede navigation of complex care pathways, contributing to lower rates of guideline-concordant imaging and therapy. While pediatric-specific data are sparse, children from racially and ethnically marginalized communities may be similarly affected by systemic barriers that delay diagnosis and limit access to specialized imaging and multidisciplinary evaluation. Emphasizing early diagnostic imaging, streamlined referral pathways, culturally and linguistically appropriate communication, and access to high volume pediatric centers may help mitigate disparities and promote more equitable outcomes for this rare but aggressive malignancy (Patel et al., 2025).

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Retinoblastoma (PEDONC-12)

Guideline Page

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Retinoblastoma – General Considerations (PEDONC-12.1)

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Criteria

- Detailed evaluation by a physician with significant training and/or experience in retinoblastoma (most commonly a pediatric ophthalmologist or pediatric oncologist) is considered medically necessary prior to considering advanced imaging.
- Whole-body MRI has shown poor sensitivity and specificity in individuals with predisposition to systemic malignancy due to germline RB1 mutations, and is not considered medically necessary.
- Individuals who carry the RB1 mutation also have increased risk of developing other cancers, such as osteosarcoma, soft tissue sarcomas, or melanoma. For more information on heritable retinoblastoma, see: **Familial Retinoblastoma Syndrome (PEDONC-2.12)**.

Background and Supporting Information

- Retinoblastoma is a malignant tumor of the developing retina that occurs almost exclusively in early childhood, with most cases diagnosed before five years of age. Approximately 60% of cases are unilateral and 40% are bilateral, with bilateral disease presenting at a younger mean age than unilateral disease (Lohmann & Gallie, 2023).
- Retinoblastoma develops through biallelic inactivation of the RB1 tumor suppressor gene. Individuals with bilateral retinoblastoma, multifocal disease, trilateral retinoblastoma, or a family history of retinoblastoma are considered at high risk for a germline RB1 pathogenic variant, while most unilateral cases without family history are non-heritable (Lohmann & Gallie, 2023).
- Disease may be unilateral, bilateral, or trilateral, the latter defined by the presence of an associated pinealoblastoma. Trilateral retinoblastoma occurs almost exclusively in individuals with a germline RB1 pathogenic variant (Lohmann & Gallie, 2023).
- Diagnosis is established by ophthalmologic examination of the retina with pupillary dilation, typically confirmed by examination under anesthesia. Biopsy is avoided due to the risk of tumor dissemination beyond the globe (Lohmann & Gallie, 2023).
- Individuals with heritable retinoblastoma have a markedly increased lifetime risk of subsequent malignant neoplasms, particularly following exposure to ionizing radiation. Avoidance of radiation exposure is a central consideration in diagnostic and surveillance imaging (Lohmann & Gallie, 2023).

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Evidence Discussion

- Clinical ophthalmologic examination, particularly examination under anesthesia in young children, is the most sensitive method for detecting intraocular disease and recurrence. Evidence supports prioritizing EUA over routine imaging (Lohmann & Gallie, 2023).
- Survivors of heritable retinoblastoma demonstrate substantially increased rates of secondary malignancies, particularly when exposed to ionizing radiation. Restricting CT and x-ray imaging reduces long-term cancer risk without compromising detection of intraocular disease (Lohmann & Gallie, 2023).
- Imaging is clinically valuable for staging, assessment of extraocular extension, and evaluation of suspected intracranial disease, but routine surveillance imaging has not shown benefit in asymptomatic patients (Lohmann & Gallie, 2023).

Health Equity Considerations

Retinoblastoma is a highly curable pediatric malignancy when detected early, making it a well-recognized tracer cancer for evaluating health system performance and equity in childhood oncology. Despite substantial global improvements in survival and reductions in mortality and disability adjusted life years (DALYs) over the past three decades, the burden of retinoblastoma remains disproportionately concentrated among children in low income and low-sociodemographic index (SDI) regions (Chen et al., 2024; Li et al., 2025; Zhu et al., 2025). Across analyses of Global Burden of Disease 2021 data, children in low SDI settings, particularly in Sub-Saharan Africa and parts of South Asia, continue to experience the highest incidence of advanced disease, mortality, and DALYs, reflecting persistent inequities in early detection, access to diagnostic imaging, and timely treatment (Chen et al., 2024; Li et al., 2025). Delayed presentation due to limited caregiver awareness, insufficient primary screening, and constrained access to ophthalmologic and imaging services often leads to diagnosis at more advanced stages, where treatment is more complex and outcomes are poorer (Zhu et al., 2025).

Socioeconomic status, geographic location, and health system capacity are key drivers of inequities along the retinoblastoma care continuum. Global analyses demonstrate strong negative correlations between SDI and retinoblastoma mortality and DALYs, indicating that children in poorer regions bear a markedly higher disease burden despite the availability of effective treatments (Chen et al., 2024; Li et al., 2025). In low resource settings, shortages of specialized imaging modalities (such as MRI for optic nerve and intracranial assessment), limited access to pediatric ocular oncology expertise, and fragmented referral pathways contribute to diagnostic delays and higher rates of enucleation and metastatic disease (Zhu et al., 2025). These disparities were further exacerbated during the COVID-19 pandemic, which disrupted routine pediatric care, diagnostic imaging, and treatment services, particularly in low and middle income countries, raising concerns about underdiagnosis and long-term survival impacts in vulnerable populations (Zhu et al., 2025).

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Evidence suggests that regions with robust screening programs, universal health coverage, coordinated referral networks, and timely access to diagnostic imaging achieve earlier detection, lower mortality, and substantially reduced DALYs (Chen et al., 2024; Li et al., 2025). Strategies such as standardized imaging protocols, expansion of pediatric ophthalmic imaging capacity, leveraging telemedicine and digital screening tools, and integration with global initiatives like the WHO Global Initiative for Childhood Cancer may help reduce inequities in diagnosis and staging (Li et al., 2025; Zhu et al., 2025). Emphasizing prompt, guideline-concordant imaging regardless of socioeconomic or geographic barriers is critical to ensuring that all children with suspected retinoblastoma have equitable opportunities for early diagnosis, vision preservation, and survival.

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Retinoblastoma – Imaging (PEDONC-12.2)

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Criteria

Initial Staging

- Tumor biopsy is NOT considered medically necessary prior to imaging in individuals with retinoblastoma
- PET is not considered medically necessary in the evaluation of retinoblastoma

Indication	Medically Necessary Imaging Study
Initial work-up of individuals with retinoblastoma	MRI Orbits (CPT® 70543) and Brain (CPT® 70553) without and with contrast • Brain imaging may be omitted or deferred at the discretion of the treating ophthalmologist or oncologist
Evidence of CNS metastasis in individuals with retinoblastoma on: • Ophthalmologic exam • MRI Brain • Lumbar CSF cytology	MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Clinical symptoms to suggest metastatic disease in individuals with retinoblastoma	CT Chest with contrast (CPT® 71260) and MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and 72197)
Ordered by the treating ophthalmologist for a specified indication in individuals with retinoblastoma	CT Orbital (contrast as requested) and orbital ultrasound
Individuals with retinoblastoma with systemic bone pain suggestive of bony metastases	Nuclear bone scan (See: **Modality General Considerations (PEDONC-1.3)**)

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Treatment Response

Indication	Medically Necessary Imaging Study
All individuals with retinoblastoma	MRI Orbits without and with contrast (CPT® 70543) and/or MRI Brain without and with contrast (CPT® 70553) every 2 cycles (~ every 6 weeks) and at the end of planned therapy
Individuals with retinoblastoma with metastatic disease	Imaging of known positive areas using the same modality as at initial staging every 2 cycles (~6 to 8 weeks) and at the end of planned therapy

Surveillance

Indication	Medically Necessary Imaging Study
Unilateral retinoblastoma	Surveillance using advanced imaging for unilateral retinoblastoma after enucleation or exenteration only for evaluation of specific clinical concerns
Bilateral retinoblastoma or individuals treated with ocular salvage approach	MRI Orbits (CPT® 70543) and Brain (CPT® 70553) for individuals undergoing ocular salvage treatment approaches every 6 months for 2 years following completion of therapy or until the age of 5 years, whichever is later

Background and Supporting Information

- Individuals with germline RB1 pathogenic variants or bilateral disease are at increased risk for pinealoblastoma (Lohmann & Gallie, 2023).

Evidence Discussion

- MRI is the preferred cross-sectional imaging modality in retinoblastoma for evaluation of optic nerve involvement, extraocular extension, and intracranial pathology. MRI avoids ionizing radiation and provides superior soft-tissue characterization compared with CT (Lohmann & Gallie, 2023).
  - MRI may be used to evaluate suspected trilateral retinoblastoma; however, routine MRI screening of the pineal gland is not considered medically necessary for unilateral, non-heritable cases (Lohmann & Gallie, 2023).

- CT is not recommended for routine evaluation of retinoblastoma, particularly in infants or individuals with suspected heritable disease, due to the increased lifetime risk of radiation-induced malignancy (Lohmann & Gallie, 2023).
- The primary method of surveillance following diagnosis and treatment is serial ophthalmologic examination under anesthesia. Routine cross-sectional imaging does not improve outcomes and increases anesthesia exposure, cost, and potential incidental findings (Lohmann & Gallie, 2023).
- Imaging strategies that minimize radiation and anesthesia exposure are particularly important given the excellent survival rates of retinoblastoma and the long life expectancy of survivors. Restrictive imaging criteria balance diagnostic benefit with long-term harm reduction (Lohmann & Gallie, 2023).

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References (PEDONC-12)

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Pediatric Nasopharyngeal Carcinoma (PEDONC-13)

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Pediatric Nasopharyngeal Carcinoma – General Considerations (PEDONC-13.1)

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Background and Supporting Information

- Pediatric nasopharyngeal carcinoma (NPC) is a rare epithelial malignancy arising from the nasopharyngeal mucosa and accounts for a minority of head and neck cancers in children and adolescents, with increasing incidence in older adolescents and distinct geographic and racial distributions (NCI, 2024).
- Due to rich lymphatic drainage of the nasopharynx, regional metastases frequently involve cervical and retropharyngeal lymph nodes. Distant metastatic disease most commonly affects bone and lung, with mediastinal and hepatic involvement occurring less frequently (NCI, 2024).
- The most common initial presentation is painless cervical lymphadenopathy. Other presenting symptoms may include nasal obstruction, epistaxis, otitis media, headache, and cranial nerve deficits related to skull base invasion (NCI, 2024).
- Pediatric NPC is strongly associated with Epstein-Barr virus (EBV), and elevated circulating EBV DNA is commonly detected at diagnosis. Pediatric cases are more likely to be nonkeratinizing or undifferentiated histologic subtypes (NCI, 2024).
- Skull base and intracranial extension are common at presentation and are associated with more advanced disease stage. Despite a high frequency of advanced locoregional disease, the incidence of distant metastasis at initial diagnosis is low (NCI, 2024).

Evidence Discussion

- Imaging strategies that define the extent of primary tumor involvement, particularly skull base invasion and nodal disease, provide essential prognostic information and direct appropriate therapy without requiring excessive serial imaging (NCI, 2024).
- Quantitative EBV DNA levels correlate with tumor burden and treatment response, allowing laboratory monitoring to complement imaging and reduce the need for frequent cross-sectional studies (NCI, 2024).
- Most pediatric NPC patients achieve long-term survival, and unnecessary imaging increases cumulative radiation exposure and risk of late effects without demonstrated improvement in outcomes (NCI, 2024).

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Health Equity Considerations

Pediatric NPC is a rare malignancy with a markedly uneven global distribution, making health equity central to timely diagnosis and optimal outcomes. Population-based analyses demonstrate that the burden of pediatric NPC is disproportionately concentrated in specific geographic and sociodemographic settings, particularly in East and Southeast Asia, North Africa, and parts of Sub-Saharan Africa, while remaining rare in most high income Western regions (Lei et al., 2024; Xu et al., 2025). Although global age standardized incidence rates have shown a modest decline over recent decades, children living in low and low middle sociodemographic index (SDI) regions continue to experience relatively higher incidence and delayed diagnosis, driven by population growth, endemic EBV exposure, environmental and dietary risk factors, and limited access to specialized pediatric care and diagnostic imaging (Lei et al., 2024; Xu et al., 2025). These structural inequities contribute to the frequent presentation of locally advanced disease at diagnosis, a pattern that is more pronounced in resource limited settings.

Socioeconomic and geographic disparities also shape access to guideline-concordant imaging, treatment planning, and survivorship care. Limited availability of advanced imaging, particularly high quality MRI for locoregional staging and evaluation of skull base or intracranial extension, can delay diagnosis and compromise accurate risk stratification in lower SDI regions, while improved imaging infrastructure and multidisciplinary oncology care in high SDI regions have contributed to declining incidence and mortality (Lei et al., 2024; Xu et al., 2025). Sex- and age-based differences, with higher incidence in adolescent males, intersect with these broader inequities and may further increase vulnerability to delayed diagnosis and treatment-related morbidity (Lei et al., 2024). Emphasizing timely access to MRI based staging, standardized imaging pathways, and strengthened referral networks to pediatric centers of expertise has the potential to reduce disparities and support earlier diagnosis, treatment de-escalation when appropriate, and improved long-term outcomes regardless of geographic location or socioeconomic status (Xu et al., 2025).

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Pediatric NPC – Imaging (PEDONC-13.2)

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Criteria

Initial Staging

Indication	Medically Necessary Imaging Study
All individuals with NPC	• MRI Brain without and with contrast (CPT® 70553) and MRI Face/Orbit/Neck without and with contrast (CPT® 70543) ◦ CT Head without and with contrast (CPT® 70470), CT Maxillofacial without and with contrast (CPT® 70488), and/or CT Neck with contrast (CPT® 70491) for individuals with documented contraindication to MRI imaging (avoidance of sedation should not be the sole reason • CT Chest with contrast (CPT® 71260)
To evaluate for distant metastases after histologic confirmation in individuals with NPC	Whole-body PET/CT (CPT® 78816) • Bone scan when PET/CT is unavailable (See: **Modality General Considerations (PEDONC-1.3)**) • Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)** • CT Abdomen with contrast (CPT® 74160) for ANY of the following if PET/CT unavailable: ◦ Initial EBV DNA load >4000 copies/mL ◦ Signs and symptoms of liver disease (including abdominal pain and elevated LFTs)

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Treatment Response

Indication	Medically Necessary Imaging Study
Response assessment in individuals with NPC at the following time points: • Following completion of neoadjuvant chemotherapy • Following completion of chemoradiotherapy	MRI Brain without and with contrast (CPT® 70553) **and** MRI Neck without and with contrast (CPT® 70543)
Individuals with NPC at the following time points: • Following completion of neoadjuvant chemotherapy only if positive at initial diagnosis • Following completion of chemoradiotherapy	CT Chest with contrast (CPT® 71260) and Whole-Body PET/CT (CPT® 78816) or bone scan (See: **Modality General Considerations (PEDONC-1.3)**)
During active treatment for recurrent pediatric cancer in rare circumstances when results are likely to result in a treatment change, including a change from active treatment to surveillance	PET/CT (CPT® 78816)

Surveillance

Indication	Medically Necessary Imaging Study
All individuals with NPC	Every 3 months for 1 year, then every 6 months for 2 years after completion of all planned therapy: • MRI Brain without and with contrast (CPT® 70553) and MRI Neck without and with contrast (CPT® 70543) • CT Chest with contrast (CPT® 71260)

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Suspected Recurrence

Indication	Medically Necessary Imaging Study
All individuals with NPC	• MRI Brain without and with contrast (CPT® 70553) and MRI Neck without and with contrast (CPT® 70543) ◦ CT Head without and with contrast (CPT® 70470), CT Maxillofacial without and with contrast (CPT® 70488), and/or CT Neck with contrast (CPT® 70491) for individuals with documented contraindication to MRI imaging • CT Chest with contrast (CPT® 71260)
• Individuals with histologically confirmed recurrence of NPC • Clarification of specified inconclusive findings seen on conventional imaging • Restaging to identify sites of disease when EBV PCR levels are abnormally high and conventional imaging is negative	Whole-body PET/CT (CPT® 78816) or bone scan (See: **Modality General Considerations (PEDONC-1.3)**)

Evidence Discussion

- MRI of the head and neck is the preferred modality for evaluating the primary tumor, parapharyngeal extension, skull base involvement, and intracranial spread due to superior soft-tissue contrast (NCI, 2024).
- Imaging of the chest and skeleton may be used at initial staging to assess for metastatic disease; however, fewer than 10% of children present with distant metastases at diagnosis (NCI, 2024).
- Evidence does not support routine PET/CT or bone scan surveillance in asymptomatic pediatric NPC survivors, as relapse is uncommon and typically detected clinically or via site-directed imaging (NCI, 2024).
- Pediatric NPC survivors are at risk for significant late effects due to combined chemotherapy and radiotherapy. Limiting radiation-based imaging reduces cumulative exposure without compromising detection of clinically meaningful recurrence (NCI, 2024).
- Selective imaging prompted by symptoms, abnormal examination findings, or rising EBV DNA levels provides a favorable balance of diagnostic yield and harm reduction (NCI, 2024).

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Pediatric Adrenocortical Carcinoma (PEDONC-14)

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Pediatric Adrenocortical Carcinoma – General Considerations (PEDONC-14.1)

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- See: **Li-Fraumeni Syndrome (LFS) (PEDONC-2.2)** and **Beckwith-Wiedemann Syndrome (BWS) (PEDONC-2.4)** for screening recommendations for individuals known to have these syndromes.

Background and Supporting Information

- Pediatric adrenocortical carcinoma (ACC) is a rare malignancy arising from the adrenal cortex, accounting for approximately 0.2%–0.3% of childhood cancers, with an estimated 25 cases diagnosed annually in the United States (NCI, 2025; NCCN, 2025).
- Pediatric ACC is aggressive but biologically distinct from adult ACC, with a spectrum ranging from adenoma to carcinoma and difficulty distinguishing benign from malignant lesions based solely on histology (NCI, 2025; NCCN, 2025).
- Most pediatric ACCs are hormonally functional, and diagnosis is commonly prompted by endocrine manifestations such as virilization, Cushing syndrome, or mixed steroid excess; nonfunctional tumors are uncommon and tend to present later (NCI, 2025; NCCN, 2025).

Evidence Discussion

- Clinical outcomes in pediatric ACC are most strongly associated with completeness of surgical resection rather than frequency of imaging, supporting guideline approaches that prioritize accurate preoperative staging while limiting repetitive imaging when disease is clinically stable (NCI, 2025; NCCN, 2025).
- Because most pediatric ACC tumors secrete hormones, biochemical markers provide sensitive indicators of disease activity and recurrence, reducing the necessity for routine imaging in the absence of clinical or laboratory concern (NCI, 2025; NCCN, 2025).
- The high prevalence of germline TP53 variants in pediatric ACC increases susceptibility to radiation-induced malignancies, supporting conservative imaging strategies that limit cumulative radiation exposure without compromising detection of clinically meaningful disease (NCI, 2025; NCCN, 2025).

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Health Equity Considerations

Pediatric ACC is a rare and aggressive malignancy with substantial global variation in incidence, presentation, and outcomes, making health equity considerations central to diagnostic and imaging guidelines. Although the overall incidence in children is low, ranging from approximately 0.2 to 2 cases per million annually, markedly higher rates occur in specific geographic and genetic clusters, most notably in Southern Brazil due to the high prevalence of germline TP53 p.R337H mutations (Lin et al., 2025; Sandru et al., 2022). Children in these high-risk regions often present at younger ages and with functional tumors, yet outcomes remain closely tied to early detection and access to specialized care. In contrast, in many low and middle income settings, limited awareness, reduced availability of endocrine evaluation, and restricted access to advanced cross-sectional imaging contribute to delayed diagnosis, larger tumor size at presentation, and a higher likelihood of metastatic disease, all of which are associated with poorer survival (Sandru et al., 2022).

Disparities in healthcare infrastructure strongly influence access to guideline-concordant imaging, staging, and longitudinal surveillance for pediatric ACC. Advanced imaging modalities, particularly contrast enhanced CT and MRI for local staging and assessment of vascular invasion, as well as chest imaging for metastatic evaluation, are essential for surgical planning and prognostication but may be inconsistently available outside high resource centers (Lin et al., 2025). Children in underserved regions are also less likely to receive timely genetic testing for hereditary cancer syndromes such as Li-Fraumeni or Beckwith-Wiedemann syndrome, which can guide imaging surveillance strategies and risk-appropriate management for patients and their families (Sandru et al., 2022). Emphasizing standardized staging protocols, early referral to multidisciplinary centers, and integration of imaging with hormonal and genetic evaluation can help mitigate structural disparities and improve the likelihood of complete resection, appropriate use of adjuvant therapy, and long-term survivorship care regardless of geographic or socioeconomic context (Lin et al., 2025; Sandru et al., 2022).

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Pediatric ACC – Imaging (PEDONC-14.2)

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Criteria

- CT Abdomen without and with contrast (CPT® 74170) increases radiation exposure and is not considered medically necessary in a child with an adrenal lesion as washout criteria have not been validated in children.
- Many ACC individuals are treated with surgery alone and do not receive adjuvant therapy. These individuals should be imaged using surveillance guidelines after surgery is completed.

Indication	Medically Necessary Imaging Study
Initial staging in individuals with ACC	• CT Chest without contrast (CPT® 71250) or with (CPT® 71260) contrast • CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183) • Nuclear bone scan (see: **PEDONC-1.3** for coding)
• Solitary adrenal mass >4cm on conventional imaging and plans for aggressive surgical resection in individuals with ACC • Inconclusive findings on conventional imaging in individuals with ACC	• FDG PET/CT scan (CPT® 78815)
After complete resection, with no plans for chemotherapy or radiotherapy in individuals with ACC	• See surveillance below

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Indication	Medically Necessary Imaging Study
Restaging, for all unresected primary or metastatic disease on chemotherapy in individuals with ACC	• Every 2 cycles (~6 weeks) during chemotherapy, and following completion of all planned chemotherapy: ◦ CT Chest without contrast (CPT® 71250) or with (CPT® 71260) contrast ◦ CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183) • If positive for distant metastases at initial diagnosis: ◦ Nuclear bone scan (see: **PEDONC-1.3** for coding) every 2 cycles (~6 weeks) during chemotherapy and following completion of all planned chemotherapy
Response assessment at the completion of radiotherapy in individuals with ACC	• CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183)
Surveillance, individuals with ACC with only localized disease at diagnosis	• Every 3 months for 2 years, then every 6 months for 3 years: ◦ CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183)
Surveillance, individuals with metastatic ACC	• Every 3 months for 2 years, then every 6 months for 3 years: ◦ CT Chest without contrast (CPT® 71250) or with (CPT® 71260) contrast ◦ CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183)
Recurrence in individuals with ACC	• CT Chest without contrast (CPT® 71250) or with (CPT® 71260) contrast • CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183)

Indication	Medically Necessary Imaging Study
Suspected bone recurrence in individuals with ACC	• Nuclear bone scan (see: **PEDONC-1.3** for coding)

Evidence Discussion

- Cross-sectional imaging at diagnosis is essential to characterize tumor size, local invasion, vascular involvement, and metastatic disease, which are critical for staging and surgical planning in pediatric ACC (NCI, 2025; NCCN, 2025).
- Contrast-enhanced CT or MRI of the abdomen is used to evaluate the primary adrenal tumor, while chest imaging is performed based on risk for pulmonary metastases. MRI is often favored in children to reduce radiation exposure (NCI, 2025; NCCN, 2025).
- Although FDG PET can identify metabolically active disease, routine PET imaging in asymptomatic pediatric ACC survivors has not been shown to improve survival or recurrence detection and contributes to unnecessary radiation exposure (NCI, 2025; NCCN, 2025).
- Post-treatment imaging is individualized based on stage, resection status, and clinical findings, with emphasis on symptom-directed imaging rather than scheduled routine whole-body studies (NCI, 2025; NCCN, 2025).

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Pediatric Melanoma and Other Skin Cancers (PEDONC-15)

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Pediatric Melanoma and Other Skin Cancers (PEDONC-15)

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Criteria

- Pediatric melanoma staging is assigned using the American Joint Committee on Cancer (AJCC) staging for adult melanoma.
- Imaging guidelines and treatment approaches are consistent with those used for adults with melanoma and other skin cancers, and these individuals should follow the imaging guidelines in section **Melanomas and Other Skin Cancers (ONC-5)** in the Oncology Imaging Guidelines.

Background and Supporting Information

- Melanoma in pediatric and adolescent/young adult populations is uncommon, and the majority of pediatric melanoma cases are localized at diagnosis, with most tumors amenable to complete surgical excision (NCCN, 2026).
- Pediatric melanoma may present diagnostic challenges due to atypical clinical and histopathologic features compared with adult melanoma, contributing to variability in management and the absence of extensive pediatric-specific prospective trial data (NCCN, 2026).
- Basal cell carcinoma and squamous cell carcinoma are extremely rare in children and are typically associated with predisposing factors such as immunosuppression, prior radiation exposure, chemotherapy, genetic syndromes, or photosensitizing medications (NCCN, 2026).
- Children and adolescents with melanoma or other skin cancers may have underlying genetic or environmental risk factors, including cancer predisposition syndromes, prior malignancy, organ transplantation, or chronic immune suppression (NCCN, 2026).

Evidence Discussion

- Imaging has low diagnostic yield in asymptomatic patients with localized melanoma, supporting imaging strategies that avoid routine staging or surveillance imaging in early-stage pediatric disease (NCCN, 2026).
- Imaging is most clinically useful in patients with higher-risk disease features, clinical symptoms, or suspected nodal or distant involvement, supporting selective imaging approaches (NCCN, 2026).

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- The avoidance of unnecessary imaging is particularly important in pediatric populations due to cumulative radiation exposure and limited evidence that routine imaging improves outcomes in low-risk skin cancers (NCCN, 2026).
- Because pediatric non-melanoma skin cancers are rare and usually localized, imaging is generally unnecessary unless there are aggressive features, recurrence, or clinical suspicion of deeper or metastatic disease (NCCN, 2026).

Health Equity Considerations

Pediatric melanoma and other skin cancers are rare but potentially aggressive malignancies in children and adolescents, with well-documented inequities in incidence, stage at diagnosis, and outcomes across racial, ethnic, and socioeconomic groups. Although melanoma is more common among non-Hispanic White children, multiple population-based studies demonstrate that Hispanic and Black children are significantly more likely to present with thicker tumors, regional or distant disease, and worse survival outcomes, reflecting delays in recognition and diagnosis (Ahuja et al., 2024; Hamilton et al., 2016). These disparities are exacerbated by atypical clinical presentations in children, including small, uniformly pigmented or amelanotic lesions that do not meet traditional ABCDE criteria, leading to frequent misdiagnosis or delayed referral, particularly in skin of color (Ahuja et al., 2024; Kuo et al., 2021). Broader sociodemographic factors also contribute to inequities in pediatric skin cancer risk and detection, including reduced access to preventive counseling on sun protection, lower perceived melanoma risk among racial and ethnic minority families, and variable access to dermatologic expertise across regions with differing sociodemographic index levels (Hamilton et al., 2016; Mackenzie et al., 2026).

Disparities in access to timely dermatologic evaluation, diagnostic biopsy, and appropriate imaging further influence outcomes for pediatric melanoma and other skin cancers. Children from lower socioeconomic backgrounds or those insured through public programs are less likely to receive subspecialty dermatology care, advanced staging procedures such as sentinel lymph node biopsy, or consistent follow-up, which may delay appropriate imaging and treatment escalation for higher risk disease (Hamilton et al., 2016; Kuo et al., 2021). Structural barriers, including limited acceptance of Medicaid by dermatology practices, geographic distance from specialized pediatric oncology and dermatology centers, language barriers, and reduced utilization of teledermatology among non-English speaking families, can disproportionately affect minority populations (Kuo et al., 2021). Providers should emphasize culturally responsive education for families and clinicians, heightened suspicion for atypical lesions in all skin types, and standardized pathways to ensure timely imaging, staging, and referral regardless of race, ethnicity, insurance status, or geographic location. Such approaches are essential to reducing avoidable delays in diagnosis and improving outcomes for children with melanoma and other skin cancers across diverse populations (Ahuja et al., 2024; Mackenzie et al., 2026).

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Pediatric Salivary Gland Tumors and Thyroid Tumors (PEDONC-16)

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Pediatric Salivary Gland Tumors and Thyroid Tumors (PEDONC-16)

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Criteria

Pediatric Salivary Gland Tumors

- American Joint Committee on Cancer (AJCC) staging is used for pediatric as well as adult salivary gland tumors.
- Imaging and treatment guidelines for malignant pediatric salivary gland tumors are consistent with those used for adults with salivary gland tumors, and these individuals should follow the imaging guidelines in section **Salivary Gland Cancers (ONC-4)** in the Oncology Imaging Guidelines.

Pediatric Thyroid Tumors

- Imaging and treatment guidelines for malignant pediatric thyroid tumors are consistent with those used for adults with thyroid tumors, and these individuals should follow the imaging guidelines in section **Thyroid Cancers (ONC-6)** in the Oncology Imaging Guidelines.

Background and Supporting Information

- Pediatric Salivary Gland Tumors
  - Pediatric salivary gland tumors are rare, with the parotid gland being the most common site of origin; tumors of the submandibular, sublingual, and minor salivary glands are less frequent but have a higher likelihood of malignancy (NCCN, 2025).
  - Approximately 75% of pediatric salivary gland tumors are benign, most commonly pleomorphic adenoma; however, malignancy rates are higher in children than in adults (NCCN, 2025).
  - The most common malignant salivary gland tumors in pediatric patients include mucoepidermoid carcinoma, acinic cell carcinoma, and adenoid cystic carcinoma, with tumor grade and histology influencing prognosis and management (NCCN, 2025).
- Pediatric Thyroid Tumors
  - Thyroid carcinoma is the most common endocrine malignancy in children, with differentiated thyroid carcinoma (DTC) accounting for the majority of cases (NCCN, 2025).

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- The most common histologic subtypes of pediatric DTC are papillary thyroid carcinoma, followed by follicular and oncocytic (Hürthle cell) carcinoma; medullary thyroid carcinoma (MTC) represents a smaller but clinically distinct subgroup (NCCN, 2025).
- Pediatric thyroid cancers frequently present with cervical lymph node involvement and, less commonly, pulmonary metastases, yet overall long-term survival remains favorable (NCCN, 2025).
- Risk stratification is based on tumor size, multifocality, extrathyroidal extension, nodal or distant metastases, histologic subtype, and molecular features; these factors guide surgical planning and adjuvant therapy decisions (NCCN, 2025).
- Medullary thyroid carcinoma is frequently associated with germline RET pathogenic variants, and management includes genetic testing, risk-based timing of surgery, and lifelong surveillance (NCCN, 2025).

Evidence Discussion

- Imaging criteria for pediatric salivary gland and thyroid tumors are designed to optimize diagnostic accuracy while minimizing radiation exposure, a critical consideration in children given long-term malignancy risk (NCCN, 2025).
- For salivary gland tumors, cross-sectional imaging (CT or MRI) provides clinically relevant information on tumor margins, deep lobe involvement, nodal disease, and perineural invasion, supporting appropriate surgical planning and reducing the risk of incomplete resection (NCCN, 2025).
- In pediatric thyroid cancer, high-resolution ultrasound is the primary imaging modality for initial staging and surveillance, as it reliably detects thyroid nodules and cervical lymph node metastases without ionizing radiation (NCCN, 2025).
  - Selective use of CT or MRI is supported in cases with suspected locally advanced disease, airway compromise, or mediastinal extension, where ultrasound alone is insufficient (NCCN, 2025).
  - Routine use of advanced imaging in low-risk pediatric thyroid cancer does not improve outcomes and may increase exposure-related harms, supporting a risk-adapted imaging strategy (NCCN, 2025).
  - For intermediate- and high-risk thyroid tumors, imaging that defines the extent of nodal or distant metastases improves surgical planning, supports appropriate use of radioactive iodine, and reduces the likelihood of repeat procedures (NCCN, 2025).
  - In medullary thyroid carcinoma, imaging recommendations combined with genetic risk stratification allow for timely surgical intervention, which is associated with improved biochemical control and reduced recurrence (NCCN, 2025).
- The potential harms of imaging, including radiation exposure, need for sedation, and false-positive findings, are mitigated through guideline-directed indications, reinforcing the balance between diagnostic benefit and patient safety (NCCN, 2025).

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Health Equity Considerations

Pediatric salivary gland tumors and thyroid tumors are rare head and neck malignancies with generally favorable survival; however, substantial inequities exist in incidence patterns, stage at presentation, access to specialty care, and treatment timeliness across racial, ethnic, socioeconomic, and geographic groups. For pediatric thyroid cancer, multiple population-based analyses demonstrate that Hispanic, Black, and other non-White children are more likely to present with larger tumors, higher disease burden, and nodal involvement than non-Hispanic White children, even after controlling for socioeconomic status (SES) (Dream, 2024; Ning et al., 2025; Sharma et al., 2023; Zhao & Wilhelm, 2023). These disparities likely reflect delayed recognition of thyroid nodules, barriers to primary and specialty care access, and differential use of diagnostic imaging and biopsy, particularly in children living in lower income neighborhoods, persistent poverty areas, or medically underserved regions (Ning et al., 2025; Zhao & Wilhelm, 2023). Geographic clustering of pediatric thyroid cancer incidence, with higher rates in the Northeast and lower rates in Southern states, further highlights structural differences in healthcare access, diagnostic intensity, and referral patterns that may influence when and how children receive imaging and definitive evaluation (Ghazy et al., 2026).

Access-related inequities also affect treatment pathways and outcomes. Children with Medicaid insurance or from lower income households frequently travel longer distances to receive thyroid surgery, experience longer wait times from diagnosis to definitive treatment, and have worse long-term survival compared with privately insured peers (Dream, 2024; Zhao & Wilhelm, 2023). Delays in surgical management are associated with increased nodal disease and higher likelihood of adjuvant radioactive iodine exposure, which carries important long-term risks for pediatric patients, including secondary malignancies and chronic health effects (Ning et al., 2025; Zhao & Wilhelm, 2023). Similar equity concerns apply to pediatric salivary gland malignancies, where diagnosis is often delayed due to slow growing, asymptomatic tumors and limited awareness among families and clinicians, particularly in younger children and underserved populations (Coleman et al., 2023). Although overall survival for pediatric major salivary gland cancers exceeds 90%, younger children and those with high stage disease or positive surgical margins experience worse outcomes, emphasizing the need for timely imaging, staging, and referral to experienced multidisciplinary teams (Coleman et al., 2023).

Equity-focused implementation of imaging guidelines for pediatric salivary gland and thyroid tumors should prioritize early and appropriate imaging evaluation for children presenting with concerning neck masses or thyroid abnormalities, irrespective of race, insurance status, or geographic location. Strategies to mitigate disparity include standardized risk-based imaging pathways, improved access to pediatric subspecialty expertise (including telemedicine to reduce travel burdens), culturally and linguistically appropriate patient education, and heightened clinician awareness of atypical presentation patterns in populations historically at risk for delayed diagnosis

(Dream, 2024; Ghazy et al., 2026; Sharma et al., 2023). Ensuring equitable utilization of high quality imaging for diagnosis, staging, and surveillance is essential to reducing preventable delays in care and improving outcomes for all children with salivary gland and thyroid tumors.

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Pediatric Breast Masses (PEDONC-17)

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Pediatric Breast Masses (PEDONC-17)

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Criteria

- Ultrasound (CPT® 76641 and CPT® 76642) is the primary and medically necessary modality used for evaluation of pediatric breast masses.
- MRI has very limited utility in evaluation of pediatric breast masses prior to biopsy but may be considered medically necessary in rare cases for surgical planning when ultrasound is non-diagnostic.
- Pediatric individuals with confirmed breast cancer should be imaged according to section **Breast Cancer (ONC-11)** in the Oncology Imaging Guidelines.

Background and Supporting Information

- Breast masses occur in approximately 2–3% of pediatric and adolescent females, and the clinical presentation, diagnostic approach, and management differ substantially from those in adults due to differences in breast development, tissue density, and malignancy risk (Knaus & Grabowski, 2021).
- Breast malignancy in pediatric patients is exceedingly rare, with an estimated incidence of approximately 0.02% in patients ≤21 years, and most malignant lesions represent metastatic or hematologic disease rather than primary breast carcinoma (Davis et al., 2021; Knaus & Grabowski, 2021).
- Fibroadenoma is the most common pediatric breast lesion, accounting for more than 80–90% of breast masses, and is typically benign, slow-growing, and self-limited (Knaus & Grabowski, 2021; Sanders et al., 2018).
- The American College of Radiology BI-RADS classification system, developed and validated in adults, has not been validated in children and may not accurately reflect malignancy risk when applied to pediatric patients (Davis et al., 2021; Knaus & Grabowski, 2021).
  - Large cohort analyses show that application of BI-RADS categories in pediatric patients significantly overestimates cancer risk, frequently assigning BI-RADS 4 or 5 to lesions that are histologically benign, leading to unnecessary biopsies and procedures (Davis et al., 2021; Knaus & Grabowski, 2021).

Evidence Discussion

- Multiple retrospective series demonstrate that routine advanced imaging or invasive evaluation of pediatric breast masses yields extremely low rates of malignancy, supporting conservative imaging approaches in low-risk patients (Davis et al., 2021; Sanders et al., 2018).

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- Ultrasound is the primary and preferred imaging modality for evaluating pediatric breast masses due to its lack of ionizing radiation and its ability to characterize lesions in dense developing breast tissue (Knaus & Grabowski, 2021; Sanders et al., 2018).
- Mammography is not recommended for routine evaluation of pediatric breast masses, as its sensitivity is limited by dense fibroglandular tissue and the radiation exposure outweighs potential diagnostic benefit in this low-risk population (Knaus & Grabowski, 2021; Davis et al., 2021).
- Observational management of small, benign-appearing breast masses has been shown to be safe, with no increase in missed malignancies, and reduces procedural complications, cost, and psychosocial distress (Sanders et al., 2018; Knaus & Grabowski, 2021).
- Core needle biopsy, while safe when indicated, should be reserved for lesions with concerning clinical features or high-risk history, as overuse may lead to unnecessary intervention in the setting of overwhelmingly benign pathology (Sanders et al., 2018; Davis et al., 2021).

Health Equity Considerations

Pediatric and adolescent breast masses are common clinical findings, most frequently representing benign conditions such as fibroadenomas, and breast malignancy in this population is exceedingly rare. Despite these reassuring epidemiologic features, breast masses can cause significant anxiety for patients and families and prompt diagnostic imaging, biopsy, or surgical intervention. Evidence from a multi-institutional cohort of pediatric patients undergoing excision of breast masses demonstrates that, within academic pediatric centers, most aspects of care, including lesion size at presentation, pathology, and time from initial imaging to surgery, do not differ significantly by race/ethnicity, insurance status, neighborhood income, or urbanicity (Knaus et al., 2022). These findings suggest a relatively equitable pathway to definitive surgical management once pediatric patients enter specialized care. However, disparities in clinical presentation were noted, including higher rates of pain at presentation among non-Hispanic Black patients, which may influence initial clinical decision making and family perceptions of urgency (Knaus et al., 2022).

Importantly, the same study identified a significant disparity in preoperative diagnostic practices; non-Hispanic White patients were substantially more likely to undergo preoperative fine needle aspiration or core needle biopsy compared with non-Hispanic Black patients, even after adjusting for lesion size and other clinical factors (Knaus et al., 2022). Although biopsy did not alter time to surgery or final pathology, this variation highlights potential inequities in diagnostic intensity, differences in provider or family preferences, and uneven exposure to additional procedures that may increase cost, discomfort, or anxiety without clear benefit in the pediatric population. Given growing evidence that most pediatric breast masses can be safely evaluated with targeted

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ultrasound surveillance and that routine biopsy may represent overtreatment, providers should emphasize standardized, evidence-based imaging algorithms and shared decision making that are consistently applied across patient populations (Knaus et al., 2022). Ensuring that imaging and diagnostic strategies minimize unnecessary interventions while remaining accessible and culturally responsive is essential to promoting equitable, high value care for children and adolescents presenting with breast masses.

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Histiocytic Disorders (PEDONC-18)

Guideline Page

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Histiocytic Disorders - General Considerations (PEDONC-18.1)

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Criteria

•
- The Non-Langerhans cell histiocytoses encompass a variety of diseases and have limited imaging considerations except as specified later in this section.
- Where there are indications for PET/CT in these guidelines, please note that indications and coding for rare circumstances where PET/MRI may be approved are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**
- PEDONC-18 applies to individuals of all ages.
- The use of PET in this guideline refers to Fluorodeoxyglucose (fluorine-18-2-fluoro-2-deoxy-D-glucose [FDG]) radiotracer only.

Background and Supporting Information

- Histiocytic disorders are rare hematologic neoplasms characterized by accumulation of clonal myeloid-derived histiocytes with associated inflammatory infiltrate, accounting for <1% of soft tissue and lymph node malignancies (NCCN, 2026).
- These disorders demonstrate marked clinical heterogeneity, ranging from localized indolent disease to disseminated multisystem involvement affecting bone, skin, lymph nodes, liver, spleen, lung, bone marrow, endocrine organs, and the central nervous system (NCCN, 2026).
- Contemporary classification has evolved beyond the traditional Langerhans versus non-Langerhans distinction and now reflects integrated clinical, radiographic, histologic, and molecular features, including grouping into Langerhans-related, cutaneous/mucocutaneous, malignant histiocytoses, RDD, and hemophagocytic syndromes (NCCN, 2026).
- Many histiocytic neoplasms share recurrent activating mutations in the MAPK signaling pathway (e.g., BRAF, MAP2K1/2, KRAS, NRAS, ARAF), supporting their classification as clonal neoplastic disorders with shared biologic mechanisms (NCCN, 2026).

Evidence Discussion

- Imaging is essential for initial characterization of disease extent and detection of multisystem involvement, particularly because histiocytic neoplasms often involve multiple organ systems with variable clinical presentation. Imaging findings are

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frequently nonspecific and overlap with inflammatory, infectious, and neoplastic processes, requiring correlation with clinical presentation, pathology, and molecular testing for accurate diagnosis (NCCN, 2026).

- Evidence supports risk-adapted imaging based on clinical presentation, organ involvement, and disease phase, rather than uniform surveillance imaging across all patients, due to lack of outcome benefit in low-risk or asymptomatic disease (NCCN, 2026).
- Limiting advanced imaging to circumstances where results impact management decisions (e.g., staging, treatment response, suspected progression) reduces unnecessary radiation exposure, incidental findings, and downstream interventions. Over-utilization of imaging, particularly serial whole-body studies, may increase radiation burden and false-positive findings without improving clinical outcomes, supporting selective imaging strategies reflected in guideline criteria (NCCN, 2026).

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Langerhans Cell Histiocytosis (LCH) (PEDONC-18.2)

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Criteria

- This guideline may be used for all ages of individuals.

Initial imaging

Indication	Medically Necessary Imaging Study
Individuals with LCH	Whole-body PET/CT (CPT® 78816) • Whole-body Tc-99m bone scan (CPT® 78306) if PET is unavailable
Suspected pulmonary LCH based on ANY of the following: • Abnormal CXR • Symptoms of pulmonary involvement and normal CXR • Clarification of pulmonary findings on PET/CT	In addition to PET/CT (CPT® 78816), CT Chest with contrast (CPT® 71260) or high-resolution CT Chest without contrast (CPT® 71250)
Individuals with LCH with ANY of the following: • Headaches or visual or neurologic disturbances • Polyuria/polydipsia or other endocrine abnormalities • Skull or craniofacial (including jaw) bone involvement • Otorrhea or hearing loss (CT Temporal Bone may be substituted if requested) • Other signs or symptoms suggesting intracranial involvement, including neurodegeneration syndrome	MRI Brain without and with contrast (CPT® 70553)
Operative planning or image guidance	CTA/MRA Head (CPT® 70496/70544)

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Indication	Medically Necessary Imaging Study
Individuals with LCH with ANY of the following: • Abdominal and/or pelvic signs and symptoms if PET/CT has not been performed or to clarify abnormal abdominal/pelvic findings on PET/CT • Abdominal and/or pelvic findings on ultrasound if PET/CT has not been performed	CT Abdomen and/or Pelvis with contrast (CPT® 74177, 74160, 72193)
Individuals with LCH with ANY of the following: • Elevated liver function tests (usually >5X upper limit of normal) • Abnormalities seen on abdominal ultrasound or CT	MRI Abdomen without and with contrast (CPT® 74183)
Individuals with LCH with ANY of the following: • Vertebral lesions seen on skeletal survey • Clinical symptoms (including back pain) suggesting spinal involvement and negative skeletal survey	MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

Treatment Response

Indication	Medically Necessary Imaging Study
For simultaneous treatment response evaluation with specific documentation showing that both are necessary (i.e., not for purpose of acquiring a PET/MRI) in individuals with LCH	Both PET/CT (CPT® 78816) and CT with contrast **and** MRI without and with contrast

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Indication	Medically Necessary Imaging Study
Restaging for all individuals with LCH	CT and/or MRI and/or PET/CT (CPT® 78816) (use modality that showed disease at initial diagnosis) • After 6-8 weeks of initial therapy • At completion of therapy (approximately 12 weeks) for individuals with persistent disease at 6-8 weeks of initial therapy • After surgical curettage (CT or MRI of involved area, not repeat PET) • After radiation therapy
Following initial phase therapy in individuals with LCH	• Treatment response evaluation of involved sites with CT with contrast or MRI with and without contrast every 3 months while receiving active treatment and at completion of therapy. ◦ PET/CT (CPT® 78816) for inconclusive conventional imaging, if disease was previously only measured/measurable on PET/CT, or for change from active treatment to surveillance ◦ Shorter interval imaging for documented signs or symptoms concerning for disease progression

Surveillance

- Surveillance imaging is determined by areas of disease involvement.
- Individuals with lung, CNS, spleen, bone marrow, and/or liver involvement warrant site-specific diagnostic imaging in addition to PET/CT as outlined below

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Indication	Medically Necessary Imaging Study
Bone involvement in individuals with LCH	• Single site bone disease ◦ Every 3 months for 1 year after completion of therapy: ▪ PET/CT (CPT® 78815 or 78816) **OR** ▪ CT or MRI contrast as requested of involved bony areas • Multifocal bone disease ◦ Every 3 months for 2 years, then no more than annually: ▪ PET/CT (CPT® 78815 or 78816) **OR** ▪ CT or MRI contrast as requested of involved bony areas • Skull or craniofacial (including jaw) bone involvement should be imaged according to CNS involvement section below
Pulmonary involvement in individuals with LCH	• CT Chest with (CPT® 71260) or without contrast (CPT® 71250) at 3 months and 6 months post-completion of therapy to complete 2 years of off-therapy surveillance for any of the following: ◦ Individuals with a history of pulmonary involvement ◦ Individuals with new respiratory or chest symptoms ◦ New findings on CXR **AND** • PET/CT (CPT® 78815 or 78816) at 3 months and 6 months post-completion of therapy, then every 6 months to complete 2 years of off-therapy surveillance, then no more than annually

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Indication	Medically Necessary Imaging Study
CNS involvement in individuals with LCH	• PET/CT (CPT® 78815 or 78816) at 3 months and 6 months post-completion of therapy, then every 6 months to complete 2 years of off-therapy surveillance, then no more than annually **AND** • MRI Brain without and with contrast (CPT® 70553) at 6 weeks, and then every 3 months for 2 years after completion of all therapy for previously documented measurable intracranial lesions ◦ If negative after two years, MRI Brain without and with contrast (CPT® 70553) at 4, 7, and 10 years after completion of all planned therapy • MRI Brain without and with contrast (CPT® 70553) for documented hypothalamic-pituitary dysfunction every 6 months for 2 years and at 4, 7, and 10 years after completion of all planned therapy ◦ MRI at any time for worsening neurologic symptoms • Intraspinal lesions imaged according to the same timeframes as brain imaging using MRI without and with contrast of all involved spine levels

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Indication	Medically Necessary Imaging Study
Liver and/or spleen involvement in individuals with LCH	• For individuals with a history of liver and/or spleen involvement, ONE of the following is considered medically necessary every at 3 months and 6 months post-completion of therapy, then every 6 months to complete 2 years of off-therapy surveillance: ◦ Ultrasound Abdomen (CPT® 76700) ◦ CT Abdomen with contrast (CPT® 74160) ◦ MRI Abdomen without and with contrast (CPT® 74183) **AND** • PET/CT (CPT® 78815 or 78816) at 3 months and 6 months post-completion of therapy, then every 6 months to complete 2 years of off-therapy surveillance
Bone marrow involvement in individuals with LCH	PET/CT (CPT® 78815 or 78816) every 3 months for 2 years, then no more than annually
Suspected recurrence or inconclusive findings on any surveillance imaging in individuals with LCH	All imaging studies considered medically necessary for initial workup including whole-body PET/CT (CPT® 78816)

Background and Supporting Information

- LCH represents the most common histiocytic disorder in children and encompasses a spectrum of disease previously described as histiocytosis X, eosinophilic granuloma, Letterer-Siwe disease, and Hand-Schüller-Christian disease (NCCN, 2026).
- Clinical presentation ranges from single-site bone or skin disease to multisystem disease with involvement of risk organs, including liver, spleen, bone marrow, and CNS (NCCN, 2026).
- Common sites of involvement include bone, skin, pituitary gland, liver, spleen, lung, lymph nodes, and CNS, with skull base or facial bone lesions conferring increased risk for CNS involvement (NCCN, 2026).
- Approximately 50% of pediatric cases harbor BRAF V600E mutations, which are associated with multisystem disease, increased recurrence risk, and neurodegenerative CNS involvement (NCCN, 2026).

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Evidence Discussion

- Evidence supports the use of imaging to establish baseline disease distribution, assess treatment response, and identify reactivation or progression, particularly in patients with multisystem or CNS-risk disease (NCCN, 2026).
- In localized or single-system LCH, studies summarized by NCCN show that observation or limited local imaging is often sufficient, avoiding unnecessary radiation exposure while maintaining excellent outcomes (NCCN, 2026).
- For multisystem or CNS-risk LCH, selective use of FDG-PET/CT, MRI, or organ-specific cross-sectional imaging improves detection of occult disease and guides therapy, with benefits outweighing imaging-related risks in these higher-risk populations (NCCN, 2026).
- Extended or intensive imaging surveillance is considered medially necessary only in subsets such as CNS-involved or recurrent LCH, where relapse rates are higher and imaging findings can precede clinical deterioration (NCCN, 2026).

Health Equity Considerations

LCH is a rare but highly heterogeneous pediatric disease that can range from self limited single system involvement to aggressive multisystem disease with significant morbidity and mortality. This clinical variability creates inherent equity challenges, as timely recognition, comprehensive imaging evaluation, and access to specialized multidisciplinary care are critical determinants of outcome. High-risk multisystem LCH disproportionately affects very young children and is strongly associated with oncogenic mutations such as BRAF p.V600E, which correlate with more aggressive disease, higher relapse rates, and resistance to conventional therapy (Mutti & Picarsic, 2025). Delays in diagnosis or incomplete staging, particularly failure to identify risk organ involvement (liver, spleen, bone marrow) or CNS-risk lesions, may worsen long-term outcomes and exacerbate disparities for patients with limited access to pediatric subspecialty imaging and molecular diagnostics (Mutti & Picarsic, 2025).

Equitable implementation of imaging guidelines for LCH requires consistent access to high quality, age-appropriate imaging across care settings, including whole body skeletal surveys, targeted MRI for CNS risk and neurodegenerative disease, and longitudinal imaging for disease surveillance and treatment response. The evolving role of molecular testing, such as sensitive detection of circulating BRAF p.V600E as a prognostic biomarker, introduces additional equity considerations, as these advanced assays may be less available in resource-limited institutions (Mutti & Picarsic, 2025). Imaging pathways should therefore emphasize standardized baseline and follow-up assessments that do not rely solely on advanced technologies to ensure early detection of high risk features. Incorporating clear criteria for imaging escalation, minimizing unnecessary invasive procedures, and facilitating referral to centers with histiocytosis expertise can help reduce disparities and support equitable outcomes for children with LCH across diverse healthcare environments (Mutti & Picarsic, 2025).

Hemophagocytic Lymphohistiocytosis (HLH) (PEDONC-18.3)

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Criteria

Indication	Medically Necessary Imaging Study
Initial evaluation of HLH	As imaging for this condition is usually done on an urgent basis, ANY or ALL of the following are considered medically necessary: • Ultrasound Abdomen (CPT® 76700) • MRI Abdomen (CPT® 74183) and/or Pelvis (CPT® 72197) without and with contrast • Chest x-ray • CT Chest with contrast (CPT® 71260) • MRI Brain without and with contrast (CPT® 70553) • CTA/MRA Head (CPT® 70496/CPT® 70544) for operative planning or image guidance • CT Sinus without or with contrast (CPT® 70486 or CPT® 70487) if clinical suspicion for sinus disease
If conventional imaging has been completed in individuals with HLH and is unrevealing	Whole-body PET/CT (CPT® 78816) for the purpose of identifying a site for tissue diagnosis of a primary source of infection or malignancy • If a malignancy is identified as the inciting factor for HLH, additional imaging decisions for that malignancy should be based on the appropriate diagnosis-specific guidelines

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Background and Supporting Information

- HLH is a hyperinflammatory syndrome driven by uncontrolled immune activation rather than a primary neoplastic histiocytic proliferation (NCCN, 2026).
- Diagnosis is primarily based on clinical features, laboratory abnormalities, and bone marrow or tissue evaluation, rather than imaging findings (NCCN, 2026).
- Organ dysfunction commonly affects the liver, spleen, bone marrow, lungs, kidneys, and CNS, which may necessitate targeted imaging for evaluation of complications (NCCN, 2026).

Evidence Discussion

- There is no established standard role for routine advanced imaging in the diagnosis or baseline evaluation of HLH, as imaging does not supplant clinical or laboratory criteria (NCCN, 2026).
- NCCN-supported evidence emphasizes that imaging should be problem-focused, used to evaluate suspected organ dysfunction or complications (e.g., CNS involvement, pulmonary compromise), rather than performed in a stepwise or routine surveillance fashion (NCCN, 2026).
- Use of PET/CT is limited to specific scenarios such as identifying occult malignancy or infection when conventional evaluation is inconclusive, balancing diagnostic yield with radiation exposure (NCCN, 2026).
- Avoiding routine PET or serial cross-sectional imaging in HLH reduces exposure to radiation and sedation while maintaining diagnostic accuracy, particularly important in acutely ill pediatric patients who may deteriorate rapidly (NCCN, 2026).

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Non-Langerhans Cell Histiocytoses (PEDONC-18.4)

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Criteria

Juvenile Xanthogranuloma (JXG)

- PET may be considered on a case-by-case basis to evaluate for suspected disease in a particular area despite negative or inconclusive MRI and for treatment response in individuals for whom PET was required to assess extent of disease at diagnosis.

Indication	Medically Necessary Imaging Study
Skin and/or cervical nodes in individuals with JXG	CT with contrast of appropriate area
Systemic JXG with multi-organ involvement	• MRI Brain (CPT® 70553) and/or Orbits (CPT® 70543) without and with contrast • CT Neck (CPT® 70491), Chest (CPT® 71260), and/or Abdomen (CPT® 74160) with contrast • CTA/MRA Head (CPT® 70496/CPT® 70544) as part of operative planning or image guidance

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Rosai-Dorfman Disease (RDD)

Indication	Medically Necessary Imaging Study
Initial imaging in individuals with RDD	• MRI Brain (CPT® 70553) and/or Orbits (CPT® 70543) without and with contrast • CTA/MRA Head (CPT® 70496/CPT® 70544) for operative planning or image guidance • CT Neck (CPT® 70491), Chest (CPT® 71260) and/or Abdomen/Pelvis (CPT® 74177) with contrast if **PET/CT not performed** or to follow up unclear findings in said body areas on PET/CT • CT Sinus without or with contrast (CPT® 70486 or CPT® 70487) if clinical suspicion for sinus disease and **PET/CT not performed** or to follow up unclear sinus findings on PET/CT • Whole-body PET/CT (CPT® 78816) after histologic confirmation of diagnosis
Treatment response in individuals with RDD	Treatment response imaging after 2-3 cycles of systemic therapy during active treatment using any modalities showing disease at diagnosis, including PET/CT • Once PET/CT is negative, conventional imaging every 2-3 cycles and/or at the end of chemotherapy and/or radiation and/or after surgical resection for subsequent restaging. One additional PET/CT is medically necessary at completion of therapy.

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Indication	Medically Necessary Imaging Study
Surveillance in individuals with RDD	PET/CT (CPT® 78816) every 3 months until stabilization of disease (two PET/CT with stable disease status) • Further surveillance imaging of single-site bone disease: ◦ Every 6 months using any modalities showing disease at initial diagnosis (excluding PET/CT) for 1 year ◦ PET/CT is not considered medically necessary for routine surveillance of RDD but is considered medically necessary if conventional imaging is inconclusive for suspected recurrence • Further surveillance imaging of multifocal bone disease: ◦ Every 6 months using any modalities showing disease at initial diagnosis (excluding PET/CT) thereafter ◦ PET/CT is not considered medically necessary for routine surveillance of RDD but is considered medically necessary if conventional imaging is inconclusive for suspected recurrence

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Erdheim-Chester Disease (ECD)

Indication	Medically Necessary Imaging Study
Initial imaging in individuals with ECD	• PET/CT Whole Body (CPT® 78816) ◦ Nuclear bone scan (See: **PEDONC-1.3: Modality General Considerations**) in lieu of PET if requested • MRI Brain without and with contrast (CPT® 70553) and/or MRI Orbits (CPT® 70543) without and with contrast for CNS symptoms, including diabetes insipidus • CTA/MRA Head (CPT® 70496/70544) for operative planning or image guidance • CT Neck (CPT® 70491), Chest (CPT® 71260) and/or Abdomen/Pelvis (CPT® 74177) with contrast **if PET/CT not performed** or if inconclusive findings in said body area on PET/CT • Cardiac MRI without and with contrast (CPT® 75561) for clinically suspected cardiac involvement ◦ If Cardiac MRI is not available, CT Chest with contrast (CPT® 71260) and/or Echocardiogram (TTE) (CPT® 93306, 93307, or 93308) • CT Sinus without or with contrast (CPT® 70486 or CPT® 70487) if clinical suspicion for sinus disease **if PET/CT not performed** or inconclusive sinus findings on PET/CT

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Indication	Medically Necessary Imaging Study
Treatment response in individuals with ECD	• Treatment response imaging every 3 months during active treatment using any modalities showing disease at initial diagnosis, including PET/CT • Once PET/CT shows no remaining FDG-avid lesions, additional PET imaging is only considered medically necessary when conventional imaging studies are inconclusive and acute treatment decisions will be made based on PET results
Surveillance in individuals with ECD	• CT **and/or** MRI **and/or** Nuclear bone scan **and/or** CTA **and/or** MRA **and/or** Cardiac MRI (if modality showed disease at initial diagnosis) every 3 months until the first year after completion of treatment and then every 6 months • PET/CT (CPT® 78816) if conventional imaging is inconclusive for suspected recurrence

Background and Supporting Information

- Non-Langerhans histiocytoses include juvenile xanthogranuloma (JXG), Rosai-Dorfman disease (RDD), and Erdheim-Chester disease (ECD), each with distinct clinical behavior and imaging requirements (NCCN, 2026).
- JXG is typically a benign, self-limited cutaneous disorder in children, often regressing spontaneously without systemic involvement (NCCN, 2026).
- RDD classically presents with bulky lymphadenopathy, frequently cervical, and may include extranodal disease affecting soft tissue, bone, or CNS (NCCN, 2026).
- ECD is an aggressive multisystem histiocytic neoplasm, characterized by long bone involvement and frequent spread to cardiovascular, retroperitoneal, pulmonary, and CNS structures (NCCN, 2026).

Evidence Discussion

- For JXG and uncomplicated RDD, evidence supports limited or no routine advanced imaging in asymptomatic patients, as outcomes are excellent and imaging rarely alters management (NCCN, 2026).

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- In RDD with symptomatic or extranodal involvement, targeted imaging is beneficial for defining disease burden and guiding therapy, while routine long-term surveillance imaging in asymptomatic patients provides minimal added benefit (NCCN, 2026).
- In ECD, robust evidence summarized by NCCN supports systemic imaging for staging and treatment response, particularly using FDG-PET/CT early in therapy; however, once metabolic remission is achieved, routine repeat PET imaging is not considered medically necessary unless results will directly influence management decisions (NCCN, 2026).
- This risk-adapted imaging approach minimizes cumulative radiation exposure while preserving the ability to detect clinically meaningful progression in aggressive non-Langerhans histiocytoses (NCCN, 2026).

Health Equity Considerations

Non-Langerhans cell histiocytoses (non LCH), including juvenile xanthogranuloma (JXG), Rosai Dorfman disease (RDD), and Erdheim Chester disease (ECD), encompass a spectrum of rare disorders with widely variable clinical behavior, ranging from self limited cutaneous disease to progressive, multisystem involvement with long-term morbidity. Equity challenges arise from the rarity and phenotypic overlap of these conditions, which frequently leads to delayed or incorrect diagnoses, particularly outside of specialized centers. Pediatric ECD is especially vulnerable to diagnostic delay due to its pathologic similarity to systemic JXG and mixed ECD/LCH presentations, as well as its historically adult-centric disease profile (Pegoraro et al., 2023; Mutti & Picarsic, 2025). Missed or delayed recognition can postpone appropriate whole body imaging and molecular characterization, increasing the risk of irreversible organ damage, including central nervous system, endocrine, renal, and orbital complications that disproportionately impact long-term quality of life in children (Pegoraro et al., 2023).

Population-based data further suggest that disparities in case identification and access to specialized imaging and molecular diagnostics may exist across racial, ethnic, and socioeconomic groups. Recent U.S. registry analyses demonstrate higher reported incidence rates of ECD among non-Hispanic Black, Hispanic, and Asian or Pacific Islander individuals, yet also highlight potential underrepresentation of lower income populations, likely reflecting barriers to advanced imaging, biopsy logistics, and subspecialty referral rather than true disease prevalence (Tessier et al., 2026). Because non-LCH disorders often require multimodality imaging, such as MRI for CNS and pituitary involvement, CT or PET for systemic disease mapping, and longitudinal surveillance, equitable imaging guidelines should emphasize standardized indications for comprehensive staging regardless of initial biopsy site or apparent disease localization. Clear guidance on when to pursue whole body imaging and referral to histiocytosis expertise centers is essential to reduce diagnostic inequities, avoid fragmented care, and ensure that children with non-LCH histiocytoses receive timely,

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consistent, and high-quality evaluation across diverse healthcare settings (Mutti & Picarsic, 2025; Pegoraro et al., 2023; Tessier et al., 2026).

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Long Term Pediatric Cancer Survivors (PEDONC-19)

Guideline Page

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Long Term Pediatric Cancer Survivors – General Considerations (PEDONC-19.1)

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Criteria

- This section applies to individuals who have passed the end of the surveillance imaging period for their specific cancer, or 5 years after completion of therapy, whichever occurs first.
- As these are long-term survivors, many individuals falling under this guideline section will have reached adult age. However, these guidelines relate specifically to late effects of childhood cancer treatment and should be applied to all long-term childhood cancer survivors regardless of current age.
- The Children's Oncology Group has published comprehensive guidelines for the management of long-term childhood cancer survivors, and these are available at: http://www.survivorshipguidelines.org.
- A summary of cancer treatment should be available for all individuals in this category and should generally include, at minimum:
  - type of cancer and stage
  - dates of diagnosis, recurrence, cancer-related surgeries, beginning and end dates of chemotherapy, radiotherapy, and/or stem cell transplant
  - protocol number used for treatment and cumulative chemotherapy drug dose exposures
  - cumulative radiation dose, fraction number, modality, and field exposure
- Annual detailed history and complete physical examination is a critical component of cancer survivorship care and along with laboratory testing serves as the primary method of screening for the majority of late effects.
- Advanced imaging for asymptomatic screening is not routinely considered medically necessary except as specified in this section.
- Imaging requests related to new clinical signs or symptoms in a long term cancer survivor not explicitly covered in this section should be reviewed according to the guideline for the individual's cancer type or the relevant non-malignant clinical problem.

Background and Supporting Information

- Advances in pediatric oncology have resulted in a growing population of long-term childhood cancer survivors, many of whom experience late effects related to prior

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chemotherapy, radiation therapy, surgery, and transplant exposures (Bashore & Hobbie, 2021; Chow et al., 2025).
- Late effects may involve cardiovascular, musculoskeletal, endocrine, neurologic, and oncologic systems, often emerging years to decades after completion of therapy (Vrooman et al., 2018; Chow et al., 2025).
- Survivorship care is guided by risk-based follow-up models, most notably the Children's Oncology Group Long-Term Follow-Up Guidelines, which tailor surveillance intensity to treatment exposures and patient-specific risk factors (Children's Oncology Group, 2023).

Evidence Discussion

- Cohort studies and guideline reviews consistently demonstrate that targeted surveillance improves early detection of clinically significant late effects, including cardiomyopathy, second malignant neoplasms, osteonecrosis, and cerebrovascular disease (Chow et al., 2025; Weil et al., 2023).
- Conversely, routine imaging of asymptomatic survivors without defined risk factors has not shown clear outcome benefit and may increase radiation exposure, incidental findings, anxiety, and downstream testing (Bashore & Hobbie, 2021; Chavhan et al., 2016).
- Evidence supports a balanced, risk-stratified imaging approach, aligning imaging frequency and modality with known late-effect risks rather than uniform lifelong surveillance (Children's Oncology Group, 2023; Chow et al., 2025).

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Cardiotoxicity and Echocardiography (PEDONC-19.2)

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Criteria

- Screening echocardiography (CPT® 93306, CPT® 93307, or CPT® 93308) is considered medically necessary for life after exposure to anthracycline chemotherapy, cardiotoxic immunotherapy, or cardiac exposure to radiotherapy

MEDICALLY NECESSARY SCREENING ECHOCARDIOGRAM INDICATIONS			
Age at Time of Exposure	**Cumulative Doxorubicin Equivalent Dose**	**Cumulative Radiation Dose to Cardiac Muscle**	**Echocardiogram Frequency**
All ages	None	None	None
0-0.99 years	≥250 mg/m2	None	Annual
	0-249 mg/m2	Any dose	Annual
	0-249 mg/m2	None	Every 2 years
1-4.99 years	≥250 mg/m2	Any dose	Annual
	0-249 mg/m2	15+ Gy	Annual
		0-14.99 Gy	Every 2 years
	None	35+ Gy	Annual
		15-34.99 Gy	Every 2 years
		0-14.99 Gy	Every 5 years
5+ years	≥250 mg/m2	Any dose	Every 2 years

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MEDICALLY NECESSARY SCREENING ECHOCARDIOGRAM INDICATIONS			
Age at Time of Exposure	**Cumulative Doxorubicin Equivalent Dose**	**Cumulative Radiation Dose to Cardiac Muscle**	**Echocardiogram Frequency**
	0-249 mg/m2	15+ Gy	Every 2 years
		0-14.99 Gy	Every 5 years
	None	35+ Gy	Every 2 years
		15-34.99 Gy	Every 5 years
		0-14.99 Gy	None
All ages with known ventricular dysfunction			Annual

- Stress echocardiography is not considered medically necessary as a screening study for anthracycline cardiotoxicity in the absence of coronary artery disease symptoms. See **Stress Testing with Imaging – Indications (CD-1.4)** for imaging guidelines.
- Female cancer survivors who are pregnant or planning to become pregnant:
  - If any of the following are present, echocardiogram is considered medically necessary as a baseline exam and in the 3rd trimester, and as clinically indicated for symptoms (see **Pregnancy – Maternal Imaging (CD-11.4)** in the Cardiology Imaging Guidelines) if ANY of the following are present:
    - Anthracycline or cardiotoxic chemotherapy/immunotherapy exposure
    - Chest radiotherapy

Background and Supporting Information

- Exposure to anthracycline chemotherapy and chest-directed radiation is a well-established risk factor for late-onset cardiomyopathy and heart failure in childhood cancer survivors (Sofia et al., 2021; Lim et al., 2025).
- Cardiac risk is influenced by age at treatment, cumulative anthracycline dose (doxorubicin-equivalent mg/m²), and presence of cardiac radiation exposure (Mansoor & Ibrahim, 2025; Sofia et al., 2021).

Evidence Discussion

- Echocardiography is the primary modality for detecting asymptomatic left ventricular dysfunction and monitoring progression over time (Sofia et al., 2021; Lim et al., 2025).

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- Long-term cohort studies demonstrate that subclinical cardiac dysfunction frequently precedes symptomatic heart failure, supporting surveillance echocardiography in at-risk survivors (Sofia et al., 2021; Lim et al., 2025).
- Early identification of reduced ventricular function allows for timely cardioprotective interventions, which may delay or prevent progression to overt heart failure (Mansoor & Ibrahim, 2025).
- Routine echocardiography in survivors without cardiotoxic exposures has not demonstrated benefit and risks overuse, reinforcing exposure-based surveillance principles (Graham et al., 2023; Lim et al., 2025).

Health Equity Considerations

Cardiotoxicity is a leading contributor to late morbidity and mortality among pediatric cancer survivors, yet access to timely cardiovascular surveillance, including echocardiography, remains uneven across populations. Survivors exposed to anthracyclines, chest-directed radiation, and newer targeted or immune-based therapies face lifelong risk for cardiomyopathy, arrhythmias, valvular disease, and heart failure, with cumulative incidence approaching 20% by 30 years after diagnosis (Mansoor& Ibrahim, 2025; Lim et al., 2025). Evidence indicates that racial and ethnic minoritized survivors, particularly non-Hispanic Black and Hispanic individuals, experience a higher burden of modifiable cardiovascular risk factors such as hypertension, obesity, and diabetes, which compound treatment-related cardiac injury (Graham, Sounderajah, & Johnson, 2023). These risks are further amplified by structural barriers, including limited access to specialty cardio-oncology care, inconsistent survivorship follow-up, insurance instability, and underrecognition of late effects in primary care, which can delay echocardiographic screening and early intervention (Graham et al., 2023; Lim et al., 2025).

Equitable implementation of cardiotoxicity surveillance guidelines requires consistent application of risk-based echocardiography protocols regardless of race, socioeconomic status, or geographic location. Updated recommendations from international and national groups emphasize stratified surveillance based on cumulative anthracycline dose, radiation exposure, age at treatment, and additional risk modifiers, with echocardiography remaining the cornerstone imaging modality for both clinical and subclinical dysfunction (Lim et al., 2025). However, disparities persist in adherence to these guidelines, with socially disadvantaged survivors more likely to be lost to follow-up and less likely to receive recommended imaging over time (Graham et al., 2023). Providers should therefore prioritize standardized surveillance pathways, clear transition planning from pediatric to adult care, and proactive management of modifiable cardiovascular risk factors. Integrating echocardiography into accessible survivorship care models and pairing imaging with education, care coordination, and culturally responsive support can help mitigate inequities and ensure that all pediatric cancer

survivors benefit from early detection and prevention of cardiotoxicity (Mansoor & Ibrahim, 2025; Lim et al., 2025).

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Second Malignant Neoplasms (SMN) (PEDONC-19.3)

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Criteria

Screening for Breast Cancer in Individuals with a History of a Prior Malignancy

Indication	Medically Necessary Imaging Study
Individuals who received therapeutic radiation exposure in the following fields while they were under 30 years of age: • Chest (thorax) • Whole lung • Mediastinal • Axilla • Mini-mantle, mantle, or extended mantle • Total (TLI) or subtotal (SLTI) lymphoid irradiation • Total body irradiation (TBI)	Clinical breast exam every 6 months supplemented with the following: • MRI Breast (CPT® 77049) annually and annual mammogram beginning at age 25 or 8 years after completion of radiotherapy (whichever occurs later - screening breast MRI is not considered medically necessary prior to age 25)

Evaluation and Screening for CNS Tumors in Individuals with a History of a Prior Malignancy

Indication	Medically Necessary Imaging Study
Individuals with NF1 or NF2	MRI Brain without and with contrast (CPT® 70553) every 2 years after completion of radiotherapy
Individual with history of brain radiotherapy and new neurologic symptoms, including simple headache	MRI Brain without and with contrast (CPT® 70553)

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Indication	Medically Necessary Imaging Study
Individual with history of spine radiotherapy and new neurologic symptoms including change in quality of pain	MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), and Lumbar Spine (CPT® 72158) without and with contrast • MRI Spine with contrast only (Cervical-CPT® 72142, Thoracic-CPT® 72147, Lumbar-CPT® 72149) if being performed immediately following a contrast-enhanced MRI Brain
Individuals with history of brain radiotherapy and persistent neurologic symptoms	MRI Brain without and with contrast (CPT® 70553) annually
Individuals with history of spine radiotherapy and persistent neurologic symptoms	MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), and Lumbar Spine (CPT® 72158) without and with contrast • MRI Spine with contrast only (Cervical-CPT® 72142, Thoracic-CPT® 72147, Lumbar-CPT® 72149) if being performed immediately following a contrast-enhanced MRI Brain

Screening for Colorectal Cancer in Individuals with a History of a Prior Malignancy

Indication	Medically Necessary Imaging Study
Individuals with ≥30Gy radiation exposure to the following fields: • Thoracic, Lumbar, Sacral, or Whole Spine • Abdomen • Pelvis • Total body irradiation (TBI)	Colonoscopy is recommended every 5 years beginning at age 30 or 5 years after radiation exposure (whichever is later)

Indication	Medically Necessary Imaging Study
Individuals with: • Personal history of ulcerative colitis, GI malignancy, adenomatous polyps, or hepatoblastoma • Familial polyposis ◦ Family history of colorectal cancer or polyps in a first degree (parent or sibling) relative	Colonoscopy is recommended every 5 years beginning at age 30 or 5 years after radiation exposure (whichever is later)

Background and Supporting Information

- Childhood cancer survivors have a substantially increased lifetime risk of second malignant neoplasms, related to prior radiation, chemotherapy, genetic susceptibility, and younger age at exposure (Chao et al., 2019; Becktell et al., 2025).
- Common SMNs include breast, CNS, hematologic, thyroid, and colorectal malignancies, with risk patterns influenced by treatment fields and dose (Hornet et al., 2026; Shah et al., 2026).
- Survivorship screening recommendations are largely derived from COG and NCCN guidelines, emphasizing modality-specific screening for high-risk populations (Children's Oncology Group, 2023; NCCN, 2026).

Evidence Discussion

- Population-based studies show that risk-adapted surveillance improves early detection of treatable SMNs, particularly breast and colorectal cancers in radiation-exposed survivors (Wong et al., 2023; Becktell et al., 2025).
- For CNS tumors, evidence does not support routine imaging surveillance in asymptomatic survivors with normal neurologic examinations, except in defined genetic or high-risk groups (Co et al., 2019; Shah et al., 2026).
- While the American Cancer Society recently added computed tomographic colonography (CTC) (AKA "Virtual Colonoscopy") as an acceptable option for colorectal cancer screening of average-risk adults, the National Comprehensive Cancer Network and United States Preventive Services Task Force concluded that data was too premature to warrant its use in screening. Colonoscopy remains the preferred screening modality for survivors at highest risk of colorectal cancer (NCCN, 2026; Wong et al., 2023).

Health Equity Considerations

SMNs represent one of the most consequential late effects of pediatric and adolescent cancer therapy, accounting for a substantial proportion of non-relapse-related mortality

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among long-term survivors. Population-based and survivor cohort studies consistently demonstrate that childhood, adolescent, and young adult cancer survivors experience a two- to six-fold higher risk of developing SMNs compared with the general population, with cumulative incidence approaching 15–20% at 30 years in high-risk groups (Becktell et al., 2025; Chao et al., 2019; Hornet et al., 2026; Shah et al., 2026). Importantly, SMN risk and outcomes are not equitably distributed. Survivors exposed to higher-intensity therapies, such as chest or cranial radiation, alkylating agents, or hematopoietic stem cell transplantation, are disproportionately affected, and these exposures historically vary by treatment era, diagnosis, and access to specialized pediatric oncology care (Becktell et al., 2025; Hornet et al., 2026). Racial and ethnic disparities in SMN outcomes have also been observed, with evidence of poorer overall survival following SMN diagnosis among minoritized populations, even after adjustment for cancer type and stage, suggesting the influence of systemic factors beyond biology alone (Brown et al., 2020; Chao et al., 2019).

Equitable implementation of imaging surveillance for SMNs requires recognition that early detection is highly dependent on consistent access to long-term survivorship care, adherence to exposure-based screening guidelines, and timely diagnostic imaging when signs or symptoms arise. Survivors from lower socioeconomic backgrounds, those who experience gaps during transition from pediatric to adult care, and those without stable insurance coverage are more likely to be lost to follow-up and less likely to receive recommended surveillance imaging, increasing the likelihood of later-stage SMN detection and worse outcomes (Becktell et al., 2025; Hornet et al., 2026). Providers should emphasize standardized, risk-adapted surveillance pathways that are applied consistently across care settings and populations, with particular attention to high-risk groups such as survivors treated at young ages, females exposed to chest radiation, and patients with underlying cancer predisposition syndromes (Shah et al., 2026). Incorporating clear indications for imaging based on treatment exposures rather than demographic or access-related factors can help mitigate inequities and support earlier SMN detection, which has been shown to improve survival and reduce treatment-related morbidity (Brown et al., 2020; Chao et al., 2019).

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Osteonecrosis in Long Term Cancer Survivors (PEDONC-19.4)

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Criteria

- See: **Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)** for information on imaging osteonecrosis in ALL during active treatment.

Indication	Medically Necessary Imaging Study
Prior to advanced imaging in individuals with suspected osteonecrosis	Plain films of symptomatic areas
Individuals with osteonecrosis with symptoms to suggest bone density issues	DEXA or Quantitative CT screening • DEXA or Quantitative CT screening is not considered medically necessary until age 18 unless a surgery, core decompression, or initiation of osteoporosis drugs will be planned based on the imaging results.
Advanced imaging is necessary for acute management decisions in individuals with osteonecrosis	MRI without contrast of the affected area(s)

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Indication	Medically Necessary Imaging Study
Surveillance imaging in individuals with osteonecrosis	Surveillance imaging of asymptomatic individuals to detect osteonecrosis is not considered medically necessary as it has not been shown to impact individual outcomes, and it is not standard to alter treatment based on imaging findings alone without symptoms • Follow-up MRI of incidentally discovered osteonecrosis findings in asymptomatic individuals has not been shown to impact individual outcomes and is not considered medically necessary • For known osteonecrosis with articular collapse on other imaging, CT without contrast of area of interest is considered medically necessary for surgical planning • Repeat advanced imaging may be considered on a case-by-case basis for individuals with osteonecrosis with specific documentation regarding how the advanced imaging will change current individual management

Background and Supporting Information

- Osteonecrosis is a recognized late effect of corticosteroid, chemotherapy, and radiation exposure, hematologic malignancies, and stem cell transplantation, with higher risk during adolescence (Rao et al., 2019; Yang et al., 2021).
- The hip, knee, shoulder, and ankle are most commonly affected, and disease may be asymptomatic in early stages (Kelly & Pottenger, 2022; Marcucci et al., 2019).
- Osteoradionecrosis of the jaw can occur in individuals receiving radiotherapy to the mandible or maxilla; those receiving ≥40Gy are at highest risk. Although unusual, it can also occur in any bone without symptoms. It is rare in other disease types (Rao et al., 2019; Yang et al., 2021).

Evidence Discussion

- Observational studies demonstrate that early MRI detection of osteonecrosis correlates with improved joint preservation options, whereas delayed diagnosis is

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associated with collapse and need for surgical intervention (Rao et al., 2019; Expert Panel on Musculoskeletal Imaging, 2022).

- Routine screening MRI in asymptomatic survivors without risk factors is not considered medically necessary and may lead to overdiagnosis and unnecessary follow-up (Marcucci et al., 2019).
- Evidence supports symptom-directed or risk-based imaging, optimizing benefit while minimizing unnecessary imaging burden (Kelly & Pottenger, 2022).

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CNS Vascular Changes in Pediatric Cancer Survivors Following CNS Radiation (PEDONC-19.5)

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Criteria

Indication	Medically Necessary Imaging Study
All individuals of any age with new neurologic symptoms or headache and a history of cranial irradiation	MRI Brain without and with contrast (CPT® 70553) and MRA Head (CPT® 70544, 70545, or 70546)
Individuals with a history of cranial irradiation and any of the following additional risk factors: • Down syndrome, sickle cell disease, or neurofibromatosis 1 or 2 • Parasellar or suprasellar tumors (i.e., craniopharyngioma) • Radiation dose >50Gy • Radiation field involving Circle of Willis	MRI Brain without and with contrast (CPT® 70553) and MRA Head (CPT® 70544, 70545, or 70546) annually for 10 years post-treatment

Background and Supporting Information

- Cranial radiation therapy in childhood is associated with long-term cerebrovascular complications, including large-vessel arteriopathy, cavernous malformations, moyamoya-like changes, and ischemic stroke These complications may occur months to years after radiation exposure (Nordstrom et al., 2018; Reulen et al., 2021).
- Radiation-induced cavernomas are common on long-term MRI but often remain clinically silent (Becker et al., 2021; Di Giannatale et al., 2014).
- Vascular injury risk increases with younger age at exposure, higher radiation dose, and longer follow-up duration (van Dijk et al., 2016; Waxer et al., 2024).

Evidence Discussion

- Long-term cohort data demonstrate increased rates of stroke and cerebrovascular disease in cranially irradiated survivors, supporting imaging evaluation when neurologic symptoms or signs emerge (Reulen et al., 2021; van Dijk et al., 2016).

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- Studies of radiation-induced cavernomas indicate that routine MRI surveillance in asymptomatic survivors rarely changes management, as most lesions remain stable and clinically insignificant (Becker et al., 2021; Dallo Gallo et al., 2023).
- Evidence supports symptom-driven neurovascular imaging, balancing early detection of significant arteriopathy with the harms of routine surveillance imaging, including incidental findings and resource utilization (Waxer et al., 2024; Nordstrom et al., 2018).

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Hematopoietic Stem Cell Transplantation (HSCT) (PEDONC-20)

Guideline Page

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General Considerations (PEDONC-20.0)

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Background and Supporting Information

- Hematopoietic stem cell transplantation (HSCT) is a therapeutic procedure involving infusion of hematopoietic stem cells to reconstitute the recipient's bone marrow and immune system after preparative conditioning with chemotherapy and/or radiation therapy (PDQ®, 2024).
- HSCT is broadly categorized as allogeneic (donor-derived stem cells) or autologous (patient-derived stem cells), with distinct goals, immune effects, and complication profiles (PDQ®, 2024; Chan et al., 2023).
  - Allogeneic ("allo"): The donor and recipient are different people, and there are multiple types depending on the source of the stem cells and degree of match between donor and recipient. This is most commonly used in diseases originating in the hematopoietic system, such as leukemias and lymphomas, and bone marrow failure syndromes or metabolic disorders. The goal is to replace the hematopoietic and immune system with healthy donor cells to treat the disease. Common types are:
    - Matched sibling donor (MSD or MRD): Donor and recipient are full siblings and HLA-matched
    - Matched unrelated donor (MUD): Donor and recipient are HLA matched but not related to each other
    - Cord blood: Donor stem cells come from frozen umbilical cord blood not related to the recipient, sometimes from multiple different donors at once
    - Haploidentical transplant (haplo): Donor is a half-HLA match to the recipient, usually a parent
  - Autologous ("auto"): The donor and recipient are the same person. The process involves delivery of high dose chemotherapy that is ablative to the bone marrow, followed by an infusion of one's own harvested stem cells. This is primarily done in the context of solid tumors and the stem cells mainly rescue hematopoiesis to facilitate high-dose chemotherapy.
- Allogeneic HSCT is associated with prolonged immunosuppression due to immune reconstitution and preventive or therapeutic immunosuppression for graft-versus-host disease (GVHD), resulting in sustained vulnerability to infectious and noninfectious complications (PDQ®, 2024; Chan et al., 2023).
- Imaging in HSCT is not directed at graft function itself, but rather at baseline assessment, detection of occult disease, and evaluation of peri-transplant and post-transplant complications affecting multiple organ systems (Chan et al., 2023).

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- Complications vary by transplant type, conditioning regimen, and time relative to transplantation, with early complications predominated by infection and toxicity and later complications dominated by chronic GVHD, organ dysfunction, and secondary malignancies (Chan et al., 2023; PDQ®, 2024).

Evidence Discussion

- Expert consensus and observational evidence support selective, indication-based imaging in HSCT patients rather than routine whole-body surveillance, reducing unnecessary radiation exposure and downstream testing without compromising detection of clinically meaningful complications (Chan et al., 2023).
- The heterogeneity of HSCT indications and complication risks necessitates imaging approaches that are risk-adapted and symptom-driven, balancing early complication detection with the cumulative burden of imaging in a medically fragile population (PDQ®, 2024; Chan et al., 2023).
- Over-imaging in immunocompromised patients may result in incidental findings that prompt invasive procedures or prolonged antimicrobial therapy without demonstrated outcome benefit (Chan et al., 2023).
- The clinical evidence supports emphasis on organ-system-specific imaging, aligned to anticipated complication patterns and informed by transplant type, clinical status, and timing relative to transplant (Chan et al., 2023; PDQ®, 2024).

Health Equity Considerations

Equitable access to HSCT in pediatric populations is shaped by biologic risk, healthcare system structure, and social determinants of health. Population-based studies from publicly funded healthcare systems suggest that when financial barriers are minimized, receipt of HSCT is largely driven by disease-related factors rather than race or neighborhood socioeconomic status. In a national Canadian cohort of children with ALL, race and neighborhood income quintile were not associated with receipt of HSCT; however, diagnosis at a transplant-performing center and greater residential distance from the treating center were modestly associated with increased odds of transplantation, indicating potential influences of referral patterns and center-level practices (Truong et al., 2019).

Despite relatively equitable access to HSCT, disparities in post-transplant outcomes remain evident. U.S. registry data demonstrate that pediatric patients with malignant disease residing in high-poverty neighborhoods experience higher transplant-related mortality, and those insured by Medicaid have inferior overall survival compared with privately insured peers, even after adjustment for clinical characteristics (Bona et al., 2021). These findings suggest that household-level socioeconomic constraints and post-transplant support needs may contribute to outcome disparities independent of access to transplant. Biologic sex also intersects with equity considerations: although

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males are more likely to present with higher-risk disease and require HSCT, sex-based differences in survival, relapse, and transplant-related complications are attenuated following transplantation, potentially reflecting the impact of conditioning regimens and hormonal suppression on pre-existing biologic disparities (De Nardi et al., 2022).

Health equity considerations extend beyond transplant recipients to include pediatric donors, who are most often siblings. Longitudinal studies indicate that pediatric donors experience transient reductions in physical and emotional functioning, with recovery over time; however, psychosocial follow-up and donor surveillance are inconsistently implemented due to resource and infrastructure limitations (Hou et al., 2023). Variability in donor support services represents an additional equity concern, particularly for families facing socioeconomic, geographic, or logistical barriers to longitudinal care. Recognition of these factors is important when interpreting imaging utilization, monitoring for transplant-related complications, and planning comprehensive, equitable care pathways for pediatric patients undergoing HSCT and their families.

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Pre-Transplant Imaging (PEDONC-20.1)

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Criteria

- Disease status assessment is considered medically necessary per individual disease guidelines if it has been 6 weeks or more between end of treatment imaging and HSCT or if there are signs and symptoms of disease progression.

Indication	Medically Necessary Imaging
Immediate pre-transplant period - screening for active or occult infection	• CT Chest with contrast or CT Chest without contrast (CPT® 71250 or CPT® 71260) ◦ CT Abdomen with contrast (CPT® 74160) for asymptomatic individuals if chest infection or disease is identified on CT Chest or for signs and symptoms of active abdominal infection including fever. ◦ For evaluation of pelvis signs or symptoms, CT Abdomen/Pelvis with contrast (CPT® 74177) is considered medically necessary. If diagnostic abdominal imaging has already been performed and only pelvic imaging is required, CT Pelvis with contrast (CPT® 72193) may be performed instead. • CT Sinus (CPT® 70486) is not considered medically necessary for screening prior to HSCT, but is considered medically necessary for signs or symptoms of infectious or disease involvement of the sinuses • For individuals receiving CAR-T, if required by institutional protocol, baseline MRI Brain without and with contrast (CPT® 70553)

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Indication	Medically Necessary Imaging
Individuals pre-transplant at risk of developing VOD, for pre-transplant baseline	• Abdominal ultrasound +/- Doppler (CPT® 76700 or 76705 +/- 93975)
Individuals pre-transplant with history of multiple blood transfusions at risk of iron overload, for iron quantification baseline	• CPT® codes as listed in **Transfusion-Associated (Secondary) Hemochromatosis (PEDAB-18.2)** in the immediate pre-transplant period even if done in the previous 12 months.
Suspected renal tubular dysfunction in individuals pre-transplant	• Nuclear renal imaging (CPT® 78700, CPT® 78701, CPT® 78707, CPT® 78708, CPT® 78709)

Background and Supporting Information

- Pre-transplant imaging serves to identify active or occult infection, establish baseline organ assessments, and evaluate transplant-related risk factors that may influence conditioning or supportive care (Chan et al., 2023).
- Immunosuppression from conditioning regimens places HSCT candidates at heightened risk for pulmonary and systemic infections, particularly invasive fungal disease, even in the absence of symptoms (Enger et al., 2023; Chan et al., 2023).
- Baseline imaging of the liver and heart may be relevant in patients with prior transfusion exposure, given the association between iron overload and adverse transplant outcomes (Chan et al., 2023).
- Sinus disease has not demonstrated a consistent association with peri-transplant outcomes in asymptomatic patients, limiting the utility of routine sinus imaging (Chan et al., 2023).

Evidence Discussion

- Pre-transplant chest CT has demonstrated utility in detecting clinically occult pulmonary infections in HSCT candidates, allowing initiation or modification of antimicrobial therapy prior to conditioning (Enger et al., 2023; Chan et al., 2023).
- Evidence does not support routine sinus CT screening, as studies show minimal impact on management decisions or surgical consultation rates, emphasizing the importance of avoiding low-yield imaging (Chan et al., 2023).
- Baseline liver ultrasound provide clinically meaningful comparators for post-transplant assessment of sinusoidal obstruction syndrome, as interval change rather than absolute measurement correlates with disease development (Chan et al., 2023).

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- MRI-based liver and cardiac iron quantification prior to transplantation correlates with post-transplant morbidity and mortality, supporting its role in selected high-risk populations while avoiding universal screening (Chan et al., 22023).
- Pre-transplant imaging practices that balance early identification of high-risk conditions with avoidance of unnecessary radiation and sedation reflect evidence-based risk stratification principles outlined in national consensus guidance (Enger et al., 2023; Chan et al., 023).

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Post-Transplant Imaging (PEDONC-20.2)

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Criteria

Early post-transplant complication (<100 days post HSCT):

Indication	Medically Necessary Imaging
For signs and symptoms of pulmonary infection or pulmonary edema or new CXR abnormalities in individuals post-transplant	• CT Chest without or with contrast (CPT® 71250 or CPT® 71260) ◦ For PET/CT requests to clarify infection vs malignancy, see disease-specific guidelines for equivocal conventional imaging
Suspected impending lung necrosis on other imaging in individuals post-transplant	• MRI Chest without and with contrast (CPT® 71552)
• EITHER of the following in individuals post-transplant: ◦ Suspected graft vs. host disease (GVHD) of chest ◦ Bronchiolitis Obliterans (BOOP/BOS)	• High-resolution CT Chest without contrast (CPT® 71250)
• ANY of the following in individuals post-transplant: ◦ Suspected intra-abdominal and or pelvic infection (including cystitis or typhlitis) ◦ Suspected small bowel GVHD	• Abdominal Ultrasound (CPT® 76700 or CPT® 76705) +/- Pelvic Ultrasound (CPT® 76856 or CPT® 76857) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) for unclear findings on ultrasound
High clinical suspicion for intra-abdominal fungal infection in individuals post-transplant	• CT Abdomen and Pelvis with contrast (CPT® 74177)
Suspected hepatic veno-occlusive disease (VOD) in individuals post-transplant	• Ultrasound elastography (CPT® 91200) or Abdominal Ultrasound with Doppler (CPT® 76700 or CPT® 76705)

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Indication	Medically Necessary Imaging
Suspected CNS complication including infection, hemorrhage, thrombosis, or encephalopathy in individuals post-transplant	• MRI Brain without contrast or MRI Brain without and with contrast (CPT® 70551 or CPT® 70553) ◦ CT Head without contrast (CPT® 70450) in the emergent setting or for suspected acute hemorrhage ▪ Acute CT should not preclude subsequent MRI imaging ◦ For additional imaging for suspected stroke, see: **Pediatric Stroke Initial Imaging (PEDHD-12.2)** in the Pediatric Head Imaging guidelines
Suspected musculoskeletal abscess or necrotizing fasciitis in individuals post-transplant	• See: **Pediatric Infection/Osteomyelitis (PEDMS-8)** in the Pediatric Musculoskeletal Imaging guidelines
Fever of unknown origin (8 or more days of temperature 38.0 C/100.4 F or higher) in individuals post-transplant	• PET/CT skull to thigh (CPT® 78815) if site-specific conventional imaging, microbiologic serologic studies, echocardiogram, urinalysis, and urine culture are all non-diagnostic
New neurologic symptoms on CAR-T	• MRI Brain without and with contrast (CPT® 70553)

Late post-transplant complication imaging (100 or more days post HSCT):

Indication	Medically Necessary Imaging
CNS complications in individuals post-transplant	Same imaging as for early post-transplant
Chest complications in individuals post-transplant	Same imaging as for early post-transplant
Hepatic VOD in individuals post-transplant	Same imaging as for early post-transplant

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Indication	Medically Necessary Imaging
Chronic GI GVHD in individuals post-transplant	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MR Enterography (CPT® 74183 and CPT® 72197) for suspected small bowel GVHD
Suspected renal tubular dysfunction in individuals post-transplant	• Nuclear renal imaging (CPT® 78700, CPT® 78701, CPT® 78707, CPT® 78708, CPT® 78709)
Suspected poor bone mineral density or osteonecrosis in individuals post-transplant	See: **Osteonecrosis in Long Term Cancer Survivors (PEDONC-19.4)**
Suspected PTLD in individuals post-transplant	See: **Post-Transplant Lymphoproliferative Disorder (PTLD) (PEDONC 5.3)**

Background and Supporting Information

- Post-transplant imaging is directed toward identifying transplant-related complications, which evolve over time and vary by organ system (Chan et al., 2023).
- Early post-transplant complications (<100 days) are dominated by infection, hemorrhage, and treatment-related toxicity, while late complications include chronic GVHD, organ fibrosis, and secondary malignancies (Chan et al., 2023; PDQ®, 2024).
- Pulmonary complications represent one of the most frequent causes of post-transplant morbidity, with radiologic findings often preceding clinical deterioration (Enger et al., 2023; Chan et al., 2023).
- Imaging selection is influenced by patient stability, need for sedation, radiation exposure concerns, and the diagnostic performance of modality-specific techniques across organ systems (Chan et al., 2023).

Evidence Discussion

- Chest CT provides high sensitivity for detecting early pulmonary infection and transplant-related lung injury, supporting its role when initial screening studies are inconclusive or clinical suspicion persists (Enger et al., 2023; Chan et al., 023).
- Ultrasound-based imaging strategies for abdominal and hepatic complications allow effective screening with minimal risk, reserving cross-sectional imaging for problem-solving and disease extent assessment (Chan et al., 2023).

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- MRI is favored for central nervous system and musculoskeletal complications due to its superior soft-tissue contrast and avoidance of ionizing radiation, particularly in long-term survivors requiring repeated evaluations (Chan et al., 2023).
- Evidence supports an imaging approach that is symptom-driven and complication-specific, minimizing cumulative imaging burden while maintaining high sensitivity for clinically actionable post-transplant complications (Chan et al., 2023; Enger et al., 2023).

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Procedure Codes Associated with Pediatric and Special Populations Oncology Imaging (PEDONC)

Guideline Page

Procedure Codes Associated with Pediatric and Special Populations Oncology Imaging (PEDONC)..... 407

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Procedure Codes Associated with Pediatric and Special Populations Oncology Imaging (PEDONC)

v1.0.2027

The inclusion of any code in this table does not imply that the code is under management or requires prior authorization. Refer to the applicable health plan for management details. Prior authorization of a code listed in this table is not a guarantee of payment. The Certificate of Coverage or Evidence of Coverage policy outlines the terms and conditions of the member's health insurance policy.

Code	Description
70450	Computed tomography, head or brain; without contrast material
70460	Computed tomography, head or brain; with contrast material(s)
70470	Computed tomography, head or brain; without contrast material, followed by contrast material(s) and further sections
70481	Computed tomography, orbit, sella, or posterior fossa or outer, middle, or inner ear; with contrast material(s)
70486	Computed tomography, maxillofacial area; without contrast material
70487	Computed tomography, maxillofacial area; with contrast material(s)
70488	Computed tomography, maxillofacial area; without contrast material, followed by contrast material(s) and further sections
70491	Computed tomography, soft tissue neck; with contrast material(s)
70496	Computed tomographic angiography, head, with contrast material(s), including noncontrast images, if performed, and image postprocessing
70543	Magnetic resonance (eg, proton) imaging, orbit, face, and/or neck; without contrast material(s), followed by contrast material(s) and further sequences
70544	Magnetic resonance angiography, head; without contrast material(s)
70545	Magnetic resonance angiography, head; with contrast material(s)
70546	Magnetic resonance angiography, head; without contrast material(s), followed by contrast material(s) and further sequences

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Code	Description
70551	Magnetic resonance (eg, proton) imaging, brain (including brain stem); without contrast material
70552	Magnetic resonance (eg, proton) imaging, brain (including brain stem); with contrast material(s)
70553	Magnetic resonance (eg, proton) imaging, brain (including brain stem); without contrast material, followed by contrast material(s) and further sequences
70554	Magnetic resonance imaging, brain, functional MRI; including test selection and administration of repetitive body part movement and/or visual stimulation, not requiring physician or psychologist administration
70555	Magnetic resonance imaging, brain, functional MRI; requiring physician or psychologist administration of entire neurofunctional testing
71250	Computed tomography, thorax, diagnostic; without contrast material
71260	Computed tomography, thorax, diagnostic; with contrast material(s)
71552	Magnetic resonance (eg, proton) imaging, chest (eg, for evaluation of hilar and mediastinal lymphadenopathy); without contrast material(s), followed by contrast material(s) and further sequences
72141	Magnetic resonance (eg, proton) imaging, spinal canal and contents, cervical; without contrast material
72142	Magnetic resonance (eg, proton) imaging, spinal canal and contents, cervical; with contrast material(s)
72147	Magnetic resonance (eg, proton) imaging, spinal canal and contents, thoracic; with contrast material(s)
72149	Magnetic resonance (eg, proton) imaging, spinal canal and contents, lumbar; with contrast material(s)
72156	Magnetic resonance (eg, proton) imaging, spinal canal and contents, without contrast material, followed by contrast material(s) and further sequences; cervical
72157	Magnetic resonance (eg, proton) imaging, spinal canal and contents, without contrast material, followed by contrast material(s) and further sequences; thoracic
72158	Magnetic resonance (eg, proton) imaging, spinal canal and contents, without contrast material, followed by contrast material(s) and further sequences; lumbar

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Code	Description
72193	Computed tomography, pelvis; with contrast material(s)
72197	Magnetic resonance (eg, proton) imaging, pelvis; without contrast material(s), followed by contrast material(s) and further sequences
73721	Magnetic resonance (eg, proton) imaging, any joint of lower extremity; without contrast material
73723	Magnetic resonance (eg, proton) imaging, any joint of lower extremity; without contrast material(s), followed by contrast material(s) and further sequences
74160	Computed tomography, abdomen; with contrast material(s)
74170	Computed tomography, abdomen; without contrast material, followed by contrast material(s) and further sections
74175	Computed tomographic angiography, abdomen, with contrast material(s), including noncontrast images, if performed, and image postprocessing
74177	Computed tomography, abdomen and pelvis; with contrast material(s)
74178	Computed tomography, abdomen and pelvis; without contrast material in one or both body regions, followed by contrast material(s) and further sections in one or both body regions
74183	Magnetic resonance (eg, proton) imaging, abdomen; without contrast material(s), followed by with contrast material(s) and further sequences
74185	Magnetic resonance angiography, abdomen, with or without contrast material(s)
75561	Cardiac magnetic resonance imaging for morphology and function without contrast material(s), followed by contrast material(s) and further sequences;
76390	Magnetic resonance spectroscopy
76498	Unlisted magnetic resonance procedure (eg, diagnostic, interventional)
76506	Echoencephalography, real time with image documentation (gray scale) (for determination of ventricular size, delineation of cerebral contents, and detection of fluid masses or other intracranial abnormalities), including A-mode encephalography as secondary component where indicated
76510	Ophthalmic ultrasound, diagnostic; B-scan and quantitative A-scan performed during the same patient encounter
76511	Ophthalmic ultrasound, diagnostic; quantitative A-scan only

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Code	Description
76512	Ophthalmic ultrasound, diagnostic; B-scan (with or without superimposed non-quantitative A-scan)
76536	Ultrasound, soft tissues of head and neck (eg, thyroid, parathyroid, parotid), real time with image documentation
76641	Ultrasound, breast, unilateral, real time with image documentation, including axilla when performed; complete
76642	Ultrasound, breast, unilateral, real time with image documentation, including axilla when performed; limited
76700	Ultrasound, abdominal, real time with image documentation; complete
76705	Ultrasound, abdominal, real time with image documentation; limited (eg, single organ, quadrant, follow-up)
76770	Ultrasound, retroperitoneal (eg, renal, aorta, nodes), real time with image documentation; complete
76856	Ultrasound, pelvic (nonobstetric), real time with image documentation; complete
76857	Ultrasound, pelvic (nonobstetric), real time with image documentation; limited or follow-up (eg, for follicles)
76870	Ultrasound, scrotum and contents
76881	Ultrasound, complete joint (ie, joint space and peri-articular soft-tissue structures), real-time with image documentation
76882	Ultrasound, limited, joint or focal evaluation of other nonvascular extremity structure(s) (eg, joint space, peri-articular tendon[s], muscle[s], nerve[s], other soft-tissue structure[s], or soft-tissue mass[es]), real-time with image documentation
77049	Magnetic resonance imaging, breast, without and with contrast material(s), including computer-aided detection (CAD real-time lesion detection, characterization and pharmacokinetic analysis), when performed; bilateral
77084	Magnetic resonance (eg, proton) imaging, bone marrow blood supply
78075	Adrenal imaging, cortex and/or medulla
78300	Bone and/or joint imaging; limited area
78305	Bone and/or joint imaging; multiple areas

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Code	Description
78306	Bone and/or joint imaging; whole body
78315	Bone and/or joint imaging; 3 phase study
78472	Cardiac blood pool imaging, gated equilibrium; planar, single study at rest or stress (exercise and/or pharmacologic), wall motion study plus ejection fraction, with or without additional quantitative processing
78608	Brain imaging, positron emission tomography (PET); metabolic evaluation
78609	Brain imaging, positron emission tomography (PET); perfusion evaluation
78700	Kidney imaging morphology
78701	Kidney imaging morphology; with vascular flow
78707	Kidney imaging morphology; with vascular flow and function, single study without pharmacological intervention
78708	Kidney imaging morphology; with vascular flow and function, single study, with pharmacological intervention (eg, angiotensin converting enzyme inhibitor and/or diuretic)
78709	Kidney imaging morphology; with vascular flow and function, multiple studies, with and without pharmacological intervention (eg, angiotensin converting enzyme inhibitor and/or diuretic)
78800	Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, single area (eg, head, neck, chest, pelvis), single day imaging
78801	Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, 2 or more areas (eg, abdomen and pelvis, head and chest), 1 or more days imaging or single area imaging over 2 or more days
78802	Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, whole body, single day imaging
78803	Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT) , single area (eg, head, neck, chest, pelvis), single day imaging

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Code	Description
78804	Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, whole body, requiring 2 or more days imaging
78813	Positron emission tomography (PET) imaging; whole body
78814	Positron emission tomography (PET) with concurrently acquired computed tomography (CT) for attenuation correction and anatomical localization imaging; limited area (eg, chest, head/neck)
78815	Positron emission tomography (PET) with concurrently acquired computed tomography (CT) for attenuation correction and anatomical localization imaging; skull base to mid-thigh
78816	Positron emission tomography (PET) with concurrently acquired computed tomography (CT) for attenuation correction and anatomical localization imaging; whole body
78830	Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT) with concurrently acquired computed tomography (CT) transmission scan for anatomical review, localization and determination/detection of pathology, single area (eg, head, neck, chest, pelvis), single day imaging
78831	Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT), minimum 2 areas (eg, pelvis and knees, chest and abdomen) or separate acquisitions (eg, lung ventilation and perfusion), single day imaging, or single area or acquisition over 2 or more days
78832	Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT) with concurrently acquired computed tomography (CT) transmission scan for anatomical review, localization and determination/detection of pathology, minimum 2 areas (eg, pelvis and knees, chest and abdomen) or separate acquisitions (eg, lung ventilation and perfusion), single day imaging, or single area or acquisition over 2 or more days
91200	Liver elastography, mechanically induced shear wave (eg, vibration), without imaging, with interpretation and report

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Code	Description
93306	Echocardiography, transthoracic, real-time with image documentation (2d), includes m-mode recording, when performed, complete, with Spectral doppler echocardiography, and with color flow doppler echocardiography
93307	Echocardiography, transthoracic, real-time with image documentation (2D), includes M-mode recording, when performed, complete, without spectral or color Doppler echocardiography
93308	Echocardiography, transthoracic, real-time with image documentation (2D), includes M-mode recording, when performed, follow-up or limited study
93321	Doppler echocardiography, pulsed wave and/or continuous wave with spectral display (List separately in addition to codes for echocardiographic imaging); follow-up or limited study (List separately in addition to codes for echocardiographic imaging)
93325	Doppler echocardiography color flow velocity mapping (List separately in addition to codes for echocardiography)
93975	Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; complete study
93976	Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; limited study

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