ABSTRACT

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Advances in the classification and treatment of pediatric brain tumors.

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PURPOSE: Recent genetic and molecular findings have impacted the diagnosis, prognosis, and in some instances, treatment strategies for children with pediatric central nervous system tumors. Herein, we review the most up-to-date molecular findings and how they have impacted tumor classification and clinical care.
RECENT FINDINGS: It is now recognized that aberrations of the mitogen-activated protein kinase pathway are present in the majority of pediatric low-grade glioma. Also, there has been the identification of recurrent histone H3 K27M mutations in diffuse intrinsic pontine and other midline gliomas. Medulloblastoma is now divided into four molecular subgroups with distinct characteristics and prognoses. The classification of other unique embryonal tumors is also highlighted. Finally, we present the newest classification of ependymoma; supratentorial ependymomas comprise two subtypes based on expression of the hhromosome 11 Open Reading Frame 95-reticuloendotheliosis Viral Oncogene Homolog A or yes-associated protein 1 fusion, whereas posterior fossa ependymomas are divided into two distinct molecular subgroups, posterior fossa-A and posterior fossa-B.
SUMMARY: These advances in the molecular classification of pediatric central nervous system tumors have not only assisted in diagnoses, but they have led to a new era of tumor classification and prognostication. They also have served as drivers for the evaluation of new targeted therapies based upon molecular aberrations with the hope for improved survival outcomes for our patients.

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