Sarcoma patients today have available an array of therapeutic resources that did not exist when Kristen Carr was diagnosed with retroperitoneal liposarcoma in 1989.

Yet it is very likely that patients and their caregivers still will not have a very good understanding of their problem: Why they have it, how it originated, or even, for that matter, some of the possible solutions.

The three essays by Kristen Ann Carr fellows in this issue of Sarcoma Update represent a treasure chest of information that she — and we, her family — would have found immeasurably important as we struggled to understand what was going on in her body, what it portended, and what we could do about it.

The last question is the most important, and surgeon Kaitlyn Kelly's article gives us an outline of the history of this particular (and particularly important) set of tumors, making it clear that the big answers are still in the future.

But her article is also full of hope — the promise that the hard work that she and all our fellows have done and are doing will, in a foreseeable future, result in patients like Kristen having a far greater likelihood of long-term survival.

Surgeon George Plitas outlines some of the most basic issues that people with such rare tumors must confront in a different way compared with issues confronted by those who have more common forms of the disease. The idea of a cancer surgeon who specializes in breast cancer but has knowledge of how to operate on sarcomas would have seemed like science fiction in 1989. Actually, it would have been science fiction.

Not any more ... and that’s the main reason we created the Kristen Ann Carr Fellowships. But the greatest gift that Dr. Plitas offers patients and their loved ones is a description of what’s happening biologically in language that’s as plain and comprehensible as possible. He presents the fundamentals of sarcoma at a human scale.

We can remember being confused by almost everything that he outlines here, and even now, the refresher course is most welcome.

Physician-scientist Sri Ambati writes about the future of sarcoma treatment. His article is a message from the land of hope and dreams. It’s a message about the future presented to all those who bear the burden of knowing the reality of sarcoma today.

Sarcoma takes, even when the outcome is successful (as in fact it often is). Dr. Ambati isn’t just hoping and dreaming. His account is as based in the real world of treatment and research as the others; it just takes on the essential, however risky, task of looking even further ahead.

We are very very proud to sponsor every issue of this newsletter, and never has that been truer than with this issue. We hope that everyone who reads these articles learns not only that there is great reason for hope, but also how hard these physicians and their colleagues work, day in and day out, to make each patient’s diverse journey arrive in the light.

— BARBARA CARR AND DAVE MARSH

**Time for Targeted Therapies in Sarcomas**

**BY SRIKANTH REDDY AMBATI**

In the 100 years that research has been done on sarcomas, we have come a long way in treating them. Before the era of chemotherapy, surgery cured less than 20 percent of patients with a localized Ewing sarcoma or osteosarcoma. Oncologists quickly understood that many relapses were caused by metastases.

Once chemotherapy was introduced, oncologists could attempt to kill metastases, in addition to controlling the primary tumor. Overall survival improved significantly, up to 60 to 70 percent for Ewing sarcoma and osteosarcoma treated by surgery combined with chemotherapy.

Since the 1980s, however, we have seen little additional progress with traditional chemotherapy.

Fortunately, recent advances in our understanding of the biology of various tumors, including sarcomas, have allowed us to develop targeted therapies. A targeted...
therapy is a drug designed to inhibit one of those specific processes or pathways.

For some tumor types, we know the specific biologic processes or pathways that have become abnormal and that allow the abnormal proliferation of cancer cells. Targeted therapy aims to inhibit cancer cell proliferation with minimal damage to the normal tissues. The classic example of a targeted therapy is imatinib (Gleevec®), a drug that targets a few related proteins; imatinib can control disease in many patients with chronic myeloid leukemia (CML) and gastrointestinal stromal tumor (GIST), a type of stomach cancer.

However, sarcomas pose a challenge for treatment with targeted agents, because they generally have numerous alterations or multiple genes that are inappropriately regulated. Therefore, shutting down the overactive machinery in a sarcoma cell will require a combination of agents. An additional challenge is that there are currently more than 50 known types of sarcomas, and a range of agents will be needed to target them all.

Knowledge of genetic dysregulation in Ewing sarcoma has led to an explosion of targeted agents, with some of them showing promise in animal models of the disease or in patients. For example, Ewing sarcoma cells depend on IGF1R for their survival and proliferation. Strategies using an antibody to inhibit IGF1R, in combination with other pathway inhibitors, have shown efficacy for some patients with Ewing sarcoma.

In cell biologist Malcolm Moore’s laboratory in the Sloan-Kettering Institute, I am working on novel targeted agents for Ewing sarcoma. Ewing sarcoma is the second most common cancer that arises in the bones, yet it can occur in soft tissues as well. The typical patient is young; more than half are adolescents.

Ewing sarcoma is very aggressive, and only 70 percent of patients survive it. Patients with relapsed tumors have extremely poor survival (around 25 percent). Such patients have already gone through one or more chemotherapies that failed to control the cancer. Therefore, we are testing targeted agents.

One type of targeted agent we are investigating is inhibitors of heat shock proteins (HSPs). HSPs are a class of related proteins present in all living organisms. They help other proteins fold into their proper shape and help maintain that shape. Proteins are more likely to unfold in stressed cells, so stressed cells increase their production of heat shock proteins. Typically, cancer cells are under extreme stress and need large amounts of HSPs, particularly HSP90 and HSP70, to survive.

HSPs are known to be overabundant in sarcomas that are resistant to chemotherapy. An additional difference between tumor cells and normal cells is that HSPs in tumor cells are in an activated state. Activated HSPs are more prone to be bound and inhibited by HSP inhibitors than are nonactivated HSPs, so tumor cells are more susceptible to HSP inhibitors.

Researchers at MSKCC have developed a new generation of HSP90 and HSP70 inhibitors that have greater effect on tumor cells and less effect on normal cells. We are currently testing these new inhibitors both on sarcoma cells grown in a culture dish and in mice bearing human tumors.

A limitation of therapy with a single drug is that the tumor may develop resistance to the drug. When we use a drug to inhibit one pathway that the tumor is using to grow and survive, a tumor cell may mutate in a way that allows it to use a different pathway. The cell, now resistant to the drug, resumes proliferation, leading to relapse.

To address the problem of drug resistance, we are testing combinations of targeted agents to inhibit multiple pathways at the same time. Understanding the activated pathways in a tumor is key in designing the appropriate combination of agents. For example, inhibitors of insulin-like growth factor-1 receptor (IGF1R) have shown efficacy in a subset of patients with Ewing sarcoma. However, some of the cancer cells become resistant to IGF1R inhibitors by turning on the mTOR pathway. When an mTOR inhibitor was added to an IGF1R therapy, more patients benefited from treatment. Another study has shown that tumor cells that become resistant to IGF1R therapy have increased numbers of HSPs.

Researchers around the country are working on various other targets in sarcomas. One of these is pazopanib (Votrient®), an agent that inhibits several different proteins known as tyrosine kinases. Pazopanib was approved in 2012 by the FDA for treatment of advanced soft tissue sarcomas. Several clinical trials are under way to study the safety and efficacy of pazopanib in combination with mTOR inhibitors or traditional chemotherapy agents.

Most of the clinical trials in progress for patients with Ewing sarcoma are specifically for patients with relapsed or refractory tumors. If the agents prove to be effective, they will be integrated into first-line therapy with the aim of preventing relapses.

In conjunction with clinical trials, researchers are identifying biomarkers that can predict whether an individual’s tumor will respond to a drug, with the goal of delivering targeted therapies to the right patient. Improvement in genetic sequencing technology is helping us to study the genome of cancer cells at a greater depth and find differences among individual patients’ tumors. We are hopeful that the current advances will make the idea of “personalized medicine” a reality.

Srikant Reddy Ambati, MD, is a pediatric oncologist at Memorial Sloan-Kettering Cancer Center and is the recipient of several awards for his research and teaching. He received his MBBS and MD degrees from Institute of Medical Sciences, Varanasi, India. He did his residency in pediatrics at Miami Children’s Hospital, in Miami, Florida. He came to Memorial Sloan-Kettering for a fellowship in pediatric hematology/oncology in 2009. He also served as chief fellow in the Department of Pediatrics at MSKCC and Weill Cornell Medical College. He is currently doing translational research in sarcomas in Malcolm Moore’s Laboratory at Memorial Sloan-Kettering. He was awarded a Kristen Ann Carr Foundation Fellowship, a Young Investigator Travel Stipend award from American Society of Pediatric Hematology/Oncology and a Ewing Sarcoma Fellow Research Fund from Margaux’s Miracle Foundation for his work on Ewing sarcoma.
The Conundrum of Retroperitoneal Sarcoma: Why Treatment Recommendations Vary and Where We Are Today

BY KAITLYN KELLY

Approximately 11,400 cases of soft tissue sarcoma are diagnosed each year in the United States. About 16 percent of soft tissue sarcomas are located in the retroperitoneum, which is the rear part of the abdomen. Retroperitoneal sarcoma (RPS) is rare, and the tumors are also diverse, with more than 50 subtypes defined by their histology (the type and appearance of the cells in the tumor).

The rarity and immense diversity of RPS make it a difficult disease to study. Most of the data we have on RPS is from retrospective studies, which look at patients treated over previous years who received whatever therapy was prescribed by their treating physicians at the time. Data from retrospective studies, while important, is not as strong as that from randomized controlled trials, where treatments are highly standardized to focus on specific experimental questions and to minimize the effect of other variables.

Retrospective data can still be used to guide treatment recommendations, but it carries inherent weaknesses that make it open to interpretation. The result for patients is that recommendations for the treatment of RPS vary dramatically among hospitals and physicians. A patient with RPS seeking a second opinion will frequently receive different treatment recommendations from the two providers. This can be very confusing for patients and their families.

Despite the inherent difficulties in studying RPS, however, multiple clinical and basic science studies have been completed in recent years, resulting in advancements in treatment for this disease.

Treatments available for RPS include surgery, radiation therapy, and chemotherapy. There is general agreement that the best therapy, when it is possible, is complete surgical removal soon after RPS is diagnosed. Radiation therapy and chemotherapy can be used to shrink the tumor to make complete resection more likely, or as the main treatment when surgical removal of the tumor is not possible. How these three treatment modalities are combined and in what order they are given is controversial.

The goal of surgery for RPS is complete removal of all visible disease. This can be difficult, as most retroperitoneal sarcomas are quite large by the time they are diagnosed. Additionally, some types of RPS, for example liposarcoma, look and feel similar to normal tissues, so the boundary between tumor and normal tissue can be difficult to determine during surgery. Additionally, even when the tumor seems to be completely removed, there are often microscopic tumor cells remaining on the borders of these very large tumors.

Surgery for RPS has historically involved removal of the tumor with a margin of surrounding normal tissue, as well as any organs directly invaded by the tumor. Several recent studies have evaluated a more aggressive surgical technique, with removal of all organs adjacent to or in any contact with the tumor. These retrospective studies have found that the more extensive organ removal is associated with a lower risk of local recurrence of the tumor, but at the cost of increased complications from surgery and with no difference in long-term survival.

Given the limitations of these data, there is currently no consensus on the best surgical approach for RPS. Our practice at Memorial Sloan-Kettering is to remove only the organs directly invaded by tumor.

When RPS recurs after surgery, it is most often in the form of local recurrence (as opposed to metastasis), so recent efforts have focused on improving local disease control. Radiation therapy, like surgery, is a form of local therapy and has an established role in the treatment of high-risk soft tissue sarcoma in the arms and legs. RPS tumors, however, are adjacent to the bowels and other vital organs, making it difficult to deliver enough radiation to tumor tissue without injuring the surrounding organs. This problem is addressed by several recent advances in radiation therapy:

- Intensity-modulated radiation therapy, which uses three-dimensional imaging and computerized mapping to deliver more radiation to high-risk areas within the tumor while minimizing radiation to surrounding tissues.
- Proton therapy, which delivers radiation in the form of protons instead of the traditional photons. Protons emit their energy over a shorter distance than photons, allowing a more focused delivery of radiation to minimize radiation to surrounding tissue.
- Intraoperative radiation therapy with electrons, which is used at the time of surgery to focus radiation on areas where the surgeon must cut close to the tumor.

Although a definitive study has not yet been performed, these advanced radiation therapies appear to have fewer adverse effects when compared with traditional radiation therapy. We also lack a definitive answer to a very basic question: whether adding preoperative radiation therapy to surgery for RPS improves cancer control and survival.

A recent retrospective study compared RPS patients treated with surgery alone to those treated with surgery and advanced-modality radiation therapy. The study demonstrated an association between radiation therapy and improved local control. A new study, called STRASS, is a randomized controlled trial that will randomly assign RPS patients to treatment with surgery alone or with intensity-modulated radiation therapy followed by surgery. This trial should definitively clarify the value of preoperative radiation therapy in the treatment of RPS.

RPS has historically been considered a chemotherapy-resistant tumor because it seldom responds to traditional chemotherapies. More recently, researchers have been developing targeted therapies, which are drugs designed to inhibit specific molecules responsible for growth and cell proliferation in different cancers. Targeted therapies can be very efficacious and have fewer side effects than traditional chemo-

CONTINUED ON PAGE 7
Blurring the Lines: Metaplastic Breast Tumors

BY GEORGE PLITAS

We start with a tumor. How it is found varies: The patient felt it, the physician felt it, a scan picked it up, or maybe it was found accidentally while looking for something else. At this point it has no name, it’s just a tumor, which is a term used for a mass of tissue that is not normal.

How does a tumor get a name? It first needs to make its way to a pathologist’s microscope. It can be a piece of the tumor from a biopsy or sometimes the whole thing, removed by a surgeon. The pathologist then begins the naming process. At first it’s a general description of size, color, and consistency, and then the examination becomes progressively more detailed and specific.

The cells and the pattern they make are carefully detailed. Sarcomas are often composed of cells that are long and thin, classically described as spindle shaped. Clustering together, these cells form sinuous whorls that appear as if they were created from the brush of Edvard Munch. A tumor at this stage of classification may graduate to be called cancer, based on numerous characteristics such as how abnormal the cells are or if they invade adjacent structures such as nerves, lymph channels, or blood vessels.

At this point, the tumor generally is given a name. Sarcoma, carcinoma, and lymphoma are among the most common, but there are numerous others with multiple subclassifications. To achieve a level of even greater certainty, the pathologist tries to determine the normal counterpart from which this tumor originated. Sarcomas for instance can come from muscle, bone, nerve, and even fat. Breast carcinoma for the most part originates from the glands of the breast, and it can be further localized to either the ducts or the lobules that together make up the functional unit of a breast gland.

Apart from the gross description and the microscopic appearance, an added level of accuracy is obtained by using special stains to determine what proteins the tumor cells are making. Why such detail? It has broad implications for prognosis and treatment. The detailed pathologic description of a sarcoma allows one to use databases created by surgeon Murray Brennan at Memorial Sloan-Kettering Cancer Center to estimate, with ever-greater accuracy, the probability of recurrence, metastasis, and survival.

Work by Memorial Sloan-Kettering investigators Samuel Singer and Aimee Crago has revealed genetic alterations that are specific to individual types of sarcoma; these alterations have been targeted both experimentally and clinically. These findings are the basis of numerous promising clinical trials that are available only to patients whose tumor has been classified and given a specific name. The naming process therefore is critical for dictating patient care.

As a breast surgeon at Memorial Sloan-Kettering, I see patients with breast carcinoma and breast sarcoma, but upon occasion the line gets blurred. There is a category of breast tumors with characteristics of both sarcoma and carcinoma. These are described as metaplastic, meaning that one form is turning into another. Some are more carcinoma like and others are more sarcoma like. For instance, they may have the characteristic waves of long spindly cells seen in sarcoma but a special stain will also indicate that they originated from a breast duct, typical of a carcinoma.

How we treat a sarcoma is very different from how we treat a carcinoma. Sarcoma rarely makes its way to the lymph nodes but carcinoma frequently will, and so further...
A Mixed Blessing

BY KIM SIMPSON

As I finished reading the Summer 2010 issue of Sarcoma Update, I saw on the back the opportunity to share experiences with other patients and their families and I thought, “Why not?”

My story has to be unique for one reason. At the time of my diagnosis, I was 12 weeks pregnant. There were two things I thought would never happen to me: getting pregnant without fertility treatment and being diagnosed with cancer. Fortunately and unfortunately, both occurred simultaneously.

In January 2008, I found out I was pregnant. My husband, Greg, and I already had two children (Koen, 2, and Avery, 1), but they were conceived with the help of fertility drugs and artificial insemination. To merely say I was ecstatic, much less surprised, would be a gross understatement. But according to the calendar, our daughter was conceived on Christmas Day 2007, and I just knew it was meant to be.

Then in February, while showering, I felt a lump on the inside of my right thigh. It felt like a muscle, and since I just tried to start working out again, I thought maybe I pulled something, although it wasn’t painful.

At my husband’s insistence, I made an appointment to see my general practitioner. My doctor suggested it could be a lipoma, something benign, and she did mention something about a soft tissue sarcoma, which she was “sure” it wouldn’t be, but she thought we should do an MRI to rule it out. Because I was pregnant, the MRI couldn’t be done until I was 16 weeks along, so I had an ultrasound instead.

Subsequently, I was referred to a surgeon to see what he thought of the matter. He was very concerned that it might be a malignant lymph node, and said a biopsy should be taken right away. My surgery was scheduled for the following day. I was scared to death because it was early in the pregnancy and because I wondered, if this was a malignant lymph node, where did it originate? Relief was short lived after the surgery. The malignant lymph node was ruled out immediately as the surgeon was able to remove the entire “gross” tumor. Three grueling weeks later, a diagnosis of a myxoid liposarcoma was returned.

What? There wasn’t a lot of information on the Internet, and what was available sounded pretty scary. I had to learn more, so a follow-up visit with the surgeon was scheduled. He told me since there weren’t clear margins from the original surgery, a second surgery would have to be done so all the sarcoma cells could be removed.

It wasn’t a fast-growing tumor, however, so we could wait until after I delivered the baby to do the second surgery. Radiation would probably be required after the procedure, so we were referred to a radiology oncologist. Most of our questions were concerning breastfeeding and being around my children after the administration of the radiation.

About a week later, I had my appointment with the radiology oncologist. I could barely see to drive home after this appointment because of the tears in my eyes. She told me these tumors are very unpredictable, and the fact that some of the cells were still present was very concerning to her. She recommended terminating the pregnancy and getting the radiation treatments as soon as possible.

I asked about doing the second surgery to remove the remaining cells before I delivered as opposed to after. She said that we could do that; however, radiation would still be needed after this surgery and the effects it would have on the baby were unknown. So I was left with this decision: end this pregnancy, which was a miracle to begin with, so I would have a greater chance of seeing my other two children grow up, or take the chance of leaving two or maybe three children motherless.

We decided to get a second opinion. I was left wondering why the surgeon seemed so relaxed about this, but the radiology oncologist so aggressive. After several more chats with the surgeon and maternal-fetal medicine experts, it was suggested that I go to MSKCC in New York City.

In the county where we live there are around 20 cases of sarcoma diagnosed annually. I needed to get to someone who was much more familiar with treating these types of tumors.

To be honest, I had never heard of Memorial Sloan-Kettering. Thankfully my husband had, and he immediately began checking them out — getting directions, booking hotels, planning the 200-plus-mile trip from our home in Hanover, Pennsylvania. I was busy, too, gathering surgical notes, ultrasound and MRI reports, and making an appointment with Memorial Sloan-Kettering sarcoma expert Samuel Singer.

It was a very overwhelming and scary time. What would he say? Would he offer me any hope or would we make this incredible journey only to return with the same decision to make? This was a torturous time for my husband and me … the fear of the unknown.

Well, there was only one way to find out. My first appointment with Dr. Singer gave me more hope and relief than I had had since finding out I had cancer. (My husband claims he was relieved the instant he saw Dr. Singer enter the examination room.) Dr. Singer told me I was his 12th case of liposarcoma that day (as opposed to the 20 or so our local doctors see in a year).

My husband and I went over the course of treatment that I was given at my local hospital. Dr. Singer told me my type of tumor isn’t even treated with radiation, as it would be an extreme overtreatment. He also told me that it was absolutely ridiculous that anyone would even suggest terminating my pregnancy over this. Moreover, he told me we could safely do surgery in a few weeks. I knew I was in the right place, and that this guy knew his stuff. The smile on his face told me so.

On May 5, 2008, I had my resection surgery at Memorial Hospital on York Avenue. The surgery revealed no additional sarcoma cells, although I lost a decent portion of my thigh. The rest of the pregnancy, however, went along just fine. And my other two little ones would have their mom.

Recovery took a couple of months. I’m sure being increasingly pregnant during this time probably extended recovery, but
that 19 years later we would ALL still be
we know that we would be so fortunate
out, and that has truly been a blessing. Did
did not accept
who have thought cancer could be funny?), and love to the whole
We assembled our “A Team”: my father
but also the long-term well-being
last, but certainly not least, my bride of seven
of course, but also the long-term well-being
of his daughter and grandchildren. Last,
and who brought lightness, brightness, a sense
of humor (who would have thought cancer
could be funny?), and love to the whole

Our first visit with Memorial Sloan-
Kettering surgeon Patrick Boland was long.
We heard a blur of cancer words and asked
about chemo, radiation, and surgery. Dr.
Boland’s answers were frightening. This
cancer needs to be cut out, and the first
thing we have to find out is how much we
are going to amputate. Amputation seemed
like such a barbaric, impractical way
to deal with cancer. We wanted the new
treatment, the cutting-edge treatment, not
the “chop off body parts” treatment.

Dr. Boland operated, removing two
fingers on my left hand along with a good
portion of the palm as well. He made no
promises, but only said, “Let’s keep an eye
on this.” In the meantime, we had to deal
with real-life issues: the lack of income
while recovering and, with that, feeding
a family of five and keeping a roof over our
heads.

Believe me, food shopping loses its
allure when, as you are putting the generic
macaroni and cheese into the wagon, you
know you will be using a credit card to
pay for it. And you have no clue when or
how you will be able to pay it off. So does
paying the mortgage while dealing with
the real concern — that we are a clinic
visit away from a death sentence. Our
family and friends flew into action, taking
care of the girls, feeding us and bringing
us groceries, and driving us back and forth
into the city for the appointments.

In the midst of all this we were afforded
the gift to appreciate life, day in and day
out, and that has truly been a blessing. Did
we know that we would be so fortunate
that 19 years later we would ALL still be
here (plus an additional daughter)? Of
course not, but here we are. Although it
was more an unconscious effort at the
time and not a life plan, we changed our
focus from money and career to family
and fun. We still never let a day go without
appreciating what we have, and being
thankful for the time we have been given.

In our family, cancer has always been
faced with humor. My children were
six, five, and five weeks old when I was
diagnosed. We had to laugh, if only to help
them feel safe. It has always amazed me
how comfortable children are at asking
scary questions and being comfortable
with the answers. When Dr. Boland
amputated my hand he left a thumb,
my ring finger, and pinky — the kids called
it “The Claw.”

As the years went on, other children
— nieces, nephews, friends of my kids —
would ask, “Hey Mr. D., what happened
to your hand?” The tall tales have ranged
from shark bites to getting it chopped off
in a poker game to biting it off when I was
hungry. Eventually I tell them the truth,
that the fingers were sick and making me
sick and I am better off without them.
The kids want to know if I have trouble tying
shoes, which I still do. We have laughed
at some of the most terrifying parts of this
because it is harder to be scared when
you laugh. We also celebrate every single
piece of good news. We go out to dinner
to celebrate or we get a small Carvel cake
and we stay grateful, because each day is
such a gift.

My family and I just participated in this
year’s Cycle for Survival, raising more than
$8,000 for research in rare cancers. I was
asked to ride the Team Fearless bike in
memory of Jennifer Goodman Linn, who
lost her battle with a rare cancer. Moments
like this remind me how very lucky I am
to be here and how much more I want to
give back to the community that helped
save my life. My daughters, now 25, 23,
18, and 17, are the most amazing girls you
could ever meet, and we all participated in
this year’s Cycle together with our whole
hearts. They make each day worth the
fight and remind me that my legacy will
continue long after I am gone.

Over the past 19 years the one issue that
has continued to trouble me was the “Why
me?” aspect of being diagnosed with a
life-threatening illness. This is something
that contributes to the survivor’s guilt that
continues to linger. I found the answer to
that when, 11 years later, my brother-in-
law’s little sister, Stacie (who helped us
care for our kids while I was sick), was
diagnosed with cancer in her spine. Stacie
had recently graduated college, started her
first job teaching in a school in September,
and was diagnosed in October.

My wife and I spoke with Stacie’s family
and recommended they see Dr. Boland.
As is true with many people, they were
concerned that getting into the city to
see a doctor would involve an exorbitant
amount of time and money. And, like
many, they clung to the familiarity that
comes with being treated by a trusted
family doctor. As part of our life’s plan,
my wife and I have made it our goal
to annoy and harass anyone who is
hesitating to “make the trip” to Memorial
Sloan-Kettering until they at least go for a
second opinion.

Dr. Boland diagnosed her with both
chondrosarcoma and osteosarcoma in her
spine. Had I not gone through my own
experience and taken the steps that put
us in the best position for a successful
recovery, then we may have never been
in the position to direct Stacie to MSKCC
and Dr. Boland. Stacie suffered through
some very painful surgeries, debilitating
treatments, and rehabilitation, all under
the care of, and with the respect and
dignity synonymous with, the staff of
MSKCC. She was an inspiration to many.
She eventually lost her fight in June 2009,
but all of what was learned about these
diseases as a result is part of her legacy.
Her smile is etched in my memory. Did
Stacie’s journey answer the question
“Why me?” In a small way it did.
therapies, which are aimed at all rapidly dividing cells.

Advances in gene-sequencing technology have enabled scientists to identify the molecules responsible for tumor growth — molecules that could serve as the targets of targeted therapy. Investigators at MSKCC are currently leading the Sarcoma Genome Project, a collaborative effort to molecularly characterize different types of soft tissue sarcoma (including RPS). Patients undergoing surgery for soft tissue sarcoma at MSKCC are asked to give consent for a portion of their tumor to be used for this genetic analysis.

As an illustration of what this analysis can accomplish, liposarcoma, the most common type of RPS, has been found to have too many copies of part of chromosome 12. One of the genes in this chromosome region is CDK4, and, because the tumor cells have too many copies of the gene, some of them produce abnormally large amounts of CDK4 protein. A drug that inhibits CDK4 is currently being tested in a clinical trial for patients with advanced well-differentiated and dedifferentiated liposarcoma.

RPS is a rare and complex disease. Because there are no standardized treatment guidelines, it is important for patients to seek treatment at high-volume hospitals where physicians have experience treating RPS. In addition, patients should seek out hospitals where they will have access to a multidisciplinary team of surgical, radiation, and medical oncologists, who can work together to give the patient full advantage of the advances in treatment methods and to determine the best course of therapy.

Kaitlyn Kelly received her medical degree from New York Medical College in 2004. She did her general surgery residency at the University of Wisconsin and a two-year basic science research fellowship at MSKCC where she worked with oncolytic viral therapy for cancer. Upon completing her surgical oncology fellowship at MSKCC in July, she will go to the University of California, San Diego, as an assistant professor of surgical oncology.

George Plitas is a surgical oncologist who specializes in the treatment of patients with breast tumors. In addition to taking care of patients he is also actively engaged in scientific research. In the laboratory, he studies ways in which the immune system can be activated to specifically target cancer. He also has an interest in broadening the understanding of breast sarcoma. The study of these unique tumors offers an opportunity to gain insight into the reasons why sarcomas form both in the breast and throughout the body.

On September 4, 2008, Libby Carroll was born; 6 pounds, 2 ounces, and just as healthy as could be. We both made it. And every time I look at her, I think of the amazing journey we made together, and I am so thankful for that second opinion. There was never a chance I was going to end her life for mine. As a mom, you sometimes have to make tough decisions.

Little did I know I was going to be blessed with being a mom for a fourth time. To our surprise, in September 2010, I found out I was pregnant again. Molly Elizabeth was born on May 29, 2011. Greg, Koen, Avery, Libby, Molly, and I still live in Hanover. Our lives are busy, but we wouldn’t have it any other way.

These life-changing events happened for a reason. I never doubted that. If it weren’t so, I never would have ended up at MSKCC. I would have had unnecessary radiation. And I never would have gotten the opportunity to share my story with you. When I was diagnosed with cancer, I feared for my own life, but especially for the life of my unborn child. Now life with a family of six has settled in to a normal, somewhat predictable routine, and I’m loving every precious minute of it!

Kim Simpson is a part-time stay-at-home mom and a part-time microbiologist. She works in a laboratory at York Hospital in York, Pennsylvania. She has worked there since 1997 and met her husband there as well. She is currently five years cancer-free and doing very well. She returns to Memorial Sloan-Kettering every year for follow-ups. In her “spare” time, she enjoys being with her family and volunteering at her children’s school.
We Want to Hear from You!

Sarcoma Update is designed to answer your questions and concerns about issues related to sarcoma. While our primary goal is to provide information regarding the medical and psychological aspects of having sarcoma, we also hope to provide a forum for patients and caregivers. We invite those readers who have had sarcoma as well as their family members and friends to share experiences with other readers. Send your stories, thoughts, comments, and concerns to:

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Or send an e-mail to: lesseri@mskcc.org.
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SARCOMA MEDICAL SERVICES

* Physician Referral Service ................................................................. 800-525-2225
  * For people who wish to come to MSKCC for prevention programs, diagnosis, treatment, or a second opinion.

  Radiation Oncology ................. Kaled Alektiar ........................................ 212-639-7981
  Surgical Oncology ................. Murray Brennan .................................. 212-639-6586
  Samuel Singer ............................................. 212-639-2940
  Aimee Crago .................................. 212-639-4807

  Medicine ......................... Mary Keohan .......................... 212-639-2809
  David D’Adamo ................. 212-639-7573
  Gary Schwartz .................. 212-639-8324
  Richard Carvajal ............. 212-639-5096
  William D. Tap .................. 212-639-5720

MSKCC SUPPORT SERVICES

  Department of Social Work ............................................................... 212-639-7017
  Resources for Life After Cancer .............................................. 646-888-4740
  Department of Psychiatry and Behavioral Sciences ................................ 646-888-0100
  Genetic Counseling ................................................................. 646-888-4050
  Integrative Medicine Center .......................................................... 646-888-0800
  Chaplaincy Service ................................................................. 212-639-5982

* Cancer Information Service .................................................. 800-4-4-CANCER
  *General information provided through a National Cancer Institute-funded program.
  Callers from outside the New York State office’s service area will reach another regional office.

* Patient Representatives ................................................................. 212-639-7202
  *For issues relating to MSKCC service to patients and families

Sarcoma Update
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New York, NY 10021