PRIMEVIEW

GLIOMA

nature REVIEWS DISEASE PRIMERS

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Gliomas are primary tumours that arise from neuroglial stem or progenitor cells and are responsible for the majority of deaths from primary brain tumours.

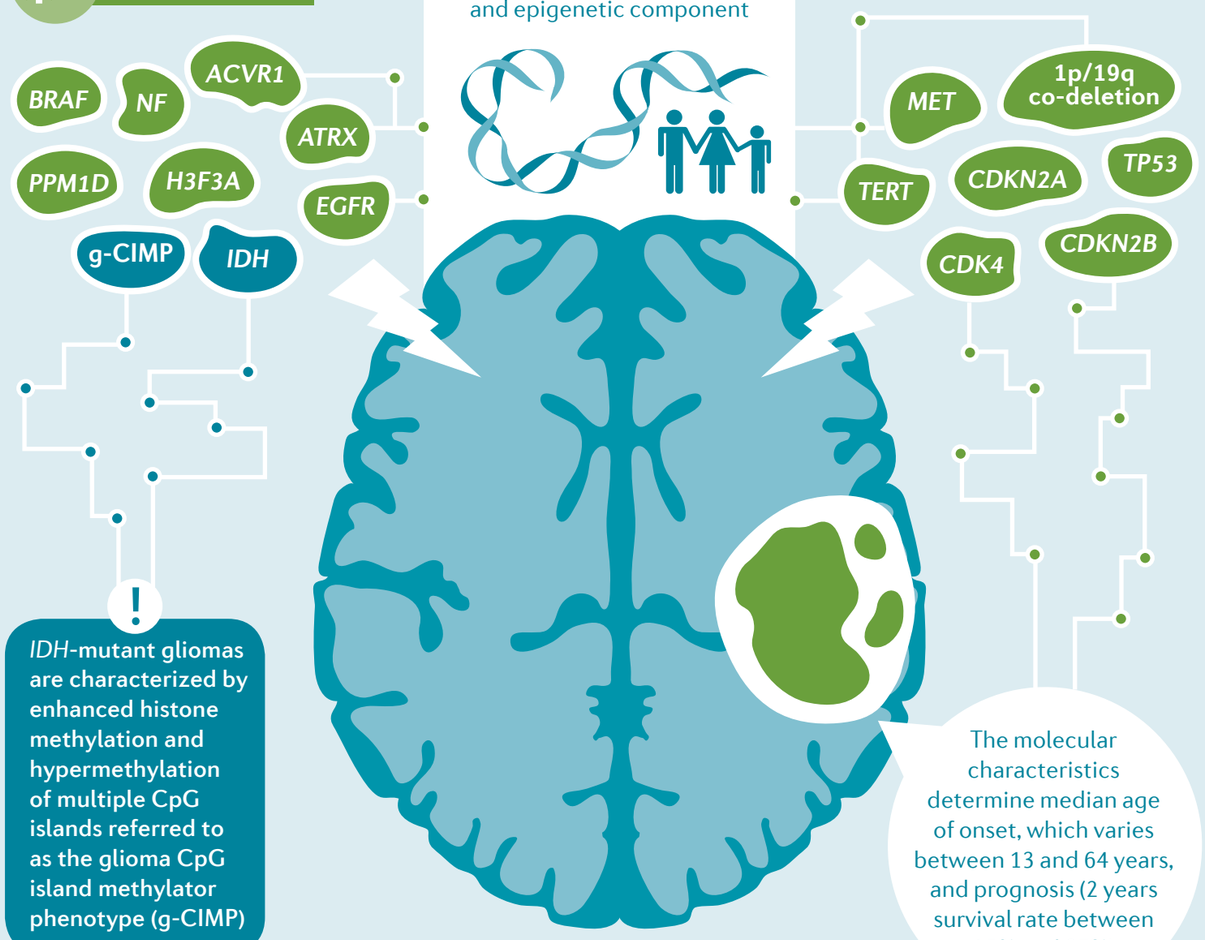
MANAGEMENT

Gliomas result in high mortality, but despite their delicate location, surgical resection is the mainstay of treatment. The need for concomitant, or follow-up, radiotherapy and/or chemotherapy is based on the individual tumour and patient characteristics. Clinical follow-up includes neurological assessment, cognitive evaluation and imaging. Other therapeutic approaches include steroids for brain oedema and anticonvulsants to control seizures.

QUALITY OF LIFE

Quality of life is impaired not only by the consequences of the brain tumour itself but also by the treatment. Consequences can include functional deficits such as motor dysfunction, impaired communication ability or decline in neurocognitive function. Almost half of patients will experience seizures.

MECHANISMS



***IDH*-mutant gliomas are characterized by enhanced histone methylation and hypermethylation of multiple CpG islands referred to as the glioma CpG island methylator phenotype (g-CIMP)**

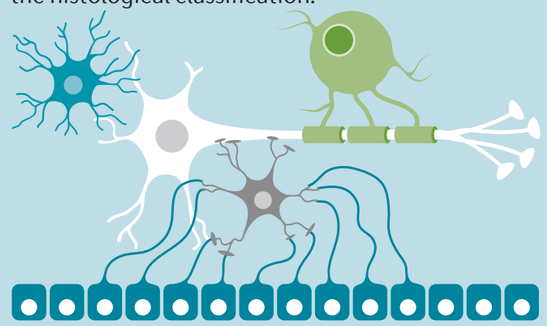
EPIDEMIOLOGY

Gliomas are the most common type of primary brain tumours (~30% of the total). The annual incidence of primary brain tumours in the United States is ~21 per 100,000 people, with ~6 per 100,000 individuals having gliomas. Geographical variation is high, with incidence rates in Japan being less than 50% of the values in the United States and Europe. The incidence also increases with age, but the biological basis of this has not yet been elucidated.

High-dose ionizing radiation (therapeutic level) is the only known exogenous risk factor

DIAGNOSIS

A neurological examination including MRI should be carried out upon the development of clinical symptoms such as sudden-onset neurological deficits and seizures. Diagnosis is based on histological classification of the resected tumour and includes the morphological resemblance to glial cells (astrocytes, oligodendrocytes or ependymal cells) and grading of malignancy (WHO grade I–IV, from low to high). Molecular profiling by DNA sequencing, methylation profiling, *in situ* hybridization, microsatellite analysis and other methods can complement the histological classification.



OUTLOOK

The continued development of high-throughput molecular profiling will lead to a better classification of gliomas, which will guide management decisions. In addition, the development of new treatment strategies will include drugs targeted to specific mutations (small-molecule inhibitors of mutant *IDH*, the MAPK pathway and EGFR inhibitors) and immunotherapy (vaccination and active cellular immunotherapy). Oncolytic viruses are also being explored as therapeutic 'weapons'. Imaging techniques will be standardized and optimized to account for treatment-specific changes that might mimic tumour progression.

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