

Children's Mercy Kansas City

SHARE @ Children's Mercy

Cancer Center Annual Reports

Oncology, Hematology and Bone Marrow
Transplant

2006

2005 Cancer Care Annual Report

Children's Mercy Hospital

Let us know how access to this publication benefits you

Follow this and additional works at: https://scholarlyexchange.childrensmc.org/cancer_center_annual_reports



Part of the [Oncology Commons](#), and the [Pediatrics Commons](#)

Recommended Citation

Children's Mercy Hospital, "2005 Cancer Care Annual Report" (2006). *Cancer Center Annual Reports*. 12. https://scholarlyexchange.childrensmc.org/cancer_center_annual_reports/12

This Book is brought to you for free and open access by the Oncology, Hematology and Bone Marrow Transplant at SHARE @ Children's Mercy. It has been accepted for inclusion in Cancer Center Annual Reports by an authorized administrator of SHARE @ Children's Mercy. For more information, please contact hlsteel@cmh.edu.

2005 CANCER CARE **ANNUAL REPORT**



table of contents

Patient Highlight: Patrick.....6

Division of Hematology/Oncology7

Patient Highlight: Petra11

Cancer Care Board Members12

Patient Highlight: Richard.....13

Cancer Registry14

Patient Highlight: Santana16

Neuroblastoma17

Patient Highlight: Satala24

Diagnostic and Treatment Support Services25

References.....27



This year's annual report focuses upon neuroblastoma, a childhood cancer that primarily occurs in the youngest of our patients, those less than 5 years of age. Over the years, research has shown that though most neuroblastoma tumors have relatively similar appearances under the microscope, there are in fact two unique forms of this cancer with widely different chances of cure. As you will read in this report, this has allowed us to identify these groups, and to reduce therapy to those who can be cured with less, and to increase therapy to those who need more for an improved cure.

This rapidly improving understanding of this childhood cancer is a result of the collaborative nature of childhood cancer specialists. In this ubiquitous clinical research culture, specific therapies are rapidly evaluated and when successful incorporated into the standard of care. The rapidity of this improvement is in direct correlation with the percentage of children participating in clinical trials. In the United States, virtually all children with cancer are treated at institutions that participate in the National Cancer Institute-sponsored Children's Oncology Group (COG). Compare this to the 1-2 percent of adults treated on clinical trials. Studies have repeatedly shown that children receiving their care at one of the COG centers as opposed to other locations have significantly better outcomes, and those treated on clinical trials have an even better chance for cure. Clinical trials allow us to provide the best known care or a therapy that may be even better which may become tomorrow's best known care. One can easily see why the cure rates in children have improved more rapidly than in adults.

The timing of this report and its subject offers us the opportunity to highlight the 50th anniversary of the work of the National Cancer Institute's Cooperative Clinical Trial Group model. For children, this is represented by COG, merged in 1999 from the prior Pediatric Oncology Group (POG) and

Children's Cancer Study Group (CCSG). The COG is a multi-institutional organization that includes over 240 institutions that provide >99 percent of the care for childhood cancer patients in the United States, Canada, Australia, New Zealand, and parts of Europe. Virtually every childhood cancer specialist in the country is a participant in this enormous clinical trial organization. It is the largest such cooperative group in the world and dwarfs similar groups in other parts of the world who have modeled their organizations after the COG and its predecessors. It incorporates all the major university and cancer specialty hospitals in these countries and as a result provides infinitely more resources to the individual patient than any single center could alone. Research scientists and clinicians from these 240+ centers work jointly and collaboratively to advance the field of childhood cancer therapy and to improve the rates of cure in our patients. This organization brings together the laboratory research of the basic scientists, and the massive numbers of patients cared for by the clinicians, allowing the clinical and translational research investigators to offer the most advanced treatment for childhood cancer victims through their local or regional childhood cancer specialists wherever they live in this country. As the COG celebrates its 50th anniversary of work towards eliminating this disease as the major cause of non-traumatic death in children, one can truly say that the care one receives in Los Angeles, Houston, New York, Rochester, or Kansas City, is the same in all centers. With this availability, parents and their child may remain close to



home and within their network of support, which is unavailable to them when far from home.

Children's Mercy is the region's largest as well as one of the larger childhood cancer centers in the country, and as result of its multi-specialty and multi-disciplinary coordinated care system, outcomes match or exceed those seen nationally. Several national and international clinical trials are led by Children's Mercy physician scientists. As an academic and clinical center, it employs a large number of people, including research data managers, pharmacists, and nurses, whose sole jobs are the operation of these cutting edge clinical trials at our hospital. I invite the reader to visit our Web site, www.childrensmercy.org, where this is further illustrated and to visit the Web sites of the National Cancer Institute, www.cancer.gov, and the Children's Oncology Group, www.curesearch.org, to learn more about these cooperative efforts and the latest laboratory and clinical trials available to children with cancer. These trials, the majority available here at Children's Mercy, offer our children with cancer the latest advances of therapy while simultaneously providing the knowledge necessary to improve the care for children of the future.

Sincerely,

A handwritten signature in dark ink, appearing to read 'Alan Gamis'.

Alan Gamis, MD, MPH
Director of Oncology
Chair, CMH Cancer Center Committee

Dear Friends,

It is with pleasure that I share with you the 2005 Cancer Care Annual Report from Children's Mercy Hospitals and Clinics. This year's report includes a special focus on our work with neuroblastoma, but also will provide you with an overview of the many different clinical and psychosocial programs we provide for children with cancer and their families.

As a parent, I can't imagine how traumatic it must be to hear a physician tell you that your child has cancer. The staff of our Division of Hematology/Oncology is well aware of how that news can impact a family, and works closely with all members of the family from the moment the diagnosis is received, through treatment and follow-up and into remission. And, thanks to the cutting-edge research in which our staff participates and our sharing of research with other pediatric programs throughout the country, we are able to continue improving the outcomes and survival rates for children with all different types of cancer.

I hope you will take a few moments to review this annual report to learn more about our latest work fighting neuroblastoma and other cancers which affect our children and teens. All of us at Children's Mercy are extremely proud of the care provided by the Division of Hematology/Oncology and the strength and hope which our patients and their families share with us.

Sincerely,

A handwritten signature in black ink, reading "Randall L. O'Donnell". The signature is fluid and cursive, with the first name "Randall" being more prominent and the last name "O'Donnell" following in a similar style.

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



patrick



As a typical 6-year-old boy, Patrick loved playing t-ball and was always causing mischief. Halfway through t-ball season, however, Patrick began complaining about feeling sick and didn't want to play. At first his parents thought he had just lost interest in the game, but they soon found out that something more was wrong.

After a month of taking antibiotics prescribed by his doctor, Patrick became severely ill while away on family vacation. His parents rushed him home and called his doctor, who referred them to Children's Mercy Hospital. Patrick's parents brought him into the hospital on July 29, 2003 and that night he was diagnosed with neuroblastoma. The next day, Patrick had a biopsy to remove part of a tumor in his abdomen.

Over the course of the next couple months Patrick underwent four rounds of chemotherapy. Then, in October of 2004, he had a major operation to remove the remainder of the large tumor that had wrapped itself around his abdomen. After more chemotherapy and radiation, Patrick had a stem cell transplant in March of 2004. He then had another operation in June of 2004 to remove a tumor in his chest

After a long fight, Patrick is now a healthy 9-year-old boy. He still makes



routine visits to Children's Mercy every three to four months, but spends the rest of his time at school or playing sports.

Keep your eye on the ball Patrick!

division of hematology/oncology

Excellent comprehensive care for children with cancer and other blood disorders is the mission of members of the Division of Hematology/Oncology at Children's Mercy Hospitals and Clinics. Children and families served by the Division receive care both in the Hematology/Oncology Clinic and on 4 Henson, the inpatient floor. Each area is specially equipped to care for patients during each phase of their treatment. Additionally, caregivers are given specialized training that continues throughout their Hematology/Oncology career. Over 40 nurses within the Division currently hold Certified Pediatric Oncology Nurse certification.

The Division of Hematology/Oncology is led by Division Chief, Gerald Woods, MD. Dr. Woods also leads the Section of Hematology and Alan Gamis, MD, MPH is the Chief of the Section of Oncology. Sue Stamm, RN, CPNP is the Manager of the Division. The following is an overview of the Division and its services.





THE SECTION OF ONCOLOGY

SECTION CHIEF - ALAN GAMIS, MD

- **Oncology Service-Director-Alan Gamis, MD, MPH:** Six physicians provide care to patients diagnosed with oncology disorders. Patients are treated according to state-of-the-art protocols endorsed by leaders in children's cancer treatment.
- **Bone Marrow Transplant Service-Director-Charles Peters, MD:** Two dedicated transplant physicians provide hematopoietic stem cell rescue to patients receiving myelo-suppressive or ablative therapy which requires stem cell rescue. Stem cell rescue is accomplished by utilizing umbilical stem cells, matched-related donor cells, and matched-unrelated donor cells. Children's Mercy Hospital participates in the National Marrow Donor Program which facilitates unrelated donor searches for patients who have such a need.

THE SECTION OF HEMATOLOGY

SECTION CHIEF - GERALD WOODS, MD

- **Sickle Cell Disease Service-Service Director-Gerald Woods, MD:** The Sickle Cell service offers specialized care for the unique diagnostic and treatment needs of children and adolescents diagnosed with sickle cell disease. The patients and families are cared for during episodes of acute pain and receive on-going management for life limiting chronic pain, and other long term issues related to their disease.
- **Regional Hemophilia Center-Center Director-Brian Wicklund, MD:** The Center supports seven sites in a five-state region to provide comprehensive treatment for both adult and pediatric patients with bleeding disorders. Additionally, this service supports the diagnosis and treatment of children with coagulation and/or thrombosis disorders.

CHILDREN'S MERCY

CANCER CARE COMMITTEE

The Children's Mercy Hospital Cancer Care Committee is a standing medical staff committee that provides oversight of the oncology services. The committee is chaired by the section chief for the Section of Oncology and has both physician and non-physician representation (see member listing). At quarterly meetings the committee reviews patient acuity, program services and quality initiatives. They also determine quality improvement needs and opportunities related to pediatric cancer prevention, diagnosis, treatment, and outreach. The committee functions in accordance with the American College of Surgeons' accreditation standards.

PEDIATRIC HEMATOLOGY/ONCOLOGY FELLOWSHIP PROGRAM

The fellowship program at Children's Mercy Hospital trains the future pediatric hematology/oncology specialists. This program requires intense clinical and research training for a minimum of three years. Each fellow at Children's Mercy Hospital enters the fellowship program as a fully trained pediatrician. The first 18 months are dedicated primarily to clinical care and becoming familiar with procedures and standards of practice. The last half of the fellowship is focused upon clinical and laboratory research. The Division of Hematology/Oncology at Children's Mercy Hospital supports three fellowship positions.



PSYCHOSOCIAL SERVICES

- **Parent-to-Parent Program:** designed to connect parents who have been through the cancer experience with parents of a newly diagnosed child with cancer. In addition, this program offers support groups on a weekly basis facilitated by parent volunteers under the oversight of a social worker.
- **Social Work and Community Services:** there are five full-time social workers dedicated to the Division of Hematology/Oncology. The primary focus of this team is to assist the patient and the family in understanding the impact the diagnosis of cancer will have on their family dynamics and life style.
- **Bereavement Support Group/Program:** this program is offered to parents who have experienced the loss of a child. The program includes staff contacts to parents/family members during significant events such as birthdays, anniversary date for the death of the child, an annual memorial service, and the composition of a memory book.
- **Child Psychologist:** a full-time psychologist provides support to the patient, parents, and siblings of those diagnosed with childhood cancer.

- **Chaplaincy:** the hospital offers chaplaincy service 24 hours each day. Within the Division, a full-time chaplain supports the spiritual needs of the patient and their families.
- **Child Life Therapist/Music Therapist:** two full-time Child Life therapists, one full-time Child Life assistant and one part-time music therapist work with patients, parents, and siblings to aid in the adjustment and ongoing support associated with diagnosis and treatment of childhood cancer.

- **Cancer Outreach/School Re-entry Services Participants:** a team of Division members including nursing and social work work to build strong connections among the hospital, parents/child, and each child's school. School staff is provided with important information and are encouraged to ask questions. This effort allows for a better level of comfort/understanding while interfacing with the child and family. Additionally, school mates are given accurate information and are able to ask questions to relieve anxieties.



CANCER CARE CONFERENCES

The Children's Mercy Cancer Center conferences provide opportunities for a multidisciplinary discussion regarding newly diagnosed patients, recurrent/relapsed patients, and unusual pediatric cancer cases. Conference participants review patient history, physical findings at diagnosis, laboratory values, radiology exams, surgical interventions, and pathological findings. This diagnostic review is followed by a thorough review of the potential treatment options, current therapy available, response to therapy and prognosis. Medical specialists from oncology, surgery, pathology, radiology, radiation oncology, neurosurgery, orthopedic surgery, endocrine, psychology, and dentistry are represented. In addition, non-physician team members are encouraged to attend. Conferences are conducted in accordance with the American College of Surgeons' accreditation standards.



CASE MANAGEMENT

Each physician within the Hematology/Oncology Division works in close collaboration with an advanced practice nurse, physician assistant or nurse in expanded role. With the assistance of an assigned social worker, these individuals coordinate every aspect of the patient's care throughout treatment and during post-treatment follow-up.

OUTREACH AND PREVENTION

In addition to providing support for current patients, members of the Division also provide preventive education/outreach on issues such as smoking cessation and sun exposure. These programs are tailored to the appropriate age group being spoken to.

RESEARCH

Opportunities to participate in research are plentiful within the Division. Research studies in all areas are always open and recruiting new participants. All studies are reviewed by the Children's Mercy Institutional Review Board to ensure that the studies are safe, equitable and hold some form of benefit. Children's Oncology Group (COG) studies comprise the majority of studies available to oncology patients. These studies are created by a cooperative group of over 175 institutions. Member researchers continue to develop research trials that bring improved care and outcome to pediatric patients. In addition to COG studies, the Division participates in studies sponsored by the National Institutes of Health, Centers for Disease Control, pharmaceutical companies, and various other research leaders. A team of research coordinators and data managers facilitate patient enrollment and follow-up during study participation.

NUTRITION/PHARMACY SERVICES

- **Nutrition:** two full-time and one part-time registered dietician support the nutrition needs of the patient/family. In addition, there is a nutrition tech that adds support to the team.
- **Pharmacy:** two full-time pharmacists and one full time technician support the operations of a satellite pharmacy Monday -Friday, 7:00 a.m. to 7:30 p.m. This satellite supports the inpatient and outpatient areas in preparing and dispensing chemotherapy. In addition, one full-time pharmacist provides clinical consultation to the oncology service and one full-time pharmacist provides clinical support to the Bone Marrow Transplant Service.



petra

After finding what looked like a shadow on her sonogram, doctors gave 37 week pregnant Candice an hour to go home and pack a bag and return to the hospital to deliver her baby. On July 8, 2005 Petra was born through C-section and diagnosed with neuroblastoma.

The next morning a transport team arrived to bring Petra to Children's Mercy.

"I held her for hours before they came to get her," says Candice. "The team patiently waited until I could bear to give her up, then they took her and strapped her into a little five point harness. I felt like the Calvary had come and she was going to be fine."



Petra spent nearly a month in the NICU at Children's Mercy. A week after her arrival Petra had surgery to remove her tumor. She was then sent home in hopes that her immune system would take care of the remainder of the cancer.

However, a couple weeks before Christmas, Petra's pediatrician sent her back to Children's Mercy after her parents noticed bruises under her eyes and small bumps on her forehead. Tests showed that the cancer had moved to her skull, and Petra was instantly moved into a high risk category and started chemotherapy in February to combat the cancer.

After eight rounds of chemotherapy, tests showed that the cancer was gone from Petra's skull. However, an antibody test showed that there might be fragments left in her bone marrow, and there was still some cancer in her liver that was receding on its own.



Today Petra is a healthy 2 year old girl, and her body has continued fighting the cancer left in her liver. Petra has developed right on schedule with other kids her age, and is even ahead of other kids in some categories.

Young Petra has lived up to her name, translated from Greek to mean rock, and is definitely a force to be reckoned with.

Stay strong Petra!

cancer care committee board members

Medical Staff Members

Andrews, Walter, MD
Burleson, Amy, DDS
Cooley, Linda, MD
Covitz-Hardy, Lynne, PhD
Dalal, Jignesh, MD

Emami, Abbas, MD
Gamis, Alan, MD, MPH Chairman
Gyves-Ray, Katherine, MD
Hetherington, Maxine, MD Assistant Chairman
Holcomb, George W., MD
Hornig, Gregory, MD
Huseman, Carolyn, MD
Lewing, Karen, MD
Manalang, Michelle, MD
Massey, Vickie, MD
Modrcin, Ann, MD
Morello, Frank, MD
Nicklaus, Pam, MD
Peters, Charlie, MD

Sharma, Mukta, MD
Shore, Richard, MD
Snyder, Charles, MD
Woods, Gerald, MD
Zwick, David, MD

Surgery
Dentistry
Cytogenetics
Developmental Medicine
Hematology/Oncology
Stem Cell Transplant
Hematology/Oncology
Hematology/Oncology
Radiology
Hematology/Oncology
Surgery
Neurosurgery
Endocrinology
Hematology/Oncology
Hematology/Oncology
Radiation Oncology
Rehabilitation Medicine
Radiology
Otorhinolaryngology
Hematology/Oncology
Stem Cell Transplant
Hematology/Oncology Fellow
Hematology/Oncology
Surgery-ACOS Liaison
Hematology/Oncology
Pathology

Non-Physician Members

Baer, Phyllis, RN
Bartholomew, Joy, RN, FNP, CPON
Brown, Pat, MS, FT
Burns, Susan, RN, CPON
Corse, Cindy, CTR
Devorin, Barb, RN, CPNP
Fournier, Julie, RN, CPON
Green, Nancy, Sr. CNS
Haynes, Cindy, MDIV, BCC
Hamlin, Julie, APRN, BC
Hutto, CJ, RN, CPON
Jones, Ronald, PA-C
Klockau, Chris, RPh, BCOP
Laurence, Kris, CCRP
Marcus, Bette, RN, CPON
Mick, Kathy, RN, CPON
Ryan, Robin, CCRP
Seal, Annie, C.L.S.
Smith, Courtney, Pharm D
Stamm, Susan, RN, MSN, PNP
Stegenga, Kristin, RN, PCNS, CPON
Thompson, Mandy, Nurse Clinician
VanStone, Jill, RN MSN, PCNS
Walter, Christy, CCRP, LPN

Hematology/Oncology
Hematology/Oncology
Hematology/Oncology
Hematology/Oncology
Cancer Registry
Neurosurgery
Hematology/Oncology
Nutrition
Chaplaincy Services
Hematology/Oncology
Senior Director Allied Health
Hematology/Oncology
Pharmacy
Hematology/Oncology
Hematology/Oncology
Education
Hematology/Oncology
Child Life
BMT/Pharmacy
Hematology/Oncology
Hematology/Oncology
Hematology/Oncology
Hematology/Oncology
Hematology/Oncology



richard

After arriving at Children's Mercy, Richard was diagnosed with neuroblastoma on February 21, 2006. He had an operation two days later to remove his adrenal gland and the majority of the tumor in his liver. Richard then started his first of eight rounds of chemotherapy.

Today Richard is an energetic 16-month-old who still sports the trademark smile that never left his face. He still has a feeding tube, but eats most of his food through his mouth and is even getting close to walking!

Keep smiling Richard!

As a 4-month-old baby boy, Richard was described as a healthy kid who always had a smile on his face. While his smile never faded, Richard's health became an issue when his mother noticed his stomach steadily enlarging.

"I thought we were just feeding him too much so I asked his doctor about it when I took him to get his shots," says Amy, Richard's mother. "The doctor sent us straight to Children's Mercy."

During Richard's second round of chemotherapy he had to be placed on a ventilator because his swollen liver had caused his lungs to become compressed. Eventually, the swelling started to go down and Richard was taken off of the ventilator. Richard continued with his chemotherapy and didn't encounter any other major problems. He finished chemotherapy in August of 2006 and was soon sent home.



cancer registry

PURPOSE

Historical Mandate: The National Cancer Act in 1971 signed by President Richard Nixon, made the “conquest of cancer a national crusade” by mobilizing the country’s resources to fight cancer. In October of 1992, congress enacted Public Law 102-515 which established the National Program of Cancer Registries. This law authorized the Centers for Disease Control and Prevention (CDC) to enhance centralized cancer registry operations in all states.

The Children’s Mercy Hospital (CMH) submissions: Confidential, protected data is reported to the Missouri State Central Registry. CMH also maintains accreditation approval from the American College of Surgeons Commission on Cancer; which requires data be submitted to the National Cancer Data Base. With this data, public health professionals are enabled to understand and address the cancer burden more effectively.

CMH Registry Goal: The ultimate purpose of the cancer registry data is to establish cancer statistics. Reporting includes incidence, mortality and pattern identifying features, in order to assist research for fighting cancer.

PROCEDURES

Quality and Supervision: The Cancer Registry operates under the guidance of the Cancer Care Committee. Quality of data is monitored by the committee with timely reports and audit reviews.

Operations: The Registrar captures a complete summary of the history of illness, diagnosis, treatment, and disease status for every cancer patient that is diagnosed or treated with a malignancy at CMH and clinics. Certain benign / borderline conditions are also collected as approved by the Cancer Care Committee. The Cancer Registry also conducts yearly follow-up contacts of all patients in the registry until age 27.

2005 REGISTRY STATISTICS

Numbers: During 2005, 152 new cases were added to the registry. During the time period from January 1, 1990 until December 31, 2005 data has been collected on 1,536 patients.

Age and Sex at Diagnosis: There were 16 patients less than 1 year of age diagnosed in 2005. 75 patients were in the 1-10 age group and 61 patients were greater than 10 years of age. During 2005 there were 83 males and 69 females diagnosed.

Histological Implications: Leukemia was the leading diagnosis in 2005 representing 28 percent of the annual caseload. This was the first year since 2002 that the leukemia diagnosis outnumbered the tumors of the central nervous system. The five most frequently occurring diagnoses for 2005 were; leukemia, CNS tumors, lymphoma, neuroblastoma and osteosarcoma. (see figure 1 and 3)

Analytic Patients: During 2005 there were 132 analytic patients. The analytic patients are those diagnosed here or elsewhere, but received all or part of the first course of treatment at CMH. The analytic patients are those eligible for inclusion in the registry’s statistical reports of treatment efficacy and survival.

Mortality: There were 22 cancer-related deaths reported during 2005, 6 of these patients were also diagnosed in 2005. Tumors of the central nervous system represented 32 percent of these mortalities. (see figure 2)



**CHILDREN'S MERCY HOSPITAL
CANCER REGISTRY**
2005 FREQUENCY OF DIAGNOSIS
FIGURE 1

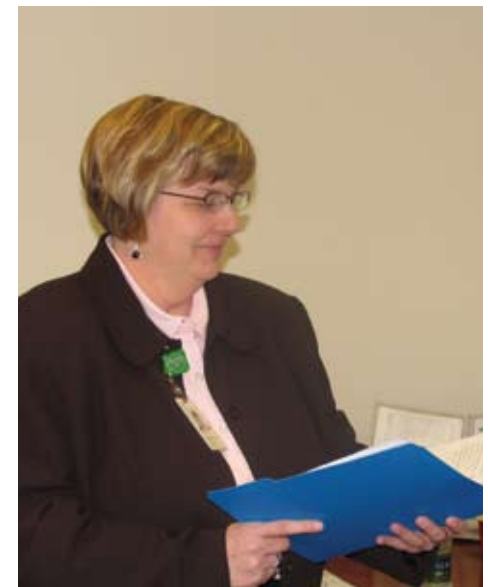
Central Nervous System	37	(24%)
Astrocytoma	7	
Glioma	7	
Medulloblastoma	3	
Ependymoma	1	
Glioblastoma Multiforme	1	
Germinoma	1	
Mixed Germ Cell of CNS	1	
Oligodendroglioma	1	
Choroid Plexus Carcinoma	1	
Benign/Borderline Tumors	14	
Leukemia	42	(28%)
ALL	33	
AML	6	
Juvenile myelomonocytic	1	
Acute promyelocytic	1	
Chronic myelogenous	1	
Lymphoma	15	(10%)
Non-Hodgkins	6	
Hodgkins	9	
Neuroblastoma	7	(5%)
Osteosarcoma	7	(5%)
Wilms Tumor	5	(3%)
Rhabdomyosarcoma	4	(3%)
Ewings Sarcoma (EFT)	2	(1%)
Other	33	(21%)
Congenital Fibrosarcoma	1	
Embryonal Carcinoma	1	
Lymphoproliferative Dis.	1	
Malignant Rhabdoid Tumor	1	
Malignant Teratoma	1	
Melanoma	1	
Mixed Germ Cell	2	
Myelodysplastic syndrome	2	
Retinoblastoma	1	
Squamous Cell Carcinoma	1	
Thyroid Carcinoma	3	
Yolk Sac Tumor	2	
Misc. Reportable conditions	16	

*The Cancer Care Committee has approved the following conditions to be included in the registry. These conditions are included in the registry due to location of tumor or because of its propensity to recur.

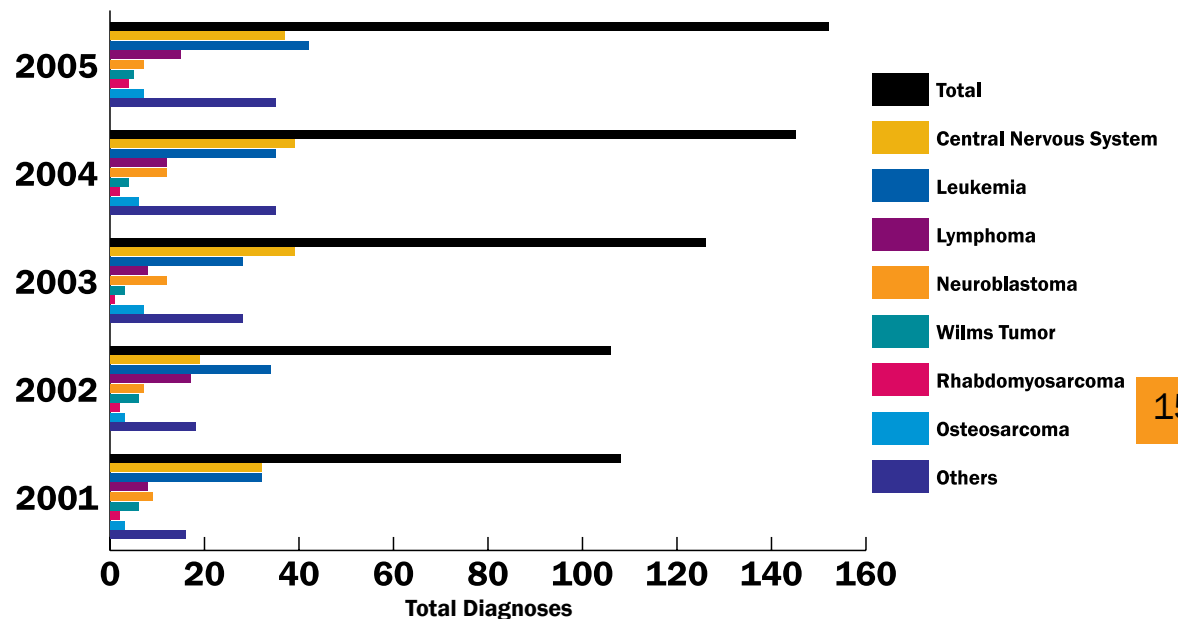
Mesoblastic nephroma
Teratomas, regardless of locations
Theca cell-granulose cell tumors
Lymphoproliferative Disease
Langerhan's Cell Histiocytosis
Hemangioendothelioma

2005 MORTALITY DATA
FIGURE 2

Central Nervous System	7
Atypical teratoid/rhabdoid	1
Choroid Plexus Carcinoma	1
Ependymoma	3
Glioblastoma Multiforme	1
Glioma	1
Leukemia	3
Wilms Tumor	2
Osteosarcoma	2
Germ Cell	1
Hepatoblastoma	1
Hodgkin Disease	1
Lymphoproliferative Disorder	1
Malignant Rhabdoid of Kidney	1
Neuroblastoma	1
Rhabdomyosarcoma	1
Undifferentiated Sarcoma	1



**A FIVE-YEAR COMPARISON OF MOST FREQUENTLY OCCURRING DIAGNOSES
AT CHILDREN'S MERCY HOSPITAL**
FIGURE 3





santana

Today Santana is a healthy 8-year-old who is full of energy. The only noticeable effects of her treatment are the hearing aids she wears and a scar from her surgery. Santana doesn't let her fight with cancer hold her back, and the compassion she shows to her horses is a reflection of her love for life and others.

Eight-year-old Santana is a vibrant young girl who enjoys participating in Girl Scouts and loves all animals, especially the miniature horses that her family raises. Today no one would expect that Santana had a long fight with cancer, but the doctors at Children's Mercy know that this little girl is a fighter!

When Santana was 18 months old her mother took her to the doctor because she had flu like symptoms, her stomach was slightly enlarged, and her lymph nodes were hard. A CAT scan found neuroblastoma, and the doctor immediately sent Santana to Children's Mercy.

Santana was admitted into Children's Mercy Hospital on Jan 10, 2000 and started chemotherapy a couple days later. After six rounds of chemotherapy Santana's tumor had decreased in size by 50 percent, and on April 24, 2000 she had surgery to remove the remainder of it.

Santana then received additional chemotherapy treatment and had a stem cell transplant on July 25, 2000. After her transplant Santana received radiation and was cleared in April of 2001.



Ride on Santana!

neuroblastoma

INTRODUCTION

Neuroblastoma (**neuro** indicates nerves, **blastoma** refers to a cancer that affects immature or developing cells) is a tumor of cells of neural crest origin that form the adrenal medulla and sympathetic ganglia. Tumors can appear anywhere along this chain from the base of the neck to the tailbone. It is the second most common solid tumor in childhood after brain tumors and accounts for approximately 8 percent of all childhood cancers. Each year about 600 children in the United States will develop neuroblastoma. The cause, like most childhood cancers is unknown. This is truly a disease of childhood as greater than 50 percent present before two years of age and it is extremely rare in adolescents and young adults. Neuroblastoma accounts for 50 percent of all tumors in infants less than one year of age. Not all autonomic nervous system tumors are malignant. There is a benign tumor called ganglioneuroma and a mixed tumor composed of benign and malignant cells called ganglioneuroblastoma. There is also a very unique form of malignant neuroblastoma, called Stage 4S (see Page 19), that is known to spontaneously regress without any therapy.

CLINICAL PRESENTATION

Most neuroblastomas (about 2/3) start in the abdomen with about 1/3 in the adrenal glands (located above the kidneys).

The rest arise primarily in the chest and neck. Metastatic spread typically involves the lymph nodes, liver, bone, bone marrow, and skin. Symptoms are typically a result of either tumor invasion or of para-neoplastic syndromes. First symptoms are often vague and may include fatigue and loss of appetite. A tumor in the abdomen may cause a swollen stomach and constipation. By contrast, infants with extensive disease are often pale and cachectic, presenting with fever, anorexia, irritability, and anemia (resulting from spread to bone marrow and bone). A tumor in the chest may cause breathing problems and suggest pneumonia as a result of bronchial obstruction. Tumors pressing on the spinal cord may cause weakness and an unstable gait, limp, or even paresis (weakness) along with bladder and/or anal sphincter dysfunction.

It is one of the few cancers in children that release hormones (catecholamines) that can cause changes in the body such as constant diarrhea. Other paraneoplastic syndromes can cause changes in some functions of the brain causing **opsoclonus** (rotary movements of the eyes) and **myoclonus** (spastic jerks of the muscles), both thought to be a result of antibody formation. Besides **“Dancing Eyes”**, one may see a **Horner’s syndrome** (sinking in of the eyeball, ptosis of the upper eyelid, slight elevation of the lower lid, constriction of the pupil, narrowing of the palpebral fissure and anhidrosis (a result of nerve compression of the upper chest or neck). As well **“Raccoon Eyes”** (periorbital edema and ecchymosis

with proptosis) is a classic finding, the result of tumor invasion into the bones of the orbits and sinuses. Skeletal involvement can present as a hard lump, pain, or refusal to bear weight. Skin involvement can present with hemorrhagic skin nodules (**Blueberry Muffin** lesions). Excessive sweating is often seen related to excessive secretion of catecholamines.

WORK UP

In addition to a detailed history and physical exam a variety of laboratory and radiological exams are performed along with pathological studies on tissue(s) obtained. These tests allow us to properly stage the degree of spread of the tumor as well as provide important markers, which are essential for treatment and prognostic reasons, as well as to monitor the affects of therapy.

LABORATORY STUDIES

One of the characteristics of Neuroblastoma is excessive production of catecholamines: **Vanillylmadelic Acid (VMA)**, and **Homovanillic Acid (HVA)**. Elevation of one or both of these can often be detected in a random (spot) sample of **urine**. **Ferritin, LDH, and Neuron-specific enolase** can also be found to be elevated in some neuroblastomas and can be measured from a **blood** sample. Anemia and thrombocytopenia are common findings due to marrow invasion.

IMAGING STUDIES

Ultra Sound uses sound waves whose echoes produce a picture. This is a relatively quick, non-invasive procedure that is often first obtained when concern arises about the possibility of the presence of a neuroblastoma, especially in the abdomen. Once a mass is felt to be present, other imaging studies are routinely done.

Computerized Axial Tomography (CAT) scan of the neck, chest, abdomen and pelvis is important to evaluate for metastatic disease as well as to better visualize the primary tumor. Evaluation of the head is also often done to better visualize boney invasion.

A **Total Body Bone Scan** involves an intravenous injection of a radioactive tracer (technetium 99). If the tumor has spread to the bones this dye travels to, and shows increased uptake in, areas of tumor spread.

A **Meta-Iodobenzylguanidine (MIBG)** scan also involves an intravenous injection. MIBG attaches to neuroblastoma cells and can indicate involvement of both organs as well as bones. Not all tumors take up MIBG, but in those that do this becomes an additional tool to monitor tumor response to treatment. In some instances (usually with resistant or recurrent disease), therapy takes advantage of MIBG sensitive tumors.

TISSUE EXAMINATION

A **Bone Marrow Aspirate and Biopsy** are performed from both the left and right posterior iliac crests (the back of the pelvis) to determine whether or not the tumor has spread to this organ. This procedure can be done with local anesthetic agents such as lidocaine and/or a numbing cream along with moderate sedation (Midazolam and Fentanyl) or deep sedation (propofol). This is done in the hospital floor or clinic procedure room along with a representative from Child Life and the option of the presence of family members. When necessary, this procedure is done in the Operating Suite with the child completely asleep using general anesthesia. This may coincide when the surgeon is removing or biopsying the primary tumor and/or providing intravenous access with the placement of a central line.

Surgery is required for both diagnostic and therapeutic reasons. If possible, it is preferable to remove all or as much of the primary tumor as possible. If this can't be done (especially at the risk of damaging vital organs or vessels) a biopsy to obtain

tissue is performed. In addition, in order to facilitate the delivery of chemotherapy in a safe and easy fashion as well as providing a means of monitoring therapy, the surgeons will place a central venous line (often at the same time, thus limiting the child to only one anesthetic interaction). Because of the need for two lumens and the small body size as well as the possibility of bone marrow transplantation, this is usually an externalized catheter, but indwelling ports are sometimes used.



STAGING

Staging is based upon the spread of tumor and the surgical resection at diagnosis. Today, the International Neuroblastoma Staging System is used (INSS) which is outlined as follows:

Stage 1: Localized tumor that is completely surgically removed at diagnosis.

Stage 2A: Localized tumor that cannot be completely surgically removed at diagnosis. It is on one side of the body. Lymph nodes on the same side of the tumor may have tumor cells present. Lymph nodes outside of the tumor are free of cancer.

Stage 2B: Localized tumor that may or may not be able to be totally removed by surgery. It is on one side of the body. Nearby lymph nodes outside the tumor contain neuroblastoma cells, but the cancer has not spread to lymph nodes on the other side of the body or elsewhere.

Stage 3: Large tumor that has spread across the middle of the body that cannot be surgically removed at diagnosis.

Stage 4: Tumor of any size that has metastasized (spread) to distant lymph nodes, bone marrow, bone or liver.

Stage 4S: Special stage that applies only to a child less than one year old. Small, localized tumor that has metastasized (spread) to liver, skin, and/or bone marrow, and specifically does not spread to the bone.

Neuroblastoma that presents as Stage 4S is a unique tumor that is known to spontaneously regress during the first year of life without therapy. Occasionally short periods of therapy are required to minimize local damage until the time that spontaneous regression begins. Despite its identical microscopic appearance to other

Children’s Mercy Neuroblastoma
Gender and Stage Distribution
10 Year Comparison

YEAR	TOTAL	MALE	FEMALE	STAGE 1	STAGE 2	STAGE 3	STAGE 4	STAGE 4S
2005	7	2	5	2	2	0	2	1
2004	13	7	6	3	3	2	5	0
2003	11	7	4	0	3	3	5	0
2002	7	3	4	1	0	0	6	0
2001	9	4	5	1	1	1	6	0
2000	4	0	4	0	0	0	4	0
1999	6	4	2	1	0	2	2	1
1998	5	3	2	2	0	0	2	1
1997	2	0	2	0	1	0	1	0
1996	8	3	5	1	3	0	4	0
TOTALS	72	33	39	11	13	8	37	3

types of neuroblastoma, this propensity to spontaneously regress (even in the face of its metastatic spread) requires its special staging designation.

PROGNOSTIC FACTORS:

Besides the stage, there are four features that have prognostic value and help determine the appropriate therapy. These are:

1) Age: Younger children (less than 18 months) are more likely to be cured than older children.

2) Tumor histology: Originally described by Dr. Shimada, the appearance of the tumor cells under the microscope are determined to be either “Favorable” or “Unfavorable” histology depending on how active the cells look. Tumors with unfavorable histology tend to behave more aggressively.

3) Tumor MYCN status: MYCN is an oncogene that regulates tumor cell growth. Tumor cells can have a **single copy** or multiple copies of this gene (called MYCN **amplification**). Tumors with MYCN amplification tend to behave more aggressively.

4) Tumor DNA index (also called Ploidy): Describes the number of chromosomes in the tumor cells, compared to the number in normal cells. Cells with about the same amount of DNA as normal cells are classified as “**Diploid**”. Neuroblastoma cells with increased amounts of DNA are termed “**Hyperdiploid**”. In infants, hyperdiploid cells tend to be associated with earlier stages of disease, respond better to chemotherapy, and usually predict a more favorable prognosis than diploid cells.

RISK GROUPS

Based on the stage along with the prognostic factors listed above, a patient is placed into one of three risk groups: **Low, Intermediate, and High**. Based on past studies, 12 months of age is used as an important cut off point, but based on review of the most recent results, it appears that in future studies this cutoff point will extend to become 18 months.

Low Risk:

- All Stage 1
- Stage 2A or 2B who are < 12 months old
- Stage 2A or 2B who are > 12 months old and have a single copy of the MYCN gene
- Stage 2A or 2B who are > 12 months old who have extra copies of MYCN gene but have favorable histology
- Any Stage 4S who have a single copy of MYCN, favorable histology and hyperdiploid

Intermediate Risk:

- Stage 3 who are < 12 months old and have a single copy of the MYCN gene
- Stage 3 who are > 12 months old and have a single copy of the MYCN and favorable histology
- Stage 4 who are < 12 months old and have a single copy of the MYCN gene
- Stage 4S who are < 12 months old and have a single copy of the MYCN gene, and either normal DNA Ploidy or has unfavorable histology

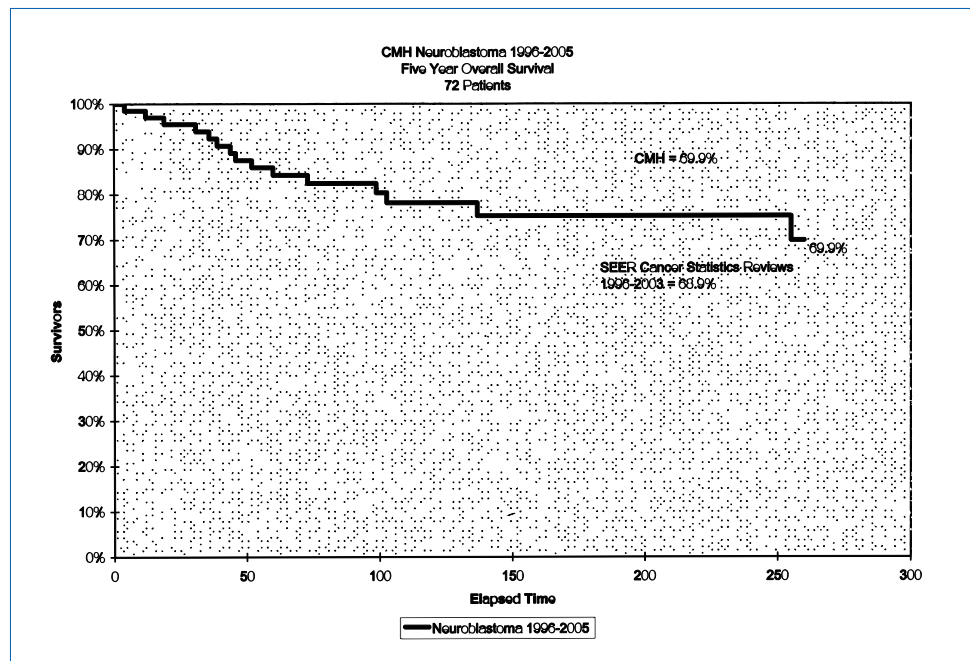
High Risk:

- Stage 2A or 2B who are > 12 months old and have extra copies of the MYCN gene and unfavorable histology
- Stage 3 who are < 12 months old and have extra copies of the MYCN gene
- Stage 3 who are > 12 months old and have a single copy of the MYCN gene and has Unfavorable histology
- Stage 3 who are > 12 months old and have extra copies of the MYCN gene
- Stage 4 who are < 12 Months old and have extra copies of the MYCN gene
- Stage 4 who are > 12 months old
- Stage 4S who are < 12 months old and have extra copies of the MYCN gene

RISK ADJUSTED TREATMENT

Therapy is adjusted according to one of three risk categories. This risk reflects the risk of the neuroblastoma recurring.

Low Risk patients are treated with **surgical resection and supportive care**. If the tumor recurs or progresses than once again the treatment of choice would be surgery and supportive care. If the tumor is not resectable, than chemotherapy agents are used. Patients will receive four (4) cycles (a group of chemotherapy agents given together over a short period of time) of chemotherapy given every three weeks. Those who recur or progress with



Intermediate Risk disease receive four (4) cycles if they have favorable histology or eight (8) cycles if they have unfavorable histology.

Intermediate Risk patients are treated with a combination of **surgery and chemotherapy**. Patients first undergo surgery to remove as much of the primary tumor and involved lymph nodes as can be safely accomplished with the minimum morbidity. Patients then receive four (4) cycles of chemotherapy if they have favorable histology and eight (8) cycles of chemotherapy if they have unfavorable histology. If the tumor had not been removed previously, then surgical removal of residual tumor is done at the completion of chemotherapy.

Radiation therapy is not routinely given to intermediate risk patients. It is used to treat in the following situations:

- Progressive clinical deterioration despite chemotherapy and/or surgery
- Partial Response (50-90 percent Reduction) after eight (8) cycles of chemotherapy in patients with unfavorable histology
- Partial Response following surgery for local recurrence more than three months after completing protocol therapy

High Risk patients are treated with a combination of therapies. As will be discussed, the prognosis for these patients is considerably less favorable than those in the Low or Intermediate Risk groups. As



with the other patients, surgery is usually the first step in treatment. Again, the goal is to remove as much of the tumor as possible without causing significant morbidity. If there is metastatic disease present, usually no attempt is made to approach this surgically. Chemotherapy is then employed.

Patients receive two (2) cycles of chemotherapy and then have their own **Peripheral Blood Stem Cells (PBSC)** collected and stored for later in their therapy. Following this, the patients receive three (3) more cycles of chemotherapy and then have their definitive surgery to remove any residual tumor (if any exists). There is then one (1) additional cycle of chemotherapy, which is then followed by the **Bone Marrow Transplant (BMT)** portion of the treatment. At this point the patient receives additional high dose chemotherapy, which is different from what the patient has seen previously. An important side effect from this therapy

is that it causes the patient's bone marrow to be ablated (destroyed), thus requiring the immediate subsequent infusion of the patient's PBSC's collected earlier to ensure recovery.

Following recovery from this process patients undergo radiation therapy to the primary tumor bed, and any metastatic areas of disease that were shown to be involved with tumor just prior to the

transplant process.

Patients then proceed to a phase where they take an oral **biologic agent, 13-cis-retinoic acid (accutane)**. Neuroblastoma cells have shown that when they are exposed to this agent, they exhibit decreased proliferation, decreased expression of MYCN, and morphologic differentiation.

Other therapies that have been used to treat these patients include the use of agents bound to MIBG and other antibodies specific to a patient's tumor. MIBG associated therapies remain limited to very few institutions. Antibody therapy is under investigation at all participating Children's Oncology Group (COG) centers including Children's Mercy.



TREATMENT RELATED SIDE EFFECTS

The following medications are the backbone of chemotherapy:

- Cyclophosphamide
- Doxorubicin
- Vincristine
- Cisplatin
- Etoposide

While effective in treating neuroblastoma these medications also can have significant side effects that affect the patient in both the short and long run.

Nausea and vomiting, hair loss, depression of the immune system, and suppression of the bone marrow are common. Infection can be life threatening and often there are hospitalizations in-between cycles of chemotherapy to treat presumed and true infections. Appetite can be suppressed to the point that weight loss requires supplementation either through their implanted central line as Total Parental Nutrition (TPN), or enterally through a Naso-Gastric (NG) tube which is a flexible tube placed through the nasal cavity down into the gastro-intestinal (GI) tract or having a “button” or gastrostomy tube (G-tube) surgically placed with a valve providing direct access to the GI tract via the stomach.

In addition, each chemotherapy agent may have some unique side effects due to the drug itself or from a break down product that occurs as the body metabolizes it.

- **Vincristine** can cause a decrease in reflexes and inhibit proprioception (the sensory nerve endings that give information concerning movements and position of the body) causing messier drawing and handwriting and increased clumsiness (especially when running). More problematic is the constipation that is caused in a population where this is already often a significant problem.
- **Cyclophosphamide** may produce bladder inflammation and blood in the urine (hemorrhagic cystitis) and/or damage to the kidneys with subsequent loss of salt and minerals in the urine. It is not uncommon for patients to need supplementation of these nutrients during and after therapy is concluded.
- **Cisplatin** may also cause some kidney damage as well as hearing loss or deafness. It is not unusual for children to require hearing aids during or after therapy is completed.
- **Doxorubicin** may cause heart damage and can cause a chemical burn to the skin if it leaks out and comes in contact.

TREATMENT OUTCOME

There is a considerable difference in outcome for the Low and Intermediate Risk patients compared to the High Risk patients. Patients with Low Risk disease have shown long term survival rates of greater than 90

percent while Intermediate Risk patients have shown long term survival rates close to the same 90 percent.

This outcome however is not what has been shown in the High Risk patients. On the positive side, there has been considerable improvement over the many years of treating these patients. From 1978 through 1985 long-term survival for this group was only about 10 percent. This rose to about 25 percent from 1986-1995. With current treatments, we have again seen improvement, but still less than desirable at only around 40-45 percent.

Through clinical studies, we have learned that those children treated with 13-cis-retinoic acid do better than those that did not receive this as part of their therapy. In addition we learned that children that underwent autologous bone marrow transplantation did better than those that only were treated with chemotherapy alone.

We also learned that screening for neuroblastoma at an earlier age and hopefully at an earlier stage resulting in an improved outcome did not pan out as was hoped. While the number of children diagnosed with neuroblastoma increased, the rate for High Risk children did not decrease as hoped. What did occur was that many children who previously would have gone undetected (due to spontaneous regression and without any consequence) were now being identified in the newborn period. Meanwhile, many parents underwent an increased incidence of anxiety and worry. It seems that while there is much similar

between these tumors, those that cause High Risk disease truly act in a different manner.

OTHER STRATEGIES AND FUTURE DIRECTIONS

As we succeed with treatment, the goal begins to switch towards decreasing treatment morbidity both in the short term and especially in the long term. While reducing therapy intensity further is not that practical in the Low or Intermediate Risk groups one likely change in the next round of therapies will be to change the prognostic cutoff age from its current 12 months of age to 18 months of age. When this was looked at with the most recent data it appears that several hundred children could be treated successfully while exposing them to significantly less therapy.

As mentioned earlier, because neuroblastoma cells tend to be radiation sensitive, there have been several therapies looking at attaching **radionuclides** to MIBG, somatostatin analogs, or anti-GD2 antibodies. These treatments are still investigational, are not readily available at most institutions, and are not active in all patients.

Neuroblastomas are often heavily vascularized tumors and the degree of vascularity often correlates with biologic features such as NMYC amplification. Agents that work against the formation of new blood vessels, **antiangiogenic agents** are being looked at but have been slow to

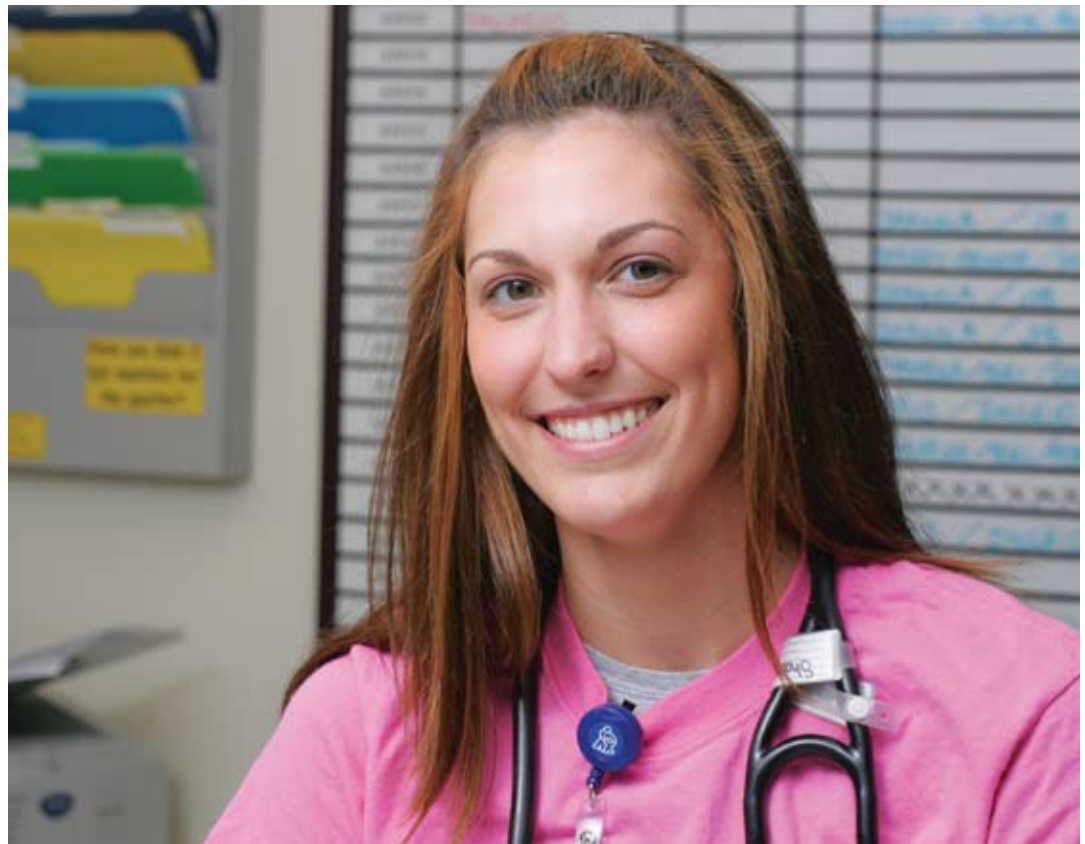
come to clinical trials.

In the laboratory, inhibiting Tyrosine Kinase activity has shown to inhibit the growth of neuroblastoma cells. Others such as the EGF receptor inhibitor gefitinib are also being looked at.

In addition, other strategies like looking at ways to prevent drug resistance are in investigation.

For now, the most likely changes will be

to explore newer agents most likely coming from experience in treating relapsed and resistant disease that has shown to be effective and moving them into frontline therapies. Ongoing participation in clinical trials will be essential to bring the most current therapies available to these patients as well as allowing us to continue to make much needed progress in treating this disease.





satala

Here at Children's Mercy we know that the children we serve today will be the leaders of tomorrow. That was no exception in the case of 3-year-old Satala.

In 2006 Satala's mother took him to see the family doctor because he had been complaining about his legs hurting for three or four months. Originally the doctor supported the mother's suspicion they were just growing pains, but took an x-ray just to be sure.

After looking at Satala's x-ray, his doctor immediately referred him to Children's Mercy where he was diagnosed, on Jan. 27, 2006, with

neuroblastoma. Young Satala started his eight month treatment on chemotherapy right away, and later received a bone marrow transplant on August 17, 2006.

After his bone marrow transplant Satala was cancer free, but continued coming to Children's Mercy two or three weeks a month to participate in an antibodies study to ensure that he remained healthy.

Today Satala is almost 5 years old. His mother refers to him as "an old soul," and the doctors at Children's Mercy can be proud in knowing that they saved the life of a very intelligent young child.



Satala loves computers and is developed way beyond his years. He currently speaks both English and Spanish, and is learning Chinese from his aunt. Satala is definitely a child with a bright future and we hope to see great things from him.

Buena Suerte Satala!

diagnostic and treatment support services

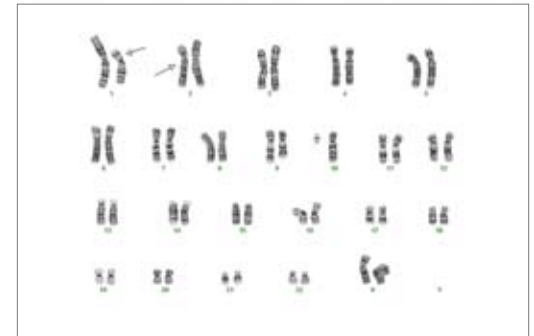
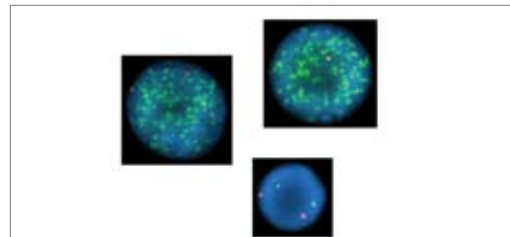
SURGERY

The pediatric surgeon is often involved in the diagnosis and treatment of neuroblastoma, as well as operative staging of the disease. More than half of NB arise in the abdominal cavity. Tumor size, location, degree of vascular and visceral involvement is assessed, and resectability determined. Although generally it is true that complete resection offers the best hope for a long term cure, biologically more favorable tumors are more likely to be amenable to complete resection. Sacrifice of major organ / vessels at attempted initial resection is not necessary. Minimally invasive surgery play only a minor role in the treatment of NB. Some centers have performed laparoscopic biopsy as an initial approach for unresectable disease.

A second look operation after adjuvant therapy is preferable to life-threatening resection initially. Operative therapy for neuroblastoma is therefore stage-based: INSS (International Neuroblastoma Staging System) Stage 1 and early Stage 2 tumors should be completely resected, with an expected survival in the 85 - 90 percent range. Advanced stage tumors undergo chemo and or radiation therapy after the diagnosis is confirmed, with delayed primary or second-look operation. Post-operative complications occur in about 10 percent of patients, dependent on the stage of the disease.

CYTOGENETICS

Neuroblastoma (NB), the most common extracranial solid tumor in children, is derived from primitive neural crest cells. NBs arise in the adrenal gland, the sympathetic nerve ganglia, and other sites. Cytogenetic anomalies distinguish prognostic groups in neuroblastoma (NB). Chromosome aberrations that correlate with a poor outcome are deletion of the chromosome 1 short arm, gain of copies of chromosome 17 long arm, MYCN gene amplification, and a diploid or near-diploid karyotype. These aberrations are closely associated with advanced stage disease. However, 1p deletions in stage I, II, and IVS tumors identify patients with high-risk disease in the absence of MYCN gene amplification. A hyperdiploid or near-triploid karyotype without the aforementioned anomalies is prognostically favorable. MYCN gene amplification manifests as double minute chromosomes (dmin) or homogeneously stained regions (hsr) at the cytogenetic level. Fluorescence in situ hybridization (FISH) analysis using touch preparations of tumor material or cytogenetically harvested cells provides a quick assessment of MYCN gene status.



The karyotype:

- 45,XX,der(1;17)(q10;q10),der(2)add(p16)del(q11.2q24.2)inv(q11.2p16),-10,+17,dmin
- Shows an unbalanced translocation between chromosomes 1 and 17 that result in loss of 1p and gain of 17q
- Shows an unbalanced rearrangement of chromosome 2 that results in loss of portions of chromosome 2 short and long arms
- Shows loss of one copy of chromosome 10
- Shows two normal copies of chromosome 17 in addition to the chromosome 17q translocated to chromosome 1; the result is 3 copies of 17q
- Shows gain of innumerable double minute chromosomes

The metaphase cell from which the karyotype was made:

- Shows the double minute chromosomes as small dots surrounding the chromosomes

The FISH of interphase nuclei:

- Shows the double minute chromosomes hybridized with the MYCN gene probe (green); two red signals show the normal copy number of chromosome 2; nucleus with two red and two green signals is a normal cell



PATHOLOGY

Children's Mercy Hospital provides a multidisciplinary team approach to the diagnosis and treatment of Neuroblastoma (NB). This team includes a pediatric pathologist that confirm or establish the diagnosis of NB, and cytogeneticist who evaluates the chromosomes and certain genes in the tumor. The pathologist's primary function is to distinguish NB from other malignant tumors that share a so-called "small round cell" microscopic appearance common to many childhood malignancies including leukemia, lymphomas, and several childhood sarcomas. A variety of monoclonal antibodies are now utilized to assist in making a reliable distinction. In addition, tumor samples are submitted for a variety of ancillary studies including determining tumor histologic grade, DNA content (Ploidy), and the number of copies of the N-Myc gene that bear on the prognosis of the patient and alter treatment accordingly.

The Pathology Laboratory processes thousands of specimens each year in the diagnosis of new and recurrent tumors. The department performs a variety of tests directly used in the diagnosis and follow-up care of cancer patients. Children's Mercy Laboratory department is staffed 24 hours each day, seven days a week and is licensed by the federal government (CLIA) and accredited by the College of American Pathologists (CAP), the Joint Commission on the Accreditation of Health Care

Organizations (JCAHO), as well as state and federal agencies.

RADIOLOGY

The mission of the Department of Radiology is to provide state-of-the-art medical imaging services in a safe and caring environment to patients, families, and referring physicians. Services are tailored to meet the patient's medical needs while treating each child as a unique individual. The Department of Radiology provides computerized tomography (CT) including the latest development in multi-detector CT, Magnetic Resonance Imaging (MRI), ultrasound, nuclear medicine, conventional and fluoroscopic studies, as well as, leading edge interventional radiology procedures. The department is staffed by fellowship trained, board certified pediatric radiologists and a dedicated group of technologists, nurses, and clerical staff. Our mission statement is taken very seriously and guides our daily practice.

One of the key medical imaging studies in the staging and follow-up evaluation of patients diagnosed with neuroblastoma is the Nuclear Medicine MIBG scan with SPECT. SPECT stands for Single Photon Emission Computed Tomography, and allows for thin section tomograms to be taken in multiple planes. This enables detection of small lesions with a high target to background ratio. MIBG also helps in distinguishing viable tumor tissue

from residual fibrosis which is a valuable prognostic indicator in patients who are undergoing or are post treatment.

RADIATION THERAPY

Radiation therapy was used following surgery to treat residual neuroblastoma in the 1950s. The use of chemotherapy changed the management of the disease since that time, and the techniques, doses and volumes of radiation therapy have continued to evolve with time. Often, the patients that receive radiation have had a stem cell transplant. That is an important consideration in determining the regions that will be treated as well as the doses that will be delivered. As our imaging studies become more sophisticated, we have modified the areas that are treated in patients. The daily doses and total doses are also being evaluated, with an emphasis on customized and tailored treatments for patients, depending on their response to treatment. The Kansas City Cancer Center has partnered with the Division of Hematology/Oncology to ensure each pediatric patient diagnosed with neuroblastoma is offered the most effective treatment option available.

references

- London, W.B., R.P. Castleberry, K.K. Matthay et al. Evidence for an Age Cutoff Greater Than 365 Days for Neuroblastoma Risk Group Stratification in the Children's Oncology Group. *J Clin Oncol* 2005 23:6459-6465.
- Rani E, George, Wendy B. London, Susan L. Cohn, John M. Maris et al. Hyperdiploidy Plus Nonamplified MYCN Confers a Favorable Prognosis in children 12 to 18 Months Old With Disseminated Neuroblastoma: A Pediatric Oncology Group Study. *J Clin Oncol* 2005 23:6466-6473.
- Schmidt, Mary Lou, Ashutosh Lal, Robert C. Seeger et al. Favorable Prognosis for Patients 12 to 18 Months of Age With Stage 4 Nonamplified MYCN Neuroblastoma: A Children's Cancer Group Study. *J Clin Oncol* 2005 23:6474-6480.
- Schnidt, Mary Lou, John N. Lukens, Robert C. Seeger et al. Biologic Factors Determine Prognosis in Infants With Stage IV Neuroblastoma: A Prospective Children's Cancer Group Study. *J Clin Oncol* 2000 18:1260-1268.
- Brodeur, Garrett M. and Maris, John M. Neuroblastoma In: Principles and Practices of Pediatric Oncology. 5th Edition. Pizzo, Phillip A and Poplack, David G. (Eds), Lippincott, Williams and Wilkins, Philadelphia 2005.
- Neuroblastoma In: Atlas of Pediatric Oncology Sinniah, D and D'Angio, Giulio J (Eds) Arnold 1996.
- Seger, RC, Brodeur GM, Sather H et al. Association of multiple copies of the N-Myc oncogene with rapid progression of neuroblastomas. *N Engl J Med* 313:1111-1116, 1985.
- Shimada H, Chatten J, Newton WA et al. Histopathologic prognostic factors in neuroblastic tumors: definition of subtypes of ganglioneuroblastoma and an age linked classification of neuroblastomas. *JNCI* 73:405-416, 1984.
- Shende, Ashok Neuroblastoma in Manual of Pediatric Hematology and Oncology. 3rd Edition. Lanzkowsky, Philip editor. Academic Press 1999.



Children's Mercy Hospitals and Clinics

2401 Gillham Road
Kansas City, Missouri 64108
816-234-3000
www.childrensmercy.org