Diffuse Intrinsic Pontine Glioma: Time for Cautious Optimism.
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Abstract
Diffuse intrinsic pontine glioma is a lethal brain cancer that arises in the pons of children. The median survival for children with diffuse intrinsic pontine glioma is less than 1 year from diagnosis, and no improvement in survival has been realized in more than 30 years. Currently, the standard of care for diffuse intrinsic pontine glioma is focal radiation therapy, which provides only temporary relief. Recent genomic analysis of tumors from biopsies and autopsies, have resulted in the discovery of K27M H3.3/H3.1 mutations in 80% and ACVR1 mutations in 25% of diffuse intrinsic pontine gliomas, providing renewed hope for future success in identifying effective therapies. In addition, as stereotactic tumor biopsies at diagnosis at specialized centers have been demonstrated to be safe, biopsies have now been incorporated into several prospective clinical trials. This article summarizes the epidemiology, clinical presentation, diagnosis, prognosis, molecular genetics, current treatment, and future therapeutic directions for diffuse intrinsic pontine glioma.

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KEYWORDS: ACVR1; K27M; brainstem glioma; diffuse intrinsic pontine glioma

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