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2008

2007 Cancer Care Annual Report

Children's Mercy Hospital

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Discovery. Research. Treatment.



2007 Cancer Care Annual Report
Focus on Osteogenic Sarcoma



Children's Mercy
HOSPITALS & CLINICS
www.childrensmercy.org

Now, slightly more than 50 years into the battle of childhood cancer with the use of chemotherapy, the cure rates have never been higher. But they are not high enough. Only through ongoing and future research can we achieve our ultimate goals of 100 percent cure and perhaps even prevention in the years to come. It was nearly 20 years ago when it was first shown in clinical trials for osteosarcoma that the use of chemotherapy in addition to surgery increased the cure from 17 to 66 percent. Children's Mercy Hospitals and Clinics is an active participant in these efforts and as such makes available to our patients the newest approaches to therapy being investigated. This is possible through the availability of a number of clinical trials available at Children's Mercy, one of the larger Childhood Cancer Centers in the Children's Oncology Group, the National Cancer Institute's cooperative organization for childhood cancer research.

This year's Annual Report for the Children's Mercy Cancer Center focuses upon our most common bone tumor in children, Osteogenic Sarcoma aka Osteosarcoma. This cancer is seen most often in adolescents but may be seen in children as young as 5 years or less. From the following discussions and the age and developmental range of the children affected by this cancer, it is clear that a multidisciplinary and multi-specialty team is critical to the ultimate successful cure and rehabilitation of the child. Just as critical is an experienced team, trained and focused in the nuances of the developing child with cancer of the bone. We are proud to have such services available at Children's Mercy Hospitals and Clinics, and invite you to read further about them in this year's report. We hope this provides you with a clear picture of all that can be gathered together here in the fight against this childhood cancer but also we invite you to contact us with any and all questions that you might have.

Our best wishes to you and the families of our patients,



Alan S Gamis, MD, MPH
Chief, Section of Oncology
Division of Hematology/Oncology &
Bone Marrow Transplantation



Dear Friends,

I can't imagine what a parent must feel when he or she hears a doctor say, "Your child has cancer." Fortunately, most of us as parents never have to deal with hearing those words. But for those who do, the oncologists and staff at Children's Mercy Hospitals and Clinics are able to offer a complete arsenal of ways to treat these diseases, as well as the emotional support and family-centered care that provides support not only for the patient, but for the entire family as well.

Our Cancer Care Annual Report tells the story of how our staff does just that. This year's report includes a special focus on our work in patients with osteogenic sarcoma, although we provide the same depth and breadth of care to all of our patients with a variety of cancer diagnoses. I hope you will take a few moments to read this report and learn more about how our nationally-recognized pediatric oncology program provides exceptional care for these children and families.

Our program not only cares for the children with cancer of today, but we are actively engaged in a wide variety of research programs to improve the treatments available to the children with cancer of tomorrow, and hopefully someday to find a cure for this disease. We also are active in educating our future pediatricians on the best ways to provide medical care for children with cancer.

There have been tremendous strides made in recent years in improving the treatments and the outcomes for children with cancer, as you will read in the following pages. And everyone in the Children's Mercy oncology program is committed to continuing that progress in the years to come.

Sincerely,



Randall L. O'Donnell, PhD
President and Chief Executive Officer



THE DIVISION OF HEMATOLOGY/ONCOLOGY

Close to 2,000 children are seen each year by the physicians and other health care providers within the Division of Hematology/Oncology. The vast majority of these are patients seen for treatment or follow-up of their cancer, but the Division's Sickle Cell and Hemophilia programs each have nearly 300 active patients that are seen. Patients with other hematological issues or diagnoses are also routinely seen by physicians.

The comprehensive services of the Division allow children and families the well-rounded care that they need during challenging times. As patients progress from diagnosis, to treatment, to follow-up care, specific needs are continually addressed by different members of the team.

During the year, ten physicians and three fellows provided care to the patients seen in the out-patient clinic and in-patient floor. During 2007 1,191 admissions occurred on the inpatient floor, and there were 8,816 clinic visits. Each physician actively participates in the many clinical trials that are available to children who are treated at Children's Mercy. Through professional organizations and research links such as the Children's Oncology Group, Centers for Disease Control, and the National Institutes of Health, physicians provide cutting edge therapy to all patients.

Advance practice nurses, each working alongside a specific physician, coordinate patient care to ensure that all aspects of their unique case is carried out efficiently and effectively. Whether it is scheduling appointments outside of the Division, sifting through insurance issues, ensuring treatment protocols are

followed, or providing hands-on clinical care, the advanced practice nurse is a vital team member for the Hematology/Oncology family.

The nursing team that cares for patients in the clinic and on the inpatient floor number over 100. Nurses provide direct care to patients, administer chemotherapy and other treatment drugs, provide administrative and clinical record-keeping, and are a comforting presence during an oftentimes stressful period. Many of the nurses within the Division have received advanced certification in their specialty. All nurses receive continuous specialized training during their tenure.

Administrative and research support complement the work that the clinical providers give. Schedulers, administrative assistants, research coordinators, and data managers work behind the scenes, but provide integral services to both patients and families. Schedulers assist in keeping patients on track with appointments with as little life disruption as possible. Administrative assistants make sure that information is exchanged appropriately among caregivers inside and outside the hospital. Research and data

management staff coordinate the more than 100 clinical trials that are offered throughout the Division.

Care and commitment for the children treated does not stop at the clinic and inpatient doors. Support throughout the hospital allows each patient access to the other specialists who can significantly impact their care. Whether it is a surgeon, endocrinologist, neurologist, or pathologist, all specialists are available to assist in the management of patients as needed. Nutritionists follow patients carefully along the treatment course. Psychologists, social workers, chaplains, music therapists and child life specialists work with the whole child and the whole family to create positive experiences and outcomes. Volunteers offer comfort and care to children, family and staff throughout the hospital.

We hope that this report highlights the work of the Division of Hematology/Oncology. We especially hope that it offers a picture of the dedicated team that is required to bring the best care to our patients. At Children's Mercy, we are lucky to have one of the best there is!

Division Chief - Gerald Woods, MD

Division Manager - Sue Stamm, RN, MSN, CPNP

The Section of Oncology

Section Chief - Alan Gamis, MD

o **Oncology Service: Director - Alan Gamis, MD**

o **Bone Marrow Transplant Service: Director - Jignesh Dalal, MD**

The Section of Hematology

Section Chief - Gerald Woods, MD

o **Sickle Cell Disease Service: Director - Gerald Woods, MD**

o **Regional Hemophilia Center: Director - Brian Wicklund, MD**





Jonathan (seated) is all smiles as he and his family celebrate at his going home party.

JONATHAN STEPP A STEPP IN THE RIGHT DIRECTION

He sang. He danced. A writer and an athlete, Jonathan Stepp a Shawnee Mission East high school student did it all. As a cross-country runner, Jonathan noticed that his leg didn't heal after a long run. After testing and the removal of a tumor, the doctor talked to his Dad, a local neurosurgeon. Jonathan was diagnosed with osteosarcoma. His initial shock was soon replaced by peace from his strong faith as he waited a week to tell his friends.

Losing 12 pounds in his first week of chemotherapy he continued with 17 more rounds. In addition to running, Jonathan had played tennis, basketball, soccer and wanted to be a football punter, but the crutches from his knee replacement surgery were one more reminder that he was benched. Instead he worked hard to keep up with his studies as he utilized the school district's Home Bound program for the next four to five months.

Jonathan's hard work paid off as he learned that he was cancer-free shortly after his 17th birthday. What a GREAT birthday present.

Jonathan has many heroes. As an avid athlete himself, Jonathan was moved by fellow athlete Lance Armstrong in his race to overcome cancer. He is also inspired by J.K. Rowling, the creator/author of the Harry Potter books as Jonathan is working on his own book about his experiences with cancer. His interest in medicine was also sparked by his own encounters and he plans to volunteer as a Care Assistant at Children's Mercy Hospital. This will give him an opportunity to see things from the other side of the bed. With restored hope, Jonathan still holds on to his dream of becoming an ESPN anchor, living and loving life cancer-free.



Cancer registration and subsequent follow-up of patients is a standard practice at Children's Mercy Hospitals and Clinics (CMH). The Cancer Registry collects data in relation to cancer cases and benign tumors that are diagnosed and/or treated at Children's Mercy Hospital. The data collection requirements of the Commission on Cancer (CoC) of the American College of Surgeons as well as the Missouri Cancer Registry are maintained. This information is then reported to the Missouri Central Cancer Registry and the National Cancer Data Base. The Children's Mercy Hospital Cancer Registry operates under the guidance of the Cancer Care Committee, which is comprised of numerous staff physicians and clinical staff.

Cancer Registry information is vital for research in the prevention and treatment of cancer. Below are some questions and answers that may be beneficial in understanding the registry procedures.

PARENTS AND PATIENTS MAY LIKE TO KNOW:

WHY is there a cancer registry?

- All health care facilities are required by federal and state law to provide data for all patients diagnosed and/or treated. CMH data is sent to the Missouri Cancer Registry located in Columbia, MO. The state registry is under the direction of the Missouri Department of Health and Senior Services and contracts with the University of Missouri for operation. Data is also reported to the National Cancer Data Base.
- Data collected by Registrars allows state and national cancer registries to analyze and determine the impact of cancer for those residing in the state.

WHAT kind of information is reported?

- Patient demographics including age, gender, race, Hispanic origin, place of birth and residence.
- The location of cancer in the body and histological type of cancer.
- The extent of disease invasion within the body.
- Treatment including surgery, radiation, chemotherapy, hormone or immunotherapy.
- Current status of patients (cancer free or recurrence).

WHY am I getting yearly follow-up contacts?

- By following our previous patients we can evaluate the best treatment methods and allow for surveillance of patient outcomes. Successful follow-up must be maintained to provide optimal patient care.

WHAT about patient confidentiality?

- The Registry at Children's Mercy follows the same strict patient confidentiality guidelines as the State Cancer Registry. According to HIPAA privacy regulations, the State Cancer Registry is a public health authority authorized by law to collect and receive information for the purpose of preventing and controlling disease, injury or disability. A covered entity such as a hospital or physician's office can disclose information to the State Cancer Registry without specific individual informed consent. However, the identities of patients or specific facilities / physicians are not released by the State Cancer Registry without written consent from the individual or facility/physician.

**CANCER REGISTRY
2007 FREQUENCY OF DIAGNOSIS
TABLE I.**

**2007 CMH CANCER REGISTRY
HIGHLIGHTS:**

During 2007, 168 newly diagnosed patients were added to the registry. The most frequent diagnoses during 2007 were tumors of the central nervous system. A snapshot of the top five diagnoses in 2007 include central nervous system tumors, leukemia, lymphoma, neuroblastoma, and hepatoblastoma. See Table I for overall percent rankings.

The median age of patients diagnosed during 2007 was 6 years. The breakdown by age of diagnosis in 2007 are as follows: 20 patients less than 1 year of age, 56 patients between 1 and 4 years, 40 patients between 5 and 9 years, 26 patients between 10 and 14 years, and 26 patients diagnosed at 15 years of age or greater. The female/male ratio was 90/78 = 1.15 for 2007.

During 2007, patients came to CMH for cancer/tumor care from 36 different counties and 3 different states. 51 percent of the patients were from Missouri counties, 47 percent from Kansas counties and two percent from other counties. All are reported to the Missouri Cancer Registry who correlates data with the Kansas Cancer Registry for Kansas residents.

There were 19 cancer-related deaths during 2007, with five of these patients diagnosed during the same year. The leading mortality diagnosis was leukemia (6), followed by neuroblastoma (5) and brain tumors (4).

Diagnosis	Totals	Percentage
Central Nervous System	44	26%
Astrocytoma	13	
Glioma	8	
Ependymoma	3	
Medulloblastoma	2	
PNET	2	
Atypical Teratoid Rhabdoid	1	
N.G. Germ Cell	1	
Astroblastoma	1	
Ganglioglioma,anaplastic	1	
Benign/Borderline	12	
Leukemia	37	22%
ALL	29	
AML	6	
Other	2	
Lymphoma	12	7%
Non-Hodgkins	8	
Hodgkins	4	
Neuroblastoma	12	7%
Hepatoblastoma	6	4%
Osteosarcoma	5	3%
Wilms Tumor	4	2%
Rhabdomyosarcoma	2	1%
Ewings Sarcoma (EFT)	4	2%
Other	15	10%
Carcinomas	7	
Germ Cell Tumors	3	
Malig. Sertoli Cell Tumor	1	
Malig. Pleomorphic Sarcoma	1	
Melanoma	1	
Myelodysplastic Syndrome	1	
Retinoblastoma	1	
Misc. Reportable Conditions	27	16%
Totals	168	100%

ABBAS EMAMI, MD HONORING A CHILDREN'S MERCY HERO

In March of 1995, Dr. Abbas Emami began working at Children's Mercy Hospitals and Clinics. While many things led him on this journey, the most important was that he loved children.

His initial interest in pediatric cancer medicine began in his residency at a children's hospital in Detroit. A challenging fellowship provided him opportunities that added to his character. It was there that Dr. Emami says "He learned to manage his emotions." He also wanted to promote trust. Through his experiences, he learned how to connect with both his patients and their families, while dealing with his own emotions. According to Dr. Emami "these are important skills to have when working with cancer patients."

During his career, Dr. Emami witnessed many advancements take place. Instead of treating osteosarcoma with surgery alone which included amputation, research has shown a critical role for systemic chemotherapy in increasing the cure rate and facilitating limb salvage reconstruction surgery in most

patients. Chances for a cure for a patient diagnosed with osteosarcoma in the 1960's and 1970's were between 30 and 40 percent. Now, the survival rate is up to 70 or 80 percent.

Cancer treatment demands a dedicated team of individuals to manage the treatment plan and provide psychosocial support. With many years of following his patients closely, Dr. Emami reflected on his indebtedness to many outstanding colleagues and friends who provided support.

When asked how cancer impacted him personally, he replied, "People of all ages have hopes, plans and dreams. Cancer is the major event that puts a stop to that as people work through treatment." Dr. Emami saw first-hand how cancer affected patients and their families. Life seemed to be put on hold for everyone as they dealt with the emotional and physical trials of treatment.

Recalling an event that held a lasting impression, Dr. Emami spoke of a 16-year-old girl who had run into

complications and found herself in the ICU on a ventilator. Her future looked bleak. Dr. Emami talked to her parents about difficult decisions that they would have to consider, but advised them to wait. The next morning, his patient improved dramatically. One week later, she was off the respirator and went on to complete her chemotherapy treatments. After leaving the hospital, he later learned that his patient had gone on to graduate from high school and enrolled in college.

Retiring late in 2007, Dr. Emami reflected on the most rewarding part of his job. In addition to receiving graduation, wedding and family photos, many of his patients would stop him in the mall or at a restaurant, exclaiming "Do you remember me?" He listened as they would fill him in on their second chance at life. As they thanked him, he smiled looking past the once sick child to a healthy lively adult.





Dr. Emami is proud to have touched the lives of so many at Children's Mercy Hospital.



It often begins with some everyday bump or fall any child has. Jana was playing during recess and her best friend Malinda ran into her knocking Jana onto the ground. She had scraped her elbow and her arm stung. A few days later, her arm still hurt and she went to see her pediatrician who ordered an x-ray. The good news: there were no broken bones. The bad news: there was a mass of some sort in her shoulder.

For Jeremy, a long distance runner looking forward to graduating and going onto college the pain in his leg was nothing too unusual but it had not gone away after several weeks. The ibuprofen that usually worked was no longer keeping the pain under control and now it was starting to bother him even when he wasn't running. As a small bump began to appear on the side of his leg, he shared this information with his parents. An X-ray of his leg revealed a mass that appeared to be growing out from his bone.

Troy loves basketball. He especially loves playing basketball. He was always better than the other kids and now something even better was happening. He was growing...taller! Puberty had started and so had his growth spurt. The pain in his leg, mostly felt at night had been occurring for several weeks. He didn't remember falling or hitting anything. There was this time two years ago when he fell out of a golf cart but other than that, nothing came to mind. He had told his mom and she reassured him that this was "growing pains". After a few weeks, she took him to the family physician who also felt that this was most likely "growing pains". Still this pain didn't get any better and mom took him back to the doctor who ordered an x-ray that showed a lesion in his leg.

These three histories, no different than what occurs everyday, converge at this point. An abnormal x-ray leads to a work up that concludes with a diagnosis of Osteogenic Sarcoma, a rare but most common primary bone tumor in children and adolescence.

Perhaps best known in the United States as the cancer Ted Kennedy's son (Ted Kennedy Jr.) had when he was 12 years old (he just celebrated his 47th birthday) or in Canada where Terry Fox ran

across his country (with just one leg), the treatment options have expanded greatly from amputation.

Today treating Osteogenic Sarcoma involves a comprehensive multi disciplinary team of physicians, technicians, therapists, nurses, and social workers, dealing with medical, surgical, emotional, school, physical, cosmetic, and functional issues and that is true without even considering possible setbacks.

This year's annual report is focusing on Osteogenic Sarcoma in honor of one of our colleagues, Dr. Abbas Emani who dedicated many years of his career treating young people with this particular cancer.

Osteogenic Sarcoma (OS) is a malignant tumor that originates from the spindle cell that produce osteoid—the building block of bones. Malignant bone tumors are rare. They make up approximately six percent of all childhood cancers. The annual incidence in children under age twenty is 8.7 per million. OS make up 56 percent of bone tumors (compared to Ewing Sarcoma 34 percent) and is the most common. In the United States, approximately 400 children are diagnosed each year with Osteogenic Sarcoma. Here at Children’s Mercy Hospital we have seen 46 children with osteogenic sarcoma in the past 10 years.

While as with most cancers, we don’t fully understand what causes it, there are some things that are known:

- The peak incidence of bone cancer is 15 – 19 years, which is coincident with the adolescent growth spurt.
- It is very unusual to diagnose OS in someone under age 10.
- The patient is usually taller than average.
- The most common sites are at the metaphyses (the most rapidly growing part of bones) of the distal femur and proximal tibia (60 percent involve this knee area) and the proximal humerus.
- There is a genetic factor in that children who have had bilateral retinoblastoma (a cancer of the eye) are at risk of developing OS regardless of any exposure to radiation or other therapies.
- Chromosome abnormalities such as the Retinoblastoma recessive oncogene (Rb) and a second recessive oncogene (p53) have been found in up to 25 percent of patients.

- There is also a relationship in families with Li-Fraumeni syndrome in which affected family members have a variety of cancers including OS.
- The most common environmental factor related to developing OS is radiation.
- About three percent of OS develop in bones that have been previously irradiated for other reasons.
- Alkylating agents which are used to treat other cancers may lead to the development of OS.
- Other bone diseases have a slightly higher incidence of OS compared to the general public.

The average length of symptoms is about three months. The most common complaint is pain (90 percent) but only about half have any local swelling at the site. Again, about half will have a decreased range of motion and while not common, about 10 percent will present with a pathologic fracture.

It is imperative that the work-up is done with the collaboration of an orthopaedic surgeon who is familiar and comfortable with treating malignant bone tumors in children.

Whether the patient is initially referred to a pediatric oncologist or the orthopaedic surgeon is not particularly important as it is essential that both have the opportunity to evaluate, discuss, and coordinate the various diagnostic and therapeutic steps and options.

Initially, more specific radiologic scanning is required to get a more precise definition of the tumor. At the time of the initial referral the diagnosis of OS is still only a serious

concern. It will take a biopsy of the tumor to confirm. A MRI is usually obtained. Again, it is essential that the surgeon who does this biopsy also be the surgeon who addresses the more formal and definitive surgical treatment options. Once the biopsy is obtained, one must then wait for the pathologist to review and record their findings. Unlike other biopsies, these results often take a little longer due to the fact that these are bone tumors and it takes longer to process compared to other tissues.

Once the diagnosis is confirmed it is now necessary to stage the tumor. Most tumors are localized to the one area of concern, but OS can have “skip lesions”. These are areas of tumor involvement that are outside the reactive rim but within the same bone (regional intraosseous spread) or across a neighboring joint (transarticular metastases). Systemic metastases have a predilection for the lungs. Spread to other bones is the second most common site for spread and usually does not occur until after spread to the lungs has occurred.

The treatment for OS involves mostly two areas of cancer treatment: chemotherapy and surgery. Radiation that is useful in many other types of malignancies and is effective in the treatment of Ewing Sarcoma has a very limited role in the treatment of OS.

Historically, it was found that if you cut the tumor out you could successfully treat many patients with OS. This led to amputation being the standard surgical approach because of the recurrence of tumor at the primary site and subsequent metastases. In order to make a proper fitting prosthetic, several weeks were required. Amputation is





not an easy thing to handle at anytime in one's life but is even more so in a young adult. In order to decrease the time from losing ones natural limb and to begin the rehabilitation with the new prosthetic limb, chemotherapy was initiated.

Not too surprisingly, in many cases the chemotherapy had an affect on the tumor. Often there was notable decrease in the swelling and a decrease in the pain. These changes were seen on scans as well. Most importantly when the tumor was evaluated after receiving this chemotherapy often some or most of the tumor was found to be necrotic or dead when the pathologist examined it.

This tumor kill was looked at and it was found to be very prognostic. Those with a high amount of tumor kill were found to have a better prognosis than those with a low amount of tumor kill.

As our ability to fabricate prosthetic limbs has dramatically improved in speed of production, cosmetic and function current treatments still incorporate this upfront chemotherapy prior to definitive surgical intervention that now takes place approximately twelve weeks after commencing chemotherapy.

In addition, the surgical options have also progressed both in techniques and materials. While prosthetic limbs often provide a functioning limb, it still is not always as good as the limb one was born with. While prosthetic limbs can now be custom painted to resemble one's natural skin color, there is still no substitute for one's natural limb. Orthopaedic surgeons began looking at ways to salvage the limb in question.

Could improved surgical techniques eliminate the concern of a local recurrence at the original site of tumor involvement and could limb salvage techniques and materials allow one to retain one's natural limb? Over time, both the techniques and materials have advanced. What for many of us was fiction not so long ago in characters like Steve Austin the Six Million Dollar Man, are a reality for many of our patients.

For those patients with a localized tumor that has been removed and found to have a good tumor kill (>90 percent) long-term survival is seen in 60-70 percent of patients. This increases to over 80 percent in those with >98 percent tumor kill. Unfortunately, these numbers significantly decrease with lower levels of tumor kill. With metastases, as suspected, the success rate is even lower.

Whether an amputation or limb salvage is performed, in addition to having to deal with all of the normal side effects of chemotherapy the need for rehabilitation and physical therapy is immense. Staying positive, active, motivated, when stays in the hospital are frequent and often prolonged is not easy.

Staying up on school work with frequent and prolonged absences from school and friends is not easy. Having a limb that doesn't function or look the same can be overwhelming.

Usually most cancers are not associated with pain. Pain is usually associated with infection or inflammation. With OS, pain is common at presentation and unfortunately may be a very real concern during treatment when it is associated with the surgical procedure, or more troublesome, when it is associated with

tumor involvement of bones. Often many different options for pain control come into play at sometime in the course of treatment. These can include simple oral medications, patches, intravenous medications, creams, massages, and indwelling pain pumps.

Finally, there is the very real fear about resistant, recurrent, and relapsed disease. These words and the discussions that follow are never easy for the patients, families or those taking care of them. Once again, the path taken is different and unique for each patient. Whether it is investigational therapy or supportive care it continues to be a multi-disciplinary approach that we offer.

Currently, Children's Mercy Hospital, through our participation with Children's Oncology Group, treats patients through several different treatment protocols, some of which are international in collaboration. All of these treatments require the cooperation of many specialties.

I invite you to learn a bit more about the multitude of services that we provide to patients and families. Years ago, I was asked what made the area of pediatric oncology unique. The "Team Effort" is never more demonstrated than in the treatment of OS. Come meet the rest of the team.



Dr. Myers now enjoys helping Children's Mercy patients every day.

ANGELA MYERS, MD FROM PATIENT TO “PAGING DR. MYERS”

From patient to doctor, Dr. Angela Myers beat cancer and now fights to keep others well.

Angie was only 16 years old when she began favoring her leg due to chronic knee pain. Angela’s parents took her to an orthopaedic surgeon who suggested more tests. On the way home from the doctor’s office, her mother’s silence left her feeling uneasy.

Angie spent the next day having those tests to help confirm the suspected diagnosis. The following morning, her parents came into her room and sat down on the bed with crutches in hand, and told her “You have bone cancer.” Overwhelmed, Angela’s mind raced back to third grade when a boy in her class had died of leukemia.

After receiving her first round of chemotherapy at the Mayo Clinic, her doctor suggested that she see Dr. Maxine Hetherington a Hematology/Oncology physician at Children’s Mercy Hospitals and Clinics. Angie began her journey with chemotherapy and surgery to receive a knee replacement. Amazingly, Angie was able to keep up with her

studies and graduate with her high school class.

Determined to be a doctor herself, she was thrilled to be accepted into the University of Missouri-Kansas City’s medical program in 1995. Although she had a few surgeries for metastatic bone cancer during her college years, she was able to graduate and complete pediatric residency at Children’s Mercy. It seemed logical to Angie and her family that she would pursue further training in pediatric oncology; but in her second year of residency, she became interested in infectious diseases. She completed a fellowship in infectious diseases here at Children’s Mercy and has stayed on as faculty. Even though she is not the primary physician for cancer patients, she appreciates the opportunity to be able to take care of these patients, and get to know some of them as a consultant.

Incidentally, Children’s Mercy is the place where she met and married her husband of eight years, who is a private practice pediatrician. Angie now has 2 children, and has remained cancer-free for the last 11 years.



OSTEOSARCOMA – A MULTI-DISCIPLINARY EFFORT OF CARE

The staff of Children’s Mercy Hospital works hard to ensure that all patients receive timely, compassionate, and world class care. These are some of the areas that are important in the care of children with Osteosarcoma.

Radiology

The Department of Radiology is an important resource in the care of children with bone tumors. The mission of the members of the radiology team is to provide the best diagnostic tools available. Their work enables the oncology and surgical teams to successfully treat these children.

A child with osteosarcoma is usually diagnosed after a plain X-ray of the bone. The tumor is then further characterized with an MRI which helps to determine the extent of the tumor and show if there are any additional areas of tumor in that bone. After osteosarcoma is diagnosed, a bone scan is used to look at the other bones in the body to evaluate for metastatic disease. A small amount of radiotracer is injected into the veins and localizes in the very active tumor sites. A chest CT is used to examine the chest for metastatic disease. Bone scans and CT are also used to monitor the tumor during therapy.

In the near future, the hospital will be opening a new state-of-the-art MRI center. This will house new 3 tesla and 1.5 tesla magnets. The scanners will include all available advanced imaging techniques and software that will improve imaging of bone, cartilage, and joints.

Surgery

The Orthopaedic Surgery section works hand in hand with oncology colleagues to diagnose, treat, and follow patients diagnosed with osteogenic sarcoma.

Most children diagnosed with osteogenic sarcoma are active, healthy adolescents. When a diagnosis is suspected, the sections of Hematology/Oncology and Orthopaedics work efficiently to arrange for a biopsy, obtain an accurate diagnosis, and initiate medical management. Effective communication between the sections ensures prompt tumor resection and reconstruction at the completion of adjuvant chemotherapy, as well as the safe resumption of adjuvant chemotherapy following surgical reconstruction.

Upon completion of reconstruction and adjuvant chemotherapy, osteosarcoma patients continue to be “co-followed” by physicians in each section to maximize function, and identify recurrent disease.

Pathology

Osteosarcoma is an aggressive cancer and it is important to make the accurate diagnosis for the sake of the patient’s management. Pathologists are involved in



making the initial diagnosis as well as gauging the treatment response. When a tumor is suspected clinically or from radiologic studies, the orthopaedic surgeon performs a biopsy of the tumor through a needle core. The biopsy specimen is sent to pathology for histological processing. The pathologist reviews the patient's material prepared on a glass slide through a microscope and determines if there is an osteosarcoma or not. The pathologist then communicates the results to the surgeon or to the treating oncologist.

Patients with osteosarcoma receive a treatment regimen that includes chemotherapy and surgical resection of the bone containing the tumor. At the time of the surgery, the pathologist examines the limb bone and confirms the biopsy diagnosis. The microscopic appearance of the tumor varies according to biology of the tumor and whether the tumor cells are predominantly forming bone (i.e. osteoblastic), cartilage (i.e. chondroblastic), or fibrous tissue (i.e. fibroblastic). Other rare histologic variants of osteosarcoma that may affect the patient's prognosis can also be identified. The pathologist will also determine the degree of tumor extension in the bone. Microscopic examination of the tumor will also reveal whether the tumor was successfully treated with chemotherapy or not. The presence of extensive tumor necrosis (non-viable tumor cells) after chemotherapy is a good sign that the patient's treatment was successful.

Cytogenetics

Osteosarcoma is the most common malignant bone tumor in childhood and adolescence. The cause of osteosarcoma is unknown. However, it is known that mutations of specific genes can predispose patients to tumors, including osteosarcoma. While the majority of osteosarcomas are sporadic and not inherited, two autosomal dominantly inherited disorders predispose to osteosarcoma, familial retinoblastoma and Li Fraumeni syndrome. A germline mutation of the RB1 gene at chromosome location 13q14 predisposes patients to retinoblastoma and osteosarcoma. A germline mutation of the TP53 gene at chromosome location 17p13 causes Li Fraumeni syndrome which predisposes patients to many tumors including osteosarcoma. Studying these genes helps to shed light on the pathogenesis of the tumors with the goal of discovering effective treatment options. Even in sporadic osteosarcomas, changes in the RB1 gene and TP53 gene are the most common recurrent abnormalities.

The cytogenetic laboratory processes living tumor tissue removed from the patient at the time of surgery in order to grow the tumor cells in culture. The cells are captured as they divide so the chromosomes can be examined. The genes reside on the chromosomes and by examining the chromosomes, the abnormal genes can be studied. Most often the chromosomes of an osteosarcoma are very mixed up with many abnormalities. This makes it difficult to determine which of the many genetic changes are responsible for the initiation of the tumor and which are responsible for the aggressive behavior that is characteristic of most osteosarcomas. Studying



osteosarcoma tumor cells gives an opportunity to identify the genes responsible for the development and behavior of these tumors. Continued study will undoubtedly lead to the identification of new treatment agents and possibly gene therapy

Social Work

Social workers are an integral part of the medical team as they demonstrate the importance we place on comprehensive, family centered care. Our social workers hold a Master's degree and each has expertise serving the needs of our children and their families. Each social worker works closely with a primary oncologist and advanced practice nurse as the patient and family's primary team.

At diagnosis, the primary team sits down with the patient's family and patient (when age appropriate). As a group, they discuss the diagnosis, treatment plan and begin to answer questions. The social worker provides supportive counseling and assists the family in organizing their own resources. They also begin identifying additional resources that could aid the family in the journey ahead.

The social worker completes a psychosocial assessment on each patient. This forms the basis for the subsequent relationship with the child and family. The assessment aids the family and social worker in determining the resources and assistance needed as well as making preparations to begin treatment.

Social workers provide counseling as patient, parents, and siblings begin to heal both physically and emotionally from the diagnosis. In addition, when appropriate, they make referrals to other psychosocial team members such as child life, chaplaincy, and child psychology.

For school-aged children, social workers assist parents and the school to ensure that the child's educational needs are being met. Social workers may attend IEP (Individual Education Plan) meetings with parents to assist with developing a plan that the patient and accomplish both during and after treatment. Social workers also provide supportive counseling to the patient and fellow students as the patient returns to school.

Social workers refer patients and their families to outside agencies for support, education, and financial assistance. Families get assistance from such groups as The American Cancer Society, National Children's Cancer Society, Turning Point and Solace House. Referrals for lodging and transportation assistance are also given with appropriate.

To provide support specifically to parents, the Hematology/Oncology Division at Children's Mercy Hospital offers the Parent to Parent program, coordinated by a social worker. Experienced parents of children with cancer volunteer and are paired with a family of a child newly diagnosed. The volunteer shares their expertise and



offers support. The program also offers bereavement services to families. This service is also staffed by parent volunteers who have lost a child to cancer.

In addition to the Parent to Parent support, social workers also offer counseling and support to families who are grieving the loss of a child. Families are guided through the funeral process and continue to receive support once outside the hospital. Both internal bereavement groups and outside resources are offered in order to best meet the needs of each individual family.

Social workers realize that treatment is not merely a series of medications, clinic appointments, and inpatient hospitalizations. It is a process where the most important outcome is that the patient and family are able to define their “new normal” after treatment ends.

For many of our patients with osteosarcoma, their lives will never be the same. They will require adjustment, assistance, equipment and opportunities to discover what their new normal looks like. As part of an ongoing partnership with each patient and family, social workers consider it their privilege to accompany patients and families on their journey and bear witness to their determination, courage and resilience.

Nutrition

Osteosarcoma presents unique nutrition challenges compared with other pediatric malignancies. This is due to the severe irritation of the gastrointestinal (GI) tract associated with chemotherapy. Chemotherapy for Osteosarcoma results in severe nausea and vomiting, often difficult to control with medications, as well as inflammation and sloughing of the tissues lining the GI tract. This irritation leads to painful sores in the mouth and throat, and often the entire length of the GI tract (stomach, intestines, colon, and anus). The combination of side effects makes eating extremely difficult, and medication or hospitalization for pain control are necessary. The majority of children and teens with Osteosarcoma also require medications to increase their appetite, in addition to a diet high in calories, protein and antioxidant vitamins from fruits and vegetables to help repair the tissue damage associated with chemotherapy. Commonly kids and teens struggle to maintain their weight and muscle strength, and eventually require supplemental medical nutrition from a temporary tube feeding or intravenous infusion of nutrients. A feeding tube inserted into the stomach (into the nose and down the throat) can help repair damaged tissue, but may not be placed in the presence of significant inflammation or sores. For those children and teens, specialized intravenous solutions of calories, protein, fat, vitamins and minerals are often required.

The medical team relies on the patient and family’s assessment of their daily level of functioning and tolerance to therapy to guide the nutrition care plan. Laboratory tests of the blood and measurements of the body are routinely monitored by a registered dietitian specializing in nutrition support along with the medical team

for continual nutrition assessment during therapy. Growth parameters are also monitored long-term by the medical team to screen for late-effects of chemotherapy on development

Behavioral Medicine

The Section of Developmental and Behavioral Sciences offers comprehensive psychological services to children and families affected by osteosarcoma, as well as other forms of cancer. Services are tailored to the needs of each child and family. Staff members provide a listening ear, as well as facilitate coping with the stressors that accompany the diagnosis and treatment. Counseling can be done both on the inpatient unit, and in the Developmental and Behavioral Sciences Clinic. Individual or family services can be offered to the patient, siblings and parents. This is part of the commitment to provide family centered care throughout CMH. In addition to a designated psychologist who works closely with the Division of Hematology/Oncology, consultative services with a psychiatrist and family therapist are also available.

Children tend to be quite resilient, and their ability to cope with life's most difficult challenges can amaze the adults around them. However, when facing cancer diagnosis and treatment, children benefit from having additional support to help them adjust to what is happening to them. Children with osteosarcoma may have to face surgeries that alter the appearance and/or functioning of their bodies, including amputation. Psychological support can help address the body image issues that accompany such changes, and that sometimes can lead to symptoms of depression, anxiety, etc. These children also face challenges in reintegrating back into their school settings, and the psychologist works with the interdisciplinary team to help the children make this transition in a healthy way.

Rehabilitation Medicine

Kids with osteosarcoma may have a hard time functioning as they should for their age, depending upon the site of the tumor and the course of treatment. Some children need surgeries to keep their limb as functional as it can be, and others need amputation either as treatment, or in order to allow them to be more functional, along with prosthetic (artificial limb) fitting. The goal of Rehabilitation involvement is to help return the child to an active, positive and functional lifestyle, and this team joins the oncology team and other health care providers whenever their expertise is needed.

The Rehabilitation Team includes a diverse group of caregivers with pediatric rehabilitation physicians, physical therapists, occupational therapists, speech therapists, recreational therapists, rehabilitation nurses, a neuropsychologist, social workers, dieticians, orthotists and prosthetists, respiratory therapists, music therapist, and child life specialists. This team of experts focuses on function for children who lose skills due to illness or injury. For each child this team works to



identify what skills and activities are more challenging, and design a plan to improve those skills. This takes a combination of hands-on therapy, assistive devices such as a brace or walker, restorative devices such as a brace or prosthesis for an arm or leg, and training in the skills needed that the child and his or her family can work on. This may take place in the hospital or after discharge, on an out-patient basis.

The Rehabilitation Team may get involved with children who have osteosarcoma at a number of different points in their treatment. With medical treatment and longer hospital stays, a child may have weakness that gets better with therapy, and play-like activities that help with strengthening. After a limb salvage surgery, it is important to start moving the knee or the affected body part almost immediately. For example, a continuous passive motion (CPM) machine may be used to improve motion after limb-salvage surgery for tumors around the knee by constantly bending and straightening the leg. It works by continuously bending and straightening the knee. Physical therapy helps the child learn to walk again, first with a walker or crutches and then later, without any assistive devices. This process may take six to 12 months following surgery.



Amputation or removal of all or part of a limb may be necessary as part of treatment. In some children with osteosarcoma of the leg, amputation provides a better chance than limb-salvage of returning a child to an active lifestyle, including play, running and sports. Our Limb Deficiency Team includes a rehabilitation doctor, an orthopaedic surgeon, physical and occupational therapy, rehabilitation nurses, and prosthetists. (A prosthetist is a professional who makes artificial limbs.) The team works in a clinic which is part of an international group of professionals dedicated to the care of children with amputations, known as ACPOC, or the Association of Children's Prosthetic Orthotic Clinics, and through that association we provide children with amputation, their families, their doctors and other care providers with information about amputation and prosthetic intervention. This team may get involved even before a planned amputation to help decide what level of limb removal will allow the best options for prosthetic fitting and overall function. They also can link children to other children with similar issues, which helps with coping and hope.

Depending on the level of the amputation and other medical factors, a new artificial leg can be fitted right away after surgery. But often a period of healing and limb preparation is needed before fitting. It usually takes at least three to six months until a young person learns to use a prosthetic (artificial) leg or arm well, and this is just the beginning of long-term physical and psychological rehabilitation. The team then has the child return twice a year to make sure the prosthesis fits, and best suits the needs of the child.

Rehabilitation efforts are tailored to the unique needs of each child with Osteosarcoma.

TYLER BARTLETT

TYLER'S TRIUMPH

The following is a letter from Tyler's mother:

Subject: Tyler and basketball

11-28-07 Last night was the best night in a long time and the worst night in a long time. We got a call Monday night that the cubs would need to be at the McEwen school on Tuesday night to help with the Hoops for Hearts. It is a fundraising for something and the cubs were to be there at 6 pm. We showed up and the kids had taken up pledges to make so much for each basket they made. The third graders went and then fourth graders were going. Tyler said he wanted to go.

Now picture this. They started two kids in center of basketball court. Each kid had to run to one end and make a basket from squares set on floor. They had 30 seconds to make as many baskets as they could. Then they got to do it again. They largest amount of baskets was the number they could use to get pledges.

Tyler kept asking me if he could play. I said no that they were doing too much too fast. He kept begging and I finally gave in. I was scared that he would fall while running, not make any baskets and be depressed and had many other thoughts running thru my mind. He walked over to the side of the gym that other kids were sitting at and sat down. The coach went over and talked to him.

Then he called Tyler's name and another girl's name and she came out. She also had leukemia a few years ago. The coach let Tyler go closer to basket and then set off the timer. He made 2 baskets during the first set. Then he made a basket during the second set.

There was an older girl who came in and finally helped him run after the ball. All the kids were yelling "Go Tyler Go" and the parents were yelling. He finished and went to sit down. Then he came over and sat on my lap and said he was ready to go home. We left he was so excited that he had been able to play. He likes basketball so much. So like I said it was the best of times and the worst of times. He was happy and I was worried. But I was so PROUD of him. He is really trying to be as normal (as normal as possible) as he can be. I really am thankful for friends. I don't know what I would do without them. My love goes out to all of you.

God bless.



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