Childs Nerv Syst. 2024 Oct 8. doi: 10.1007/s00381-024-06639-7. Online ahead of print.

Molecular markers for pediatric low-grade glioma

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PMID: 39379532 DOI: 10.1007/s00381-024-06639-7

Abstract

Over the past decade, our understanding of the molecular drivers of pediatric low-grade glioma (PLGG) has expanded dramatically. These tumors are predominantly driven by RAS/MAPK pathway activating alterations (fusions and point mutations), most frequently in BRAF, FGFR1, and NF1. Furthermore, additional second hits in tumor suppressor genes (TP53, ATRX, CDKN2A) can portend more aggressive behaviour. Accordingly, comprehensive molecular profiling-specifically genetic sequencing, often plus copy number profiling-has become critical for guiding the diagnosis and management of PLGG. In this review, we discuss the most important genetic alterations that inform on classification and prognosis of PLGG, highlighting their diagnostic and therapeutic relevance.

Keywords: Glioneuronal tumor; Neurofibromatosis; Pilocytic astrocytoma; Targeted therapy.