

CANCER OBSERVATORY

Volume II

HEMATOLOGICAL NEOPLASMS

2000-2022

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Cancer Observatory Hematological Neoplasms

Hospital Cancer Registry

A.C. Camargo Cancer Center

2000 to 2022

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INDEX

| | |
|---|----|
| Acknowledgments..... | 8 |
| Introduction | 9 |
| 1. A.C.Camargo Cancer Center Cancer Observatory | 10 |
| 2. Hospital Cancer Registry | |
| 2.1. Concept | 11 |
| 2.2. Objectives. | 11 |
| 2.3. The HCR team's working methods. | 11 |
| 3. HCR Quality Indicators..... | 13 |
| 4. Cancer Observatory Methods | |
| 4.1. Descriptive analysis | 14 |
| 4.2. Survival analysis. | 17 |
| 5. Summary of Hematological Neoplasms. | 18 |
| 6. Observatory Results for Hematological Neoplasms | |
| 6.1. Descriptive results. | 22 |
| 6.2. Survival results..... | 25 |
| 7. Conclusions. | 33 |
| 8. The Hematological Neoplasms Reference Center Team..... | 34 |
| 9. The HCR Team. | 35 |
| 10. HCR Database Access. | 36 |
| 11. Contacts at the Hospital Cancer Registry | 37 |
| References. | 38 |
| Appendices..... | 43 |

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Diego Rodrigues Mendonça e Silva, Ph.D.
HCR Coordinator

Introduction

The A.C.Camargo Cancer Center's (ACCCC's) Cancer Observatory for malignant hematological neoplasms describes the epidemiological profiles of and estimated overall survival rates for leukemias, lymphomas, and multiple myeloma (MM), as well as chronic myeloproliferative neoplasms (CMPNs) and myelodysplastic syndromes.

Hematological neoplasms are a group of rare cancers representing 3-4% of all cancers. The diagnosis and treatment of these neoplasms are challenging because of the complexity of the circulatory system. However, advances in hematological oncology have increased the ability to diagnose and treat these cancers, resulting in improved prognoses for affected patients.

In this second volume of the ACCCC's Hospital Cancer Registry (HCR) Cancer Observatory, we describe the sociodemographic profile of patients treated for hematological neoplasms between 2000 and 2022, and the overall survival rates for selected malignant hematological neoplasms for the period of 2000-2019.

The aims of the Cancer Observatory for hematological neoplasms are to amplify the knowledge of the results achieved by the institution and to share the epidemiological characteristics and survival rates, with a vision to contributing to the dissemination of information about rare neoplasms. In so doing, we are collaborating with broader scientific research and thus contributing to healthcare and management indicators that benefit cancer patients' treatment.

1 A.C. Camargo Cancer Center Cancer Observatory

The Cancer Observatory is an annual publication that organizes, analyzes, and shares the epidemiological profiles and survival results for patients with cancer diagnosed and/or treated at the ACCCC over time.

The HCR database is the source of information for the Cancer Observatory. This database has been in continuous operation at our institution since 2000. Currently, the HCR team is working on adding cases diagnosed in 2023. The database contains more than 150,000 new cases of primary (unique and multiple) tumors, including cases of skin cancer.

In the first volume of the Cancer Observatory, published in 2023, we described the profile of about 98,000 cancer cases treated at the ACCCC between 2000 and 2020. These profiles represented all types of cancer, including non-melanoma skin cancers, and contained overall survival rates for the most frequent types of cancer treated at the Institution between 2000 and 2017.

Below, we describe the HCR's activities, and the methodology used for the groups of hematological neoplasms analyzed.

2 Hospital Cancer Registry

2.1. Concept

The Hospital Cancer Registry (HCR) work group is responsible for and specialized in the analysis, coding, validation, and recording of information on patients diagnosed and/or treated for cancer at a specific institution. The record for each cancer case contains information on the patient's sociodemographic characteristics, clinical diagnosis, staging, treatment, and follow-up (ACCCC, 2023).

2.2. Objectives

- To describe cancer cases treated each year at ACCCC;
- To apply the International Classification of Diseases for Oncology-3rd edition (ICD-O3) criteria when recording cases;
- To describe the sociodemographic, epidemiological, and clinical characteristics of the cases;
- To describe treatment and staging;
- To estimate overall patient survival rates.

Records team

The HCR team consists of registrars trained in the extraction, classification, and coding of data on cancer cases according to the ICD-O3.

2.3. The HCR team's working methods

The HCR contains records of all malignant tumors diagnosed in a given year. A given patient may have one more tumors, and all of them are recorded according to the date of diagnosis. Diagnoses of hematological neoplasms are confirmed through blood counts, myelograms, lymph-node biopsies, and/or bone marrow or molecular markers. Cases are recorded with a delay of about 6-12 months.

The HCR team first identifies a new case according to the patient's point of entry into the Institution, such as the Department of Anatomical Pathology, Chemotherapy, or Radiation Therapy or the health/cancer clinic (Figure 1). They then work to create a final and definitive HCR record by recording data on all essential variables (Appendices).

The morphological classifications and topographic codes for malignant neoplasms are selected from the 2012 version of the ICD-O3. For each case, the HCR team also collects data about the clinical stage and pathology, presence of metastases, treatment performed, and the patient's vital status (alive or dead) based on the latest available information.

To input patient information, the HCR team uses the SISHCR© 2007 software (version 6.72) developed by the Fundação Oncocentro do Estado de São Paulo (FOSP). After the creation of a definitive case record, the team verifies the consistency of the information and performs validation using the IARCcrgTools© 2018 software. Duplicate cases are identified and managed using the Link Plus 2018 software (version 2.0) from the Centers for Disease Prevention and Control.

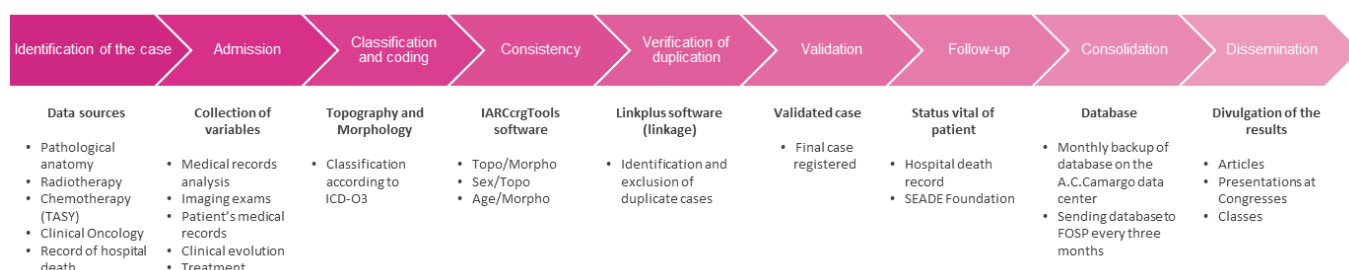


Figure 1. Flowchart of the HCR team's recording of cancer cases/ A.C. Camargo Cancer Center, São Paulo, 2024.

3 HCR Quality Indicators

The following indicators are used to assess the quality of the HCR: the proportion of cases with confirmation via histological (microscopic) analysis; the proportion of “malignant neoplasm, not otherwise specified (NOS), 8000/3” cases; the proportion of “primary site unknown, C80.9” cases; and the proportion of cases without clinical staging. Annual monitoring of these indicators enables the implementation of effective actions to improve the accuracy of the information contained in the HCR.

The HCR quality indicators are described in the Brazilian National Cancer Institute (INCA) Cancer Registry Manual (MS/INCA, 2010) and applied by the FOSP to all HCRs in the State of São Paulo (Table 1).

Table 1. Data quality indicators for the Hospital Cancer Registry, A.C. Camargo Cancer Center, according to the INCA/MS and FOSP for 2000 to 2022.

| Indicator | A.C. CAMARGO | Fundação Oncocentro São Paulo (FOSP) ^a | Brazilian National Cancer Institute (INCA) ^b |
|--|-----------------|--|---|
| Proportion of cases confirmed by microscopy (histology) | 98.9% | 98.3% | ≥95.0%–<100% |
| Proportion of cases coded as “malignant neoplasm, not otherwise specified” (NOS; 8000/3) | 0.2% | 0.9% | <3.0% |
| Proportion of cases coded as “unknown primary site” (C80.9) | 0.6% | 1.3% | <2.5% |
| Proportion of cases without staging (code “X”) | 4.7% | 4.0% | <10.0 % |

a) HCRs in the State of São Paulo, 2000-2020. Source: FOSP, March/2024.

b) National Cancer Institute (Brazil): Hospital Cancer Registries - planning and management. 2nd ed. rev. Rio de Janeiro: INCA, 2010.

4 Cancer Observatory Methods

4.1. Descriptive analysis

We included cases of malignant hematological neoplasms diagnosed between 2000 and 2022, extracted from the HCR database on January 9, 2024.

Malignant hematological tumors are described in terms of their annual distribution, patient age group (5-year intervals), and patient sex, and distributed into seven hematological groups [acute leukemias (AL), aggressive lymphomas, indolent lymphomas, Hodgkin lymphoma, monoclonal gammopathies, CMPNs, and myelodysplastic syndromes] according to ICD-O3 morphological codes (Chart 1).

| Chart 1. Groups of hematological neoplasms with the morphological code (International Classification of Diseases for Oncology, 3rd edition) included in this study, 2000 to 2022. | |
|---|--|
| Hematological neoplasm group | Morphology (ICD-O3) |
| Acute leukemias | 97273 Precursor cell lymphoblastic lymphoma, NOS 97283 Precursor B-cell lymphoblastic lymphoma 97293 Precursor T-cell lymphoblastic lymphoma 98013 Acute leukemia, NOS 98053 Acute biphenotypic leukemia 98203 Lymphoid leukemia, NOS 98213 Acute lymphoblastic leukemia, NOS 98223 Subacute lymphoid leukemia 98353 Precursor cell lymphoblastic leukemia, NOS 98363 Precursor B-cell lymphoblastic leukemia 98373 Precursor T-cell lymphoblastic leukemia 98403 Acute myeloid leukemia, M6 type 98603 Myeloid leukemia, NOS 98613 Acute myeloid leukemia, NOS 98663 Acute promyelocytic leukemia, T(15;17)(q22;q11-12) 98673 Acute myelomonocytic leukemia 98723 Acute myeloid leukemia, minimal differentiation |

| Hematological neoplasm group | Morphology (ICD-O3) |
|------------------------------|---|
| Acute leukemias | 98913 Acute monocytic leukemia 98953 Acute myeloid leukemia with multilineage dysplasia 98963 Acute myeloid leukemia, t(8;21)(q22;q22) 99303 Myeloid sarcoma 99313 Acute panmyelosis with myelofibrosis 99483 Aggressive NK-cell leukemia |
| Aggressive lymphoma | 95903 Malignant lymphoma, NOS 95913 Diffuse malignant lymphoma, NOS 95963 Composite lymphoma: Hodgkin and non-Hodgkin 96733 Mantle cell lymphoma 96753 Malignant lymphoma, mixed small and large cell, diffuse 96793 Mediastinal large B-cell lymphoma (C38.3) 96803 Malignant lymphoma, large B-cell, diffuse, NOS 96813 Malignant lymphoma, large cleaved cells, diffuse 96823 Malignant lymphoma, noncleaved, diffuse 96843 Malignant large B-cell lymphoma, diffuse, immunoblastic 96853 Malignant lymphoma, convoluted cell 96873 Burkitt lymphoma, NOS 97013 Sézary syndrome 97023 Mature T-cell lymphoma, NOS 97033 T-zone lymphoma 97053 Angioimmunoblastic T-cell lymphoma 97073 Peripheral T-cell lymphoma, pleomorphic medium and large cell 97083 Subcutaneous panniculitis-like T-cell lymphoma 97093 Cutaneous T-cell lymphoma, NOS 97113 Monocytoid B-cell lymphoma 97143 Anaplastic large-cell lymphoma, T and NULL 97173 Intestinal T-cell lymphoma 97193 Nasal and T/NK cell nasal lymphoma 98263 Burkitt lymphoma 98273 Adult T-cell leukemia/lymphoma (HTLV-1 positive) |
| Indolent lymphomas | 95913 Non-Hodgkin malignant lymphoma, NOS 96703 Malignant lymphoma, small B lymphocytic, NOS 96713 Malignant lymphoma, lymphoplasmacytic 96893 Splenic marginal zone B-cell lymphoma 96903 Follicular lymphoma, NOS 96913 Follicular lymphoma, grade 2 96923 Malignant centroblastic-centrocytic lymphoma 96933 Malignant lymphoma, lymphocytic, well-defined, nodular 96953 Follicular lymphoma grade |

| Hematological neoplasm group | Morphology (ICD-O3) |
|---|--|
| Indolent lymphomas | 96973 Malignant lymphoma, centroblastic, follicular 96983 Follicular lymphoma grade 3 96993 Marginal zone B-cell lymphoma, NOS 97003 Mycosis fungoides 97183 Primary cutaneous CD30+ T-cell lymphoproliferative disorder 97233 True histiocytic lymphoma 97613 Waldenstrom macroglobulinemia 98233 B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma 98313 T-cell large granular lymphocytic leukemia 98323 Prolymphocytic leukemia, NOS 98343 Prolymphocytic leukemia, T-cell type 99403 Hairy cell leukemia |
| Hodgkin lymphoma | 96503 Hodgkin lymphoma, NOS 96513 Hodgkin lymphoma, lymphocyte rich 96523 Hodgkin lymphoma, mixed cellularity, NOS 96533 Hodgkin lymphoma, lymphocyte depletion, NOS 96593 Hodgkin lymphoma, nodular lymphocyte predominance 96633 Hodgkin lymphoma, nodular sclerosis, NOS 96643 Hodgkin lymphoma, nodular sclerosis, cellular phase 96653 Hodgkin lymphoma, nodular sclerosis, grade I 96673 Hodgkin lymphoma, nodular sclerosis, grade II |
| Monoclonal gammopathies | 97323 Multiple myeloma 97313 Plasmacytoma, NOS 97343 Plasmacytoma, extramedullary (exc. bone 9731/3) |
| Chronic myeloproliferative neoplasms | 98633 Chronic myeloid leukemia, NOS 9 98683 Chronic myelomonocytic leukemia 98803 Eosinophilic leukemia 99453 Chronic myelomonocytic leukemia, NOS 99503 Polycythemia vera 99603 Chronic myeloproliferative disease, NOS 99613 Myelosclerosis with myeloid metaplasia 99621 Idiopathic thrombocythemia 99623 Essential thrombocythemia 99643 Hypereosinophilic syndrome |

| Hematological neoplasm group | Morphology (ICD-O3) |
|---|---|
| Myelodysplastic syndromes | 99821 Refractory anemia with sideroblasts 99831 Refractory anemia with excess blasts 99863 Myelodysplastic syndrome with 5q deletion (5q-) syndrome 99873 Therapy-related myelodysplastic syndrome, NOS 99891 Myelodysplastic syndrome 99893 Myelodysplastic syndrome, NOS |
| Excluded (not belonging in any group analyzed) | 97203 Malignant histiocytosis 97503 Malignant histiocytosis 97511 Langerhans cell histiocytosis, NOS 97521 Unifocal Langerhans cell histiocytosis 97543 Langerhans cell histiocytosis, disseminated 97553 Histiocytic sarcoma 97573 Interdigitating dendritic cell sarcoma 97583 Follicular dendritic cell sarcoma 97651 Monoclonal gammopathy of undetermined significance 97691 Immunoglobulin deposition disease 99701 Lymphoproliferative disorder, NOS |

4.2. Survival analysis

In the analysis of overall survival, we considered the malignant hematological tumors treated at the ACCCC between 2000 and 2019 shown in Chart 1.

The survival time was calculated as the difference between the date of diagnosis and the date of death (of whatever cause) or the most recent date for which information was available, with tracking updated up to December 31, 2023. The overall probability of survival was calculated for four 5-year periods (2000-2004, 2005-2009, 2010-2014, and 2015-2019) and two 10-year periods (2000-2009 and 2010-2019) for both sexes.

For MM, we analyzed the 5-year overall survival probability over two periods with the patients divided into those who did and did not undergo Bone Marrow Transplantation (BMT).

Survival curves were estimated using the Kaplan-Meier method and IBM® SPSS Statistics (version 23) and compared using the log-rank test with a significance level of $p < 0.05$.

5 Summary of Hematological Neoplasms

Acute leukemias

AL are aggressive neoplasms that arise from the abnormal proliferation of immature cells of hematopoietic lineages (Health Organization, 2022; Alaggio et al., 2022). Acute myeloid leukemia (AML) affects people aged ≥ 60 years old, and acute lymphoblastic leukemia (ALL) is more frequent in the 2-5-year age group (Tebbi, 2021). AL treatments combine chemotherapy, targeted therapies, and allogenic bone-marrow transplantation (allo-BMT) (Gökbuget et al., 2024).

Advances in recent decades have led to the development of chemotherapy-free treatments for ALL that reduce the disease risk with cellular [e.g., chimeric antigen receptor (CAR) T-cell] therapies (Jabbour et al., 2023). First-line therapies for AML that involve less toxicity and provide greater disease control and access to potentially curative therapies, such as allo-BMT, have also been developed (DiNardo et al., 2020; Gökbuget et al., 2024). Rare ALs, such as mixed-phenotype acute leukemia, were also included in the present analysis.

Aggressive lymphomas

Aggressive non-Hodgkin lymphomas (NHLs) include subtypes of B-, T-, and natural killer (NK)-cell lymphomas with rapid growth and varying responses to treatment, which involves chemotherapy, immunotherapy, autologous bone-marrow transplantation (auto-BMT), and allo-BMT (Alaggio et al., 2022).

Diffuse large B-cell lymphoma is the most common lymphoma in this group (Alaggio et al., 2022), and it can potentially be cured with chemoimmunotherapy (CoifGer et al., 2002). With recent advances, we have entered a new phase in the first-line treatment of this disease (Tilly et al., 2022) and in the handling of recurrent/refractory cases using bispecific antibodies and CAR T-cell therapies (Locke et al., 2022). Mantle cell lymphoma, despite its varied behavior, is also included in this group.

Progress in the treatment of T-cell lymphomas has been slower, with few treatments approved in recent decades (O'Connor et al., 2015; Horwitz et al., 2019). Allo-BMT still offers the best chance for a restricted set of patients. The largest representative of this class is peripheral T-cell lymphoma, NOS (Zain & Hanona, 2021).

Indolent lymphomas

Indolent lymphomas constitute a diverse group of slow-growing neoplasms of varying aggressiveness. These diseases cannot be cured with currently available first-line treatments and are marked by late recurrence, which does not always require immediate treatment.

Among B-cell lymphomas, follicular lymphoma is the most common subtype, representing 20% of NHL cases (Swerdlow, et al., 2016). The use of well-defined criteria for treatment initiation continues to be an essential part of decision making for this disease (Brice et al., 1997).

Chronic lymphocytic leukemia is the leukemic form of mature B-cell lymphocyte neoplasms (Alaggio et al., 2022), and for this reason is also included in this group. The combined use of immunotherapy with anti-CD20 antibodies and chemotherapy has significantly increased response rates for indolent B-cell lymphomas (Marcus et al., 2005; Salles et al., 2011). With other molecules, such as Bruton's tyrosine kinase inhibitors (TKIs) and B-cell lymphoma-2 inhibitors, however, highly efficient treatment sequencing remains challenging (Burger et al., 2015; Jain et al., 2019).

T-cell lymphomas are rare. The main indolent form is mycosis fungoides (MF). It presents clinically as skin lesions, and the difficulty of determining clonality using the usual methods frequently contributes to its late diagnosis. Unlike B-cell lymphomas, a minority of MFs are treated with chemotherapy, while the majority of cases are monitored and treated topically by dermatologists (Wilcox, 2014).

Hodgkin lymphoma

Classic Hodgkin lymphoma (cHL) is characterized by the presence of inflammatory cells associated with rare Reed-Sternberg cells in the nodal region. These cells originate from B-lymphocytes that have lost the expression of various lineage markers, inactivating a large part of the immune response in their microenvironments (Connors et al., 2020).

Nodular lymphocyte-predominant Hodgkin lymphoma does not arise from the same cells and has distinct characteristics and response rates, which are similar to those of NHL (Eichenauer & Engert, 2017). In this analysis, we included both forms in the Hodgkin lymphoma group.

cHL has >90% response rates to the majority of conventional chemotherapy protocols and a high first-line cure rate (Connors et al., 2020). The addition of brentuximab vedotin, an antibody-drug conjugate targeting CD-30, to the therapeutic regimen is an option, even in older patients and those who cannot tolerate chemotherapy (Connors et al., 2018). The use of anti-programmed death ligand-1 (anti-PDL-1) treatment is an option for recurrent/refractory disease (Moskowitz et al., 2021).

Multiple myeloma

MM is a plasma-cell neoplasm involving the anomalous production of immunoglobulins and consequent lesion formation in target organs. It can cause anemia, hypercalcemia, lytic lesions, and kidney failure. MM makes up 1% of all cancers and 10% of blood cancers. It is the third most common hematological neoplasm (Sung et al., 2021).

First-line treatments can achieve excellent long-term control of the disease, but they cannot cure it. As a result, recurrence and retreatment are frequent (Moreau et al., 2019; Mateos et al., 2022; Sonneveld et al., 2024).

Most of the younger patients who have recently been diagnosed with MM are offered high-dose chemotherapy, rather than auto-BMT (Moreau et al., 2019; Sonneveld et al., 2024), alongside maintenance therapy. Older patients and those with prohibitive comorbidities are offered continuous first-line therapies (Facon et al., 2021). Overall MM survival rates have improved internationally with the greater availability of and access to new therapies, such as those implemented with anti-CD38 antibodies, CAR T cells, and bispecific antibodies (Puertas et al., 2023). Data for Latin America remains scarce and show poorer outcomes than in developed countries (Hungria et al., 2017).

Chronic myeloproliferative neoplasms

CMPNs are heterogenous. Among them, Philadelphia chromosome (Ph)-negative myeloproliferative disorders are represented by chronic myeloid leukemia (CML). This disease occurs with the translocation of chromosomes 9 and 22, resulting in the creation of the BCR-ABL1 fusion gene in hematopoietic stem cells, which is translocated to the tyrosine-kinase protein, the great orchestrator of cell proliferation related to physiopathology. The treatment of CML has been revolutionized by the discovery of TKIs (O'Brien et al., 2003; Hochhaus et al., 2020; Cross et al., 2023). Today, the overall survival rate for patients with access to these drugs is similar to that of healthy adults in the same age group.

The non-Ph CPMN group encompasses diseases not characterized by specific cytogenetic changes, each of which has its own distinct profile. It includes polycythemia vera (PV), essential thrombocythemia, primary myelofibrosis, chronic myelomonocytic leukemia, and other rarer forms (Khoury et al., 2022).

In general, these pathologies develop with the accelerated proliferation of myeloid (erythrocytic, megakaryocytic, and erythrocytic) lineages. Some forms interface with myelodysplastic syndrome (MDS) neoplasms, with a greater risk of clonal evolution into AL. PV treatment frequently involves bleeding, cytoreduction, and control of the thrombosis risk (Tefferi et al., 2018).

Myelodysplastic syndromes

MDSs form a heterogeneous group of myeloid diseases with varying risks of evolution into bone marrow failure and AML (Garcia-Manero, 2023). Their clinical presentation is marked by the presence of myeloid-series cytopenias, such as anemia, leukopenia (at the expense of granulocytes), and/or thrombocytopenia. The detection of dysplasia on bone marrow evaluation is essential for their diagnosis (Khoury et al., 2022).

The use of prognostic instruments that bring together clinical, cytogenetic, and molecular data is essential to determine the best path forward in these cases, especially for patients who are candidates for therapies involving allo-BMT (Greenberg et al., 2012; Garcia-Manero, 2023; Bernard, 2023). Therapies frequently used for low- and very low-risk MDSs involve the reversal of cytopenia using erythropoietin and granulocyte colony-stimulating factors, as well as the use of immunomodulating drugs and targeted therapies such as luspatercept (Garcia-Manero, 2023).

For high- and very high-risk MDSs, therapies such as hypomethylating agents and chemotherapy followed by allo-BMT achieve better outcomes in candidates for these options (Garcia-Manero, 2023). However, the prognosis for these diseases appears to have evolved very little in recent decades (Gangat et al., 2016).

6 Observatory Results for Hematological Neoplasms

6.1. Descriptive results

For the period of January 2000 to December 2022, we recorded 114,623 new cancer cases in the ACCCC's HCR. Of these, 4,637 (4%) cases were hematological neoplasms; 2,397 (51.7%) were in men and 2,240 (48.3%) were in women (Figure 2).

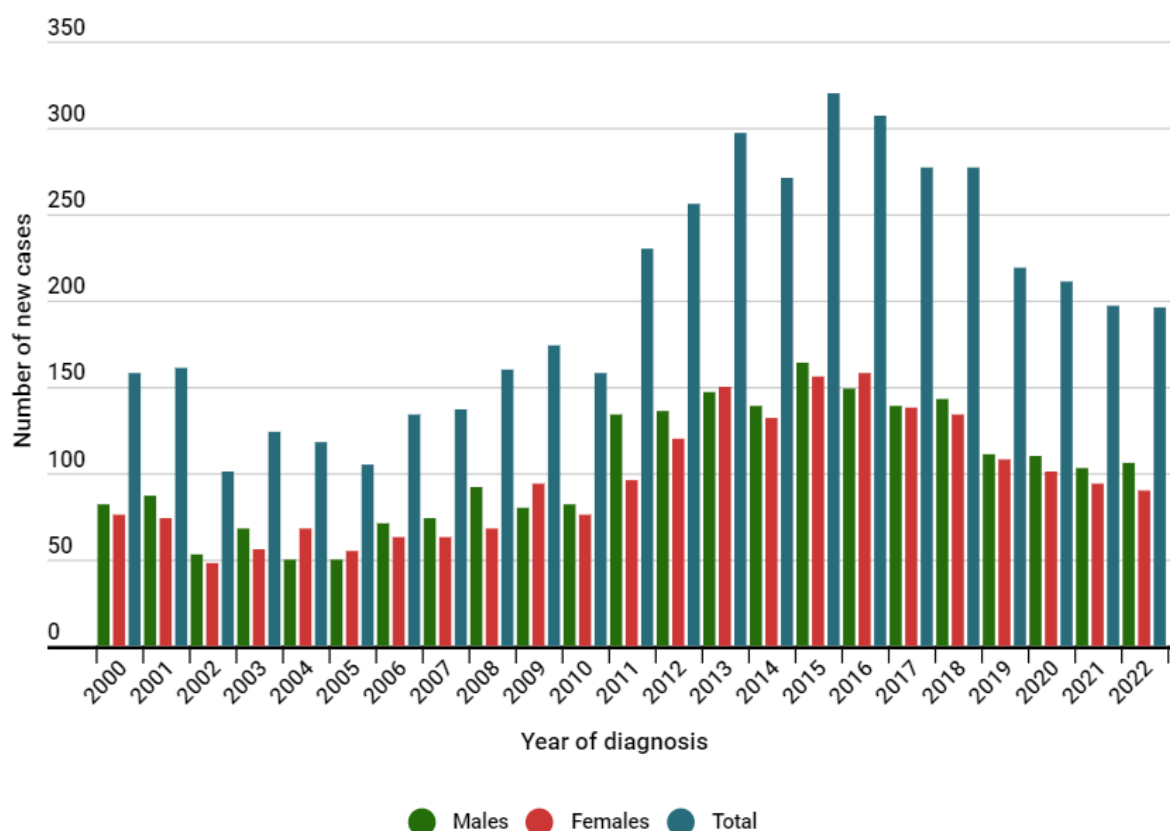


Figure 2. Distribution of new cases of hematological neoplasms overall and by year of diagnosis and patient sex. HCR/A.C. Camargo, 2000 to 2022.

Among males, 12% (n = 291) of the cases occurred in children and adolescents (aged 0-19 years old), 17% (n = 417) in young adults (aged 20-39 years old), 31% (n = 737) in adults aged 40-59 years old, and 39% (n = 942) occurred in those aged ≥ 60 years old. Among females, 10% (n = 213) of the cases occurred in children and adolescents, 19% (n = 419) occurred in young adults, 31% (n = 696) occurred in adults aged 40-59 years old, and 41% (n = 912) occurred in older adults (Figure 3).

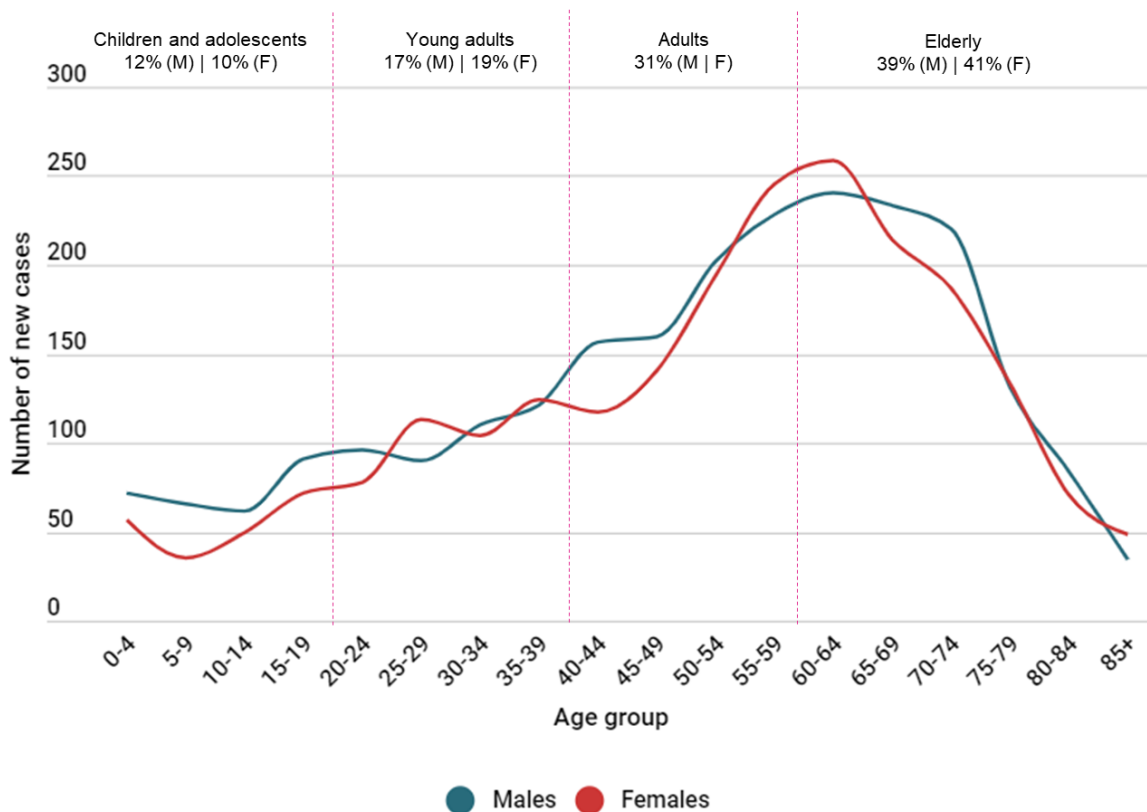


Figure 3. Distribution of the 4,637 cases of hematological neoplasms by age group (children and adolescents, young adults, and older adults) and sex. HCR/A.C.Camargo, 2000 to 2022.

Of all hematological neoplasms, 30% (n = 1,376) were indolent lymphomas, 26% (n = 1,210) were aggressive lymphomas, 15% (n = 689) were Hodgkin lymphomas, 12% (n = 533) were AL, 12% were monoclonal gammopathies (MM, n = 496; plasmacytoma, n = 49), 4% (n = 182) were CMPNs, and 2% (n = 102) were MDSs (Figure 4).

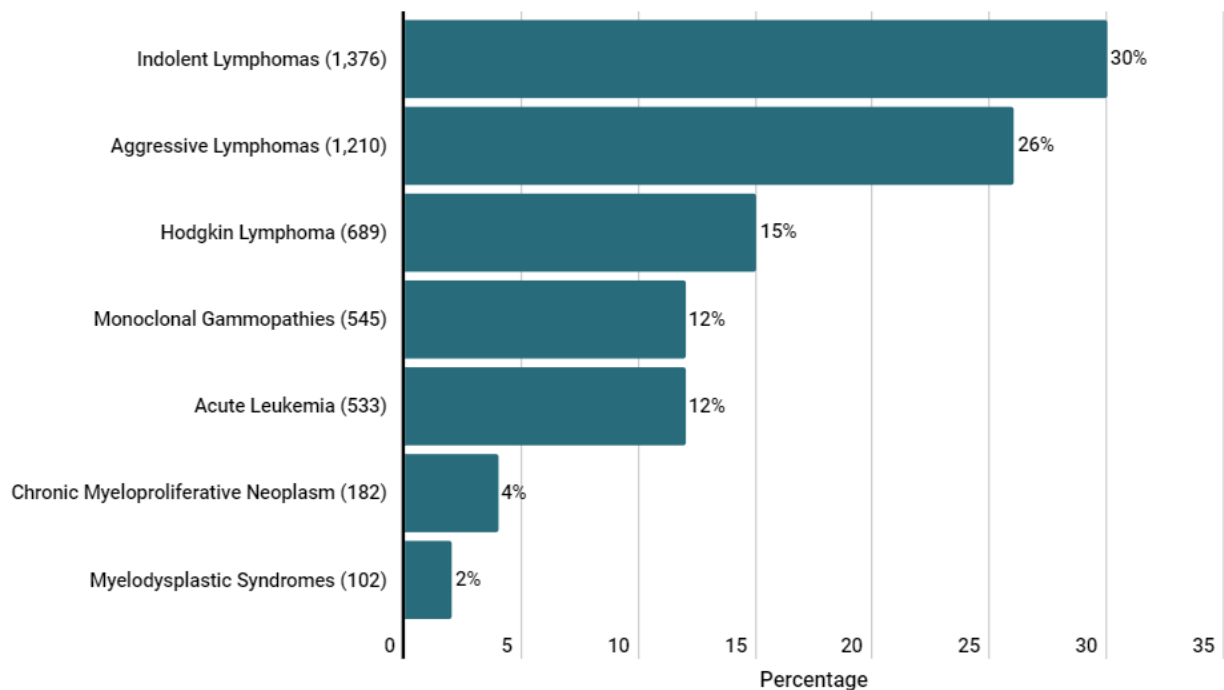


Figure 4. Distribution of 4,637 cases of hematological cancer. HCR/A.C. Camargo, 2000 to 2022.

The majority of the hematological neoplasms occurred mostly in males, with the exception of indolent lymphomas (48.5%) and MDSs (45.1%; Table 2).

Table 2. Distribution of hematological neoplasms by sex and age group, A.C. Camargo Cancer Center, 2000–2022, HCR/A.C. Camargo.

| Variable | Acute leukemia | Aggressive lymphomas | Indolent lymphomas | Hodgkin lymphoma | Monoclonal gammopathies | | CMPN | Myelodysplastic syndromes |
|-------------------|-------------------|-------------------------|-----------------------|---------------------|----------------------------|--------------|-------------|------------------------------|
| | | | | | Multiple myeloma | Plasmacytoma | | |
| | N (%) | N (%) | N (%) | N (%) | N (%) | N (%) | N (%) | N (%) |
| Sex | | | | | | | | |
| Male | 273 (51.2) | 664 (54.9) | 667 (48.5) | 348 (50.5) | 259 (52.2) | 27 (55.1) | 113 (62.1) | 46 (45.1) |
| Female | 260 (48.8) | 546 (45.1) | 709 (51.5) | 341 (49.5) | 237 (47.8) | 22 (44.9) | 69 (37.9) | 56 (54.9) |
| Age group (years) | | | | | | | | |
| 0–19 | 224 (42.0) | 96 (7.9) | 33 (2.4) | 129 (18.7) | - | 15 (30.6) | 7 (3.8) | 15 (14.7) |
| 20–59 | 190 (35.6) | 576 (47.6) | 659 (47.9) | 492 (71.4) | 207 (41.7) | 27 (55.1) | 107 (58.8) | 22 (21.6) |
| 60+ | 119 (22.3) | 538 (44.5) | 684 (49.7) | 68 (9.9) | 289 (58.3) | 7 (14.3) | 68 (63.7) | 65 (63.7) |
| Total | 533 (100.0) | 1,210 (100.0) | 1,376 (100.0) | 689 (100.0) | 496 (100.0) | 49 (100.0) | 182 (100.0) | 102 (100.0) |

CMPN, chronic myeloproliferative neoplasm.

6.2. Survival results

Overall survival by period

During the period of 2000-2019, the greatest probability of 5-year survival was observed for Hodgkin lymphoma (88.0%), followed by indolent lymphomas (82.1%). The lowest probabilities were for AL (51.1%) and MDSs (48.6%; Figure 5, Table 3).

Table 3. Estimated probability of 5-year overall survival of hematological neoplasms (both sexes). Hospital Cancer Registry, A.C.Camargo Cancer Center, 2000–2019.

| Hematological group | 2000–2019 | |
|--------------------------------------|-----------|-----------------------|
| | Deaths/N | % survival at 5-years |
| Acute leukemia | 225/467 | 51.1 |
| Aggressive lymphomas | 336/1042 | 66.5 |
| Indolent lymphomas | 209/1232 | 82.1 |
| Hodgkin lymphoma | 67/598 | 88.0 |
| Multiple myeloma | 169/403 | 56.0 |
| Chronic myeloproliferative neoplasms | 31/157 | 79.5 |
| Myelodysplastic syndromes | 42/85 | 48.6 |

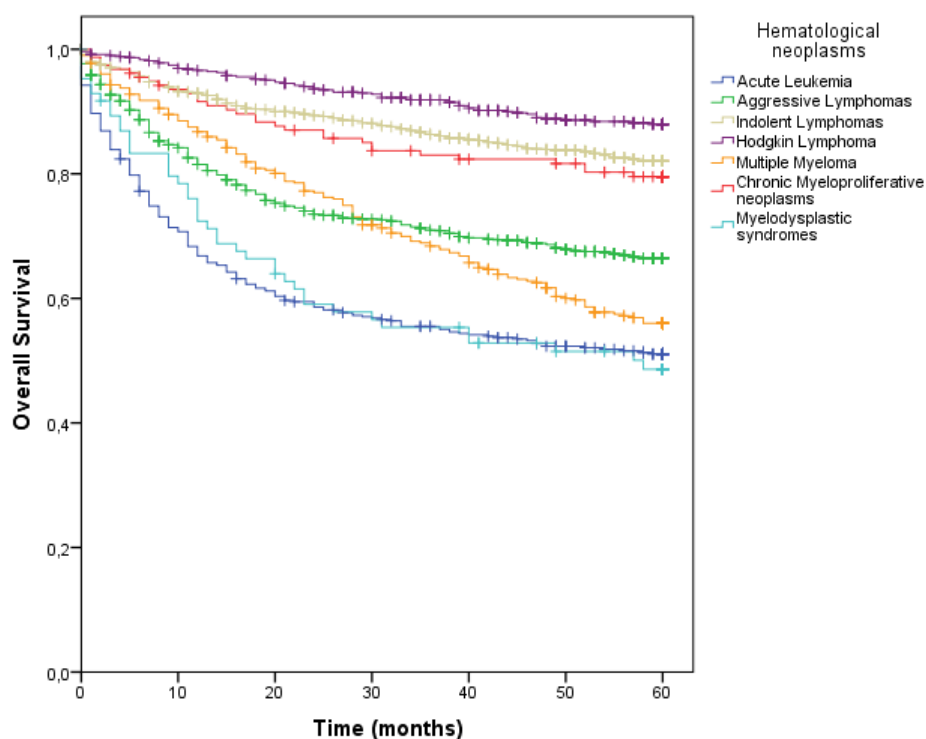


Figure 5. Probability of 5-year overall survival for selected hematological neoplasms, 2000-2019. HCR/A.C.Camargo.

Overall survival by sex

Significant sex differences in the 5-year survival rate were observed for indolent lymphomas (84.3% for women vs. 79.7% for men, $p = 0.032$) and CMPNs (89.7% for women vs. 73.3% for men, $p = 0.019$; Table 4, Figures 6-10).

Table 4. Probability of 5-year overall survival of hematological neoplasms by sex. Hospital Cancer Registry/A.C. Camargo Cancer Center, 2000–2019

| Hematological neoplasm | Males | | Females | | Log-rank test p value |
|--------------------------------------|----------|------|----------|------|-------------------------|
| | Deaths/N | % | Deaths/N | % | |
| Acute leukemia | 121/243 | 49.5 | 104/224 | 52.8 | 0.612 |
| Aggressive lymphomas | 191/568 | 65.0 | 145/474 | 68.2 | 0.384 |
| Indolent lymphomas | 115/597 | 79.7 | 94/635 | 84.3 | 0.032 |
| Hodgkin lymphoma | 31/304 | 89.2 | 36/294 | 86.7 | 0.436 |
| Multiple myeloma | 81/205 | 58.8 | 88/198 | 53.1 | 0.333 |
| Chronic myeloproliferative neoplasms | 25/97 | 73.3 | 06/60 | 89.7 | 0.019 |
| Myelodysplastic syndromes | 23/37 | 36.2 | 19/48 | 58.8 | 0.094 |

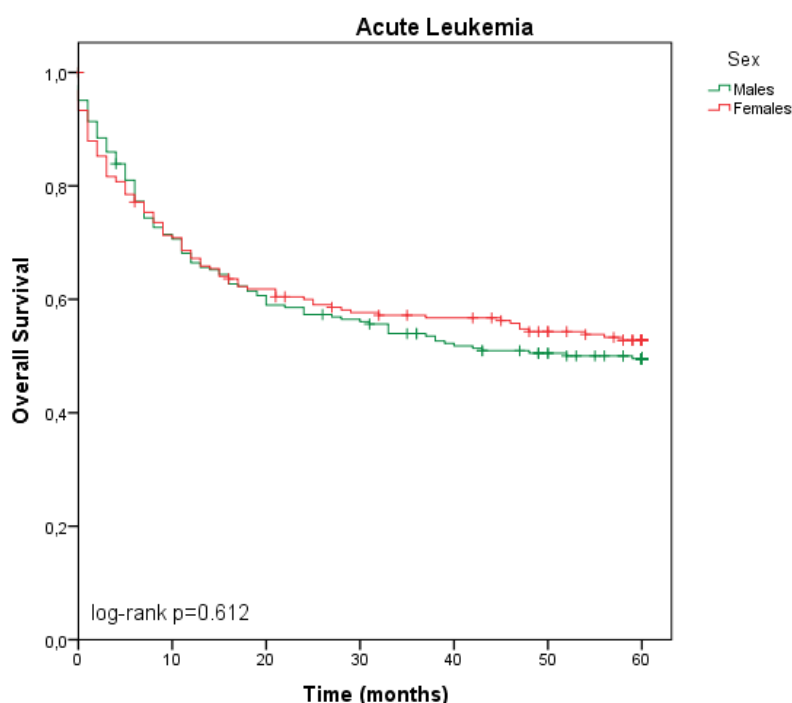


Figure 6. Probability of 5-year overall survival of acute leukemia by sex, 2000–2019. HCR/A.C. Camargo.

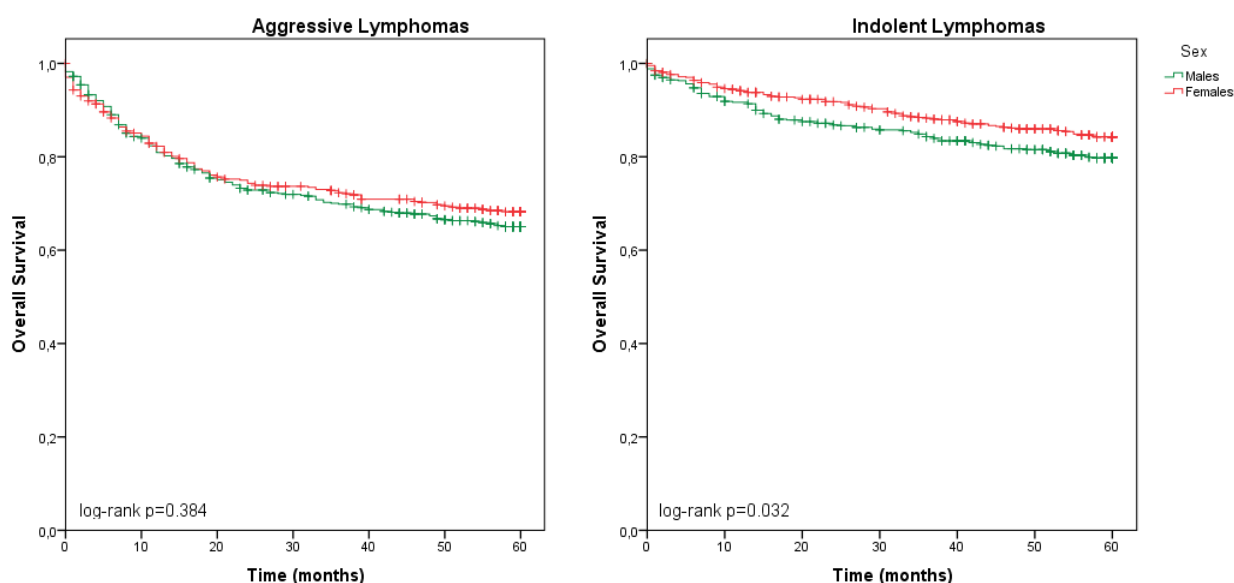


Figure 7. Probability of 5-year overall survival of aggressive and indolent lymphomas by sex, 2000-2019. HCR/A.C.Camargo.

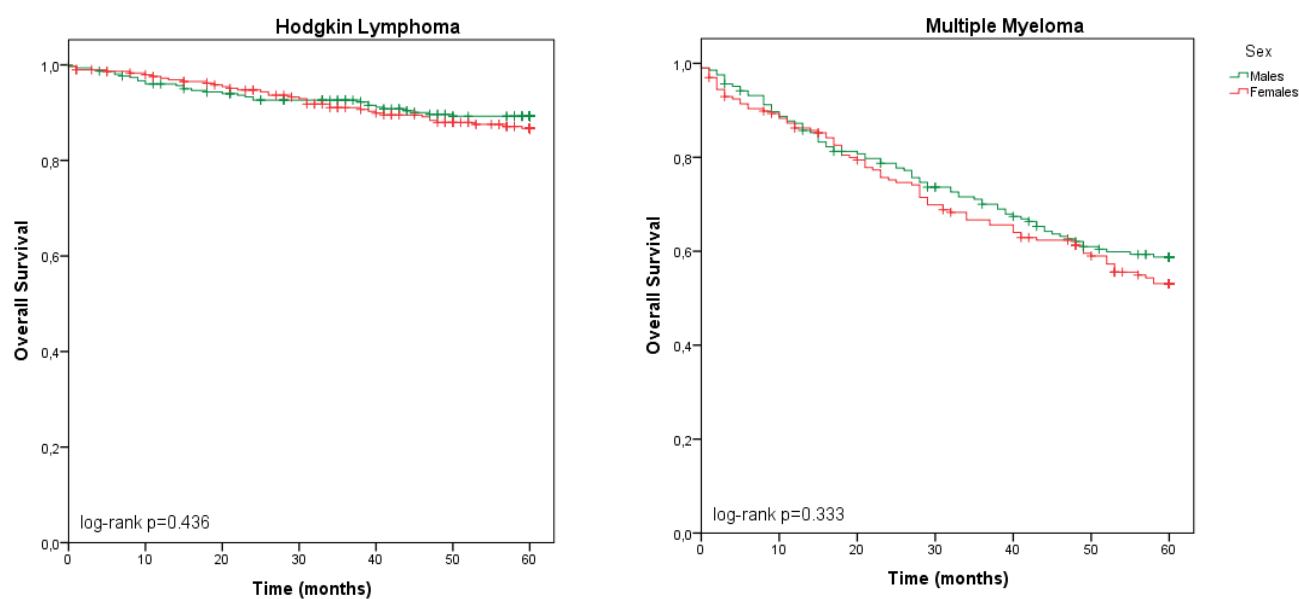


Figure 8. Probability of 5-year overall survival of Hodgkin lymphoma by sex, 2000-2019. HCR/A.C.Camargo.

Figure 9. Probability of 5-year overall survival of multiple myeloma by sex, 2000-2019. HCR/A.C.Camargo.

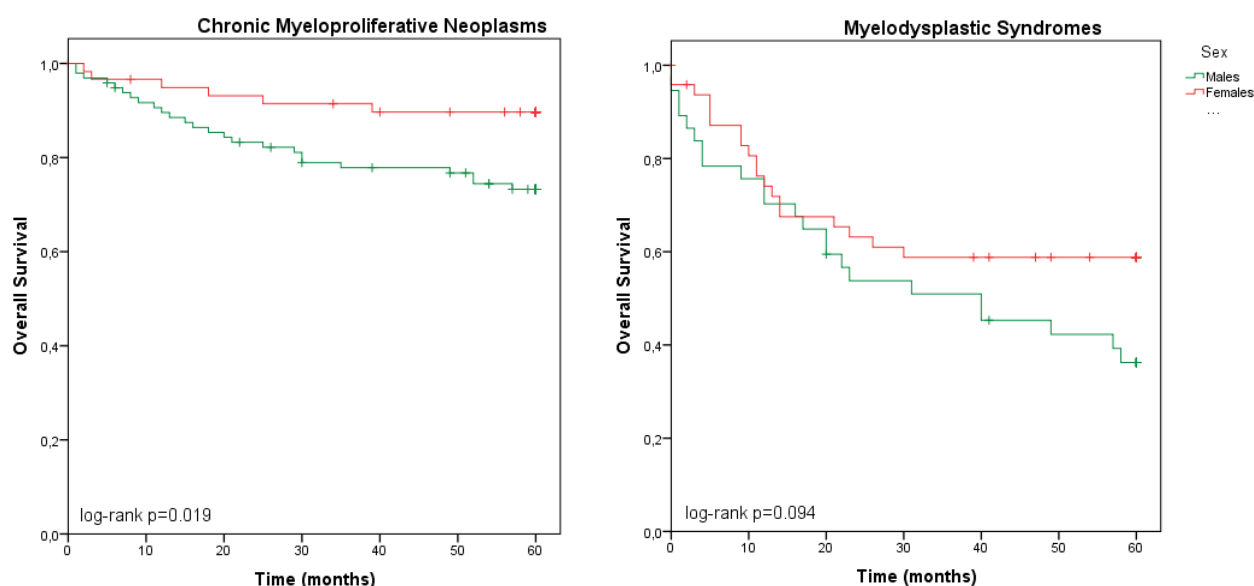


Figure 10. Probability of 5-year overall survival of chronic myeloproliferative neoplasms and myelodysplastic syndromes by sex, 2000-2019. HCR/A.C. Camargo.

Overall survival by age group

Overall survival of the hematological neoplasms for both sexes revealed significant differences among age groups (0-19, 20-59, and ≥ 60 years), except for MDSs (Table 5).

Table 5. Probability of 5-year overall survival of hematological neoplasms for both sexes by age group. Hospital Cancer Registry, A.C. Camargo Cancer Center, 2000-2019.

| Hematological neoplasm | 5-year overall probability of survival | | | | | | Log-rank test |
|--------------------------------------|--|------|--------------|------|----------|------|------------------|
| | 0–19 | | 20–59 | | 60+ | | |
| | Deaths/ N | % | Deaths/ N | % | Deaths/N | % | <i>p</i> value |
| Acute leukemia | 68/218 | 68.7 | 85/156 | 44.1 | 72/93 | 19.5 | <0.001 |
| Aggressive lymphomas | 17/92 | 81.3 | 126/506 | 74.2 | 193/444 | 54.5 | <0.001 |
| Indolent lymphomas | 10/32 | 67.5 | 64/601 | 88.7 | 135/599 | 76.2 | <0.001 |
| Hodgkin lymphoma | 14/123 | 88.0 | 35/424 | 91.1 | 18/51 | 62.5 | <0.001 |
| Multiple myeloma | - | - | 53/168 | 66.8 | 116/235 | 48.3 | <0.001 |
| Chronic myeloproliferative neoplasms | 2/7 | 71.4 | 8/92 | 91.0 | 21/58 | 61.6 | <0.001 |
| Myelodysplastic syndromes | 7/14 | 50.0 | 7/17 | 58.2 | 28/54 | 45.0 | 0.661 |

Overall survival by period

5-year overall survival rates for the hematological neoplasms in 5-year intervals between 2000 and 2019 are shown in Table 6 and Figures 11-15.

Table 6. 5-year overall probability of survival of hematological neoplasms for both sexes by 5-year interval. Hospital Cancer Registry, A.C.Camargo Cancer Center, 2000–2019.

| Hematological neoplasms | 5-year overall probability of survival | | | | | | | | Log-rank test |
|--------------------------------------|--|------|-----------|------|-----------|------|-----------|------|------------------|
| | 2000–2004 | | 2005–2009 | | 2010–2014 | | 2015–2019 | | |
| | Deaths/N | % | Deaths/N | % | Deaths/N | % | Deaths/N | % | |
| Acute leukemia | 59/139 | 57.3 | 47/85 | 44.6 | 57/94 | 38.7 | 62/149 | 56.7 | 0.003 |
| Aggressive lymphomas | 65/178 | 63.1 | 65/207 | 68.6 | 116/314 | 61.7 | 90/343 | 71.5 | 0.023 |
| Indolent lymphomas | 59/158 | 62.6 | 28/212 | 86.7 | 57/407 | 85.0 | 65/455 | 84.1 | <0.001 |
| Hodgkin lymphoma | 17/116 | 85.3 | 15/108 | 86.0 | 25/189 | 85.9 | 10/185 | 93.2 | 0.081 |
| Multiple myeloma | 32/42 | 23.8 | 34/61 | 44.3 | 46/133 | 63.4 | 57/167 | 62.9 | <0.001 |
| Chronic myeloproliferative neoplasms | 6/13 | 53.8 | 4/19 | 78.9 | 9/49 | 81.3 | 12/76 | 83.0 | 0.054 |
| Myelodysplastic syndromes | 10/16 | 37.5 | 11/18 | 38.9 | 8/26 | 68.6 | 13/25 | 38.8 | 0.053 |

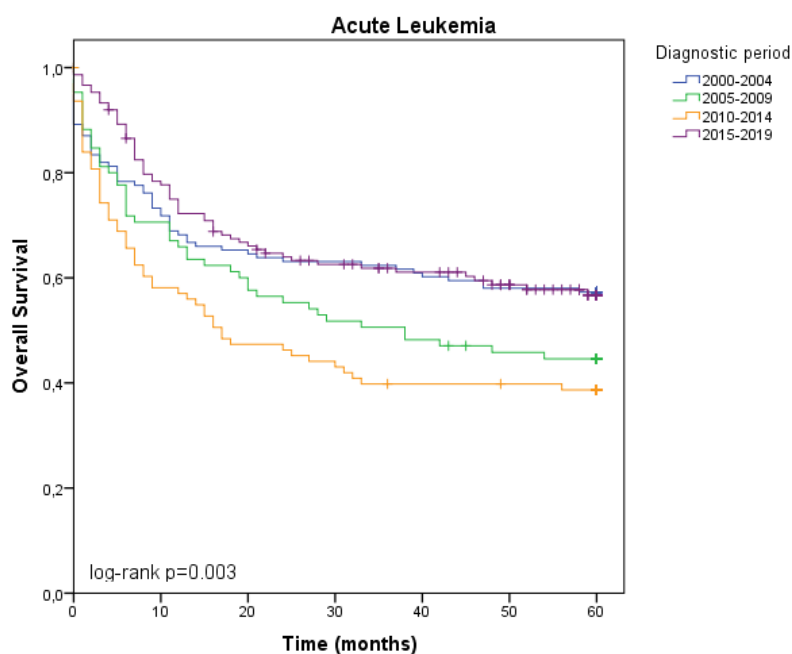


Figure 11. Probability of 5-year overall survival of acute leukemia for both sexes by diagnostic period between 2000 and 2019. HCR/A.C.Cargo.

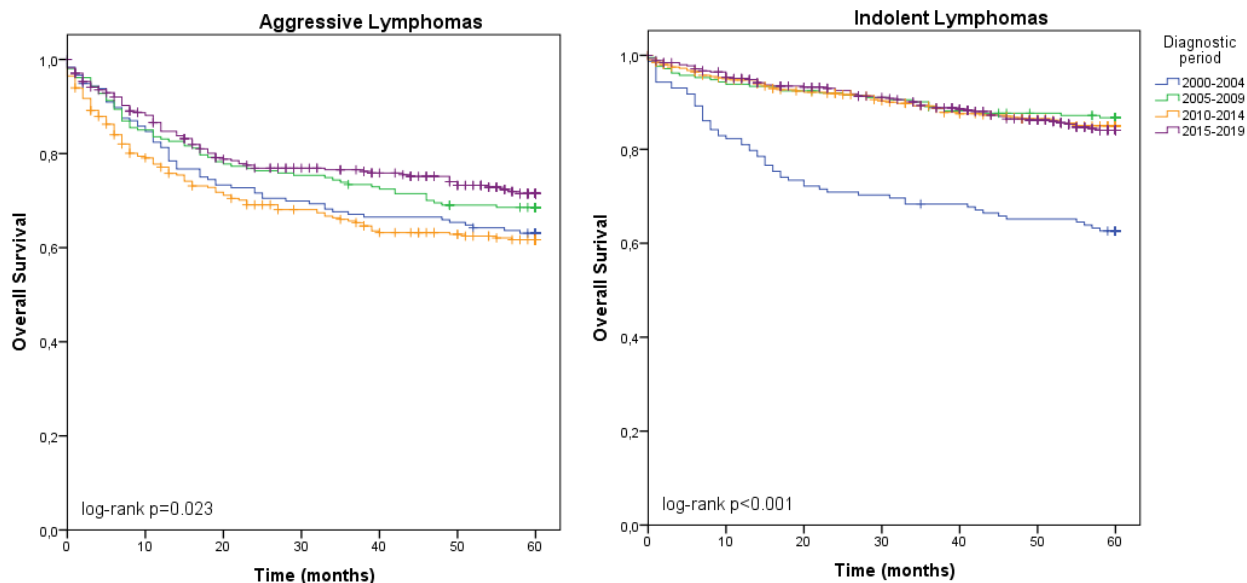


Figure 12. Probability of 5-year overall survival of aggressive and indolent lymphomas for both sexes by diagnostic period between 2000 and 2019. HCR/A.C. Camargo.

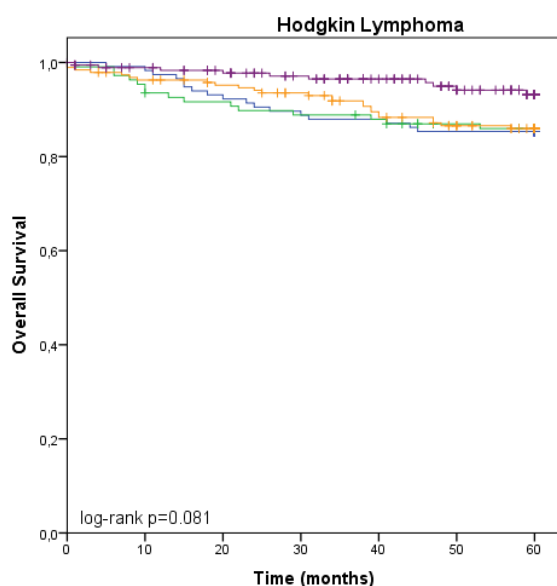


Figure 13. Probability of 5-year overall survival of Hodgkin lymphoma for both sexes by diagnostic period between 2000 and 2019. HCR/A.C. Camargo.

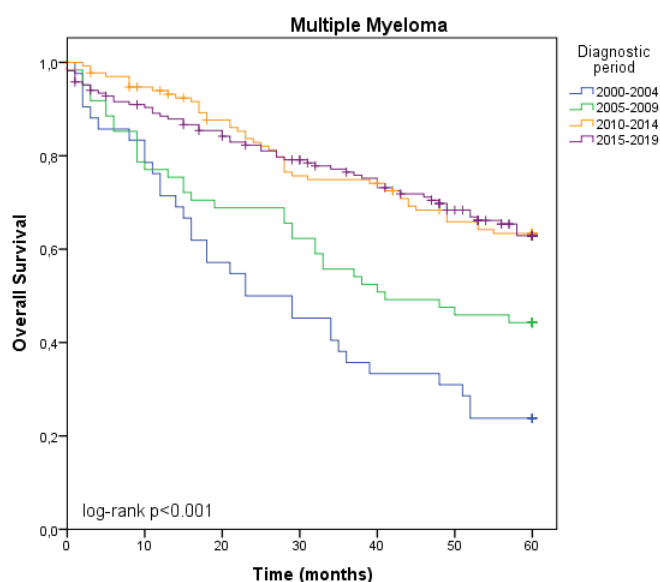


Figure 14. Probability of 5-year overall survival of multiple myeloma for both sexes by diagnostic period between 2000 and 2019. HCR/A.C. Camargo.

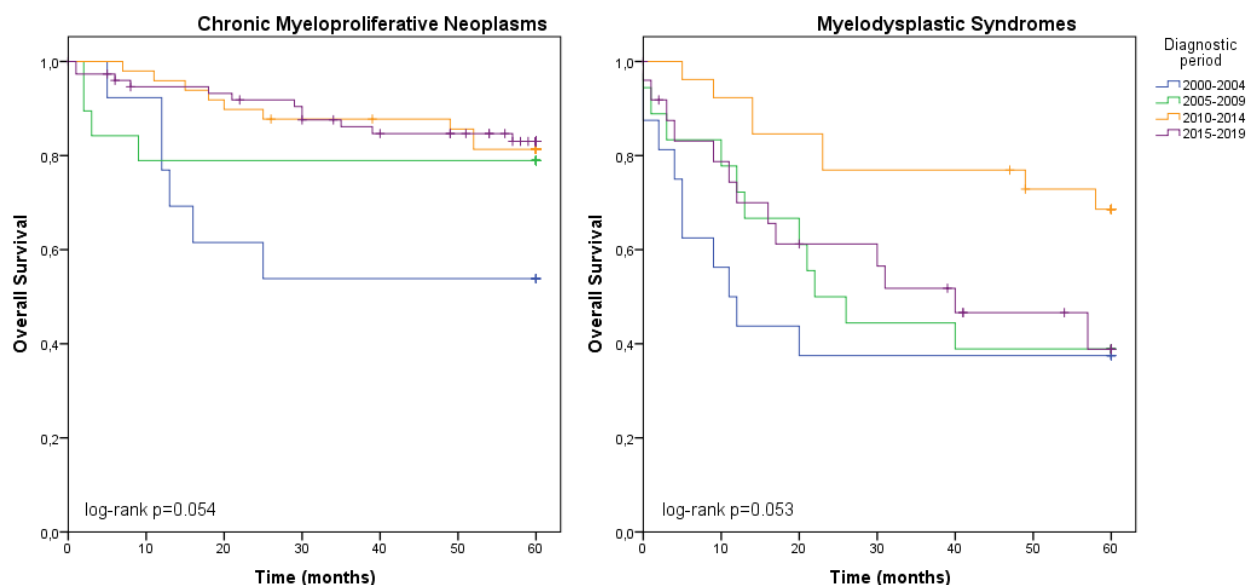


Figure 15. Probability of 5-year overall survival of chronic myeloproliferative neoplasms and myelodysplastic syndromes for both sexes by diagnostic period between 2000 and 2019. HCR/A.C.Camargo.

In the results for the 5-year overall probability of survival by hematological neoplasm for the two 10-year periods between 2000 and 2019, we observed significant increases in survival for indolent lymphomas, MM, CMPNs, and MDSs (Table 7).

Table 7. Probability of 5-year overall survival of hematological neoplasms for both sexes by decade of diagnosis. Hospital Cancer Registry, A.C.Camargo Cancer Center, 2000–2019.

| Hematological neoplasm | 5-year overall probability of survival | | | | Log-rank test |
|--------------------------------------|--|------|--------------|------|------------------|
| | 2000–2009 | | 2010–2019 | | |
| | Deaths/N | % | Deaths/ N | % | |
| Acute leukemia | 106/224 | 52.5 | 119/243 | 49.8 | 0.699 |
| Aggressive lymphomas | 130/385 | 66.0 | 206/657 | 66.8 | 0.908 |
| Indolent lymphomas | 87/370 | 76.4 | 122/862 | 84.6 | <0.001 |
| Hodgkin lymphoma | 32/224 | 85.6 | 35/374 | 89.4 | 0.133 |
| Multiple myeloma | 66/103 | 35.9 | 103/300 | 63.3 | <0.001 |
| Chronic myeloproliferative neoplasms | 10/32 | 68.8 | 21/125 | 82.3 | 0.048 |
| Myelodysplastic syndromes | 21/34 | 38.2 | 21/51 | 55.3 | 0.043 |

Survival improved in both groups, with better results observed for patients who underwent BMT (Table 8, Figure 16).

Table 8. Probability of 5-year overall survival of multiple myeloma by diagnostic period and BMT. Hospital Cancer Registry, A.C. Camargo Cancer Center, 2000–2019.

| Multiple myeloma | 2000–2009 | | 2010–2019 | |
|--|-----------|------|-----------|------|
| | Deaths/N | % | Deaths/N | % |
| Bone Marrow Transplantation (BMT) | | | | |
| No | 53/71 | 25.4 | 75/160 | 49.8 |
| Yes | 13/32 | 59.4 | 28/140 | 78.3 |

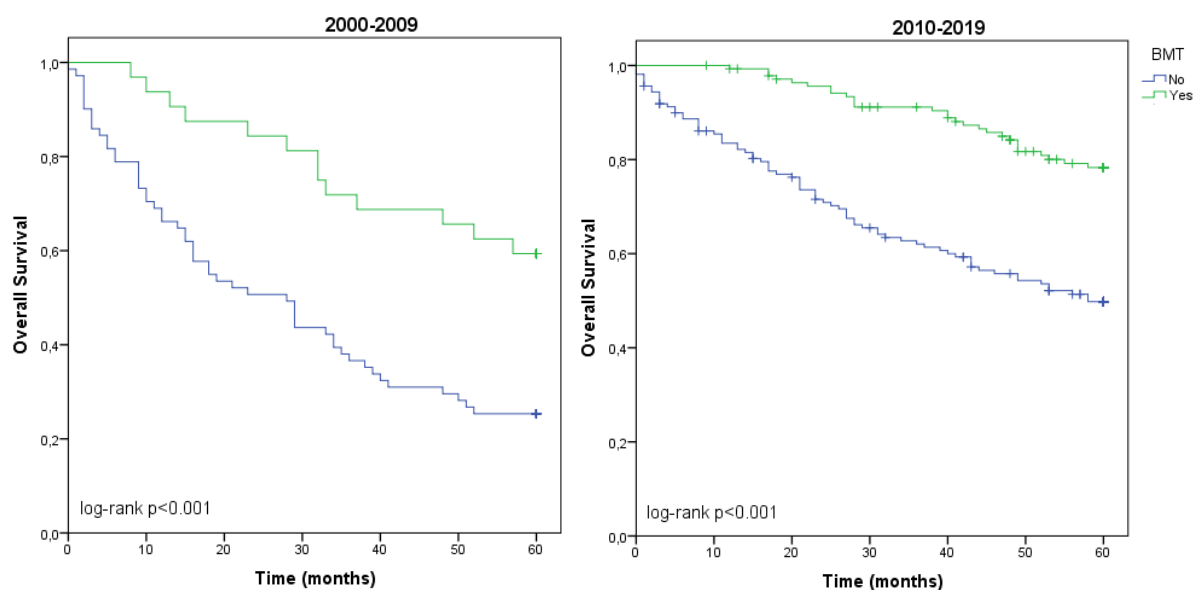


Figure 16. Probability of 5-year overall survival of multiple myeloma for both sexes by diagnostic period and bone marrow transplantation between 2000 and 2019. HCR/A.C. Camargo.

7 Conclusions

Over two decades, from 2000 to 2022, more than 4,600 cases of hematological neoplasms, representing 4% of all cases of cancer, were treated at the Institution. The most common types were lymphomas (indolent, aggressive and Hodgkin), followed by monoclonal gammopathies (MM and plasmacytoma) and AL.

Between 2000 and 2019, the most improvement in overall survival was observed for MM, which is attributable to therapeutic advances and the adoption of BMT as a treatment component. In both periods analyzed, survival results were better for patients who underwent BMT than for those who did not.

Overall survival rates also increased for aggressive and indolent lymphomas, whereas the effective treatment of acute lymphomas remained challenging across the whole period. Survival results for Hodgkin lymphoma were excellent across the whole period.

Many factors could explain these gains in survival, including early diagnosis, the use of new therapies, and the optimization of existing treatments. The implementation of multimodal therapies including chemotherapy, radiation therapy, BMT, immunotherapy, and/or targeted therapies has been fundamental for the significant improvement in survival results for many patients. These advances provide a positive outlook for the continued evolution of survival for these types of cancer.

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10 HCR Database Access

Access to this information is available on the A.C.Camargo's internet through the Tabnet tool at:

<https://tabnet-RHC.accamargo.org.br>



The Hospital Cancer Registry database provides access to the profiles of the patients treated at the ACCCC since 2000. It makes available data on sociodemographic, clinical, and epidemiological characteristics; tumor morphological characteristics, clinical stages, and treatments; and patients' vital status. The data are anonymized and can be consulted via Tabnet, a tool that allows users to build tables according to the following variables of interest: diagnosis/prior treatment, federative unit of birth, federative unit of residence, age group, sex, educational level, and tumor properties (topography, morphology, clinical stage, and treatment).

The consultation of non-anonymized data is permitted only for specific research projects and requires Research Ethics Committee approval.

To request access, please email the named HCR officer (attaching Research Ethics Committee approval).

11 Contacts at the Hospital Cancer Registry

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Our contact information is also available:

1. On the Institutional Intranet (Institucional > Recursos > RHC):
<https://intranet.accamargo.org.br/institucional/recursos/registro-hospitalar-de-cancer-RHC>
2. On the ACCCC HCR webpage:
<https://accamargo.org.br/pesquisa/registro-hospitalar-de-cancer>

References

- A.C.Camargo Cancer Center; Diego Rodrigues Mendonça e Silva, Maria Paula Curado, José Humberto Tavares Guerreiro Fregnani. Observatório do câncer: Registro Hospitalar de Câncer do A.C.Camargo Cancer Center 2000 a 2020. São Paulo: Fundação Antônio Prudente, 2023. Disponível em: https://accamargo.org.br/sites/default/files/2023/05/observatorio_cancer_v1.1.pdf.
- Alaggio R, Amador C, Anagnostopoulos I, Attygalle AD, Araujo IBO, Berti E, et al. The 5th edition of the World Health Organization Classification of Haematolymphoid Tumours: Lymphoid Neoplasms. *Leukemia*. 2022 Jul;36(7):1720-1748.
- Bernard E. Molecular international prognostic scoring system for myelodysplastic syndromes. *NEJM Evid*. 2023;2(6)
- Burger JA, Tedeschi A, Barr PM, Robak T, Owen C, Ghia P, et al.; RESONATE-2 Investigators. Ibrutinib as Initial Therapy for Patients with Chronic Lymphocytic Leukemia. *N Engl J Med*. 2015 Dec 17;373(25):2425-37.
- Brice P, Bastion Y, Lepage E, Brousse N, Haioun C, Moreau P, et al. Comparison in low-tumor-burden follicular lymphomas between an initial no-treatment policy, prednimustine, or interferon alfa: a randomized study from the Groupe d'Etude des Lymphomes Folliculaires. Groupe d'Etude des Lymphomes de l'Adulte. *J Clin Oncol*. 1997 Mar;15(3):1110-7.
- Coifman B, Lepage E, Briere J, Herbrecht R, Tilly H, Bouabdallah R, et al. CHOP chemotherapy plus rituximab compared with CHOP alone in elderly patients with diffuse large-B-cell lymphoma. *N Engl J Med*. 2002 Jan 24;346(4):235-42.
- Connors JM, Jurczak W, Straus DJ, Ansell SM, Kim WS, Gallamini A, et al.; ECHELON-1 Study Group. Brentuximab Vedotin with Chemotherapy for Stage III or IV Hodgkin's Lymphoma. *N Engl J Med*. 2018 Jan 25;378(4):331-344.
- Connors JM, Cozen W, Steidl C, Carbone A, Hoppe RT, Flechtner HH, Bartlett NL. Hodgkin lymphoma. *Nat Rev Dis Primers*. 2020 Jul 23;6(1):61. doi: 10.1038/s41572-020-0189-6. Erratum in: *Nat Rev Dis Primers*. 2021 Oct 20;7(1):79.
- Cross NCP, Ernst T, Branford S, Cayuela JM, Deininger M, Fabarius A, et al. European LeukemiaNet laboratory recommendations for the diagnosis and management of chronic myeloid leukemia. *Leukemia*. 2023 Nov;37(11):2150-2167.

DiNardo CD, Jonas BA, Pullarkat V, Thirman MJ, Garcia JS, Wei AH, et al. Azacitidine and Venetoclax in Previously Untreated Acute Myeloid Leukemia. *N Engl J Med*. 2020 Aug 13;383(7):617-629.

Eichenauer DA, Engert A. Nodular lymphocyte-predominant Hodgkin lymphoma: a unique disease deserving unique management. *Hematology Am Soc Hematol Educ Program*. 2017 Dec 8;2017(1):324-328. doi: 10.1182/asheducation-2017.1.324.

Facon T, Kumar SK, Plesner T, Orlowski RZ, Moreau P, Bahlis N, et al. Daratumumab, lenalidomide, and dexamethasone versus lenalidomide and dexamethasone alone in newly diagnosed multiple myeloma (MAIA): overall survival results from a randomised, open-label, phase 3 trial. *Lancet Oncol*. 2021 Nov;22(11):1582-1596.

Gangat N, Patnaik MM, Begna K, Al-Kali A, Litzow MR, Ketterling RP, et al. Survival trends in primary myelodysplastic syndromes: a comparative analysis of 1000 patients by year of diagnosis and treatment. *Blood Cancer J*. 2016 Apr 8;6(4):e414.

Garcia-Manero G. Myelodysplastic syndromes: 2023 update on diagnosis, risk-stratification, and management. *Am J Hematol*. 2023 Aug;98(8):1307-1325.

Gökbuget N, Boissel N, Chiaretti S, Dombret H, Doubek M, Gelding AK, et al. Management of ALL in Adults: 2023 ELN Recommendations from a European Expert Panel. *Blood*. 2024 Feb 2;blood.2023023568.

Greenberg PL, Tuechler H, Schanz J, Sanz G, Garcia-Manero G, Solé F, et al. Revised international prognostic scoring system for myelodysplastic syndromes. *Blood*. 2012 Sep 20;120(12):2454-65.

Health Organization Classification of Haematolymphoid Tumours: Myeloid and Histiocytic/Dendritic Neoplasms. *Leukemia*. 2022 Jul;36(7):1703-1719.

Hochhaus A, Baccarani M, Silver RT, Schiffer C, Apperley JF, Cervantes F, et al. European LeukemiaNet 2020 recommendations for treating chronic myeloid leukemia. *Leukemia*. 2020 Apr;34(4):966-984.

Horwitz S, O'Connor OA, Pro B, Illidge T, Fanale M, Advani R, et al.; ECHELON-2 Study Group. Brentuximab vedotin with chemotherapy for CD30-positive peripheral T-cell lymphoma (ECHELON-2): a global, double-blind, randomised, phase 3 trial. *Lancet*. 2019 Jan 19;393(10168):229-240.

Hungria VT, Maiolino A, Martinez G, Duarte GO, Bittencourt R, Peters L, et al.; International Myeloma Working Group Latin America. Observational study of multiple myeloma in Latin America. *Ann Hematol*. 2017 Jan;96(1):65-72.

Jain N, Keating M, Thompson P, Ferrajoli A, Burger J, Borthakur G, et al. Ibrutinib and Venetoclax for First-Line Treatment of CLL. *N Engl J Med*. 2019 May 30;380(22):2095-2103.

Khoury JD, Solary E, Ablan O, Akkari Y, Alaggio R, Apperley JF, et al. The 5th edition of the World Health Organization Classification of Haematolymphoid Tumours: Myeloid and Histiocytic/Dendritic Neoplasms. *Leukemia*. 2022 Jul;36(7):1703-1719.

Locke FL, Miklos DB, Jacobson CA, Perales MA, Kersten MJ, Oluwole OO, et al.; ALL ZUMA-7 Investigators and Contributing Kite Members. Axicabtagene Ciloleucel as Second-Line Therapy for Large B-Cell Lymphoma. *N Engl J Med*. 2022 Feb 17;386(7):640-654.

Marcus R, Imrie K, Belch A, et al. CVP chemotherapy plus rituximab compared with CVP as first-line treatment for advanced follicular lymphoma. *Blood*. 2005;105:1417-1423.

Mateos MV, Weisel K, De Stefano V, Goldschmidt H, Delforge M, Mohty M, et al. LocoMMotion: a prospective, non-interventional, multinational study of real-life current standards of care in patients with relapsed and/or refractory multiple myeloma. *Leukemia*. 2022 May;36(5):1371-1376.

Ministério da Saúde, Gabinete do Ministro. Portaria nº 3.535, de 2 de setembro de 1998 – estabelece os critérios para cadastramento de centros de atendimento em oncologia. Disponível em: https://bvsms.saude.gov.br/bvs/saudelegis/gm/1998/prt3535_02_09_1998_revog.html Acesso em 30 ago. 2022.

Ministério da Saúde, Instituto Nacional de Câncer José Alencar Gomes da Silva. Registros hospitalares de câncer: planejamento e gestão. 2ª ed. Rio de Janeiro: INCA, 2010.

Moreau P, Attal M, Hulin C, Arnulf B, Belhadj K, Benboubker L, et al. Bortezomib, thalidomide, and dexamethasone with or without daratumumab before and after autologous stem-cell transplantation for newly diagnosed multiple myeloma (CASSIOPEIA): a randomised, open-label, phase 3 study. *Lancet*. 2019 Jul 6;394(10192):29-38.

Moskowitz AJ, Shah G, Schöder H, Ganesan N, Drill E, Hancock H, et al. Phase II Trial of Pembrolizumab Plus Gemcitabine, Vinorelbine, and Liposomal Doxorubicin as Second-Line Therapy for Relapsed or Refractory Classical Hodgkin Lymphoma. *J Clin Oncol*. 2021 Oct 1;39(28):3109-3117.

National Program of Cancer Registries. Registry Plus™ Link Plus Features and Future Plans [Internet]. Atlanta: Centers for Disease Control and Prevention; 2018 [acessado em 11 jan. 2019]. Disponível em: https://www.cdc.gov/cancer/npcr/tools/registryplus/lp_features.htm Disponível em:

O'Brien SG, Guilhot F, Larson RA, Gathmann I, Baccarani M, Cervantes F, et al.; IRIS Investigators. Imatinib compared with interferon and low-dose cytarabine for newly diagnosed chronic-phase chronic myeloid leukemia. *N Engl J Med*. 2003 Mar 13;348(11):994-1004.

O'Connor OA, Horwitz S, Masszi T, Van Hoof A, Brown P, Doorduijn J, et al. Belinostat in Patients With Relapsed or Refractory Peripheral T-Cell Lymphoma: Results of the Pivotal Phase II BELIEF (CLN-19) Study. *J Clin Oncol*. 2015 Aug 10;33(23):2492-9.

Organização Mundial da Saúde. CID-O Classificação internacional de Doenças para Oncologia. 3ª Ed. EDUSP. São Paulo, 2012.

Puertas B, González-Calle V, Sobejano-Fuertes E, Escalante F, Queizán JA, Báñez A, et al. Novel Agents as Main Drivers for Continued Improvement in Survival in Multiple Myeloma. *Cancers (Basel)*. 2023 Mar 2;15(5):1558.

Salles G, Seymour JF, Offner F, López-Guillermo A, Belada D, Xerri L, et al. Rituximab maintenance for 2 years in patients with high tumour burden follicular lymphoma responding to rituximab plus chemotherapy (PRIMA): a phase 3, randomised controlled trial. *Lancet*. 2011 Jan 1;377(9759):42-51.

Secretaria de Estado da Saúde de São Paulo (SESSP). Fundação Oncocentro de São Paulo. Resolução SS-15, de 27 de janeiro de 2000. Disponível: <http://www.fosp.saude.sp.gov.br/publicacoes/resolucaoss15> Acesso em 15 fev. 2021.

Secretaria de Estado da Saúde de São Paulo (SESSP). Fundação Oncocentro de São Paulo. Registro Hospitalar de Câncer: conceitos, rotinas e instruções de preenchimento. 2ª ed. São Paulo, 2013. Disponível em: <http://www.fosp.saude.sp.gov.br/publicacoes/sisCHR>. Acesso em 06 fev. 2022.

Secretaria de Estado da Saúde de São Paulo (SESSP). Fundação Oncocentro de São Paulo. Hospital Cancer Register de São Paulo: análise dos dados (janeiro/2000 a março/2022) e indicadores de qualidade (2000 a 2016). Disponível em: <http://www.fosp.saude.sp.gov.br/fosp/diretoria-adjunta-de-informacao-e-epidemiologia/HCR-registro-hospitalar-de-cancer/dados-de-cancer/> Acesso em 06 fev. 2022.

Sonneveld P, Dimopoulos MA, Boccadoro M, Quach H, Ho PJ, Beksac M, et al.; PERSEUS Trial Investigators. Daratumumab, Bortezomib, Lenalidomide, and Dexamethasone for Multiple Myeloma. *N Engl J Med*. 2024 Jan 25;390(4):301-313.

Sung H, Ferlay J, Siegel RL, Laversanne M, Soerjomataram I, Jemal A, Bray F. Global Cancer Statistics 2020: GLOBOCAN Estimates of Incidence and Mortality Worldwide for 36 Cancers in 185 Countries. *CA Cancer J Clin*. 2021 May;71(3):209-249.

Swerdlow SH, Campo E, Pileri SA, et al. The 2016 revision of the World Health Organization classification of lymphoid neoplasms. *Blood*. 2016;127:2375-90.

Tebbi CK. Etiology of Acute Leukemia: A Review. *Cancers (Basel)*. 2021 May 8;13(9):2256.

Tefferi A, Vannucchi AM, Barbui T. Polycythemia vera treatment algorithm 2018. *Blood Cancer J*. 2018 Jan 10;8(1):3.

Tilly H, Morschhauser F, Sehn LH, Friedberg JW, Trněný M, Sharman JP, et al. Polatuzumab Vedotin in Previously Untreated Diffuse Large B-Cell Lymphoma. *N Engl J Med*. 2022 Jan 27;386(4):351-363.

Zain JM, Hanona P. Aggressive T-cell lymphomas: 2021 Updates on diagnosis, risk stratification and management. *Am J Hematol*. 2021 Aug 1;96(8):1027-1046.

Wilcox RA. Cutaneous T-cell lymphoma: 2014 update on diagnosis, risk-stratification, and management. *Am J Hematol*. 2014;89:837-851.

Appendices



Secretaria de Estado da Saúde
Fundação Oncocentro de São Paulo
Registro Hospitalar de Câncer

FICHA DE ADMISSÃO

Data : 20/02/2013
pág.: 1 / 2

Instituição: _____ Número RHC: _____

IDENTIFICAÇÃO DO PACIENTE

Prontuário: _____ Categoria Atend.: ☐ 1. SUS / 2. Convênio / 3. Particular Data de Nascimento: ____ / ____ / ____
Sexo: ☐ 1. Masculino / 2. Feminino Documento: ☐ 1. PIS/PASEP / 2. RG / 3. Certidão de Nascimento N°: _____ Idade: _____
4. CPF / 5. Cartão SUS

Nome: _____

Nome da mãe: _____

Escolaridade: ☐ 1. Analfabeto / 2. Ens. Fundamental incompleto / 3. Ens. Fundamental completo / 4. Ensino Médio completo / 5. Superior completo / 9. Ignorado

Estado/País de nascimento: _____

Residência atual

Logradouro: _____ Nº: _____
Complemento: _____ Tel.: _____ CEP: _____ - _____
Cidade: _____ UF: _____

SITUAÇÃO DO PACIENTE À ADMISSÃO

Data da primeira consulta: ____ / ____ / ____ Clínica de atendimento: _____

Diagnóstico/tratamento anterior: 1. sem diagnóstico/sem tratamento / 2. com diagnóstico/sem tratamento / 3. com diagnóstico/com tratamento / 4. outros

Instituição de origem: _____

INFORMAÇÕES SOBRE A DOENÇA

Data do 1º diagnóstico: ____ / ____ / ____

Base para realização do diagnóstico: 1. exame clínico / 2. recursos auxiliares não microscópicos / 3. confirmação microscópica / 9. sem informação

Caracterização do tumor principal

Localização primária: _____ Lateralidade: ☐ 0 - Não se aplica
1 - Direita
2 - Esquerda

Tipo histológico: _____

Estadio clínico:

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 Outro estadiamento: _____
Fatores de Risco:

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Estadio pós-cirúrgico: PT:

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 pM: _____

Metástases: _____

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Secretaria de Estado da Saúde
Fundação Oncocentro de São Paulo
Registro Hospitalar de Câncer

FICHA DE ADMISSÃO

Data : 20/02/2013
pág.: 2 / 2

Instituição: _____ Número RHC: _____

EVENTOS DESSE ATENDIMENTO

Data de início do tratamento no hospital: ____ / ____ / ____

Tratamento recebido no Hospital:

- ☐ Nenhum ☐ Cirurgia
☐ Radioterapia ☐ Quimioterapia
 Hormonioterapia ☐ TMO
 Imunoterapia ☐ Outros

Fora do hospital antes da admissão:

- ☐ Nenhum ☐ Cirurgia
☐ Radioterapia ☐ Quimioterapia
 Hormonioterapia ☐ TMO
 Imunoterapia ☐ Outros
☐ Sem informação

Fora do hospital durante e/ou após admissão:

- ☐ Nenhum ☐ Cirurgia
☐ Radioterapia ☐ Quimioterapia
 Hormonioterapia ☐ TMO
 Imunoterapia ☐ Outros
☐ Sem informação

Razão para não realização do tratamento no hospital :

- ☐ 1. Recusa do tratamento/ 2. Doença avançada, Falta de condições clínicas/ 3. Outras doenças associadas
 4. Abandono tratamento/ 5. Óbito por câncer/ 6. Óbito por outras causas, SOE/ 7. Outras/ 8. Não se aplica
 9. Sem informação

Estado da doença ao final do tratamento:

- ☐ 1. Sem evidência da doença / 2. Remissão parcial / 3. Doença estável/ 4. Doença em progressão
 5. Fora de possibilidade/ 6. Óbito por câncer/ 7. Óbito por outras causas, SOE/ 8. Tratamento não concluído
 9. Não se aplica/ 10. Sem informação

Data do óbito: ____ / ____ / ____ Data de preenchimento: ____ / ____ / ____ Registrador: _____

OBSERVAÇÕES



Secretaria de Estado da Saúde
Fundação Oncocentro de São Paulo
Registro Hospitalar de Câncer

Data : 20/02/2013

FICHA DE SEGUIMENTO

Instituição: _____

Número RHC: _____

IDENTIFICAÇÃO DO PACIENTE

Prontuário: _____ Documento: ☐ 1. PIS/PASEP / 2. RG / 3. Certidão de Nascimento N°: _____
4. CPF / 5. Cartão SUS
Dt. Nascimento: ____ / ____ / ____ Sexo: ☐ 1. Masculino / 2. Feminino
Nome: _____

ÚLTIMA INFORMAÇÃO DO PACIENTE

Situação Atual: ☐ 1. Vivo com câncer / 2. Vivo, SOE / 3. Óbito por câncer / 4. Óbito, SOE

Data da Informação: ____ / ____ / ____

Tratamento Realizado no hospital: Nenhum Cirurgia Radioterapia Quimioterapia
Hormonioterapia TMO Imunoterapia Outros

Tratamento Realizado fora do hospital: ☐ Nenhum ☐ Cirurgia ☐ Radioterapia ☐ Quimioterapia ☐ Hormonioterapia
☐ TMO ☐ Imunoterapia ☐ Outros ☐ Sem informação

Recidiva: ☐ 1. Local / 2. Regional / 3. Não / 9. Sem Informação

Data da Recidiva/Metástase: ____ / ____ / ____

Metástase:

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Data do Óbito: ____ / ____ / ____

Data do Preenchimento: ____ / ____ / ____

Registrador: _____

OBSERVAÇÕES



GABARITO DO ARQUIVO DBF

| CAMPO | DESCRIÇÃO |
|-------------------|---|
| IBGE | Código do município brasileiro, segundo o IBGE, onde reside o paciente |
| ESCOLARI | Escolaridade: 1 – Analfabeto 2 – Ensino Fundamental Incompleto 3 – Ensino Fundamental completo 4 – Ensino Médio Completo 5 – Ensino Superior 9 – Ignorado |
| IDADE | Idade do paciente |
| DTNASC | Data de nascimento |
| SEXO | Sexo do paciente: 1 – Masculino 2 – Feminino |
| UFNASC | Estado (UF) de nascimento |
| UFRESID | Estado (UF) da residência |
| DTOBITO | Data do óbito |
| DIAGPREV | Diagnóstico e tratamento anterior: 1 – Sem diagnóstico e sem tratamento 2 – Com diagnóstico e sem tratamento |
| CATATEND | Categoria de atendimento à admissão: forma do atendimento realizado no hospital, no momento da admissão, de acordo com: SUS Convênio Particular (a partir de março/2013) |
| SITUACAO | Estado atual após o primeiro tratamento: 01 – Sem evidência da doença 06 – Óbito por câncer 02 – Remissão parcial 07 – Óbito por outras causas 03 – Doença estável 08 – Tratamento não concluído 04 – Doença em progressão 09 – Não se aplica 05 – Fora de possibilidades terapêuticas 10 – Sem informação |
| NAOTRAT | Razão para a não realização do tratamento: 1 – Recusa do tratamento 6 – Óbito por outras causas 2 – Doença avançada, falta de condições clínicas 7 – Outras razões 3 – Outras doenças associadas 8 – Não se aplica 4 – Abandono do tratamento 9 – Sem informação 5 – Óbito por câncer |
| BASEDIAG | Base utilizada para o diagnóstico: 1 – Exame clínico 3 – Confirmação microscópica 2 – Recursos auxiliares não microscópicos 9 – Sem informação |
| TOPO | Topografia. Localização do tumor primário (CID-O, 2ª e 3ª edições) |
| MORFO | Morfologia. Tipo histológico do tumor primário (CID-O, 2ª e 3ª edições) |
| DTCONSULTA | Data da primeira consulta |
| DTDIAG | Data do diagnóstico do tumor |
| EC | Estádio clínico (TNM, 5ª e 6ª edições) |
| T | Código T (TNM, 5ª e 6ª edições) |
| N | Código N (TNM, 5ª e 6ª edições) |
| M | Código M (TNM, 5ª e 6ª edições) |



| | |
|---|---|
| DTTRAT | Data de início do primeiro tratamento |
| TRATAMENTO | Tipo(s) de tratamento(s) proposto(s): A – Cirurgia B – Radioterapia C – Quimioterapia D – Cirurgia + Radioterapia E – Cirurgia + Quimioterapia F – Radioterapia + Quimioterapia G – Cirurgia + Radio + Químio H – Cirurgia+Radio+Químio+Hormônio I – Outras combinações de tratamento J – Nenhum tratamento realizado K – Sem informação do tratamento |
| META01 | Localização da metástase (CID-O, 2ª e 3ª edições) |
| META02 | Localização da metástase (CID-O, 2ª e 3ª edições) |
| META03 | Localização da metástase (CID-O, 2ª e 3ª edições) |
| META04 | Localização da metástase (CID-O 2ª e 3ª edições) |
| DTULTSEG | Data do último seguimento informado |
| A) SITULTSEG (até dez/2011) | Situação do paciente no último seguimento informado: 1 – Vivo com câncer 2 – Vivo, SOE 3 – Óbito por câncer 4 – Óbito, SOE 5 – Liberado de seguimento 9 – Sem informação |
| B) SITULTSEG (a partir de mar/2012) | Situação do paciente no último seguimento informado: 1 – Vivo com câncer 2 – Vivo, SOE 3 – Óbito por câncer 4 – Óbito, SOE 5 – Perda de seguimento |
| ANODIAG | Ano do diagnóstico do tumor |
| ECGRUP | Estadiamento TNM agrupado |
| TOPOGRUP | Topografia. Localização do tumor primário (CID-O, 2ª e 3ª edições), com 3 dígitos |
| CICI | Estadiamento Infantil – Grupo + Subgrupo |
| CICIGRU | Descrição do Estadiamento Infantil (CICI) – Grupo |
| CICISUBGRU | Descrição do Estadiamento Infantil (CICI) – Subgrupo |
| FAIXAETARI | Faixa etária (de 10 em 10 anos) |
| A) LATERALI | Localização (lateralidade) de tumores em órgãos, glândulas e cavidades em pares 1 – Direita 2 – Esquerda 3 – Indiferente |
| B) LATERALI (a partir de set/2011) | Localização (lateralidade) de tumores em órgãos, glândulas e cavidades em pares 1 – Direita 2 – Esquerda 3 – Não se aplica |
| DSCTOPO | Descrição da topografia. Localização do tumor primário (CID-O, 2ª e 3ª edições) |
| DSCMORFO | Descrição da morfologia. Tipo histológico do tumor primário (CID-O, 2ª e 3ª edições) |



CANCER OBSERVATORY