# Childhood Brain and Spinal Cord Tumors Summary Index (PDQ®)–Health Professional Version

## General Information About Childhood Brain and Spinal Cord Tumors

Primary brain tumors are a diverse group of diseases that together constitute the most common solid tumor of childhood. The Central Brain Tumor Registry of the United States (CBTRUS) estimates that approximately 4,300 U.S. children are diagnosed each year.[1]

Brain tumors are classified by histology, but tumor location and extent of spread are also important factors that affect treatment and prognosis. Immunohistochemical analysis, cytogenetic and molecular genetic findings, and measures of proliferative activity are increasingly used in tumor diagnosis and classification.[2]

***References***

1. Ostrom QT, Gittleman H, Farah P, et al.: CBTRUS statistical report: Primary brain and central nervous system tumors diagnosed in the United States in 2006-2010. Neuro Oncol 15 (Suppl 2): ii1-56, 2013. [PUBMED Abstract]
2. Louis DN, Perry A, Reifenberger G, et al.: The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary. Acta Neuropathol 131 (6): 803-20, 2016. [PUBMED Abstract]

## Type of Childhood Brain and Spinal Cord Tumors

For information about the type of childhood brain and spinal cord tumor and its related PDQ summary, see the table below. If a tumor type is not listed, a corresponding PDQ treatment summary is not available.

### CNS Tumor Type, Pathological Subtype, and Its Related PDQ Treatment Summary

| Tumor Type (Based on the 2021 WHO Classification)[a] | Pathological Subtype (Based on the 2021 WHO Classification)[a] | Related PDQ Treatment Summary |
|---|---|---|
| **Pediatric-type diffuse high-grade gliomas** | Diffuse pediatric-type high-grade glioma, H3-wild type and *IDH*-wild type | Childhood Astrocytomas, Other Gliomas, and Glioneuronal/Neuronal Tumors Treatment |
| | Diffuse midline glioma, H3 K27-altered | |
| | Diffuse hemispheric glioma, H3 G34-mutant | |
| | Infant-type hemispheric glioma | |
| **Circumscribed astrocytic gliomas** | Pilocytic astrocytoma | Childhood Astrocytomas, Other Gliomas, and Glioneuronal/Neuronal Tumors Treatment |
| | High-grade astrocytoma with piloid features | |
| | Subependymal giant cell astrocytoma | |
| | Pleomorphic xanthoastrocytoma | |
| | Choroid glioma | |
| | Astroblastoma, *MN1*-altered | |
| **Ependymal tumors** | Subependymoma | Childhood Ependymoma Treatment |
| | Myxopapillary ependymoma | |
| | Supratentorial ependymoma, NOS | |

CNS = central nervous system; NEC = not elsewhere classified; NOS = not otherwise specified; WHO = World Health Organization.

[a]WHO classification adapted from Louis et al.[1]

| Tumor Type (Based on the 2021 WHO Classification)[a] | Pathological Subtype (Based on the 2021 WHO Classification)[a] | Related PDQ Treatment Summary |
|---|---|---|
| | Supratentorial ependymoma, *ZFTA* fusion–positive | |
| | Supratentorial ependymoma, *YAP1* fusion–positive | |
| | Posterior fossa ependymoma, NOS | |
| | Posterior fossa group A (PFA) ependymoma | |
| | Posterior fossa group B (PFB) ependymoma | |
| | Spinal ependymoma, NOS | |
| | Spinal ependymoma, *MYCN*-amplified | |
| **Pediatric-type diffuse low-grade glioma** | Diffuse astrocytoma, *MYB*- or *MYBL1*-altered | Childhood Astrocytomas, Other Gliomas, and Glioneuronal/Neuronal Tumors Treatment |
| | Angiocentric glioma | |
| | Polymorphous low-grade neuroepithelial tumor of the young | |
| | Diffuse low-grade glioma, MAPK pathway-altered | |
| **Glioneuronal and neuronal tumors** | Dysembryoplastic neuroepithelial tumor | Childhood Astrocytomas, Other Gliomas, and Glioneuronal/Neuronal Tumors Treatment |
| | Ganglioglioma | |

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[a]WHO classification adapted from Louis et al.[1]

| Tumor Type (Based on the 2021 WHO Classification)[a] | Pathological Subtype (Based on the 2021 WHO Classification)[a] | Related PDQ Treatment Summary |
|---|---|---|
| | Gangliocytoma | |
| | Desmoplastic infantile ganglioglioma | |
| | Desmoplastic infantile astrocytoma | |
| | Diffuse glioneuronal tumor with oligodendroglioma-like features and nuclear clusters (provisional entity) | |
| | Papillary glioneuronal tumor | |
| | Rosette-forming glioneuronal tumor | |
| | Myxoid glioneuronal tumor | |
| | Diffuse leptomeningeal glioneuronal tumor | |
| | Multinodular and vacuolating neuronal tumor | |
| | Dysplastic cerebellar gangliocytoma (Lhermitte-Duclos disease) | |
| | Extraventricular neurocytoma | |
| | Cerebellar liponeurocytoma | |
| | Central neurocytoma | |

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[a]WHO classification adapted from Louis et al.[1]

| Tumor Type (Based on the 2021 WHO Classification)[a] | Pathological Subtype (Based on the 2021 WHO Classification)[a] | Related PDQ Treatment Summary |
|---|---|---|
| **Tumors of the pineal region** | Pineoblastoma | Childhood Medulloblastoma and Other Central Nervous System Embryonal Tumors Treatment |
| **Embryonal tumors** | Medulloblastoma, *WNT*-activated | Childhood Medulloblastoma and Other Central Nervous System Embryonal Tumors Treatment |
| | Medulloblastoma, SHH-activated and *TP53*-mutant; Medulloblastoma, SHH-activated and *TP53*-wildtype | |
| | Medulloblastoma, non-*WNT*/non-SHH | |
| | Medulloblastoma, histologically defined | |
| | Desmoplastic nodular medulloblastoma | |
| | Medulloblastoma with extensive nodularity | |
| | Large cell medulloblastoma | |
| | Anaplastic medulloblastoma | |
| | Embryonal tumor with multilayered rosettes, *C19MC*-altered | |
| | CNS tumor with *BCOR* internal tandem duplication | |
| | CNS neuroblastoma, *FOXR2*-activated | |

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[a]WHO classification adapted from Louis et al.[1]

| Tumor Type (Based on the 2021 WHO Classification)[a] | Pathological Subtype (Based on the 2021 WHO Classification)[a] | Related PDQ Treatment Summary |
|---|---|---|
| | CNS embryonal tumor, NEC/NOS | |
| | Atypical teratoid/rhabdoid tumor | Childhood Central Nervous System Atypical Teratoid/Rhabdoid Tumor Treatment |
| **Germ cell tumors** | Germinoma | Childhood Central Nervous System Germ Cell Tumors Treatment |
| | Embryonal carcinoma | |
| | Yolk sac tumor | |
| | Choriocarcinoma | |
| | Mature teratoma | |
| | Immature teratoma | |
| | Teratoma with somatic-type malignancy | |
| | Mixed germ cell tumor | |
| **Tumors of the sellar region** | Adamantinomatous craniopharyngioma | Childhood Craniopharyngioma Treatment |
| | Papillary craniopharyngioma | |

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[a]WHO classification adapted from Louis et al.[1]

### *References*


1. Louis DN, Perry A, Wesseling P, et al.: The 2021 WHO Classification of Tumors of the Central Nervous System: a summary. Neuro Oncol 23 (8): 1231-1251, 2021. [PUBMED Abstract]

## Latest Updates to This Summary (12/19/2023)

The PDQ cancer information summaries are reviewed regularly and updated as new information becomes available. This section describes the latest changes made to this summary as of the date above.

This summary was comprehensively reviewed and extensively revised.

This summary was renamed from Childhood Brain and Spinal Cord Tumors Treatment Overview.

This summary is written and maintained by the PDQ Pediatric Treatment Editorial Board, which is editorially independent of NCI. The summary reflects an independent review of the literature and does not represent a policy statement of NCI or NIH. More information about summary policies and the role of the PDQ Editorial Boards in maintaining the PDQ summaries can be found on the About This PDQ Summary and PDQ® Cancer Information for Health Professionals pages.

# About This PDQ Summary

## Purpose of This Summary

This PDQ cancer information summary for health professionals provides comprehensive, peer-reviewed, evidence-based information about the treatment of childhood brain and spinal cord tumors. It is intended as a resource to inform and assist clinicians in the care of their patients. It does not provide formal guidelines or recommendations for making health care decisions.

## Reviewers and Updates

This summary is reviewed regularly and updated as necessary by the PDQ Pediatric Treatment Editorial Board, which is editorially independent of the National Cancer Institute (NCI). The summary reflects an independent review of the literature and does not represent a policy statement of NCI or the National Institutes of Health (NIH).

Board members review recently published articles each month to determine whether an article should:

- be discussed at a meeting,
- be cited with text, or
- replace or update an existing article that is already cited.

Changes to the summaries are made through a consensus process in which Board members evaluate the strength of the evidence in the published articles and determine how the article should be included in the summary.

The lead reviewers for Childhood Brain and Spinal Cord Tumors Summary Index are:

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- Louis S. Constine, MD (James P. Wilmot Cancer Center at University of Rochester Medical Center)
- Roger J. Packer, MD (Children's National Hospital)
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## Levels of Evidence

Some of the reference citations in this summary are accompanied by a level-of-evidence designation. These designations are intended to help readers assess the strength of the evidence supporting the use of specific interventions or approaches. The PDQ Pediatric Treatment Editorial Board uses a formal evidence ranking system in developing its level-of-evidence designations.

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## Disclaimer

Based on the strength of the available evidence, treatment options may be described as either “standard” or “under clinical evaluation.” These classifications should not be used as a basis for insurance reimbursement determinations. More information on insurance coverage is available on Cancer.gov on the Managing Cancer Care page.

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