

PHYSICIAN'S

September
2008

UPDATE

Comprehensive Care for
Complex Cases

BMT, Sickle Cell,
Coag and Oncology

Updates



Children's Mercy
HOSPITALS & CLINICS
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Leukemia, the most common form of childhood cancer, occurs in the tissues within the body that produce the body's blood cells and bone marrow. About one-third of all cancers in children under the age of 15 are diagnosed as leukemia, as are about one-fourth of all cancers in people under 20.

Fortunately, the cure rate for Children's Mercy patients diagnosed with ALL (Acute Lymphocytic Leukemia)—the most common type of cancer in children—is 80 to 90 percent, with a 99 percent remission rate. The hospital's

"What drives us is that we can cure most of the kids we see."

Alan Gamis, MD, Section Chief, Oncology Chair, Myeloid Cancer Section, Children's Oncology Group

AML cure rate is around 60 percent, with 90 percent of patients going into remission. This is significant as AML is one of the deadliest forms of childhood cancer and occurs in 20 percent of patients.

"Finding better outcomes is a key focus area," says Alan Gamis, MD, Children's Mercy Oncology Section Chief, and Professor of Pediatrics, UMKC School of Medicine. "As the largest pediatric cancer center in the region, Children's Mercy is actively involved in ongoing research, participating in leading-edge clinical trials."

Children's Mercy is expanding the research effort toward finding a cure for ALL and many other cancers. With Children's Mercy diagnosing almost 170 new cases of cancer every year, there is a commitment to improved cure rates, daily interaction with oncology experts around the country, and looking ahead for new treatments.

As chair of the Myeloid Cancer section of the National Cancer Institute's only pediatric cooperative group, the Children's Oncology Group (COG), Dr. Gamis oversees most of the childhood AML (Acute Myelogenous Leukemia) research in the U.S. Participation in the COG also means that Children's Mercy patients have access to the same treatment that is provided at Sloan Kettering, MD Anderson, St. Jude's and other well known cancer centers.

"Children's Mercy is actively involved in more than 100 national trials, assessing new patients or following up patients previously enrolled," says Dr. Gamis. "All of our patients have access to enrolling in and benefiting from those studies. There is no need to send children anywhere else, because the gold standard of care is right here."

The Children's Mercy Hematology/Oncology team evaluates and treats patients up to 21 years of age who have all forms of acute and chronic leukemia, lymphoma, or other diseases that may involve the bone marrow.

Treatment at Children's Mercy, depending on the type of leukemia, can include chemotherapy infusions, bone marrow transplant, or

radiation. Certain features of a child's leukemia, such as age and initial white blood cell count, are used in determining the intensity of treatment needed to achieve the best chance for cure. Additionally, the genetic and molecular nature of the leukemia are increasingly becoming critical factors in prognosis. Children's Mercy's laboratories have the latest and most advanced methods used in these analyses and serve as reference labs for patients from many other centers. After treatment begins, the goal is remission of the leukemia.

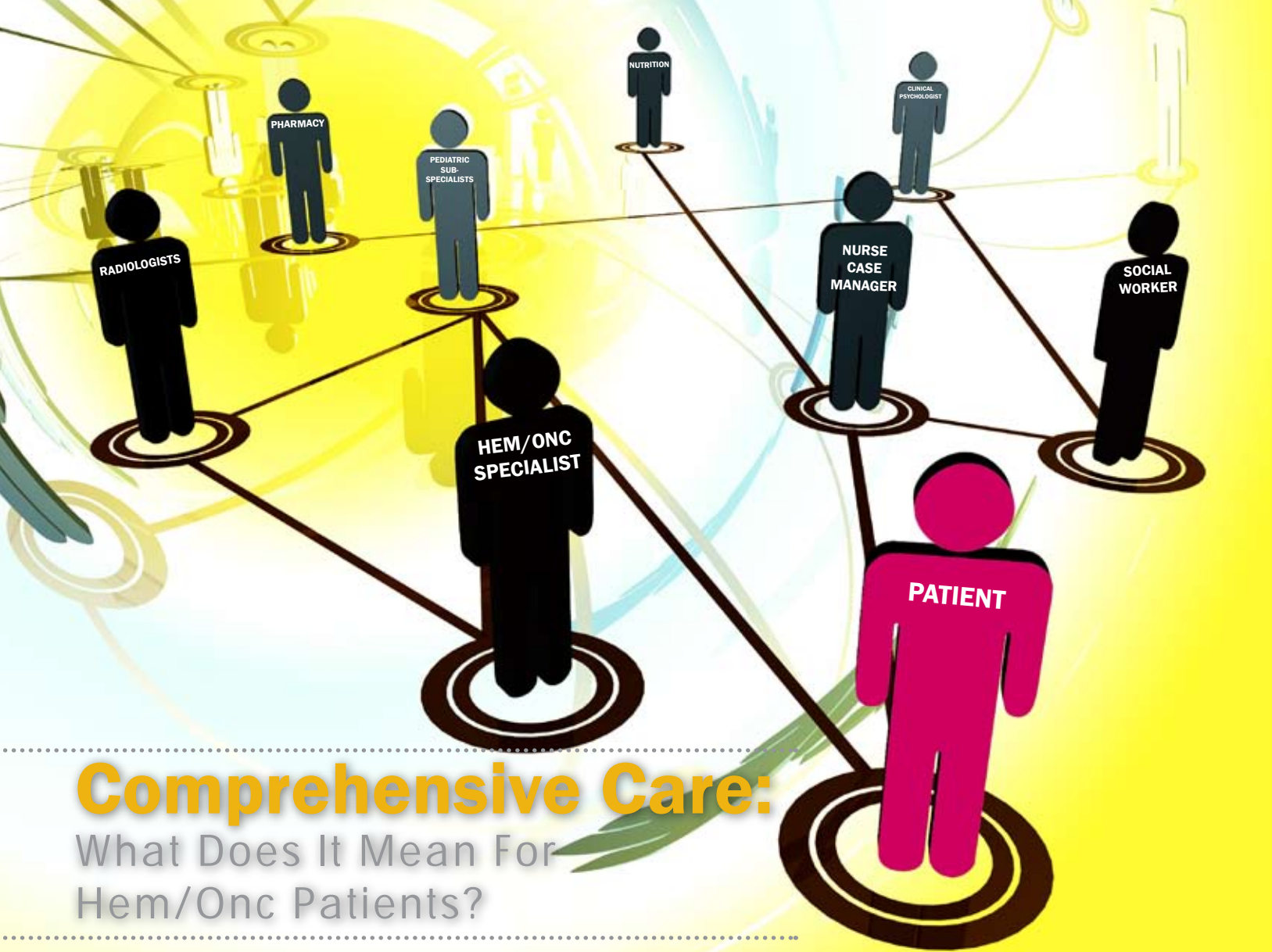
"Children's Mercy is leading the way—we are motivated to find a cure and to lessen the suffering of our patients. As much as Children's Mercy is ramping up the research, which is critical, it's also important to note that we dedicate that same level of energy to every child," Dr. Gamis says.

He adds, "What drives us is that we can cure most of the kids we see. With a continued, persistent focus to be a key participator in research, as well as our multidisciplinary approach, Children's Mercy is making a difference."

Alan Gamis, MD, with patient Kellen Bolling



CLINICAL HIGHLIGHT: Leukemia



Comprehensive Care:

What Does It Mean For Hem/Onc Patients?

One hour. For cancer patients with depressed immune systems, one hour without appropriate treatment can be the difference between life and death.

That's why Children's Mercy patients who live further than one hour away from the hospital are provided with an antibiotic kit to take to their local emergency department or physician in case of emergencies. Upon discharge from the hospital, the patient's local emergency department and physician are notified and provided with instructions for use of the kit. To the knowledge of Oncology Section Chief Alan Gamis, MD, Children's Mercy is the only hospital in the country to provide this service.

It is just one example of the comprehensive, multidisciplinary care provided by the Hematology/Oncology Division at Children's Mercy. Children's Mercy is one of the largest childhood cancer and blood disorder centers in the country. Last year the center treated 150 newly diagnosed cancer patients and approximately 200 children with newly diagnosed hematology disorders.

Children's Mercy has 10 board certified hematology/oncology specialists on staff, and in some cases such as

hemophilia, the only physicians in their specialty within the region. But caring for children with these types of diagnoses requires more than just physicians.

"The highly complex nature of hematology disorders and cancer care requires a well-coordinated multidisciplinary team approach," says Gerald Woods, MD, Hematology/Oncology Division Chief, and Professor of Pediatrics, UMKC School of Medicine. "What makes our program special is that we take a holistic approach – we look at the whole child, from top to bottom, and ensure that each one is meeting with every section of the hospital as needed to manage the disease."

Case Management

Every patient is assigned a nurse case manager, usually an advanced practice nurse or physician's assistant teamed with a social worker and a hematology/oncology specialist, who oversees each child's case from diagnosis through follow-up care. These case managers provide education on the child's diagnosis, the plan of care, medications, side effects and treatment schedule, and help coordinate admissions, tests and checkups, and visits to the primary hematology/oncology care doctor and additional sub-specialists. They may also provide physical exams, prescribe common medications and perform certain procedures.

FAMILY SUPPORT AND RESOURCES

Also, every patient is assigned a social worker to help address the psychosocial, emotional needs of the child, as well as assist in coordinating family and community services, including supportive counseling, anticipatory guidance, school reentry, linkage to financial support, and grief and loss counseling.

“Throughout their care our hematology/oncology patients will have the same case team. The case manager is the first contact for the family whenever they have questions or concerns,” says nurse practitioner Sue Stamm, Hematology/Oncology Division Manager. “The benefit is that they have consistent people who know their child and family and based on our experience, we believe that increases continuity of care and patient satisfaction.”

Treatment plans are developed based on the child’s unique medical and supportive needs and developmental level.

Care is provided by a team of specialists that extends well beyond nurses and hematology/oncology physicians.

- **Radiologists** - Board certified pediatric radiologists using a new 3 Tesla MRI unit, two 1.5 tesla magnets and three multi-slice CT scanners, help ensure effective and safe surgical and radiosurgical planning.
- **Pharmacy** - Two full-time pharmacists and a technician support the inpatient and outpatient areas in preparing and dispensing chemotherapy. Full-time pharmacists also provide consultation for the hematology/oncology and bone marrow transplant services.
- **Nutrition** - Dedicated registered dietitians and techs support the unique nutrition needs of patients.
- **Child Life Specialists** - Four child life specialists and a music therapist work with patients, parents and siblings to aid in the adjustment and ongoing support associated with the child’s diagnosis and treatment.
- **Clinical Psychologist** - A dedicated, full-time psychologist provides support to families of children diagnosed with childhood cancer or blood disorders.
- **Discharge Planner** - Discharge planners assist with arrangements for home infusion, nursing visits, medical supplies, prescriptions and other post-hospitalization treatment arrangements.
- **Pediatric Subspecialists** - Conferences provide opportunities for multidisciplinary discussion of patient care with a comprehensive array of specialists including hematology/oncology, surgery, pathology, radiology, radiation oncology, neurosurgery, orthopaedic surgery, endocrine, psychology, and dentistry.
- **Chaplaincy** services are offered 24/7 to provide comfort and assist families with the emotions and spiritual questions that often accompany a diagnosis of cancer, or blood disorder.

“When you are dealing with childhood cancer and blood disorders, you are dealing with illnesses that will affect patients for the rest of their lives,” says Dr. Woods. “We have a tremendous responsibility and obligation to provide the children we treat and their families with the highest level of care possible and that care can only be provided through a coordinated, multidisciplinary approach.”

When children are diagnosed with cancer and blood disorders, it affects the entire family. Children’s Mercy offers a family-centered approach designed to encourage parents and families to make informed decisions regarding their child’s care and quality of life.

The **Parent-to-Parent program** is designed to connect parents who have gone through the cancer experience with parents of a newly diagnosed child and offers weekly support gatherings.

Children’s Mercy contracts with the Kansas City Missouri School District to have three **full-time teachers** based at the hospital to help students keep up with school assignments. Teachers work with students from all school districts and grades.



The **Ronald McDonald House** offers a place to stay for family members who must travel more than 35 miles for their child’s treatment. Families can use the house during initial hospital visits and for trips associated with ongoing or follow-up therapy.

Camps - Children’s Mercy patients and staff participate in several camps that allow children to interact with others who have had similar diagnoses and experiences. These camps include Camp Courage (Sickle Cell), Camp Hope (Oncology), Camp Quality (Oncology), Camp Wilderness (Hemophilia), Hole in the Wall Gang Camp, and Camp Barnabas.



ADVANCING CARE THROUGH RESEARCH

Through partnerships and collaborations with national groups, Children's Mercy Hematology/Oncology patients are able to participate in hundreds of research studies. In addition, Children's Mercy faculty and research scientists are also pursuing their own innovative research studies.

Two areas of particular interest are the role of genetics in how children respond to treatment and how a patient's quality of life is affected by cancer or blood disorder.

Individualizing Cancer Treatment

Despite the widespread use of cyclophosphamide to treat a wide variety of cancers, relatively little is known about how it is metabolized in infants and children. Some of the complicated metabolic reactions of this drug cause tumor cells to die while others cause severe side effects.

Jignesh Dalal, MD, Bone Marrow Transplantation, and Andrea Gaedigk, MS, PhD, Clinical Pharmacology, are conducting a study to better understand how cyclophosphamide is metabolized in children. The study is funded by a grant from the Tom Keaveny Foundation.

The amount of enzymes that metabolize certain drugs varies considerably from person to person, which is mostly due to a person's genetic make-up. An additional layer of complexity is that many of these drug-metabolizing enzymes 'turn on' at variable times after birth, usually during the first year of life.

"This research will help give us a better understanding of how this drug should be dosed for children," says Dr. Gaedigk. "Since it can make them very

sick, every bit that we can reduce the amount they get and still receive the benefits will help."

Quality of Life

Not only are the professionals of the Division of Hematology/Oncology concerned about the treatments that their patients receive, but also the impact that the diagnosis of cancer or a blood disorder has on their development and quality of life and ways to mitigate the negative psychosocial effects of these disorders.

In collaboration with other researchers at Indiana University School of Nursing and across the country, Kristin Stegenga, RN, PhD, is studying ways to promote resilience and quality of life in adolescents and young adults undergoing stem cell transplant. This study is open at a limited number of institutions across the county and is funded both through the National Institutes of Health's National Institute of Nursing Research and the Children's Oncology Group. It is the first of its kind for the Children's Oncology Group as a supportive care study.

Dr. Stegenga and colleagues have also looked at quality of life issues related to the effect of cancer diagnosis on adolescents and decision making, and the impact of treatment to prevent strokes on the quality of life of children with sickle cell disease.

TREATING SICKLE CELL DISEASE

Despite significant advances during the past two decades, sickle cell disease remains a painful, unpredictable chronic disease. A painful "episode" or "crisis" is the most common symptom of sickle cell disease and the top reason that people with the disease go to the emergency room or hospital.

Children's Mercy currently follows approximately 300 children with sickle cell disease. In the past year, there were 258 admissions and several hundred Emergency Room visits due to complications related to sickle cell disease.

While progress has been made, Children's Mercy remains dedicated to improving quality of life for these patients. The hospital has participated in numerous studies during the past 20 years and is currently involved in research studies evaluating pain and stroke treatment.

"In the 1970s, the average life span of a patient with sickle cell disease was approximately 20 years; however, advances in the management of sickle cell disease have dramatically changed the outlook for our patients," says Gerald Woods, MD, Division Chief, Hematology/Oncology, Sickle Cell Program Director, and Professor of Pediatrics, UMKC School of Medicine.

Treatment

Treatment of complications often includes antibiotics, narcotics, intravenous fluids, blood transfusion and surgery, all backed by psychosocial support. Like all patients with chronic disease, sickle cell patients are best managed in a comprehensive multi-disciplinary program of care.

Blood transfusions help benefit sickle cell patients by reducing risk of stroke and other complications. Patients who receive repeated blood transfusions can accumulate iron in the body until it reaches toxic levels, so treatments are available to eliminate iron overload.

In search for a substance that can prevent red blood cells from sickling without causing harm to other parts of the body, Hydroxyurea was found to reduce the frequency of severe pain, acute chest syndrome and the need for blood transfusions in adult patients with sickle cell anemia. Children's Mercy is currently participating in a phase III study (SWITCH) to investigate the effectiveness of Hydroxyurea in children.

"Hydroxyurea is a chemotherapy agent with potent effects on the bone marrow, which was used for many years to treat people with certain malignancies before being used for sickle cell disease. To date, Hydroxyurea is the first agent that can prevent complications of sickle cell anemia," says Dr. Woods.

Children's Mercy also offers bone marrow transplantation, which is the only cure for sickle cell disease. Bone marrow transplantation has been shown to be effective by replacing short-lived sickle cells with longer-lived healthy red blood cells.

"Twenty years ago, the prognosis for patients with sickle cell was not promising. Now with comprehensive care, clinical knowledge, and active research, our patients are living longer, more productive lives," says Dr. Woods.



Quality Improvement

Anticoagulation Management Program Seeks to Improve Safety

Heparin and Warfarin are the workhorses of anticoagulation medications, but both come with significant risks as well as rewards. The risk is such that The Joint Commission has mandated that hospitals implement an anticoagulation program to monitor and guide the use of these medications. There has been national media attention to this after the Dennis Quaid twins received excessive doses of heparin while hospitalized in Los Angeles.

“Heparin and Warfarin are high risk because of the difficulty in determining the correct dosage for each patient, the complexity of monitoring these drugs and the risk of bleeding if too much drug is given,” says Brian Wicklund, MDCM, MPH, Director of the Hemophilia/Coagulation/Thrombosis program at Children’s Mercy and Associate Professor of Pediatrics, UMKC School of Medicine.

Dr. Wicklund, Christine Walsh-Kelly, MD, Pediatric Emergency Medicine specialist and Professor of Pediatrics, UMKC School of Medicine, and Susan Feldman, Clinical Safety Officer, are leading the quality improvement initiative at Children’s Mercy. The program will be piloted in the NICU and the Pediatric Intensive Care Unit this fall and then rolled out to other units by January 2009.

The challenges lie in how each drug works.

Heparin response is variable and works through augmenting the effect of a naturally occurring protein in the blood. Response to heparin is very rapid. If too much is given, bleeding becomes a problem.

Warfarin dosing is slow to affect the body and may take up to two weeks to get into the proper therapeutic range. It is a vitamin

K antagonist and is sensitive to foods and medications ingested. Two genetic mutations have also been identified which influence how the body reacts to Warfarin.

“Heparin is one of the most widely used drugs in the hospital,” says Dr. Wicklund. “We would love to get rid of Warfarin, but it is the only oral anticoagulant we have right now.”

The QI initiative starts with standard protocols based on national recommendations and guidelines for the use of these drugs in pediatric patients, with the goal of getting everyone to dose and monitor use in the same way. The program is hospital-wide and involves physicians, nurses, pharmacists and dietary personnel. Special guidelines will be developed specific to areas such as cardiovascular surgery and cardiology, ECMO and hemodialysis. Education of prescribers, staff, parents and patients is also part of the program.

Quarterly audits will evaluate time to achieve therapeutic range, percentage of time in the therapeutic range, complications and adverse events, education provided and other clinically useful measures.

“This is really a national patient safety goal,” says Dr. Wicklund. “We want to establish protocols to guide services and monitor their delivery, so that everyone is dosing and monitoring in the same way.”



NOTABLES

In 2006, Children’s Mercy was the only pediatric hospital in the nation to receive the Outstanding Achievement Award from the American College of Surgeons Commission on Cancer. Only 31 hospitals in the entire nation received this prestigious award, which recognizes the significant commitment made in providing high quality cancer care to our patients and exceeding the standards of excellence set by the Commission on Cancer Approvals Program.



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A PATIENT'S STORY

Brandon Cobb

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Diagnosed at an early age with sickle cell disease, Brandon Cobb recognized that his health was something he had to work at harder to maintain than other children his age. It was this realization that would drive Brandon to be a survivor, versus a victim, of the chronic disease.

With regular visits to his primary care physician and Gerald Woods, MD, at Children's Mercy, Brandon focused on a care regimen that enabled him to feel like a regular kid, including playing on his high school football team.

One of Brandon's key motivators was attending Camp Courage, a weeklong overnight camp for children with sickle cell disease. As a camper for eight years, Brandon met other children with sickle cell. His experience motivated him to give back, so Brandon became a camp counselor at age 18.

"As a young adult with sickle cell, I am a role model for the other kids by showing them that they can lead a happy, healthy, and productive life."

Currently, Brandon is a student at the University of Missouri-Kansas City, working toward a PhD in psychology. "Children's Mercy was a wonderful medical home for me and Dr. Woods always encouraged me to live my life to the fullest—and I'm doing just that."



HOW TO REFER

For Transport, Inpatient Admission or for Consult
1-800 GO MERCY
(1-800-466-3729)

Call this number 24 hours a day to mobilize the in-house neonatal or pediatric transport teams, consult with a specialist, or admit a patient directly to Children's Mercy Hospital or Children's Mercy South.

For Specialty Clinic Appointments
(816) 234-3700 or
toll free 1-800 800-7300

Nurses with our Physician Appointment line can assist you with scheduling clinic appointments for Children's Mercy Hospital, Children's Mercy South and Children's Mercy Northland.

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