# CIGNA MEDICAL COVERAGE POLICIES – RADIOLOGY

## Pediatric and Special Populations Oncology Imaging Guidelines

Effective Date: February 3, 2026

**Instructions for use**

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

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

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

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

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

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

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# 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 |

# General Guidelines (PEDONC-1.0)

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- 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 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 medically necessary.
- Routine imaging of brain, spine, neck, chest, abdomen, pelvis, bones, or other body areas is not 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 routinely supported without signs or symptoms of systemic involvement of cancer.

  - Repeat imaging studies are not generally 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 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 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<br>• Screening using advancing imaging is only supported 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<br>  ◦ CT Chest prior to anesthesia for biopsy or resection of solid tumors and CTs of other involved body areas are generally indicated and should be performed concurrently<br>  ◦ 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<br>◦ Unless otherwise stated in the diagnosis-specific guidelines, imaging for treatment response is 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<br>◦ PET imaging is not 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<br>• Following documented recurrence of childhood cancer, any studies recommended for initial staging of that cancer type in the diagnosis-specific imaging guideline section should be approved<br>◦ 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 medically necessary according to the treatment response imaging in the diagnosis-specific guideline section<br>▪ Always refer to the diagnosis specific guideline for PET indications in recurrence. |

- Brain imaging is performed for signs or symptoms of brain disease.
  - 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 medically necessary to further characterize the lesion.

  - CT Head without and with contrast (CPT® 70470) is 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 medically necessary.
  - Radiopharmaceutical Localization scan SPECT (CPT® 78803 or CPT® 78831) or SPECT/CT (CPT® 78830 or CPT® 78832) is medically necessary as an add-on test for further evaluation of a specific area of interest.
  - CPT® codes 78300 (Nuclear bone scan limited), 78305 (Nuclear bone scan multiple areas), or 78315 do not have any indications in oncology nuclear medicine imaging.
- Bone scan supplemented by plain x-rays are the initial imaging modalities for suspected malignant bone pain. For specific imaging indications, see also:
  - **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)**
  - **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 prior 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.
- PET/CT may be indicated if:
  - Conventional imaging (CT, MRI, or bone scan) reveals findings that are inconclusive or negative, with continued suspicion for recurrence
- **Unless specified in diagnosis-specific guideline section PET/CT Imaging is NOT 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 8 mm 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 medically necessary.

### 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.

# 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** |
| ≥18 years old at initial diagnosis | • General Oncology Imaging Guidelines, except where directed otherwise by a specific guideline section |
| <18 years old at initial diagnosis | • Pediatric Oncology Imaging Guidelines, except where directed otherwise by a specific guideline section |
| 15 to 39 years old 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 Oncology, AYA individuals should be imaged according to the guideline section for their specific cancer type, regardless of the individual's age<br>• When unique guidelines for a specific cancer type exist in both Oncology and Pediatric Oncology, AYA individuals should be imaged according to the age rule in the previous bullet |

# Appropriate Clinical Evaluations (PEDONC-1.2)

**ONCP.GG.0001.2.C**

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- 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 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 medically necessary unless the imaging is being performed solely to address a study objective and would not be indicated 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-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 indicated 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 |
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| Screening | • Imaging requested for individuals at increased risk for a particular cancer in the absence of known clinical signs or symptoms<br>• Screening using advancing imaging is only supported 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<br>◦ CT Chest prior to anesthesia for biopsy or resection of solid tumors and CTs of other involved body areas are generally indicated and should be performed concurrently<br>◦ Metastatic CNS imaging and nuclear medicine imaging are generally deferred until after a histologic diagnosis is made, unless otherwise indicated by diagnosis-specific guideline |
| 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<br>◦ Unless otherwise stated in the diagnosis-specific guidelines, imaging for treatment response is 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 |

| Phases of Oncology Imaging | Definition |
|---|---|
| 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<br>◦ PET imaging is not 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<br>• Following documented recurrence of childhood cancer, any studies recommended for initial staging of that cancer type in the diagnosis-specific imaging guideline section should be approved<br>◦ 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 medically necessary according to the treatment response imaging in the diagnosis-specific guideline section<br>▪ Always refer to the diagnosis specific guideline for PET indications in recurrence. |

**Cardiac Function Assessment in Pediatric Oncology During Active Treatment:**

| 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:<br>◦ Cumulative cardiotoxic therapy received to date<br>◦ Individual's age and gender<br>◦ Most recent echocardiogram results<br>◦ New or worsening cardiac symptoms | • Echocardiography (CPT® 93306, CPT® 93307, or CPT® 93308) |
| • For either of the following:<br>◦ Echocardiography yielded a borderline shortening fraction (<30%) and additional left ventricular function data are necessary to make a chemotherapy decision OR<br>◦ Echocardiography windowing is suboptimal due to body habitus or tumor location | • Multi-gated acquisition (MUGA, CPT® 78472) blood pool nuclear medicine scanning |

**Background and Supporting Information**

**Immunosuppression 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 should be approved immediately
  - 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 should be approved 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 (ONC-29)**.
- Due to the high risk of invasive infections, frequent CT may be indicated to evaluate known sites of invasive fungal infection, and in general these should be approved as requested
  - Surveillance imaging of asymptomatic individuals to detect invasive fungal infection has not been shown to impact individual outcomes
    - Imaging requests are indicated when acute clinical decisions will be made based on the imaging

**Hematopoietic Stem Cell Transplant (HSCT) 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 (ONC-29)**.

# Modality General Considerations (PEDONC-1.3)

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- 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 with and without 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 without contrast 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 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 can be approved

◦ 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 may be approved 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 considerations 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:<br>◦ CPT® 78300<br>◦ CPT® 78305<br>◦ CPT® 78306<br>◦ CPT® 78803, 78830, or 78832<br>▪ May be approved alone or in combination with:<br>- CPT® 78305<br>- CPT® 78306 |
| $^{123}$I-metaiodobenzylguanidine (MIBG) scintigraphy | • ANY one of the following codes can be approved:<br>◦ CPT® 78801<br>◦ CPT® 78802<br>◦ CPT® 78804<br>• ANY one of the following codes may also be approved, individual or in combination with CPT® 78801, 78802, or 78804:<br>◦ CPT® 78803<br>◦ CPT® 78830<br>◦ CPT® 78831<br>◦ CPT® 78832<br>◦ 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.**

- 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 supported for a vast majority of oncologic conditions due to lack of standardization in imaging technique and interpretation. However, it is 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 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 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 medically necessary.
- PET imaging is not reliable for the detection of anatomic lesions smaller than 8 mm 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
  - Covered:





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- 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 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 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.
- PET is medically necessary if **all** of the following apply:
  - Conventional imaging (CT, MRI, US, plain film) reveals findings that are equivocal or suspicious
  - No other specific metabolic imaging (MIBG, octreotide, technetium, etc.) Is appropriate 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 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.C**

**v1.0.2026**

- 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 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.

# References (PEDONC-1)

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1. Krishnamurthy R, Daldrup-Link HE, Jones JY, et al. Imaging studies in the diagnosis and management of pediatric malignancies. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:185-238.
2. Allen-Rhoades W, Steuber CP. Clinical assessment and differential diagnosis of the child with suspected cancer. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:101-112.
3. Gottschalk S, Naik S, Hegde M, et al. Hematopoietic stem cell transplantation in pediatric oncology. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:419-440.
4. Freedman JL, Rheingold SR, and Fisher MJ. Oncologic emergencies. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:967-991.
5. Andrews J, Galel SA, Wong W, et al. Hematologic supportive care for children with cancer. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:992-1009.
6. Ardura MI, Koh AY. Infectious complications in pediatric cancer patients. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:1010-1057.
7. Shaikh R, Prabhu SP, Voss SD. Imaging in the evaluation and management of childhood cancer. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. *Nathan and Oski's Hematology and Oncology of Infancy and Childhood*. 8th ed. Philadelphia, PA: Elsevier Saunders; 2015:2146-2254.
8. Sung L, Fisher BT, Koh AY. Infectious Disease in the pediatric cancer patient. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. *Nathan and Oski's Hematology and Oncology of Infancy and Childhood*. 8th ed. Philadelphia, PA: Elsevier Saunders; 2015:2257-2266.
9. Mullen EA, Gratias E. Oncologic emergencies. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. *Nathan and Oski's Hematology and Oncology of Infancy and Childhood*. 8th ed. Philadelphia, PA: Elsevier Saunders; 2015:2267-2291.
10. Weiser DA, Kaste SC, Siegel MJ, et al. Imaging in childhood cancer: A society for Pediatric Radiology and Children's Oncology Group joint task force report. *Pediatr Blood Cancer*. 2013;60(8):1253-1260. doi:10.1002/pbc.24533.
11. American College of Radiology. ACR-ASER-SCBT-MR-SPR Practice Parameter for the Performance of Pediatric Computed Tomography (CT). 2019. Accesible from: https://gravitas.acr.org/PPTS.
12. Smith EA, Dillman JR. Current role of body MRI in pediatric oncology. *Pediatr Radiol*. 2016;46(6):873-880. doi:10.1007/s00247-016-3560-8.
13. Uslu L, Doing J, Link M, et al. Value of 18F-FDG PET and PET/CT for evaluation of pediatric malignancies. *J Nucl Med*. 2015;56(2):274-286. doi:10.2967/jnumed.114.146290.
14. McCarville MB. PET-CT imaging in pediatric oncology. *Cancer Imaging*. 2009(1);9:35-43. doi:10.1102/1470-7330.2009.0008.
15. Matthews JD, Forsythe AV, Brady Z, et al. Cancer risk in 680 000 people exposed to computed tomography scans in childhood or adolescence: data linkage study of 11 million Australians. *BMJ*. 2013;346:f2360. doi:10.1136/bmj.f2360.
16. Erdi YE. Limits of tumor detectability in nuclear medicine and PET. *Mol Imaging Radio Nucl Ther*. 2012;21(1):23-28. doi:10.4274/Mirt.138.
17. Brody AS, Guillerman RP. Don't let radiation scare trump patient care: 10 ways you can harm your patients by fear of radiation-induced cancer from diagnostic imaging. *Thorax*. 2014;69(8):782-784. doi:10.1136/thoraxjnl-2014-205499.
18. Meulepas JM, Ronckers CM, Smets AMJB, et al. Radiation exposure from pediatric CT scans and subsequent cancer risk in the Netherlands, *JNCI J Natl Cancer Ins*t. 2019;111:256-2634. doi: 10.1093/jnci/djy104.

19. Bhatia S, Pappo AS, Acquazzino M, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 2.2025—September 24, 2024, Adolescent and Young Adult (AYA) Oncology, available at: https://www.nccn.org/professionals/physician_gls/pdf/aya.pdf, referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Adolescent and Young Adult (AYA) Oncology V2.2025 09/24/2024. ©2024 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
20. States LJ, Reid JR. Whole-Body PET/MRI applications in pediatric oncology. *AJR Am J Roentgenol*. 2020;215(3):713-725. doi:10.2214/AJR.19.22677.
21. Mayerhoefer ME, Archibald SJ, Messiou C, Staudenherz A, Berzaczy D, Schöder H. MRI and PET/MRI in hematologic malignancies. *J Magn Reson Imaging*. 2020;51(5):1325-1335. doi:10.1002/jmri.26848.
22. Sher AC, Seghers V, Paldino MJ, et. al. Assessment of sequential PET/MRI in comparison with PET/CT of pediatric lymphoma: a prospective study. *AJR Am J Roentgenol*. 2016;206(3):623-31. doi:10.2214/AJR.15.15083.
23. Reighard C, Junaid S, Jackson WM, Arif A, Waddington H, Whitehouse AJO, Ing C. Anesthetic exposure during childhood and neurodevelopmental outcomes: a systematic review and meta-analysis. *JAMA Netw Open*. 2022;5(6):e2217427. doi:10.1001/jamanetworkopen.2022.17427.

# Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2)







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

**ONCP.SC.0002.1.C**

**v1.0.2026**

- 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 may be approved 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 may be approved on a case-by-case basis





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

**ONCP.SC.0002.2.C**
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**The following imaging studies should be considered medically necessary in individuals with LFS:**

| Indication | Medically Necessary Imaging Study |
|---|---|
| All individuals | • BOTH of the following, annually:<br>◦ MRI Brain without and with contrast (CPT® 70553)<br>◦ Whole-body MRI (WBMRI, CPT® 76498)<br>▪ CPT® 76498 is the only code for whole-body MRI at this time.<br>• Every 3 months from birth:<br>◦ Abdominal (CPT® 76700) and pelvic (CPT® 76856) ultrasound<br>• Beginning at age 20:<br>◦ Annual MRI Breast (CPT® 77049), alternating every 6 months with breast ultrasound, in addition to the previously noted annual MRI studies. |
| 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 - syndrome inherited in an autosomal dominant manner (50% risk to offspring) associated with germline mutations in *TP53* resulted in an increased susceptibility to a variety of cancers.
  - Eighty percent of individuals will have germline *TP53* mutation:
    - Tumor-specific *TP53* mutations are much more common than germline *TP53* mutations and are not associated with an increased risk for subsequent cancers.
    - If *TP53*-negative, formal diagnosis of LFS should be assigned by a physician with significant training and/or experience in LFS (most commonly a geneticist or oncologist) based on specified clinical criteria prior to beginning a screening imaging program.

    - *TP53* mutations may be present in 50% to 80% of pediatric adrenocortical carcinoma, 10% of pediatric rhabdomyosarcoma, and 10% of pediatric osteosarcoma.
- Individuals with LFS have an increased sensitivity to ionizing radiation, so screening strategies resulting in significant radiation exposure are not appropriate (CT and nuclear medicine).
- When a specific malignancy is suspected, the individual should be imaged according to the imaging guideline specific to the suspected cancer type.
- Annual complete detailed physical examinations, complete blood counts, and urinalyses form the backbone of LFS cancer screening.

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

**ONCP.SC.0002.3.C**
**v1.0.2026**

**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<br>◦ 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 | • MRI Brain (CPT® 70553) and Orbits (CPT® 70543) without and with contrast |
| • Clinical symptoms suggestive of change in a known plexiform neurofibroma<br>◦ Examples include: pain, rapid growth, and neurologic dysfunction | • MRI without and with contrast |
| • Clinical symptoms concerning for malignant transformation of a known plexiform neurofibroma, and ALL of the following are met:<br>◦ Recent MRI is inconclusive regarding transformation or progression<br>◦ Negative PET will result in a decision to avoid biopsy in a difficult or morbid location | • PET/CT (CPT® 78815 or CPT® 78816) |
| • Baseline tumor burden at age 16 or older<br>(one-time imaging) | • WBMBRI (CPT® 76498) |





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| Indication | Medically Necessary Imaging Study |
|---|---|
| • New soft tissue mass(es) | • See: Soft tissue mass information in **Soft Tissue Mass and Morton's Neuroma (MS 10.1)** 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) | • See: **Bone Tumors - General Considerations (PEDONC-9.1)** |
| • Documented optic pathway gliomas | • See: **Intracranial Low Grade Gliomas (PEDONC-4.2)** |
| • Known plexiform neurofibromas | • MRI without and with contrast of a known body area containing a neurofibroma is indicated for any of the following:<br>◦ Every 3 months for treatment response in individuals receiving active treatment<br>◦ New or worsening clinical symptoms suggesting progression<br>◦ Preoperative planning |
| • Biopsy-proven MPNST in individuals with known NF-1 or NF-2 | • For adult individuals, see: **Sarcomas - Bone, Soft Tissue, and GIST (ONC-12)**<br>• For pediatric individuals, see: **Non-Rhabdomyosarcoma Soft Tissue Sarcomas (NRSTS) (PEDONC-8.3)** |

**Background and Supporting Information**

**NF1:**

- Common syndrome inherited in an autosomal dominant manner (50% risk to offspring) affecting 1 in 2500 people. The diagnosis is commonly made based on

established clinical criteria including café-au-lait spots, Lisch nodules of the iris, axillary freckling, family history, and the presence of NF-associated tumors.
- The majority of tumors are benign in nature, but malignant degeneration can occur.
- NF1-affected persons have increased sensitivity to ionizing radiation, so CT and nuclear medicine imaging are not appropriate screening or surveillance studies for these individuals.
- 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 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 warranted in the absence of acute symptoms suggesting new or worsening intracranial disease
- Individuals with NF1 are at increased risk for plexiform neurofibromas (PN) and malignant peripheral nerve sheath tumors (MPNST— a high grade sarcoma)
  - Screening imaging of asymptomatic individuals for these tumors is not supported by evidence. PET imaging is not supported for PN surveillance in asymptomatic individuals at this time as the positive predictive value is only 60% to 65% even in symptomatic individuals.
- Although PET imaging has a positive predictive value of only 61% to 63% in NF1 individuals with suspected transformation to MPNST, the negative predictive value is high (96% to 99%)
  - Considerations for PET/CT coding and indications/coding for PET/MRI are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**
  - Repeat PET studies are not indicated due to the poor positive predictive value in this setting.
- 2017 AACR recommendations support a single baseline Whole-Body MRI to assess tumor burden in late adolescence or young adulthood. Further imaging should be based on focused MRI for symptomatic changes or pre-operative planning, and further surveillance WBMRI are not supported.

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

| Indication | Medically Necessary Imaging Study |
|---|---|
| • All individuals, beginning at age 10 years | • Annual MRI Brain without and with contrast (CPT® 70553) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| • All individuals 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) every 3 years |
| • All individuals 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 | • MRI Brain without and with contrast (CPT® 70553) |
| • Any of the following:<br>◦ Clinical symptoms suggestive of new or progressive spinal or paraspinal tumors, including uncomplicated back pain or radiculopathy<br>◦ 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 known vestibular schwannoma | • Annual MRI Brain without and with contrast (CPT® 70553) |
| • Known vestibular schwannomas and any of the following:<br>◦ New or worsening clinical symptoms including hearing loss<br>◦ Preoperative planning | • MRI Brain without and with contrast (CPT® 70553) |
| • Known meningioma | • See: **Meningiomas (ONC-2.8)** in the Oncology Imaging Guidelines |
| • Known ependymoma | • See: **Ependymoma (PEDONC-4.8)** |

## Background and Supporting Information

**NF2:**

- NF2 is substantially less common than NF1. It is inherited in an autosomal dominant manner (50% risk to offspring) affecting ~1 in 25000 people. NF2 is associated with increased risk for meningiomas (50% of affected individuals), vestibular schwannomas, and spinal tumors (75% of affected individuals).

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

**ONCP.SC.0002.4.A**
**v1.0.2026**

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

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

| Indication | Medically Necessary Imaging Study |
|---|---|
| • Known renal tumors | • See: **Pediatric Renal Tumors (PEDONC-7)** |
| • Known hepatoblastoma | • See: **Hepatoblastoma (PEDONC-11.2)** |
| • Known neuroblastoma | • See: **Neuroblastoma (PEDONC-6)** |
| • Known adrenocortical carcinoma | • See: **Pediatric Adrenocortical Carcinoma (PEDONC-14)** |
| • Known pheochromocytoma | • See: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines |

**Background and Supporting Information**

- Inherited syndrome characterized by macroglossia, hemihypertrophy, macrosomia, organomegaly, and neonatal hypoglycemia. Individuals with isolated hemihypertrophy are also imaged according to this guideline.
- Caused by mutation at chromosome 11p15, affected children are predisposed to Wilms tumor, hepatoblastoma, rhabdomyosarcoma, and adrenal tumors.

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

**ONCP.SC.0002.5.A**
**v1.0.2026**

**The following imaging studies should be considered appropriate in individuals with DDS:**

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

**Background and Supporting Information**

- Characterized by pseudohermaphroditism, early renal failure, and >90% risk of Wilms tumor development in each kidney. Associated with mutations at 11p13, risk of renal failure after detection of symptomatic Wilms tumor is 62%, so early detection may allow for renal-sparing surgical approaches.





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

**ONCP.SC.0002.6.A**
**v1.0.2026**

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

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

**Background and Supporting Information**

- Named for the components of the disorder, it is associated with mutations at 11p13. As the name suggests, individuals are predisposed to Wilms tumor, with 57% of individuals in one cohort developing Wilms tumor. Risk of renal failure after detection of symptomatic Wilms tumor is 38%, so early detection may allow for renal-sparing surgical approaches.

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

**ONCP.SC.0002.7.A**
**v1.0.2026**

- 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, from birth to the 6th birthday | • Abdominal ultrasound (CPT® 76700) every 3 months |
| • All individuals, beginning at age 12 years | • Annual thyroid ultrasound (CPT® 76536) |
| • All individuals, beginning at age 30 years | • Annual pelvic ultrasound (CPT® 76856) |
| • Family history of desmoid tumors | • Abdominal ultrasound (CPT® 76700) annually for life after age 6 |
| • Individuals with Spigelman Stage III or IV or if duodenectomy is being planned<br>• 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) |
| • Known colorectal tumors | • See: **Colorectal Cancer (ONC-16)** in the Oncology Imaging Guidelines |
| • Known desmoid tumors | • See: **Non-Rhabdomyosarcoma Soft Tissue Sarcomas (NRSTS) (PEDONC-8.3)** |

**Background and Supporting Information**

- Inherited in an autosomal dominant manner (50% risk to offspring), it is also known as Adenomatous Polyposis Coli (APC). It is associated with the development of thousands of colonic polyps by age 20 and >90% risk of colorectal carcinoma. Prophylactic total colectomy is recommended by age 20 for most individuals. FAP is also associated with hepatoblastoma, tumors of the pancreas and small bowel, medulloblastoma, and thyroid cancer.

# Multiple Endocrine Neoplasias (MEN) (PEDONC-2.8)

**ONCP.SC.0002.8.C**
**v1.0.2026**

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

| Indication | Medically Necessary Imaging Study |
|---|---|
| All individuals, beginning at age 5 years | • Annual MRI Brain without and with contrast (CPT® 70553)<br>• Annual MRI Abdomen without and with contrast (CPT® 74183), CT Abdomen with contrast (CPT® 74160), or ultrasound (CPT® 76700)<br>• Annual Octreotide study (see: **PEDONC-1.3** for coding) |
| All individuals, beginning at age 15 years | • Annual MRI Chest without and with contrast (CPT® 71552) or CT Chest with contrast (CPT® 71260) |
| Known thyroid cancer | • See: **Thyroid Cancer (ONC-6)** in the Oncology Imaging Guidelines |
| Known pheochromocytoma | • 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:**

| Indication | Medically Necessary Imaging Study |
|---|---|
| All individuals, beginning at age 5 years | • MRI Abdomen without and with contrast (CPT® 74183) every 3 years |
| • One of the following:<br>◦ Elevated catecholamines<br>◦ Inconclusive adrenal mass on MRI | • ONE of the following:<br>◦ Octreotide study (See: **PEDONC-1.3** for coding)<br>◦ Adrenal Nuclear Imaging (CPT® 78075) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| • Known pheochromocytoma | • See: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines |

**Background and Supporting Information**

- Inherited in an autosomal dominant manner (50% risk to offspring)
- MEN1 is characterized by parathyroid, pancreatic islet cell, and pituitary gland tumors (3 P's), as well as carcinoid tumors in the chest and abdomen, and 28% of individuals will develop at least one tumor by age 15.
- MEN2a is characterized by medullary thyroid carcinoma, parathyroid adenomas, and pheochromocytomas.
- MEN2b is characterized by ganglioneuromas of the GI tract and skeletal abnormalities presenting in infancy.
- Recommended cancer screening includes:
  - MEN2a and MEN2b: Annual measurement of catecholamines for pheochromocytoma screening

# Tuberous Sclerosis Complex (TSC) (PEDONC-2.9)

**ONCP.SC.0002.9.C**

**v1.0.2026**

- Abdominal MRI (CPT® 74183) is 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, at the time of suspected diagnosis until age 25 years | • Annual Brain MRI without and with contrast (CPT® 70553) |
| • All individuals at diagnosis | • Single baseline MRI Abdomen without and with contrast (CPT® 74183) |
| • If no renal lesions seen on baseline MRI Abdomen | • From diagnosis through age 11:<br>◦ Annual Renal US (CPT® 76770)<br>• Age 12 (or 10 years earlier than the youngest family member with renal cell carcinoma, whichever comes earlier):<br>◦ MRI Abdomen without and with contrast (CPT® 74183) annually in lieu of ultrasound |
| • For documented renal lesions on baseline MRI Abdomen or any ultrasound | • MRI Abdomen without and with contrast (CPT® 74183) annually |
| • All individuals, beginning at age 18 years | • CT Chest without contrast (CPT® 71250) every 5 years |
| • All individuals, 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 |
| • Individuals with documented abnormalities on baseline CT Chest | • Additional CT Chest without contrast or with contrast (CPT® 71250 or CPT® 71260) every 1 year |

| Indication | Medically Necessary Imaging Study |
|---|---|
| • Any of the following:<br>◦ New pulmonary symptoms<br>◦ Worsening pulmonary function testing | • CT Chest without contrast (CPT® 71250) |
| • Known SEGA tumors | • See: **Intracranial Low Grade Gliomas (LGG) (PEDONC-4.2)** |
| • Known renal cell carcinoma | • See: **Pediatric Renal Cell Carcinoma (RCC) (PEDONC-7.4)** |

**Background and Supporting Information**

- Inherited in an autosomal dominant manner (50% risk to offspring), affecting ~1 in 6000 individuals, it is associated with benign tumors, hypopigmented skin macules (ash leaf spots), pulmonary lymphangioleiomyomatosis, developmental delay, and epilepsy.
- Malignancies associated with this syndrome include:
  - Subependymal giant cell astrocytomas (SEGA tumors)
    - Historically, early surgery was important to reduce morbidity related to these tumors
    - More recently, everolimus has been successfully used to treat these tumors without surgery, and early detection remains an important feature for success
  - Renal cell carcinoma
  - Cardiac rhabdomyosarcoma
  - Pulmonary lymphangioleiomyomatosis
- Recommended cancer screening includes:
  - Ophthalmologic and dermatologic evaluation annually

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

**ONCP.SC.0002.10.A**
**v1.0.2026**

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

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

| Indication | Medically Necessary Imaging Study |
|---|---|
| • Known (based on imaging) CNS hemangioblastoma | • See: **Intracranial Low Grade Gliomas (LGG) (PEDONC-4.2)** |
| • Known renal cell carcinoma | • See: **Pediatric Renal Cell Carcinoma (RCC) (PEDONC-7.4)** |
| • Known pheochromocytoma or other neuroendocrine tumor | • See: **Neuroendocrine Cancers And Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines |

**Background and Supporting Information**

- Inherited in an autosomal dominant manner (50% risk to offspring), it is associated with CNS hemangioblastomas, retinal angiomas, endolymphatic sac tumors (ELST), gastrointestinal stromal tumor (GIST), renal cell carcinoma (RCC), and pheochromocytomas and other neuroendocrine tumors (NETs). Pediatric individuals are at risk of developing hemangioblastomas and pheochromocytomas that can remain clinically occult until symptoms become severe. Historically, substantial mortality was attributable to RCC, pancreatic NET, and CNS hemangioblastoma.
- Recommended cancer screening includes:
  - Annual ophthalmologic evaluation beginning at birth
  - Annual measurement of catecholamines beginning at age 2
  - Audiology assessment every 2 years beginning at age 5

# Rhabdoid Tumor Predisposition Syndrome (PEDONC-2.11)

**ONCP.SC.0002.11.A**
**v1.0.2026**

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

| Indication | Medically Necessary Imaging Study |
|---|---|
| • All individuals, at diagnosis, as early as birth if requested | • Whole-body MRI (WBMRI) (CPT® 76498) |
| • All individuals, from birth to 6 months of age | • ALL of the following, monthly:<br>◦ US Head (CPT® 76506)<br>◦ US Abdomen (CPT® 76700)<br>◦ US Pelvis (CPT® 76856)<br>◦ US Neck (CPT® 76536)<br>◦ MRI with and without contrast of areas of concern found on baseline WBMRI |
| • All individuals, from age 7 months to 5 years | • ALL of the following, every 3 months:<br>◦ US Abdomen (CPT® 76700)<br>◦ US Pelvis (CPT® 76856)<br>◦ US Neck (CPT® 76536)<br>◦ MRI Brain (CPT® 70553)<br>◦ MRI Spine (CPT® 72156, CPT® 72157, and CPT® 72158) without and with contrast<br>◦ MRI with and without contrast of areas of concern found on baseline WBMRI |
| • All individuals, after age 5 years | • Annual WBMRI (CPT® 76498) |
| • Clinical symptoms or WBMRI findings suggesting malignancy | • Targeted advanced imaging of the suspected disease site (CT with or without contrast, or MRI without and with contrast) |





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| Indication | Medically Necessary Imaging Study |
|---|---|
| • Inconclusive findings on ultrasound | • MRI with and without contrast of the inconclusive US site<br>◦ MRI is then medically necessary in place of ultrasound for remainder of planned screening |

**Background and Supporting Information**

- Inherited in an autosomal dominant manner (50% risk to offspring), it is associated with malignant rhabdoid tumors of the kidney and extrarenal locations, and atypical teratoid/rhabdoid tumors (ATRT) of the CNS. It is caused by a germline mutation in *INI1* or *SMARCB1*, and is associated with a more variable prognosis than de novo rhabdoid tumors.

# Familial Retinoblastoma Syndrome (PEDONC-2.12)

**ONCP.SC.0002.12.C**
**v1.0.2026**

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

| Indication | Medically Necessary Imaging Study |
|---|---|
| • Individuals with retinomas (premalignant retinal lesions) | • Annual MRI Orbits (CPT® 70543) |
| • Either of the following:<br>◦ Inconclusive EUA findings<br>◦ New symptoms | • US Orbits (CPT® 76512, 76510, or 76511) or MRI Orbits (CPT® 70543)<br>◦ These studies should be used if at all possible in lieu of CT or nuclear imaging to avoid radiation exposure |

**Background and Supporting Information**

- This syndrome is inherited in an autosomal dominant manner (50% risk to offspring). As the name suggests, it is associated with retinoblastoma, as well as osteosarcoma, pediatric melanoma, and a significantly increased risk for radiation-related malignancies.
- 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.

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

**ONCP.SC.0002.13.A**
**v1.0.2026**

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

| Indication | Medically Necessary Imaging Study |
|---|---|
| • All individuals with SDHx, MAX, TMEM127 mutations, beginning at age 6 | • Every 2 years, ONE of the following sets of imaging:<br>◦ Whole-body MRI (CPT® 76498)<br>▪ CPT® 76498 is the only code for whole-body MRI at this time<br>OR<br>◦ 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)<br>▪ 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)<br>▪ MRI Neck imaging should include skull base, thus separate MRI Brain (CPT® 70553) is only medically necessary if there are CNS-specific symptoms. |
| • Initial screening for individuals diagnosed at age 18 or older | • One-time PET/CT (CPT® 78815 or 78816), with 68Ga-DOTA-SSAs<br>◦ FDG may be substituted if Dotatate radiotracers are not available |
| • Known pheochromocytoma or other neuroendocrine tumors | • See: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines |

**Background and Supporting Information**

- Caused by mutations in *SDHx* and related genes, this syndrome is inherited in an autosomal dominant manner (50% risk to offspring), and is associated with pheochromocytomas and paragangliomas.
- Individuals with multiple endocrine neoplasias should not use this guideline and should be imaged according to **Multiple Endocrine Neoplasias (MEN) (PEDONC-2.8)**.
- MRI is preferred to CT to minimize radiation exposure given these individuals' lifelong need for screening
- All individuals with HPP receive annual measurement of catecholamines
- EANM–SNMMI joint guidelines propose the use of 68Ga-DOTA-SSAs PET as the first-choice functional imaging modality in adult SDHx mutation carriers and the use of 18F-FDG when 68Ga-DOTA-SSAs PET is not available.

# Costello Syndrome (PEDONC-2.14)

**ONCP.SC.0002.14.A**
**v1.0.2026**

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

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

**Background and Supporting Information**

- Caused by mutations in *HRAS* genes, this syndrome is inherited in an autosomal dominant manner (50% risk to offspring), and is associated with rhabdomyosarcoma and neuroblastoma in early childhood, and transitional cell cancer of the bladder in older children and adults.

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

**ONCP.SC.0002.15.C**
**v1.0.2026**

**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, beginning at age 4 years | • Annual esophagogastroduodenoscopy and colonoscopy |
| • All individuals, beginning at age 6 years | • Annual whole-body MRI (CPT® 76498)<br>◦ CPT® 76498 is the only code for whole-body MRI at this time |

**Background and Supporting Information**

- A highly penetrant and aggressive cancer predisposing syndrome resulting from autosomal recessive inheritance of biallelic mutations in mismatch repair genes, CMMRD syndrome leads to substantial risk for several commonly fatal childhood malignancies - high-grade CNS tumors (glioma, PNET, medulloblastoma) and hematologic malignancies (non-Hodgkin lymphoma, acute lymphoblastic leukemia). CMMRD individuals are also at increased risk for gastrointestinal tumors.





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

**ONCP.SC.0002.16.A**
**v1.0.2026**

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

**Background and Supporting Information**

- Hereditary leiomyomatosis and renal cell cancer (HLRCC) is a disorder in which affected individuals tend to develop benign tumors containing smooth muscle tissue (leiomyomas) in the skin and, in females, the uterus. Approximately 20 percent of people with HLRCC develop renal cell cancer. People with HLRCC are commonly diagnosed with kidney cancer in their forties but cases have been reported in individuals as young as 11.
- In 2019, Forde et. al. published the first large, prospective study of screening for HLRCC and showed that most symptomatic individuals present with stage 3 or 4 RCC with a high risk of death and one life is saved for every 5 individuals on an MRI screening protocol.

# Other Renal Cell Cancer Predisposition Syndromes (PEDONC-2.17)

ONCP.SC.0002.17.C
**v1.0.2026**

### Birt-Hogg-Dube Syndrome (BHDS)

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

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

**Background and Supporting Information**

- Birt-Hogg-Dube Syndrome (BHDS) in an autosomal dominant disorder with a heterozygous pathogenic variant in FLCN gene.
- These individuals have, or may have:
  - Cutaneous manifestations (fibrofolliculomas, acrochordons, angiofibromas, oral papules, cutaneous collagenomas, and epidermal cysts)
  - Pulmonary cysts/history of pneumothorax
  - A seven-fold increase in renal tumors
    - The most common renal tumors are oncocytoma, chromophobe, and a hybrid of these two.
    - Renal tumors may be multifocal and bilateral.
    - Median age of renal tumor diagnosis is 48 years.
- Recommended cancer screening includes:
  - Screening colonoscopy starting at age 40
    - Earlier colonoscopy may be considered for those with a family history of colorectal cancer earlier than age 40.

### BAP1 Tumor Predisposition Syndrome

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

| Indication | Medically Necessary Imaging Study |
|---|---|
| • All individuals, beginning at age 30 years | • MRI Abdomen with and without contrast (CPT® 74183) every 2 years |

**Background and Supporting Information**

BAP1 tumor predisposition syndrome (TPDS) is an autosomal dominant condition with a predisposition to melanoma (ocular and cutaneous), mesothelioma, clear cell renal cancer, and chromophobe renal cancer.

**Hereditary Papillary Renal Carcinoma (HPRC)**

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

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

**Background and Supporting Information**

- Hereditary papillary renal carcinoma (HPRC) is an autosomal dominant condition involving the MET gene and predisposes individuals to multifocal, bilateral renal tumors.

**DICER1**

**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:<br>◦ CT Chest (contrast as requested)<br>• 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:<br>◦ Abdominal US (CPT® 76700)<br>• After age 12, it is 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) |

**Other Renal Predisposition Syndromes**

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

**Background and Supporting Information**

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

# Infantile Myofibromatosis (PEDONC-2.18)

**ONCP.SC.0002.18.A**
**v1.0.2026**

**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 | • WBMRI (CPT® 76498) once within 6 months of diagnosis |
| • Pre-operative planning or<br>• Signs or symptoms suggesting progression 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 | • MRI with and without contrast of unresected visceral involved sites, every 6 months until the age of two years |
| • Post-operative evaluation and both of the following:<br>◦ Adequacy of resection is unclear and<br>◦ Re-excision is being considered | • One-time MRI with and without contrast |
| • Surveillance for emergence of visceral disease | • Every 6 months, until the age of two years:<br>◦ CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250)<br>◦ CT Abdomen and Pelvis (CPT® 74177)<br>• CT should not duplicate simultaneous MRI imaging of involved sites |
| • Individuals requiring chemotherapy | • See: **PEDONC-8.3** |

**Background and Supporting Information**

- Infantile myofibromatosis is a benign condition characterized by soft tissue tumors, 90% of which present in the first 2 years of life. 75% of cases present as solitary lesions affecting the skin and/or muscles of the head, neck and trunk. Other patterns of inheritance include congenital multicentric disease limited to skin and muscle, congenital multicentric with a single visceral site, and congenital with multiple visceral site involvement.
- The condition is most commonly sporadic, though familial cases associated with *PDGFRB* and *NOTCH3* have been described
- The majority of cases with skin or muscle only involvement regress spontaneously.
- Visceral cases are lethal in the absence of therapy in 75% of cases due to organ compression, particularly with cardiopulmonary involvement.
- If vital structures are involved or significant symptoms occur, treatment is generally radical resection. Chemotherapy is sometimes utilized for progressive or multifocal life threatening lesions.
- Infants may present with a single lesion and develop further lesions in the first two years of life. This scenario, along with the inability of infants to readily express symptoms, impacts the imaging studies that may be approved.
- Routine surveillance of cutaneous/muscular sites is not supported as spontaneous regression is common.
- Surveillance beyond the age of two years is not supported.

# Bloom Syndrome (PEDONC-2.19)

**ONCP.SC.0002.19.A**
**v1.0.2026**

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

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





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# References (PEDONC-2)

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1. Plon SE, Malkin D. Childhood Cancer and Heredity. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:13-31.
2. Aplan PD, Shern JF, Khan J. Molecular and genetic basis of childhood cancer. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:32-62.
3. Zelley K, Lindell RB, Schiffman JD. et al. Genetic predisposition to cancer. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th ed. Philadelphia, PA: Elsevier Saunders; 2015:1270-1293.
4. Garber JE, Offit K. Hereditary cancer predisposition syndromes. *J Clin Oncol*. 2005;23(2):276-292. doi:10.1200/JCO.2005.10.042.
5. Holman JD, Dyer JA. Genodermatoses with malignant potential. *Current Opin Pediatr*. 2007;19(4):446-454. doi:10.1097/MOP.0b013e3282495939. .
6. Foulkes WD. Inherited susceptibility to common cancers. *N Engl J Med*. 2008;359:2143-2153. doi:10.1056/NEJMra0802968.
7. Schiffman JD, Geller JI, Mundt E, et al. Update on pediatric cancer predisposition syndromes. *Pediatr Blood Cancer*. 2013;60(8):1247-1252. doi:10.1002/pbc.24555.
8. American Academy of Pediatrics. Ethical and policy issues in genetic testing and screening of children. *Pediatrics*. 2013;131(3):620-622. doi:10.1542/peds.2012-3680.
9. Monsalve J, Kapur J, Malkin D, Babyn PS. Imaging of cancer predisposition syndromes in children. *Radiographics*. 2011;31(1):263-280. doi:10.1148/rg.311105099.
10. D'Orazio JA. Inherited cancer syndromes in children and young adults. *J Pediatr Hematol Oncol*. 2010;32(3):195-228. doi:10.1097/MPH.0b013e3181ced34c.
11. Lu KH, Wodd ME, Daniels M, et al. American Society of Clinical Oncology Expert Statement: collection and use of a cancer family history for oncology providers. *J Clin Oncol*. 2014;32(8):833-841. doi:10.1200/JCO.2013.50.9257.
12. Farid M, Ngeow J. Sarcomas associated with genetic cancer predisposition syndromes: a review. *Oncologist*. 2016;21(8):1002-1013. doi:10.1634/theoncologist.2016-0079.
13. Tiwari R, Singh AK, Somwaru AS, et al. Radiologist's primer on imaging of common hereditary cancer syndromes. *RadioGraphics*. 2019;39:759-778. doi:10.1148/rg.2019180171.
14. Schneider K, Zelley K, Nichols KE, Levine AS, Garber J. Li-Fraumeni Syndrome. In: Adam MP, Ardinger HH, Pagon RA, et al., ed. GeneReviews® [Internet]. Seattle, WA: University of Washington, Seattle; 1999-2025. https://www.ncbi.nlm.nih.gov/books/NBK1311/.
15. Villani A, Tabori U, Schiffman J, et al. Biochemical and imaging surveillance in germline TP53 mutation carriers with Li-Fraumeni syndrome: a prospective observational study. *Lancet Oncol*. 2011;12(6):559-567. doi:10.1016/S1470-2045(11)70119-X.
16. Villani A, Shore A, Wasserman JD, et al. Biochemical and imaging surveillance in germline TP53 mutation carriers with Li-Fraumeni syndrome: 11 year follow-up of a prospective observational study. *Lancet Oncol*. 2016;17(9):1295-1305. doi:10.1016/S1470-2045(16)30249-2.
17. Ballinger ML, Best A, Mai PL, et al. Baseline surveillance in Li-Fraumeni Syndrome using whole-body magnetic resonance imaging a meta analysis. *JAMA Oncol*. 2017;3(12):1634-1639. doi:10.1001/jamaoncol.2017.1968.
18. Ruijs MWG, Loo CE, van Buchem CAJM, et al. Surveillance of Dutch patients with Li-Fraumeni Syndrome: the life-guard study. *JAMA Oncol*. 2017;3(12):1733-1734. doi:10.1001/jamaoncol.2017.1346.
19. Mai PL, Khincha PP, Loud JT, et al. Prevalence of Cancer at Baseline screening in the National Cancer Institute Li-Fraumeni Syndrome Cohort. *JAMA Oncol*. 2017;3(12):1640-1645. doi:10.1001/jamaoncol.2017.1350.
20. Kratz CP, Achatz MI, Brugiéres L, et al. Cancer screening recommendations for individuals with Li-Fraumeni Syndrome. *Clin Cancer Res*. 2017;23(11):e38-e45. doi:10.1158/1078-0432.CCR-17-0408.
21. Greer MC, Voss SD, States LJ. Pediatric cancer predisposition imaging: focus on whole-body MRI. *Clin Cancer Res*. 2017;23(11):e6-e13. doi:10.1158/1078-0432.CCR-17-0515.

22. Asdahl PH, Ojha RP, Hasle H. Cancer screening in Li-Fraumeni Syndrome. *JAMA Oncol*. 2017;3(12):1645-1646. doi:10.1001/jamaoncol.2017.2459.
23. Eutsler EP, Khanna G. Whole-body magnetic resonance imaging in children: technique and clinical applications. *Pediatr Radiol*. 2016;46(6):858-872. doi: 10.1007/s00247-016-3586-y.
24. Gottumukkalla RV, Gee MS, Hampilos PJ, Greer MC. Current and emerging roles of whole-body MRI in evaluation of pediatric cancer patients. *RadioGraphics*. 2019;39:516-534. doi:10.1148/rg.2019180130.
25. Daly MB, Pal T, AlHilli Z, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 1.2026 —July 10, 2025. Genetic/Familial High-Risk Assessment: Breast, Ovarian, Pancreatic, and Prostate available at: https://www.nccn.org/professionals/physician_gls/pdf/bopp.pdf. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Genetic/Familial High-Risk Assessment: Breast, Ovarian, Pancreatic, and Prostate V1.2026 7/10/2025 ©2024 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
26. Miller DT, Freedenberg D, Schorry E, et al. Health supervision for children with neurofibromatosis type 1. *Pediatrics*. 2019;143:e20190660. doi:10.1541/peds.2019-0660.
27. Sahin M. Neurocutaneous syndromes. In: Kliegman RM, Stanton BF, St. Geme JW, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. Philadelphia, PA: Elsevier Saunders; 2011:2046-2048.
28. Ullrich NJ. Neurocutaneous syndromes and brain tumors. *J Child Neurol.* 2016;31(12):1399-1411. doi:10.1177/0883073815604220.
29. Korf BR and Bebin EM. Neurocutaneous disorders in Children. *Pediatr Rev*. 2017; 38(3):119-127. doi:10.1542/pir.2015-0118.
30. Campian J, Gutmann DH. CNS tumors in neurofibromatosis. *J Clin Oncol*. 2017; 35(21):2378-2385. doi:10.1200/JCO.2016.71.7199.
31. Friedman JM. Neurofibromatosis 1. In: Pagon RA, Adam MP, Ardinger HH et al., eds. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle;1998-2025. https://www.ncbi.nlm.nih.gov/books/NBK1109/.
32. Karajannis MA, Ferner RE. Neurofibromatosis-related tumors: emerging biology and therapies. *Curr Opin Pediatr*. 2015;27(1):26-33. doi:10.1097/MOP.0000000000000169.
33. Meany H, Dombi E, Reynolds J, et al. 18-Fluorodeoxyglucode-Positron Emission Tomography (FDG-PET) evaluation of nodular lesions in patients with neurofibromatosis Type 1 and Plexiform Neurofibromas (PN) or Malignant Peripheral Nerve Sheath Tumors (MPNST). *Pediatr Blood Cancer*. 2013;60(1):59-64. doi:10.1002/pbc.24212.
34. Tsai LL, Druback L, Fahey F, et al. [18F]-Fluorodeoxyglucose positron emission tomography in children with neurofibromatosis type 1 and plexiform neurofibromas: correlation with malignant transformation. *J Neuro Oncol*. 2012;108(3):469-475. doi:10.1007/s11060-012-0840-5.
35. Corbemale P, Valeyrie-Allanore L, Giammarile F, et al. Utility of 18F-FDG PET with a semi-quantitative index in the detection of sarcomatous transformation in patients with neurofibromatosis Type 1. *PLoS ONE*. 2014;9(2):e85954. doi:10.1371/journal.pone.0085954.
36. Chirindel A, Chaudhry M, Blakeley JO, Wahl R. 18F-FDG PET/CT Qualitative and quantitative evaluation in neurofibromatosis Type 1 Patients for detection of malignant transformation: comparison of early to delayed imaging with and without liver activity normalization. *J Nucl Med*. 2015;56(3):379-385. doi:10.2967/jnumed.114.142372.
37. Prada CE, Hufnagel RB, Hummel TR, et al. The use of magnetic resonance imaging screening for optic pathway gliomas in children with neurofibromatosis Type 1. *J Pediatr*. 2015;167(4):851-856. doi:10.1016/j.jpeds.2015.07.001.
38. Blakeley JO, Evans DG, Adler J, et al. Consensus recommendations for current treatments and accelerating clinical trials for patients with neurofibromatosis Type 2. *Am J Med Genet A*. 2012;158A(1):24-41. doi:10.1002/ajmg.a.34359.
39. Evans DG. Neurofibromatosis 2. In: Pagon RA, Adam MP, Bird TD et al., eds. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle; 1998-2023. . https://www.ncbi.nlm.nih.gov/books/NBK1201/.
40. Evans DG. Neurofibromatosis type 2 (NF2): a clinical and molecular review. *Orphanet J Rare Dis*. 2009;4:16. doi:10.1186/1750-1172-4-16.

41. Ardern-Holmes S, Fisher G, North K. Neurofibromatosis Type 2: presentation, major complications, and management, with a focus on the pediatric age group. *J Child Neurol*. 2016;32(1):9-22. doi:10.1177/0883073816666736.
42. Shuman C, Beckwith JB, Weksberg R. Beckwith-Wiedemann syndrome. In: Pagon RA, Adam MP, Bird TD et al., eds. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle; 2000-2023. https://www.ncbi.nlm.nih.gov/pubmed/20301568.
43. Choyke PL, Siegel MJ, Craft AW, Green DM, DeBaun MR. Screening for Wilms Tumor in children with Beckwith-Wiedemann syndrome or idiopathic hemihypertrophy. *Med Pediatr Oncol*. 1999;32(3):196-200. doi:10.1002/(SICI)1096-911X(199903)32:3<196::AID-MPO6>3.0.CO;2-9.
44. Mussa A, Molinatto C, Baldassare G, et al. Cancer risk in Beckwith-Wiedemann syndrome: A systematic review and meta-analysis outlining a novel (epi)genotype specific histotype targeted screening protocol. *J Pediatr*. 2016;176:142-149. doi:10.1016/j.jpeds.2016.05.038.
45. MacFarland SP, Mostoufi-Moab S, Zelley K, et al. Management of adrenal masses in patients with Beckwith-Wiedemann syndrome. *Pediatr Blood Cancer*. 2017;64(8):e26432. doi:10.1002/pbc.26432.
46. Dome JS and Huff V. Wilms tumor predisposition. In: Pagon RA, Adam MP, Bird TD et al., eds. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle; 2003-2022. https://www.ncbi.nlm.nih.gov/pubmed/20301471
47. Fischbach BV, Trout KL, Lewis J, Luis CA, Sika M. WAGR Syndrome: A clinical review of 54 Cases. *Pediatrics*. 2005;116(4):984-988. doi:10.1542/peds.2004-0467.
48. Gupta S, Weiss JM, Axell L, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 1.2025 —June 13, 2025, Genetic/Familial High-Risk Assessment: Colorectal, Endometrial, and Gastric available at: https://www.nccn.org/professionals/physician_gls/pdf/genetics_ceg.pdf. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Genetic/Familial High-Risk Assessment: Colorectal, Endometrial, and Gastric V1.2025 June 13, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
49. Kennedy RD, Potter DD, Moir CR, Mounif E. The natural history of familial adenomatous polyposis syndrome: A 24 year review of a single center experience in screening, diagnosis, and outcomes, *J Pediatr Surg*. 2014;49(1):82-86. doi:10.1016/j.jpedsurg.2013.09.033.
50. Jasperson KW, Patel SG, Ahnen DJ. APC-Associated Polyposis Conditions. In: Pagon RA, Adam MP, Bird TD, et al., ed. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle; 1998-2022. https://www.ncbi.nlm.nih.gov/books/NBK1345/.
51. Stoffel EM, Mangu PB, Gruber SB, et al. Hereditary colorectal cancer syndromes: American Society of Clinical Oncology clinical practice guideline endorsement of the familial risk-colorectal cancer: European Society for Medical Oncology clinical practice guidelines. *J Clin Oncol*. 2015;33(2):209-217. doi:10.1200/JCO.2014.58.1322.
52. Giusti F, Marini F, Brandi ML. Multiple endocrine neoplasia Type 1. In: Pagon RA, Adam MP, Ardinger HH et al, ed. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle; 2005-2022. https://www.ncbi.nlm.nih.gov/books/NBK1538/.
53. Thakker RV, Newey PJ, Walls GV, et al. Clinical Practice guidelines for multiple endocrine neoplasia type 1 (MEN1). *J Clin Endocrinol Metab*. 2012;97(9):2990-3011. doi:10.1210/jc.2012-1230
54. Giri D, McKay V, Weber A, Blair JC. Multiple endocrine neoplasia syndromes 1 and 2: manifestations and management in childhood and adolescence. *Arch Dis Child*. 2015;100(10):994-999. doi:10.1136/archdischild-2014-307028.
55. Eng C, Plitt G. Multiple endocrine neoplasia Type 2. In: Pagon RA, Adam MP, Ardinger HH et al, eds. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle; 1993-2023. https://www.ncbi.nlm.nih.gov/books/NBK1257/.
56. Northrup H, Koenig MK, Pearson DA, Au KS. Tuberous Sclerosis Complex. In: Pagon RA, Adam MP, Ardinger HH et al, eds. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle; 1993-2024. https://www.ncbi.nlm.nih.gov/pubmed/20301399.
57. Krueger DA, Care MM, Agricola K, Tudor C, Mays M, Franz DN. Everolimus long-term safety and efficacy in subependymal giant cell astrocytoma. *Neurology*. 2013;80(6):574-580. doi:10.1212/WNL.0b013e3182815428.

58. Wheless JW, Klimo P. Subependymal giant cell astrocytomas in patients with tuberous sclerosis complex: considerations for surgical or pharmacotherapeutic intervention. *J Child Neurol*. 2014;29(11):1562-1571. doi:10.1177/0883073813501870.
59. van Leeuwaarde RS, Ahmad S, Links TP, Giles RH. Von Hippel-Lindau Syndrome. In: Pagon RA, Adam MP, Ardinger HH et al, eds. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle;2000-2025. https://www.ncbi.nlm.nih.gov/books/NBK1463/.
60. Nielsen SM, Rhodes L, Blanco I, et al. Von Hippel-Lindau Disease: genetics and role of genetic counseling in a Multiple Neoplasia Syndrome. *J Clin Oncol*. 2016;34(18):2172-2181.
61. Rednam SP, Erez A, Druker H, et al. Von Hippel-Lindau and hereditary pheochromocytoma/paraganglioma syndromes: clinical features, genetics, and surveillance recommendations in childhood. *Clin Cancer Res*. 2017;23:e68-e75. doi: 10.1158/1078-0432.CCR-17-0547.
62. Kordes U, Bartelheim K, Modena P, et al. Favorable outcome of patients affected by rhabdoid tumors due to Rhabdoid Tumor Predisposition Syndrome (RTPS). *Pediatr Blood Cancer*. 2014;61(5):919-921. doi:10.1002/pbc.24793.
63. Sredni ST, Tomita T. Rhabdoid Tumor Predisposition Syndrome. *Pediatr Dev Pathol*. 2015;18(1):49-58. doi:10.2350/14-07-1531-MISC.1.
64. Nemes K, Bens S, Bourdeaut F, et al. Rhabdoid tumor predisposition syndrome. In: Pagon RA, Adam MP, Ardinger HH et al, eds. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle; 2017-2022. https://www.ncbi.nlm.nih.gov/books/NBK469816/.
65. Lohmann DR, Gallie BL. Retinoblastoma. In: Pagon RA, Adam MP, Ardinger HH et al, eds. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle; 2000-2023. https://www.ncbi.nlm.nih.gov/books/NBK1452/.
66. Else T, Greenberg S, Fishbein L. Hereditary Paraganglioma-Pheochromocytoma Syndromes. In: Adam MP, Everman DB, Mirzaa GM, Pagon RA, Wallace SE, Bean LJH, Gripp KW, Amemiya A, editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 2008-2023. https://www.ncbi.nlm.nih.gov/pubmed/20301715.
67. Gripp KW, Weaver KN. Costello Syndrome. In: Pagon RA, Adam MP, Ardinger HH et al, eds. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle; 2006-2023. https://www.ncbi.nlm.nih.gov/books/NBK1507/.
68. Tabori U, Hansford JR, Achatz MI, et al. Clinical management and tumor surveillance recommendations of inherited mismatch repair deficiency in childhood. *Clin Cancer Res*. 2017;23:e32-e37. doi: 10.1158/1078-0432.CCR-17-0574.
69. Krueger DA, Northrup H. Tuberous sclerosis complex surveillance and management: recommendations of the 2012 International Tuberous Sclerosis Complex Consensus Conference. *Pediatr Neurol*. 2013;49(4):255–265. doi:10.1016/j.pediatrneurol.2013.08.002.
70. Campen CJ, Gutmann DH. Optic pathway gliomas in neurofibromatosis type 1. *J Child Neurol*. 2018;33(1):73–81. doi:10.1177/0883073817739509.
71. Rosser T. Neurocutaneous disorders. *CONTINUUM*. 2018;24(1, Child Neurology):96-129. doi:10.1212/CON0000000000000562.
72. Motzer RJ, Jonasch E, Agarwal N, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 1.2026 – July 24, 2025. Kidney Cancer, available at: https://www.nccn.org/professionals/physician_gls/pdf/kidney.pdf. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Kidney Cancer V1.2026 – July 24, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
73. Tuberous Sclerosis Alliance. Diagnosis, surveillance, and management of individuals with tuberous sclerosis complex. Silver Spring, MD. *Tuberous Sclerosus Alliance*; 2014. https://www.tsalliance.org/healthcare-professionals/diagnosis/.
74. Lam H, Siroky BJ, Henske EP. Renal disease in tuberous sclerosis complex: pathogenesis and therapy. *Nat Rev Nephrol*. 2018;14(11):704-706. doi:10.1038/s41581-018-0059-6.
75. Sattler EC, Ortud KS. Birt-Hogg-Dube Syndrome. In: Pagon RA, Adam MP, Ardinger HH et al, eds. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle;2006-2024. https://www.ncbi.nlm.nih.gov/books/NBK1522/.

76. Pilarski R, Rai K, Cebulla C, et al. BAP1 Tumor Predisposition Syndrome. In: Pagon RA, Adam MP, Ardinger HH et al, eds. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle;2016-2024. https://www.ncbi.nlm.nih.gov/books/NBK390611/.
77. Gläsker S, Vergauwen E, Koch CA, Kutikov A, Vortmeyer AO. Von Hippel-Lindau disease: current challenges and future prospects. *Onco Targets Ther*. 2020:13;5669-5690. doi:10.2147/OTT.S190753.
78. VHL Alliance. VHLA Suggested Active Surveillance Guidelines. Revised April 24, 2020. Accessible from https://www.vhl.org/.
79. Balis F, Green DM, Armstrong A, et. al. National Comprehensive Cancer Network (NCCN) Guidelines Version 2.2025 – June 11, 2025. Wilms Tumor (Nephroblastoma), available at: https://www.nccn.org/professionals/physician_gls/pdf/wilms_tumor.pdf, Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Wilms Tumor (Nephroblastoma) V2.2025 – June 11, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
80. Todd K, Kim HK, Szabo S, et. al. Solitary myofibroma preceding the development of multicentric myofibromatosis: A report of two cases with surveillance recommendations. *Pediatr Blood Cancer*. 2020;67(10):e28266. doi:10.1002/pbc.28266.
81. Orbach, D, Bodemer C. Infantile myofibromatosis. *Orphanet*. 2019. https://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=GB&Expert=2591
82. National Cancer Institute. *Infantile myofibromatosis*. 2020. https://www.cancer.gov/pediatric-adult-rare-tumor/rare-tumors/rare-soft-tissue-tumors/infantile-myofibromatosis.
83. Tang ER, Khalatbari H, Parisi MT. Utility of 18F-FDG PET/CT in infantile myofibromatosis. *Clin Nucl Med*. 2019;44:676-679. doi:10.1097/RLU.0000000000002627.
84. Evans DGR, Salvador H, Chang VY, et al. Cancer and Central Nervous System Tumor Surveillance in Pediatric Neurofibromatosis 1. *Clin Cancer Res*. 2017;23(12):e46-e53. doi: 10.1158/1078-0432.CCR-17-0589.
85. Gottumukkala RV, Gee MS, Hampilos PJ, et al. Current and Emerging Roles of Whole-Body MRI in Evaluation of Pediatric Cancer Patients . *Radiographics*. 2019;39(2):516-534. doi:10.1148/rg.2019180130.
86. Al-Sarhani H, Gottumukkala RV, Grasparil ADS 2nd, Tung EL, Gee MS, Greer MC. Screening of cancer predisposition syndromes. *Pediatr Radiol*. 2021. doi:10.1007/s00247-021-05023-w.
87. Grasparil AD 2nd, Gottumukkala RV, Greer MC, Gee MS. Whole-Body MRI surveillance of cancer predisposition syndromes: current best practice guidelines for use, performance, and interpretation. *AJR Am J Roentgenol*. 2020;215(4):1002-1011. doi:10.2214/AJR.19.22399.
88. Yang J, Gurudu SR, Koptiuch C, et.al. American Society for Gastrointestinal Endoscopy guideline on the role of endoscopy in familial adenomatous polyposis syndromes. *Gastrointestinal Endoscopy*. 2020;91(5):963-82. doi:10.1016/j.gie.2020.01.028.
89. PDQ® Cancer Genetics Editorial Board. PDQ Birt-Hogg-Dubé Syndrome. Bethesda, MD: National Cancer Institute. Updated 12/16/2024. Available at: https://www.cancer.gov/types/kidney/hp/renal-cell-carcinoma-genetics/bhd-syndrome.
90. Frühwald MC, Nemes K, Boztug H, et al. Current recommendations for clinical surveillance and genetic testing in rhabdoid tumor predisposition: a report from the SIOPE Host Genome Working Group. *Fam Cancer*. 2021;20(4):305-316. doi:10.1007/s10689-021-00229-1.
91. Wolters WPG, Dreijerink KMA, Giles RH, et al. Multidisciplinary integrated care pathway for von Hippel-Lindau disease. *Cancer*. 2022;128(15):2871-2879. doi:10.1002/cncr.34265.
92. Schultz KAP, Williams GM, Kamihara J, et. al. DICER1 and Associated conditions: identification of at-risk individuals and recommended surveillance strategies. *Clin Cancer Res*. 2018;24(10):2251-2261. doi:10.1158/1078-0432.CCR-17-3089.
93. Flanagan M, Cunniff CM. Bloom Syndrome. In: Adam MP, Mirzaa GM, Pagon RA, Wallace SE, Bean LJH, Gripp KW, Amemiya A, editors. *GeneReviews®[Internet]*. Seattle (WA): University of Washington, Seattle; 2006–2023. https://www.ncbi.nlm.nih.gov/books/NBK1398/
94. Cunniff C, Djavid AR, Carrubba S, et. al. Health supervision for people with Bloom syndrome. *Am J Med Genet A*. 2018;176(9):1872-1881. doi:10.1002/ajmg.a.40374.
95. Taieb D, Jha A, Treglia G, Pacak K. Molecular imaging and radionuclide therapy of pheochromocytoma and paraganglioma in the era of genomic characterization of disease subgroups. *Endocr Relat Cancer*. 2019;26(11):R627-R652. doi:10.1530/ERC-19-0165.

96. Amar L, Pacak K, Steichen O, et. al. International consensus on initial screening and follow-up of asymptomatic SDHx mutation carriers. *Nature Reviews Endocrinology*. 2021;17:435-445. doi:10.1038/s41574-021-00492-3.

# Pediatric Leukemias (PEDONC-3)

Guideline Page







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# Pediatric Leukemia General Considerations (PEDONC-3.1)

**ONCP.LE.0003.1.A**

**v1.0.2026**

- MRI Brain without and with contrast (CPT® 70553) is medically necessary in individuals exhibiting CNS symptoms
  - CT Head without or with contrast (CPT® 70450 or CPT® 70460) is 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 (ONC-29)** for imaging guidelines related to transplant

**Background and Supporting Information**

- The overwhelming majority of leukemias occurring in children are acute. Chronic myelogenous leukemia (CML) is rare in children, and the occurrence of chronic lymphocytic leukemia (CLL) appears to have only been reported once in pediatric individuals to date
- Routine advanced imaging is not indicated in the evaluation and management of chronic myeloid leukemia in the absence of specific localizing clinical symptoms or clearance for hematopoietic stem cell transplantation

# Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)

**ONCP.LE.0003.2.C**
**v1.0.2026**

- 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
- Scrotal Ultrasound CPT® 76870 and/or doppler ultrasound of the scrotum CPT® 93975 or 93976 is medically necessary for suspected testicular involvement.
- Chest x-ray should be performed to evaluate for mediastinal mass in suspected cases or upon initial diagnosis.
  - CT Chest with contrast (CPT® 71260) is medically necessary to immediately to evaluate for airway compression and anesthesia safety prior to attempting histologic diagnosis if mediastinal widening is seen on chest x-ray
  - In individuals with known or strongly suspected T-cell histology or other suspected lymphoblastic lymphoma involvement, EITHER of the following is medically necessary for initial staging purposes:
    - 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 medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**
- MRI Brain without and with contrast (CPT® 70553) is medically necessary for individuals exhibiting CNS symptoms.
  - Imaging due to CSF tumor burden has not been shown to improve the detection of CNS involvement compared with CSF alone.
    - CT Head without or with contrast (CPT® 70450 or CPT® 70460) is medically necessary for urgent concerns where MRI would delay care (i.e., suspected CNS bleeding)

**Additional imaging in lymphoblastic lymphoma/lymphomatous extramedullary disease:**

- CT is medically necessary to assess response to therapy **only** for individuals with known bulky nodal disease (usually with T-cell histology) at the end of induction (4

to 6 weeks). CT is medically necessary for individuals with residual masses every new therapy phase (consolidation, interim maintenance, etc., generally every 8 to 12 weeks) until disease resolution is seen
- PET/CT (CPT® 78815) is medically necessary when residual mass ≥8 mm 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.
- Chest x-ray or Abdominal ultrasound (CPT® 76700) **only**, as indicated by site(s) of bulky disease present at diagnosis, is medically necessary for further surveillance, once CT imaging shows no evidence of disease.
- CT of all involved bulky nodal areas is medically necessary for individuals with persistent residual masses performed as part of an end of therapy evaluation

**Immunosuppression during ALL therapy:**

- CT or MRI is medically necessary 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
- 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 medically necessary
- MRA/MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) is medically necessary for the following:
  - To rule out bleeding associated with sinus venous thrombosis in individuals treated with asparaginase

**Imaging during therapy for relapsed ALL:**

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

**Imaging of known or suspected osteonecrosis in ALL:**

- MRI without contrast or without and with contrast of the affected joint(s) is medically necessary for symptoms suggesting osteonecrosis
  - CT without contrast is medically necessary when MRI is contraindicated or unavailable, or for diagnosis of suspected subchondral fracture

- MRI Bilateral Hips (CPT® 73721 or CPT® 73723 with modifier -50) is medically necessary once at 6 to 9 months after diagnosis for individuals age ≥11 years
- Repeat MRI without contrast of the affected joint(s) is medically necessary every 2 cycles of maintenance (every 6 months) if reintroduction of corticosteroids is being considered in individuals whose symptoms have resolved and are still receiving active treatment
- MRI without contrast of the affected joint(s) is medically necessary for preoperative planning for individuals undergoing core decompression
- See: **Osteonecrosis In Long Term Cancer Survivors (PEDONC-19.4)** for information on osteonecrosis in ALL individuals who have completed therapy

**Background and Supporting Information**

- The majority of individuals with ALL have B-precursor ALL and routine advanced imaging is not necessary.
- Individuals with ALL who relapse are treated with very intensive chemotherapy regimens and most spend the majority of their chemotherapy treatment phase in the hospital
- Individuals may have therapy-induced hypogammaglobulinemia which requires supplemental intravenous immune globulin (IVIG) during maintenance therapy. Those receiving supplemental IVIG should be treated similarly to those with ANC <500 with regards to imaging for infectious disease
- Osteonecrosis (ON) in individuals with ALL is a relatively common complication of ALL and its treatment, primary corticosteroids. Approximately 3% of younger children and 12 to 15% of adolescents are affected by ON at some point during therapy. The peak incidence occurs approximately one year from the time of diagnosis
- Screening MRI of asymptomatic individuals age ≤10 years to detect osteonecrosis has not been shown to impact outcomes, and it is not standard to alter treatment based on imaging findings alone without symptoms
- If osteonecrosis is detected on initial MRI, corticosteroids are often withheld during maintenance chemotherapy (but continued in earlier phases of therapy).

# Acute Myeloid Leukemia (AML) (PEDONC-3.3)

**ONCP.LE.0003.3.A**

**v1.0.2026**

- Frequent CT or MRI imaging is medically necessary to evaluate known sites of invasive fungal infection
- Surveillance imaging of asymptomatic individuals is 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 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**

- The majority of AML individuals do not have any bulky disease and routine advanced imaging is not necessary
- AML individuals are treated with very intensive chemotherapy regimens and spend the majority of their chemotherapy treatment phase in the hospital





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# References (PEDONC-3)

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1. Inaba H, Teachey D, Annesley C, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 1.2026—August 11, 2025 Pediatric Acute Lymphoblastic Leukemia, available at: https://www.nccn.org/professionals/physician_gls/pdf/ped_all.pdf. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Pediatric Acute Lymphoblastic Leukemia V1.2026 August 11, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
2. Ranta S, Palomäki M, Levinsen M, et al. Role of neuroimaging in children with Acute Lymphoblastic Leukemia and central nervous system involvement at diagnosis. *Pediatr Blood Cancer*. 2016;64:64-70. doi:10.1002/pbc.26182/epdf.
3. Pui C-H, Yang JJ, Hunger SP, et al. Childhood Acute Lymphoblastic Leukemia: progress through collaboration. *J Clin Oncol*. 2015;33(27):2938-2948. doi: 10.1200/JCO.2014.59.1636.
4. Kawedia JD, Kaste SC, Pei D, et al. Pharmacokinetic, pharmacodynamic, and pharmacogenetic determinants of osteonecrosis in children with acute lymphoblastic leukemia. *Blood Journal*. 2011;117(8):2340-2347. doi:10.1182/blood-2010-10-311969.
5. Marcucci G, Beltrami G, Tamburini A, et al. Bone health in childhood cancer: review of the literature and recommendations for the management of bone health in childhood cancer survivors. *Ann Oncol* .2019;30:908-920. doi:10.1093/annonc/mdz120.
6. Vora A. Management of osteonecrosis in children and young adults with acute lymphoblastic leukaemia. *Br J Haematol*. 2011;155(5):549-560. doi:10.1111/j.1365-2141.2011.08871.x.
7. Kaste SC, Pei D, Cheng C, et al. Utility of early screening magnetic resonance imaging for extensive hip osteonecrosis in pediatric patients treated with glucocorticoids. *J Clin Oncol*. 2015;33(6):610-615. doi:10.1200/JCO.2014.57.5480.
8. Niinimäki T, Harila-Saari A, Niinimäki R. The diagnosis and classification of osteonecrosis in patients with childhood Leukemia. *Pediatr Blood Cancer*. 2015;62(2):198-203. doi:10.1002/pbc.25295.
9. Murphey MD, Roberts CC, Bencardino JT, et al. Osteonecrosis of the hip. ACR Appropriateness Criteria® 2015;1-12. https://acsearch.acr.org/docs/69420/Narrative/.
10. Karol SE, Mattano LA, Yang W, et al. Genetic risk factors for the development of osteonecrosis in children under age 10 treated for acute lymphoblastic leukemia. *Blood Journal*. 2016;127(5):558-564. doi:10.1182/blood-2015-10-673848.
11. Chavhan GB, Babyn PS, Nathan PC, et al. Imaging of acute and subacute toxicities of cancer therapy in children. *Pediatr Radiol*. 2016;46(1):9-20. doi:10.1007/s00247-015-3454-1.
12. Lauer M, Kernen E, Schwabe D, Lehrnbecher T, Porto L. The role of magnetic resonance imaging in the diagnosis of central nervous system involvement in children with acute lymphoblastic leukemia. *Pediatr Blood Cancer*. 2020;e28294. doi:10.1002/pbc.28294.

# CNS Tumors (PEDONC-4)

Guideline Page







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# CNS Tumors General Considerations (PEDONC-4.1)

**ONCP.CT.0004.1.A**

**v1.0.2026**

- 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
  - 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 indicated, if being performed immediately following a contrast-enhanced MRI Brain
- Functional MRI (fMRI) (CPT® 70555 or CPT® 70554) is medically necessary to depict spatial relationships between eloquent cortex and neoplasms for preoperative planning and to promote safe resections (following baseline MRI Brain).
- Isotropic volumetric MRI (CPT® 76376, 76377, 0865T, or 0866T) is medically necessary for preoperative planning to accurately localize tumors.
- MRI Orbits without and with contrast (CPT® 70543) is medically necessary in individuals who present with papilledema, altered vision, strabismus, nystagmus, anisocoria, proptosis, ocular cranial nerve palsies, coloboma, or leukocoria.

**CT Considerations**

- CT for evaluation of ventriculomegaly or other operative considerations, or for children who cannot undergo MRI safely
  - CT for evaluation of headaches related to head trauma or evaluation of skull or facial bone abnormalities
  - Post-contrast CT is generally not indicated
- 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 **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** 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
- MRS is considered not medically necessary for all other histologies and indications not listed in a diagnosis-specific guideline section
- MR spectroscopy is not medically necessary for routine surveillance

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

- PET Brain Metabolic imaging (CPT® 78608) is **only** 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 medically necessary for routine surveillance
- PET Brain Metabolic imaging is considered not medically necessary for all other histologies and indications not listed in a diagnosis-specific guideline section
- PET Brain Perfusion imaging (CPT® 78609) is not medically necessary in the evaluation or management of primary CNS tumors
- Fusion PET/CT studies (CPT® 78814, CPT® 78815, or CPT® 78816) are not 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 medically necessary
  - MRI Brain without and with contrast (CPT® 70553) is 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
    - 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 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.

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

**ONCP.CT.0004.2.C**
**v1.0.2026**

- MR Spectroscopy and PET Brain Metabolic are not medically necessary for routine surveillance.
- MRI is generally superior to CT for staging and restaging CNS malignancies, but CT is medically necessary in accordance with these guidelines where MRI is contraindicated.

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging of all LGG | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)<br>• 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 | Either or both of the following:<br>• Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T)<br>• Functional MRI (fMRI) (CPT® 70555 or 70554) |
| • At any time, for ANY of the following:<br>◦ For rapid assessment in the acute setting<br>◦ Evaluation of acute intracranial hemorrhage<br>◦ Evaluation of ventriculomegaly<br>◦ Evaluation of shunt-related issues | • CT Head without contrast (CPT® 70450) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| • ANY of the following:<br>◦ Determining the need for biopsy when transformation to high grade glioma is suspected based on clinical symptoms or recent MRI findings<br>◦ 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:<br>◦ Distinguish low grade from high grade gliomas<br>◦ Evaluate a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed<br>◦ Distinguish radiation-induced tumor necrosis from progressive disease within 18 months of completing radiotherapy | • MR Spectroscopy (MRS, CPT® 76390) |
| Baseline imaging after resection, to assess degree of resection | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI with and without contrast at level of resected spinal site<br>• If orbital resection, MRI Orbits without and with contrast (CPT® 70543) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Treatment response at the completion of radiotherapy | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI with and without contrast at level of irradiated spinal site<br>• If prior orbital involvement, MRI Orbits without and with contrast (CPT® 70543) |
| Treatment response on chemotherapy | • MRI Brain without and with contrast (CPT® 70553) approved every 2 cycles during active treatment and at the end of planned chemotherapy<br>• If prior orbital involvement, MRI Orbits without and with contrast (CPT® 70543) |
| Additional treatment response imaging during induction chemotherapy for individuals 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 |
| Surveillance, including individuals with unresected disease on observation only | • For individuals with intracranial primary:<br>◦ MRI Brain without and with contrast (CPT® 70553) every 3 months for 2 years, then every 6 months for 3 years, then annually thereafter<br>◦ If prior orbital involvement, MRI Orbits without and with contrast (CPT® 70543)<br>◦ MRI Spine is not medically necessary during surveillance in individuals without prior history of spinal involvement except to evaluate symptoms suspicious for spinal cord recurrence<br>• For individuals with a prior history of spine primary tumor or metastatic spinal involvement:<br>◦ 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 |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Surveillance imaging for individuals with optic pathway glioma and a history of NF1 | • Every 3 months for 2 years, then every 6 months for 3 years, then annually thereafter:<br>◦ MRI Brain without and with contrast (CPT® 70553) and<br>◦ MRI Orbits without and with contrast (CPT® 70543) |
| Suspected intracranial or intraspinal recurrence | • All imaging supported in initial staging criteria |

## Background and Supporting Information

| Includes the following tumors: |
|---|
| • Pediatric-type diffuse low-grade gliomas<br>◦ Diffuse astrocytoma, MYB- or MYBL1-altered<br>◦ Angiocentric glioma<br>◦ Polymorphus low-grade neuroepithelial tumor of the young<br>◦ Diffuse low-grade glioma, MAPK pathway-altered<br>• Circumscribed astrocytic gliomas<br>◦ Pilocytic astrocytoma<br>◦ High-grade astrocystoma with piloid features<br>◦ Pleomorphic xanthoastrocytoma<br>◦ Subependymal giant cell astrocystoma (SEGA)<br>◦ Choroid glioma<br>◦ Astroblastoma, MN<br>• Glioneuronal and neuronal tumors<br>◦ Gangliоglioma<br>◦ Desmoplastic infantile gangliоglioma/desmoplastic infantile astrocystoma<br>◦ Dysembryoplastic neuroepithelial tumor<br>◦ Diffuse glioneuronal tumor with oligodendroglioma-like features and nuclear clusters<br>◦ Papillary glioneuronal tumor<br>◦ Rosette-forming glioneuronal tumor<br>◦ Myxoid glioneuronal tumor<br>◦ Diffuse leptomeningeal glioneuronal tumor<br>◦ Gangliocytoma<br>◦ Multinodular and vacuolating neuronal tumor<br>◦ Dysplastic cerebellar gangliocytoma (Lhermitte-Duclos disease)<br>◦ Central neurocytoma<br>◦ Extraventricular neurocytoma<br>◦ Cerebellar liponeurocytoma<br>• Any other glial tumor with a WHO grade I or II classification |

**General Considerations:**

- Account for 40 to 60% of pediatric CNS tumors.
- These tumors are defined as having a WHO grade of I or II (out of IV), can occur anywhere in the CNS

**Treatment Considerations**

- Children with neurofibromatosis and small optic pathway tumors may not undergo biopsy or resection and will proceed directly to treatment or surveillance
- Children on observation without specific treatment should be imaged according to surveillance guidelines for LGG
- Individuals who undergo complete resection should be imaged according to surveillance guidelines after post-resection imaging
- Individuals age >10 years with incompletely resected tumors usually receive adjuvant radiation therapy
- Individuals age ≤10 years with incompletely resected tumors are commonly treated with chemotherapy





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

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| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging of all HGG | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| In addition to the above imaging, for preoperative planning | Either or both of the following:<br>• Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T)<br>• Functional MRI (fMRI) (CPT® 70555 or 70554) |
| • At any time, for ANY of the following:<br>◦ For rapid assessment in the acute setting<br>◦ Evaluation of acute intracranial hemorrhage<br>◦ Evaluation of ventriculomegaly<br>◦ Evaluation of shunt-related issues | • CT Head without contrast (CPT® 70450) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| • ANY of the following:<br>◦ Distinguishing radiation-induced tumor necrosis from progressive disease within 18 months of completing radiotherapy<br>◦ 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<br>◦ 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:<br>◦ To distinguish low grade from high grade gliomas<br>◦ To evaluate a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed<br>◦ 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 | • Functional MRI (fMRI) (CPT® 70544 or CPT® 70555) |
| Baseline imaging following resection, to assess degree of resection | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast of resected area, or whole spine if requested (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Treatment response at the completion of radiotherapy | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast of irradiated area, or whole spine if requested (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| Treatment response on chemotherapy | • MRI Brain without and with contrast (CPT® 70553) every 2 cycles during active treatment and at the end of planned chemotherapy |
| Additional treatment response imaging during induction chemotherapy for individuals 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 |
| Signs or symptoms of recurrence or progression | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |

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

## Background and Supporting Information

| Includes the following tumors: |
|---|
| • Diffuse midline glioma, H3 K27-altered<br>• Diffuse hemispheric glioma, H3 G34-mutant<br>• Diffuse pediatric-type high-grade glioma, H3-wildtype and IDH-wildtype<br>• Infant-type hemispheric glioma<br>• Any other glial tumor with a WHO grade of III or IV classification |

**General Considerations:**

- Rare in children compared with the adult population, but represent 10% to 20% of pediatric CNS tumors
- Prognosis is very poor, and survival significantly beyond 3 years from diagnosis is rare, even with complete surgical resection at initial diagnosis
- These tumors are defined as having a WHO histologic grade of III or IV (out of IV) can occur anywhere in the CNS (though the majority occur in the brain)

**Treatment Considerations:**

- Individuals who undergo complete resection should be imaged according to surveillance guidelines after post-resection imaging
- Individuals with incompletely resected tumors are commonly treated with chemotherapy





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

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- MR Spectroscopy and PET Brain Metabolic are not medically necessary for routine surveillance.

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging for all individuals, preoperatively | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| In addition to the above imaging, for preoperative planning | Either or both of the following:<br>• Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0867T, or 0866T)<br>• Functional MRI (fMRI) (CPT®) 70555 or 70554) |
| • At any time, for ANY of the following:<br>◦ For rapid assessment in the acute setting<br>◦ Evaluation of acute intracranial hemorrhage<br>◦ Evaluation of ventriculomegaly<br>◦ Evaluation of shunt-related issues | • CT Head without contrast (CPT® 70450) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| • ANY of the following:<br>◦ To distinguish radiation-induced tumor necrosis from progressive disease within 18 months of completing radiotherapy<br>◦ 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<br>◦ 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) |
| • ANY of the following:<br>◦ 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 | • MRI Brain without and with contrast (CPT® 70553)<br>• 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 | • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Treatment response | • At the start of adjuvant chemotherapy and every 2 cycles until therapy is completed:<br>◦ MRI Brain without and with contrast (CPT® 70553)<br>◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic CPT® 72157, Lumbar-CPT® 72158) |
| Children age <3 years 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. |
| End of treatment evaluation | • MRI Brain without and with contrast (CPT® 70553)<br>• 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 | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Surveillance | For low- or average-risk individuals:<br>• Every 3 months for 2 years, then every 6 months for 3 years:<br>◦ MRI Brain without and with contrast (CPT® 70553)<br>• Every 6 months for 2 years:<br>◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)<br>For high- or very high-risk individuals:<br>• Every 3 months for 2 years, then every 6 months for 3 years:<br>◦ MRI Brain without and with contrast (CPT® 70553)<br>• Every 3 months for 2 years, then every year for 3 years:<br>◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)<br>• 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)** |





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## Background and Supporting Information

| Includes the Following Tumors: |
|---|
| • Medulloblastoma<br>◦ Molecularly-defined Medulloblastoma<br>▪ WNT-activated Medulloblastoma<br>▪ SHH-activated and TP53-wildtype Medulloblastoma<br>▪ SHH-activated and TP53-mutant Medulloblastoma<br>▪ Non-WNT/Non-SHH<br>◦ Medulloblastoma, histologically defined<br>• Other CNS Embryonal Tumors (previously supratentorial primitive neuro-ectodermal tumors)<br>◦ CNS embryonal tumor<br>◦ Cribriform neuroepithelial tumor<br>◦ Embryonal tumor with multilayered rosettes<br>◦ CNS neuroblastoma<br>◦ FOXR2-activated CNS tumor with BCOR internal tandem duplication<br>• Pineoblastoma |
| **Risk Assessment is Important in Determining Optimal Treatment**<br>**High-Risk Features Include the Following:** |
| • Spinal metastasis (including cytology positive only)<br>• Multifocal intracranial tumors<br>• Anaplastic histology<br>• All other CNS embryonal tumors and pineoblastomas<br>• >1.5 $cm^2$ residual tumor area on postoperative MRI and age <3 years |
| **Individuals without any high-risk features are considered "Average Risk"** |

**General Considerations:**

- Account for 15% to 25% of pediatric CNS tumors
- Prognosis is generally favorable
- Leptomeningeal spread is common and can occur after initial diagnosis

**Treatment Considerations:**

- Individuals generally proceed to chemoradiotherapy within 31 days of surgical resection

- All individuals receive adjuvant chemotherapy lasting 6 to 12 months that begins ~6 weeks after completion of chemoradiotherapy

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

**ONCP.CT.0004.5.C**

**v1.0.2026**

- PET Brain Metabolic does not have a defined role in the evaluation of ATRT at this time and is, therefore, not medically necessary.
- MR Spectroscopy is not medically necessary for routine surveillance.

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging for all individuals, preoperatively | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)<br>• Renal US (CPT® 76770)<br>◦ 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 | Either or both of the following:<br>• Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0867T, or 0866T)<br>• Functional MRI (fMRI) (CPT® 70555 or 70554) |
| • At any time, for ANY of the following:<br>◦ For rapid assessment in the acute setting<br>◦ Evaluation of acute intracranial hemorrhage<br>◦ Evaluation of ventriculomegaly<br>◦ Evaluation of shunt-related issues | • CT Head without contrast (CPT® 70450) |

| 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 | • MR Spectroscopy (CPT® 76390) |
| Postoperative (preferably within 48 hours of surgery) to quantify residual tumor volume | • MRI Brain without and with contrast (CPT® 70553) |
| Within 28 days post-op, if spinal imaging was not performed preoperatively | • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| Treatment response to induction chemotherapy | • After every 2 cycles:<br>◦ MRI Brain without and with contrast (CPT® 70553)<br>◦ 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 | • Disease evaluation is 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 |
| End of treatment evaluation | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| Signs or symptoms of recurrence or progression | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Surveillance | • Every 3 months for 2 years, then every 6 months for 3 years:<br>◦ MRI Brain without and with contrast (CPT® 70553)<br>◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)<br>• Further imaging only for signs and symptoms of recurrence<br>• 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**

**General Considerations:**

- Highly aggressive tumor occurring primarily in very young children that has a clinical presentation very similar to medulloblastoma with a much higher rate of leptomeningeal spread.
- Metastases can occur outside the CNS, and associated tumors can also arise in the kidneys (Malignant Rhabdoid Tumor of the Kidney, MRT).
  - 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)**
- Overall prognosis is poor, with <20% of individuals surviving beyond 2 years from the diagnosis.
- Individuals generally proceed to induction chemotherapy shortly following surgical resection or biopsy.
- Following completion of chemotherapy some individuals will proceed to radiotherapy.
  - MRI performed at the end of consolidation therapy should serve as the diagnostic MRI prior to radiotherapy.





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# Pineocytomas and Pineal Parenchymal Tumors (PEDONC-4.6)

**ONCP.CT.0004.6.C**
**v1.0.2026**

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging for all individuals | • MRI Brain without and with contrast (CPT® 70553) |
| In addition to the above imaging, for preoperative planning | Either or both of the following:<br>• Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, 0866T)<br>• Functional MRI (fMRI) (CPT® 70555 or 70554) |
| • Additional initial staging imaging for individuals with:<br>◦ Multicentric tumors<br>◦ Atypical histology including pineoblastoma-like elements (grade 2 or 3 pineal parenchymal tumors which have not been considered a pineoblastoma)<br>◦ Clinical signs or symptoms suggesting spinal cord involvement | • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| Baseline imaging following resection | • MRI Brain without and with contrast (CPT® 70553) |
| End of radiotherapy | • MRI Brain without and with contrast (CPT® 70553) |
| Additional imaging at end of radiotherapy for individuals with measurable spinal cord disease on MRI | • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Surveillance | • 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 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 | • MRI Brain without and with contrast (CPT® 70553) |
| Suspected spinal cord recurrence or progression | • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |

**Background and Supporting Information**

**General Considerations:**

- Low grade malignancy that is similar in presentation to low grade glioma (LGG)
- PET Brain Metabolic imaging and MR Spectroscopy do not have a defined role in the evaluation of pineocytoma
- Surgical resection is curative for most individuals
  - Individuals with a complete resections should then be imaged according to surveillance guidelines
- Individuals with incompletely resected tumors may receive adjuvant radiation therapy
  - After end of radiotherapy imaging, these individuals should be imaged according to surveillance guidelines

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

**ONCP.CT.0004.7.A**

**v1.0.2026**

- PET Metabolic Brain imaging does not have a defined role in the evaluation of CNS GCT and is, therefore, considered not medically necessary.

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging for all individuals | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| In addition to the above imaging, for preoperative planning | Either or both of the following:<br>• Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T)<br>• Functional MRI (fMRI) (CPT® 70555 or 70554) |
| • At any time, for ANY of the following:<br>◦ For rapid assessment in the acute setting<br>◦ Evaluation of acute intracranial hemorrhage<br>◦ Evaluation of ventriculomegaly<br>◦ 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 | • MR Spectroscopy (CPT® 76390) |
| Treatment response to induction chemotherapy | • MRI Brain without and with contrast (CPT® 70553) every 2 cycles |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Additional treatment response to induction chemotherapy for individuals 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 |
| End of induction chemotherapy for individuals with localized intracranial tumors | • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| Prior to second-look surgery | • MRI of all known sites of measurable disease |
| Prior to radiotherapy | • MRI of all known sites of measurable disease |
| At the end of all planned therapy | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine (with or without and with contrast) |
| Signs or symptoms of recurrence or progression | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| Surveillance | • 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:<br>◦ MRI Brain without and with contrast (CPT® 70553)<br>◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)<br>• 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)** |





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| Indication | Medically Necessary Imaging Study |
|---|---|
| Suspected recurrence - new or worsening neurologic symptoms (including worsening of diabetes insipidus) | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |

**Background and Supporting Information**

| Includes the following tumors: |
|---|
| • CNS Germinoma<br>• Non-Germinomatous Germ Cell Tumors (NGGCT)<br>◦ Embryonal carcinoma<br>◦ Yolk sac tumor<br>◦ Choriocarcinoma<br>◦ Immature Teratoma<br>◦ Mature Teratoma<br>◦ Mixed germ cell tumor |

**General Considerations:**

- More common in older school age children and younger adolescents, but can occur throughout the pediatric age range
- Although leptomeningeal spread is common, prognosis is excellent due to high sensitivity to chemotherapy and radiotherapy
- Individuals generally proceed to chemotherapy shortly following surgical resection or biopsy and will usually receive 2 to 4 cycles
- Following completion of chemotherapy, individuals with residual disease will proceed to second-look surgery and/or radiotherapy

# Ependymal Tumors (Ependymoma) (PEDONC-4.8)

**ONCP.CT.0004.8.A**

**v1.0.2026**

- 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 medically necessary for routine surveillance

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging for all individuals | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| In addition to the above imaging, for preoperative planning | Either or both of the following:<br>• Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T)<br>• Functional MRI (fMRI) (CPT® 70555 or 70554) |
| Evaluation of 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) |
| Baseline imaging following resection | • MRI Brain without and with contrast (CPT® 70553) or MRI without and with contrast of involved spinal level(s) |
| Completion of radiotherapy | • MRI Brain without and with contrast (CPT® 70553) or MRI without and with contrast of involved spinal level(s) |
| Prior to radiotherapy | • MRI of all known sites of measurable disease |
| Treatment response to induction chemotherapy | • MRI Brain without and with contrast (CPT® 70553) or MRI without and with contrast of involved spinal level(s) every 2 cycles |

| Indication | Medically Necessary Imaging Study |
|---|---|
| End of induction chemotherapy and again at end of all therapy | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| Prior to second-look surgery | • MRI of all known sites of measurable disease |
| Signs or symptoms of recurrence or progression | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| 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<br>◦ 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)**<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) can be approved annually for 2 years |

| Indication | Medically Necessary Imaging Study |
|---|---|
| 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<br>◦ 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)**<br>• 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 |
| 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<br>• 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

**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

**General Considerations:**

- Occur primarily intracranially, roughly 2/3 in the posterior fossa
- Overall prognosis is very good, with supratentorial tumors faring better
- Primary spinal tumors can also occur, and are more common in adult individuals than pediatric individuals.
- Surgery is the primary treatment modality
- Individuals with a complete resection should then be imaged according to surveillance guidelines
- Radiotherapy +/- chemotherapy is used for:
  - Incompletely resected tumors
  - Anaplastic histology
  - Infratentorial location
    - Individuals with incomplete resection or high-risk histology that receive adjuvant radiation therapy should then be imaged according to surveillance guidelines after end of radiotherapy imaging.
- Individuals with gain of chromosome 1q have worse progression-free survival and overall survival outcomes
- RELA-fusion supratentorial ependymoma did not portend worse overall survival outcomes in recent Clinical Oncology Group studies, and is generally not considered an indication for more frequent surveillance imaging

# Malignant Tumors of the Spinal Cord (PEDONC-4.9)

**ONCP.CT.0004.9.A**
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- If a disease-specific guideline exists, image according to the guidance found in that disease-specific guideline section.
- 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.

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging for all individuals | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| In addition to the above imaging, for preoperative planning | Either or both of the following:<br>• Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T)<br>• Functional MRI (fMRI) (CPT® 70555 or 70554) |
| Treatment response, every 2 cycles | • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)<br>• Additionally, MRI Brain without and with contrast (CPT® 70553) for known intracranial disease |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Signs or symptoms of recurrence or progression | • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)<br>• Additionally, MRI Brain without and with contrast (CPT® 70553) for known intracranial disease |
| Surveillance of rare histologies is highly individualized.<br><br>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 indicated in these rare individuals. | • MRI Brain and/or MRI Spine may be considered |

**Background and Supporting Information**

- Treatment principles are the same as tumors of the brain, and should follow imaging guidelines according to the specific histologic type
- Multiple spinal cord tumors should raise suspicion for neurofibromatosis
- Asymptomatic surveillance imaging should generally end at the time point appropriate for the specific tumor type





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

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- 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.

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging for all individuals | • MRI Brain without and with contrast (CPT® 70553)<br>◦ Concurrent CT Head without contrast (CPT® 70450) in addition to MRI if craniopharyngioma is suspected |
| In addition to the above imaging, for preoperative planning | Either or both of the following:<br>• Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T)<br>• Functional MRI (fMRI) (CPT® 70555 or 70554) |
| • Additional initial staging for individuals with:<br>◦ Multicentric tumors<br>◦ 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 | • MRA Head (CPT® 70544, 70545, or 70546)<br>OR<br>• CTA Head (CPT® 70496) |
| Baseline imaging following resection | • MRI Brain without and with contrast (CPT® 70553) |
| Completion of radiotherapy | • MRI Brain without and with contrast (CPT® 70553) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Treatment response to chemotherapy | • 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 measurable spinal cord disease on MRI | • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 2 cycles |
| Signs or symptoms of recurrence or progression | • MRI Brain without and with contrast (CPT® 70553)<br>◦ Concurrent CT Head without contrast (CPT® 70450) in addition to MRI for craniopharyngioma<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) for signs or symptoms suggesting spinal cord involvement |
| Surveillance | • 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<br>◦ 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)** |
| Suspected spinal cord recurrence | • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |

## Background and Supporting Information

| Includes the following tumors: |
|---|
| • Craniopharyngioma<br>• Papillary craniopharyngioma<br>• Pituicytoma, granular cell tumor of the sellar region, and spindle cell oncocytoma<br>• PitNET<br>• Pituitary blastoma |

**General Considerations:**

- Imaging guidelines and treatment approaches for pediatric pituitary tumors other than those listed above 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
- Craniopharyngiomas are less common, accounting for 6% to 8% of pediatric CNS tumors.
- Most commonly affects children in the pre-adolescent ages
- 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.
- 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**)

**Treatment Considerations:**

- Surgical resection is curative for many individuals
  - Those with a complete resection should then be imaged according to surveillance guidelines after post-resection imaging is completed
- Individuals 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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# Primary CNS Lymphoma (PEDONC-4.11)

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- 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.
- 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 a solitary or multifocal mass occurring in the brain without evidence of systemic (bone marrow or lymph node) involvement.
- 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.

# Meningiomas (PEDONC-4.12)

**ONCP.CT.0004.12.A**

**v1.0.2026**

- 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**

- Account for 1% to 3% of pediatric CNS tumors
- Usually associated with neurofibromatosis type 2 (NF-2) or prior therapeutic radiation exposure to the brain
  - Lifetime risk may be as high as 20% for young children receiving whole brain radiotherapy, most commonly occurring 15 to 20 years after radiation exposure.

# Choroid Plexus Tumors (PEDONC-4.13)

ONCP.CT.0004.13.A
**v1.0.2026**

- PET Metabolic Brain imaging does not have a defined role in the evaluation of choroid plexus tumors and is, therefore, considered not medically necessary.

**Choroid Plexus Papilloma:**

| Indication | Medically Necessary Imaging Study |
|---|---|
| Suspected/Diagnosis | • MRI Brain without and with contrast (CPT® 70553) |
| In addition to the above imaging, for preoperative planning | Either or both of the following:<br>• Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T)<br>• Functional MRI (fMRI) (CPT® 70555 or 70554) |
| Evaluation of 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) |
| Suspected return of hydrocephalus, or return of hydrocephalus seen on CT imaging | • MRI Brain without and with contrast (CPT® 70553) |

**Choroid Plexus Adenoma or Atypical Choroid Plexus Papilloma:**

| Indication | Medically Necessary Imaging Study |
|---|---|
| Suspected/Diagnosis | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| In addition to the above imaging, for preoperative planning | Either or both of the following:<br>• Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T<br>• Functional MRI (fMRI) (CPT® 70555 or 70554) |
| Evaluation of 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) |
| Suspected return of hydrocephalus, or return of hydrocephalus seen on CT imaging | • MRI Brain without and with contrast (CPT® 70553) |

**Choroid Plexus Carcinoma:**

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging of all individuals | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| In addition to the above imaging, for preoperative planning | Either or both of the following:<br>• Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T)<br>• Functional MRI (fMRI) (CPT® 70555 or 70554) |
| Evaluation of 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) |
| Baseline imaging following resection | • MRI Brain without and with contrast (CPT® 70553) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Prior to radiotherapy | • 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 | • MRI Brain without and with contrast (CPT® 70553) |
| Treatment response to chemotherapy | • MRI Brain without and with contrast (CPT® 70553) every 2 cycles during active treatment |
| Additional treatment response to chemotherapy for individuals 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 | • MRI of all known sites of measurable disease |
| End of all planned therapy | • MRI Brain without and with contrast (CPT® 70553)<br>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) |
| Signs or symptoms of recurrence or progression | • MRI Brain without and with contrast (CPT® 70553)<br>• 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, no history of spinal cord involvement | • 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<br>◦ 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)**<br>• 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 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<br>◦ 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)**<br>• 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**

- As a group these account for 1% to 4% of pediatric CNS tumors, and 70% of choroid plexus tumors present within the first 2 years of life
- Includes the following tumors:
  - Choroid plexus papilloma
  - Choroid plexus adenoma, or atypical choroid plexus papilloma
  - Choroid plexus carcinoma
- Choroid plexus papillomas
  - Choroid plexus papillomas outnumber other choroid plexus tumors by 4 to 5 times.

  - These ventricular tumors commonly present with hydrocephalus caused by increased CSF production, resulting in signs of increased intracranial pressure.
  - Appearance on MRI is typical, and they are usually treated by excision.
  - Regrowth is rare
- Choroid Plexus Adenoma or Atypical Choroid Plexus Papilloma
  - These are extremely rare tumors with features midway in the malignant spectrum between papillomas and carcinomas
  - They are more prone to local invasion but rarely to metastasis
  - Presenting symptoms are similar to papillomas
  - Appearance on MRI is typical, and they are usually treated by excision
  - Spinal imaging may be approved if requested at initial diagnosis
  - Regrowth is rare
- Choroid Plexus Carcinoma
  - This is a very aggressive malignancy, with high rates of metastasis to other parts of the CNS
    - Overall incidence of metastases in choroid plexus carcinoma is 12%–50%, which is associated with a worse outcome
  - Prognosis is significantly less favorable than for papillomas with overall survival rates of 35% to 40%
  - *TP53* mutations and alternative lengthening telomeres (ALT) are common in individuals with choroid plexus carcinoma
  - Surgical gross total resection is curative for many individuals
  - Individuals with confirmed gross total resection should then 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 indicated for routine surveillance

# References (PEDONC-4)

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1. Nabors LB, Portnow J, Baehring J, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 1.2025—June 3, 2025. Central Nervous System Cancers, available at: https://www.nccn.org/professionals/physician_gls/pdf/cns.pdf, referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Central Nervous System Cancers V1.2025 June 3, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
2. Gajjar A, Mahajan A, Bale T, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 2.2025—January 17, 2025. Pediatric Central Nervous System Cancers, available at: https://www.nccn.org/professionals/physician_gls/pdf/ped_cns.pdf, referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Central Nervous System Cancers V2.2025 1/17/25. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org
3. Zukotynski K, Fahey F, Kocak M, et al. 18F-FDG PET and MR Imaging associations across a spectrum of pediatric brain tumors: a report from the Pediatric Brain Tumor Consortium. *J Nucl Med*. 2014;55(9):1473-1480. doi:10.2967/jnumed.114.139626.
4. Gajjar A, Bowers DC, Karajannis MA, Leary S, Witt H, Gottardo NG. Pediatric brain tumors: innovative genomic information is transforming the diagnostic and clinical landscape. *J Clin Oncol*. 2015;33(27):2986-2998. doi:10.1200/JCO.2014.59.9217.
5. Brandão LA, Poussaint TY. Pediatric brain tumors. *Neuroimag Clin N Am*. 2013 August;23(3):499-525. Accessed January 3, 2018. http://www.neuroimaging.theclinics.com/article/S1052-5149(13)00017-8/abstract.https://www.ncbi.nlm.nih.gov/pubmed/23928202
6. Brandão LA and Castillo M. Adult brain tumors: clinical applications of magnetic resonance spectroscopy. *Neuroimag Clin N Am*. 2013;23(3):527-555. doi:10.1016/j.nic.2013.03.003
7. Chamdine O, Broniscer A, Wu S, Gajjar A, Qaddoumi I. Metastatic low-grade gliomas in children: 20 years' experience at St. Jude Children's Research Hospital. *Pediatr Blood Cancer*. 2016;63(1):62-70. doi:10.1002/pbc.25731. https://doi.org/10.1007/s11060-009-9922-4
8. Chalil A, Ramaswamy V. Low grade gliomas in children. *J Child Neurol*. 2016;31(4):517-522. doi:10.1177/0883073815599259.
9. Krishnatry R, Zhukova N, Stucklin ASG, et al. Clinical and treatment factors determining long-term outcomes for adult survivors of childhood low-grade glioma: a population-based study. *Cancer*. 2016;122(8):1261-1269. doi:10.1002/cncr.29907.
10. Ullrich NJ. Neurocutaneous Syndromes and brain tumors. *J Child Neurol*. 2016;31(12):1399-1411. doi:10.1177/0883073815604220.
11. Campian J, Gutmann DH. CNS tumors in neurofibromatosis. *J Clin Oncol*. 2017; 35(21):2378-2385. doi:10.1200/JCO.2016.71.7199.
12. Morris EB, Gajjar A, Okuma JO, et al. Survival and late mortality in long-term survivors of pediatric CNS tumors. *J Clin Oncol*. 2007;25(12):1532-1538. doi:10.1200/JCO.2006.09.8194.
13. Karthigeyan M, Gupta K, Salunke P. Pediatric central neurocytoma: a short series with literature review. *J Child Neurol*. 2016;32(1):53-59. Accessed January 3, 2018 doi:10.1177/0883073816668994.
14. Greenfield JP, Heredia AC, George E, Keiran MW, Morales La Madrid A. Gliomatosis cerebri: a consensus summary report from the First International Gliomatosis cerebri Group Meeting, March 26-27, 2015, Paris, France. *Pediatr Blood Cancer*. 2016;63(12):2072-2077. doi:10.1002/pbc.26169.
15. Zaky W, Dhall G, Ji L, et al. Intensive induction chemotherapy followed by myeloablative chemotherapy with autologous hematopoietic progenitor cell rescue for young children newly-diagnosed with central nervous system atypical teratoid/rhabdoid tumors: the head start III experience. *Pediatr Blood Cancer*. 2014;61(1):95-101. doi:10.1002/pbc.24648.

16. Millard NE and DeBraganca KC. Medulloblastoma. *J Child Neurol*. 2016;31(12):1341-1353. doi:10.1177/0883073815600866.
17. Koschmann C, Bloom K, Upadhyaya S, Geyer JR. Survival after relapse of medulloblastoma. *J Pediatr Hematol. Oncol*. 2016;38(4):269-273. doi:10.1097/MPH.0000000000000547.
18. Martinex S, Khakoo Y, Giheeney S, et al. Marker (+) CNS germ cell tumors in remission: are surveillance MRI scans necessary? *Pediatr Blood Cancer*. 2014;61(5):853-854. Accessed January 3, 2018. doi:10.1002/pbc.24888.
19. Abu Arja MH, Bouffet E, Finlay JL, AbdelBaki MS. Critical review of the management of primary central nervous system germ cell tumors. *Pediatr Blood Cancer*. 2019;66:e27658. doi:10.1002/pbc.27658.
20. Norris GA, Garcia J, Hankinson TC, et al. Diagnostic accuracy of neuroimaging in pediatric optic chiasm/suprasellar tumors. *Pediatr Blood Cancer*. 2019;66:e27860. doi:10.1002/pbc.27680.
21. Grommes C, DeAngelis LM. Primary CNS lymphoma. *J Clin Oncol*. 2017;35(21):2410-2418. doi:10.1200/JCO.2017.72.7602.
22. Li Z, Li H, Wang S, et al. Pediatric skull base meningiomas: clinical features and surgical outcomes. *J Child Neurol*. 2016;31(14):1523-1527. doi:10.1177/0883073816664669.
23. Horská A, Barker PB. Imaging of brain tumors: MR Spectroscopy and metabolic imaging. *Neuroimaging Clin N Am*. 2010;20(3):293-310. doi:10.1016/j.nic.2010.04.003.
24. Zaky W, Dhall G, Khatua S, et al. Choroid plexus carcinoma in children: the head start experience. *Pediatr Blood Cancer*. 2015;62(5):784-789. doi:10.1002/pbc.25436.
25. Zaky W, Finlay JL, Pediatric choroid plexus carcinoma: Biologically and clinically in need of new perspectives, *Pediatr Blood Cancer*. 2018;65:e27031. doi:10.1002/pbc.27031.
26. Merchant TE, Bendel AE, Sabin ND, et.al. Conformal radiation therapy for pediatric ependymoma, chemotherapy for incompletely resected ependymoma, and observation for completely resected, supratentorial ependymoma. *J Clin Oncol*. 2019;37(12):974-983. doi:10.1200/JCO.18.01765.
27. PDQ® Pediatric Treatment Editorial Board. PDQ Childhood Medulloblastoma and Other Central Nervous System Embryonal Tumors Treatment. Bethesda, MD: National Cancer Institute. Updated 4/12/2023. Available at: https://www.cancer.gov/types/brain/hp/child-cns-embryonal-treatment-pdq. [PMID: 26389418].
28. Burns J, Policeni B, Bykowski J, et al. ACR Appropriateness Criteria® Neuroendocrine Imaging. Available at https://acsearch.acr.org/docs/69485/Narrative/. American College of Radiology. Accessed 7/21/2020.
29. Klawinski D, Indelicato DJ, Hossain J, Sandler E. Surveillance imaging in pediatric ependymoma. *Pediatr Blood Cancer.* 2020;67(11):e28622. doi:10.1002/pbc.28622.
30. Massimino M, Barretta F, Modena P, et al. Pediatric intracranial ependymoma: correlating signs and symptoms at recurrence with outcome in the second prospective AIEOP protocol follow-up. *J Neurooncol.* 2018;140(2):457-465. doi:10.1007/s11060-018-2974-6.
31. Jaju A, Li Y, Dahmoush H, et. al. Imaging of pediatric brain tumors: A COG Diagnostic Imaging Committee/SPR Oncology Committee/ASPNR white paper. *Pediatr Blood Cancer*. 2023;70 Suppl 4:e30147. doi:10.1002/pbc.30147.
32. Louis DN, Perry A, Wesseling P. et al. The 2021 WHO classification of tumors of the central nervous system: a summary. *Neuro-Oncology*. 2021;23(8):1231–1251.doi:10.1093/neuonc/noab106.
33. Rootman MS, Dotan G, Konen O. Neuroimaging in children with ophthalmological complaints: a review. *J Neuroimaging*. 2021;31(3):446-458. doi:10.1111/jon.12842.

# Pediatric Lymphomas (PEDONC-5)

Guideline Page







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# Pediatric Lymphoma – General Considerations (PEDONC-5.1)

**ONCP.HL.0005.1.A**

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- Lymphoma mostly 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.
  - Noncontrast CT imaging has not been shown to be beneficial in the management of pediatric lymphomas
  - Given the limited utility of noncontrast CT imaging in pediatric lymphomas, MRI without **or** without and with contrast is recommended in place of CT for individuals who cannot tolerate CT contrast due to allergy or impaired renal function.
- MRI without and with contrast of symptomatic or previously involved bony areas is 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 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 medically necessary when MRI is contraindicated.

# Pediatric Hodgkin Lymphoma (HL) (PEDONC-5.2)

**ONCP.HL.0005.2.C**
**v1.0.2026**

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging | ANY or ALL of the following:<br>• CT Neck with contrast (CPT® 70491)<br>◦ MRI Neck without and with contrast (CPT® 70543) as a substitute for CT to limit radiation exposure if requested<br>• CT Chest with contrast (CPT® 71260)<br>◦ MRI is not a substitution for CT Chest<br>• CT Abdomen and Pelvis with contrast (CPT® 74177)<br>◦ 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<br>• MRI Brain with and without contrast (CPT® 70553) for known CNS involvement or new signs or symptoms suggesting intracranial disease<br>• PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498)<br>◦ Can be performed prior to biopsy if necessary for individual scheduling<br>• 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 | • ALL of the following, as often as every 2 cycles of chemotherapy:<br>◦ CT of previously involved visceral areas<br>▪ In individuals on treatment for recurrent or refractory Hodgkin Lymphoma, all of the following:<br>- CT Neck with contrast (CPT® 70491)<br>- CT Chest with contrast (CPT® 71260)<br>- CT Abdomen and Pelvis with contrast (CPT® 74177)<br>◦ MRI Brain without and with contrast (CPT® 70553) for known CNS involvement<br>AND<br>• After cycles 2 and 4* of chemotherapy and at end of therapy:<br>◦ PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (only using combination of CPT® 78813 and 76498)<br>▪ *For individuals with low-risk stage IA or IIA mixed cellularity Hodgkin lymphoma, this restaging PET/CT is medically necessary after cycles 1 and 3 (instead of cycles 2 and 4) if requested. End of therapy PET/CT or PET/MRI is medically necessary for all individuals with Hodgkin lymphoma.<br>◦ Diagnostic CT or MRI of previously involved areas may be combined with PET at end of therapy.<br>• In addition to the above studies:<br>◦ 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 CPT® 78816) or PET/MRI is medically necessary >12 weeks after radiation therapy to confirm complete response. |
| Surveillance for individuals with no history of recurrent HL | • Imaging studies are only medically necessary when relapse is clinically suspected, because most individuals will clinically declare themselves and there is no survival advantage in pre-emptive imaging.<br>• **Routine surveillance imaging is not medically necessary in individuals without a prior history of recurrent disease** |

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

**Background and Supporting Information**

- Most individuals experiencing recurrence are detected based on physical findings, and frequent CT surveillance imaging of Hodgkin lymphoma after completion of therapy does not improve post-recurrence overall survival. The primary determinant of survival at recurrence is time to relapse, regardless of whether relapse is detected

clinically or via imaging. NCCN pediatric HL guidelines recommend no pre-emptive surveillance imaging.

- Pediatric individuals have a high rate of neck involvement with Hodgkin lymphoma
- Early treatment response evaluations involve both PET and CT as decisions about chemotherapy drug selection and radiation treatment are frequently made based on both anatomic (CT-based) and metabolic (PET/CT-based) responses.
- Indications and coding for rare circumstances where PET/MRI may be approved are found in **PET imaging in Pediatric Oncology (PEDONC-1.4)**

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

**ONCP.HL.0005.3.A**
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| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging | ANY or ALL of the following:<br>• CT Neck with contrast (CPT® 70491)<br>◦ MRI Neck without and with contrast (CPT® 70543) in place of CT Neck, if requested<br>• CT Chest with contrast (CPT® 71260)<br>• CT Abdomen and Pelvis with contrast (CPT® 74177)<br>◦ 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.<br>◦ Abdominal ultrasound (CPT® 76700 or 76705) at initial presentation if CT/MRI not available.<br>• CT with contrast or MRI without and with contrast any other symptomatic body area<br>• 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 if symptoms or extent of disease suggest intracranial extension or metastasis | • MRI Brain without and with contrast (CPT® 70553) |
| Additional initial staging if symptoms or extent of disease suggest intraspinal extension or metastasis (back pain) | • MRI of suspected spinal level involvement without and with contrast<br>• MRI of whole spine without and with contrast if there is suspected leptomeningeal disease or if sedation will be required |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Restaging for treatment response (following initial response evaluation) | • ANY OR ALL of the following, as often as every cycle of chemotherapy (~every 3 weeks):<br>◦ CT with contrast or MRI without and with contrast (should be same modality as initial diagnosis if possible) of previously involved areas<br>◦ PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) until a negative PET is obtained<br>▪ Whole-body PET/CT (CPT® 78816) may be approved if there is clinical suspicion of, or known, skull or distal lower extremity involvement.<br>▪ 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) | • 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 |
| End of therapy PET with Deauville 4-5 avidity | ONCE, at least 6-8 weeks after end of therapy PET:<br>• PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498)<br>OR<br>• 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 | • PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Individuals 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)<br>◦ US, x-ray, or other red flags are not required prior to CT |
| Surveillance of asymptomatic individuals 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. |
| Clinical symptoms or laboratory findings suggesting recurrence | • MRI Brain without and with (CPT® 70553) for history of CNS involvement or new signs or symptoms suggesting intracranial disease<br>And ANY OR ALL of the following:<br>• CT Neck with contrast (CPT® 70491) **and**<br>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**<br>• 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 | • PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) |

**Background and Supporting Information**

- Aggressive mature B-Cell NHL includes all of the following diagnoses, all of which should be imaged according to this section:
  - Burkitt's lymphoma/leukemia (BL)
  - Diffuse Large B-Cell Lymphoma (DLBCL)
  - Primary Mediastinal Large B-Cell Lymphoma (PMBCL)

  - Post-transplant Lymphoproliferative Disorder (PTLD)
    - Most commonly occurs following solid organ or stem cell transplantation
  - Viral-associated lymphoproliferative disorders
    - Most commonly occurs following hematopoietic stem cell transplantation or in individuals with primary immunodeficiency
- Initial treatment is usually 7 days of low intensity therapy, with early response evaluation determining next steps in therapy using CT with contrast or MRI without and with contrast of previously involved areas performed around day 6
  - Individuals are customarily still inpatient for this evaluation so outpatient requests should be rare for this time point
- Routine asymptomatic surveillance with advanced imaging has not been found to impact individual outcomes as the majority of these individuals present clinically at relapse due to the highly aggressive nature of these lymphomas
- Indications and coding for rare circumstances where PET/MRI may be approved are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

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

**ONCP.HL.0005.4.C**
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| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging | • ANY or ALL of the following:<br>◦ CT Neck with contrast (CPT® 70491), CT Chest with contrast (CPT® 71260), and CT Abdomen and Pelvis with contrast (CPT® 74177)<br>◦ MRI without and with contrast of affected area<br>▪ Substituted for CT in cases of paraspinal or soft tissue extremity primary tumors<br>◦ CT with contrast or MRI without and with contrast any other symptomatic body area<br>◦ PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and 76498) |
| Additional initial staging for individuals 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) | • 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 treated with cytotoxic chemotherapy | • Every 2 cycles:<br>◦ CT of previously involved areas<br>▪ If CT is performed for primary treatment response, PET/CT (CPT® 78815 or CPT® 78816) to clarify inconclusive findings detected on conventional imaging<br>OR<br>◦ PET/CT or PET/MRI (CPT® 78813 and CPT® 76498) until a negative PET is obtained<br>▪ 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) | • CT with contrast of previously involved areas |
| Surveillance | • 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 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 | • 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 |
| Clarify inconclusive findings on conventional imaging to evaluate the need for biopsy to establish recurrence | • PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) |

**Background and Supporting Information**

- ALCL is similar in presentation to Hodgkin Lymphoma, and may be indistinguishable until immunocytology and molecular studies are complete
- Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

# References (PEDONC-5)

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1. Barth M, Xavier AC, Acquazzino M, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 2.2025 — April 28, 2025, Pediatric Aggressive Mature B-Cell Lymphomas, available at: https://www.nccn.org/professionals/physician_gls/pdf/ped_b-cell.pdf. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Pediatric Aggressive Mature B-Cell Lymphomas V2.2025 April 28, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
2. Metzger ML, Krasin MJ, Choi JK, et al. Hodgkin lymphoma. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:568-586.
3. Allen CE, Kamdar KY, Bollard CM, et al. Malignant Non-Hodgkin lymphomas in Children. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:587-603.
4. Hayashi RJ, Wistinghausen B, Shiramizu B. Lymphoproliferative Disorders and Malignancies Related To Immunodeficiencies. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:604-616.
5. Alexander S, Ferrando AA. Pediatric lymphoma. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th ed. Philadelphia, PA: Elsevier Saunders; 2015:1626-1671.
6. Lee Chong A, Grant RM, Ahmed BA, Thomas KE, Connolly BL, Greenberg M. Imaging in pediatric patients: time to think again about surveillance. *Pediatr Blood Cancer*. 2010;55(3):407-413. doi:10.1002/pbc.22575.
7. Ceppi F, Pope E, Ngan B, Abla O. Primary cutaneous lymphomas in children and adolescents. *Pediatr Blood Cancer*. 2016;63(11):1886-1894. doi:10.1002/pbc.26076.
8. Flerlage JE, Kelly KM, Beishuizen A, et al. Staging evaluation and response criteria harmonization (SEARCH) for childhood, adolescent, and young adult Hodgkin Lymphoma (CAYAHL): Methodology statement. *Pediatr Blood Cancer*. 2017;64(7):e26421. doi:10.1002/pbc.26421.
9. Friedman DL, Chen L, Wolden S, et al. Dose-intensive response-based chemotherapy and radiation therapy for children and adolescents with newly diagnosed intermediate-risk Hodgkin lymphoma: a report from the Children's Oncology Group Study AHOD0031. *J Clin Oncol*. 2014;32(32):3651-3658. doi:10.1200/JCO.2013.52.5410.
10. Schwartz CL, Chen L, McCarten K, et al. Childhood Hodgkin International Prognostic Score (CHIPS) predicts event-free survival in Hodgkin lymphoma: a report from the Children's Oncology Group. *Pediatr Blood Cancer*. 2017;64(4):e26278. doi:10.1002/pbc.26278.
11. Friedmann AM, Wolfson JA, Hudson MM, et al. Relapse after treatment of Hodgkin lymphoma: outcome and role of surveillance after therapy. *Pediatr Blood Cancer*. 2013;60(9):1458-1463. doi:10.1002/pbc.24568.
12. Mauz-Körholz C, Metzger ML, Kelly KM, et al. Pediatric Hodgkin lymphoma. *J Clin Oncol*. 2015;33(27):2975-2985. doi:10.1200/JCO.2014.59.4853.
13. Voss SD, Chen L, Constine LS, et al. Surveillance computed tomography imaging and detection of relapse in intermediate- and advanced-stage pediatric Hodgkin's lymphoma: A report from the Children's Oncology Group. *J Clin Oncol*. 2012;30(21):2635-2640. doi:10.1200/JCO.2011.40.7841.
14. Voss SD. Surveillance Imaging in Pediatric Hodgkin Lymphoma. *Curr Hematol Malig Rep*. 2013;8(3):218-225. doi:10.1007/s11899-013-0168-z.
15. Appel BE, Chen L, Buxton AB, et al. Minimal treatment of low-risk, pediatric lymphocyte-predominant Hodgkin lymphoma: a report from the Children's Oncology Group. *J Clin Oncol*. 2016;34(20):2372-2379. doi:10.1200/JCO.2015.65.3469.
16. Rosolen A, Perkins SL, Pinkerton CR, et al. Revised international pediatric non-Hodgkin lymphoma staging system. *J Clin Oncol*. 2015;33:2112-2118. doi:10.1200/JCO.2014.59.7203.
17. Sandlund JT, Guillerman RP, Perkins SL, et al. International pediatric Non-Hodgkin lymphoma response criteria. *J Clin Oncol*. 2015;33(18):2106-2111. doi:10.1200/JCO.2014.59.0745.

18. Eissa HM, Allen CE, Kamdar K, et al. Pediatric Burkitt's lymphoma and Diffuse B-cell lymphoma: are surveillance scans required? *Pediatr Hematol Oncol*. 2014;31(3):253-257. doi:10.3109/08880018.2013.834400.
19. Minard-Colin V, Brugieres L, Reiter A, et al. Non-Hodgkin lymphoma in children and adolescents: progress through effective collaboration, current knowledge, and challenges ahead. *J Clin Oncol*. 2015;33(27):2963-2974. doi:10.1200/JCO.2014.59.5827.
20. Vali R, Punnett A, Bajno L, Moineddin R, Shammas A. The value of 18F-FDG PET in pediatric patients with post-transplant lymphoproliferative disorder at initial diagnosis. *Pediatr Transplant*. 2015;19(8):932-939. doi:10.1111/petr.12611.
21. Hochberg J, Flower A, Brugieres L, Cairo MS. NHL in adolescents and young adults: a unique population. *Pediatr Blood Cancer*. 2018;65:e27073. doi.org/10.1002/pbc.27073.
22. Buhtoiarov IN. Pediatric lymphoma. *Pediatr In Rev*. 2017;38(9):410-423. doi:10.1542/pir.2016-0152.
23. Kelly K, Hiniker SM, Armenian S, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 2.2025, June 9, 2025. Pediatric Hodgkin Lymphoma, available at: https://www.nccn.org/professionals/physician_gls/pdf/ped_hodgkin.pdf, Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Pediatric Hodgkin Lymphoma V2.2025, June 9, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
24. Mhlanga J, Alazraki A, Cho SY, et. al. Imaging recommendations in pediatric lymphoma: A COG Diagnostic Imaging Committee/SPR Oncology Committee white paper. *Pediatr Blood Cancer*. 2022:e29968. doi:10.1002/pbc.2996.

# Neuroblastoma (PEDONC-6)

Guideline Page







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# Neuroblastoma – General Considerations (PEDONC-6.1)

**ONCP.NP.0006.1.C**
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- **Neuroblastoma (PEDONC-6)** should be used to review neuroblastoma, ganglioneuroblastoma, and ganglioglioma 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 can be approved:<br>- CPT® 78300<br>- CPT® 78305<br>- CPT® 78306<br>- CPT® 78803, 78830, or 78832<br>• May be approved alone or in combination with:<br>◦ CPT® 78305<br>◦ CPT® 78306 |

| Study Type | Coding |
|---|---|
| $^{123}$I-metaiodobenzylguanidine (MIBG) scintigraphy | ▪ Any one of the following codes can be approved:<br>- CPT® 78801<br>- CPT® 78802<br>- CPT® 78804<br>▪ Any one of the following codes may also be approved, individual or in combination with CPT® 78801, 78802, or 78804<br>- CPT® 78803<br>- CPT® 78830<br>- CPT® 78831<br>- CPT® 78832<br>- 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 |

- MIBG remains the standard of care metabolic imaging in neuroblastoma, 18F-FDG PET/CT is not 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 medically necessary at this time:

| Indication | Medically Necessary Imaging Study |
|---|---|
| - ANY of the following:<br>• Individuals with MIBG-negativity documented at initial diagnosis<br>• Individuals with discordant findings on MIBG and conventional imaging (i.e., it is suspected there is more active disease than is visible on MIBG)<br>• 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)<br>- In scenarios where PET/CT is supported, it does not preclude the other diagnostic imaging studies supported throughout **PEDONC 6**. PET/CT is viewed as replacing MIBG in these scenarios.<br>- Indications and coding for rare circumstances where PET/MRI is 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:<br>• Tricyclic antidepressants (amitriptyline, imipramine, etc.)<br>• Selective serotonin reuptake inhibitors (SSRI's, sertraline, paroxetine, escitolapram, etc.)<br>• Neuroleptics (risperidone, haloperidol, etc.)<br>• Antihypertensive drugs (alpha or beta blockers, calcium channel blockers)<br>• Decongestants (phenylephrine, ephedrine, pseudoephedrine)<br>• Stimulants (methylphenidate, dextroamphetamine, etc. | - 18F-FDG PET/CT (CPT® 78816) is only medically necessary for this indication when specific documentation of the medication interaction is included with the current PET imaging request.<br>- Indications and coding for rare circumstances where PET/MRI is 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 generally arises from the adrenal gland or along the sympathetic chain. Neuroblastoma staging has recently changed to better incorporate the prognostic impact of biological and genetic characteristics, particularly segmental chromosome aberrations (SCA) as an additional genomic marker. SCAs of 1p or 11g are poor prognostic indicators.
- 90% to 95% of neuroblastomas secrete homovanillic acid (HVA) and vannilylmandelic acid (VMA) in the urine, and urine HVA/VMA should be performed at every disease evaluation for individuals with positive HVA or VMA at diagnosis
- 99mTc-MDP bone scan does not identify foci of disease that affect staging or clinical management and provides no advantage over MIBG scintigraphy and is not used for evaluation of most individuals with neuroblastoma

# Staging and Risk Grouping – Neuroblastoma (PEDONC-6.2)

**ONCP.NP.0006.2.A**

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- 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.

# Neuroblastoma – Initial Staging (PEDONC-6.3)

**ONCP.NP.0006.3.A**
**v1.0.2026**

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

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging for all individuals | • $^{123}$I-metaiodobenzylguanidine (see table in **Neuroblastoma – General Considerations (PEDONC-6.1)** for MIBG coding details) scintigraphy<br>AND<br>• ONE of the following sets of imaging:<br>◦ CT Neck with contrast (CPT® 70491), CT Chest with contrast (CPT® 71260), and CT Abdomen and Pelvis with contrast (CPT® 74177)<br>OR<br>• 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 paraspinal tumors<br>• Individuals with back pain or cord compression symptoms | In addition to the above imaging:<br>• MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), and Lumbar (CPT® 72158) spine without and with contrast |
| Evaluation of suspected adrenal neuroblastoma, ganglioneuroblastoma, or ganglioneuroma when CT or MRI is inconclusive for an adrenal lesion. | • Adrenal nuclear imaging (CPT® 78075) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Clinical signs/symptoms suggest brain and/or orbital involvement | Any or all of the following:<br>• MRI Brain without and with contrast (CPT® 70553)<br>• CT Head with contrast (CPT® 70470)<br>• CT Orbits/Maxillofacial with contrast (CPT® 70487) |

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

**Background and Supporting Information**

- MIBG provides superior sensitivity and sensitivity for detecting viable osseous disease compared with bone scintigraphy so technetium bone scan is not necessary when MIBG is utilized. MIBG is positive in 90% to 95% of neuroblastomas.
  - Most MIBG imaging studies are SPECT/CT studies using CT for localization only. Separate diagnostic CT codes should not be approved for this purpose
  - Occasionally MIBG cannot be performed prior to initiation of therapy. In this circumstance, MIBG should be completed within 3 weeks of therapy initiation as the reduction in MIBG avidity in response to chemotherapy is not immediate. Inability to complete MIBG before starting therapy is not an indication to approve PET imaging

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

**ONCP.NP.0006.4.A**

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- 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, 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 medically necessary.<br>◦ Ultrasound is medically necessary if requested in place of CT or MRI to avoid radiation and/or anesthesia exposure in low risk individuals |

### Background and Supporting Information

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

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

| Indication | Medically Necessary Imaging Study |
|---|---|
| Prior to surgical resection | • Restaging imaging (MIBG and CT or MRI, as performed at initial diagnosis)<br>• PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met.<br>• Additional imaging for individuals with paraspinal disease:<br>◦ 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) | • CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177)<br>OR<br>• 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)<br>AND<br>• CT or MRI of other sites with prior measurable disease<br>• Additional imaging for individuals with paraspinal disease:<br>◦ 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 | • MIBG scan (see table in **Neuroblastoma - General Considerations (PEDONC-6.1)** for MIBG coding details)<br>• 18F-FDG PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met |

### Background and Supporting Information

- Individuals generally receive 2 to 12 cycles of moderate-intensity chemotherapy depending on response to treatment.

- Surgical resection may occur prior to or following chemotherapy depending on disease stage. Restaging prior to surgery is appropriate.

**High-Risk Neuroblastoma:**

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

**Background and Supporting Information**

- This group of individuals receives highly aggressive therapy using sequential chemotherapy, surgery, high dose chemotherapy with stem cell rescue, radiotherapy, monoclonal antibody (mAb) immunotherapy, and biologic therapy.

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

**ONCP.NP.0006.5.C**
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**Very Low Risk and Low Risk Neuroblastoma:**

| Indication | Medically Necessary Imaging Study |
|---|---|
| All individuals | • At 3, 6, 9, 12, 18, 24, 30, and 36 months after surgery:<br>◦ Ultrasound of involved areas<br>OR<br>• 3, 6, 9, 12, 18, 24, 30, and 36 months after surgery or to clarify unclear findings on ultrasound:<br>◦ 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 | • MIBG (see table in **Neuroblastoma - General Considerations (PEDONC-6.1)** for MIBG coding details)<br>• 18F-FDG PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met. |

**Intermediate Risk Neuroblastoma:**

| Indication | Medically Necessary Imaging Study |
|---|---|
| All individuals | • Every 3 months for 1 year, then every 6 months for 1 year, and then at 36 months after surgery:<br>◦ CT with contrast or MRI without and with contrast of the primary tumor<br>▪ If the primary tumor is paraspinal:<br>- MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158)<br>◦ If primary site is abdomen or pelvis, both body sites are medically necessary (CPT® 74177 or the combination of CPT® 71297 and CPT® 74183)<br>◦ If negative at 36 months, ultrasound at 48 and 60 months after surgery to complete 5 years surveillance. |
| • BOTH of the following:<br>◦ Individuals with stage 4, or M disease, or 4S, or MS disease AND<br>◦ 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:<br>◦ MIBG scan (see table in **Neuroblastoma – General Considerations (PEDONC-6.1)** for MIBG coding details)<br>◦ Once a negative MIBG is achieved, no further MIBG imaging is necessary.<br>• MIBG is not medically necessary for all other intermediate risk individuals.<br>• 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 | • Every 3 months for 1 year, then every 6 months for 1 year, and then annually to complete 6 years surveillance:<br>◦ CT with contrast or MRI without and with contrast of the primary tumor site AND<br>◦ MIBG scan (see table in **Neuroblastoma – General Considerations (PEDONC-6.1)** for MIBG coding details)<br>▪ 18F-FDG PET/CT or PET/MRI is indicated if exceptions noted in **PEDONC-6.1** are met<br>◦ If primary site is abdomen or pelvis, both body sites are medically necessary (CPT® 74177 or the combination of CPT® 71297 and CPT® 74183)<br>◦ For history of paraspinal disease, MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158)<br>◦ If negative at 6 years, no further advanced imaging. |
| Suspected recurrence | • 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<br>• MRI Brain with and without contrast (CPT® 70553) for signs or symptoms of brain involvement<br>• 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.<br>• MIBG scan (see table in **Neuroblastoma - General Considerations (PEDONC-6.1)** for MIBG coding details)<br>• 18F-FDG PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met. |

**Background and Supporting Information**

- Very Low Risk and Low Risk Neuroblastoma:
  - CT Chest is not indicated in asymptomatic surveillance imaging of neuroblastoma individuals with no prior history of thoracic disease

- High-Risk Neuroblastoma:
  - Early detection of recurrence with $^{123}$I-MIBG has been shown to improve post-relapse outcomes in high-risk neuroblastoma





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# References (PEDONC-6)

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1. Brodeur GM, Hogarty MD, Bagatell R, et al. Neuroblastoma. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:772-797.
2. Shusterman S and George RE. Neuroblastoma. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th ed. Philadelphia, PA: Elsevier Saunders; 2015:1675-1713.
3. Sharp SE, Gelfand MJ, Shulkin BL. Pediatrics: diagnosis of neuroblastoma. *Semin Nucl Med*. 2011;41(5):345-353. doi:10.1053/j.semnuclmed.2011.05.001.
4. Kushner BH, Kramer K, Modak S, et al. Sensitivity of surveillance studies for detecting asymptomatic and unsuspected relapse of high-risk neuroblastoma. *J Clin Oncol*. 2009;27(7):1041-1046. doi:10.1200/JCO.2008.17.6107.
5. Nuchtern JG, London WB, Barnewolt CE, et al. A prospective study of expectant observation as primary therapy for neuroblastoma in young infants: A Children's Oncology Group Study. *Ann Surg*. 2012;256(4):573-580. doi:10.1097/SLA.0b013e31826cbbbd.
6. GE Healthcare. AdreView™ Iobenguane I 123 Injection prescribing information. Revised September 2008.
7. Bombardieri E, Giammarile F, Aktolun C, et al. 131I/123I-Metaiodobenzylguanidine (MIBG) scintigraphy: procedure guidelines for tumor imaging. *Eur J Nucl Med Mol Imaging*. 2010;37(12):2436-2446.
8. Mueller WP, Coppenrath E, Pfluger T. Nuclear medicine and multimodality imaging of pediatric neuroblastoma. *Pediatr Radiol*. 2013;43(4):418-427. doi:10.1007/s00247-012-2512-1.
9. Park JR, Bagatell R, Cohn SL, et al. Revisions to the International Neuroblastoma Response Criteria: A consensus statement from the National Cancer Institute clinical trials planning meeting. *J Clin Oncol*. 2017;35(22):2580-2587. doi:10.1200/JCO.2016.72.0177.
10. Gauguet J-M, Pace-Emerson T, Grant FD, et al. Evaluation of the utility of 99mTC-MDP bone scintigraphy versus MIBG scintigraphy and cross-sectional imaging for staging patients with neuroblastoma. *Pediatr Blood Cancer*. 2017;64:e26601. doi:10.1002/pbc.26601.
11. Venkatramani R, Pan H, Furman WL, et al. Multimodality treatment of pediatric esthesioneuroblastoma. *Pediatr Blood Cancer*. 2016;63(3):465-470. doi:10.1002/pbc.25817.
12. Owens C, Li BK, Thomas KE, et al. Surveillance imaging and radiation exposure, in the detection of relapsed neuroblastoma. *Pediatr Blood Cancer*. 2016:63(10);1786-1793. doi:10.1002/pbc.26099.
13. Allen-Rhoades W, Whittle SB, Rainusso N. Pediatric solid tumors of infancy: an overview. *Pediatr In Rev*. 2018;39(2):57-67. doi:10.1542/pir.2017-0057.
14. Park JR, Kreissman SG, London WB. Effect of tandem autologous stem cell transplant vs single transplant on event-free survival in patients with high-risk neuroblastoma. *JAMA*. doi:10.1001/jama.2019.11642.
15. Irwin MS, Naranjo A, Zhang FF, et al. Revised neuroblastoma risk classification system: a report from the Children's Oncology Group. *J Clin Oncol*. 2021;39(29):3229-3241. doi:10.1200/JCO.21.00278.
16. Maaz AUR, O'Doherty J, Djekidel M.68Ga-DOTATATE PET/CT for neuroblastoma staging: utility for clinical use. *J Nucl Med Technol*. 2021;49(3):265-268. doi:10.2967/jnmt.120.258939.
17. Bar-Sever Z, Biassoni L, Shulkin B, et. al. Guidelines on nuclear medicine imaging in neuroblastoma. *Eur J Nucl Med Mol Imaging.* 2018;45(11):2009-2024. doi:10.1007/s00259-018-4070-8.
18. Bleeker G, Tytgat GAM, Adam JA, et. al. 123I-MIBG and 18F-FDGPET imaging for diagnosing neuroblastoma. *Cochrane Database of Systematic Reviews*. 2015;9. doi:10.1002/14651858.CD009263.pub2.
19. Lai HA, Sharp SE, Bhatia A, et. al. Imaging of pediatric neuroblastoma: A COG Diagnostic Imaging Committee/SPR Oncology Committee white paper. *Pediatr Blood Cancer*. 2022;e29974. doi:10.1002/pbc.29974.
20. Vo KT, DuBois SG, Neuhaus J, et. al. Pattern and predictors of sites of relapse in neuroblastoma: A report from the International Neuroblastoma Risk Group (INRG) project. *Pediatr Blood Cancer*. 2022;69(9):e29616. doi:10.1002/pbc.29616.
21. Bagatell R, Park JR, Archarya S, et. al. National Comprehensive Cancer Network (NCCN) Guidelines Version 1.2025 – April 16, 2025. Neuroblastoma, available at: https://www.nccn.org/professionals/physician_gls/pdf/neuroblastoma.pdf, Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology

# Pediatric Renal Tumors (PEDONC-7)

Guideline Page







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# Pediatric Renal Tumors – General Considerations (PEDONC-7.1)

**ONCP.RC.0007.1.C**
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- 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 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.

# Unilateral Wilms Tumor (UWT) (PEDONC-7.2)

**ONCP.RC.0007.2.C**
**v1.0.2026**

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial Staging | ONE of the following:<br>• MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)<br>OR<br>• CT Abdomen with contrast (CPT® 74160) or CT Abdomen and Pelvis with contrast (CPT® 74177)<br>◦ Doppler ultrasound to evaluate for tumor thrombus is not necessary unless CT findings are inconclusive<br>AND<br>• CT Chest with (CPT® 71260) or without contrast (CPT® 71250)<br>◦ Should be completed prior to anesthesia exposure if possible<br>• PET is not medically necessary in the initial staging of any pediatric renal tumor |
| Bilateral renal lesions noted on ultrasound or CT | • MRI Abdomen (CPT® 74183) and Pelvis (CPT® 72197) without and with contrast |
| Additional initial staging imaging for any individual with neurologic signs or symptoms raising suspicion of CNS metastases | • MRI Brain without and with contrast (CPT® 70553) |
| Additional initial staging imaging for any individual 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 ~every 2 cycles during treatment and at the end of planned therapy | • CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250)<br>• 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 | • PET/CT (CPT® 78815) |
| Surveillance | • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250), or chest x-ray AND CT Abdomen with contrast (CPT® 74160) or Abdominal US (CPT® 76700)<br>◦ Every 3 months for 2 years, then every 6 months for 2 additional years, to complete 4 years surveillance<br>• 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. | |

**Background and Supporting Information**

- Many individuals will present with an asymptomatic abdominal mass, and will undergo ultrasound as a primary evaluation.
- Only ~0.5% of individuals with Wilms tumor will ever develop brain metastases
- The majority of individuals will receive chemotherapy with or without XRT, beginning within 14 days of initial surgery

# Bilateral Wilms Tumor (BWT) (PEDONC-7.3)

**ONCP.RC.0007.3.C**
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| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial Staging | ONE of the following:<br>• MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)<br>OR<br>• CT Abdomen and Pelvis with contrast (CPT® 74177)<br>◦ 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<br>▪ Doppler ultrasound to evaluate for tumor thrombus is not medically necessary unless CT findings are inconclusive<br>AND<br>• CT Chest with (CPT® 71260) or without contrast (CPT® 71250)<br>◦ Should be completed prior to anesthesia exposure if possible<br>• PET is not routinely medically necessary in the initial staging of any pediatric renal tumor |
| Additional initial staging imaging for any individual 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 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 | • MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)<br>◦ CT Abdomen and Pelvis with contrast (CPT® 74177) for individuals with a contraindication to MRI<br>• 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 | • PET/CT (CPT® 78815) |
| Surveillance | • Every 3 months for 2 years, then every 6 months for 2 additional years, to complete 4 years surveillance:<br>◦ CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) or chest x-ray AND<br>◦ CT Abdomen with contrast (CPT® 74160) or Abdominal US (CPT® 76700)<br>• 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 medically necessary to assess treatment response or for surveillance in Wilms tumor. | |

**Background and Supporting Information**

- Many individuals will present with an asymptomatic abdominal mass, and will undergo ultrasound as a primary evaluation.
- Individuals with bilateral Wilms Tumor may begin therapy without a histologic diagnosis to preserve a localized disease stage and attempt to shrink the tumors to allow for renal-sparing surgical approaches.
- nly ~0.5% of Wilms Tumor individuals will ever develop brain metastases





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

**ONCP.RC.0007.4.C**
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| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial Staging | • CT Abdomen and Pelvis with contrast (CPT® 74177)<br>◦ If bilateral renal lesions are noted on ultrasound or CT, MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) should be strongly considered<br>• CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250)<br>◦ Should be completed prior to anesthesia exposure if possible<br>• PET scan is not routinely medically necessary in the initial staging of any pediatric renal tumor |
| Additional initial staging for any individual 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 signs or symptoms raising suspicion of bony metastases | • Bone scan (See: **PEDONC-1.3** for coding) |
| Treatment response in individuals with residual measurable disease after initial surgery and receiving adjuvant medical therapy | • Every 2 cycles during active treatment:<br>◦ CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) and<br>◦ CT Abdomen with contrast (CPT® 74160)<br>▪ Pelvic imaging is not medically necessary unless prior pelvic involvement has been documented |

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

**Background and Supporting Information**

- A majority of pediatric cases have a novel subtype involving TFE3 or TFEB translocations, which have a different natural history than “adult type” RCC
- Individuals of any age with TFE3 or TFEB translocated RCC should be imaged according to this guideline section.
- 40% to 45% of pediatric RCC cases have similar histologies to adult RCC (clear cell, papillary, chromophobe, etc.) and imaging decisions will be similar to general oncology guidelines.
  - Individuals with all other subtypes of RCC should be imaged according to **Renal Cell Cancer (RCC) (ONC-17)** in the Oncology Imaging Guidelines

- Many individuals will present with an asymptomatic abdominal mass, and will undergo ultrasound as a primary evaluation. 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.
- Most individuals will have surgical resection of all disease at the time of diagnosis and will enter directly into surveillance

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

**ONCP.RC.0007.5.C**
**v1.0.2026**

**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 | • CT Abdomen and Pelvis with contrast (CPT® 74177)<br>◦ Doppler ultrasound to evaluate for tumor thrombus is not medically necessary unless CT findings are inconclusive<br>• CT Chest with (CPT® 71260) or without contrast (CPT® 71250)<br>◦ Should be completed prior to anesthesia exposure if possible<br>• Bone scan (see: **PEDONC-1.3** for coding)<br>• MRI Brain without and with contrast (CPT® 70553)<br>• 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 | • 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 | • CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250)<br>• CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)<br>• Additionally, for individuals with CNS metastases at initial staging:<br>◦ MRI Brain without and with contrast (CPT® 70553) Bone scan (see: **PEDONC-1.3** for coding) at the end of planned therapy |

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

**Background and Supporting Information**

- Many individuals will present with an asymptomatic abdominal mass, and will undergo ultrasound as a primary evaluation. 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.

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

**ONCP.RC.0007.6.C**
**v1.0.2026**

Be careful not to confuse the diagnosis with rhabdomyosarcoma. See: **Rhabdomyosarcoma (RMS) (PEDONC-8.2)** for Imaging Guidelines

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

| Indication | Medically Necessary Imaging Study |
|---|---|
| Treatment response ~every 2 cycles during treatment and at the end of planned therapy | • CT Chest with (CPT® 71260) *or* CT Chest without contrast (CPT® 71250)<br>• CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)<br>◦ 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<br>• MRI Brain without and with contrast (CPT® 70553):<br>◦ Every 2 cycles during treatment for individuals with CNS metastases at initial staging<br>◦ At the end of planned therapy for all individuals<br>• 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 | • PET/CT (CPT® 78815)<br>• Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)** |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Surveillance | • Every 3 months for 2 years after completion of all therapy:<br>◦ CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250)<br>• Every 3 months for 3 years after completion of all therapy:<br>◦ CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)<br>▪ If primary site other than kidney, perform CT with contrast or MRI without and with contrast of primary site in place of abdominal imaging<br>• Every 3 months for 1 year, then every 6 months for 1 year after completion of all therapy:<br>◦ MRI Brain without and with contrast (CPT® 70553)<br>• If bone scan positive at initial diagnosis:<br>◦ Every 3 months for 1 year, then every 6 months for 2 years:<br>▪ Bone scan (see: **Modality General Considerations (PEDONC-1.3)** for coding)<br>◦ If negative at 36 months, no further advanced imaging<br>• 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**

- MRT is a highly aggressive histologic variant that can also occur in other locations and all non-CNS sites should follow these guidelines.
- Primary CNS rhabdoid malignancies should be imaged according to **Atypical Teratoid/Rhabdoid Tumors (ATRT) PEDONC-4.5)**
- Many individuals will present with an asymptomatic abdominal mass, and will undergo ultrasound as a primary evaluation.

# Congenital Mesoblastic Nephroma (CMN) (PEDONC-7.7)

**ONCP.RC.0007.7.C**
**v1.0.2026**

### Congenital Mesoblastic Nephroma Initial Staging

- CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary in all individuals
- CT Chest with (CPT® 71260) is medically necessary to evaluate inconclusive findings on chest x-ray

### Congenital Mesoblastic Nephroma Treatment Response

- CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary once following resection to establish baseline imaging, and those with a complete resection should then be imaged according to surveillance guidelines
- CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary for individuals receiving preoperative chemotherapy every 2 cycles until surgery and then should be imaged according to surveillance guidelines after their postoperative baseline imaging study

### Congenital Mesoblastic Nephroma Surveillance Imaging

- Ultrasound is medically necessary as it is the preferred surveillance imaging modality to avoid radiation and anesthesia exposures
- CT Abdomen and Pelvis with contrast (CPT® 74177) **or** MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) is medically necessary for residual abnormalities present on post-operative imaging or inconclusive findings on ultrasound every 3 months for 1 year after completion of all therapy

### Background and Supporting Information

- This is the most common primary renal tumor occurring in young infants, and the overall prognosis is very good
- Complete surgical removal is curative in most cases, and histologically confirmed metastatic disease or bilateral disease has never been reported
- Many individuals will present with an asymptomatic abdominal mass at the time of birth or abnormal prenatal ultrasound, and will undergo ultrasound as a primary evaluation.
- PET is not indicated in the initial staging of any pediatric renal tumor
- Recurrences are rare but most occur within 12 months of diagnosis

# References (PEDONC-7)

**v1.0.2026**


1. Fernandez C, Geller JI, Ehrlich PF, et al. Renal tumors. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:753-771.
2. Dome JS, Mullen EA, and Argani P. Pediatric renal tumors. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th ed. Philadelphia, PA: Elsevier Saunders; 2015:1714-1746.
3. Kaste SC, Brady SL, Yee B, et al. Is routine pelvic surveillance imaging necessary in patients with Wilms tumor? *Cancer*. 2013;119(1):182-188. doi:10.1002/cncr.27687.
4. Khanna G, Rosen N, Anderson JR, et al. Evaluation of diagnostic performance of CT for detection of tumor thrombus in children with Wilms tumor: a report from the Children's Oncology Group. *Pediatr Blood Cancer*. 2012;58(4):551-555. doi:10.1002/pbc.23222.
5. Misch D, Steffen IG, Schönberger S, et al. Use of positron emission tomography for staging, preoperative response assessment and post therapeutic evaluation in children with Wilms tumor. *Eur J Nucl Med Mol Imaging*. 2008;35(9):1642-1650. doi:10.1007/s00259-008-0819-9.
6. Servaes S, Khanna G, Naranjo A, et al. Comparison of diagnostic performance of CT and MRI for abdominal staging of pediatric renal tumors: a report from the Children's Oncology Group. *Pediatr Radiol*. 2015;45(2):166-172. doi:10.1007/s00247-014-3138-2.
7. Servaes SE, Hoffer FA, Smith EA, Khanna G. Imaging of Wilms tumor: an update. *Pediatr Radiol*. 2019;49(11):1441-1452. doi:10.1007/s00247-019-04423-3.
8. Uslu L, Doing J, Link M, Rosenburg J, Quon A, Daldrup-Link HE. Value of 18F-FDG PET and PET/CT for Evaluation of Pediatric Malignancies. *J Nucl Med*. 2015;56(2):274-286. doi:10.2967/jnumed.114.146290.
9. Dome JS, Graf N, Geller JI, et al. Advances in Wilms tumor treatment and biology: progress through international collaboration. *J Clin Oncol*. 2015;33(27):2999-3007. doi:10.1200/JCO.2015.62.1888.
10. Venkatramani R, Chi Y-Y, Coppes MJ, et al. Outcome of patients with intracranial relapse enrolled on National Wilms Tumor Study Group clinical trials. *Pediatr Blood Cancer*. 2017;64(7):e26406. doi:10.1002/pbc.26406.
11. Mullen EA, Chi Y-Y, Hibbitts E, et al. Impact of surveillance imaging modality on survival after recurrence in patients with favorable-histology Wilms tumor: a report from the Children's Oncology Group. *J Clin Oncol*. 2018;36:3396-3403. doi:10.1200/JCO.18.00076.
12. Ehrlich P, Chi Y-Y, Chintagumpala MM, et al. results of the first prospective multi-institutional treatment study in children with bilateral Wilms tumor (AREN0534): a report from the Children's Oncology Group. *Ann Surg*. 2017;266(3):470-478. doi:10.1097/SLA.0000000000002356.
13. Geller JI, Ehrlich PF, Cost NG, et al. Characterization of adolescent and pediatric renal cell carcinoma: a report from the Children's Oncology Group study AREN03B2. *Cancer*. 2015 July; 121(14):2457-2464. doi:10.1002/cncr.29368.
14. Rialon KL, Gulack BC, Englum BR, Routh JC, Rice HE. Factors impacting survival in children with renal cell carcinoma. J *Pediatr Surg*. 2015;50(6):1014-1018. doi:10.1016/j.jpedsurg.2015.03.027.
15. Gooskens SL, Houwing ME, Vujanic GM, et al. Congenital mesoblastic nephroma 50 years after its recognition: a narrative review. *Pediatr Blood Cancer*. 2017; 64(7):e26437. doi:10.1002/pbc.26437.
16. Seibel NL, Chi YY, et al. Impact of cyclophosphamide and etoposide on outcome of clear cell sarcoma of the kidney treated on the National Wilms Tumor Study-5 (NWTS-5). *Pediatr Blood Cancer*. 2019;66(1):e27450. doi:10.1002/pbc.27450.
17. Balis F, Green DM, Armstrong A, et. al. National Comprehensive Cancer Network (NCCN) Guidelines Version 2.2025 – June 11, 2025. Wilms Tumor (Nephroblastoma), available at: https://www.nccn.org/professionals/physician_gls/pdf/wilms_tumor.pdf. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Wilms Tumor (Nephroblastoma) V2.2025 – June 11, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.

18. Servaes SE, Hoffer FA, Smith EA, Khanna G. Imaging of Wilms tumor: an update. *Pediatric Radiol*. 2019;49(11):1441–1452. doi:10.1007/s00247-019-04423-3.
19. Dix DB, Seibel NL, Chi Y, et. al. Treatment of stage IV favorable histology Wilms tumor with lung metastases: a report from the Children's Oncology Group AREN0533 study. *J Clin Oncol*. 2018;36(16):1564-1570. doi:10.1200/JCO.2017.77.1931.

# Pediatric Soft Tissue Sarcomas (PEDONC-8)

Guideline Page







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

**ONCP.SS.0008.1.C**

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- Unless specified below, individuals age <18 years old should be imaged according to this guideline section. Exceptions include:
  - Rhabdomyosarcoma in individuals (except uterine rhabdoymyosarcoma) of all ages should be imaged according to guidelines in **Rhabdomyosarcoma (RMS) (PEDONC-8.2)**
  - Uterine rhabdomyosarcoma 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 age 0 to 17 years, see: **Soft Tissue and Bone Masses (PEDMS-3)**in the Pediatric Musculoskeletal Imaging Guidelines
  - Individuals age ≥18 years, 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 medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**





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# Rhabdomyosarcoma (RMS) (PEDONC-8.2)

**ONCP.SS.0008.2.A**

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- 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 | ALL of the following:<br>• MRI without and with contrast of primary site<br>◦ CT with contrast if MRI is contraindicated<br>• CT Chest with contrast or CT Chest without contrast<br>◦ Should be completed prior to anesthesia exposure if possible<br>• Whole-Body PET/CT (CPT® 78816)<br>◦ Bone scan substituted for PET, if PET not available |
| Additional initial staging for ANY of the following:<br>• Evaluation of inconclusive PET findings in the abdomen or pelvis<br>• Primary site of abdomen or pelvis<br>• Lower extremity primary site | • CT Abdomen and Pelvis with contrast (CPT 74177) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Additional initial staging for ANY of the following:<br>• Primary site involving the paraspinal or paravertebral region<br>• PET or bone scan-avid lesions in skull, neck, vertebrae<br>• Neurologic signs or symptoms raising suspicion of CNS metastases | ALL of the following:<br>• MRI Brain (CPT® 70553)<br>• 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 | ALL of the following:<br>• CT Chest with (CPT® 71260) contrast or CT Chest without contrast (CPT® 71250)<br>• MRI without and with contrast of primary site<br>◦ CT if used at initial imaging for MRI contraindication<br>• 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:<br>• Response assessment prior to local control surgery or radiation therapy<br>• Evaluation of residual mass visible on conventional imaging as part of end of therapy evaluation<br>• Response assessment of disease visible on PET but not conventional imaging<br>• 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)<br>◦ 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 medically necessary for continued disease monitoring or surveillance unless one of the exceptions in section **General Guidelines (PEDONC-1.0)** applies. |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Restaging following local control surgery | ALL of the following:<br>• MRI without and with contrast of primary site<br>◦ CT if used at initial imaging for MRI contraindication<br>• 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, localized RMS | ALL of the following, every 3 months for 1 year, then every 4 months for 2 years, then every 6 months for 1 year after completion of all therapy:<br>• MRI without or without and with contrast of primary tumor site<br>◦ CT if MRI contraindicated<br>• Chest x-ray<br>◦ 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 |
| Surveillance, metastatic RMS | ALL of the following, every 3 months for 1 year, then every 4 months for 2 years, then every 6 months for 1 year after completion of all therapy:<br>• MRI without or without and with contrast of primary tumor site<br>• CT if contraindication to MRI<br>• CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250)<br>• CT with or without contrast of all known metastatic sites<br>• Nuclear bone scan (see: **PEDONC-1.3** for coding) for surveillance of known bony metastases |
| Suspected recurrence | • Repeat conventional imaging as per initial staging for all individuals |

| Indication | Medically Necessary Imaging Study |
|---|---|
| ONE of the following:<br>• Biopsy-proven recurrence<br>• 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<br>• Rare circumstances where obvious clinical symptoms show strong evidence suggesting recurrence, and PET would replace conventional imaging modalities. | • Whole-body PET/CT (CPT® 78816) |

**Background and Supporting Information**

- Soft tissue sarcomas occur in both adult and pediatric individuals, but some are more common in one age group than the other.
- Pediatric soft tissue sarcomas are divided into **two** groups:
  - Rhabdomyosarcoma (RMS) accounts for ~60% of soft tissue sarcomas in young individuals, but only ~25% of soft tissue sarcomas in adolescents
  - Non-rhabdomyosarcoma soft tissue sarcomas (NRSTS) which encompasses all other histologic subtypes
- Because RMS can arise from any muscle tissue, the presenting symptoms and primary tumor sites vary widely and strongly influence the appropriate imaging decisions
- CT with contrast is the preferred primary site imaging modality for RMS arising in the thoracic cavity (not the chest wall)
- MRI without and with contrast is the preferred primary site imaging modality for RMS occurring in all other anatomic locations, including the chest wall.
- PET/CT is superior to conventional imaging for detection of nodal and bony metastases in pediatric RMS and is indicated in the initial staging of all individuals after histologic diagnosis is established
- CT Abdomen and Pelvis with contrast (CPT® 74177) is not routinely indicated in the initial metastatic staging of pediatric RMS
- PET is not routinely utilized to assess treatment response in RMS

- PET is generally not indicated during active treatment for recurrent pediatric cancer
- Residual mass that has not changed in size since the last conventional imaging does not justify PET imaging during surveillance
- PET avidity in a residual mass at the end of planned therapy is not an indication for PET imaging during surveillance

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

**ONCP.SS.0008.3.A**
**v1.0.2026**

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging of all individuals | ALL of the following:<br>• MRI without and with contrast of primary site<br>◦ CT with contrast if contraindication to MRI<br>• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250)<br>◦ Should be completed prior to anesthesia if possible<br>• 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)<br>• PET/CT (CPT® 78815)<br>◦ Whole-body PET/CT (CPT® 78816) if there is clinical suspicion of skull or distal lower extremity involvement<br>• Nuclear bone scan (see: **PEDONC-1.3** for coding) if PET is not available |

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

| Indication | Medically Necessary Imaging Study |
|---|---|
| Treatment response, every 2 cycles of treatment, prior to local control, and at the end of planned therapy | Either of the following:<br>• CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250), and<br>• MRI without and with contrast of primary site<br>◦ CT if contraindication to MRI<br>and<br>• CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/ pelvic imaging for initial staging<br>• Imaging of all known metastatic sites using the same conventional imaging modality as per initial staging<br>OR<br>• PET/CT (CPT® 78815) if disease visible on initial staging PET but not on conventional imaging<br>◦ 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:<br>• Assessment prior to local control surgery or radiation therapy<br>• Evaluation of residual mass visible on conventional imaging as part of end of therapy evaluation<br>• 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 (CPT® 78815)<br>◦ Whole-body PET/CT (CPT® 78816) if there is clinical suspicion of skull or distal lower extremity involvement<br>◦ 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 medically necessary for continued disease monitoring or surveillance unless one of the exceptions in section **General Guidelines (PEDONC-1.0)** applies. |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Restaging following local control surgery | ALL of the following:<br>• MRI without and with contrast of the primary site<br>◦ CT if contraindication to MRI<br>• Imaging of all known metastatic sites using the same conventional imaging modality as per initial staging<br>• 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 | 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:<br>• MRI without and with contrast of the primary site<br>• CT if MRI contraindicated<br>AND<br>Every 6 months for 3 years, then annually thereafter:<br>• CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/ pelvic imaging for initial staging<br>• CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250) |
| Surveillance all other NRSTS | Every 3 months for the first 2 years, then every 6 months in years 3 and 4, and then annually:<br>• MRI without and with contrast of the primary site<br>◦ CT if MRI contraindicated<br>◦ Nuclear bone scan (see: **PEDONC-1.3** for coding) for surveillance of known bony metastases<br>• CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250)<br>• CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/ pelvic imaging for initial staging |

| Indication | Medically Necessary Imaging Study |
|---|---|
| ANY of the following:<br>• Angiosarcoma<br>• Alveolar soft part sarcoma<br>• Cardiac sarcoma | • In addition to the above studies:<br>◦ MRI Brain without and with contrast (CPT® 70553) annually<br>For surveillance of individuals with known brain metastases, see: **Brain Metastases (ONC-31.3)** |
| Surveillance, 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):<br>• CT Chest (CPT® 71260) with contrast<br>• CT with contrast or MRI without and with contrast of the primary site<br>• CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/ pelvic imaging for initial staging |
| ANY of the following:<br>• 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<br>• Obvious clinical symptoms show strong evidence suggesting recurrent and PET would replace conventional imaging modalities | • PET/CT (CPT® 78815)<br>◦ Whole-body PET/CT (CPT® 78816) if there is clinical suspicion of skull or distal lower extremity involvement |

**Background and Supporting Information**

- All soft tissue sarcomas other than RMS fall into this category
- Because soft tissue sarcomas can arise from any soft tissue, the presenting symptoms and primary tumor sites vary widely and strongly influence the appropriate imaging decisions

- CT Abdomen and Pelvis with contrast (CPT® 74177) is not routinely indicated in the initial metastatic staging of pediatric NRSTS
- Many individuals with NRSTS will be treated with surgical resection alone, and these individuals enter immediately into surveillance
- PET imaging is not routinely utilized to assess treatment response in NRSTS
- PET imaging is generally not indicated during active treatment for recurrent pediatric cancer.
- Residual mass that has not changed in size since the last conventional imaging does not justify PET
- PET avidity in a residual mass at the end of planned therapy is not an indication for PET imaging during surveillance
- Given that pediatric-specific resources are limited in regards to surveillance of NRSTS, this guideline combines published pediatric literature with NCCN recommendations for the same histologies.

# References (PEDONC-8)

**v1.0.2026**


1. Wexler LH, Skapek SX, Helman LJ. Rhabdomyosarcoma. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th edition. Philadelphia, PA: Wolters Kluwer; 2016:798-826.
2. Spunt SL, Million L, and Coffin C. The Nonrhabdomyosarcoma soft tissue sarcomas. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th edition. Philadelphia, PA: Wolters Kluwer; 2016:827-854.
3. Womer RB, Barr FG, Linardic CM. Rhabdomyosarcoma. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th edition. Philadelphia, PA: Elsevier Saunders; 2015:1906-1945.
4. Davis IJ, Perez-Atayde AR, Fisher DE. Nonrhabdomyosarcoma soft tissue sarcomas and other soft tissue tumors. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th edition. Philadelphia, PA: Elsevier Saunders; 2015:1946-1982.
5. McCarville MB. What MRI can tell us about neurogenic tumors and rhabdomyosarcoma. *Pediatr Radiol*. 2016;46(6):881-890. doi:10.1007/s00247-016-3572-4.
6. Federico SM, Spunt SL, Krasin MJ, et al. Comparison of PET-CT and Conventional Imaging in staging pediatric rhabdomyosarcoma. *Pediatr Blood Cancer*. 2013;60(7):1128-1134. doi:10.1002/pbc.24430.
7. Arnold MA, Anderson JA, Gastier-Foster JM, et al. Histology, fusion status, and outcome in alveolar rhabdomyosarcoma with low-risk clinical features: a report from the Children's Oncology Group. *Pediatr Blood Cancer*. 2016;63(4):634-639. doi:10.1002/pbc.25862.
8. Völker T, Denecke T, Steffen I, et al. Positron Emission Tomography for staging of pediatric sarcoma patients: results of a prospective multicenter trial. *J Clin Oncol*. 2007; 25(34):5435-5441. doi:10.1200/JCO.2007.12.2473.
9. Ostermeier A, McCarville MB, Navid F, et al. FDG PET/CT imaging of desmoplastic small round cell tumor: findings at staging, during treatment and at follow-up. *Pediatr Radiol*. 2015;45(9):1308-1315. doi:10.1007/s00247-015-3315-y.
10. Uslu L, Doing J, Link M, Rosenberg J, Quon A, Daldrup-Link HE. Value of 18F-FDG PET and PET/CT for evaluation of pediatric malignancies. *J Nucl Med*. 2015;56(2):274-286. doi:10.2967/jnumed.114.146290.
11. Roberts CC, Kransdorf MJ, Beaman FD, et al. Follow-Up of malignant or aggressive musculoskeletal tumors. ACR Appropriateness Criteria® 2015:1-15. https://acsearch.acr.org/docs/69428/Narrative/.
12. Ferrari A, Chi Y-Y, De Salvo GL, et al. Surgery alone is sufficient therapy for children and adolescents with low-risk synovial sarcoma: a joint analysis from the European paediatric soft tissue sarcoma Study Group and the Children's Oncology Group. *Eur J Cancer*. 2017;78:1-6. doi:10.1016/j.ejca.2017.03.003.
13. Spunt SL, Million L, Chi YY, et. al. A risk-based treatment strategy for non-rhabdomyosarcoma soft-tissue sarcomas in patients younger than 30 years (ARST0332): a Children's Oncology Group prospective study. *Lancet Oncol*. 2020;21(1):145-161. doi:10.1016/S1470-2045(19)30672-2.
14. Acord MR, Pace E, El-Ali A, et. al. Imaging of pediatric extremity soft tissue tumors: A COG Diagnostic Imaging Committee/SPR Oncology Committee white paper. *Pediatr Blood Cancer*. 2022:e29944. doi:10.1002/pbc.29944.
15. Abramson ZR, Nagaraj UD, Lai LM, et. al. Imaging of pediatric head and neck tumors: A COG Diagnostic Imaging Committee/SPR Oncology Committee/ASPNR white paper. *Pediatr Blood Cancer*. 2023;70 Suppl 4:e30151. doi:10.1002/pbc.30151.
16. Von Mehren M, Kane III JM, Armstrong SA, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 1.2025 – May 2, 2025. Soft Tissue Sarcoma, available at: https://www.nccn.org/ professionals/physician_gls/pdf/sarcoma.pdf. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Soft Tissue Sarcoma V1.2025– May 2, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.

# Bone Tumors (PEDONC-9)

Guideline Page







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# Bone Tumors – General Considerations (PEDONC-9.1)

**ONCP.BT.0009.1.C**
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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 individuals of all ages should be imaged according to guidelines in **Osteogenic Sarcoma (OS) (PEDONC-9.3)**
  - Ewing Sarcoma and Primitive Neuroectodermal Tumor individuals of all ages should be imaged according to guidelines in **Ewing Sarcoma and Primitive Neuroectodermal Tumors (ESFT) (PEDONC-9.4)**
  - Chondrosarcoma individuals of all ages should be imaged according to guidelines in **Bone Sarcomas – Initial Work-up/Staging (ONC-12.6)** in the Oncology Imaging Guidelines
  - Chordoma individuals of all ages should be imaged according to guidelines in **Bone Sarcomas – Initial Work-up/Staging (ONC-12.6)** in the Oncology Imaging Guidelines
  - Giant cell tumor of bone and enchondroma individuals of all ages should be imaged according to guidelines in **Benign Bone Tumors – General Considerations (ONC-12.9)** in the Oncology Imaging Guidelines
  - 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 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 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 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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# Benign Bone Tumors (PEDONC-9.2)

**ONCP.BT.0009.2.A**
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- Osteochondroma

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

- Osteoid osteoma

| Indication | Medically Necessary Imaging Study |
|---|---|
| Suspected, based on clinical history and plain film findings | • CT without contrast |
| • ANY of the following:<br>◦ CTs are not characteristic for diagnosis<br>◦ Individual has bone pain not localized to the area of findings on CT or x-ray | • ONE of the following:<br>◦ Triple phase bone scan (CPT® 78315)<br>◦ SPECT (CPT® 78803)<br>◦ Hybrid SPECT/CT (CPT® 78830) |
| • Individuals 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
  - Refer to **Mass Involving Bone (Including suspected Lytic and Blastic Metastatic Disease) (PEDMS-3.4)**

**Background and Supporting Information**

- Plain x-ray appearance is diagnostic for many benign bone tumors, and advanced imaging is generally unnecessary except for preoperative planning
- Plain x-ray appearance is diagnostic for osteochondroma for the majority of individuals and advanced imaging is generally unnecessary

# Osteogenic Sarcoma (OS) (PEDONC-9.3)

**ONCP.BT.0009.3.A**
**v1.0.2026**

| Indication | Medically Necessary Imaging Study |
|---|---|
| Suspected or Biopsy Proven Diagnosis | • MRI without and with contrast of the primary site OR<br>• CT, contrast as request, of the primary site if there is a contraindication to MRI<br>AND<br>• CT Chest with (CPT® 71260) or without contrast (CPT® 71250)<br>◦ CT Chest is medically necessary in initial workup of all suspected malignant bone tumors in children and should be completed prior to anesthesia exposure if possible<br>• These studies are medically necessary even when PET/CT has already been authorized or performed |
| Initial Staging of Biopsy Proven Disease | • 18F-FDG PET/CT whole-body (CPT® 78816)<br>◦ Nuclear bone scan (see: **(PEDONC-1.3)** for coding) if PET not available<br>◦ PET/CT is medically necessary in addition to the conventional imaging listed in the suspected/ diagnosis sections<br>• CT Abdomen and Pelvis with contrast (CPT® 74177) is not routinely medically necessary in the initial metastatic staging of pediatric OS but is medically necessary in the following situations:<br>◦ Evaluation of inconclusive PET findings<br>◦ Primary site of abdomen or pelvis |
| Suspected bony metastatic sites noted on PET or bone scan | • MRI without and with contrast of the suspected metastatic site OR<br>• CT with contrast of the suspected metastatic site |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Restaging after 10 to 12 weeks of neoadjuvant chemotherapy prior to local control surgery | • MRI without and with contrast of primary site<br>• CT Chest with (CPT® 71260) or without contrast (CPT® 71250)<br>• 18F-FDG PET/CT whole-body (CPT® 78816) or bone scan (see: **(PEDONC-1.3)** for coding) |
| Individuals with metastatic disease undergoing current chemotherapy | • CT Chest with (CPT®) or without contrast (CPT® 71250) can be performed every 2 cycles during treatment and at the end of planned chemotherapy<br>• MRI without and with contrast of primary site can be performed every 2 cycles during treatment and at the end of planned chemotherapy<br>• 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<br>• Imaging may be indicated more frequently around the time of surgical resection of primary or metastatic lesions to assess for resectability |
| Preoperative planning for local control surgery | • CT, contrast as requested<br>• MRA and/or CTA may rarely be indicated for complicated surgical resections, and is medically necessary after evaluation by the operating surgeon to clarify inconclusive MRI findings for preoperative planning<br>• See: **General Considerations (PEDONC-9.1)** for advancing imaging requests related to prosthetic planning |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Following local control surgery, until the end of planned adjuvant chemotherapy | • MRI without and with contrast of primary site ~6 weeks after surgical procedure, and at the end of planned chemotherapy<br>• Plain x-rays of the primary site and chest immediately after local control and then every 2 months between the supported, listed advanced imaging studies.<br>• CT Chest with (CPT® 71260) or without contrast (CPT® 71250):<br>◦ Measurable pulmonary metastases: every 6 weeks, and at the end of planned chemotherapy<br>◦ No measurable pulmonary metastases: every 4 months, and at the end of planned chemotherapy<br>• Bone scan (see: **(PEDONC-1.3)** for coding) every 4 months, and at the end of planned chemotherapy<br>◦ 18F-FDG PET/CT whole-body (CPT® 78816) in place of bone scan, if positive for distant bone metastases at initial diagnosis |
| Recurrent metastatic or recurrent unresectable disease on treatment | • The following every 2 cycles of treatment, and at the end of planned chemotherapy:<br>◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250)<br>◦ MRI without and with contrast of primary site<br>◦ PET is generally not medically necessary during active treatment for recurrent pediatric cancer. In rare circumstances, 18F-FDG PET/CT imaging with CPT® 78816 may be appropriate when results are likely to result in a treatment change for the individual, including a change from active treatment to surveillance. |

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

| Indication | Medically Necessary Imaging Study |
|---|---|
| • For suspected recurrence, ANY of the following:<br>◦ Conventional imaging reveals findings that are inconclusive or suspicious for recurrence and PET avidity will determine whether biopsy or continued observation is appropriate<br>◦ 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 | • PET/CT whole-body (CPT® 78816) for biopsy-proven recurrence<br>◦ If disease is considered potentially resectable, follow osteosarcoma treatment response as previous |

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

**ONCP.BT.0009.4.A**
**v1.0.2026**

| Indication | Medically Necessary Imaging Study |
|---|---|
| Suspected/Diagnosis – 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 – Primary site | • MRI without and with contrast<br>◦ CT, contrast as requested, if there is a contraindication to MRI<br>• MRA and/or CTA may rarely be indicated for complicated surgical resections, and is medically necessary after evaluation by the operating surgeon to clarify inconclusive MRI findings for preoperative planning<br>• CT Chest with (CPT® 71260) or without contrast (CPT® 71250)<br>◦ CT Chest is 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<br>• These studies are medically necessary even when PET/CT has already been authorized or performed |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Suspected or Biopsy Proven Diagnosis – Chest wall primary | • MRI Chest without and with contrast AND<br>• CT Chest with (CPT® 71260) or without contrast (CPT® 71250)<br>◦ CT Chest is 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<br>• These studies are medically necessary even when PET/CT has already been authorized or performed |
| Initial Staging (additional imaging after biopsy confirmed disease) | • PET/CT whole-body (CPT® 78816)<br>◦ Bone scan (see: **(PEDONC-1.3)** for coding) substituted for PET imaging if PET not available<br>◦ PET/CT is medically necessary in addition to the conventional imaging listed in the suspected/ diagnosis sections |
| • For ANY of the following:<br>◦ Evaluation of inconclusive PET findings<br>◦ Primary site involving the abdomen or pelvis | • CT Abdomen and Pelvis with contrast (CPT® 74177) |
| Restaging after 10 to 12 weeks of neoadjuvant chemotherapy prior to local control surgery | Imaging modality should be the same as used for initial staging, any or all from the list below as described:<br>• MRI without and with contrast of the primary site<br>• CT (contrast as requested) if requested per valuation by the operating surgeon for pre-operative planning<br>◦ See: **General Considerations (PEDONC-9.1)** for imaging requests related to prosthetic planning<br>• CT Chest with (CPT® 71260) or without contrast (CPT® 71250)<br>• ONE of the following:<br>◦ PET/CT whole-body (CPT® 78816)<br>◦ Whole-body bone scan (see: **PEDONC-1.3** for coding)<br>◦ MRI bone marrow blood supply/diffusion-weighted MRI with ADC (CPT® 77084) |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Treatment response following local control surgery | • MRI without and with contrast of primary site 3 months after surgical procedure and at the end of planned chemotherapy<br>• Plain x-rays of the primary site and chest immediately after local control then every 3 months<br>• CT Chest with (CPT® 71260) or without contrast (CPT® 71250):<br>◦ Measurable pulmonary metastases: every 6 weeks and at the end of planned chemotherapy<br>◦ No measurable pulmonary metastases: every 3 months and at the end of planned chemotherapy<br>• Whole-body bone scan (see: **PEDONC-1.3** for coding) at the end of planned chemotherapy<br>• PET/CT whole-body (CPT® 78816) for clinical or imaging findings suggestive of local recurrence |
| Individuals with metastatic disease undergoing current chemotherapy | • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) every 2 cycles during treatment and at the end of planned chemotherapy<br>• MRI without and with contrast of primary site every 2 cycles during treatment and at the end of planned chemotherapy<br>• 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<br>• Imaging may be indicated more frequently around the time of surgical resection of primary or metastatic lesions to assess for resectability |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Recurrent metastatic or recurrent unresectable disease on treatment | • Every 2 cycles of treatment, and at the end of planned chemotherapy:<br>◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250)<br>◦ MRI without and with contrast of primary site<br>◦ PET is generally not medically necessary during active treatment for recurrent pediatric cancer. In rare circumstances, PET imaging may be appropriate when results are likely to result in a treatment change for the individual, including a change from active treatment to surveillance. |
| Surveillance for all disease **other than** low grade, stage I disease | • 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:<br>◦ 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):<br>◦ Bone scan (see: **(PEDONC-1.3)** for coding)<br>• Every 3 months for year 1 and 2, then every 4 months in year 3, then every 6 months in year 4 and 5:<br>◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250)<br>◦ After year 5, CXR should be used for surveillance |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Surveillance for low grade stage I disease only | • 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:<br>◦ 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)<br>◦ Bone scan (see: **PEDONC-1.3** for coding)<br>• Every 3 months for year 1 and 2<br>◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250)<br>◦ After 24 months off therapy, CXR should be used for surveillance with CT supported for new or inconclusive findings on CXR |
| Suspected Recurrence | • MRI without and with contrast of the primary site and/or site of suspected recurrence based on symptoms or other imaging, OR<br>• 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<br>• CT Chest with (CPT® 71260) or without contrast (CPT® 71250)<br>◦ CT Chest is medically necessary in initial workup of all suspected malignant bone tumors in children and should be completed prior to anesthesia exposure if possible<br>• CT Abdomen and Pelvis with contrast (CPT® 74177) is not routinely medically necessary in the metastatic staging of pediatric EWS but is medically necessary in the following situations:<br>◦ Evaluation of inconclusive PET findings<br>◦ Primary site of abdomen or pelvis<br>• These studies are medically necessary even when PET/CT has already been authorized or performed |

| Indication | Medically Necessary Imaging Study |
|---|---|
| • For suspected recurrence, any of the following:<br>◦ Conventional imaging reveals findings that are inconclusive or suspicious for recurrence and PET avidity will determine whether biopsy or continued observation is appropriate<br>◦ 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 | • Refer to ESFT initial imaging for studies for suspected recurrence.<br>• PET/CT whole-body (CPT® 78816)<br>• If disease is considered potentially resectable, follow ESFT treatment response as previously noted |

**Background and Supporting Information**

- Bone and bone marrow metastases can occur in ESFT, and cause a significant change in treatment approach.
- 18F-FDG PET/CT can replace bone scan and bone marrow biopsy in ESFT individuals and is indicated in the initial staging of all ESFT individuals after histologic diagnosis is established

# References (PEDONC-9)

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1. Hawkins DS, Brennan BMD, Bölling T, et al. Ewing Sarcoma. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th edition. Philadelphia, PA: Wolters Kluwer; 2016:855-876.
2. Gorlick R, Janeway K, and Marina N. Osteosarcoma. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th edition. Philadelphia, PA: Wolters Kluwer; 2016:877-898.
3. DuBois SG, Grier HE, and Lessnick SL. Ewing Sarcoma. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th edition. Philadelphia, PA: Elsevier Saunders; 2015:1983-2017.
4. Janeway KA. Osteosarcoma. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th edition. Philadelphia, PA: Elsevier Saunders; 2015:2018-2055.
5. Azouz EM. Magnetic resonance imaging of benign bone lesions: cysts and tumors. *Top Magn Reson Imaging*. 2002 August;13(4):219-230.
6. Wolf M. Knee pain in children, part III: stress injuries, benign bone tumors, growing pains. *Pediatr Rev*. 2016;37(3):114-119. doi:10.1542/pir.2015-0042.
7. Uslu L, Doing J, Link M, Rosenberg J, Quon A, Daldrup-Link HE. Value of 18F-FDG PET and PET/CT for Evaluation of Pediatric Malignancies. *J Nucl Med*. 2015;56(2):274-286. doi:10.2967/jnumed.114.146290.
8. Reed DR, Hayashi M, Wagner L, et al. Treatment pathway of bone sarcoma in children, adolescents, and young adults. *Cancer*. 2017 March;123(12):2206-2218.
9. Hurley C, McCarville MB, Shulkin BL, et al. Comparison of 18F-FDG-PET-CT and Bone Scintigraphy for evaluation of osseous metastases in newly diagnosed and recurrent osteosarcoma. *Pediatr Blood Cancer*. 2016;63(8):1381-1386. Accessed January 4, 2018. doi:10.1002/pbc.26014.
10. Byun BH, Kong CB, Lim I, et al. Comparison of (18)F-FDG PET/CT and (99m)TC-MDP bone scintigraphy for detection of bone metastasis in osteosarcoma. *Skeletal Radiol*. 2013;42(12):1673-1681.
11. Quartuccio N, Treglia G, Saisano M, et al. The role of Fluorine-18-Fluorodeoxyglucose positron emission tomography in staging and restaging of patients with osteosarcoma. *Radiol Oncol*. 2013;47(2):97-102. doi:10.2478/raon-2013-0017.
12. Isakoff MS, Bielack SS, Meltzer P, Gorlick R. Osteosarcoma: current treatment and a collaborative pathway to success. *J Clin Oncol*. 2015;33(27):3029-3035. doi:10.1200/JCO.2014.59.4895.
13. Newman EN, Jones RL, Hawkins DS. An evaluation of [F-18]-Fluorodeoxy-D-Glucose positron emission tomography, bone scan, and bone marrow aspiration/biopsy as staging investigations in Ewing sarcoma. *Pediatr Blood Cancer*. 2013;60(7):1113-1117. doi:10.1002/pbc.24406.
14. Roberts CC, Kransdorf MJ, Beaman FD, et al. Follow-up of malignant or aggressive musculoskeletal tumors. ACR Appropriateness Criteria® 2015:1-15. https://acsearch.acr.org/docs/69428/Narrative/.
15. Meyer JS, Nadel HR, Marina N, et al. Imaging guidelines for children with Ewing sarcoma and osteosarcoma: a report from the Children's Oncology Group Bone Tumor Committee. *Pediatr Blood Cancer*. 2008;51(2):163-170. doi:10.1002/pbc.21596.
16. Gaspar N, Hawkins DS, Dirksen U, et al. Ewing sarcoma: current management and future approaches through collaboration. *J Clin Oncol*. 2015;33(27):3036-3046. doi:10.1200/JCO.2014.59.5256.
17. Mascarenhas L, Felgenhauer JL, Bond MC, et al. Pilot study of adding vincristine, topotecan, and cyclophosphamide to interval-compressed chemotherapy in newly diagnosed patients with localized Ewing sarcoma: a report from the Children's Oncology Group. *Pediatr Blood Cancer*. 2016;63(3):493-498. doi:10.1002/pbc.25837.
18. Weiser DA, Kaste SC, Siegel MJ, et al. Imaging in childhood cancer: a society for pediatric radiology and children's oncology group joint task force report. *Pediatr Blood Cancer*. 2013;60(8):1253-1260. doi:10.1002/pbc.24533.
19. Dimitrakopoulou-Strauss A, Strauss LG, Heichel T, et al. The role of quantitative 18F-FDG PET studies for the differentiation of malignant and benign bone lesions. *J Nucl Med*. 2002;43(4):510-518.

20. Bestic JM, Wessell DE, Beaman FD, et al. Primary bone tumors. ACR Appropriateness Criteria®, 2019;1-13. Date of origin: 1995 Last review date: 2019 https://acsearch.acr.org/docs/69421/Narrative/.
21. Biermann JS, Hirbe A, Ahlawat S, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 2.2025 – February 28, 2025. Bone cancer, available at: https://www.nccn.org/professionals/physician_gls/pdf/bone.pdf. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Bone Cancer V2.2025– February 28, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
22. Jeys L, Abudu A, Grimer R. Expandable prostheses. In: Wiesel SW, ed. Operative Techniques in Orthopaedic Surgical Oncology. Philadelphia, PA: Lippincott Williams & Wilkins; 2015:1-9.
23. PDQ® Pediatric Treatment Editorial Board. PDQ Osteosarcoma and Malignant Fibrous Histiocytoma of Bone Treatment. Bethesda, MD: National Cancer Institute. Updated 12/2/2024. Available at: https://www.cancer.gov/types/bone/hp/osteosarcoma-treatment-pdq. [PMID: 26389179].
24. Demertzis JL, Wessell DE, Lenchik L, et. al. ACR Appropriateness Criteria® malignant or aggressive primary musculoskeletal tumor-staging and surveillance: 2022 update. *J Am Coll Radiol*. 2022;19(11S):S374-S389. doi:10.1016/j.jacr.2022.09.015.
25. Cederberg KB, Iyer RS, Chaturvedi A, et. al. Imaging of pediatric bone tumors: A COG Diagnostic Imaging Committee/SPR Oncology Committee white paper. *Pediatr Blood Cancer*. 2023;70 Suppl 4:e30000. doi:10.1002/pbc.30000.

# Pediatric Germ Cell Tumors (PEDONC-10)

Guideline Page







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# Pediatric Germ Cell Tumors (PEDONC-10)

**ONCP.GC.0010.A**
**v1.0.2026**

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging for all individuals | • CT Abdomen and Pelvis with contrast (CPT® 74177)<br>◦<br>◦ For testicular masses, MRI Abdomen without and with contrast (74183) and CT Pelvis without and with contrast (CPT® 72193) are medically necessary instead, if requested<br>• CT Chest with contrast (CPT® 71260)<br>◦ Should be completed prior to anesthesia exposure if possible |
| • Initial staging, in lieu of CT Abdomen and Pelvis with contrast (above), for ANY of the following:<br>◦ Proven or highly suspected ovarian neoplasm that was already imaged with ultrasound<br>◦ Immature sacrococcygeal teratoma suspected on other imaging<br>◦ Contraindication to CT contrast<br>OR<br>• To clarify inconclusive findings on initial staging CT | • MRI Abdomen and Pelvis without and with contrast (CPT® 74183 or CPT® 72197) |





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| Indication | Medically Necessary Imaging Study |
|---|---|
| • Ovarian masses that are <10 cm 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 medically necessary |
| • Additional initial evaluation for individuals with ANY of the following:<br>◦ Symptoms suggesting CNS metastases<br>◦ Choriocarcinoma syndrome (hemorrhagic metastatic disease to lung with extremely elevated HCG) HCG >10,000miU/ml | • MRI Brain without and with contrast (CPT® 70553) |
| Additional initial evaluation for individuals with systemic symptoms or bone pain | • Nuclear bone scan (See: **PEDONC-1.3** for coding) |
| Restaging in individuals with disease not completely resected at initial diagnosis | • Every 2 cycles (~every 6 weeks) and at the end of planned therapy:<br>◦ CT Chest/Abdomen/Pelvis (CPT® 71260 and CPT® 74177) with contrast<br>▪ CT imaging may be indicated more frequently to assess for surgical resectability in individuals who have received more than 4 cycles of chemotherapy<br>▪ 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<br>◦ Imaging of any metastatic sites with the same modality used during initial staging |

| Indication | Medically Necessary Imaging Study |
|---|---|
| • Suspicious lesion seen on CT and BOTH of the following:<br>◦ Relapse risk is determined to be low by the treating physician<br>◦ Biopsy would cause unnecessary morbidity for the individual | • Short-interval CT study of the involved area |
| Stage I individuals age 0-10 years 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<br>◦ For those with primary mediastinal tumors at diagnosis, CT Chest with contrast (CPT® 71260) in lieu of CXR according to the above schedule.<br>• Every 3 months for 1 year, then every 6 months in year two, to complete two years surveillance after completion of all therapy:<br>◦ CT Abdomen/Pelvis with contrast (CPT® 74177)<br>▪ 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 I individuals ages 11+ years 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:<br>◦ Chest x-ray<br>◦ For individuals with primary mediastinal tumors at diagnosis, CT Chest with contrast (CPT® 71260) in lieu of chest x-ray on the above schedule.<br>• 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<br>◦ CT Abdomen and Pelvis with contrast (CPT® 74177)<br>▪ 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 II-IV individuals | • 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.<br>◦ 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<br>• 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 |

| Indication | Medically Necessary Imaging Study |
|---|---|
| • Suspected Recurrence:<br>◦ Any clinically significant rise in tumor markers<br>◦ Symptoms suggesting recurrent disease<br>◦ Abnormal chest x-ray | • CT Chest with contrast (CPT® 72160) and CT Abdomen and Pelvis with contrast (CPT® 74177)<br>◦ 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<br>• Whole-body bone scan (CPT® 78306) for individuals with a history of involvement or with bone pain |
| Clarify inconclusive findings on conventional imaging | PET/CT (CPT® 78815) |

**Background and Supporting Information**

**General Considerations:**

- Malignant pediatric germ cell tumors commonly include one of four histologic subtypes:
  - Yolk sac tumor
  - Choriocarcinoma
  - Embryonal carcinoma
  - Mixed histology (including immature sacrococcygeal teratoma)
- Tumors can occur in testicular, ovarian or extragonadal primary locations
- 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)**.

**Treatment Considerations:**

- Overall treatment strategies are similar for all malignant germ cell tumors.
- Individuals with localized GCT are often cured with surgery alone and do not receive adjuvant therapy.
  - These individuals should be imaged using surveillance guidelines after surgery is completed.
- Individuals receiving adjuvant chemotherapy are usually treated with 4 to 6 cycles of combination chemotherapy.

**Imaging Modality Considerations:**

- Initial evaluation of: Ovarian, testicular, and abdominal extragonadal suspected GCT should be completed by ultrasound and tumor markers (AFP, β-hCG)
  - Once a primary mass suspected to be GCT is discovered, initial staging is indicated prior to histologic confirmation
  - The degree of abdominal exploration and node sampling necessary for adequate staging is determined in part by imaging findings and is required for preoperative planning
- The primary method of response assessment is by tumor marker decrease
- The primary method of surveillance in pediatric GCT is frequent assessment of serum tumor markers, unless tumor markers were not elevated at diagnosis
- Surveillance imaging of the chest in disease stages I-IV should generally be performed using chest x-ray
  - See surveillance indications for specific imaging recommendations
- There has been no published evidence to date supporting the routine use of PET/CT in the evaluation of pediatric GCT
  - Additionally, PET has been found to have similar efficacy to CT imaging in initial staging of adults with non-seminomatous GCT (the majority of pediatric GCT are non-seminomatous)
- PET as a marker of treatment response has been shown not to be predictive of individual outcomes in GCT and should not be approved
  - Suspicious lesions seen on conventional imaging should be biopsied to confirm active disease

# References (PEDONC-10)

**v1.0.2026**


1. Frazier AL, Olson TA, Schneider DT, et al. Germ cell tumors. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th edition. Philadelphia, PA: Wolters Kluwer; 2016:899-918.
2. Frazier AL, Billmire D, Amatruda J. Pediatric germ cell tumors. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th edition. Philadelphia, PA: Elsevier Saunders; 2015:2056-2099.
3. Gilligan T, Lin DW, Adra N, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 2.2025—March 25, 2025, Testicular Cancer, available at: https://www.nccn.org/professionals/physician_gls/pdf/testicular.pdf, Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Testicular Cancer V2.2025 March 25, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org
4. Armstrong DK, Alvarez RD, Backes FJ, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 3.2025—July 16, 2025 Ovarian Cancer Including Fallopian Tube Cancer and Primary Peritoneal Cancer, available at: https://www.nccn.org/professionals/physician_gls/pdf/ovarian.pdf, Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Ovarian Cancer V3.2025 July 16, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
5. de Wit M, Brenner W, Hartmann M, et al. [18F]-FDG-PET in clinical stage I/II non-seminomatous germ cell tumors: results of the German multicentre trial. *Ann Oncol*. 2008;19(9):1619-1623. doi:10.1093/annonc/mdn170.
6. Huddart RA, O'Doherty MJ, Padhani A, et al. 18Flourodeoxyglucose positron emission tomography in the prediction of relapse in patients with high-risk, clinical stage I nonseminomatous germ cell tumors: preliminary report of MRC trial TE22--the NCRI testis tumor clinical study group. *J Clin Oncol*. 2007;25(21):3090-3095. doi:10.1200/JCO.2006.09.3831.
7. Papic JC, Finnell MA, Slaven JE, et al. Predictors of ovarian malignancy in children: overcoming clinical barriers of ovarian preservation. *J Pediatr Surg*. 2014;49(1):144-148. doi:10.1016/j.jpedsurg.2013.09.068.
8. Olson TA, Murray MJ, Rodriguez-Galindo C, et al. Pediatric and adolescent extracranial germ cell tumors: the road to collaboration. *J Clin Oncol*. 2015;33(27):3018-3028. doi:10.1200/JCO.2014.60.5337.
9. Fresneau B, Orbach D, Faure-Conter C, et al. Sex-cord stromal tumors in children and teenagers: results of the TGM-95 study. *Pediatr Blood Cancer*. 2015;62(12):2114-2119. doi:10.1002/pbc.25614.
10. Fonseca A, Xia C, Lorenzo AJ, et al. Detection of Relapse by Tumor Markers Versus Imaging in Children and Adolescents With Nongerminomatous Malignant Germ Cell Tumors: A Report From the Children's Oncology Group. *J Clin Oncol*. 2019;37:396-402. doi:10.1200/JCO.18.00790.
11. Behr GG, Morani AC, Artunduaga M, et. al. Imaging of pediatric ovarian tumors: A COG Diagnostic Imaging Committee/SPR Oncology Committee white paper. *Pediatr Blood Cancer*. 2022:e29995. doi:10.1002/pbc.29995.
12. Behr GG, Morani AC, Artunduaga M, et. al. Imaging of pediatric testicular tumors: A COG Diagnostic Imaging Committee/SPR Oncology Committee white paper. *Pediatr Blood Cancer*. 2022:e29988. doi:10.1002/pbc.29988.
13. Keenan C, Ramirez N, Elijovich L, et. al. A rare manifestation of choriocarcinoma syndrome in a child with primary intracranial germ cell tumor and extracranial metastases: A case report and review of the literature. *Pediatr Blood Cancer*. 2021;68(6):e29000. doi:10.1002/pbc.29000.
14. Pace E, Johnson TS, Kao SC, et. al. Imaging of pediatric extragonadal pelvic soft tissue tumors: A COG Diagnostic Imaging Committee/SPR Oncology Committee white paper. *Pediatr Blood Cancer*. 2023;70 Suppl 4:e29966. doi:10.1002/pbc.29966.

# Pediatric Liver and Pancreatic Tumors (PEDONC-11)







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

**ONCP.LT.0011.1.C**

**v1.0.2026**

- 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: **Transplant (AB-42)** in the Abdomen Imaging Guidelines.

# Hepatoblastoma (PEDONC-11.2)

**ONCP.LT.0011.2.C**
**v1.0.2026**

**Hepatoblastoma Initial Staging:**

- Ultrasound imaging for most suspected liver tumors as initial evaluation
- Ultrasound is medically necessary even after MRI or CT imaging in order to allow evaluation for tumor thrombus
- Once a primary liver mass is discovered, definitive imaging is medically necessary prior to histologic diagnosis, and may involve 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 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 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
- CT Chest with (CPT® 71260) **or** CT Chest without contrast (CPT® 71250) is medically necessary in the initial workup of all pediatric liver tumors and should be completed prior to anesthesia exposure if possible
- Bone scan (See: **Modality General Considerations (PEDONC-1.3)**) is medically necessary for initial evaluation of bony metastases only with systemic symptoms or bone pain.
- MRI Brain without and with contrast (CPT® 70553) is medically necessary only for symptoms suggesting CNS metastases
- PET/CT is medically necessary only in very rare circumstances for preoperative planning when MRI and CT scans are insufficient for surgical decision-making.
  - Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

**Hepatoblastoma Treatment Response:**

- CT Chest with (CPT® 71260) **or** CT Chest without contrast (CPT® 71250) is medically necessary every 2 cycles **and** at the end of planned therapy for individuals with incomplete resection at initial diagnosis





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- MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) is medically necessary every 2 cycles **and** at the end of planned therapy for individuals with incomplete resection at initial diagnosis
  - 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 medically necessary if MRI is not available, contraindicated, or sedation is required but not available, or to clarify specific inconclusive areas on MRI.
- 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
- MRA Abdomen (CPT® 74185) or CTA Abdomen (CPT® 74175) is medically necessary to evaluate vascular invasion
- Imaging of any metastatic sites with the same modality used during initial staging is medically necessary every 2 cycles and at the end of planned therapy for individuals with incomplete resection at initial diagnosis
- Imaging is medically necessary more frequently to assess for surgical resectability in individuals who have received more than 4 cycles of chemotherapy.
- Abdominal ultrasound is medically necessary if tumor thrombus was detected at initial diagnosis.
  - If no tumor thrombus was present, continued ultrasound evaluations are not medically necessary without a specific reason documented in the clinical records
- PET/CT is medically necessary only in very rare circumstances for preoperative planning when MRI and CT scans are insufficient for surgical decision making.

**Hepatoblastoma Surveillance Imaging:**

- For surveillance in individuals with an AFP of >100 ng/mL, the following imaging is medically necessary:
  - 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 medically necessary if MRI is not available, contraindicated, or sedation is required but not available, or to clarify specific inconclusive areas on MRI.
- For individuals with AFP ≤100 ng/mL at diagnosis or recurrence, the following imaging is medically necessary:
  - 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**

- Pediatric liver tumors primarily include hepatoblastoma and hepatocellular carcinoma, but hepatic germ cell tumors and primary hepatic sarcomas occur with some frequency. Tumor markers are useful for initial evaluation as well as treatment response, particularly in hepatoblastoma. Early consideration of liver transplant may be undertaken in children and adolescents with unresectable localized disease, provided that the disease remains confined to the liver.
- Hepatoblastoma occurs most commonly in very young children (median diagnosis age of 19 months). Most cases of hepatoblastoma are sporadic, but some are associated with genetic abnormalities, including Beckwith-Wiedemann syndrome, familial adenomatous polyposis, and trisomy 18. Most suspected liver tumors will have ultrasound and tumor markers (AFP, β-hCG, CEA) as part of the initial evaluation.
- There has been no published evidence to date supporting the routine use of PET/CT imaging in the evaluation of pediatric hepatoblastoma during initial imaging, treatment response, or surveillance.
- PET/CT should not be approved in lieu of biopsy of suspicious lesions
- Individuals with localized hepatoblastoma of pure fetal histology are often cured with surgery alone and do not receive adjuvant therapy. These individuals should be imaged using surveillance guidelines after surgery is completed.
- Individuals receiving adjuvant chemotherapy are usually treated with 2 to 8 cycles of combination chemotherapy. Tumor marker decrease is important in response assessment but does not eliminate the need for advanced imaging in individuals with unresected hepatoblastoma.
- The primary method of surveillance in hepatoblastoma is frequent assessment of serum tumor markers (primarily AFP).
- No specific imaging for surveillance in individuals with an AFP of >100 ng/mL at diagnosis or recurrence.

# Pediatric Hepatocellular Carcinoma (HCC) (PEDONC-11.3)

**ONCP.LT.0011.3.C**
**v1.0.2026**

**Pediatric HCC Initial Staging:**

- Ultrasound for most suspected liver tumors as initial evaluation
- Ultrasound is medically necessary even after MRI or CT imaging in order to allow evaluation for tumor thrombus
- Once a primary liver mass is discovered, definitive imaging prior to histologic diagnosis including ANY of the following medically necessary studies:
  - 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 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 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
- CT Chest with (CPT® 71260) **or** CT Chest without contrast (CPT® 71250) is medically necessary in the initial workup of all pediatric liver tumors and should be completed prior to anesthesia exposure if possible
- MRI Brain without and with contrast (CPT® 70553) is medically necessary only for symptoms suggesting CNS metastases
- Nuclear bone scan (See: **Modality General Considerations (PEDONC-1.3)**) is medically necessary for initial evaluation of bony metastases only in individuals with systemic symptoms or bone pain
- PET/CT is medically necessary in very rare circumstances for preoperative planning when MRI and CT are insufficient for surgical decision-making.
  - Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

**Pediatric HCC Treatment Response:**

- For individuals with disease not completely resected at initial diagnosis, the following imaging is medically necessary 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)
    - 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
    - CT Abdomen and Pelvis with contrast (CPT® 74177) is inferior to MRI for pediatric liver malignancies and is only 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
- Imaging of any metastatic sites with the same modality used during initial staging is medically necessary
- Abdominal ultrasound is medically necessary if tumor thrombus was detected at initial diagnosis
  - If no tumor thrombus was present, continued ultrasound evaluations are not medically necessary without a specific reason documented in the clinical records
- PET/CT is medically necessary in very rare circumstances for preoperative planning when MRI and CT scans are insufficient for surgical decision-making.

**Pediatric HCC Surveillance Imaging:**

- MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) is medically necessary every 3 months for 1 year, then every 6 months for 1 year, then annually for 3 years after completion of all therapy
- Chest x-ray or CT Chest with contrast (CPT® 71260) is medically necessary every 3 months for 1 year, then every 6 months for 1 year, then annually for 3 years after completion of all therapy

**Background and Supporting Information**

- 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.
- HCC, including its rare histologically distinct variant fibrolamellar hepatocellular carcinoma (FL-HCC), occurs mostly in older children and adolescents. Despite recent advances in treatment, overall survival of pediatric HCC diagnosed in advanced stages remains exceedingly poor, with five-year survival of only 17% to 22% for

all stages of pediatric HCC (and FL-HCC). Most suspected liver tumors will have ultrasound and tumor markers (AFP, β-hCG, CEA) as initial evaluation.
- PET/CT should not be approved in lieu of biopsy of suspicious lesions
- The majority of hepatocellular carcinoma individuals are treated with surgery alone and do not receive adjuvant therapy. Individuals with successful upfront gross total resection should be imaged using surveillance guidelines after surgery is completed.
- PET/CT has no documented role in the surveillance evaluation of pediatric hepatocellular carcinoma.





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# Pediatric Pancreatic Carcinoma (PEDONC-11.4)

**ONCP.LT.0011.4.A**
**v1.0.2026**

- 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 | ▪ 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® 72917)<br>▪ CT Chest with (CPT® 71260) or without (CPT® 71250) contrast<br>▪ $^{68}$Ga-Dotatate PET/CT whole-body (CPT® 78816)<br>- 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 | ▪ CT or MRI as used at time of initial imaging |
| If conventional imaging for treatment response is equivocal | ▪ Neuroendocrine tumors: $^{68}$Ga Dotatate PET/CT skull to thighs or whole-body (CPT® 78815 or 78816)<br>▪ Non-neuroendocrine tumors: FDG PET/CT skull to thighs or whole-body (CPT® 78815 or 78816) |
| Assess candidacy for PRRT therapy | ▪ $^{68}$Ga Dotatate PET/CT whole body CPT® 78816 to assess candidacy for PRRT therapy |





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| Indication | Medically Necessary Imaging Study |
|---|---|
| Surveillance | ▪ 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 | ▪ Repeat all imaging as noted in initial staging section |

**Background and Supporting Information**

- Pancreatic tumors in children are exceedingly rare. The most common are solid pseudopapillary neoplasm (SPN), a low-grade epithelial malignancy. Pancreatoblastoma is the second most common.

# References (PEDONC-11)

**v1.0.2026**


1. Meyers RL, Trobaugh-Lotario AD, Malogolowkin MH, et al. Pediatric liver tumors. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th edition. Philadelphia, PA: Wolters Kluwer; 2016:726-752.
2. Tomlinson GE. Hepatoblastoma and other liver tumors in children. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th edition. Philadelphia, PA: Elsevier Saunders; 2015:1886-1905.
3. Pugmire BS, Towbin AJ. Magnetic resonance imaging of primary pediatric liver tumors. *Pediatr Radiol*. 2016;46:764-777.doi:10.1007/s00247-016-3612-0.
4. Chavhan GB, Shelmerdine S, Jhaveri K, Babyn PS. Liver MR Imaging in children: current concepts and technique. *RadioGraphics*. 2016;36(5):1517-1532. doi:10.1148/rg.2016160017.
5. Rai P, Feusner H. Cerebral metastasis of hepatoblastoma: a review. *J Pediatr Hematol Oncol*. 2016;38(4):279-282. doi:10.1097/MPH.0000000000000554.
6. Allen-Rhoades W, Whittle SB, Rainusso N. Pediatric solid tumors of infancy: an overview. *Pediatr Rev*. 2018;39:57-67. doi:10.1542/pir.2017-0057.
7. Allan BJ, Wang B, Davis JS, et al. A review of 218 pediatric cases of hepatocellular carcinoma. *J Pediatr Surg*. 2014;49(1):166-171. doi:10.1016/j.jpedsurg.2013.09.050.
8. Pappo AS, Furman WL, Schulz KA, Ferrari A, Helman L, Krailo MD. Rare tumors in children: progress through collaboration. *J Clin Oncol*. 2015;33(27):3047-3054. doi:10.1200/JCO.2014.59.3632.
9. Benson AB, D'Angelica MI, Abrams T, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 1.2025—March 20, 2025, Hepatocellular Carcinoma, available at: https://www.nccn.org/professionals/physician_gls/pdf/hcc.pdf, Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Hepatocellular Carcinoma V1.2025 March 20, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
10. Benson AB, D'Angelica MI, Abrams T, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 2.2025—July 2, 2025, Biliary Tract Cancers, available at: https://www.nccn.org/professionals/physician_gls/pdf/btc.pdf, Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Biliary Tract Cancers V2.2025 July 2, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org
11. D'Souza AM, Shah R, Gupta A, et al, Surgical management of children and adolescents with upfront completely resected hepatocellular carcinoma, Pediatr Blood Cancer. 2019;65:e27293. doi:10.1002/pbc.27293.
12. Weeda VB, Aronson DC, Verheij J, and Lamers WH, Is hepatocellular carcinoma the same disease in children and adults? Comparison of histology, molecular background, and treatment in pediatric and adult patients, Pediatr Blood Cancer. 2019;66:e27475. doi:10.1002/pbc.27475.
13. Eklund MJ, States LJ, Acord MR, et. al. Imaging of pediatric pancreas tumors: A COG Diagnostic Imaging Committee/SPR Oncology Committee White Paper. Pediatr Blood Cancer. 2022;e29975. doi:10.1002/pbc.29975.
14. Schooler GR, Infante JC, Acord M, et. al. Imaging of pediatric liver tumors: A COG Diagnostic Imaging Committee/SPR Oncology Committee white paper. Pediatr Blood Cancer. 2023;70 Suppl 4:e29965. doi:10.1002/pbc.29965. .

# Retinoblastoma (PEDONC-12)

Guideline Page







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# Retinoblastoma – General Considerations (PEDONC-12.1)

**ONCP.EC.0012.1.C**
**v1.0.2026**

- Retinoblastoma (RB) is primarily a disease of the infant and young child, and presents with leukocoria (loss of red reflex).
- About 75% of individuals are diagnosed before the age of two years (bilateral RB presents at 12 months of age).
- Retinoblastoma can occur as heritable (25% of cases) or nonheritable (75%) disease.
  - Heritable RB is associated with a germline mutation in the RB1 gene often resulting typically in bilateral disease.
  - 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)**.
  - Whole-body MRI has shown poor sensitivity and specificity in individuals with predisposition to systemic malignancy due to germline RB1 mutations, and is not supported.
- Detailed evaluation by a physician with significant training and/or experience in retinoblastoma (most commonly a pediatric ophthalmologist or pediatric oncologist) prior to considering advanced imaging.
- Retinoblastoma can be unilateral, bilateral, or trilateral (involving the pineal gland). Extraocular spread of retinoblastoma is rare and generally confined to the brain.

# Retinoblastoma – Imaging (PEDONC-12.2)

**ONCP.EC.0012.2.C**
**v1.0.2026**

**Retinoblastoma Medically Necessary Initial Staging Imaging:**

- Tumor biopsy is NOT required prior to imaging
- MRI Orbits (CPT® 70543) and Brain (CPT® 70553) without and with contrast in the initial workup
  - Brain imaging may be omitted or deferred at the discretion of the treating ophthalmologist or oncologist
- MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) if there is evidence of CNS metastasis on:
  - Ophthalmologic exam
  - MRI Brain
  - Lumbar CSF cytology
- CT Chest (CPT® 71260) **and** MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) with clinical symptoms to suggest metastatic disease
- CT Orbital (contrast as requested) and Orbital ultrasound if ordered by the treating ophthalmologist for a specified indication
- Nuclear bone scan (See: **Modality General Considerations (PEDONC-1.3)**) for systemic bone pain suggestive of bony metastases

**Retinoblastoma Medically Necessary Treatment Response Imaging:**

- MRI Orbits (CPT® 70543) **and/or** Brain (CPT® 70553) every 2 cycles (~every 6 weeks) **and** at the end of planned therapy
- For individuals with metastatic disease, imaging of known positive areas using the same modality at initial staging every 2 cycles (~6 to 8 weeks) **and** at the end of planned therapy

**Retinoblastoma Medically Necessary Surveillance Imaging:**

- 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**

- CT should generally be avoided in retinoblastoma individuals under one year of age or with family history of retinoblastoma (heritable) due to substantially increased risks for secondary malignancy
- PET has no documented role in the evaluation of retinoblastoma
- The primary method of surveillance in retinoblastoma is examination under anesthesia (EUA). Although some older children can be sufficiently evaluated by exam without anesthesia (EWA).
- Surveillance using advanced imaging is generally not indicated for unilateral retinoblastoma after enucleation or exenteration but can be approved for evaluation of specific clinical concerns.
- Individuals with bilateral retinoblastoma or germline mutation in RB1 are at increased risk for subsequent pineoblastoma
- Routine MRI follow up for pineal disease is not currently supported by evidence in unilateral retinoblastoma individuals without germline RB1 mutations

# References (PEDONC-12)

**v1.0.2026**


1. Hurwitz RL, Shields CL, Shields JA, et al. Retinoblastoma. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:700-725.
2. Rodriguez-Galindo C, Wilson MW, and Dyer M. Retinoblastoma. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th edition. Philadelphia, PA: Elsevier Saunders; 2015:1747-1778.
3. de Jong MC, Kors WA, de Graaf P, Castelijns JA, Kivelä T, Moll AC. Trilateral retinoblastoma: a systematic review and meta-analysis. *Lancet Oncol*. 2014;15(10):1157-1167. doi:10.1016/S1470-2045(14)70336-5.
4. De Graaf P, Göricke S, Rodjan F, et al. Guidelines for imaging retinoblastoma: imaging principles and MRI standardization. *Pediatr Radiol*. 2012;42(1):2-14. doi:10.1007/s00247-011-2201-5.
5. Allen-Rhoades W, Whittle SB, Rainusso N. Pediatric Solid Tumors of Infancy: An Overview, *Pediatr Rev*. 2018;39:57-67. doi:10.1542/pir.2017-0057.
6. Lohmann DR, Gallie BL. Retinoblastoma. In: Pagon RA, Adam MP, Ardinger HH et al, eds. *GeneReviews™ [Internet]*. Seattle, WA: University of Washington, Seattle; 2000-2023. https://www.ncbi.nlm.nih.gov/books/NBK1452/.
7. Freidman DN, Lis E, Sklar CA, et al. Whole-body magnetic resonance imaging (WB-MRI) as surveillance for subsequent malignancies in survivors of hereditary retinoblastoma: a pilot study. *Pediatr Blood Cancer*. 2014;61(8):1440-4. doi:10.1002/pbc.24835.
8. Kamihara J, Bourdeaut F, Foulkes WD, et al. Retinoblastoma and neuroblastoma predisposition and surveillance. *Clin Cancer Res*. 2017;23(13):e98-e106. doi:10.1158/1078-0432.CCR-17-0652.

# Pediatric Nasopharyngeal Carcinoma (PEDONC-13)

Guideline Page







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# Pediatric Nasopharyngeal Carcinoma – General Considerations (PEDONC-13.1)

**ONCP.NC.0013.1.A**

**v1.0.2026**

- Pediatric nasopharyngeal carcinoma (NPC) is rare in comparison to adult NPC but is responsible for up to 50% of nasopharyngeal cancers in children and has higher rates of aggressive type III EBV-associated histology than adult NPC.
- Standard upfront treatment in pediatric NPC consists of 3 to 4 cycles of neoadjuvant chemotherapy followed by definitive chemoradiotherapy. Rare individuals with lower stage disease may be treated with radiotherapy alone.

# Pediatric NPC – Imaging (PEDONC-13.2)

**ONCP.NC.0013.2.C**
**v1.0.2026**

**Pediatric NPC Medically Necessary Initial Staging Imaging:**

- 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 (avoidance of sedation should not be the sole reason)
- CT Chest with contrast (CPT® 71260) in initial staging
- Whole-body PET/CT (CPT® 78816) after histologic confirmation of NPC to evaluate for distant metastases
  - 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)

**Pediatric NPC Medically Necessary Treatment Response Imaging:**

- MRI Brain without and with contrast (CPT® 70553) **and** MRI Neck without and with contrast (CPT® 70543) for response assessment at the following time points:
  - Following completion of neoadjuvant chemotherapy
  - 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)**) at the following time points:
  - Following completion of neoadjuvant chemotherapy only if positive at initial diagnosis
  - Following completion of chemoradiotherapy
- PET 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.

**Pediatric NPC Medically Necessary Surveillance Imaging:**

- MRI Brain without and with contrast (CPT® 70553) **and** MRI Neck without and with contrast (CPT® 70543) every 3 months for 1 year, then every 6 months for 2 years after completion of all planned therapy
- CT Chest with contrast (CPT® 71260) every 3 months for 1 year, then every 6 months for 2 years after completion of all planned therapy

**Pediatric NPC Medically Necessary Suspected Recurrence Imaging:**

- 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)
- Whole-body PET/CT (CPT® 78816) or bone scan (See: **Modality General Considerations (PEDONC-1.3)**) for histologically confirmed recurrence of NPC. These studies are also medically necessary for:
  - 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

**Background and Supporting Information**

- Metastasis frequently occurs in cervical lymph nodes and retropharyngeal space. Distal metastasis usually appears in bones, lungs, mediastinum, and rarely, in the liver. In many individuals, the initial presentation is a cervical adenopathy, and diagnosis is made with a lymph node biopsy.
- Quantitative EBV DNA PCR measured at initial diagnosis can serve as an effective tumor marker if elevated at initial diagnosis.
- Skull base invasion is common in pediatric NPC and has a dramatic impact on prognosis, and is more easily recognized on MRI imaging
- PET is generally not indicated during active treatment for recurrent pediatric cancer
  - Whole-body PET/CT (CPT® 78816) or bone scan (See: **Modality General Considerations (PEDONC-1.3)**) are not indicated for routine surveillance in asymptomatic individuals

# References (PEDONC-13)

**v1.0.2026**


1. Pappo AS, Rodriguez-Galindo C, and Furman WL. Management of infrequent cancers of childhood. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th edition. Philadelphia, PA: Wolters Kluwer; 2016:1098-1123.
2. Goldberg JM, Pappo AS, and Bishop M. Rare tumors of childhood. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th edition. Philadelphia, PA: Elsevier Saunders; 2015:2123-2145.
3. Cheuk DKL, Sabin ND, Hossain M, et al. Positron emission tomography-computed tomography for staging and follow-up of pediatric nasopharyngeal carcinoma. *Eur J Nucl Med Mol Imaging*. 2012;39(7):1097-1106. doi:10.1007/s00259-012-2091-2.
4. Stambuk HE, Patel SG, Mosier KM, Wolder SL, Holodny AI. Nasopharyngeal carcinoma: recognizing the radiographic features in children. *Am J Neuroradiol*. 2005;26(6):1575-1579.
5. Sahai P, Mohanti BK, Sharma A, et al. Clinical outcome and morbidity in pediatric patients with nasopharyngeal cancer treated with chemotherapy. *Pediatr Blood Cancer*. 2017;64(2):259-266. doi:10.1002/pbc.26240.
6. PDQ® Pediatric Treatment Editorial Board. PDQ Childhood Nasopharyngeal Cancer Treatment – Health Professional Version. Bethesda, MD: National Cancer Institute. Updated 08/23/2024. Available at: https://www.cancer.gov/types/head-and-neck/hp/child/nasopharyngeal-treatment-pdq.
7. Zhang, Lu et al. Plasma Epstein-Barr viral DNA complements TNM classification of nasopharyngeal carcinoma in the era of intensity-modulated radiotherapy. *ONcotarget*. 2015;7(5):6221-6231.

# Pediatric Adrenocortical Carcinoma (PEDONC-14)

Guideline Page







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# Pediatric Adrenocortical Carcinoma – General Considerations (PEDONC-14.1)

**ONCP.AC.0014.1.C**

**v1.0.2026**

- Pediatric Adrenocortical Carcinoma (ACC) is a rare but aggressive tumor, with fewer than 25 cases diagnosed each year.
- Most individuals are diagnosed because of virilizing symptoms, Cushing syndrome, and rarely with feminization and hyperaldosteronism or detection on screening imaging recommended for specified cancer predisposition syndromes.
- 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.

# Pediatric ACC – Imaging (PEDONC-14.2)

**ONCP.AC.0014.2.C**
**v1.0.2026**

- CT Abdomen without and with contrast increases radiation exposure and should not be routinely performed in a child with an adrenal lesion as washout criteria have not been validated in children

| Indication | Medically Necessary Imaging Study |
|---|---|
| Initial staging | • CT Chest without contrast (CPT® 71250) or with (CPT® 71260) contrast<br>• CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183)<br>• Nuclear bone scan (see: **PEDONC-1.3** for coding) |
| • Solitary adrenal mass >4 cm on conventional imaging and plans for aggressive surgical resection<br>• Inconclusive findings on conventional imaging | • FDG PET/CT scan (CPT® 78815) |
| After complete resection, with no plans for chemotherapy or radiotherapy | • See surveillance below |
| Restaging, for all unresected primary or metastatic disease on chemotherapy | • Every 2 cycles (~6 weeks) during chemotherapy, and following completion of all planned chemotherapy:<br>◦ CT Chest without contrast (CPT® 71250) or with (CPT® 71260) contrast<br>◦ CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183)<br>• If positive for distant metastases at initial diagnosis:<br>◦ Nuclear bone scan (see: **PEDONC-1.3** for coding) every 2 cycles (~6 weeks) during chemotherapy and following completion of all planned chemotherapy |

| Indication | Medically Necessary Imaging Study |
|---|---|
| Response assessment at the completion of radiotherapy | • CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183) |
| Surveillance, individuals with only localized disease at diagnosis | • Every 3 months for 2 years, then every 6 months for 3 years:<br>◦ CT Abdomen with contrast (CPT®7 4160) 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:<br>◦ CT Chest without contrast (CPT® 71250) or with (CPT® 71260) contrast<br>◦ CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183) |
| Recurrence | • CT Chest without contrast (CPT® 71250) or with (CPT® 71260) contrast<br>• CT Abdomen without and with contrast (CPT® 74170) or MRI Abdomen without and with contrast (CPT® 74183) |
| Suspected bone recurrence | • Nuclear bone scan (see: **PEDONC-1.3** for coding) |

**Background and Supporting Information**

- The mainstay of treatment is surgery.
  - Chemotherapy, adrenal suppression, and radiotherapy typically follow resection.
- 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.

# References (PEDONC-14)

**v1.0.2026**


1. Waguespack SG, Huh WW, and Bauer AJ. Endocrine tumors. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th edition. Philadelphia, PA: Wolters Kluwer; 2016:919-945.
2. Michalkiewicz E, Sandrini R, Figueiredo B, et al. Clinical and outcome characteristics of children with adrenocortical tumors: a report from The International Pediatric Adrenocortical Tumor Registry. *J Clin Oncol*. 2004:22(5):838-845. doi:10.1200/JCO.2004.08.085.
3. Flynt KA, Dillman JR, Davenport MS, et al. Pediatric adrenocortical neoplasms: can imaging reliably discriminate adenomas from carcinomas? *Pediatr Radiol*. 2015;45(8):1160-1168. doi:10.1007/s00247-015-3308-x.
4. Gupta N, Rivera M, Novotny P, et al, Adrenocortical Carcinoma in Children: A Clinicopathological Analysis of 41 Patients at the Mayo Clinical from 1950 to 2017. *Horm Res Paediatr*. 2018;90:8-18. doi:10.1159/000488855.
5. PDQ® Pediatric Treatment Editorial Board. PDQ Childhood Adrenocortical Carcinoma Treatment. Bethesda, MD: National Cancer Institute. Updated 04/03/2025. Available at: https://www.cancer.gov/types/adrenocortical/hp/child-adrenocortical-treatment-pdq.
6. Wasserman JD, Novokmet A, Eichler-Jonsson C, et al. Prevalence and functional consequence of TP53 mutations in pediatric adrenocortical carcinoma: a children's oncology group study. *J Clin Oncol*. 2015:33(6):602-9. doi:10.1200/JCO.2013.52.6863.
7. Bergsland E, Rose JB, Benson III AB, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 2.2025 – May 28, 2025. Neuroendocrine tumors, available at: https://www.nccn.org/professionals/physician_gls/pdf/neuroendocrine.pdf Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Neuroendocrine and Adrenal tumors V2.2025 – May 28, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org
8. Rees MA, Morin CE, Behr GG, et. al. Imaging of pediatric adrenal tumors: A COG Diagnostic Imaging Committee/SPR Oncology Committee white paper. *Pediatr Blood Cancer*. 2022:e29973. doi:10.1002/pbc.29973. .

# Pediatric Melanoma and Other Skin Cancers (PEDONC-15)

Guideline Page







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# Pediatric Melanoma and Other Skin Cancers (PEDONC-15)

**ONCP.SK.0015.C**

**v1.0.2026**

- Pediatric melanoma staging is assigned using the American Joint Committee on Cancer (AJCC) staging for adult melanoma. Most cases of melanoma arising in children and AYAs (~75%) are localized at diagnosis, and approximately 90% of individuals with pediatric melanoma are amenable to radical excision. The clinical management of adolescents and young adults with melanoma is still challenging and evolving because it is difficult to diagnose, and there is no standard treatment.
- Non-melanoma skin cancers (mostly basal cell carcinoma and squamous cell carcinoma) are extremely rare in pediatric individuals. In many cases, predisposing factors such as prolonged immunosuppression, radiation therapy, chemotherapy, voriconazole use, or a combination of the factors are present, and established age-specific guidelines for management of these skin tumors do not exist.
- 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.





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# References (PEDONC-15)

**v1.0.2026**


1. Rodriguez-Galindo C, Furman WL, Pappo AS. Rare pediatric tumors. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th edition. Philadelphia, PA: Wolters Kluwer; 2016:946-966.
2. Goldberg JM, Pappo AS, Bishop M. Rare tumors of childhood. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th edition. Philadelphia, PA: Elsevier Saunders; 2015:2123-2145.
3. Swetter SM, Johnson D, Albertini MR, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 2.2025—January 28, 2025, Melanoma: Cutaneous, available at: https://www.nccn.org/professionals/physician_gls/pdf/cutaneous_melanoma.pdf, Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Melanoma: Cutaneous V2.2025 January 28, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org
4. Senerchia AA, Ribiero KB, Rodriguez-Galindo C. Trends in incidence of primary cutaneous malignancies in children, adolescents, and young adults: a population-based study. *Pediatr Blood Cancer*. 2014;61(2):211-216. doi:10.1002/pbc.24639.
5. Kolandijan NA, Wei C, Burke A, Bedikian AY. Malignant melanoma in teenagers and young adults. *J Pediatr Hematol Oncol*. 2014;36(7):552-558. doi:10.1097/MPH.0000000000000231.
6. Brecht IB, De Paoli A, Bisogno G, et al. Pediatric patients with cutaneous melanoma: A European study. *Pediatr Blood Cancer*. 2018;65:e26974. doi:10.1002/pbc.26974.
7. Indini A, Brecht I, Del Vecchio M, et al, Cutaneous melanoma in adolescents and young adults. *Pediatr Blood Cancer*. 2018;65:e27292. doi:10.1002/pbc.27292.
8. Kohsravi H, Schmidt B, and Huang JT, Characteristics and outcomes of nonmelanoma skin cancer (NMSC) in children and young adults. *J Am Acad Dermatol*. 2015;73:785-790. doi:10.1016/j.aad.2015.08.007. .

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

Guideline Page







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

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**Pediatric Salivary Gland Tumors**

- The majority of pediatric salivary gland tumors arise in the parotid gland. Approximately 10% to 15% of tumors arise in the submandibular, sublingual, or minor salivary glands.
- Roughly 75% of pediatric salivary gland tumors are benign, most commonly pleomorphic adenoma.
- The most common malignant tumors occurring in the salivary glands are mucoepidermoid carcinoma, adenoid cystic carcinoma, acinic cell carcinoma, undifferentiated carcinoma, and rarely adenocarcinoma.
- 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**

- Differentiated thyroid cancers (DTC): Papillary, Follicular and Hürthle Cell are the most common childhood thyroid malignancy. Standard treatment is thyroidectomy and radioactive iodine (RAI).
- 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.

# References (PEDONC-16)

**v1.0.2026**


1. Rodriguez-Galindo C, Furman WL, and Pappo AS. Rare pediatric tumors. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th edition. Philadelphia, PA: Wolters Kluwer; 2016:946-966.
2. Goldberg JM, Pappo AS, and Bishop M. Rare tumors of childhood. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th edition. Philadelphia, PA: Elsevier Saunders; 2015:2123-2145.
3. Pfister DG, Spencer S, Adkins D, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 4.2025—June 20, 2025, Head and Neck Cancers, available at: https://www.nccn.org/professionals/physician_gls/pdf/head-and-neck.pdf, referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Head and Neck Cancers V4.2025 June 20, 2025 ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org
4. Chiaravalli S, Guzzo M, Bisogno G, et al. Salivary gland carcinomas in children and adolescents: The Italian TREP project experience. *Pediatr Blood Cancer*. 2014;61(11):1961-1968.
5. Rebours C, Couloigner V, Galmiche L, et al. Pediatric salivary gland carcinomas: diagnostic and therapeutic management. *Laryngoscope*. 2017;127:140-147.
6. Zamani M, Gronhoj C, Jensen JS, et al. Survival and characteristrics of pediatric salivary gland cancer: A systematic review and meta-analysis. *Pediatr Blood Cancer*. 2019;66:e27543. doi:10.1002/pbc.27543.
7. Prasad PK, Mahajan P, Hawkins DS, Mostoufi-Moab S, Venkatramani R. Management of pediatric differentiated thyroid cancer: An overview for the pediatric oncologist. *Pediatr Blood Cancer*. 2020;67(6):e28141. doi:10.1002/pbc.28141.
8. Haddad RI, Bischoff L, Salgado SA, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 1.2025 – March 27, 2025 Thyroid carcinoma, available at: https://www.nccn.org/professionals/physician_gls/pdf/thyroid.pdf. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Thyroid carcinoma V1.2025–March 27, 2025 ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
9. Squires JH, Martinez-Rios C, Davis JC, et. al. Imaging of pediatric thyroid tumors: A COG Diagnostic Imaging Committee/SPR Oncology Committee white paper. *Pediatr Blood Cancer*. 2023;70 Suppl 4:e29957. doi:10.1002/pbc.29957.

# Pediatric Breast Masses (PEDONC-17)

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# Pediatric Breast Masses (PEDONC-17)

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- 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 indicated 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**

- Less than 1% of pediatric breast lesions are malignant, and advanced imaging is generally not recommended without histological confirmation of malignancy.
- Mammography has limited utility in pediatric breast mass evaluation due to the high mammographic breast density in this age group, and the risk of the radiation exposure outweighs the benefit of this modality. As a result, mammography is NOT recommended for evaluation of pediatric or adolescent breast masses.
  - BI-RADS classification may overstate the risk of malignancy or need for biopsy in pediatric individuals.

# References (PEDONC-17)

**v1.0.2026**


1. Rodriguez-Galindo C, Furman WL, Pappo AS. Rare pediatric tumors. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th edition. Philadelphia, PA: Wolters Kluwer; 2016:946-966.
2. Goldberg JM, Pappo AS, Bishop M. Rare Tumors of Childhood. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th edition. Philadelphia, PA: Elsevier Saunders; 2015:2123-2145.
3. Kaneda HJ, Mack J, Kasales CJ, Schetter S. Pediatric and adolescent breast masses: a review of pathophysiology, imaging, diagnosis, and treatment. *AJR Am J Roentgenol*. 2013;200(2):W204-W212. doi:10.2214/AJR.12.9560.
4. Koning JL, Davenport KP, Poole PS, Kruk PG, Grabowski JE. Breast Imaging-Reporting and Data System (BI-RADS) classification in 51 excised palpable pediatric breast masses. *J Pediatr Surg*. 2015;50(10):1746-1750. doi:10.1016/j.jpedsurg.2015.02.062.
5. Siegel MJ, Chung EM. Breast. In: Seigel MJ, ed. Pediatric Sonography. 5th ed. Philadelphia, PA: Wolters Kluwer; 2019:196-210.
6. Siegel MJ, Chung EM. Breast masses in children and adolescents. *Applied Radiology*. 2017;46(9):12-17.
7. Johnson RH, Anders CK, Litton JK, Ruddy KJ, Bleyer A. Breast cancer in adolescents and young adults. *Pediatr Blood Cancer*. 2018;65:e27397. doi:10.1002/pmb.27397.
8. Sanders LM, Sharma P, El Madany M, et al. Clinical breast concerns in low-risk pediatric patients: practice review with proposed recommendations. *Pediatr Radiol*. 2018;48:186-195. doi:10.1007/s00247-017-4007-6.






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# Histiocytic Disorders (PEDONC-18)

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# Histiocytic Disorders – General Considerations (PEDONC-18.1)

**ONCP.HC.0018.1.A**

**v1.0.2026**

- The majority of histiocytic disorders occurring in the pediatric population are either Langerhans Cell Histiocytosis (LCH) or Hemophagocytic Lymphohistiocytosis (HLH).
- 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.





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# Langerhans Cell Histiocytosis (LCH) (PEDONC-18.2)

**ONCP.HC.0018.2.C**
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**LCH Medically Necessary Initial imaging:**

- Whole-body PET/CT (CPT® 78816)
  - Whole-body Tc-99m bone scan (CPT® 78306) in lieu of PET if PET is unavailable
- CT Chest with contrast (CPT® 71260) or high-resolution CT Chest without contrast (CPT® 71250) in addition to PET/CT for 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
- MRI Brain without and with contrast (CPT® 70553) for 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
- CTA/MRA Head (CPT® 70496/CPT® 70544) for operative planning or image guidance
- CT Abdomen and/or Pelvis with contrast (CPT® 74177, 74160, or 72193) for 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
- MRI Abdomen without and with contrast (CPT® 74183) for ANY of the following:
  - Elevated liver function tests (usually >5X upper limit of normal)
  - Abnormalities seen on abdominal ultrasound or CT
- MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) for ANY of the following **if PET/CT has not been performed**:
  - Vertebral lesions seen on skeletal survey
  - Clinical symptoms (including back pain) suggesting spinal involvement and negative skeletal survey

**LCH Medically Necessary Treatment Response Imaging:**

- Both PET/CT and CT with contrast **and** MRI without and with contrast **only** for simultaneous treatment response evaluation with specific documentation showing that both are necessary (i.e., not for purpose of acquiring a PET/MRI)
- CT and/or MRI and/or PET/CT (if modality showed disease at initial diagnosis):
  - After 2-3 cycles of treatment
  - At completion of therapy (approximately 12 weeks) for individuals with persistent disease on cycle 2-3 imaging
  - After surgical curettage (CT or MRI of involved area, not repeat PET)
  - After radiation therapy
- Following the initial phase, treatment response evaluation of **involved sites** with CT with contrast or MRI without and with contrast, every 3 months while receiving active treatment and at completion of therapy.
  - PET/CTfor 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

**LCH Medically Necessary Surveillance Imaging:**

- Surveillance imaging is determined by areas of disease involvement.
  - Bone involvement
    - Single site bone disease
      - Every 3 months for 1 year after completion of therapy:
        - PET/CT (CPT® 78815 or CPT® 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 CPT® 78816) OR
      - CT or MRI contrast as requested of involved bony areas every 6 months for 2 years
    - Skull or craniofacial (including jaw) bone involvement should be imaged according to CNS involvement section below.

  Patients with lung, CNS, and/or liver involvement are considered to have high-risk disease and warrant site-specific diagnostic imaging in addition to PET/CT as outlined below:
- Pulmonary involvement
  - CT Chest with (CPT® 71260) **or** CT Chest without contrast (CPT® 71250) every 6 months for the first 2 years post completion of therapy 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 CPT® 78816) every 6 months for 2 years, then no more than annually
- CNS involvement
  - PET/CT (CPT® 78815 or CPT® 78816) every 6 months for 2 years, then no more than annually

    AND
  - MRI Brain without and with contrast (CPT® 70553) at 6 weeks, and then every 6 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
    - If residual measurable intracranial lesions are present at 6 months, imaging repeated every 3 months until negative or unchanged on two consecutive studies, at which time the schedule in the previous bullet should begin
  - MRI Brain without and with contrast (CPT® 70553) for documented hypothalamic-pituitary dysfunction at 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
- Liver involvement
  - For individuals with a history of liver involvement, ONE of the following every 6 months for 2 years after completion of all therapy:
    - Ultrasound Abdomen (CPT® 76700)
    - CT Abdomen with contrast (CPT® 74160)
    - MRI Abdomen without and with contrast (CPT® 74183)

    AND
  - PET/CT (CPT® 78815 or CPT® 78816) every 6 months for 2 years, then no more than annually
- Suspected recurrence or inconclusive findings on any surveillance imaging:
  - All imaging studies supported for initial workup, including whole-body PET/CT (CPT® 78816)

**Background and Supporting Information**

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

- LCH includes a heterogeneous group of disorders formerly known by other names, including histiocytosis X, eosinophilic granuloma, Letterer-Siwe Disease, Hand-Schuller-Christian Disease, and diffuse reticuloendotheliosis. LCH has a widely variable clinical presentation, ranging from single indolent lesions to disseminated multisystem disease.
- Most common sites of involvement are skin, bones, liver, lung, and pituitary, though other sites are possible.
- Individuals with localized or single site disease treated only with local therapies or observed should be imaged according to surveillance guidelines
- Individuals receiving systemic therapy will usually undergo treatment for ~12 months. Treatment response is assessed using any modalities showing disease at initial diagnosis after ~6 weeks of treatment.
- Skull or craniofacial (including jaw) bone involvement at diagnosis are at higher risk for CNS recurrence
- CNS LCH has a particularly high rate of refractory and recurrent disease, and requires longer imaging surveillance
- Liver involvement
  - Persistent liver involvement is rare, and imaging after completion of LCH therapy will be highly individualized depending on degree of liver dysfunction and plans for supportive therapy or liver transplant

# Hemophagocytic Lymphohistiocytosis (HLH) (PEDONC-18.3)

**ONCP.HC.0018.3.A**

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- As imaging for this condition is usually done on an urgent basis, ANY or ALL of the following are medically necessary for the initial evaluation of HLH include:
  - Ultrasound Abdomen (CPT® 76700)
  - CT Abdomen and/or Pelvis (contrast as requested)
  - 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
- 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 conventional imaging has been completed and is unrevealing
  - 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

**Background and Supporting Information**

- There are no standard imaging studies required for the diagnosis and initial evaluation of HLH. Most cases are diagnosed with a combination of physical findings, laboratory testing, and bone marrow evaluation. Advanced imaging studies may be necessary to assess organ dysfunction as HLH commonly affects the liver, spleen, and bone marrow, and less commonly the kidneys, lungs, and brain
- It is NOT required to perform ultrasound or plain film in a stepwise fashion if CT or MRI is planned as individuals with HLH can deteriorate rapidly
- There is no established standard role for PET in the diagnosis or treatment response evaluation of HLH
  - Secondary HLH is very difficult to treat if the primary cause is not concurrently treated

# Non-Langerhans Cell Histiocytoses (PEDONC-18.4)

**ONCP.HC.0018.4.C**
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**Juvenile Xanthogranuloma (JXG) medically necessary imaging:**

- Skin and/or cervical nodules:
  - CT with contrast of involved nodal areas
- 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) may be approved as part of operative planning or image guidance
- There is no established role for PET in the diagnosis or treatment of JXG

**Rosai-Dorfman Disease (RDD) medically necessary imaging:**

- RDD Initial Imaging Studies:
  - 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 and 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 histological confirmation of diagnosis
- RDD Treatment Response:
  - 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 with other modalities that revealed disease at presentation for subsequent restaging at completion of chemotherapy and/or radiation and/or after surgical resection.
- RDD Surveillance Imaging:
  - 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 medically necessary for routine surveillance of RDD but is 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 medically necessary for routine surveillance of RDD but is medically necessary if conventional imaging is inconclusive for suspected recurrence.

**Erdheim-Chester Disease (ECD) medically necessary imaging:**

- ECD Initial imaging studies:
  - PET/CT Whole Body (CPT® 78816)
    - Nuclear bone scan (See: **PEDONC-1.3: Modality General Considerations**) may be approved 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/CPT® 70544) for operative planning or image guidance
  - CT Neck (CPT® 70491), Chest (CPT® 71260) **and/or** Abdomen and 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
  - 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
- ECD Treatment Response:
  - 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 when conventional imaging studies are inconclusive and acute treatment decisions will be made based on PET results.
- ECD Surveillance Imaging:
  - 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 if conventional imaging is inconclusive for suspected recurrence.

**Background and Supporting Information**

- Non-Langerhans Cell histiocytoses includes diagnoses such as juvenile xanthogranuloma (JXG), sinus histiocytosis with lymphadenopathy (Rosai-Dorfman disease, RDD), and Erdheim-Chester disease (ECD)
- In general, these are localized cutaneous or nodal disease without need for regular advanced imaging, but important exceptions are listed in this section.
- Juvenile Xanthogranuloma (JXG):
  - Generally involves only skin or cervical nodes, and involutes spontaneously
- Rosai-Dorfman Disease (RDD):
  - Characterized by bulky adenopathy (usually cervical) with frequent systemic involvement
  - There is no established role for routine surveillance imaging of asymptomatic individuals after treatment for RDD
- Erdheim-Chester Disease (ECD):
  - An aggressive histiocytic disorder with overall poor prognosis that is characterized by long bone involvement with frequent spread to multiple organs
  - Most individuals will receive systemic therapy
  - Once PET/CT shows no remaining FDG-avid lesions, additional PET imaging is not indicated unless conventional imaging studies are inconclusive and acute treatment decisions will be made based on PET results.

# References (PEDONC-18)

**v1.0.2026**


1. McClain KL, Allen CE, Hicks MJ. Histiocytic diseases. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:617-627.
2. Degar BA, Fleming MD, Rollins BJ, et al. Histiocytoses. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th ed. Philadelphia, PA: Elsevier Saunders; 2015:2100-2122.
3. Haupt R, Minkov M, Astigarraga I, et al. Langerhans cell histiocytosis (LCH): guidelines for diagnosis, clinical work-up, and treatment for patients till the age of 18 years. *Pediatr Blood Cancer*. 2013;60(2):175-184. doi:10.1002/pbc.24367.
4. Chandrakasan S, Filipovich AH. Hemophagocytic lymphohistiocytosis: advances in pathophysiology, diagnosis, and treatment. *J Pediatr*. 2013;163(5):1253-1259. doi:10.1016/j.jpeds.2013.06.053.
5. Phillips M, Allen C, Gerson P, et al. Comparison of FDG-PET scans to conventional radiography and bone scans in management of Langerhans cell histiocytosis. *Pediatr Blood Cancer*. 2009;52(1):97-101. doi:10.1002/pbc.21782.
6. Chellapandian D, Shaiki F, van den Bos C, et al. Management and outcome of patients with langerhans cell histiocytosis and single-bone CNS-risk lesions: a multi-institutional retrospective study. *Pediatr Blood Cancer*. 2015;62(12):2162-2166. doi:10.1002/pbc.25645.
7. Allen CE, Ladisch S, McClain KL. How I treat Langerhans cell histiocytosis. *Blood*. 2015;126(1):26-35. doi:10.1182/blood-2014-12-569301.
8. Schram AM, Berliner N. How I treat Hemophagocytic lymphohistiocytosis in the adult patient. *Blood*. 2015;125(19):2908-2914. doi:10.1182/blood-2015-01-551622.
9. Weitzman S, Jaffe R. Uncommon histiocytic disorders: the non-Langerhans cell histiocytoses. *Pediatr Blood Cancer*. 2005;45(3):256-264. doi:10.1002/pbc.20246.
10. Diamond EL, Dagna L, Hyman DM, et al. Consensus guidelines for the diagnosis and clinical management of Erdheim-Chester disease. *Blood*. 2014;124(4):483-492. doi:10.1182/blood-2014-03-561381.
11. Go RS, Jacobsen E, Baiocchi R, et. al. National Comprehensive Cancer Network (NCCN) Guidelines Version 1.2025 – June 20, 2025. Histiocytic Neoplasms, available at: https://www.nccn.org/professionals/physician_gls/pdf/histiocytic_neoplasms.pdf. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Histiocytic Neoplasms V1.2025 –June 20, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
12. Salama HA, Jazieh AR, Alhejazi AY, et. al. Highlights of the management of adult histiocytic disorders: Langerhans cell histiocytosis, Erdheim-Chester disease, Rosai-Dorfman disease, and hemophagocytic lymphohistiocytosis. *Clinical Lymphoma, Myeloma & Leukemia*. 2020;21(1):e66-75. doi:10.1016/j.dml.2020.08.007.
13. Goyal G, Young JR, Koster MJ, et. al. The Mayo Clinical Histiocytosis Working Group consensus statement for the diagnosis and evaluation of adult patients with histiocytic neoplasms: Erdheim-Chester disease, Langerhans cell histiocytosis, and Rosai-Dorfman disease. *Mayo Clinical Proc*. 2019;94(10):2054-2071. doi:10.1016/j.mayocp.2019.02.023.
14. Allen CE, Merad M, McClain KL. Langerhans-Cell histiocytosis. *N Engl J Med*. 2018;379:856-68. doi:10.1056/NEJMra1607548.
15. Abla O, Jacobsen E, Picarsic J, et. al. consensus recommendations for the diagnosis and clinical management of Rosai-Dorfman-Destombes disease. *Blood*. 2018;131(26):2877-2890. doi:10.1182/blood-2018-03-839753.
16. Ferrell J, Sharp S, Kumar A, Jordan M, Picarsic J, Nelson A. Discrepancies between F-18-FDG PET/CT findings and conventional imaging in Langerhans cell histiocytosis. *Pediatr Blood Cancer*. 2021;68(4):e28891. doi:10.1002/pbc.28891.
17. Campbell KM, Shulman DS, Grier HE, DuBois SG. Role of bone marrow biopsy for staging new patients with Ewing sarcoma: A systematic review. *Pediatr Blood Cancer*. 2021 Feb;68(2):e28807. doi: 10.1002/pbc.28807. Epub 2020 Nov 21. PMID: 33219750.

# Long Term Pediatric Cancer Survivors (PEDONC-19)

Guideline Page







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

**ONCP.CS.0019.1.A**

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- 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 indicated 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.

# Cardiotoxicity and Echocardiography (PEDONC-19.2)

**ONCP.CS.0019.2.C**

**v1.0.2026**

- Screening echocardiography (CPT® 93306, CPT® 93307, or CPT® 93308) 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 |
| | 0-249 mg/m2 | 15+ Gy | Every 2 years |

| MEDICALLY NECESSARY SCREENING ECHOCARDIOGRAM INDICATIONS | | | |
|---|---|---|---|
| **Age at Time of Exposure** | **Cumulative Doxorubicin Equivalent Dose** | **Cumulative Radiation Dose to Cardiac Muscle** | **Echocardiogram Frequency** |
| | | 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 medically necessary as a screening study for anthracyclines cardiotoxicity in the absence of coronary artery disease symptoms. See: **Stress Testing with Imaging – Indications (CD-1.4)** in the Cardiac Imaging Guidelines.
- Female cancer survivors who are pregnant or planning to become pregnant:
  - Echocardiogram is 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 cardiotoxic anthracycline chemotherapy agents is common in pediatric oncology due to the high success rate of this drug class in the treatment of pediatric cancers.
- Cardiac risk is assessed based on the age of the individual at the time of treatment initiation, the cumulative drug exposure expressed as doxorubicin equivalent mg/m$^2$, and the presence or absence of radiotherapy exposure to cardiac muscle.

# Second Malignant Neoplasms (SMN) (PEDONC-19.3)

**ONCP.CS.0019.3.C**
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### SMN—Breast Cancer

Clinical breast exam every 6 months supplemented with the following medically necessary imaging:

- 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 supported prior to age 25) for 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)

### SMN – CNS Tumors

- MRI Brain without and with contrast (CPT® 70553) is medically necessary every 2 years after completion of radiotherapy for individuals with NF1 or NF2.
- MRI Brain without and with contrast (CPT® 70553) is medically necessary for any individual with history of brain radiotherapy and new neurologic symptoms, including simple headache.
- MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), and Lumbar Spine (CPT® 72158) without and with contrast are medically necessary for any individual with history of spine radiotherapy and new neurologic symptoms including change in quality of pain.
  - MRI Spine with contrast only (Cervical-CPT® 72142, Thoracic-CPT® 72147, Lumbar-CPT® 72149)is medically necessary if being performed immediately following a contrast-enhanced MRI Brain.
- MRI Brain without and with contrast (CPT® 70553) is medically necessary annually for individuals with history of brain radiotherapy and persistent neurologic symptoms.

- MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), and Lumbar Spine (CPT® 72158) without and with contrast is medically necessary annually for individuals with history of spine radiotherapy and persistent neurologic symptoms.
  - MRI Spine with contrast only (Cervical-CPT® 72142, Thoracic-CPT® 72147, Lumbar-CPT® 72149) is medically necessary if being performed immediately following a contrast-enhanced MRI Brain.

**SMN—Colorectal Cancer**

- Advanced imaging is not medically necessary. See: *Background and Supporting Information* section for recommended surveillance.

**Background and Supporting Information**

- SMN—Breast Cancer:
  - Clinical breast exam every 6 months supplemented with advanced imaging
- SMN-CNS Tumors:
  - These are associated with radiation exposure to the brain and with neurofibromatosis
  - Routine surveillance of asymptomatic individuals with normal neurologic exams is not supported by evidence, with the exception of NF1 and NF2 listed above
- SMN—Colorectal Cancer:
  - Colonoscopy is recommended every 5 years beginning at age 30 or 5 years after radiation exposure (whichever is later) for individuals with ≥30 Gy radiation exposure to the following fields:
    - Thoracic, Lumbar, Sacral, or Whole Spine
    - Abdomen
    - Pelvis
    - Total body irradiation (TBI)
- Colonoscopy is also recommended every 5 years beginning at age 30 or 5 years after radiation exposure (whichever is later) for 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
- 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

# Osteonecrosis in Long Term Cancer Survivors (PEDONC-19.4)

**ONCP.CS.0019.4.C**
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- Plain films of symptomatic areas are indicated prior to advanced imaging.
- DEXA or Quantitative CT screening is medically necessary **only** for those with symptoms to suggest bone density issues.
  - DEXA or Quantitative CT screening is not medically necessary until age 18 unless a surgery, core decompression, or initiation of osteoporosis drugs will be planned based on the imaging results.
- Serial advanced imaging is **only** medically necessary in osteonecrosis with specific documentation regarding how the advanced imaging will change current management.
  - MRI without contrast of the affected area(s) when advanced imaging is medically necessary for acute management decisions
  - For known osteonecrosis with articular collapse on other imaging, CT without contrast of area of interest is medically necessary for surgical planning.
- See: **Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)** for information on imaging osteonecrosis in ALL individuals during active treatment.

**Background and Supporting Information**

- Osteonecrosis is associated with corticosteroid, chemotherapy, and radiation exposure during treatment for ALL, NHL, and allogeneic HSCT in pediatrics. Osteonecrosis occurs primarily in hips, knees, and ankles and is frequently multifocal.
- Osteoradionecrosis of the jaw can occur in individuals receiving radiotherapy to the mandible or maxilla; those receiving ≥ 40 Gy are at highest risk. Although unusual, it can also occur in any bone without symptoms. It is rare in other disease types.
- Routine bone density screening using DEXA or Quantitative CT screening has not been well normalized in the pediatric population
- Surveillance imaging of asymptomatic individuals to detect osteonecrosis has not been shown to impact 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 necessary

# CNS vascular changes in pediatric cancer survivors following CNS radiation (PEDONC-19.5)

**ONCP.CS.0019.5.C**

**v1.0.2026**

- MRI Brain without and with contrast (CPT® 70553) and MRA Head (CPT® 70544, 70545, or 70546) are medically necessary for 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) are medically necessary annually for 10 years post treatment in 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 >50 Gy
  - Radiation field involving Circle of Willis

### Background and Supporting Information

Children receiving cranial radiation are at increased risk of cerebrovascular complications, including: hemorrhagic and ischemic stroke, moyamoya, occlusive vasculopathy, and cavernomas. These complications may occur months to years after radiation exposure.





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# References (PEDONC-19)

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1. Landier W, Armenian SH, Meadows AT, et al. Late effects of childhood cancer and its treatment. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th ed. Philadelphia, PA: Wolters Kluwer; 2016:1173-1196.
2. Vrooman L, Diller L, Kenney LB. Childhood cancer survivorship. In: Orkin SH, Fisher DE, Ginsburg D, Look AT, Lux SE, Nathan DG, eds. Nathan and Oski's Hematology and Oncology of Infancy and Childhood. 8th ed. Philadelphia, PA: Elsevier Saunders; 2015:2397-2434.
3. Children's Oncology Group. Long-term follow up guidelines for survivors of childhood, adolescent and young adult cancers, version 5.0. Monrovia, CA: Children's Oncology Group; October 2018; Available on-line: www.survivorshipguidelines.org
4. Ryerson AB, Border WL, Wasilewski-Masker K, et al. Assessing anthracycline-treated childhood cancer survivors with advanced stress echocardiography. *Pediatr Blood Cancer.* 2015;62(3):502-508. doi:10.1002/pbc.25328.
5. Tieu MT, Cigsar C, Ahmed S, et al. Breast cancer detection among young survivors of pediatric Hodgkin lymphoma with screening magnetic resonance imaging. *Cancer*. 2014;120(16):2507-2513. doi:10.1002/cncr.28747.
6. Lange JM, Takashima JR, Peterson SM, Kalapurakal JA, Green DM, Breslow NE. Breast cancer in female survivors of Wilms tumor: a report from the national Wilms tumor late effects study. *Cancer.* 2014;120(23):3722-3730. doi:10.1002/cncr.28908.
7. Bevers RB, Niell BL, Aminololama-Shakeri S, et al. National Comprehensive Cancer Network (NCCN) Guidelines Version 2.2025—March 28, 2025. Breast Cancer Screening and Diagnosis, available at: https://www.nccn.org/professionals/physician_gls/pdf/breast-screening.pdf, Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines™) for Breast Cancer Screening and Diagnosis V2.2025 March 28, 2025. ©2025 National Comprehensive Cancer Network, Inc. All rights reserved. The NCCN Guidelines™ and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines™, go online to NCCN.org.
8. Recht M, Mostoufi-Moab S, McFadden J, et al. Bone health in pediatric hematology-oncology. In: American Society of Pediatric Hematology/Oncology Annual Meeting. Chicago; 2014.
9. Bhatia S, Armenian SH, Armstrong GT, et al. Collaborative research in childhood cancer survivorship: the current landscape. *J Clin Oncol*. 2015;33(27):3055-3064. doi:10.1200/JCO.2014.59.8052.
10. Chavhan GB, Babyn PS, Nathan PC, Kaste SC. Imaging of acute and subacute toxicities of cancer therapy in children. *Pediatr Radiol*. 2016;46(1):9-20. doi:10.1007/s00247-015-3454-1.
11. Karol SE, Mattano LA, Yang W, et al. Genetic risk factors for the development of osteonecrosis in children under age 10 treated for acute lymphoblastic leukemia. *Blood Journal.* 2016;127(5):558-564. doi:10.1182/blood-2015-10-673848.
12. Marcucci G, Beltrami G, Tamburini A, et al. Bone health in childhood cancer: review of the literature and recommendations for the management of bone health in childhood cancer survivors. *Ann Oncol.* 2019;30:908-920. doi:10.1093/annonc/mdz120.
13. Sanders LM, Sharma S, El Madany M, King AB, Goodman KS, Sanders AE. Clinical breast concerns in low-risk pediatric patients: practice review with proposed recommendations. *Pediatr Radiol*. 2018(48):186–195. doi:10.1007/s00247-017-4007-6.
14. Murphy ES, Xie H, Merchant TE, Yu JS, Chao ST, Suh JH. Review of cranial radiotherapy-induced vasculopathy. *J Neurooncol.* 2015;122(3):421-9. doi:10.1007/s11060-015-1732-2. Epub 2015 Feb 12.
15. Baumgartner JE, Ater JL, Ha CS, et. al. Pathologically proven cavernous angiomas of the brain following radiation therapy for pediatric brain tumors. *Pediatr Neurosurg*. 2003;39(4):201-7. doi:10.1159/000072472.
16. Di Giannatale A, Morana G, Rossi A, et. al. Natural history of cavernous malformations in children with brain tumors treated with radiotherapy and chemotherapy. *J Neurooncol*. 2014;117(2):311-20. doi:10.1007/s11060-014-1390-9. Epub 2014 Feb 11.

17. Bowers DC, Liu Y, Leisenring W, et. al. Late-occurring stroke among long-term survivors of childhood leukemia and brain tumors: a report from the Childhood Cancer Survivor Study. *J Clin Oncol*. 2006;24:5277–5282.
18. Morris B, Partap S, Yeom K, Gibbs IC, Fisher PG, King AA. Cerebrovascular disease in childhood cancer survivors: a Children's Oncology Group report. *Neurology*. 2009;73:1906–1913
19. Laitt RD, Chambers EJ, Goddard PR, Wakeley CJ, Duncan AW, Foreman NK. Magnetic resonance imaging and magnetic resonance angiography in long term survivors of acute lymphoblastic leukemia treated with cranial irradiation. *Cancer.* 1995;76:1846–1852
20. Gastelum E, Sear K, Hills N, et. al. Rates and characteristics of radiographically detected intracerebral cavernous malformations after cranial radiation therapy in pediatric cancer patients. *J Child Neurol*. 2015;30(7):842–849. doi:10.1177/0883073814544364.
21. ACR-ASNR-SPR Practice Parameter for the Performance and Interpretation of Cervicocerebral Computed Tomography (CTA). (Resolution 42). 2020.
22. Rao SS, El Abiad JM, Puvanesarajah V, Levin AS, Jones LC, Morris CD. Osteonecrosis in pediatric cancer survivors: epidemiology, risk factors, and treatment. *Surgical Oncology*. 2019;28:214-221. doi:10.1016/j.suronc.2019.02.001.
23. Nordstrom M, Felton E, Sear K, et. al. Large vessel arteriopathy after cranial radiation therapy in pediatric brain tumor survivors. *J Child Neurol.* 2018;33(5):359-366. doi:10.1177/0883073818756729.
24. Ha AS, Chang EY, Bartolotta RJ, et. al. Expert Panel on Musculoskeletal Imaging. ACR Appropriateness Criteria® Osteonecrosis. *Am Coll Radiol* (ACR); Date of Origin: 2016. Revised: 2022. https://acsearch.acr.org/docs/69402/Narrative/.

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

Guideline Page







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# General Considerations (PEDONC-20.0)

**ONCP.HT.0020.0.A**
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**Transplant Types:**

- 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 results in a much greater degree of immunosuppression than autologous HSCT because of the need to allow the new immune system to chimerize with the recipient's body. Immune reconstitution commonly takes more than a year for individuals who receive allogeneic HSCT, and individuals remain at high risk for invasive infections until that has occurred. In addition, patients may require prolonged immunosuppression for prevention and management of graft-vs-host disease.
- Recipients of autologous transplant are deeply immunosuppressed until complete count recovery, which may take several months. Graft-vs-host disease is not a consideration in auto transplants because the host is also the donor.

# Pre-Transplant Imaging (PEDONC-20.1)

**ONCP.HT.0020.1.A**
**v1.0.2026**

- Disease status assessment may be approved 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)<br>◦ 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.<br>◦ CT Pelvis for pelvic signs and symptoms with CPT® 74177 or CPT® 72193 if abdominal imaging has already been performed.<br>• CT Sinus (CPT® 70486) is not medically necessary for screening prior to HSCT, but is medically necessary for signs or symptoms of infectious or disease involvement of the sinuses |
| Individuals at risk of developing VOD, for pre-transplant baseline | • Abdominal ultrasound +/- Doppler (CPT® 76700 or 76705 +/- 93975) |
| Individuals 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 | • Nuclear renal imaging (CPT® 78700, CPT® 78701, CPT® 78707, CPT® 78708, CPT® 78709) |





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**Background and Supporting Information**

- CT Sinus screening is not routinely recommended as two studies showed no utility and no effect on surgical consults.
- Changes from baseline abdominal ultrasound with regard to veno-occlusive disease (VOD) are more predictive than absolute measurements and thus baseline imaging is supported.
- Pre-transplant liver and cardiac iron levels are associated with adverse transplant outcomes and a baseline is supported for further management.

# Post-Transplant Imaging (PEDONC-20.2)

**ONCP.HT.0020.2.A**
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**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 | • CT Chest without or with contrast (CPT® 71250 or CPT® 71260)<br>◦ For PET/CT requests to clarify infection vs malignancy, see disease-specific guidelines for equivocal conventional imaging |
| Suspected impending lung necrosis on other imaging | • MRI Chest without and with contrast (CPT® 71552) |
| • EITHER of the following:<br>◦ Suspected graft vs. host disease (GVHD) of chest<br>◦ Bronchiolitis Obliterans (BOOP/BOS) | • High-resolution CT Chest without contrast (CPT® 71250) |
| • ANY of the following:<br>◦ Suspected intra-abdominal and or pelvic infection (including cystitis or typhlitis)<br>◦ Suspected small bowel GVHD | • Abdominal Ultrasound (CPT® 76700 or CPT® 76705) +/- Pelvic Ultrasound (CPT® 76856 or CPT® 76857)<br>◦ CT Abdomen and Pelvis with contrast (CPT® 74177) for unclear findings on ultrasound |
| High clinical suspicion for intra-abdominal fungal infection | • CT Abdomen and Pelvis with contrast (CPT® 74177) |
| Suspected hepatic veno-occlusive disease (VOD) | • 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 | • MRI Brain without contrast or MRI Brain without and with contrast (CPT® 70551 or CPT® 70553)<br>◦ CT Head without contrast (CPT® 70450) in the emergent setting or for suspected acute hemorrhage<br>▪ Acute CT should not preclude subsequent MRI imaging<br>◦ 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 | • 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) | • PET/CT skull to thigh (CPT® 78815) if site-specific conventional imaging, microbiologic serologic studies, echocardiogram, urinalysis, and urine culture are all non-diagnostic |

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

| Indication | Medically Necessary Imaging |
|---|---|
| CNS complications | Same imaging as for early post-transplant |
| Chest complications | Same imaging as for early post-transplant |
| Hepatic VOD | Same imaging as for early post-transplant |
| Chronic GI GVHD | • CT Abdomen and Pelvis (CPT® 74177)<br>◦ MR Enterography (CPT® 74183 and CPT® 72197) for suspected small bowel GVHD |
| Suspected renal tubular dysfunction | • Nuclear renal imaging (CPT® 78700, CPT® 78701, CPT® 78707, CPT® 78708, CPT® 78709) |

| Indication | Medically Necessary Imaging |
|---|---|
| Suspected poor bone mineral density or osteonecrosis | See: **Osteonecrosis in Long Term Cancer Survivors (PEDONC-19.4)** |
| Suspected PTLD | See: **Post-Transplant Lymphoproliferative Disorder (PTLD) (PEDONC 5.3)** |





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# References (PEDONC-20)

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1. Chan SS, Coblentz A, Bhatia A, et. al. Imaging of pediatric hematopoietic stem cell transplant recipients: A COG Diagnostic Imaging Committee/SPR Oncology Committee white paper. *Pediatr Blood Cancer*. 2023;70 Suppl 4:e30013. doi: 10.1002/pbc.30013.
2. Kaste SC, Kaufman RA, Sunkara A, et. al. Routine pre- and post-hematopoietic stem cell transplant computed tomography of the abdomen for detecting invasive fungal infection has limited value. *Biol Blood Marrow Transplant*. 2015;21(6):1132-5. doi: 10.1016/j.bbmt.2015.02.023.
3. PDQ® Pediatric Treatment Editorial Board. PDQ Pediatric Hematopoietic Stem Cell Transplantation and Cellular Therapy for Cancer. Bethesda, MD: National Cancer Institute. Updated 06/13/2024. Available at: https://www.cancer.gov/types/childhood-cancers/hp-stem-cell-transplant.