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HOPE DIES LAST

A 37-year-old father of two faces the dilemma of a rare cancerous disease
One of the hardest things for Max Rose to face is that he no longer has the strength to keep up with his two preschoolers, including carrying them up the stairs of the home he built in North Vancouver.

It's especially painful for the 37-year-old construction superintendent to deal with the fact one of his sons will soon be learning to ride a bicycle.

"I won't be able to run after him. That's going to be tough. I think of all the things I'd like to do with my sons, and hopefully I'll be able to. But still."

Max, who was supervising more than 100 trades workers on Vancouver's Olympic village project

before he took medical leave late last year, is receiving intravenous chemotherapy for an extremely rare and virulent disease.

It's called adrenal cortical cancer.

Its rarity is part of what makes it so devastating. Since so few people are attacked by adrenal cancer, there has been virtually no research into targeted ways to treat it.

Adrenal cortical cancer strikes only one or two people per million. There are only about 600 patients throughout Canada and the U.S.

Adrenal cancer is considered an "orphan" disease because pharmaceutical companies calculate there is not enough money to be made researching drugs to combat it.

Still, U.S. Democratic Senator Edward Kennedy, who has brain cancer, is among the people leading legislative efforts that might address the dilemma of those battling rare and ultrarare cancers.

The fight against the clock is on for Max and those who love him.

Last November, when doctors finally figured out why Max was suddenly gaining weight, feeling tired and bruising easily, his adrenal cancer was discovered to be stage four, the most serious stage.

Since the cancerous tumours were too advanced for surgery, he was told it would be unusual for him to live more than a year. His treatment is considered palliative.

Max was undergoing his third round of intravenous chemotherapy when I talked to him and his wife, Stefanie WyerRose, in the combined livingroom/kitchen of their airy home. "I'm trying to stay as positive as I can. I can do that or I can be negative and curl up in the fetal position and give up, which I'm not going to do," said Max, sitting on a couch with energetic Cooper, 2, and Fisher, 1, bustling nearby.



Members of Max's extended family, including his father Chris Rose, a former Vancouver Sun editor, are doing everything they can to improve his chances.

Max's wife of six years, who overcame her own bout of skin cancer almost five years ago, has been doggedly researching adrenal cancer and pressing for more research and better treatment regimens.

But Max and those who care about him are coming face-to-face with a major obstacle in the Canadian and international medical communities. It relates to the ethics of triage.

That's the system of medical rationing that began on battlefields, where emergency doctors, nurses and resources were often in short supply.

Triage ethics developed with harsh simplicity: The soldiers considered most treatable were helped first. Those with the most grievous wounds were left until last, since their chances of survival were already poor.

Triage thinking continues to influence the apportioning of medical resources in North American society, where dollars are not infinite.

Sometimes it's called the ethics of scarcity. Still, people like Stefanie are working hard for Max and others with adrenal and related rare cancers — to raise their odds.

They have allies throughout the world. They also have ideas for reform.

Random misfortune

There are so many difficult things about the life-and-death struggle Max and his family are enduring. Most are existential: the pure random misfortune of being struck with this unusual cancer of the adrenal endocrine glands, which are located above the kidneys.

Max's adrenal tumours were found last fall to have spread to his left liver and lungs.

Other difficulties relate to the medical system. For reasons related to the rarity of adrenal cancer, Max wasn't diagnosed early.

If medical personnel had discovered sooner that his symptoms, including Cushing's syndrome, were signalling something more serious, the cancer would have been more treatable.

When Max was interviewed in late March in his home in south Lonsdale, a cherry tree was blossoming in the yard. Corners of the shiny hardwood floors were covered with the toy trucks and diggers that, true to their dad's vocation, Max's sons like so much.

Max misses his career at Metro-Can Construction Ltd. He has supervised the building of Vancouver's Chan Centre for the Performing Arts, SkyTrain extensions and other high-profile projects.

"It's really hard not working. I worked with those guys for 15 years. I made longtime friendships."

The cheery brightness of the living room, and the innocent lack of awareness of the children, belied Max and Stefanie's inner struggles.

Vancouver Sun Digital - The Vancouver Sun - 23 May 2009 - HOPE DIES LAST Page 4 of

4

<http://>

Adrenal cancer is not well understood

[From page C1](#) Despite his fatigue, Max was gracious, quick-witted and brave. And he and Stefanie had one bit of good news to pass on, the first in months, a CT scan had shown that one of Max's adrenal gland tumours had shrunk by three centimetres. Another one, on his liver, may also have decreased.

The fairly standard cancer treatment Max has been receiving from his oncologist, Dr. Sasha Smiljanic, was having some effect. But the treatment is not particularly targeted to adrenal cancer, since the disease is not well understood.

Max's prognosis remains grim. "It's like somebody pulled a rug out from underneath your feet." To add to the extended family's plight, Max's stepfather has grave prostate cancer. Stefanie, who works as a judicial assistant at B.C. Supreme Court, wears a purple "AC Warriors" bracelet to highlight the cause of adrenal cancer.

The family has been joined in the campaign to raise awareness for it and other rare diseases by two of the few British Columbians struggling with adrenal cancer. Megan McNeil, 18, of North Delta, has recorded a song with Nickelback producer Garth Richardson to support cancer victims. It's titled, *The Will to Survive*. (Unlike Max, McNeil has been able to benefit from surgery.)

Another British Columbian with adrenal cancer, Victoriabased wildlife biologist Karen Truman, is raising money for the B.C. Cancer Agency, but said she's disappointed she's not allowed to divert the money to rare-cancer programs.

Both Max's family and Truman discovered on their own that a scientist at the University of Michigan, Dr. Gary Hammer, is one of the few experts on adrenal cancer in the world. Truman, whose stage two adrenal cancer is not as advanced as Max's, paid out of her own pocket to fly to Michigan. Max paid to have his pathology report sent there. Hammer, who is experimenting with genetically based treatments for adrenal cancer, provided the medical analysis that led to Max's current chemotherapy regimen, which he receives at Lions Gate Hospital.

Smiljanic, the oncologist who treats Max, says "there is a really heated debate" about whether rare-cancer patients like Max should receive travel costs to visit specialized treatment centres.

"In the policy area, the question is where do you draw the line?" said Smiljanic.



Dr. Gary Hammer at the cancer centre at the University of Michigan. Dr. Hammer is an expert on adrenal cancer.



The B.C. Cancer Agency, said Smiljanic, has 40 to 50 oncologists treating patients with breast, colon and other common cancers with programs “that are the envy of other parts of the world.”

But Canadians with rare cancers aren’t in as strong a position as those with common cancers, in part because they “don’t have a strong lobby group,” said Smiljanic, who never had a patient with adrenal cancer until he met Max.

If Smiljanic was in Max’s shoes, he’d consider enrolling in an experimental clinical trial for adrenal cancer, such as those at the University of Michigan. Max, however, would have to pay for his own travel and accommodation.

‘A price on life’

Simon Fraser University applied ethicist Mark Wexler has nothing but empathy for Max and his family.

They are doing their duty, he said, which is to do whatever they can to quickly get the best possible treatment.

However, Max is running into a wider ethical conflict: Even though our culture says every human life is “invaluable,” society still places limits on how much to spend on an ailing individual.

“Some people ask, ‘Isn’t it hypocritical to admit we actually put a price on life?’ ” said Wexler.

But the hard reality is that society does put an economic value on a human life — in a way reminiscent of so-called lifeboat ethics.

If there is only so much room in a lifeboat to save those who are drowning, if there is only so much money to help the sick, Wexler said society is forced to decide who is allowed on the lifeboat.

University of Victoria medical ethicist Eike Kluge agrees with Wexler that health care systems can’t devote unlimited resources to people with rare cancers, while taking away from broader programs.

Kluge, who is being treated for prostate cancer, feels wholehearted sympathy for Max. Still, the professor of applied ethics said: “I often have to tell my students that not everything that is tragic is unethical.”

Dr. Charles Blanke, who heads the B.C. Cancer Agency’s systemic therapy program, said one of the hardest questions the organization has to face is how to fairly parcel out limited resources “so everybody gets at least a share of the pie.”

Even though the B.C. Cancer Agency is covering Max’s chemotherapy and will pay for out-of-province treatment, Blanke said the B.C. government won’t pay for Max’s travel and accommodation costs to attend clinical trials.

If Max isn’t able to finance his own travel to specialized treatment programs, Blanke said Max could complete his chemotherapy treatment and then, depending on the outcome, seek a referral to the Vancouver Cancer Centre.

Some oncologists at the Vancouver Cancer Centre focus on rare cancers, Blanke said, and could, “on a case-by-case basis,” try to provide targeted or even “highly experimental” treatment.

Still, the problem remains that adrenal cancers, unlike common cancers, don’t have proven treatment programs.

Level the playing field

In a North American society that so strongly emphasizes economic gain, patients with rare diseases can be left behind, said the University of Michigan's Hammer.

In an effort to aid those with rare cancers, Hammer has been busily consulting with officials connected to the proposed Cancer Alert Act in the U.S.

The Alert Act is being brought before the House of Congress by Kennedy and Republican Senator Kay Hutchinson. It calls for increased funding for research and treatment of cancer, including rare and ultra-rare cancers.

It makes sense, Hammer said, that armies of researchers around the world are focusing on common cancers, like that of the breast and prostate. Breast cancer takes the lives of about 45,000 North Americans a year, while prostate cancer kills roughly 30,000.

"But it leaves other patients with rare cancers as orphans, with little hope for research into their diseases let alone treatment. How do you level the playing field somewhat?"

"And how do you do that without breaking the bank? Our society and medical system shouldn't be based on only the survival of the fittest, on only social Darwinism."

Hammer has a series of arguments for increasing society's emphasis on rare cancers, which include those of the adrenal glands, saliva glands and tongue.

Hammer's first argument is that some of the most important advances in the field of cancer have been made studying rare cancers.

Discovering a targeted genetic-based therapy for adrenal cancer, which Hammer is working on through two clinical trials, could also help those with testicular and ovarian cancers.

Secondly, Hammer believes financial incentives could encourage research into uncommon diseases.

Government regulators, for instance, could approve rare-cancer drugs that are proven to shrink tumours even if they do not ensure long-term survival.

Pharmaceutical companies, he added, could also be given longer times to hold exclusive patents on drugs they discover to treat rare diseases.

Perhaps most importantly, Hammer calls for the establishment of rare-cancer "centres of excellence."

Since so few doctors in the world specialize in rare cancers, he thinks society's research and treatment efforts should be focused on leading international centres.

"Patients could be sent to wherever there is a centre of excellence. There could be reciprocal agreements between centres and even states and countries."

Some insurance companies have already financed sending adrenal-cancer patients to Hammer in Michigan. Other patients have paid out of their pockets.

Even though Dr. Smiljanic would support Max going to the University of Michigan, he cautioned it would not necessarily solve Max's rare-cancer dilemma. That's because taking part in clinical trials can be hit and miss.

It's possible, Smiljanic said, Max could end up, without his knowledge, in a "blind" control group that simply receives the "old treatment" for cancer — not the targeted genetics-based treatments, which are themselves unproven.

Still, such clinical programs are important, at least for others.

"Sometimes the people who take part in clinical trials don't get to reap the benefits themselves," Smiljanic said. "But they will benefit someone in their situation in the future."

Fight for his life

Despite the ethical, financial and political complexities around finding a targeted treatment for people with rare cancers, SFU ethicist Wexler is “totally supportive” of Max’s family’s efforts to fight for his life.

Families, Wexler said, operate by an unwritten code that says parents should die before their children. Max’s loved ones, he said, are utterly justified in seeking help from whatever “deep pockets” they can, including government and private donors.

For his part, Max is not about to give up.

And he says he’s not afraid. His attitude, and that of those around him, is reminiscent of the Russian proverb, “Hope is the last to die.”

Max notes with a smile that doctors are surprised he has not lost his thick red hair from his chemotherapy treatments. “I tell them it’s because I’m too stubborn.”

Max also appreciates “the one good thing” to come with his excruciating diagnosis — he’s able to spend extra time with his sons.

Max knows what he’s up against with his little-understood disease. He knows how medical rationing works. He’s been told his chances.

But he’s looking forward to a fishing trip this June near Merritt with his dad, stepbrother and father-in-law. And he’s treasuring every moment with his family.

Everyone who cares about Max is yearning for him, and others like him.

They are realistic, but they will never stop hoping.