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Low-Grade Gliomas: A New Mutation, New Targeted Therapy, and Many Questions

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Abstract

The discovery in 2008 that many adult gliomas harbor a hitherto unknown mutation in the metabolic gene isocitrate dehydrogenase (IDH) initiated revolutionary advances in our understanding of the biology, and correspondingly our classification, of gliomas. IDH mutations are found in most nonglioblastoma adult gliomas and portend a better prognosis. Massive efforts have unraveled many of the pleiotropic cellular effects of these mutations and spawned several lines of investigation to target the effect to therapeutic benefit. In this article are reviewed the implications of the IDH mutation in gliomas, in particular focusing on recent studies that have culminated in a rare positive phase 3 trial in these generally refractory tumors.

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