

Review J Neurooncol. 2024 Feb 12. doi: 10.1007/s11060-024-04584-8. Online ahead of print.

# Diffuse Midline H3K27-Altered Gliomas in the Spinal Cord: A Systematic Review

Gina Watanabe <sup>1</sup>, Jennifer Manyu Wong <sup>1</sup>, Bradley Estes <sup>2</sup>, Mohammad Faizan Khan <sup>3</sup>,  
Christian Ogasawara <sup>4</sup>, Giuseppe E Umana <sup>5</sup>, Allan R Martin <sup>6</sup>, Orin Bloch <sup>6</sup>, Paolo Palmisciano <sup>7</sup>

Affiliations

PMID: 38342826 DOI: [10.1007/s11060-024-04584-8](https://doi.org/10.1007/s11060-024-04584-8)

## Abstract

**Purpose:** To systematically review the clinical features, management, and outcomes of diffuse midline H3K27-altered gliomas of the spinal cord (DMG-SCs).

**Methods:** PubMed, Ovid EMBASE, Scopus, and Web of Science were searched from database inception to 23 September 2023 for histologically confirmed cases of DMG-SC. Patient demographics, tumor characteristics, management information, and survival outcomes were extracted and analyzed.

**Results:** A total of 279 patients from 39 studies were collected. Patients were mostly male (61%), with an average age of 32 years. Patients were treated with surgery, radiotherapy, and chemotherapy combined (31%) or surgery only (24%), and extent of resection was most often subtotal (38%). Temozolomide was the most common chemotherapeutic agent (81%). Radiation therapy was delivered with mean dose of 47 Gy in 23 fractions. At mean follow-up time of 21 months, 13% of patients were alive. Average median overall survival was 24 months (range of 13 to 40 months) with a median progression-free survival of 14 months. Historical WHO grades of 2 or 3 appeared to exhibit a longer average median overall survival time than that of grade 4 DMG-SCs (32 vs. 23 months,  $p = 0.009$ ).

**Conclusions:** Outcomes for DMG-SCs are poor overall but appear to be favorable compared to intracranial DMGs. Despite the recent WHO 2021 grade 4 classification for all DMGs, given the differences in overall survival reported based on historical grading systems, future studies on DMG-SCs are needed to further define if DMG-SCs may represent a heterogeneous group of tumors with different prognoses.

**Keywords:** Diffuse midline glioma; Intramedullary; Spinal cord; Systematic review; Tumor.

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