

Ependymoma

- [Optimizing outcomes in intracranial ependymoma: a contemporary review](#)
- [Machine learning-driven national analysis for predicting adverse outcomes in intramedullary spinal cord tumor surgery](#)
- [How long should survivors of pediatric medulloblastoma and ependymoma be screened for recurrence? A retrospective cohort study](#)
- [Recurrent parietal lobe supratentorial ependymoma, ZFTA fusion-positive, CNS WHO grade 3, with new dural and calvarial reactive changes in a child: illustrative case](#)
- [Radiosurgery for pediatric central nervous system lesions - initial report and insights from a multicenter registry](#)
- [Comprehensive Genome Analysis Facilitates the Diagnosis of Ependymoma With ZFTA::NCOA2 Fusion](#)
- [Intramedullary schwannoma mimicking ependymoma: illustrative case](#)
- [Diagnostic Accuracy of MRI and CT Scan Features in Differentiation of Pediatric Ependymoma from Medulloblastoma](#)

Ependymomas are rare [neuroepithelial tumors](#) thought to arise from precursor [radial glial cells](#) lining the walls of the [ventricles](#) and central canal of the [brain](#) and [spinal cord](#), respectively.

Ependymoma is a [ependymal tumor](#).

Epidemiology

see [Ependymoma Epidemiology](#).

Classification

see [Ependymoma Classification](#).

Etiology

Fetal ependymal [tanycytes](#) directly give rise to mature [ependymocytes](#), whereas other tanycytic populations mature and remain as ependymal tanycytes within selective regions of the ventricular system, particularly the hypothalamic region of the third ventricle and within circumventricular organs. Specialized ependyma of the circumventricular organs and choroid plexus cells are additional highly specialized ependymal cells that ultimately derive from this developmental pathway.

Genomics

The most substantial magnitude of differentially expressed genes (DEGs) in ependymoma might be [HOX genes](#). However, whether the differential expression of these genes is the cause or consequence

of the disease remains to be elucidated in a larger prospective study ¹⁾.

Loss of [chromosome 22](#) and gain of 1q are the most frequent genomic aberrations in [ependymomas](#), indicating that genes mapping to these regions are critical in their pathogenesis. Using real-time quantitative PCR, Karakoula et al. measured relative copy numbers of 10 genes mapping to 22q12.3-q13.33 and 10 genes at 1q21-32 in a series of 47 pediatric [intracranial ependymomas](#). Loss of one or more of the genes on 22 was detected in 81% of cases, with RAC2 and C22ORF2 at 22q12-q13.1 being deleted most frequently in 38% and 32% of ependymoma samples, respectively. Combined analysis of quantitative-PCR with methylation-specific PCR and bisulphite sequencing revealed a high rate (>60% ependymoma) of transcriptional inactivation of C22ORF2, indicating its potential importance in the development of pediatric ependymomas. Increase of relative copy numbers of at least one gene on 1q were detected in 61% of cases, with TPR at 1q25 displaying relative copy number gains in 38% of cases. Patient age was identified as a significant adverse prognostic factor, as a significantly shorter overall survival time ($P = 0.0056$) was observed in patients <2 years of age compared with patients who were >2 years of age. Loss of RAC2 at 22q13 or amplification of TPR at 1q25 was significantly associated with shorter overall survival in these younger patients ($P = 0.0492$ and $P = < 0.0001$, respectively). This study identifies candidate target genes within 1q and 22q that are potentially important in the pathogenesis of intracranial pediatric ependymomas ²⁾.

Clinical features

The nonspecific clinical presentation of a spinal cord tumor frequently results in the delay of diagnosis with opposing outcomes.

[Intracranial ependymoma clinical features](#)

Diagnosis

[Intracranial ependymoma diagnosis](#)

[Spinal ependymoma diagnosis](#)

Differential Diagnosis

[Ependymoma Differential Diagnosis.](#)

Outcome

Ependymoma typically has a better overall survival rate than most [gliomas](#).

Histopathological classification is not sufficient to show variable outcomes, and fails to show prognostic markers of the diverse outcomes; hence, it is essential to understand biological

mechanisms.

These tumors have a distinct propensity for metastases, both within and outside the CNS. However, dissemination at the time of first presentation and retrograde dissemination of the tumor is rare.

Case series

[Ependymoma case series.](#)

Case reports

Simultaneous supratentorial anaplastic and infratentorial low grade ependymomas with distinct genetic profiles ³⁾.

References

¹⁾

Kim KT, Lee CH, Chung CK, Kim JH. Is NF2 a Key Player of the Differentially Expressed Gene Between Spinal Cord Ependymoma and Intracranial Ependymoma? *World Neurosurg.* 2018 Jul 18. pii: S1878-8750(18)31574-2. doi: 10.1016/j.wneu.2018.07.096. [Epub ahead of print] PubMed PMID: 30031196.

²⁾

Karakoula K, Suarez-Merino B, Ward S, Phipps KP, Harkness W, Hayward R, Thompson D, Jacques TS, Harding B, Beck J, Thomas DG, Warr TJ. Real-time quantitative PCR analysis of pediatric ependymomas identifies novel candidate genes including TPR at 1q25 and CHIBBY at 22q12-q13. *Genes Chromosomes Cancer.* 2008 Nov;47(11):1005-22. doi: 10.1002/gcc.20607. PubMed PMID: 18663750.

³⁾

Wang JL, Hong CS, Henderson EY, Salma A, Heerema NA, Ray-Chaudhury A, Elder JB. Simultaneous supratentorial anaplastic and infratentorial low grade ependymomas with distinct genetic profiles. *J Neurosurg Sci.* 2018 Aug;62(4):526-529. doi: 10.23736/S0390-5616.17.03941-8. PubMed PMID: 29938482.

From:

<https://neurosurgerywiki.com/wiki/> - **Neurosurgery Wiki**

Permanent link:

<https://neurosurgerywiki.com/wiki/doku.php?id=ependymoma>

Last update: **2024/06/07 02:54**

