Unmet Needs and Evidence Gaps in Relapsed/Refractory Marginal Zone Lymphoma: Findings From a Systematic Literature Review

Leyla Mohseninejad, PhD¹, Sohan Deshpande, MSc², Katarzyna Borkowska, MSc², Priscilla Wittkopf, PhD², Keri Yang, PhD, MBA, MPH, MS, CPM, BSPharm³, Dirk Weber, MD⁴

¹BeiGene Netherlands B.V., Schiphol, the Netherlands; ²Evidera, London, UK; ³BeiGene USA, Inc, San Mateo, CA, USA; ⁴BeiGene Switzerland, GmbH, Basel, Switzerland

OBJECTIVES: Marginal zone lymphoma (MZL) is a rare disease accounting for 8% to 12% of all non-Hodgkin lymphomas. Many patients experience relapse, and with limited treatment options, their risk increases over time. This review aimed to identify evidence on the clinical, epidemiologic, humanistic, and economic outcomes of R/R MZL.

METHODS: Searches were conducted in MEDLINE, EMBASE, Cochrane Library, EconLit, PsycInfo, and grey literature in November 2022 with no time limit. Two reviewers independently screened the articles. Extraction was performed by a single reviewer, with validation conducted by a second reviewer.

RESULTS: A total of 31 studies were identified. Among 21 clinical studies that were included, 17 were single-arm trials, 2 were randomized controlled trials (RCTs), and 2 were real-world evidence (RWE) studies. None of the RCTs was conducted specifically in patients with R/R MZL. Of the 19 systemic regimens identified, only 1 regimen (zanubrutinib) is currently approved in Europe for R/R MZL. Evidence from RWE studies was limited, and survival rates were generally lower than in clinical trials, indicating a lack of treatment options for patients with R/R MZL. None of the 3 epidemiology studies reported true incidence and prevalence estimates. Humanistic burden associated with MZL was underreported, with only 1 study identified. Similarly, evidence on economic evaluations and economic burden was scarce, originating mainly from only a few US-based analyses. The results, although limited, suggest a substantial economic burden of MZL, encompassing direct and indirect costs.

CONCLUSIONS: Review of clinical evidence highlights a lack of RCTs conducted in the R/R MZL population and sparsity of treatment options available in the real-world setting. More research, both RCTs and RWE studies, is needed in the MZL population and its subgroups. A significant evidence gap exists for the epidemiologic, humanistic, and economic burden in the R/R MZL population, with very little evidence available from European countries.