

# Central Nervous System Hemophagocytic Lymphohistiocytosis

Dana T. Lounder

Division of Bone Marrow Transplantation and Immune Deficiency, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio

## Abstract

Hemophagocytic lymphohistiocytosis (HLH) is an immune regulatory disorder that commonly presents with central nervous system (CNS) involvement. The only cure for genetic HLH is hematopoietic stem cell transplantation (HSCT), typically treated with reduced-intensity conditioning (RIC) regimens. We sought to estimate the incidence of CNS relapse after RIC HSCT, determine risk factors, and evaluate outcomes. We performed a retrospective chart review of 94 consecutive children and young adults with primary HLH who received RIC HSCT. CNS relapse within 1 year after transplantation was diagnosed by review of clinical symptoms, cerebral spinal fluid (CSF), and radiologic findings. Four (4.25%) patients developed symptoms of possible CNS HLH after HSCT and 3 patients were diagnosed. Eight patients underwent screening lumbar puncture because of history of active CNS disease at the onset of the conditioning regimen and 4 had evidence of continued disease. The overall incidence of CNS relapse and continued CNS disease after RIC HSCT was 8%. All patients with CNS disease after HSCT responded to CNS-directed therapy. Whole blood donor chimerism at the time of CNS relapse was low at 1% to 34%, but it remained high at 88% to 100% for patients with continued CNS disease. Overall survival for patients with CNS relapse was 50%, compared with 75% for patients without CNS disease ( $P = .079$ ). Our data suggest that a low level of donor chimerism or active CNS disease at the time of transplantation increase the risk of CNS HLH after HSCT. Surveillance CSF evaluation after allogeneic RIC HSCT should be considered in patients with risk factors and CNS-directed treatment should be initiated if appropriate.

**Received:** January 11, 2022; **Accepted:** January 16, 2022; **Published:** January 20, 2022

## Biography

Hossein Pakdaman graduated in Neurology from the Pennsylvania and Henry Ford University. He is Professor of Nervous affiliated to Shahid Beheshti University School of

Medicine, President of Iranian Neurological Association, and Director of Iranian Neurological Board Examination. He has also published more than 40 papers in international journals and is Chairman of Iranian Journal of Nervous system.