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# Treatment of Pediatric Low-Grade Gliomas

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## Abstract

**Purpose of review:** Pediatric low-grade gliomas and glioneuronal tumors (pLGG) account for approximately 30% of pediatric CNS neoplasms, encompassing a heterogeneous group of tumors of primarily glial or mixed neuronal-glial histology. This article reviews the treatment of pLGG with emphasis on an individualized approach incorporating multidisciplinary input from surgery, radiation oncology, neuroradiology, neuropathology, and pediatric oncology to carefully weigh the risks and benefits of specific interventions against tumor-related morbidity. Complete surgical resection can be curative for cerebellar and hemispheric lesions, while use of radiotherapy is restricted to older patients or those refractory to medical therapy. Chemotherapy remains the preferred first-line therapy for adjuvant treatment of the majority of recurrent or progressive pLGG.

**Recent findings:** Technologic advances offer the potential to limit volume of normal brain exposed to low doses of radiation when treating pLGG with either conformal photon or proton RT. Recent neurosurgical techniques such as laser interstitial thermal therapy offer a "dual" diagnostic and therapeutic treatment modality for pLGG in specific surgically inaccessible anatomical locations. The emergence of novel molecular diagnostic tools has enabled scientific discoveries elucidating driver alterations in mitogen-activated protein kinase (MAPK) pathway components and enhanced our understanding of the natural history (oncogenic senescence). Molecular characterization strongly supplements the clinical risk stratification (age, extent of resection, histological grade) to improve diagnostic precision and accuracy, prognostication, and can lead to the identification of patients who stand to benefit from precision medicine treatment approaches. The success of molecular targeted therapy (BRAF inhibitors and/or MEK inhibitors) in the recurrent setting has led to a gradual and yet significant paradigm shift in the treatment of pLGG. Ongoing randomized trials comparing targeted therapy to standard of care chemotherapy are anticipated to further inform the approach to upfront management of pLGG patients.

**Keywords:** BRAF inhibitors; Chemotherapy; Glioneuronal tumors; MEK inhibitors; Molecular targeted therapy; Neurosurgery; Pediatric low-grade gliomas; Radiotherapy.



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