Chronic Lymphocytic Leukemia

B-cell Chronic Lymphocytic Leukemia (CLL) and Small Lymphocytic Lymphoma (SLL) are cancers of the white blood cells that are characterized by a proliferation of B cell lymphocytes. According to Statistics Canada (2006), about 1900 new patients are diagnosed every year. SLL primarily involves the lymph nodes while CLL affects the blood, bone marrow and lymph nodes. CLL and SLL are now considered to be different manifestations of the same disease. Resulting anemia, low platelets and frequent infections because of low immunity are common.

Most cases of CLL are diagnosed in people over age 50 and its incidence increases with age. CLL tends to be indolent in the early stages and is often diagnosed as a result of routine blood tests. In many cases, a watchful waiting period begins as there are little benefits to early treatment. This period often lasts several years during which patients often complain of various levels of fatigue and anxiety. Proper diagnosis is critical as a subset of patients have an aggressive form of CLL and need to make the best therapy choices quickly.

CLL symptoms leading to treatment include fever and/or chills, severe weight loss, soaking night sweats and overwhelming fatigue. Chemotherapy agents such as fludarabine (Fludara) and cyclophosphamide (Procytox, Cytoxan) are typically used in combination with monoclonal antibodies such as rituximab (Rituxan) in order to reduce these symptoms. This combination, known as FCR has become the gold standard for treatment. However it tends to be quite toxic and typically leaves patients in a more immune-compromised state. Younger or high-risk patients may be offered an allogeneic stem cell transplant. Clinical trials are one way patients can receive emerging drug combinations while waiting for provinces to fund drugs officially approved by Health Canada. These trials tend to be restricted to patients who meet stringent criteria and who have access to a cancer centre offering the trial. Many patients are frustrated that there is no standard of practice for CLL in Canada as treatment depends on which province you live in.

CLL is due to DNA genetic damage during cell division, as part of the B-cell regenerative process. Defective B-cells accumulate and can eventually lead to bone marrow failure, if left untreated. Genetic biomarker testing such as FISH (Fluorescent In Situ Hybridization) and the more recent introduction of Chromosomal Microarray Analysis known as array Comparative Genomic Hybridization (aCGH) are expanding the field of genetic markers in CLL. FISH testing is done for patients in clinical trials but they are generally not given the results. The aCGH is currently not available to Canadian patients unless they make their own arrangements to have it completed out of the country. Advances in treatment, a better understanding of how the cancer functions and mapping of the chromosome damage that causes CLL has led to greatly increased response rates and durations of response. Treatments can be repeated but they typically result in shorter remissions. Not much advance has been made in extending overall survival, however.

Canada does not have a Centre of Excellence for CLL. We are fortunate to have a dedicated group of clinicians and researchers who form the Canadian CLL group and meet in Manitoba every year. CLLPAG is a national volunteer organization of patients committed to advocacy, awareness, education and equal access to care on behalf of Canadians affected by CLL and SLL. The Leukemia and Lymphoma Society of Canada, Lymphoma Foundation of Canada and Juravinski Cancer Centre in Hamilton, ON support CLLPAG in organizing an International Conference on CLL every few years. This unique event, organized by CLL patients for CLL patients and practitioners, provides a world-class forum to learn about the latest developments on the treatment of this complex, heterogeneous disease.

As we learn more about the genetic variations in CLL/SLL, there will be a growing need for individualized treatment and advanced biomarker testing. These will require further treatment approvals based on new criteria, new ways of thinking about the disease, new technologies and a greater emphasis on targeted therapies that closely match the needs of the patient.

Personalized medicine will save money by reducing ineffective or unnecessary treatments. Canadians should be able to receive optimal health care no matter where they live. It is imperative that the cancer care agencies of Canada and provincial and federal health ministries support research into new treatments and technologies as well as fast-track approval of selective inhibitors which are newer, innovative, less toxic treatments. A great deal of progress has been achieved with the approval of FCR as the therapy of choice for most untreated CLL patients. Approval of FCR or FR for repeat therapy and select alternatives, such as bendamustine (Treanda), for older patients is paramount.

CLL
PATIENT ADVOCACY GROUP
GROUPE DE DÉFENSE DES DROITS DES PATIENTS ATTEINTS DE LLC

Submitted by members of the CLL Patient Advocacy Group *www.cllpag.ca*

Ovarian Cancer

While considered rare compared to some cancers, Canadian women have a 1 in 70 lifetime risk of developing ovarian cancer. Approximately 2,600 women are diagnosed and there are 1,750 deaths from the disease each year. Seventy per cent of women with ovarian cancer do not survive five years—making the disease Canada's most fatal gynecologic cancer. Although ovarian cancer is most common among women over the age of 50, it also affects younger women. About 10 per cent of ovarian cancers are hereditary—usually due to a mutation to the BRCA1 or BRCA2 (Breast

Cancer 1 or 2) gene. One in 50 Ashkenazi Jews carries one of these gene mutations that increases their risk for breast, ovarian and related cancers. Segments of the French Canadian population may also be at increased risk.

Elisabeth Ross

As the country's sole charity dedicated to overcoming ovarian cancer, Ovarian Cancer Canada has a continuous dialogue with women and families living with the disease, other families who have lost loved ones, health professionals who deliver care and researchers who study ovarian cancer. Our stakeholders speak with one voice when they say that their biggest frustration is the absence of a screening test for the early detection of ovarian cancer. The need for increased resources to support Canada's most promising research in this area cannot be overstated. When detected and treated at an early stage, five-year survival of ovarian cancer can be as high as 90 per cent. Unfortunately, the lack of a screening test and the fact that symptoms can be vague and attributed to other causes means ovarian cancer is usually diagnosed in later stages, when the disease has already spread to other parts of the body.

The signs and symptoms of ovarian cancer—including swelling or bloating of the abdomen, pelvic discomfort or heaviness, difficulty eating or feeling full quickly and emptying the bladder frequently—are not well known among Canadian women and many primary health care providers. This lack of awareness and the absence of a screening test for early detection can result in a longer wait for a correct diagnosis compared to other cancers. Until an early detection test is available, knowledge and awareness of the signs and symptoms are the best defence against this disease. A woman who experiences one or more symptoms that persist for three weeks should see her doctor for a full investigation.

If ovarian cancer is suspected, the family doctor or gynecologist should refer the patient to a gynecologic oncologist, a specialist with five years of postgraduate training in obstetrics and gynecology plus an additional two years of cancer training. The gynecologic oncologist will manage treatment—usually surgery and chemotherapy. Studies have shown that there are better outcomes for ovarian cancer when surgery is performed by a gynecologic oncologist. Canada has 82 of these specialists and more are needed to meet the growing demand for care.

Women with ovarian cancer tell us that the wait to see a gynecologic oncologist, and the wait for surgery and other treatment to begin, can be very stressful. A shortage of specialists plus other challenges, such as limited access to operating rooms and inconsistencies in available therapies across the country—intraperitoneal chemotherapy, for example—are other frustrations they experience.

It is estimated that the incidence of gynecologic cancers in Canada will rise by 47 per cent between 2001 and 2014. The Society of Gynecologic Oncology of Canada (GOC), in partnership with Ovarian Cancer Canada, is now conducting a study of wait times and treatment protocols for gynecologic cancers across the country with an aim to improve care for women requiring treatment.

Women across Canada also tell us that they experience high stress in the period immediately following completion of their treatment. By this point in their ovarian cancer journey, they have educated themselves about the disease and know that they have to live with the possibility of recurrence. Issues ranging from survivorship to recurrence, palliative care and end-of-life care mean women with ovarian cancer and their families need support services close to home.

Since our organization was founded 13 years ago, Ovarian Cancer Canada has worked diligently with survivors, volunteers and the cancer community toward the goal of overcoming ovarian cancer by:

- supporting women and families living with the disease
- educating the public and health professionals about ovarian cancer
- and raising funds for research into early detection and, ultimately, a cure.

We believe that success can best be achieved through a collaborative national cancer strategy that brings greater focus and improved research opportunities for diseases that are as lethal as ovarian cancer. We also believe that a coordinated national approach can best address the human and financial resource issues, and ensure equal access to the best evidence-based care and support for all Canadian women and their families living with ovarian cancer.

Elisabeth Ross is Chief Executive Officer of Ovarian Cancer Canada.
www.ovariancanada.org
She also co-chairs the National Survivorship Working Group of the Canadian Partnership Against Cancer.

Testicular Cancer

Testicular cancer starts in the cells of a testicle. The testicles are part of a man's reproductive system. They are the two egg-shaped organs found in the sac of loose skin (scrotum) at the base of the penis. The testicles are held in the scrotum by the spermatic cord. The spermatic cord contains the ductus deferens, some lymph nodes, veins and nerves.

Testicles make the male sex hormone testosterone and sperm. Sperm begins to form in "germ" cells inside the testicles. Most testicular cancers start in the germ cells and are called germ cell tumours.

There are two main types of germ cell tumours: seminomas and non-seminomas. Each type grows differently and is treated differently. Both types can be treated successfully.

Testicular cancer, although a rare cancer in the spectrum of cancers overall, is still the most common cancer for males from 15–29 years of age. On average, one out of every 273 males in Canada will be diagnosed

with testicular cancer in their lifetime.

The highest overall incidence of testicular cancer occurs in Alberta, then Nova Scotia and Saskatchewan. However, it is the age-specific incidence rates that reveal the poignant truth about the impact of this disease.

Peter Laneas

The highest rate of testicular cancer in one age group occurs in Nova Scotia, for men 25–29 years of age. Across the country, the highest incidence occurs in young men between the age of 25 and 34, with only Manitoba having a higher rate in the 35–39 age group. The incidence of testicular cancer increases from age 15 and decreases after age 40.[1]

While this cancer has the highest curability rate for all men's cancers at 97 per cent, the aftershock following diagnosis causes deeper problems and complications for survivors.

The stigma of an orchiectomy (semi or full surgical castration), creates typically unaddressed issues for the survivor and his sense of masculinity. Akin to breast cancer mastectomy and reconstruction, testicular cancer survivors who opt for a testicular implant do have a quick recovery time and generally are sent home to recover, either with a surety of having been cured or getting a finite chemotherapy schedule as the next, final step to being cured. What is left untreated is the how the patient feels as a man.

The psychological impact of testicular cancer is as relevant as any other cancer, regardless of the curability factor. The emotional effect following an orchiectomy is parallel to a mastectomy for a woman. Only in moments of intimacy or examination would someone know the man in front of them has had testicular cancer.

The typical responses by TC survivors are either, "sweep it under the carpet"—living in denial that it ever happened—or a gradual acceptance of the new reality, depending on the survivor's personal sense of security and whether a social support system is in place.

Cancer is a catalyst for confronting one's mortality. It offers a perspective like no other, bringing issues to the forefront. What once seemed important and/or tolerable becomes insignificant and/or intolerable, leaving you with your world upside down, asking "where do I go from here?"

In men, this experience is difficult to adjust to, as we do not often express feelings liberally and honestly. When combined with the age group involved—often as young as 15— the impact on personal, emotional development can be severe.

TCTCA offers a peer support system for survivors to network via email, telephone, or in person. As the only registered non-profit charity for Canadian testicular cancer awareness, if every TC survivor were to approach our group at once, we would be overwhelmed accommodating an entire country of males in need.

Testicular cancer is one face to the young adult cancer issue in Canada, where we are effectively "a lost generation." With so much effort focused on paediatric/geriatric cancer care and support programs, hospitals and cancer treatment centres do not have as many young adult survivor outreach programs in place yet. This is mostly due to the lack of a young adult approach within the cancer system, stemming from the insecurity and fear of patients who retreat from drawing attention.

Encouraging an open forum for testicular cancer patients at the time of diagnosis is just as important as the diagnosis itself. Hopefully this leads to easier adjustment in developing self-identity through any/all treatments, approaching friends and/or family for emotional support and security as men following cancer's touch.

Peter Laneas is a two-time testicular cancer survivor and the national spokesperson for The Canadian Testicular Cancer Association. For more information on testicular cancer, www.tctca.org.

References

1 Sources: Surveillance and Risk Assessment Division, CCDPC, Health Canada; Statistics Canada and the Canadian Council of Cancer Registries

UPDATE

Chronic Myelogenous Leukemia

Key issues last year were disease resistance and patient intolerance to current front line therapy, imatinib (Gleevec). Newer second-generation drugs are now available to treat these issues. In August 2010, Health Canada approved nilotinib (Tasigna), which joined dasatinib (Sprycel) as second line therapies. The approval of nilotinib is good news, but because each province must approve the drug for

Cheryl-Anne Simoneau

reimbursement on their provincial formularies, it has been very difficult for some Canadian patients to access this drug. In BC, Alberta, Saskatchewan and Quebec, nilotinib has been approved on provincial formularies as a second line treatment. However, in Ontario, nilotinib funding is only available for third line treatment, setting up a potential disaster for some patients. Some patients linger longer on the first line treatment while their health deteriorates to the point that when they may be able to access sec-

ond and third line therapies, neither of them offers any improvement. We must be able to trust our doctors to protect our health.

The next issue we addressed was purchasing our oral cancer therapy through private employer insurance, with some assistance from provincial drug plans. For some patients, the gap leaves them paying $1,500 that is not reimbursable. Why are we subjecting these patients to something that sounds like a "cancer tax"? Additionally, colleagues frown upon co-workers with diseases on "designer" priced drugs, as premiums are increased or benefits are scaled back to meet the costs. BC and Alberta continue to be the only provinces that provide oral cancer drugs through their cancer and rare disease drug programs.

Our third issue was asking the government to step in and get involved in supporting drug combination trials or stopping drug trials for CML. This is happening in Europe. The data from Europe indicate that 10 per cent of CML patients on the current front line therapy, imatinib, may be candidates to safely stop taking drugs. This figure could be as high as 40 per cent if we factor in the superior responses that patients achieve on the second line drugs such as dasatinib and nilotinib. We hear rumours that industry is stepping up to the plate, but where is our government?

We would like the governments to support a personalized medicine approach with regards to CML. If we could allow our physicians to conduct appropriate diagnostic tests up front for CML patients, such as immunophenotying, mutation detection combined with cytogenetics, PCR, FISH and standard blood tests, as well as other tests that may be deemed appropriate, we may be able to ensure that the right patient gets matched with the right drug, at the right dose, at the right time.

Next year we hope to report that these critical issues are resolved, with improvements in patients' quality of life and return to good health.

Cheryl-Anne Simoneau is President of the CML Society of Canada.

Multiple Myeloma

Have there been improvements in our access to drug treatments? This year's answer—maybe yes, maybe no.

There are two parts of the equation. First there is the funding approval and then, the challenge of physically getting the drug.

The Pan Canadian Oncology Drug Review is to be a more transparent, accountable system of cancer drug review. pCODR is a step forward as more public inclusion has been desired and requested—including patient submissions. It is a modest step, but we welcome it and look forward to further openness and understanding.

Carolyn Henry

Then each province and territory will make their own funding decisions based on the pCODR evaluation of effectiveness and cost-benefit. Even assuming, with great optimism, that the provinces and territories all accept a positive recommendation from pCODR and decide to fund a new cancer drug, there are always conditions and restrictions.

The second part of this equation is receiving the needed drug therapy.

The actual receipt of a needed drug may be either a simple or an onerous task—most stressful and unfair when one's life is hanging in the balance. Ontario's Expanded Access Program puts restrictions on access with the intent of protecting the patient. A perfect example is lenalidomide (Revlimid). Processes must be and have been put in place because of the historical birth defect issue. So this is good. However, the organization or administration of this program is a convoluted nightmare.

An application to receive a drug under the EAP is supposed to receive a decision within two weeks. Approvals have been taking up to two months. This system has forced some physicians to spend extraordinary amounts of time on paperwork rather than providing patient care. Hospitals already hire "reimbursement navigators" to help patients and doctors find a way to obtain the many treatments not covered by the provincial drug plan.

Would you, as a patient with an advancing cancer, want to wait two months simply because of paperwork to find out if your EAP application is approved? Whether or not the health ministry feels any sense of urgency, the cancer patients certainly do.

Physicians who work with this system 24/7 are strong proponents for positive change. But patients also can, and must, have a voice. It is the responsibility of the patient population to become more informed, more involved and more outspoken.

Carolyn Henry is a myeloma patient and co-group leader, London and District Myeloma Support Group, London, ON. www.myelomacanada.ca

Neuroendocrine Tumours

Neuroendocrine tumours, now known more commonly as NET cancers, is the umbrella term for a group of unusual, often slow-growing cancers, which develop from cells in the diffuse endocrine system. They are found most commonly in the lung or gastrointestinal system, but they can also originate in other parts of the body such as the pancreas, ovary, and testes, among other sites.

Increased awareness of NET cancer has developed as a result of media coverage of Apple founder Steve Jobs' pancreatic NET cancer. We are now recognized as the fastest growing cancer community worldwide (BBC, Belfast, September, 2010).

Despite increased media exposure, NET cancer still remains, for the most part, a no-name cancer. Although the

Canadian Cancer Encyclopedia now boasts a Neuroendocrine Cancer chapter, it is filed under the category of "other endocrine." Usually NET Cancers are enmeshed inside the gastrointestinal category. We NET Cancer patients would dearly love to see our own category emerge on official cancer sites.

Maureen Coleman

The problem seems to be that NET cancers do not originate from a specific site. This seems to confound provincial health ministries and cancer agencies when it comes to identifying us properly.

An important NET cancer diagnostic scan, the octreoscan, still has no billable code, which means hospitals have to individually foot the bill for our scans, making us an expensive and unattractive patient group for hospitals worried about their bottom line.

For most of last year, the only centre in Canada accepting patients from all over the country were the Neuroendocrine Cancer Centres in Calgary and Edmonton. The Cross Cancer Centre in Edmonton was actually the first location in Canada to begin to treat patients with the not-yet-approved rare nuclear isotope Lutetium 177, which targets all NET cancer cells and is regarded as a significant treatment option for a substantial segment of our community.

The London Regional Cancer Centre, which in the past had accepted patients from all over Canada, had been closed to patients outside the Local Health Integrated Network, (LHIN) for 2009 and much of 2010, but in the second half of 2010 Ontario bowed to patient pressure and the London HSC opened its doors again.

Currently Cancer Care Ontario has an Expert Panel and Steering Committee looking at the delivery of radioisotope treatment for qualifying patients in Ontario. CNETS Canada is acting in a consultative capacity.

Our Vancouver International Conference in May 2010, with 35 faculty from six countries, focused on the best and most innovative practices worldwide and our DVDs are available for distribution across Canada to medical and patient libraries everywhere.

On June 2, 2010, CNETS Canada spoke of the need for radioisotope treatment at the Health Committee of the House of Commons. On October 10, 2010 the Canadian NET Cancers community took part in the first World NET Cancer Awareness Day by hosting information tables in key hospitals and making sure our newly published information leaflets were in the stacks. We will host more hospital information tables and put leaflets into many more hands in 2011.

Maureen Coleman is the President of CNETS Canada. www.cnetscanada.org

GIST

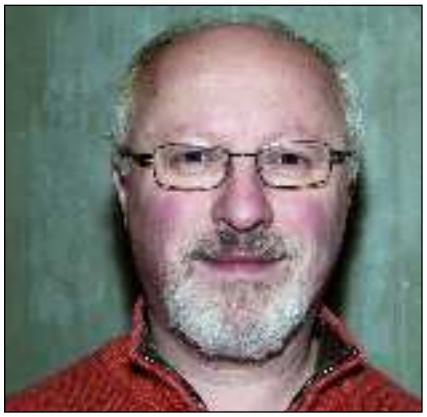
In last year's Report Card on Cancer in Canada, Life Raft Group Canada noted the many difficulties faced by patients living with GIST (gastrointestinal stromal tumour), a rare sarcoma of the gastrointestinal tract.

Drug access is a particular problem for patients with rare cancers. Imatinib (Gleevec) was the first approved targeted chemotherapy for GIST. Subsequently, several other drugs, including sunitinib (Sutent), nilotinib (Tasigna), and dasatinib (Sprycel), have either been approved or are in trial for GIST. All of these drugs cost thousands of dollars per month, and provincial drug plans have been slow to provide coverage.

There are unacceptable differences in drug coverage between provinces: a "patchwork quilt" or "postal code lottery." A particular problem right now is access to "adjuvant" Gleevec, that is, treatment intended to delay or prevent recurrence of GIST, following surgery. This treatment is known to be highly effective, based on results of several published clinical trials, but remains unavailable to many Canadian GIST patients, because of financial barriers. Novartis Pharma-ceuticals had provided access, through its compassionate-use program, but this program was shut down to all new patients, as of Feb. 1, 2011.

So, we monitor the provincial funding decisions and we press the case for improved access.

- In B.C., funding approval for adjuvant Gleevec was given, with some restrictions, in 2009.
- In Quebec, funding can be provided through the Exceptional Patient Program.
- In N.S., funding was approved in Dec. 2010.
- One month later, in N.B., funding was denied.
- In Ontario and Alberta, decisions have not yet been made. Ontario is a key province, and its pending decision could make a huge difference. For now, many patients needing adjuvant Gleevec have no options, and can only wait and hope that their cancer does not return.



David Josephy

These capricious and arbitrary differences in drug funding across Canada must end. The provinces must harmonize their coverage at the Best Practices/highest standard of care that can be achieved. Our patients cannot be left without access to needed drugs!

David Josephy is the President, GIST Sarcoma Life Raft Group Canada. www.liferaftgroup.ca