

From the paucity of treatment options to difficulties in obtaining expensive life-saving drugs, multiple myeloma patients face big obstacles to a rare cancer.

Spotlight on an Orphan Cancer

FRUSTRATION IN SMALL NUMBERS

In July 1999, George Boudreau went to his doctor for a routine medical. He felt fine, and everything looked good—heart, lungs and cholesterol levels all checked out. A week later, Boudreau, then 52, got the dreaded call: “You’ve got terminal cancer.” A blood workup had revealed—purely by accident—that Boudreau had multiple myeloma, a form of cancer that attacks the bone marrow. The Kelowna, B.C. resident was stunned. “I didn’t have any symptoms. I’d never even heard of multiple myeloma. I stormed into the doctor’s office and demanded some answers.”

Few were forthcoming, even from Kelowna’s new state-of-the-art cancer clinic and the regional cancer agency. Instead, over the next few months Boudreau got a runaround from the medical community that he describes as “ugly, draining, horrifying. The lack of concern, the lack of answers, the bullshit, were unbelievable.”

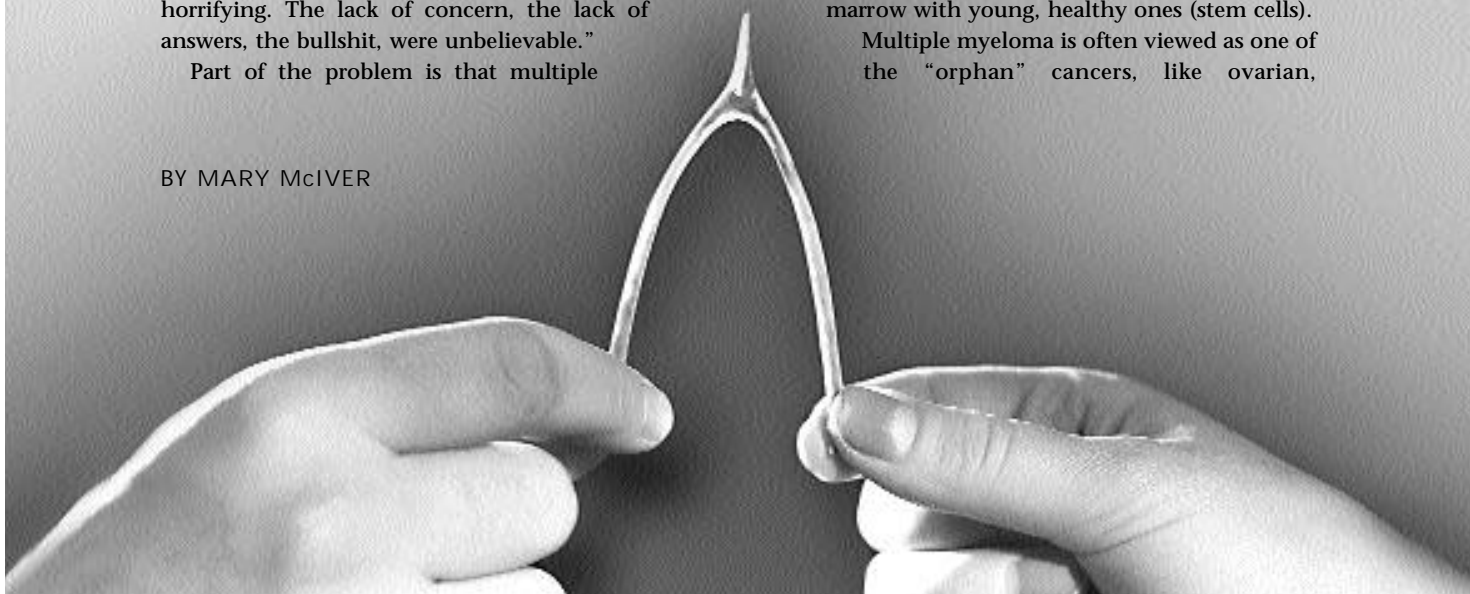
Part of the problem is that multiple

myeloma is a relatively rare (and consequently unfamiliar) disease, constituting only one per cent of cancer incidence. Specifically, it’s a proliferating malignancy of plasma cells, the blood-producing tissues in the bone marrow. The most common symptoms are fatigue, prompted by the anemia that occurs when myeloma cells replace oxygen-carrying red blood cells, and pain in the bones caused by their gradual disintegration. Other symptoms can include constipation, nausea, vomiting, loss of appetite and excessive thirst and urination. Calcium from affected bones may dissolve, creating an elevated level of calcium in the blood. It’s also common to find a protein abnormality in the blood and urine. Both of these conditions can contribute to kidney failure.

There is no cure for multiple myeloma, but the disease can be kept under control with treatments that include radiation, chemotherapy and bone marrow transplants. In Canada, the most effective treatment to date is a heavy course of chemotherapy taken in conjunction with a stem cell transplant, a procedure that replaces diseased blood cells in the marrow with young, healthy ones (stem cells).

Multiple myeloma is often viewed as one of the “orphan” cancers, like ovarian,

BY MARY McIVER



esophageal and testicular, since it doesn't have the incidence level of the so-called "Big Four": lung, breast, prostate and colorectal. However, its profile is rising, simply because of the numbers. Says Dr. Shelley Naiman, head of the hematology laboratory at St. Paul's Hospital in Vancouver: "From my observations, the number of cases is not only increasing, but the people contracting the disease are getting younger."

Like Greg Orr, from Stouffville, Ont., who was 49 when he was diagnosed with the disease. And George Boudreau. "When I was diagnosed," he says, "I was told that multiple myeloma was a rarity under 60."

Boudreau was also shocked by the lack of options available. He was told that he had just one: a stem cell transplant. He agreed to have it; otherwise, he says, "I'd have been dead in 24 months." Essential to the procedure is a drug called Neupogen® (filgrastim), which triggers the bone marrow into producing stem cells, and teases them out of the bone and into the blood so they can be harvested. But since the drug, which costs in the neighbourhood of \$3,000, wasn't officially approved, it wasn't officially covered by British Columbia's health plan. Boudreau, who didn't have that kind of money, hit the panic button. "I knew I had to get it or else. I knew I had to fight for it." Suddenly, mysteriously, after he'd

gone to the wall more times than he cares to count, Boudreau got the drug.

What followed was a draining course of events, which Boudreau describes as being "as close as you can get to death without it killing you." Heavy doses of chemotherapy preceded the Neupogen® injections, followed by more chemotherapy. The effects of the chemotherapy were particularly debilitating, says Boudreau. "Chemo dries you out; you need liquids constantly. I was close to kidney failure at one point." Finally, in December of 1999, Boudreau successfully underwent the stem cell transplant and is currently in remission. But his battle is just beginning. "I have responded superbly to the transplant but I'm still under a death sentence. What happens when I go out of remission?" After intense research into the disease, he's finding answers south of the border where, he says, there are "dozens and dozens" of alternative



George Boudreau, a multiple myeloma patient from Kelowna, B.C., was shocked by the lack of options available in Canada to treat this disease.

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Multiple myeloma patient George Boudreau has little patience with such wait-and-see attitudes.



treatments. “Last year, I attended a symposium in California for people with multiple myeloma. Of the 300 people there, only two had had stem cell transplants. The rest were exercising other options.”

One of the most promising developments on this front is in the area of angiogenesis inhibitors. Angiogenesis literally means the growth of new blood vessels; as it applies to multiple myeloma, it refers to the growth of existing but virtually dormant capillaries. Normally these are no bigger than a hair, but a seemingly insatiable need for nourishment on the part of the myeloma cells prompts them to flourish. The idea behind the inhibitors, most commonly derived from thalidomide, is to attack the myeloma environment, discouraging the growth of these blood vessels and making it difficult, or even impossible, for the malignant cells to thrive. Starve them out, in other words. In pioneering clinical trials, these drugs had an impressive record of stabilizing or improving a patient’s condition. Other drugs in the works are designed to attack the myeloma cell itself, keeping it from bonding with the bone marrow, or to pump up the patient’s immune system.

In the U.S., several of these new drugs have either been approved by the Federal Drug Administration or are available on compassionate grounds. In Canada, the medical community is proceeding with caution. Dr. Keith Stewart, an oncologist at Princess Margaret Hospital in Toronto and a leading Canadian specialist in multiple myeloma, says some cutting-edge drugs are available here but certain conditions apply. “They’re only available as part of a clinical trial or through the emergency drug release program for people who have failed other treatments.” Currently Dr. Stewart is overseeing 12 active clinical trials, with three more in the offing, to test, among other things, the efficacy of anti-cholesterol drugs, angiogenesis inhibitors and immune-boosting vaccines. Meanwhile, he says, there are advances every day on the multiple myeloma front in terms of microbiological research, new therapeutic agents and supportive care.

Events aren’t moving quickly enough for George Boudreau. He’s determined to draw attention to the paucity of options in Canada, which he says behaves “like a third world country” when it comes to the diagnosis and treatment of cancer. It sounds paradoxical, but apart from his disease and a compromised

immune system due to the chemotherapy, Boudreau is in excellent health. His blood and urine are normal, there are no discernible tumours and “doctors everywhere are baffled by my good bones,” he says, not without irony. “I’m lucky. I’m well enough to fight, and I’m going to fight this every step of the way.”

In contrast, all the fight’s gone out of Stouffville’s Greg Orr. Since contracting the disease, he’s lost height and weight and is currently undergoing dialysis for kidney damage. He suffers from pain and constant fatigue, and he can’t stand up for more than an hour at a time. Not that this happened all at once. In fact, says his partner, Barbara Righton, the sequence of events has been an endless nightmare. “At every turn,” she says, “there was another complication, which turned into eight, which turned into 20. Every step had an octopus of ramifications.”

Orr’s troubles began one day in 1998, when he jumped up for a pop fly during a game of catch and thought he’d put his back out. He consulted a chiropractor, underwent physiotherapy and seemed to be getting better—until an MRI scan revealed that a growth of some kind was pressing on his spine. An operation was ordered at once: over 12 hours later, surgeons had removed the growth and the vertebra it was wrapped around, replacing the vertebra with a steel plate. The growth turned out to be malignant, Orr was diagnosed with multiple myeloma, and a stem cell transplant was recommended two months down the line. Orr decided to put the procedure off for a while, because, “I felt so good. Why go through months of agony, half a year out of my life?” It was a decision approved by his oncologist, who said that his myeloma was not yet compromising. “We were told that Greg’s disease was operating at a slow burn,” says Righton. Over the next couple of years, Orr took a fortifying agent to strengthen his bones, steroids to make him feel perkier and a mild course of chemotherapy to bring his white blood cell count down. He continued to work at his job at a car dealership, and experienced few symptoms. “Greg was tired at the end of the day but not dramatically so,” says Righton. Then, in late July of last year, the slow burn burst into flames. Orr was lifting some tires and suddenly experienced excruciating pain. “My bones were starting to break,” he explains.

Plans for the transplant got underway immediately. Orr was put on a stepped-up course of chemother-

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apy, and Neupogen® was doled out after a lot of annoying red tape. The stem cell harvesting was completed early this year, but shortly after that, Orr’s kidneys began to shut down. The transplant was postponed, and Orr was taken off chemo. As he gets weaker, Orr has been in and out of hospital; recently he was turned away because there were no beds available. Meanwhile, he’s been ordered to have his heart checked out regularly, which strikes Righton as the height of absurdity. “They put him on a treadmill to check his ‘pulmonary function,’ and the poor guy can hardly walk.”

Orr was once invited to participate in one of Keith Stewart’s clinical trials. He passed, because he felt too sick.

Orr’s situation illustrates an irony that colours the entire cancer landscape: the people who have the greatest stake in the fight against cancer—the patients—have the smallest voice. If they’re not too sick to do battle, they’re stymied by lack of information, delays in treatment, unavailability of new medications, inaccessibility to clinical trials. Individuals with cancer and concerned citizens alike must be granted a stronger role in the funding, research and policy-making decisions that affect their lives. ♦

Postscript: A month after being interviewed for this article, Greg Orr died. George Boudreau still shows no signs of cancerous activity.

The Multiple Myeloma Research Foundation and the University of Toronto are sponsoring a program on Multiple Myeloma. “Novel Therapeutic Approaches in the Treatment of Multiple Myeloma,” a free symposium for patients and their families, will take place on Thursday, September 20, 3:30 to 8:45 at Mount Sinai Hospital, 18th floor auditorium. Dr. Keith Stewart from Toronto is program director. For more information and to register email: themmrf@themmrf.org or call 203-972-1250.