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1: [Semin Oncol](#). 2009 Aug;36(4 Suppl 2):S2-S16.



Central nervous system disease in hematologic malignancies: historical perspective and practical applications.

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Acute lymphoblastic leukemia (ALL) 5-year survival rates are approaching 90% in children and 50% in adults who are receiving contemporary risk-directed treatment protocols. Current efforts focus not only on further improving cure rate but also on patient quality of life. Hence, all protocols decrease or limit the use of cranial irradiation as central nervous system (CNS)-directed therapy, even in patients with high-risk presenting features, such as the presence of leukemia cells in the cerebrospinal fluid (even resulting from traumatic lumbar puncture), adverse genetic features, T-cell immunophenotype, and a large leukemia cell burden. Current strategies for CNS-directed therapy involve effective systemic chemotherapy (eg, dexamethasone, high-dose methotrexate, intensive asparaginase) and early intensification and optimization of intrathecal therapy. Options under investigation for the treatment of relapsed or refractory CNS leukemia in ALL patients include thiotepea and intrathecal liposomal cytarabine. CNS involvement in non-Hodgkin lymphoma (NHL) is associated with young age, advanced stage, number of extranodal sites, elevated lactate dehydrogenase, and International Prognostic Index score. Refractory CNS lymphoma in patients with NHL carries a poor prognosis, with a median survival of 2 to 6 months; the most promising treatment, autologous stem cell transplant, can extend median survival from 10 to 26 months. CNS prophylaxis is required during the initial treatment of NHL subtypes that carry a high risk of CNS relapse, such as B-cell ALL, Burkitt lymphoma, and lymphoblastic lymphoma. The use of CNS prophylaxis in the treatment of diffuse large B-cell lymphoma is controversial because of the low risk of CNS relapse (approximately 5%) in this population. In this article, we review current and past practice of intrathecal therapy in ALL and NHL and the risk models that aim to identify predictors of CNS relapse in NHL.

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