

## ***Treatment and prognosis of Primary CNS lymphoma***

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### **SUMMARY AND RECOMMENDATIONS**

- Primary central nervous system lymphoma (PCNSL) is an uncommon variant of extranodal non-Hodgkin lymphoma that can involve the brain, leptomeninges, eyes, or spinal cord without evidence of systemic disease.
- The pretreatment evaluation of patients with PCNSL both determines the extent of the disease and provides information about the individual's comorbidities that are likely to have an impact on treatment options.
- For patients who are candidates for chemotherapy, we recommend high-dose systemic MTX-based induction therapy, rather than WBRT alone ([Grade 1B](#)). For patients with good performance status (ie, ECOG  $\leq 3$ ) who choose not to participate in a clinical trial, we suggest a MTX-based combination regimen rather than MTX alone ([Grade 2B](#)). In addition to chemotherapy, we suggest treating with [rituximab](#) ([Grade 2B](#)).
- Optimal consolidation therapy in patients who achieve a complete response has not been defined, and at least half of these patients will eventually relapse. Options for consolidation include non-myeloablative chemotherapy ([etoposide](#), [cytarabine](#)), high dose chemotherapy with autologous hematopoietic cell transplantation (HCT), or WBRT. The choice among these depends upon multiple factors, including the age of the patient, overall functional status, comorbidities, and patient preferences.
  - For younger patients who achieve a complete response with systemic high-dose chemotherapy, we suggest consolidation with chemotherapy (e.g. non-myeloablative chemotherapy or high-dose chemotherapy with HCT), ideally within the context of a clinical trial, rather than delivering whole brain radiation (WBRT) at the completion of induction chemotherapy ([Grade 2B](#)).
  - Patients over age 60 are at high risk of radiation-induced symptomatic neurotoxicity. For these patients, we suggest postponing WBRT until there is disease progression, rather than delivering it after completion of chemotherapy ([Grade 2B](#)). The optimal consolidation strategy in these patients is unknown. We generally consider a high-dose chemotherapy strategy in patients up to the age of 70 years, provided they have a good functional status and no contraindications to further chemotherapy.
- For younger patients who fail to achieve a complete response with adequate MTX-based induction chemotherapy, a second chemotherapy regimen e.g. [cytarabine](#), [etoposide](#))

can be given. HDT/HCT is an acceptable alternative for patients with an initial partial response. For such patients, we also consider the addition of WBRT after completion of chemotherapy rather than waiting until the development of progressive disease.

- For patients who are unable to tolerate high-dose systemic MTX for induction therapy, options include: alternative chemotherapy regimens (e.g. [temozolomide](#) and [rituximab](#); high-dose [cytarabine](#); or an anti-folate agent other than MTX) or palliative approaches. Palliation can be provided with WBRT or corticosteroids alone.
- The general treatment principles for PCNSL apply to most disease presentations and patient populations. However, involvement of the eye, cerebrospinal fluid, or nerve roots may have unique therapeutic implications.
- After completion of the initially planned treatment of PCNSL, it should not be assumed that a “cure” has been established. Patients should be evaluated to determine the disease response to treatment and should be followed longitudinally for relapse and long-term treatment toxicities. We follow the consensus-based guidelines published by the International PCNSL Collaborative Group (IPCG).
- Therapeutic options at relapse include retreatment with high-dose MTX if there had been a prior complete remission with this agent, alternative chemotherapy regimens ([cytarabine](#) and [etoposide](#)), HDT/HCT and WBRT.

## REFERENCES

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