

Patterns of survival in lymphoplasmacytic lymphoma/Waldenström macroglobulinemia: A population-based study of 1,555 patients diagnosed in Sweden from 1980 to 2005.

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BACKGROUND:

Clinical management of lymphoplasmacytic lymphoma (LPL)/Waldenström macroglobulinemia (WM) has changed considerably over the last years, reflected in the use of new therapeutic agents and procedures (such as purine analogues, monoclonal antibodies, thalidomide-, and bortezomib-based therapies), increased use of prognostic markers, as well as supportive care measures. Due to the rarity of LPL/WM, few large phase III randomized clinical trials have been performed to assess overall survival in this disorder. Additionally, clinical trials are associated to a certain degree with a selection of patients, especially in the elderly population. Population-based studies can thus serve as a complement to clinical trials to evaluate survival patterns.

AIMS:

To establish patterns of survival in LPL/WM patients in a population-based setting in Sweden.

DESIGN AND METHODS:

Using population-based data from Sweden, we assessed variations in survival among patients diagnosed with LPL/WM from 1980 to 2005, with follow-up data until 2007. Relative survival ratios (RSR) and excess mortality rate ratios (EMRR) were computed as measures of survival.

RESULTS:

A total of 1,555 LPL/WM patients were identified (LPL n=755 and WM n=800); 58% were males and the median age at diagnosis was 72 years. There was a significant overall excess mortality in patients with LPL/WM compared to controls. Survival of patients with LPL/WM has improved significantly ($p=0.007$) over time with 5-year RSR being 0.57 (95% confidence interval (CI) 0.46-0.68), 0.65 (0.57-0.73), 0.74 (0.68-0.80), 0.72 (0.66-0.77), and 0.78 (0.71-0.85) for the calendar periods 1980-1985, 1986-1990, 1991-1995, 1996-2000, and 2001-2005, respectively (Figure). This was reflected in a 34% lower EMRR (0.66, 95% CI 0.46-0.95) in 2001-2005 compared to 1991-1995. When analyzing LPL and WM separately, WM had superior survival compared to LPL (EMRR = 0.38; 95% CI 0.30-0.48; Figure). However, there was still a significant improvement in survival over time in both disorders. Older age at LPL/WM diagnosis was consistently associated with a poorer survival. ($p<0.001$). Females had a better survival than men, EMRR 0.79 (95% CI 0.64-0.97; $p=0.027$).

CONCLUSIONS:

In this large population-based study including over 1,500 LPL/WM patients, we found a significant improvement in survival in recent years. This is probably explained by the introduction of effective therapeutic developments in recent years and better supportive care. It can not be excluded that earlier establishment of LPL/WM diagnosis in more recent years may partly contribute to our findings. LPL was associated with an inferior survival, which may reflect different biology, prognostic factors or treatment-related issues. New effective agents with a more favourable toxicity profile are needed to improve survival further, particularly in the elderly.

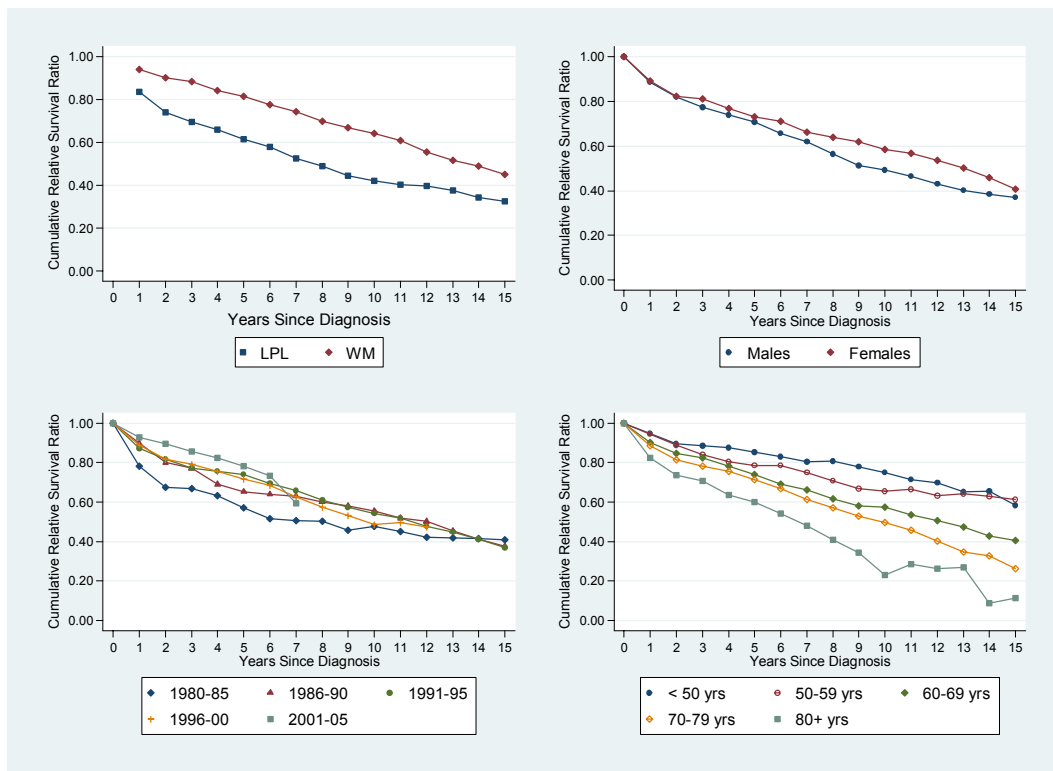


Figure. Relative survival ratios of lymphoplasmacytic lymphoma/Waldenström macroglobulinemia patients stratified by subtype, sex, calendar period, and age at diagnosis.