Childs Nerv Syst. 2024 May 5. doi: 10.1007/s00381-024-06430-8. Online ahead of print.

Low-grade glioma in children with neurofibromatosis type 1: surveillance, treatment indications, management, and future directions

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PMID: 38704493 DOI: 10.1007/s00381-024-06430-8

Abstract

Neurofibromatosis type 1 (NF1) is an autosomal dominant cancer predisposition syndrome characterized by the development of both central and peripheral nervous system tumors. Low-grade glioma (LGG) is the most prevalent central nervous system tumor occurring in children with NF1, arising most frequently within the optic pathway, followed by the brainstem. Historically, treatment of NF1-LGG has been limited to conventional cytotoxic chemotherapy and surgery. Despite treatment with chemotherapy, a subset of children with NF1-LGG fail initial therapy, have a continued decline in function, or recur. The recent development of several preclinical models has allowed for the identification of novel, molecularly targeted therapies. At present, exploration of these novel precision-based therapies is ongoing in the preclinical setting and through larger, collaborative clinical trials. Herein, we review the approach to surveillance and management of NF1-LGG in children and discuss upcoming novel therapies and treatment protocols.

Keywords: Low-grade glioma; Neurofibromatosis type 1; Optic pathway glioma; Pediatric oncology.