LEUKEMIA & LYMPHOMA SOCIETY OF CANADA®
fighting blood cancers

someday is today

Blood Cancer in Canada

Facts & Stats

2016

Note: the major source of information for this report is the Canadian Cancer Statistics Report, 2016. Other data sources were used to address gaps in the information provided about some populations or on some blood cancers.

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About Blood Cancers

There are to 137 types of blood cancers and related disorders. These cancers involve blood cells, the bone marrow, the lymph nodes and other parts of the lymphatic system. The main types of blood cancers include:

- Leukemia
- Hodgkin and non-Hodgkin lymphoma
- Myeloma
- Other, less common, blood cancers, such as myelodysplastic syndromes and myeloproliferative neoplasms.

Stem cells are a class of cells that have the ability to develop into any of several specialized types of cells, such as red or white blood cells or platelets. Most blood cancers result from an acquired (as opposed to a genetic or inherited) mutation to the DNA of a stem cell that produces lymphatic or blood-forming cells. With blood cancers, abnormal cells multiply much more rapidly – and are less likely to die naturally – than healthy cells. The accumulation of abnormal cells in the bone marrow, blood, and/or lymphatic tissue interferes with the production and healthy functioning of:

- Red blood cells that transport oxygen to the cells throughout the body and waste gases back to the lungs to be exhaled. When there are abnormal or too few red blood cells, the person is at risk of anemia.
- White blood cells that are critical for fighting infections and disease
- Platelets that play an essential role in blood clotting.

Advances in the understanding of the genetics and mechanisms of blood cancers have opened new and exciting avenues in diagnosing, treating and managing blood cancers. More information is provided in each of the specific blood cancer sections which follow.

Highlights

Prevalence

An estimated 138,100 people in Canada are living with, or are in remission from, the following forms of blood cancer:

- 43,335 with lymphoma
- 22,510 with leukemia
- 7,455 with myeloma
- Up to 40,000 with myelodysplastic syndromes
- 14,300 with polycythemia vera
- 8,700 with essential thrombocythermia
- 1,800 with myelofibrosis

Did you know: More Canadians than ever are living with the effects of a blood cancer. The number of Canadians living with, or in remission from blood cancer has increased by 25% from 2014 to 2016.

New Cases (Incidence)

In 2016, approximately 22,340 Canadians of all ages were diagnosed with a form of blood cancer:

- 9,000 cases of lymphoma
- 5,900 cases of leukemia
- 2,700 cases of myeloma
- 3,850 cases of myelodysplastic syndrome
- 400 cases of polycythemia vera (PV)
- 290 cases of essential (primary) thrombocythemia (ET/PT)
- 200 cases of myelofibrosis

Figure 1: Estimated new annual cases of blood cancer in Canada, 2016

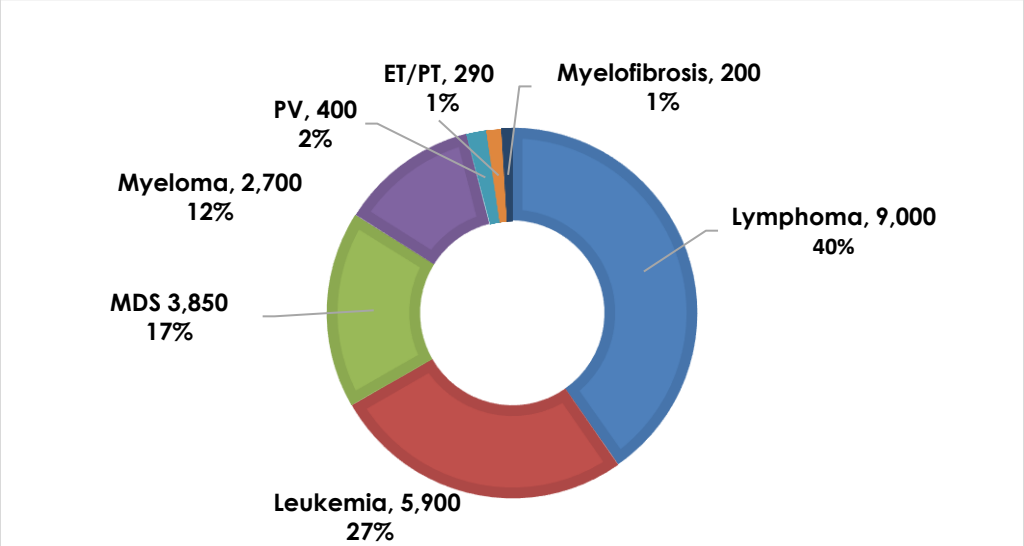


Figure 2: Estimated number of new annual cancer cases, all cancers 2016

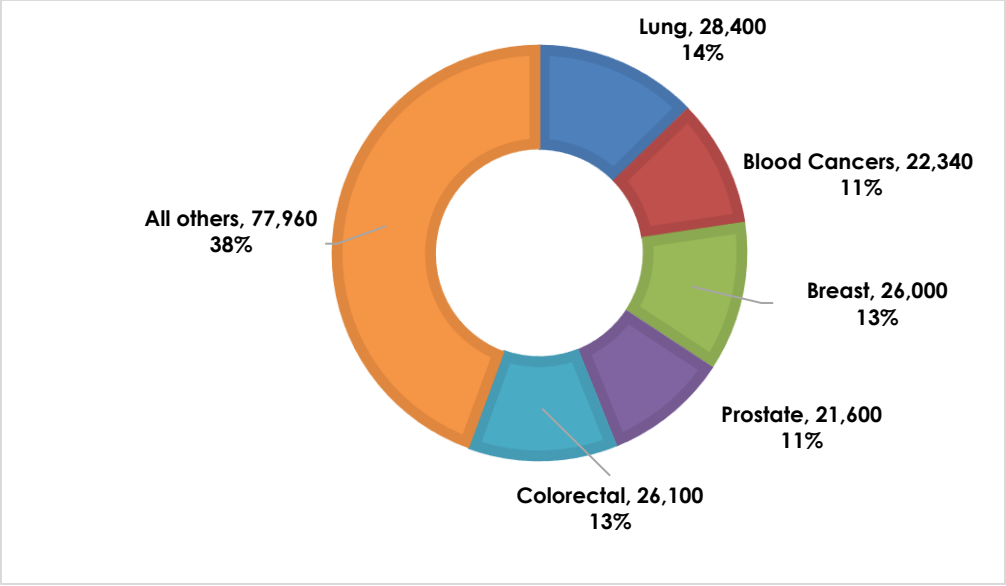


Table 1: Rate for new cases (per 100,000 population)

Type of Blood Cancer	Rate per 100,000 2014	Rate per 100,000 2016
Non-Hodgkin lymphoma	17	21
Leukemia	13	15
Myeloma	5	7
Hodgkin lymphoma	3	3

Table 2: Lifetime probability of developing a blood cancer by gender

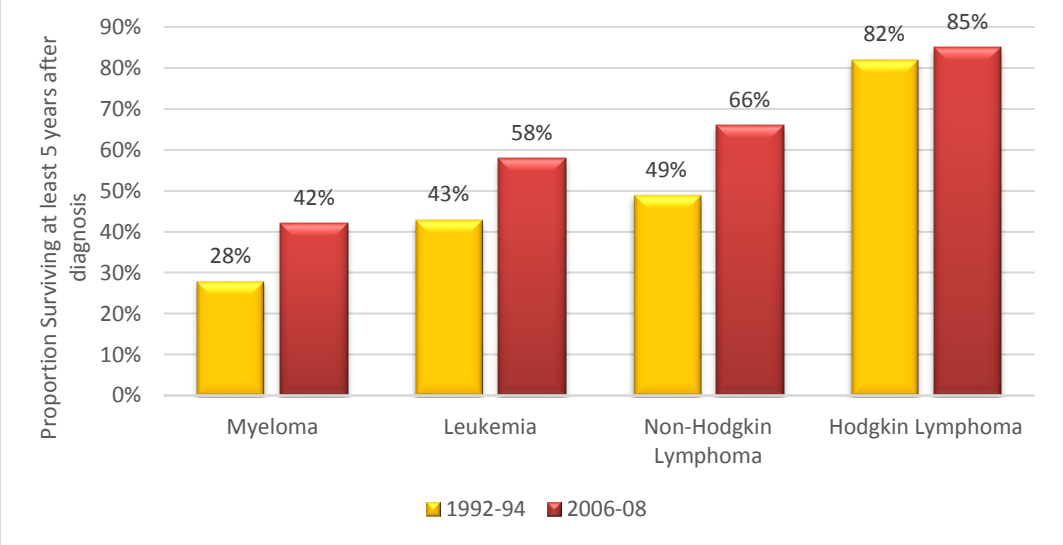
Type of blood cancer	Males		Females	
	2014	2016	2014	2016
Non-Hodgkin lymphoma	1 in 44	1 in 43	1 in 51	1 in 50
Leukemia	1 in 54	1 in 53	1 in 72	1 in 72
Myeloma	1 in 123	1 in 118	1 in 159	1 in 143
Hodgkin lymphoma	Not reported	1 in 432	Not reported	1 in 498

Did you know: Blood cancers are the fourth most commonly diagnosed cancer in Canada. Although there has been little change in the lifetime probability of developing most forms of blood cancer between 2014 and 2016, there have been increases in the rate at which Canadians are diagnosed with non-Hodgkin lymphoma, leukemia and myeloma, due to impact of an ageing population.

Survival

- For most types of blood cancer, the proportion of people who live five or more years after diagnosis has increased significantly over the past decade. The 5 year survival rates range from 42% for myeloma to 85% for Hodgkin Lymphoma.

Figure 3: Changes in observed 5 year survival rates



Did you know: Between the early 1990s and the mid-2000s, research and advances in treatment have made possible a 50% increase in the five-year survival rate for myeloma and 35% increases for leukemia and non-Hodgkin lymphoma. Despite these advances, the five-year survival rate for most blood cancers are much lower than many other cancers.

Mortality

- Leukemia, lymphoma and myeloma are estimated to have caused the deaths of almost 7,200 Canadians in 2016, compared to 7,000 in 2014. This means that approximately every 73 minutes someone in Canada dies from one of these diseases, or almost 20 Canadians per day.
- Blood cancers are the third leading cause of cancer death in Canadian men and the fourth in women.
- These three forms of blood cancer account for 9 percent of the 78,800 cancer deaths in Canada in 2016.
- Due to advances in diagnosis and treatment, the age-standardized mortality rates for leukemia, non-Hodgkin lymphoma and myeloma fell between 2003 and 2012.

Did you know: In 2016 an estimated 7,200 Canadians died from leukemia, lymphoma and myeloma. This represents a 3% increase over 2014, due to an ageing population. On an age standardized basis, mortality rates for leukemia, non-Hodgkin lymphoma and myeloma fell between 2003 and 2012 due to advances in diagnosis and treatment.

Leukemia

The term "leukemia" refers to a cancer of the blood cells. There are four major types of leukemia. Because of differences in the characteristics of the different types of leukemia, there are also differences in how they are treated.

In understanding the four major types of leukemia, it is helpful to understand the following terms.

- **Acute vs. chronic**: when the term "acute" is used, it refers to a type of cancer that – without treatment – may advance quite rapidly, such as within months. In contrast, the term "chronic" refers to a form of cancer that typically progress much more slowly.
- **Lymphoblastic vs. myeloid**: these terms refer to the type of blood cell that is involved. "Lymphoblastic" or "lymphocytic" refers to a cancer affecting lymphocytes, white blood cells such as the B lymphocytes, T lymphocytes or natural killer cells. In contrast, "myeloid" or "myelogenous" refers to a cancer that involves a type of stem cell that has the potential to develop into a red blood cell, non-lymphocytic white blood cells such as granulocytes, or platelets.

In all forms of leukemia, the proliferation of abnormal, non-functional cells in the bone marrow and blood interferes with the production of normal, fully-functional red blood cells, white blood

cells, or platelets. As a result, people with leukemia may develop anemia, have a reduced ability to fight infections, and may experience blood clotting disorders.

Keeping this in mind, the four major types of leukemia are:

1. **Acute lymphoblastic leukemia (ALL)** – a fast-growing cancer of lymphocytes (white blood cell) that results in the accumulation of immature, malfunctioning cells in the bone marrow and blood.
2. **Chronic lymphocytic leukemia (CLL)** – a more slowly-progressing cancer of the lymphocyte cells
3. **Acute myeloid leukemia (AML)** – also known as acute myelogenous leukemia or acute non-lymphocytic leukemia, AML is a rapidly-progressing cancer of myeloid stem cells.
4. **Chronic myeloid leukemia (CML)** – a more slowly-progressing cancer of the myeloid stem cells.

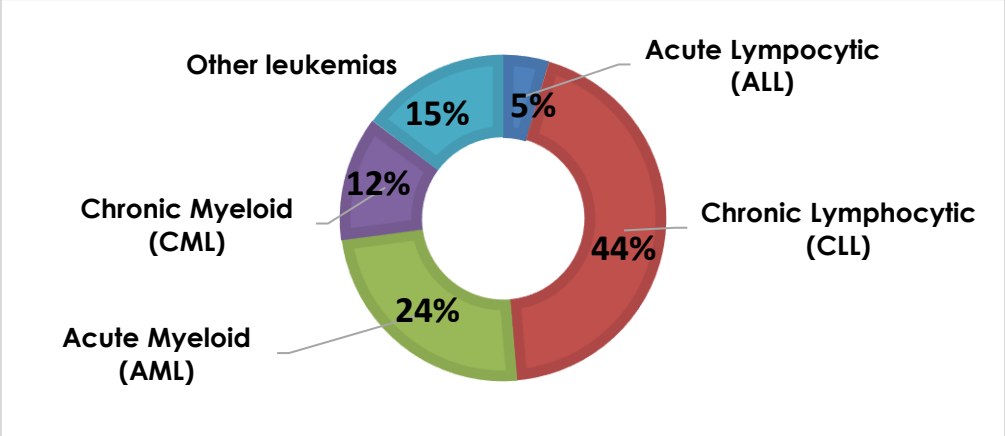
Treatment

Within each type of leukemia, there can be several sub-types depending upon the cancer cells involved, how mature they are, and how different they are from normal cells. Treatment options vary according to the type of leukemia and other factors, such as age and general health. The main treatment options include chemotherapy, stem cell transplant, or targeted therapy using drugs designed to only attach to specific antibodies or proteins on cancer cells. In special circumstances, other treatments may be used, such as surgery, radiation therapy, leukapheresis (removing white blood cells from the blood), or treatment with monoclonal antibodies. Advances in understanding the genetics of leukemia and how they influence the progress of the disease and response to treatment is opening new doors in individualizing treatment.

Prevalence & Incidence

- Approximately 22,510 Canadians are living with, or are in remission from leukemia: 13,040 males and 9,470 females.
- An estimated 5,900 people were diagnosed with leukemia in 2016.
- The most common types of leukemia in adults are AML and CLL. The median age at the time of diagnosis is 67 for AML and 71 for CLL.
- One in 53 men and one in 72 women will develop leukemia in their lifetime

Figure 4: Distribution by type of Leukemia in Canada 1992-2008*



*excluding Quebec ages 15-99 years

Survival

- The age-standardized five-year survival rate for leukemia is 58% for males and 59% for females. In comparison, the five-year survival rate is 95% for thyroid cancer, 81% for prostate cancer, 79% for melanoma and 80% for breast cancer.

Mortality

- An estimated 2,900 Canadians died from leukemia in 2016.
- One in 96 men and one in 132 women will die of Leukemia in their lifetime.

Table 4: Median age at time of diagnosis

Type of Leukemia	Age most frequently diagnosed	Median Age at Diagnosis
Acute lymphocytic (ALL)	< 20 yrs.	15
Acute myeloid (AML)	65-74 yrs.	67
Chronic lymphocytic (CLL)	65-74 yrs.	71
Chronic myeloid(CML)	65-74 yrs.	64

Did you know: The age standardized mortality of Leukemia has declined by 1.4% a year between 2003 and 2012 due to advances in research and treatment options.

Lymphoma

"Lymphoma" refers to a cancer of the lymphocytes, a type of white blood cell found in the lymphatic system. The lymphatic system is part of the circulatory system and drains a clear fluid called lymph from the tissues into the blood. The lymphatic system plays a key role in the body's immune response that protects us against disease and infections. In lymphoma, an abnormal lymphocyte reproduces uncontrollable and may form masses in the lymph nodes, liver, spleen and/or other parts of the body.

Lymphoma can occur in both children and adults. There are two main types:

- **Hodgkin lymphoma** – in this type of lymphoma, there is a particular type of large, abnormal lymphocyte, called a Reed-Sternberg cell, in the lymph nodes
- **Non-Hodgkin lymphoma (NHL)** – the main feature distinguishing a non-Hodgkin lymphoma is the absence of Reed-Sternberg calls. There are many different types of non-Hodgkin lymphoma, as they can develop from abnormal B-cells (B-lymphocytes), T-cells (T-lymphocytes) or natural killer cells (another form of lymphocyte). Some forms of non-Hodgkin lymphoma are slow-growing (indolent) whereas others are aggressive or fast-growing. Waldenstrom macroglobulienmia is a form of non-Hodgkin lymphoma which involves a large protein called a macroglobulin. Sometimes, a non-Hodgkin lymphoma can start in the lymphatic tissue in the skin; this is referred to a skin or cutaneous lymphoma.

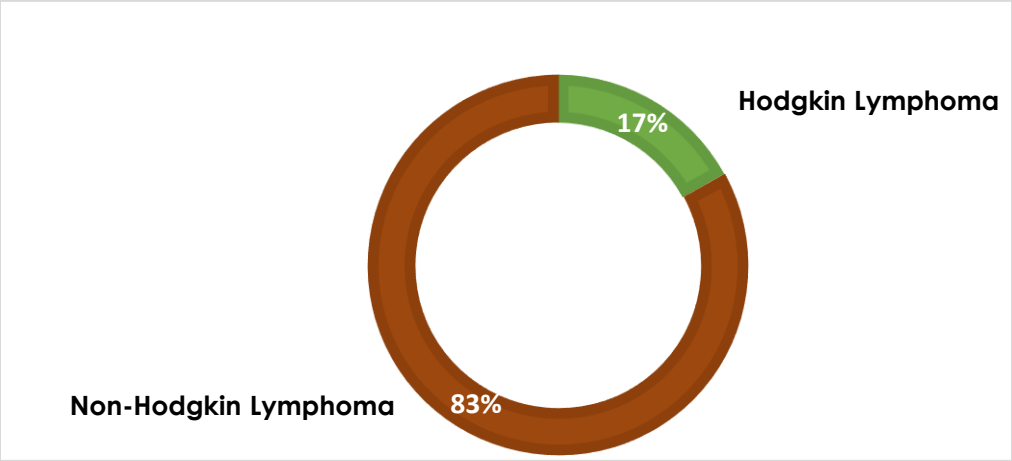
Treatment

Treatment for Hodgkin lymphoma depends upon the stage of the disease and may include chemotherapy, radiation therapy, immunotherapy (treatment with drugs to stimulate the immune system to recognize and destroy cancer cells), or high-dose chemotherapy followed by stem cell transplant. For non-Hodgkin lymphoma the treatment options are similar (chemotherapy, immunotherapy, radiation and stem cell transplant) but may also include targeted therapies (new drugs that attached only to cancer cells) or, in rare cases, surgery to remove a mass.

Prevalence & Incidence

- Approximately 43,335 Canadians are living with, or are in remission from a lymphoma: 36,175 with non-Hodgkin lymphoma and 7,160 with Hodgkin lymphoma.
- An estimated 9,000 new cases of lymphoma were diagnosed in Canada in 2016: 8,000 cases of non-Hodgkin lymphoma and 1,000 cases of Hodgkin lymphoma.
- The median age at diagnosis of Hodgkin lymphoma is 39 years and 66 years for non-Hodgkin lymphoma.
- NHL is the sixth most commonly diagnosed cancer in Canada.
- The lifetime probability of developing non-Hodgkin lymphoma is one in 43 for men and one in 50 for women in their lifetime.
- The lifetime probability of developing Hodgkin lymphoma is one in 432 for a men and one in 498 for a female.

Figure 5: Distribution by type of lymphoma



Survival

- The five-year age-standardized survival rate for Hodgkin lymphoma is 85%.
- The five-year age-standardized survival rate for non-Hodgkin lymphoma is only 66%.

Mortality

- 2,830 people are estimated to have died from lymphoma in 2016: 2,700 from non-Hodgkin lymphoma and 130 from Hodgkin lymphoma.

- The lifetime probability of dying from lymphoma is one on 99 for men and one in 118 for women.

Did you know: Lymphoma is the most common form of blood cancer overall and the third-most common form of cancer in children 0 to 14 years in Canada.

Myeloma

Myeloma (also referred to as multiple myeloma) is a cancer of the plasma cells. Plasma cells are found in the bone marrow and produce antibodies (immunoglobulins) that protect you against disease. Myeloma is defined by the type of immunoglobulin that reproduces uncontrollably (e.g., IgG, IgA, etc.) and how aggressive the disease is (e.g., "smoldering" or "indolent" myeloma compared to symptomatic or active disease).

Cancerous myeloma cells can:

- disrupt normal blood production, leading to anemia
- interfere with the functioning of the immune system, resulting in frequent or aggressive infections
- damage normal bone tissue (osteolytic or bone lesions) that can result in pain, fractures or collapse of a vertebra
- damage the kidneys (renal disease)
- form into masses (plasmacytoma)

Treatment

During the past several decades, advances have been made in the diagnosis, staging and treatment of myeloma. Approved treatments for myeloma include a tailored combination of:

- Careful observation or watchful waiting for asymptomatic (smoldering or indolent) myeloma,
- Radiotherapy
- Chemotherapy and high dose chemotherapy for future stem cell transplantation
- Corticosteroids, often in combination with chemotherapy
- Immunomodulatory therapy with drugs that interfere with the underlying processes that promote the growth and reproduction of myeloma cells
- Proteasome inhibitors, which are drugs that inhibit plasma cell growth and reproduction and promote the death of abnormal plasma cells.

New developments in understanding the genetics of myeloma are also opening opportunities to tailor treatment in order to optimize the impact and minimize side effects.

Prevalence & Incidence

- Approximately 7,455 Canadians are living with, or are in remission from myeloma
- An estimated 2,700 Canadians were diagnosed with myeloma in 2016.
- More men than women are diagnosed with myeloma each year: 1,600 men compared to 1,150 women.
- The median age at diagnosis is 69 years; myeloma rarely occurs in people under age 45.
- The lifetime probability of developing myeloma is one in 118 for men and one in 143 for women in Canada.

Survival

- The 5 year age standardized survival rate for myeloma is a little over 42%.

Mortality

- An estimated 1,450 (800 men and 650 women) died from myeloma in 2016.
- The age-standardized mortality rate of myeloma fell 1.8% per year for women but only 0.9% per year for men between 2003 and 2012.

Did you know: While the rate of increase of myeloma diagnoses is small, only 36% of myeloma patients live five or more years after diagnosis, making it one of the most daunting forms of cancer.

Myelodysplastic Syndromes

Myelodysplastic syndromes (MDS) are a group of diseases that are often referred to a bone marrow failure disorders. In MDS, immature blood cells (referred to as blasts) are abnormal and build up in the bone marrow and the blood. Because of the proliferation of abnormal, immature cells, there are fewer healthy, functioning red and white blood cells and platelets. Sometimes, MDS has been referred to as "smoldering leukemia" or "preleukemia" as about a third may develop Acute Myeloid Leukemia. However, these terms can be misleading as MDS is a serious health problem even if it doesn't progress to active leukemia.

MDS usually develop in older people and are more common in men than women.

Treatment

Treatment options for myelodysplastic syndromes include chemotherapy, immunotherapy, stem cell or bone marrow transplantation, and supportive care (treatment to relieve symptoms and improve quality of life). Treatment will depend upon the type and stage of myelodysplastic syndrome. New types of treatments are being developed and tested in clinical trials.

Prevalence & Incidence

There is some evidence MDS may be under-diagnosed, particularly among older Canadians. It has been estimated there may be between 10,000 and 40,000 Canadians 65 and over who have been diagnosed with, or are living with, MDS.

Among adults aged 65 and older, the incidence has been estimated to range from 75 to 162 per 100,000.

Using these rates, it is estimated that between1,800 and 5,900 new cases of MDS are diagnosed in Canada per year.

Survival

- Depending upon the severity of the disease, the median survival for MDS after diagnosis ranges from 0.4 to 5.7 years.

Did you know: Researchers and physicians suspect MDS is seriously under-diagnosed, particularly among older Canadians. New approaches to diagnosis and treatments are needed, as the median survival after diagnosis currently ranges from less than a year to 6 years.

Myeloproliferative Neoplasms

Myeloproliferative neoplasms (MPN) are types of blood cancer that begin with an abnormal mutation (change) in a stem cell in the bone marrow. The change leads to an overproduction of any combination of white cells, red cells and platelets.

This group of blood disorders includes polycythemia vera, essential (primary) thrombocythemia and myelofibrosis.

Polycythemia Vera (PV)

In PV, there is an increase in the number of red blood cells which can cause the blood to "thicken" and increase the risk of blood clots. Treatment options include:

- Phlebotomy – removing blood to reduce the number of blood cells
- Medications to reduce the risk of blood clots, such as low-dose aspirin, hydroxyurea, Jakafi (ruxolitinib)
- Pergylated interferon

Prevalence & Incidence

- The prevalence of PV in North American has been estimated to range from a low of 22 to a high of 57 per 100,000 people. This suggests the number of Canadians who have had, or are living with, PV may range from roughly 8,000 to almost 21,000.
- It can be estimated that 180 to 600 Canadians are diagnosed with PV each year.
- The average age at which PV is diagnosed is about 60 to 65 years old. Only 10% of patients are under the age 40.

Did you know: Each year up to 600 Canadians with PV are at risk of serious complications such as blood clots.

Essential or Primary Thrombocythemia (ET/PT)

ET/PT is characterized by an increased number of platelets, the cells that play a key role in blood coagulation or clotting. If not treated, it can lead to serious bleeding or the formation of blood clots (thrombosis).

Some people with ET may not require treatment but others may need to take low-dose aspirin, hyroxyurea, anagrelide, or interferon. Novel new treatments are currently being developed and tested as a result of breakthroughs on the genetics of the disease.

Prevalence & Incidence

- It is estimated there may be between 36 to 544 new cases of ET in Canada each year, using available global incidence data as a guide.
- There are approximately 8,700 Canadians who are living with, or have had, ET
- Canadians over 50 are more likely to develop ET, although 20% of patients are under the age of 40.
- The annual new incidence rate for ET has been estimated to range from 0.1 to 1.5 per 100,000. An even higher estimate – of up to 2.2 per 100,000 people – has also been suggested.
- The prevalence of ET has been estimated to be 24 per 100,000 people.

Did you know: Essential or primary thrombocythemia increases the risk of bleeding disorders.

Myelofibrosis (MF)

In MF, there is excessive scar tissue in the bone marrow, which impairs the ability of the marrow to produce blood cells. This can result in a number of blood-associated disorders and problems. Treatment depends upon the type of blood cells that are affected by the disease.

Prevalence & Incidence

- It is estimated that between 36 and 360 Canadians are diagnosed with MF each year.
- An estimated 1,400 and 2,177 Canadians have, or have had, myelofibrosis.
- The median age at the time of diagnosis is 69 years but about 15% of people are under 50.
- In North America, the prevalence of people living with MF ranges between 4 and 6 per 100,000 people.

Did you know: Myelofibrosis impairs the functioning of the bone marrow and can lead to a number of serious complications.

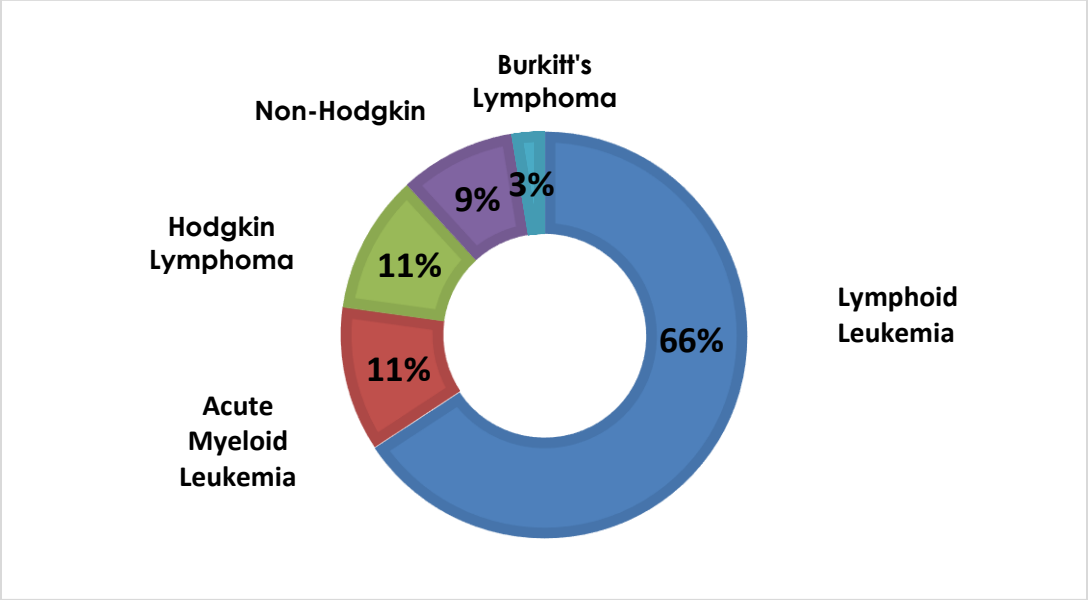
Pediatric Blood Cancers - Children Age 0-14

Prevalence & Incidence

- Leukemia and Lymphoma account for over 40% all new cancers diagnosed in children 0-14 years.
- Leukemia is the most common form of cancer in children 0-14 years of age and the 6th most common among young adults 15 to 29 years.
- Over 900 children, 0-14 years of age are diagnosed with cancer each year in Canada.
    - about 300 are diagnosed with leukemia accounting for nearly a third of all cancer diagnoses in children.
    - about 100 are diagnosed with lymphoma, accounting for 11% of all childhood cancer.
- Between 1992 and 2010 the incidence of leukemia in children increased an average of 0.6% a year.

- The most common type of leukemia in children age 0-14 years is ALL. Three out of 4 children diagnosed with leukemia are diagnosed with ALL.
- Between 1992 and 2010 the incidence of lymphomas in children had a non-significant increase of 0.5% per year.

Figure 6: Type of blood cancers diagnosed in children 0 - 14: Canada 2006 – 2010



Survival

- The observed five-year survival rate for children with leukemia, myeloproliferative disease or myelodysplastic diseases is 88%: 91% for lymphoid leukemia and 73% for acute myeloid leukemia.
- The observed five-year survival for children with lymphoma is 92%: 98% for Hodgkin lymphoma, 88% for non-Hodgkin lymphoma and 92% for Burkitt lymphoma.

Mortality

- Fortunately, thanks to advances in treatment, between 1992 and 2010 the age-standardized mortality for children with leukemia decreased an average of 3.5% per year.
- Between 1992 and 2010 the age-standardized mortality for children with lymphoma fell an average of 4.5% per year.

Did you know: Due to advances in research and treatments survival rate for children suffering from leukemia or lymphoma is as high as 98%. At the same time it is estimated that at least two-thirds of pediatric cancer survivors will experience persistent long term effects from chemotherapy or radiation. These late effects can affect the physical and emotional health of a child, as well as their long-term mortality. More research and gentler effective therapies are needed to reduce late long-term health effects.

Adolescent Blood Cancers - Age 15-19

Prevalence

- Each year, an average of 400 adolescents 15-19 years are diagnosed with cancer in Canada and 74 die from the disease.
- More boys are diagnosed than girls.
- Lymphomas are the most common type of adolescent cancer, accounting for 29% of all cases.
- Leukemia accounts for 12% of adolescent cancer cases.
- Adolescents have not benefited from the same gains in survival observed in younger children and it's not clear why this gap exists.

Did you know: Adolescents have not benefited from the same gains in blood cancer survival observed in younger children and it's not clear why this gap exists.

Table 5: Number of New Cases of Blood Cancer by Province, 2016

	Lymphoma by Type		Total Lymphoma*	Leukemia*	Myeloma*	Estimates of all others **	Total
	Non-Hodgkin Lymphoma*	**Hodgkin Lymphoma ***					
CANADA	**8,000**	**1,000**	**9,000**	**5,900**	**2,700**	**4740**	**22,340**
Newfld & Labrador	160	15	175	130	45	69	419
PEI	35	0	35	25	15	20	95
Nova Scotia	260	25	285	220	80	124	709
NB	215	15	230	200	65	98	593
Quebec	1,870	260	2,130	1,670	660	1088	5,548
Ontario	3,150	380	3,530	2,970	1,100	1827	9,427
Manitoba	280	30	310	295	86	173	864
Saskatchewan	240	40	280	265	75	150	770
Alberta	800	110	910	815	260	555	2,541
BC	1,040	115	1,155	995	360	620	3,131

* Numbers from Canadian Cancer Society 2016 Canadian Cancer Statistics
** Total number of cases of myelodysplastic syndrome, polycythemia vera, essential/primary thrombocythemia and myelofibrosis estimated from the literature.