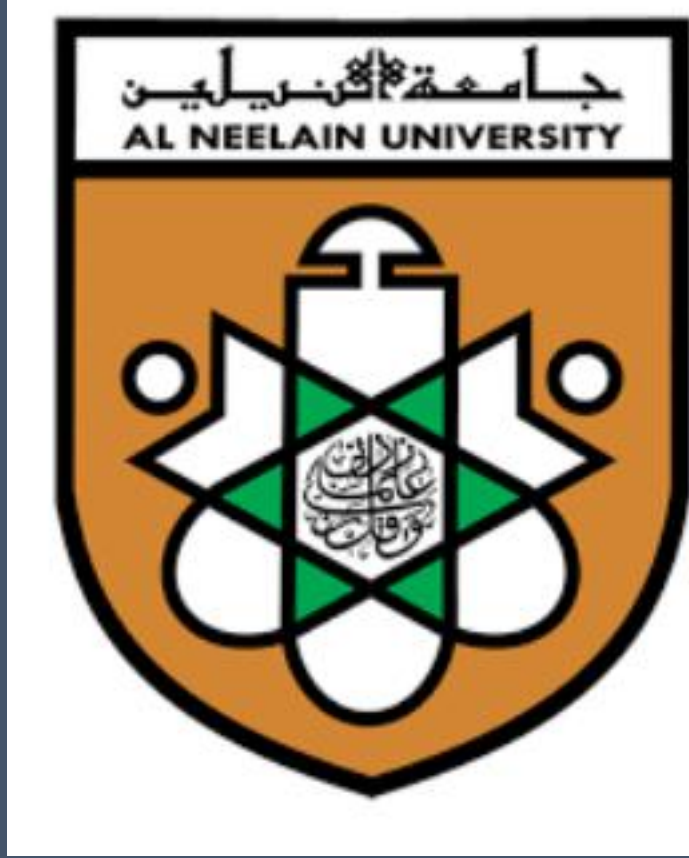


LIFE EXPECTANCY OF HAIRY CELL LEUKEMIA PATIENTS APPROACHES THAT OF THE GENERAL POPULATION: AN UPDATED ANALYSIS OF 5669 CASES FROM THE SEER DATABASE (1978-2020)



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INTRODUCTION

Hairy cell leukemia (HCL) is a rare lymphoproliferative disorder affecting mature B-cells, predominantly observed in males. It is recognized by cytopenia, splenomegaly, myelofibrosis, and the presence of atypical lymphoid cells exhibiting distinctive cytoplasmic hairy projections.

METHOD

This is a population-based study that used updated HCL clinical data from the Surveillance, Epidemiology, and End Results (SEER) database. This study identified patients with HCL diagnosed between 1978 and 2020. A total of 5669 patients were included after data screening by inclusion and exclusion criteria. Univariate and multivariate Cox regression models were used to study the factors that affect overall survival (OS).

RESULTS

Estimated median survival for patients with HCL aged less than 40 years was 39.3 years (95% confidence interval [CI], 38.9–39.7 years) compared with 8.8 years (95% CI, 8.0–9.4 years) for patients with HCL older than 65 years, which closely approaches the life expectancy of the general population. Also, patients diagnosed by multiple advanced methods, such as immunophenotyping and genetic studies, exhibited a reduced hazard ratio (0.52; 95% CI, 0.38–0.72) compared with those diagnosed using histology alone.

CONCLUSIONS

This study found that the life expectancy of patients with HCL is very close to that of the general population. Improved OS is associated with recent diagnoses (post 2006), chemotherapy treatment, and use of multiple advanced diagnostic methods, such as immunophenotyping and genetic studies.

TABLES AND FIGURES

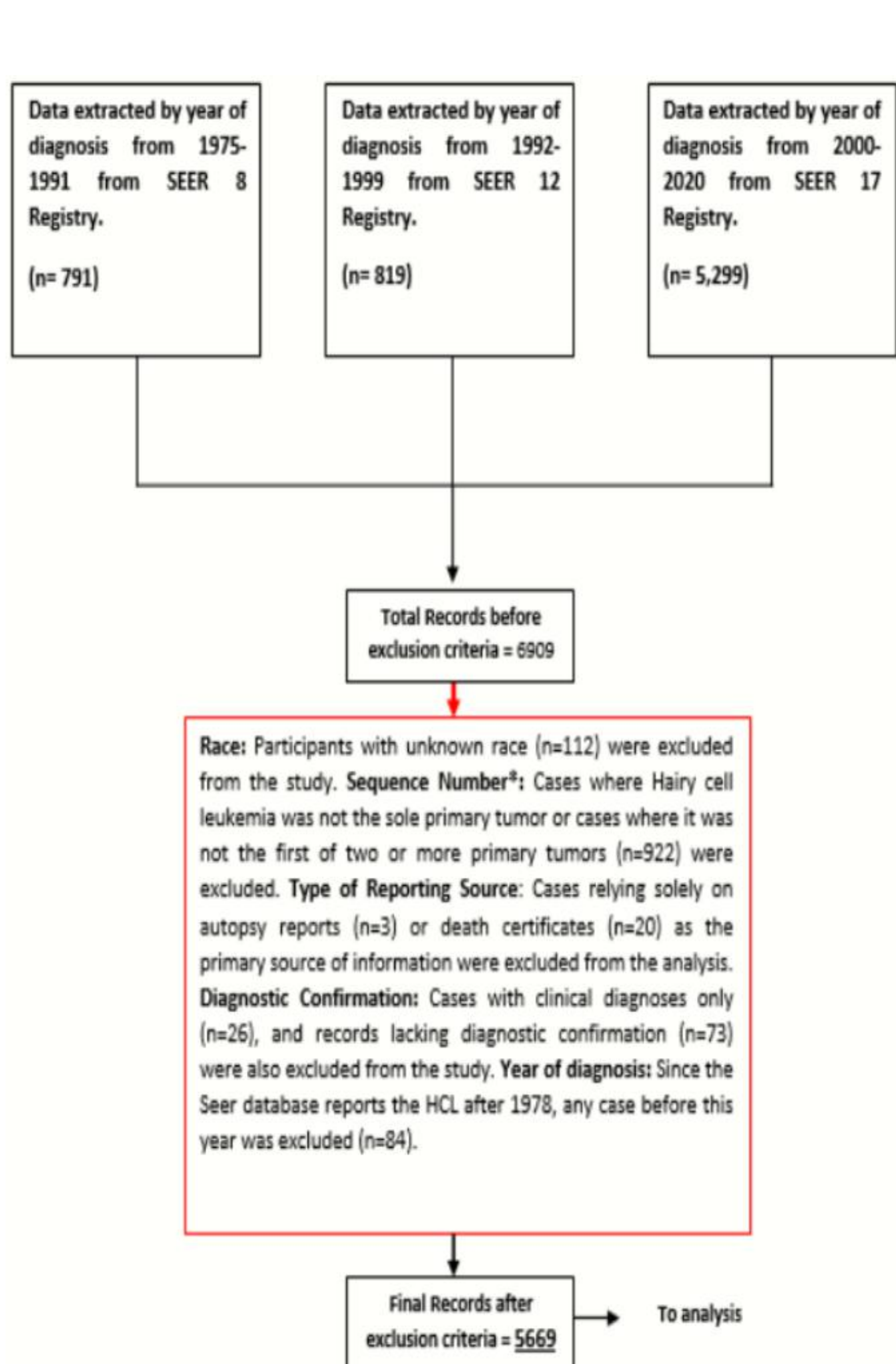


Figure 1: The figure shows the flow chart of the exclusion criteria, *Sequence Number: The sequence number indicates the sequence of all reportable neoplasms over the lifetime of the patient.

Table 1: Clinicopathological variables of HCL patients

		Count	N %
Age	15-39 Years	539	9.5%
	40-64 Years	3256	57.4%
	Older Than 65	1874	33.1%
Sex	Male	4475	78.9%
	Female	1194	21.1%
Race	White	5275	93.0%
	Non-White	394	7.0%
Year of diagnosis	Diagnosed before 1991	573	10.1%
	Diagnosed Between 1991-2006	2177	38.4%
	Diagnosed After 2006	2919	51.5%
Median household income inflation-adjusted to 2021	Less than \$54,999	616	10.9%
	\$55,000 To \$74,999	2114	37.3%
	\$75,000+	2420	42.7%
	Unknown	519	9.2%
Type of reporting	Hospital inpatient/outpatient or clinic	5170	91.2%
	Other	499	8.8%
Diagnostic conformation	Positive histology	4789	84.5%
	Positive histology plus (Immunophenotyping AND/OR Pos Genetic Studies)	748	13.2%
	Other	132	2.3%
Marital status	Single (never married)	802	14.1%
	Married (Including Common Law)	3623	63.9%
	Divorced or Separated	399	7.0%
	Widowed	313	5.5%
	Unknown and Other	532	9.4%
Primary site labeled	Bone marrow	5663	99.9%
	Spleen and Blood	6	0.1%
Radiation	Beam Radiation	20	0.4%
	None/Unknown and Other	5649	99.6%
Surgery	Surgery Performed	240	4.2%
	Not Recommended	4893	86.3%
	Recommended	451	8.0%
	Unknown	85	1.5%
	No/Unknown	2113	37.3%
Chemotherapy	Yes	3556	62.7%
	No/Unknown	2113	37.3%
Vital status	Live	3728	65.8%
	Dead	1941	34.2%

Table 2: Univariate and multivariate Cox regression analysis of the prognostic factors for WN patients

	Multivariate Cox Regression		Univariate Cox Regression	
	P-value	Hazard Ratio [95.0% CI]	P-value	Hazard Ratio [95.0% CI]
Age	<0.001		<0.001	
15-39 years (Reference)				
40-64 years	<0.001	2.16 [1.56-2.99]	<0.001	2.86 [2.18-3.77]
Older than 65 years	<0.001	5.22 [3.73-7.30]	<0.001	15.33 [11.65-20.16]
Sex				
Male (Reference)				
Female	0.04	0.86 [0.74-1.00]	0.02	1.14 [1.02-1.27]
Race				
White (Reference)				
Non-white	0.94	1.01 [0.80-1.27]	0.13	1.15 [0.96-1.37]
Year of diagnosis	<0.001		<0.001	
Diagnosed before 1991 (Reference)				
Diagnosed between 1991-2006	0.14	0.71 [0.44-1.12]	<0.001	0.66 [0.59-0.75]
Diagnosed after 2006	<0.001	0.09 [0.06-0.15]	<0.001	0.54 [0.47-0.62]
Median household income inflation-adjusted to 2021	0.51		<0.001	
Less than \$54,999 (Reference)				
\$55,000 to \$74,999	0.28	0.89 [0.72-1.10]	0.02	0.83 [0.71-0.98]
\$75,000+	0.16	0.86 [0.69-1.06]	<0.001	0.73 [0.62-0.85]
Unknown	0.34	0.78 [0.47-1.30]	<0.001	1.29 [1.08-1.54]
Type of reporting				
Hospital inpatient/outpatient or clinic (Reference)				
Other			0.49	1.06 [0.90-1.25]
Diagnostic confirmation	<0.001		0.02	
Positive histology (Reference)				
Positive histology AND immunophenotyping AND/OR positive genetic studies	<0.001	0.52 [0.38-0.72]	0.08	0.82 [0.66-1.02]
Other	0.81	0.94 [0.60-1.50]	0.03	1.38 [1.02-1.85]
Marital status	0.02		<0.001	
Single (Reference)				
Married	0.04	0.82 [0.67-0.99]	0.21	1.10 [0.95-1.27]
Divorced or separated	1.00	1.00 [0.76-1.32]	0.01	1.31 [1.06-1.62]
Widowed	0.97	1.00 [0.76-1.32]	<0.001	4.33 [3.60-5.21]
Unknown and other	0.02	0.66 [0.47-0.92]	0.46	1.08 [0.88-1.33]
Radiation				
Beam radiation (Reference)				
None/Unknown and other	0.64	0.87 [0.49-1.55]	<0.001	0.22 [0.14-0.37]
Surgery	0.20		<0.001	
Surgery performed (Reference)				
Not Recommended	0.17	0.83 [0.64-1.08]	<0.001	0.69 [0.58-0.80]
recommended	0.23	0.85 [0.65-1.11]	0.84	1.02 [0.85-1.23]
Unknown	0.19	1.56 [0.81-3.01]	0.39	1.13 [0.85-1.50]
Chemotherapy				
Yes (Reference)				
No/Unknown	0.06	1.22 [0.99-1.51]	<0.001	1.92 [1.76-2.11]
Follow-up time	<0.001	0.12 [0.11-0.14]	<0.001	0.18 [0.17-0.19]
Months from diagnosis to treatment	0.61	1.01 [0.98-1.03]	<0.001	1.04 [1.02-1.06]

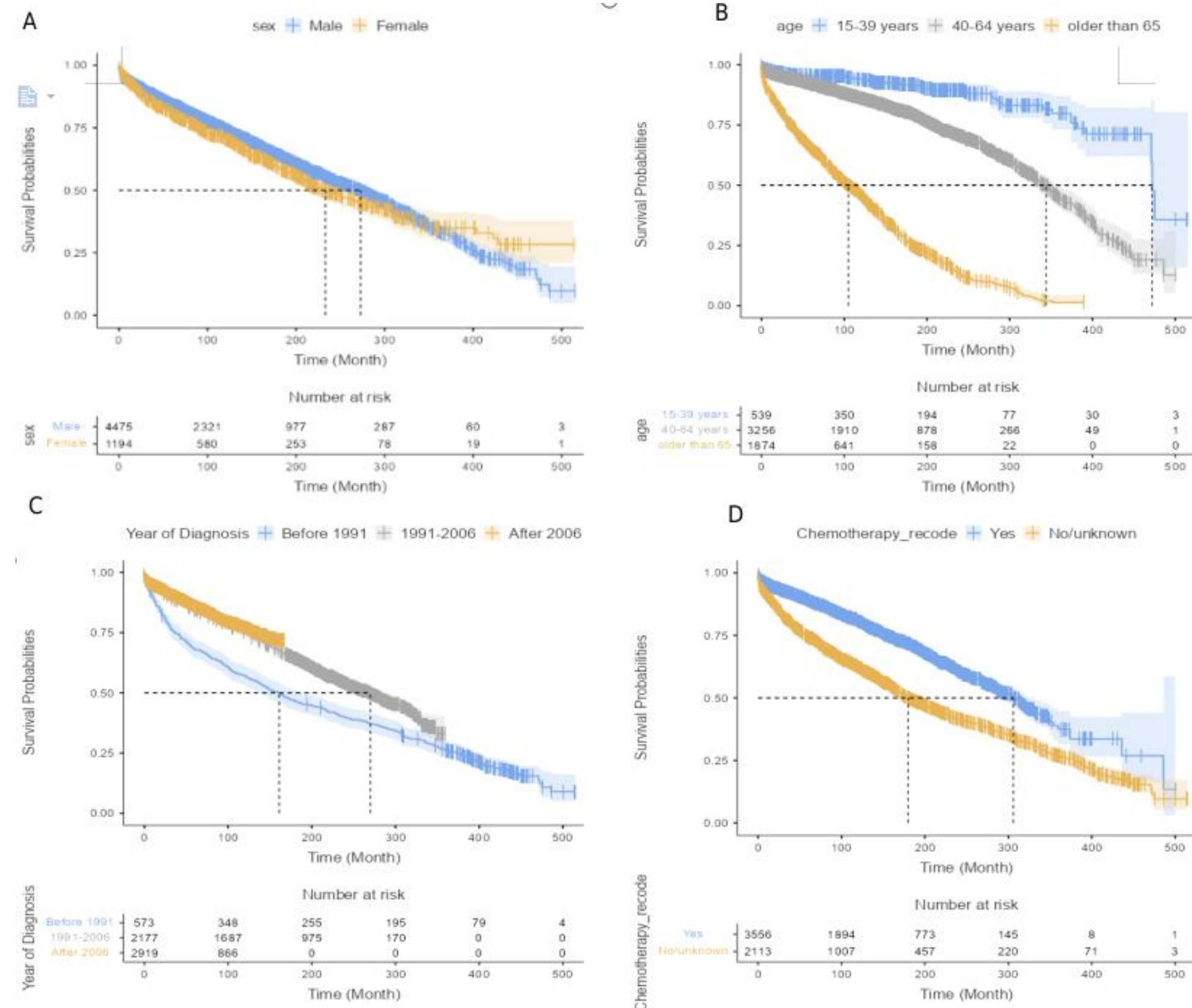


Figure 2: This figure shows four Kaplan-Meier survival curves of patients with Hairy Cell Leukemia (HCL) stratified according to sex (Figure 2a), age (Figure 2b), year of diagnosis (Figure 2c) and chemotherapy treatment (Figure 2d).

Table 3: Cause of death in HCL patients

Cause of Death	count	%
Lymphoid leukemias (excluding CLL/SLL)	452	23.3%
Ischemic heart disease	200	10.3%
Other and unspecific disorders of the circulatory system	99	5.1%
Other leukemias	86	4.4%
Lung and bronchus	79	4.1%
Non-Hodgkin lymphoma	77	4.0%
Cerebrovascular diseases	57	2.9%
Miscellaneous Neoplasms	55	2.8%
Other causes of death	836	43.1%

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