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What's New in Grade II and Grade III Gliomas?

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The majority of World Health Organization grade II and grade III gliomas harbor heterozygous mutations in the metabolic enzyme isocitrate dehydrogenase 1 (IDH1), and tumors with an IDH wild-type status show molecular features of a glioblastoma and simply may constitute a separate disease entity. This discovery has led to a profound shift in the way that gliomas are classified and, consequently, how treatment decisions are made. We will review the current understanding of *IDH*-mutant gliomagenesis and the preclinical models being used to investigate the underlying biology of these tumors and to explore new therapeutic options for these patients. We further summarize the results of recent pivotal trials addressing treatment of grade II and grade III gliomas and highlight promising *IDH*-mutant-specific therapies on the horizon.

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