PubMed

**Format**: Abstract

Semin Neurol. 2018 Feb;38(1):104-111. doi: 10.1055/s-0038-1636503. Epub 2018 Mar 16.

# Ependymoma.

Gerstner ER[1], Pajtler KW[2,3,4].

1 Division of Neuro-oncology, Massachusetts General Hospital Cancer Center, Boston, Massachusetts.
2 Hopp-Children's Cancer Center, NCT Heidelberg (KiTZ), Heidelberg, Germany.
3 Division of Pediatric Neurooncology, German Cancer Research Center (DKFZ), German Cancer Consortium (DKTK), Heidelberg, Germany.
4 Department of Pediatric Hematology and Oncology, Heidelberg University Hospital, Heidelberg, Germany.

Ependymoma can arise throughout the whole neuraxis. In children, tumors predominantly occur intracranially, whereas the spine is the most prevalent location in adults. Significant variance in the grade II versus grade III distinction of ependymomas has led to the acknowledgment that the clinical utility of histopathological classification is limited. Epigenomic profiling efforts have identified molecularly distinct groups of ependymomas that adequately reflect the biological, clinical, and histopathological heterogeneities across anatomical compartments, age groups, and grades. The recent update of the World Health Organization classification of central nervous system tumors has already integrated one of these groups, and molecular classification will be part of future clinical trials to improve risk stratification. Clinical management of this rare disease is challenging, making professional experience and intensified multidisciplinary cooperation pivotal factors for treatment success. Novel research strategies are currently applied for target discovery in ependymomas since for most molecular groups, genetic drivers are unknown.

PMID: **29548057** DOI: 10.1055/s-0038-1636503