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2009 Cancer Care Annual Report

Children's Mercy Hospital

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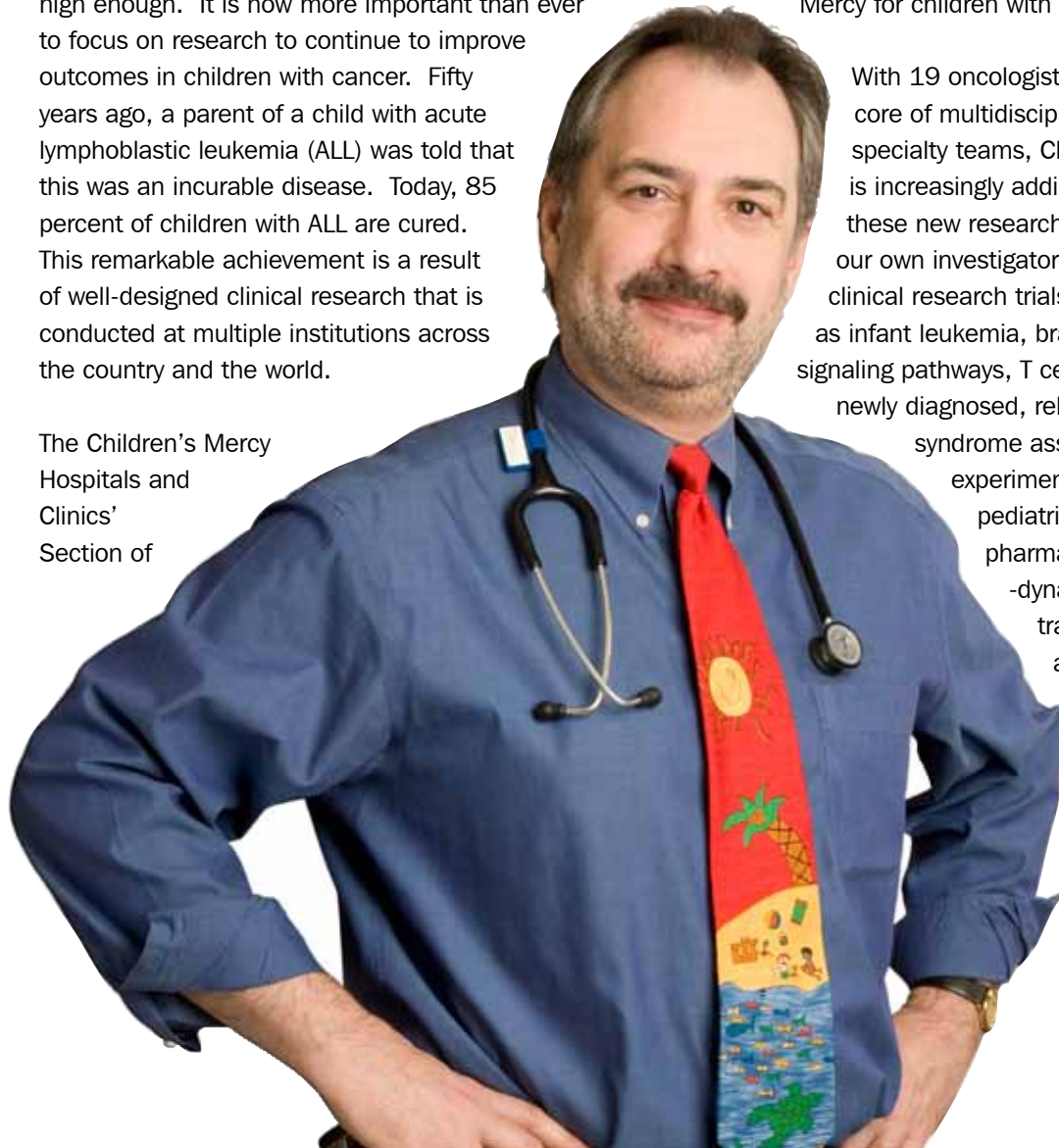
2009
Cancer Care
Annual Report

Focus on ALL



Over 75 percent of children diagnosed with cancer in 2010 will be cured and survive into adulthood. That is encouraging news for a family struggling with the news that their child is one of 12,000 children diagnosed with cancer every year in the United States. Cure rates today are dramatically higher than they were just 20 years ago. But they are not high enough. It is now more important than ever to focus on research to continue to improve outcomes in children with cancer. Fifty years ago, a parent of a child with acute lymphoblastic leukemia (ALL) was told that this was an incurable disease. Today, 85 percent of children with ALL are cured. This remarkable achievement is a result of well-designed clinical research that is conducted at multiple institutions across the country and the world.

The Children's Mercy
Hospitals and
Clinics'
Section of



Oncology is proud to be an important part of these accomplishments. We are the sole regional site for, and indeed one of the larger Childhood Cancer Centers in the Children's Oncology Group, the National Cancer Institute's cooperative organization for childhood cancer research. Through this network, nearly 100 clinical trials are available at Children's Mercy for children with cancer.

With 19 oncologists forming the core of multidisciplinary and multi-specialty teams, Children's Mercy is increasingly adding leadership to these new research efforts through our own investigator-led laboratory and clinical research trials in areas such as infant leukemia, brain tumor cancer signaling pathways, T cell & relapsed ALL, newly diagnosed, relapsed, and Down syndrome associated AML, experimental drug discovery, pediatric drug formulation, pharmaco-kinetics and -dynamics, bone marrow transplantation, adoptive immune

and gene therapies, infectious complications, supportive care, and survivor quality of life.

This year's annual report for the Children's Mercy Cancer Center focuses on the most common type of cancer seen in children, acute lymphoblastic leukemia, commonly referred to as ALL. ALL can affect children of all ages, but is most commonly diagnosed in younger children. As with all childhood cancer, the treatment of ALL requires a multidisciplinary team with the knowledge and experience necessary to handle all aspects of care.

At Children's Mercy Hospitals and Clinics, we are fortunate to have many dedicated people that are focused on providing the best possible care for children with cancer. This year's annual report highlights all of the people that are involved, and emphasizes the role each plays in treating children of all ages with ALL. We hope this report will serve as an introduction to our team and the resources we can provide to help children and families that are diagnosed with cancer. 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
Professor of Pediatrics, UMKC School of Medicine

Dear Friends,

All of us at Children's Mercy were so pleased and proud when our oncology program again was awarded the Outstanding Achievement Award by the American College of Surgeons Commission on Cancer in 2009. This is the second time in a row that we have received this prestigious three-year designation, which is awarded to fewer than 20 percent of all adult or pediatric cancer programs nationwide.

As you read through this annual report, you'll understand why the Children's Mercy oncology program continues to receive these types of honors. Our physicians and nurses, our support staff and our researchers are all exceptionally committed to providing the finest state-of-the-art care for children with cancer and support services for their families, as well as cutting-edge research to help find new treatments and cures for this disease.

We have one of the largest pediatric hematology and oncology faculties anywhere in the nation, with more than nine times as many pediatric oncologists as any other program in our region and survival rates equal to or better than the national averages. A number of our physicians serve in national leadership roles in organizations and research programs working with childhood cancers.

This Cancer Care Annual Report focuses on Acute Lymphoblastic Leukemia and the strides we have made in providing care for children with this diagnosis. It also provides you with a more complete overview of the wide array of services, technology, staff and quality that we are able to offer these children and families at a truly challenging time in their lives.

I hope you will take a few minutes to read this annual report and learn more about how Children's Mercy is caring for children with ALL and working as a national leader in treating and researching cures for all types of childhood cancers. I offer my sincere congratulations and my deepest gratitude to everyone in our Division of Hematology/Oncology for the extraordinary work they do.

Sincerely,

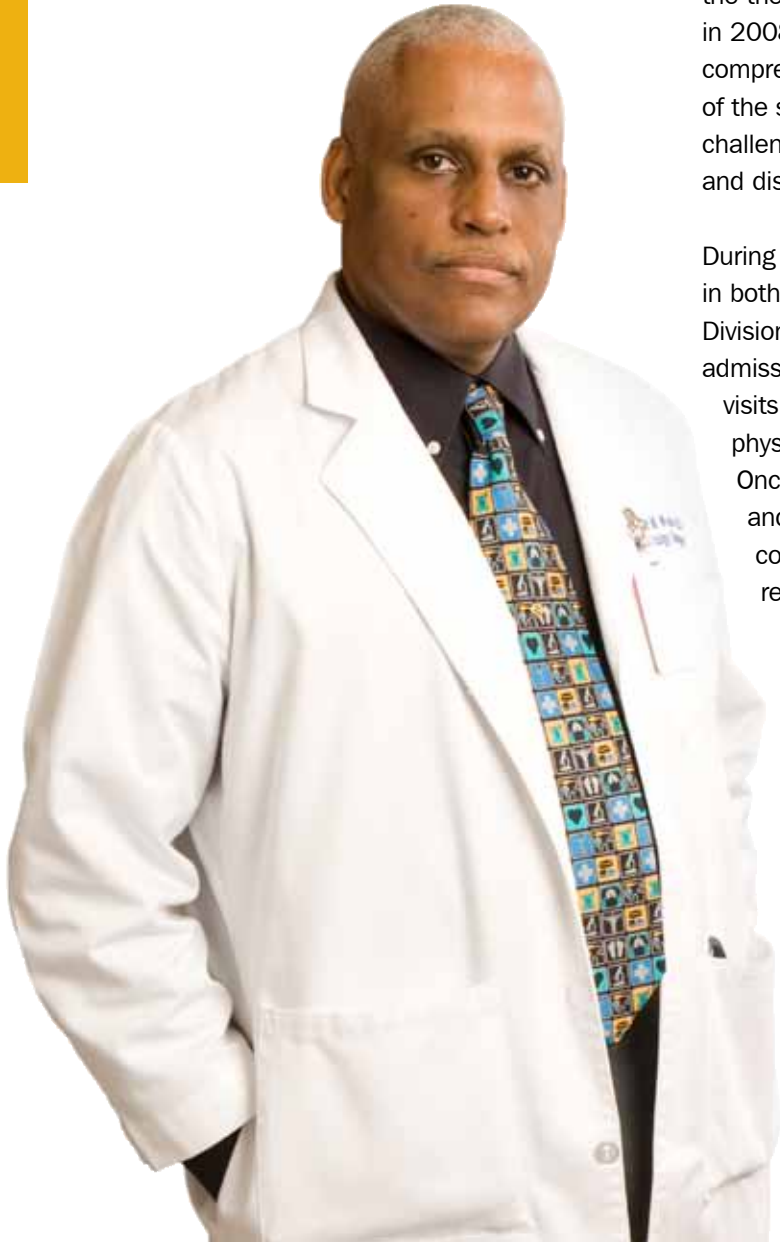


Randall L. O'Donnell, PhD

President and Chief Executive Officer



THE DIVISION OF HEMATOLOGY-ONCOLOGY



Growth in programs, services and personnel was the theme for the Division of Hematology/Oncology in 2008 and 2009. Dedication to excellence in comprehensive care continues to be the main focus of the staff working with patients who are experiencing challenges with oncologic and hematologic diseases and disorders.

During 2008-2009, 16 physicians provided care in both the inpatient and outpatient settings. The Division continues to have over 1,000 inpatient admissions and approximately 10,000 outpatient visits annually. Research efforts are led by the physicians with their involvement in the Children's Oncology Group, Centers for Disease Control and National Institutes of Health. In addition, collaboration on research occurs locally, regionally and nationally.

The team approach to comprehensive care is the model that is practiced within the division. Eighteen Advanced Practice Nurses work alongside a physician to coordinate

the many details of patient care. From scheduling appointments with other specialists, ensuring that chemotherapy protocols are followed, dealing with insurance issues and providing hands-on patient care and education about diseases and treatment; the Advanced Practice Nurses are essential members of the team.

Nurses in both the inpatient and outpatient setting provide safe and effective family-centered care. All staff nurses receive extensive training in pediatric hematology-oncology disorders, treatment modalities and potential effects of treatment. Currently, more than 40 staff nurses are Certified Pediatric Oncology Nurses.

Because our patients require complex care that involves many disciplines, the division operates closely with other specialties such as Surgery, Endocrinology, Neurology, Pathology and Radiology. Nutritionists and pharmacists monitor each patient closely throughout their course of treatment. Many additional team members work behind the scenes to ensure that

Gerald Woods, MD

Division Chief, Hematology/Oncology

Section Chief, Hematology

Director, Sickle Cell Program

Professor of Pediatrics, UMKC School of Medicine

the necessary support is in place for the direct care providers, patients and their families. Support services are essential to the care of these patients who have many complex needs. Social workers assist families with counseling, serve as an advocate for the patient and family and provide expertise with resource utilization. Psychology, Family Therapy, Child Life Therapists and Music Therapists provide opportunities for ongoing developmental growth and psychosocial support for children that are experiencing serious illness. Chaplaincy is available 24/7 to support the spiritual needs of our patients, families and the staff. Volunteers offer a listening ear and many other comfort measures such as meals in the parent room and special events during various times of the year.

Research and data managers who coordinate many clinical trials, administrative assistants who maintain communication both within the division and outside the hospital, as well as schedulers who organize appointments all serve as crucial members of the team.

The Division added two new programs in 2009: the Survive & Thrive Late Effects Clinic for childhood cancer survivors and the Experimental Therapeutics in Pediatric Cancer Program that is working to develop and test new treatments for children with cancer.

We hope this report provides insight into the recent efforts of the Division of Hematology/Oncology. We especially hope that it demonstrates that the Division offers many of the “million reasons” why Children’s Mercy Hospitals and Clinics is one of the best places to seek care for kids.



Madison's a-Mazing Marrow Match



Madison with brother Steven

A child diagnosed with leukemia is a jolt to any family, but when the disease is treated and then returns, the impact can even be a more powerful one. Madison's family found out that she had ALL when she was 5. Her mother remembers that it felt like it was the end of the world. At the time, she knew nothing of leukemia and didn't even know "if it was fixable." The family credits the doctors and nurses who cared for Madison with helping the family understand what their treatment would be and what they could expect in the months ahead.

After Madison completed her treatment, she went about a year without any problems and was doing great. That was when the one thing Madison's mom didn't really expect to happen, happened. Madison's disease was back.

Again, Madison's mom, Samantha, remembers how Dr. Karen Lewing talked to her about the options for treating her 9-year-old daughter. One of the options was a hematopoietic stem cell transplant. Because of the timing of Madison's relapse, they would only be able to do a transplant if Madison had a sibling who matched her type of bone marrow. With only a one in four chance of being a match, the family anxiously awaited the results of her brother Steven's testing. When the news came that he matched, the family knew that their next goal was to keep Madison healthy until it was time to get her transplant. She remembers, "That was a struggle, but we made it."

For transplant, Madison had to switch treatment teams from her oncologist, Dr. Lewing, to a doctor that specializes in transplant. Samantha remembers

not being sure about that, but recalls her relief that Dr. Doug Myers could not have been better. "He explained everything in a way that we could understand and both the kids really liked him."

Madison entered the special bone marrow transplant unit at Children's Mercy several days before her transplant. At that time she had received chemotherapy that had made her disease go into remission again. Once in the hospital, she would receive very strong chemotherapy to get her body ready for the new bone marrow. Without the strong chemotherapy, her body would probably attack the new bone marrow and not let it grow. The day of the transplant, Steven went into surgery with a big smile on his face. His mom remembers that he "wasn't one bit scared and this was his first time having done anything like that." Madison received her brother's bone marrow in a procedure very much like a blood transfusion. After getting the cells, she had to stay in the hospital until the new bone marrow grew enough to make a brand new immune system that would now protect her.

Madison is now more than two years past her transplant and she is a happy, healthy girl. She likes to draw and sing. She has a great sense of humor and likes to tell jokes. Samantha is unable to determine how Madison has or has not changed because of what she has gone through. "It all started when she was so young. What I do know is my family is very thankful to Children's Mercy Hospital. I don't think we've ever been treated so well!"

CANCER REGISTRY

The Cancer Registry at Children's Mercy Hospital plays a vital role in the fight against cancer. Data is collected and maintained on all patients with malignancies and certain benign tumors. This confidential information will assist health care providers and Researchers concerning treatment methods, prevention, recurrence and survival.

The Cancer Registry operates under the guidance of the Cancer Care Committee and follows the data collection requirements established by the Commission on Cancer of the American College of Surgeons and the State of Missouri. In October 2009 the Commission on Cancer surveyed the Children's Mercy Hospital Cancer Program for the continued performance of outstanding high quality patient care. We were pleased to again receive the Approval Award with a three-year commendation, in addition to the prestigious Outstanding Achievement

Award from the Commission on Cancer of the American College of Surgeons. This award was only achieved by 18 percent of the 432 cancer programs surveyed during 2009.

FAST FACTS - 2008 and 2009

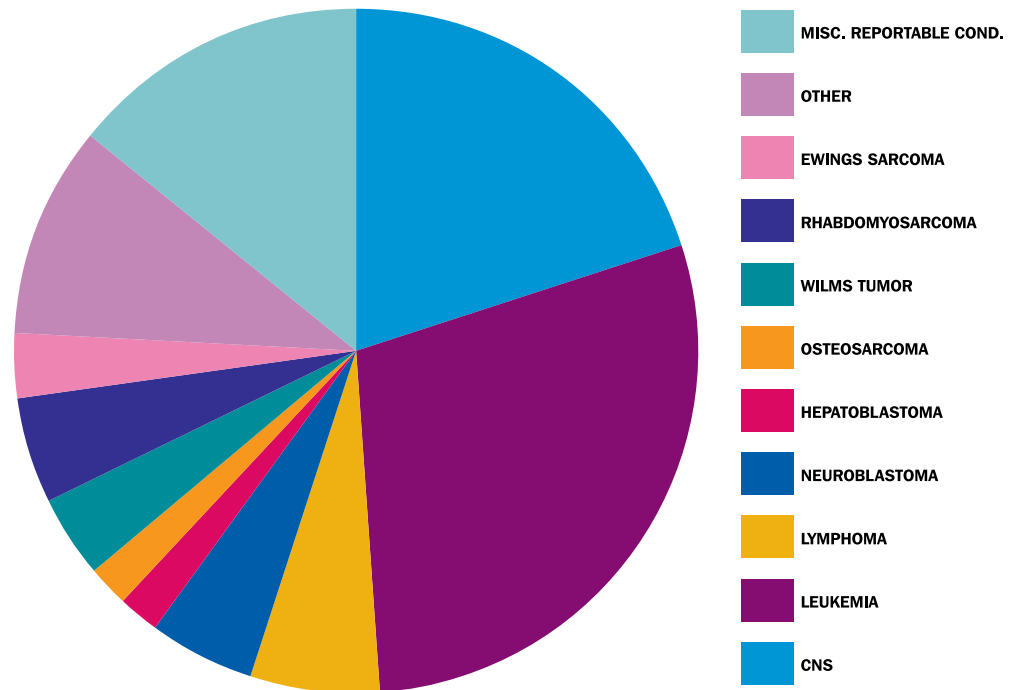
- The median age of patients diagnosed during 2008 and 2009 was 10.5 years
- The male/female ratio was 1:1.1
- Race distribution included 255 Caucasians and 33 African Americans, with 28 patients designated as other race. Within the 255 Caucasian patients, 27 patients were of the Spanish/Hispanic ethnicity.
- Geographically patients came from 67 different counties. Fifty two percent of the patients were from Missouri counties and 45 percent were from Kansas counties. Two additional states with corresponding counties made up three percent of the total patients.

During 2008 and 2009 the Cancer Registry added 316 patients to the database. Of these patients there were 259 malignancies, 14 benign or borderline diseases of the central nervous system and 43 reportable conditions. The reportable conditions are among a group of disease processes that the Cancer Care Committee has agreed should be collected because they are of local interest. Leukemia is the most frequent diagnosis during 2008 and 2009, followed by tumors of the Central Nervous System. The diseases of Lymphoma, Neuroblastoma, and Rhabdomyosarcoma made up the remainder of the top

five diagnoses during this two-year timeframe.

**CANCER REGISTRY
FREQUENCY OF DIAGNOSIS BY YEAR**

DIAGNOSIS BY YEAR	2008	2009
Central Nervous System	35	28
Astrocytoma	6	9
Glioma	11	4
Ependymoma	4	2
Medulloblastoma	3	5
N.G. Germ Cell	2	0
Pineoblastoma	0	1
Desmoplastic Small Round Cell	0	1
Choroid Plexus Carcinoma	0	1
Benign / Borderline CNS	9	5
Leukemia	44	48
ALL	33	35
AML	9	11
Other	2	2
Lymphoma	7	13
Non-Hodgkins	4	6
Hodgkins	3	7
Neuroblastoma	10	6
Hepatoblastoma	2	3
Osteosarcoma	3	4
Wilms Tumor	7	7
Rhabdomyosarcoma	4	11
Ewings Sarcoma (EFT)	5	5
Other	14	17
Carcinoma	5	4
Sarcomas	2	4
Retinoblastoma	2	3
Germ Cell Tumors	3	1
Misc. Malignant Conditions	2	5
Misc. Reportable Conditions	20	23
TOTAL BY PER YEAR	151	165



Quinton knew that the summer of 2010 was going to be one of new experiences. He had just graduated from Leavenworth High School and was ready for a new chapter of his life to begin. Quinton spent his senior year in the prestigious position of Battalion Chaplain of his school's ROTC program. He was awarded this position after having gone through a rigorous application process involving his peers and instructors. In the end, Quinton was chosen for the role because his commanders knew that he would be a moral and spiritual leader that the other cadets would turn to for advice and support. Quinton excelled in the position. Because of his exemplary high school performance, he was now preparing for a four-year stay at his dream school, Kansas State, his tuition being paid by a full Army ROTC scholarship and acceptance into the Smith Scholarship House.

Soon after graduation, Quinton had gone with friends on a service trip to Colorado Springs where they worked to help military families who had loved ones out of the country. During the times when he and his buddies were chopping and hauling wood and joking about their extraordinary strength, Quinton was also feeling run-down and not himself. He was used to always being full of energy – he ran cross-country track, loved other sports and was always on the go.

Back home in Leavenworth, Quinton's parents took him to see a doctor who thought he had mononucleosis. When he didn't get better, more tests were done and he was sent to Children's Mercy Hospital where he was diagnosed with Acute Lymphoblastic Leukemia. Quinton spent close to two weeks in a hospital bed during June. During that time his body hurt, his head ached, and he was so tired

that he had to be coaxed to get out of bed and to use his muscles. He got chemotherapy; he got blood transfusions; and he thought a lot about how all of this would impact his plans for the future.

Now, several months after hearing the news, Quinton has accepted his diagnosis. His doctor, nurse practitioner and social worker have spent time with Quinton and his family to make sure they understand what treatment will be like and what they can expect over the next three and a half years that his treatment will last. Because most of the treatment can be given as an outpatient, Quinton now spends much of his time in the Hematology/Oncology Clinic. Not going into the hospital is great for him, but the clinic days are long when they involve having blood work done, waiting for the results, and getting chemotherapy that can take a long time to infuse into the catheter that he has implanted in his chest.

When asked what it is like to come to Children's Mercy as one of the older patients, Quinton says it makes him glad. He says that the people at Children's

Mercy have a gentle touch that he appreciates. He also says that he has learned more about himself during this and sees what he can adjust to and get through. Quinton says that he has many people "praying their eyeballs out for him," and he thinks that one day he will be able to use his own experience to help other kids with cancer. When asked where he sees himself in 10 years, he thinks that maybe he will be an officer in the army, but he expects that he will be in a role where he can teach and he can listen. He says that he knows he "can't make everything all better, but I can be a person who will listen and hear."



Quinton's Quest for a Cure

Quinton will miss going to Kansas State in the fall. He may lose his scholarship due to health requirements of his reward. Talking to Quinton, however, you would not detect anger, resentment or any other negative emotion that would almost seem natural. What you see is a strong young man who lives every day with the faith that he can make it a good one.



ACUTE LYMPHOBLASTIC LEUKEMIA (ALL)

Acute lymphoblastic leukemia (ALL) is the most commonly diagnosed cancer in children. Every year, there are about 2,400 children diagnosed with ALL in the United States. Approximately one quarter of the children seen at Children's Mercy Hospital and Clinics for cancer treatment have ALL.

Leukemia is a cancer of white blood cells. ALL begins within the bone marrow in a specific type of white blood cell called a lymphocyte. Normal lymphocytes within the body help to fight infections and we cannot survive without them. In ALL, the growth of certain lymphocytes becomes abnormal, and they begin to grow and divide very quickly. Because these cells are growing so rapidly, the production of other cells that normally grow in the bone marrow such as red blood cells, platelets, and healthy white blood cells is diminished. This results in many of the symptoms that are seen at diagnosis. By the time ALL is diagnosed, there are over a trillion leukemia cells in the circulation.

ALL can present with a variety of symptoms and is often initially misdiagnosed. Symptoms seen in patients with ALL include fever, fatigue, anemia, bruising, bleeding, enlarged lymph nodes, an enlarged liver or spleen, or bone pain. Diagnosis is often made after a complete blood count (CBC) is ordered and abnormal white blood cells called lymphoblasts are seen.

Little is known about the causes of ALL in children and the vast majority of cases occur in healthy children. ALL is seen more commonly in males, and the most likely age at diagnosis is between 2 and 5 years. ALL is more common in white children when compared to black children. It is seen more frequently in children of families with higher socioeconomic status, for reasons that are not well understood. There are certain genetic syndromes that are predisposed to developing ALL, the most common of which is Down syndrome. Other syndromes that can predispose to the development of ALL include: Bloom syndrome, neurofibromatosis, Klinefelter syndrome, and ataxia-telangiectasia.

The improvement in survival for children with ALL over the last 40 years is one of the true success stories of modern medicine. As recently as the 1960's, the diagnosis of leukemia for a child was considered terminal. With current therapies, a child diagnosed with ALL today will have above an 80 percent chance of cure.

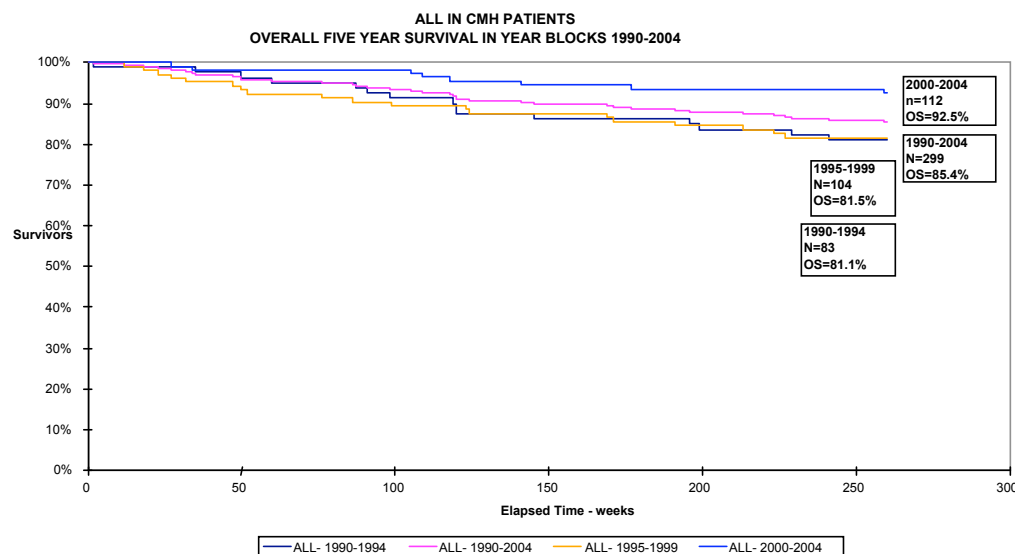
ALL is treated with chemotherapy, and treatment can last as long as three and a half years. One of the most remarkable aspects of the improvements in the outcome of patients with ALL is that they have occurred without the benefit of new medicines, but with improved knowledge and understanding of medicines that have been around for a long period of time.

The earliest attempts at the treatment of ALL included one chemotherapy drug. This treatment would result in some remissions, but all patients would eventually relapse. This led to the use of multiple chemotherapy drugs in combination, and improvements in the number of patients that respond and even long term cures were seen. Now, almost all patients are able to achieve remission after one month of induction treatment that includes three to four chemotherapy medications.

in the CNS, but the long-term effects of this treatment were substantial. For young children that are still developing, radiation directed at the CNS could cause learning disabilities, growth defects, and abnormalities of hormone production. Over the past 20 years, preventive treatment of CNS relapses has shifted from radiation to chemotherapy administered directly into the cerebrospinal fluid. Using chemotherapy has proven to be just as effective at preventing CNS relapses with a lower incidence of severe side effects,

half years for girls and two and a half years for boys.

Other improvements in the treatment of ALL in children have come from a better understanding of the disease and the knowledge of specific risk factors that are present at the time of diagnosis. Although ALL cannot be staged like other kinds of cancer, there is a lot of information that is used to classify patients into risk groups, with higher risk patients receiving more aggressive therapy to prevent relapse.



and is now used for all but the highest risk patients.

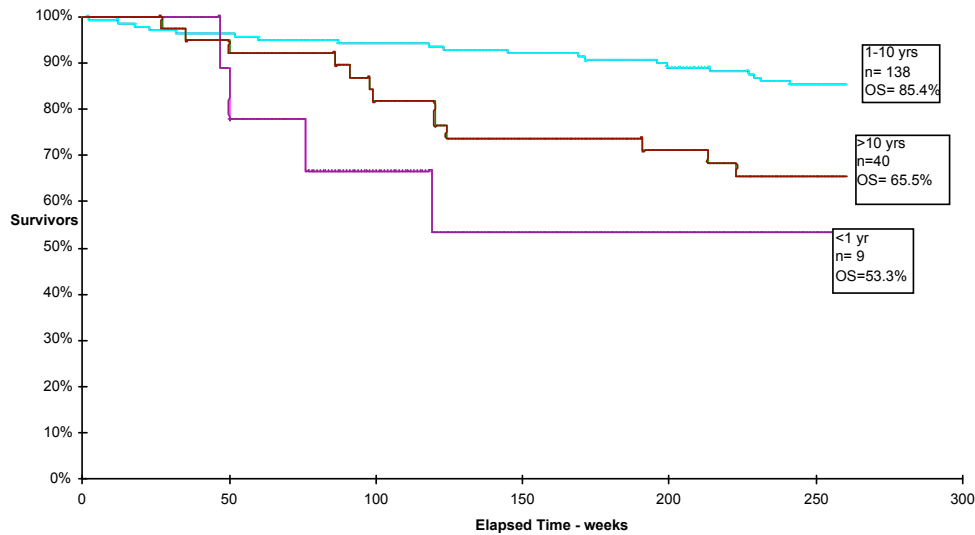
Other improvements in the delivery of chemotherapy include the use of an additional phase of intensified treatment and the use of low dose chemotherapy for an extended

Age is an important risk factor in ALL. Children diagnosed with ALL have much better chance of survival than adults with the same diagnosis. Along those lines, younger children have an improved chance of cure compared to older ones. The exception to this rule is that infants less than one year of age with ALL typically have a specific genetic abnormality in their leukemia cells that makes them high risk. Because of this, they are treated more aggressively. Currently in the United States, patients older than 10 years are considered to be high risk.

One of the earliest lessons learned when more and more children were being cured of ALL is that leukemia cells that were present in the cerebrospinal fluid were protected against the effects of chemotherapy, and many relapses occurred in the central nervous system (CNS). CNS directed therapy initially included treatment with radiation directed at the brain and spinal cord. This led to fewer relapses

period of time, referred to as maintenance therapy. Patients that are receiving maintenance therapy take medicine at home every day, and are seen in clinic on a monthly basis for laboratory monitoring and an intravenous dose of chemotherapy. Patients in this part of their treatment are just like other children; they regrow their hair and attend school daily. This maintenance phase of treatment lasts for one and a

**ALL Overall Five Year Survival by AGE
1990-1999**



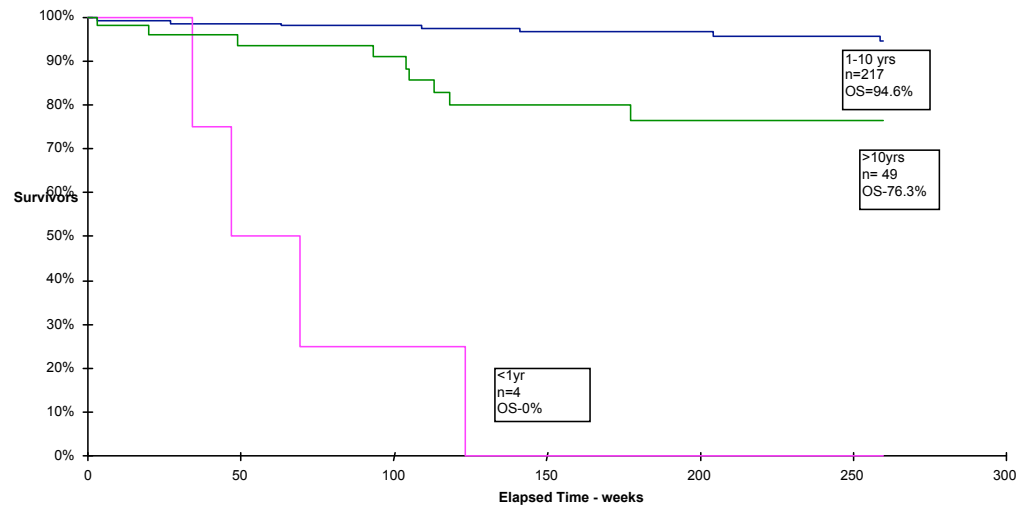
target this mutation, dramatic improvements have been made in the survival of these children. These ‘targeted’ treatments are an important focus of new research in oncology. The success achieved in ALL with the Philadelphia chromosome utilizing targeted therapeutics serves as a model for integrating new advances in our understanding of the biology of ALL into improved outcomes for patients.

The remarkable improvement in cure rates for children with ALL is looked at as one of the most impressive achievements of modern medicine. Through clinical research trials that include a great number of children, improvements continue to be made. Children’s Mercy Hospital is a member of the Children’s Oncology Group, an international collaboration focused on conducting clinical trials in childhood cancer. Currently, there are multiple open clinical trials at

White blood cell count at diagnosis is also a risk factor. Since the majority of white blood cells in patients with ALL are leukemia cells, this number represents the amount of leukemia present within the patient. A white blood count of greater than 50,000 cells per microliter of blood qualifies as high risk.

Whenever a patient is diagnosed with ALL, the DNA of the leukemia cells is analyzed. There are specific genetic mutations that when detected can tell us whether the leukemia cells may be more or less susceptible to treatment. These genetic risk factors are taken into account when planning treatment. One particular genetic abnormality called the Philadelphia chromosome is seen in a small percentage of children with ALL, but heralds a much more guarded prognosis. By integrating newer treatments that

**ALL Overall Survival by Age
2000-2009**



**ACUTE LYMPHOCYTIC LEUKEMIA / AGE 0-19
5-YEAR SURVIVAL COMPARISON RATE**

SEER 5-YEAR RELATIVE SURVIVAL RATES (PERCENT)*
CMH 5-YEAR KAPLAN-MEIER ADJUSTED SURVIVAL RATES (PERCENT) **

	1990-1992	1993-1995	1996-1998	1999-2006
	1990-1992	1993-1995	1996-1998	1999-2006
CMH	71.9 (n= 41)	79.8 (n=66)	76.3 (n=55)	91.1- (n=199)
SEER	80.0	81.4	84.2	85.9

SEER follow-up of patients into 2007

For Children's Mercy patients we calculated 5-year survival for the time periods noted. For this group of patients, the overall median length of follow-up was 376 weeks.

* Table 28.8 SEER Cancer Statistics Review 1975-2007 (National Cancer Institute)

** Cancer Registry Management: Principles and Practice- pg 431 comparison of Relative And Kaplan-Meier Adjusted Survival Modules

Children's Mercy Hospital for ALL, both for patients when they are first diagnosed and for when relapses occur.

The future for ALL treatment in children is bright. Improvements continue to be made in cure rates, and our goal of curing 100 percent of children is within reach. Newer technologies are being utilized to learn much more about the genetics of ALL, and

how differences in the genetics of individual patients can dictate how they respond to chemotherapy. By integrating this new information into treatments, their effectiveness can be enhanced while reducing the incidence of short and long term side effects of chemotherapy.



Carly's Courage

Carly Bachman was 3 years old when she was diagnosed with ALL. When asked about her memories of that time, she says “I don’t remember much, but I do remember coming [to the hospital] in the middle of the night and bringing all of my beanie babies.” Her mom remembers the event very differently. What they were told that night would drastically change the family’s life.

Looking at the healthy 17 year old today, you can



understand why her family calls her the “Miracle Child.” Carly is an active premier soccer player who looks forward to continuing her soccer playing into college. In her words, soccer brings Carly “joy and happiness.” When she chooses a career, she wants to work with kids. Her mom, Rachel, agrees that Carly will be great at this because she loves to play with kids and the kids love her.

Carly’s memories of her diagnosis and treatment are not as clear as her mom’s. She does remember that she did not like the shots she had to get in her legs several times a week. She was good at keeping track of which leg had most recently been used for the shot so the nurses could use the other one. She remembers her dad coming up with the name “Buddy” for her port-a-cath. Buddy allowed her to get her chemotherapy without a new IV being put in each time. Carly remembers the days in the hospital with some fondness. Being in the hospital meant lots of visitors bringing lots of goodies! A goody

that she does not remember fondly is strawberries. After not eating all day because she was having a procedure, she eagerly gobbled up the big bowl of strawberries one of her nurses brought in for her. Carly laughs, remembering getting sick and not liking strawberries any more after that. She also doesn’t have the same love of French fries. She ate lots of them when she was at her most vulnerable to infection. During those times, when she couldn’t go out into public, her mom would bundle her into the car for a drive

around the neighborhood and finish with a trip through the drive-through.

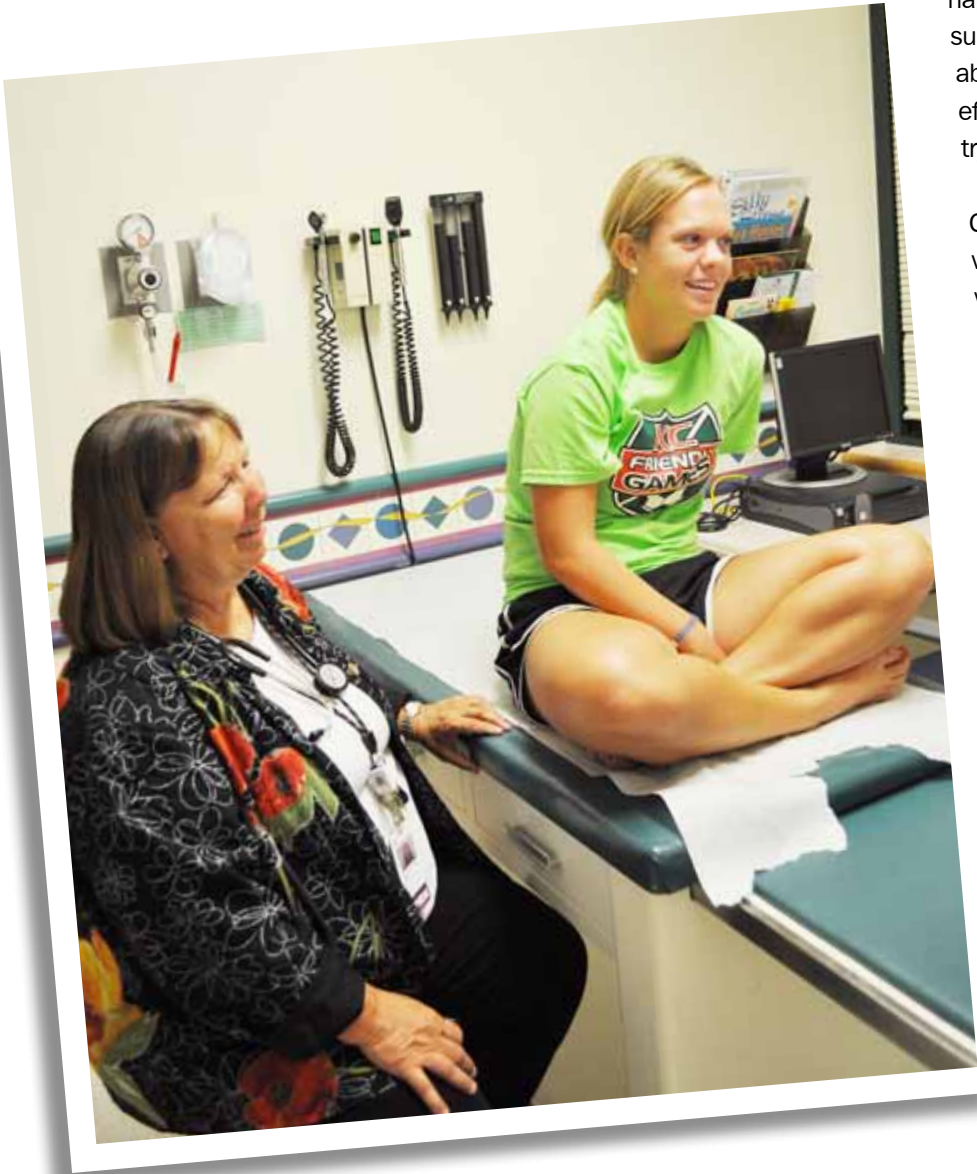
Carly does not talk much about her fight and victory over leukemia. Carly’s nurse, Cathy Burks, says that is just the way Carly is. She will not draw attention to herself. She has some friends who know what she went through as a child, but not all do. When they find out, they are surprised, but not at all surprised that Carly beat her cancer. After all, the word that she says that they would use to describe her is “strong.”

Nowadays, Carly is among the growing number of childhood cancer survivors. Sitting and talking with Carly, her mom, and Nurse Practitioner, Cathy Burks, it feels more like friends catching up after an absence than a clinic appointment. Last year for the first time, Carly attended the new “Survive and Thrive Clinic” rather than her usual yearly appointment with her treating doctor, Gerald Woods. During her appointment at the Survive and Thrive clinic, Carly had her usual physical exam, but special attention was paid to those areas of concern for people who received the type of therapy she did. She also got to spend time with other specialists like an oncology social worker and registered dietician. When she left the clinic, Carly received lots of information that would help her make health decisions as she aged out of the Children’s Mercy population.

Almost 15 years have passed since Carly and her family made the trip to Children’s Mercy that would change the way each of them looks at life. She is definitely strong, but she is also a friendly, humble, charming teenager – who also happens to have once had childhood ALL.

CATHY BURKS, RN, PCNS, CPON

Advanced Practice Nurse working with Gerald Woods, MD



Cathy Burks has worked in pediatric Hematology-Oncology for 30 years. Much has happened during those years, but Cathy especially marvels at the improvement in survival rates. Cathy notes that, “It is exciting that our treatment protocols have been able to respond to the cure rate being so high that now we look at the long term effects and try to amend our treatment to lessen those! In the 1970’s we were just trying to buy time. Worrying about late effects was pointless.”

Cathy is also grateful for the changes in techniques for giving drugs. Central lines for venous access help to avoid the often numerous pokes to find a vein at each clinic visit. There are also better drugs for nausea and pain, as well as better antibiotics. Our ability to deal with side effects and toxicities has improved dramatically. The more kid-friendly and appealing space is also a great improvement. She finds that kids now stroll into clinic knowing they will have fun if at all possible.

Cathy reflects on her career and states how very aware she is of how fortunate she and her husband are. Raising their daughters without the challenges of chronic or life-threatening illnesses is something she cannot take for granted when she knows so many families who have had such different journeys.

Cathy says she is in awe of how well the child and family handle the obstacles and challenges they face on a daily basis. She admires the strength of a family called on to deal with cancer and how they often find a way to be closer, stronger, and more appreciative of things in life the rest of us may well take for granted.

Cathy credits the special people in the Hematology-Oncology Division for creating special connections with the families served. “We have the honor to help the families that come to us. Our staff pulls together to help a child that we are honored to know.”

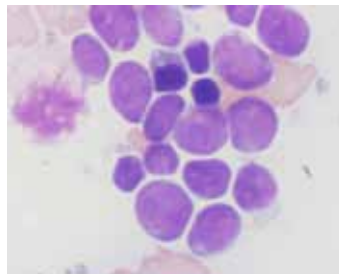
DIAGNOSTIC TREATMENT AND SUPPORT SERVICES

Pathology

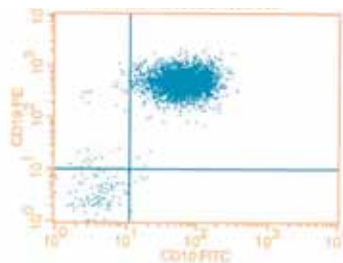
When a child comes to Children’s Mercy Hospital with a suspected diagnosis of ALL, the diagnosis is confirmed with routine and special pathology studies on peripheral blood, bone marrow, and/or spinal fluid. The number and type of leukemia blast cells is determined by the pathologist who looks at the cells through a microscope. In addition, flow cytometry tests are done that employ tagged antibodies to confirm specific lymphocyte lineage (either T or B lymphocyte lineage), as well as establish the stage of T or B cell maturation (i.e. early, common or middle, and late or mature). Stages of maturation carry prognostic significance. Common pre-B stage leukemia is generally favorable, especially compared to the B-cell ALL that lacks the common childhood ALL marker, CD10. Similarly, T-cell ALL that express the common thymocyte marker, CD1a, or coexpress CD4 and CD8, is prognostically more favorable than T-ALL that does not express these cell surface markers. Bone marrow is collected at points during treatment to allow the pathologist to determine how the child has responded to therapy.

The Pathology Department of Children’s Mercy Hospital also employs flow cytometry testing to detect residual ALL in bone marrow when the blast level falls below conventional morphologic means of detection (minimal residual disease [MRD] testing). Normal bone marrow contains about 1-3 percent blasts that ultimately give rise to, and mature into normal peripheral blood cells. On the basis of results

from a comprehensive panel of markers done on the initial leukemia cells prior to treatment, a special set of informative markers are selected for each patient that distinguish the leukemic cells from normal progenitors. These markers are again tested for on post-treatment bone marrow samples. MRD is a very useful tool for monitoring treatment response when patients are in, what is conventionally defined as, “remission” (i.e. less than 5 percent blasts in the marrow). MRD is also useful for both predicting a potential for a higher risk of relapse (if there are more than 0.01 percent MRD positive cells at the end of induction therapy), and for detecting relapse before the disease reaches conventionally high levels.



Predominance of lymphoblasts and few normal lymphocytes



Blasts that have dual expression of CD19 (B cell marker) and CD10 (Childhood acute lymphoblastic leukemia antigen)

Cytogenetics

Current treatment strategies adjust therapy of leukemia patients according to the genetic subtype of the individual tumor. Cytogenetic analysis plays a vital role in defining this genetic subtype of leukemia cells. Cells from each newly diagnosed leukemia patient are studied in order to optimize therapy using the least toxic, most effective therapeutic regimen.

A blood or bone marrow sample is sent to the Cytogenetics Laboratory at diagnosis. The leukemia cells from the sample are cultured and harvested during the metaphase portion of the cell division cycle (a seven minute window). The harvested cells are applied to glass slides and the chromosomes are stained to show the band pattern. This allows each chromosome to be identified and analyzed by skilled technologists.

Metaphase chromosomes are analyzed for any and all genetic defects, including the number of chromosomes (normal is 46) and any structural defect. Structural rearrangement of chromosomes frequently identifies cancer-specific genes. Research has shown that some of these genes can be targeted by mutation specific therapies. An example is the t (9;22) that results in rearrangement of the ABL1 gene on chromosome 9 with the BCR gene on chromosome 22. This rearrangement changes a normal tyrosine kinase gene into one which drives cell proliferation and leukemogenesis. Tyrosine kinase inhibitors have been identified to target the abnormal BCR/ABL1

fusion product. Targeted therapies are less toxic than conventional cell cycle chemotherapy and more effective since they target the specific genetic defect.

The Cytogenetics Laboratory submits the cytogenetic data to the Children's Oncology Group (COG) Cytogenetics committee when a patient is enrolled on a COG study. This information is used to determine which arm of a COG therapy protocol is most appropriate for each child. COG collects this data from many institutions that enroll their patients on study. The collected data is analyzed at the end of the study to correlate between genetics and patient outcomes. This information provides valuable guidance on the most effective therapy for specific genetic abnormalities.

In addition to the conventional chromosome analysis, the Cytogenetics Lab performs FISH (fluorescence in situ hybridization) analysis to detect submicroscopic chromosome abnormalities and to confirm the molecular rearrangements caused by chromosome translocations. All information gathered from these methods is utilized in determining the most effective therapy for each patient.

Follow-up cytogenetic or FISH analyses are done to determine the presence or absence of residual disease after treatment. Residual disease has prognostic significance. If there is a relapse of disease, conventional chromosome analysis will determine if there are changes in the genetics of the leukemia (i.e. clonal evolution).

The goal of the Cytogenetics Lab is to determine, with the greatest accuracy, what the genetics are for

each child's leukemia so the child can receive optimal therapy and the best chance for long-term survival.

Pharmacy

Over last decade, chemotherapy for ALL has become more specialized. Patient treatment - both the number and dose intensity of chemotherapy agents - is based on each patient's risk for relapse. Many new drugs have also been incorporated into the treatment of ALL. These drugs are given based on the type of leukemia or specific genetic abnormalities. The tyrosine kinase inhibitors such as imatinib and dasatinib (both oral agents) are now included in the treatment regimens for patients with t(9;22) translocation. Other new agents that have been used in the treatment of ALL include nelarabine for T-cell ALL and clofarabine, the first drug within a decade to be FDA-approved specifically for the treatment of a pediatric cancer.

The Pharmacy Department is integral to the complete care of oncology patients. The Children's Mercy Pharmacy staff and facilities have evolved over the past decade with a goal of providing the best pharmaceutical care possible for all patients, including those with cancer. The distinct teams within this department include Inpatient/Clinic Operations, Clinical Specialist Pharmacists, Investigational Drug Service, Home Care and Outpatient Operations. Within the Hematology/Oncology Division, three clinical pharmacy specialists work with patient families and assist the primary team in optimizing patient medications based on drug interactions, disease states, and organ function. In addition to our pharmacy specialists, three pharmacists and a technician are dedicated to the review of

chemotherapy orders and safe production and distribution of chemotherapy to all patients within the Children's Mercy system. Our investigational drug service works with over 100 open drug trials for our patients, including many phase II oncology drug studies. Our outpatient pharmacy is able to compound many prescriptions that are not commercially available.

Our Hem/Onc pharmacists are dedicated to providing education to pharmacy students through clinical rotations and lectures at our local school of pharmacy. In addition, our Pharmacy Department has two nationally accredited post-graduate residency programs. In 2008 we became only the second program in the nation for post-graduate residency training of pediatric hematology/oncology pharmacists. This residency program provides pharmacists with focused and intensive training in the care of pediatric patients with cancers.

Inpatient Nursing

Nurses at the bedside have seen many changes and growth in the treatment of ALL in the last decade. With the use of Gleevec for High Risk ALL, nurses have witnessed more positive outcomes for their patients. Relapsed ALL patients now have more options for treatment, including the possibility of a bone marrow transplant. With the improvements in treatment, Standard Risk ALL patients are rarely seen inpatient for treatment or side effects. Bedside nurses are provided more resources and education to assist families at the time of diagnosis. This has allowed us to prepare our families to care for their children and understand the treatment.

While the physicians and residents are responsible for all aspects of diagnosis, nursing input is welcome and valued. It is usually the inpatient nurse's responsibility to coordinate care between the multi-disciplinary care teams. Inpatient nurses are the front line of daily care, alerting the care team to changes in patient status. Patients and their families rely on the nursing staff for new diagnosis education as well as assisting the social workers with psychosocial care during a stressful time of life.

Clinic

The Children's Mercy Hospitals and Clinics Hematology/Oncology Clinic nursing team are the first line providers of patient care in the outpatient setting. Every patient has a primary care team that consists of an attending physician, advanced practice nurse, and social worker. Nursing follows this model. Having a primary nurse during a lengthy treatment allows for consistency in care and optimal patient and family experiences. Each nurse works with a set of patients and families with whom they will develop professional relationships that foster understanding and trust and a reduction in overall anxiety. Our nursing team is able to provide the supportive care, education and anticipatory guidance appropriate for each individual patient and family member. This is based on their knowledge and understanding of each patient's diagnosis, treatment plan, educational needs, and family dynamic.

As the first health care providers to access patients in the Hematology/Oncology Clinic, nurses are responsible for medication and allergy histories, obtaining a history of symptoms, and for providing nursing interventions, such as

medication administration. At Children's Mercy, the Hematology/Oncology nurse is required to be certified in the following: Chemotherapy and Biotherapy administration, Basic Life Support, Pediatric Advanced Life Support, central line care, and sedation. In addition, nurses are required to take and pass the General Anesthesia Registered Nurse course provided by the hospital and a Bone Marrow Transplant course provided by the Division of Hematology/Oncology. In addition to Children's Mercy requirements, many of our nursing staff members voluntarily test to become certified in pediatric oncology (Certified Pediatric Oncology Nurse), demonstrating an expert knowledge base in this specialty. Because of this, nurses are equipped to provide the same treatment that would be otherwise be provided in an inpatient setting. Treatment provided includes: chemotherapy, high risk biotherapies and monoclonal antibodies, stem cell infusions, administration of blood products, and sedation for procedures. Nursing also provides supportive care for the Hematology/Oncology patients who require anesthesia for procedures, but are not candidates for sedation. candidates and require anesthesia for procedures. Nurses in the clinic must also be prepared to care for children that present with oncologic emergencies, such as anaphylaxis and sepsis. This means knowing what resources are available and accessing them appropriately.

FACT Team

Although there have been many advancements in treatment for children with ALL, caring for a child diagnosed with ALL remains a daunting road for parents, siblings, family members and friends. The Hematology/Oncology Family Care Team (FaCT) recognizes that treatment is not merely a series of

chemotherapy, clinic appointments and inpatient admissions. It is a process where the most important outcome is that the patient and family are able to redefine their 'new normal' during treatment and after treatment ends. For many patients and families experiencing leukemia, life will never again be the same. They require continued adjustment with each new developmental stage of their life.

The FaCT team consists of social workers, child life specialists, music therapists, a family therapist, a psychologist, a chaplain and a hospital -based school teacher. Each of these highly specialized staff members is equipped to support families through each step of the process, including diagnosis, active treatment, end of treatment, and late effects. The overarching goal of the FaCT team is to assist each patient and family member by providing support and guidance on an individual basis.

Since ALL treatment lasts two to three years, the impact on children and families is large. Members of the FaCT team are available to provide support to the patient, families and school staff in order for the patient to stay connected with their peers during treatment. Team members can meet with siblings and classmates to provide education about the diagnosis and treatment and facilitate discussion of how they can support the patient in their treatment process. As part of an ongoing relationship with each patient and family, the family care team considers it a privilege to accompany patients and families on their journey and bear witness to their determination, courage and resilience.

Nutrition

A child's appetite and weight change while undergoing therapy for ALL is similar to riding a roller coaster. Families are frequently frustrated when the thing that their child was craving is pushed aside after only a couple bites.

Nutrition therapy for a child with ALL centers around assessing the child's usual diet and the family's health habits. Nutritionists provide anticipatory guidance related to taste changes, steroid-induced weight gain, and increased vitamin and mineral requirements while on chemotherapy.

The consequence of prolonged inadequate intake is nutrient deficiencies that can lead to increased chemo toxicity. Prolonged inactivity decreases energy expenditure which can lead to further decreased appetite as well as later risk for obesity. At other times children struggle with excessive cravings and eating caused by the steroid component of therapy, which leads to a concerning pattern of obesity. Children with ALL who are not following age-appropriate growth rates, based on their growth chart, are referred for nutrition assessment and recommendations.

Changes in nutrition therapy for children with ALL over the past decade relate to vitamin/mineral needs and obesity prevention. Nutrients found in fruits and vegetables, called phytochemicals, are now strongly encouraged at age-appropriate amounts (5-9 servings per day) throughout ALL therapy, unlike previous recommendations for neutropenic diets (only frozen or canned fruits & veggies). Safe food handling, including appropriate washing of fresh fruits and vegetables, has replaced neutropenic diet recommendations

during most cancer treatments. Increased fruit and vegetable intake throughout ALL therapy is associated with fewer hospitalizations for blood stream infections and lessened chemotherapy-related toxicities. This helps children finish their lengthy course of therapy on time.

Another major nutritional concern is decreased bone mineral density and risk of osteopenia or osteoporosis. Children undergoing treatment with steroids have increased calcium and vitamin D needs in order to maintain normal growth and development. Nutritionists evaluate diets and the child's current intake levels of these important nutrients. Education is provided to families with tips for dietary changes or appropriate supplements as indicated.

Nutrition education for children with ALL also deals with the increased incidence of obesity in adult survivors of childhood leukemias. This is especially seen in those who required CNS radiation and/or stem cell transplant. Screening for obesity risk factors throughout therapy and annually after completion of therapy is essential to identifying children and young adults who would benefit from additional nutrition assessment and education.

Stem Cell Transplant

The Stem Cell Transplant team is comprised of specially trained pediatric hematologist/oncologists, advanced practice nurses, physician assistants, pharmacists, transplant coordinators, and research staff. Each team member plays a part in taking care of children who are referred to the team for a transplant. Indications for a transplant following a diagnosis of leukemia are relapse and disease that

does not respond to chemotherapy. Because each child is unique, a full consultation is done with the family members before it is decided how or if to move forward with transplant.

Children's Mercy offers families the opportunity to have a gold standard transplant within the specially designed transplant unit of the inpatient floor. Care is provided by trained caregivers who understand the intricacies of caring for these patients.

Research

Progress in the treatment of ALL is one of the leading success stories in all of cancer research. The dramatic breakthroughs, for the most part, have come through clinical trials. Most of the clinical trials offered to patients with ALL in the United States come from a cooperative effort with the Children's Oncology Group (COG), an association of institutions dedicated to research in pediatric oncology. Lessons learned from the treatment of childhood ALL have been applied to other less curable cancers, even those that strike adults.

COG offers clinical trials to study every gamut of ALL. Current areas of research include:

- Epidemiology and Biology
- Treatment protocols
- The use of investigational new drugs
- Relapse/Refractory Disease
- Late effects and long-term follow up

Biology/Laboratory Research

Over the past 10-15 years, researchers have made progress in the basic scientific understanding of ALL. A single cell in the body develops some mistakes



in the genes crucial for controlling cell growth and development. Those mistakes (called mutations) make the cancer multiply. By enrolling patients on biology studies, we are learning why these mistakes occur and how knowledge of these abnormalities can be used to improve treatment for children and adolescents with ALL.

Treatment Protocols

Clinical trials for the treatment of ALL are focusing on risk-adapted therapy. This helps low-risk patients (patients predicted to have a very low risk of relapse) avoid unnecessary treatment and side effects, while giving higher-risk patients stronger treatment to improve their chance of being cured. Therapy is tailored to the individual patient based on a series of clinical (age, gender) and biological features (white blood cell count, chromosome content of blasts) at diagnosis as well as an assessment of how quickly the leukemia responds to treatment.

Treatment of Relapsed and Refractory ALL

Despite the progress made in treating children with

ALL, children still relapse. Since it is more difficult to cure a child who has relapsed, better treatments are being sought for these patients. Laboratory studies are helping us to understand why some patients with ALL relapse, but others do not. New drugs are being rapidly integrated into conventional chemotherapy protocols to improve outcome. Clinical

trials have been designed to evaluate new drugs in the context of more conventional therapy to speed the development of promising new agents. In addition, COG is aiming to improve outcomes after stem cell transplant by decreasing the burden of leukemia prior to transplant and piloting the use of drugs that simultaneously combat the leukemia and prevent graft vs. host disease.

Long Term Follow up and Late Effects

Ongoing clinical research is determining which patients are at greatest risk for known side effects and intervention trials are now planned to decrease the psychological burden on patients and families, to prevent neuro-psychological deficits, and to decrease problems with bone and joint development and muscle strength and flexibility.

Survive & Thrive

Survival is now a reality for almost 80 percent of children diagnosed with cancer. In the U.S today there are over 270,000 childhood cancer survivors.

While surviving is a major accomplishment in itself, many of these children will have the challenge of some kind of health problem related to their treatment for cancer, even years after that treatment has ended.

These “late effects” of childhood cancer can include heart and lung dysfunction, growth issues, psychological and educational concerns, as well as the possibility of a second cancer. The Division of Hematology-Oncology created the Survive & Thrive Late Effects program to provide comprehensive physical and psychosocial assessment. The program provides a treatment summary for each patient and educates them on what late effects they may be at risk for, what type of health care follow-up is necessary and how to make healthy lifestyle choices. The Survive & Thrive Clinic is staffed by a medical director, nurse coordinator, clinical social worker and registered dietician. We also work closely with a radiation oncologist and endocrinology physicians. Other specialty services are consulted as needed. An additional goal of the clinic is to assist patients in making the transition from pediatric care to adult health care, ensuring that they will continue to get the long term follow up they need.

The Survive & Thrive Late Effects program is important because even though these kids have survived their cancer diagnosis and treatment, they can still face incredible challenges in their lives with many chronic and debilitating health issues. Our goal is to identify these issues and provide them with the resources they need to live the healthiest and most productive life possible.

Children's Mercy



Cancer Center
Tough Fight! Tough Kids!

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