

2006 CANCER CARE ANNUAL REPORT

Focus on Tumors of the
Central Nervous System



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

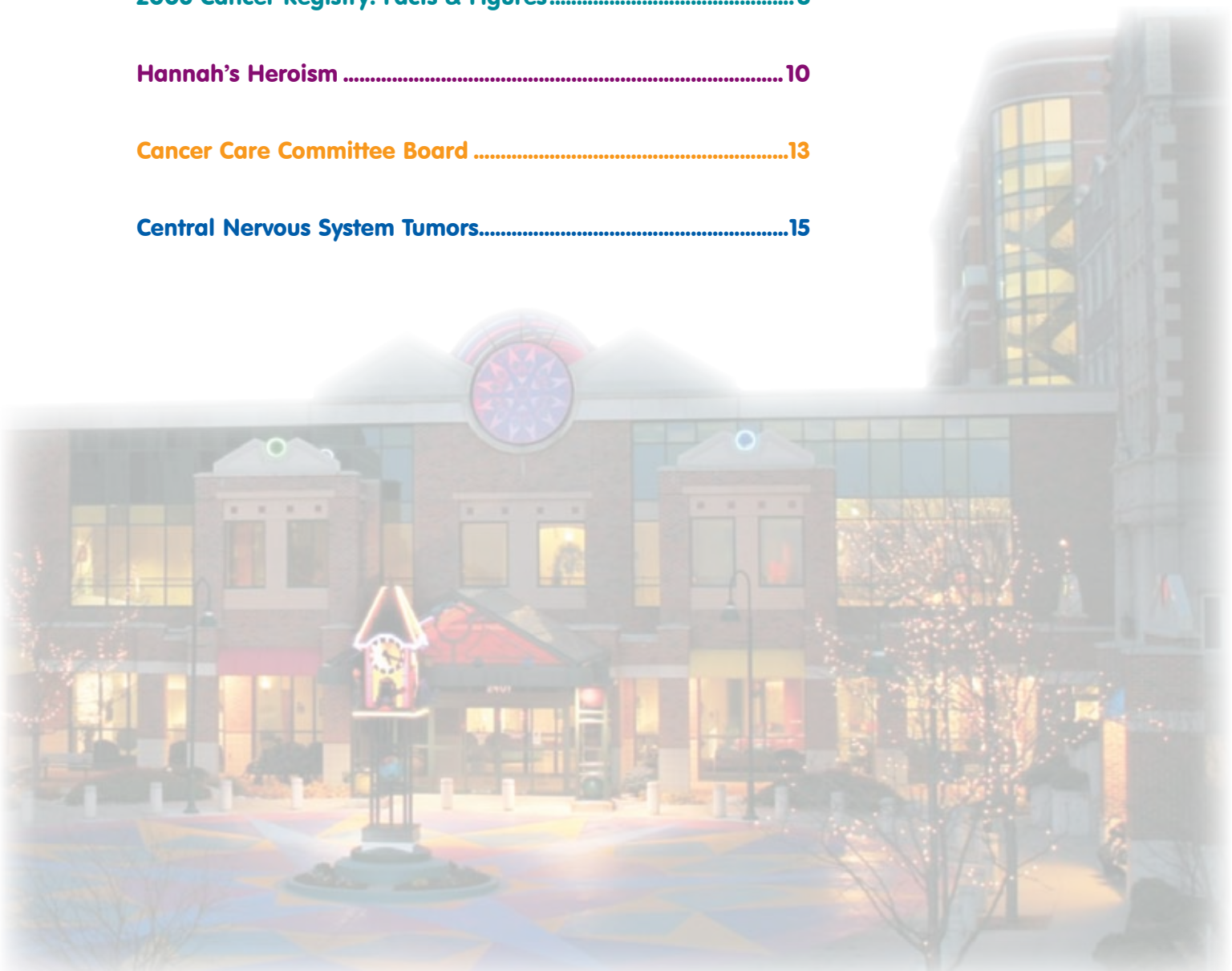
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Dear Friends,

I am pleased to share with you our 2006 Cancer Care Annual Report, which provides a comprehensive overview of the outstanding pediatric oncology program offered at Children's Mercy Hospitals and Clinics.

This year's issue includes a special focus on tumors of the central nervous system and the ground-breaking work that Children's Mercy is doing in this area.

This has been a particularly stellar year for our oncology program, as 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. I offer my heartfelt congratulations and appreciation to Drs. Woods and Gamis, and the entire Hematology/Oncology staff for this notable achievement.

I hope you will take a few minutes to enjoy this year's annual report and find out more about why our oncology program has received such well-deserved national recognition. We certainly enjoy receiving these types of awards. However, the greatest reward our oncology staff receives throughout the year is the ability to provide both cutting-edge and supportive care to ensure bright futures for hundreds of children with cancer and their families.



Sincerely,

A handwritten signature in black ink that reads "Randall L. O'Donnell". The signature is written in a cursive, flowing style.

Randall L. O'Donnell, PhD
President and Chief Executive Officer
Children's Mercy Hospitals and Clinics

Dear Friends,

This year's Children's Mercy Hospitals and Clinics Cancer Center Annual Report focuses upon the second most common tumor in childhood, Brain Tumors. Of all the tumors seen in childhood, these are among the most complex due to their location, their significant impact upon the child even before diagnosis, and the limitations placed upon therapy options due to the young age of most patients at the time of diagnosis. This latter item, age at diagnosis, plays a significant role in the ultimate outcome of these children, both as it relates to cure as well as to quality of life after cure. The average age of children diagnosed with a brain tumor is eight years, with 25 percent diagnosed <3 yrs of age, 27 percent between 3-8 yrs of age, and 48 percent >8 yrs of age. The child's brain is going through tremendous development and maturation during the years prior to age 3. Even up to age 8 there is significant change. Damage, either from the tumor itself, surgery needed to remove the tumor, radiation therapy, or chemotherapy, can arrest any further development or worse yet, cause loss of development or function. Thus, therapy is greatly altered in the youngest ages, moderately so in the middle childhood ages, and minimally so in those older than age 8. Due to this, these patients require a significant array of services – from diagnosis through the complex time of therapy, and throughout the years that follow – that are pediatric-specific. Children, based on their tolerance, go through an intense battery of age-specific tests prior to therapy. These tests are repeated continuously throughout and after therapy. For

these to be beneficial, pediatric-trained professionals are critically important. These professionals must maintain their expertise, both through continual education and via large populations of children with these disorders. The Neuro-Oncology team of pediatric professionals at Children's Mercy includes oncologists, neurosurgeons, radiation therapists, neurologists, rehabilitation medicine, neuropsychology, endocrinologists, ophthalmologists, pathologists, and radiologists. Couple these physicians with pediatric-specific facilities, dedicated pediatric oncology units and clinics and pediatric trained caregivers including Oncology dedicated nurses, advanced nurse specialists, pharmacists, dieticians, social workers, child life specialists, and research professionals, and you get a comprehensive program second to none in the region.

Children's Mercy Hospital is one of the larger Childhood Cancer Centers, not only in the region, but also in the country with over 150 children diagnosed annually. Nearly one fourth of these children are diagnosed with a brain tumor. The large volume of children seen with the tumors leads to an expertise that is rarely found elsewhere. With this volume, comes an efficient organization with excellent coordination of care under the auspices of case managers who are highly trained and specialty specific advance nurse practitioners. Each child has their own APN/PA case manager who works intimately with their oncologist to oversee and coordinate all aspects the child's care and the family's support. Be sure to

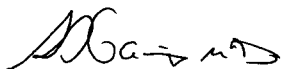
learn more about us, not only on our Web site, www.childrensmercy.org, but also as a featured hospital on the Children's Oncology Group's Web site, www.curesearch.org.

This year we take great pride in announcing that Children's Mercy is the recipient of this past year's American College of Surgeon's Commission on Cancer's Outstanding Achievement Award, given to the top six percent of all Cancer Centers in the country. Our center submits itself to on-site inspections/surveys every



three years by this commission to ensure that our continuing goal of providing the best possible care for our children is not just words, but an achieved and measurable reality. This survey identified that among all critical areas of care and oversight, our center achieved outstanding marks, not just in the year of the survey, but in all three reviewed years of activity. This award carries even more significance as Children's Mercy was the only children's hospital in the United States to be awarded this honor. Our belief that we provide

a level of care to our patients that we would want for our own children, and that this level of care is among the best in the country, has been substantiated by this award. It has been, and remains our mission to continue this high level of care for our current and future patients. I hope this year's Annual Report provides you a glimpse of this as we review our program for the care of children with Brain Tumors.



Alan S Gamis, MD, MPH

Chief, Section of Oncology
Division of Hematology/
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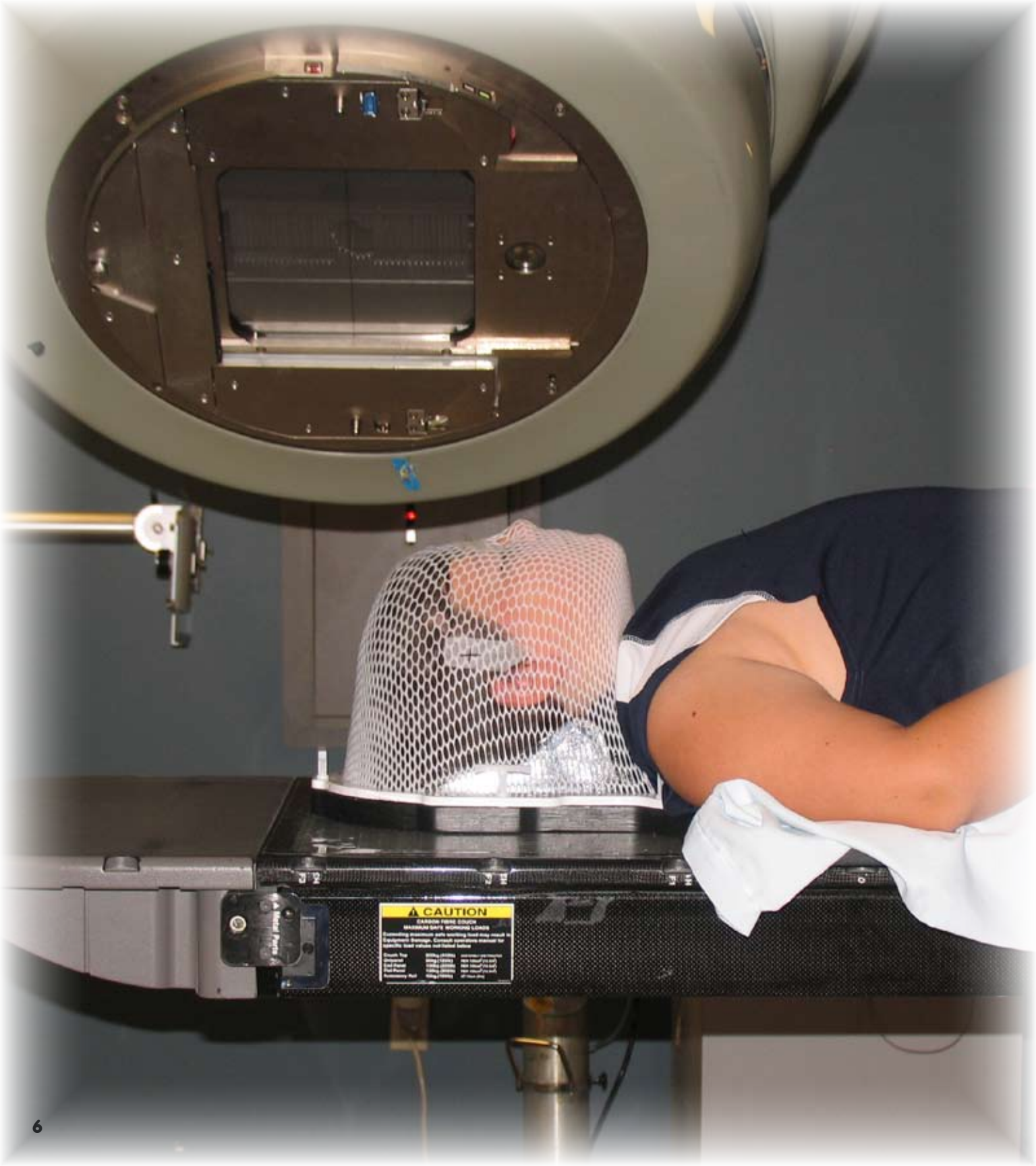
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CAUTION
CARBON FIBRE Couch
Maximum static bending warning
Excessive pressure or flexing load may result in permanent damage. Consult the manual for specific load ratings and labels below.

| Couch Top | Weight Capacity | Maximum Pressure |
|-----------|-----------------|------------------|
| Couch Pad | 150kg (330lb) | 100kPa (14.5psi) |
| Couch Pad | 150kg (330lb) | 100kPa (14.5psi) |
| Couch Pad | 150kg (330lb) | 100kPa (14.5psi) |

The Division of Hematology/ Oncology

Division Chief

Gerald Woods, MD

Division Manager

Sue Stamm, RN, MSN, CPNP

Children seen in the Division of Hematology/Oncology are cared for in the outpatient clinic and on the dedicated Hematology/Oncology Unit in the Henson Inpatient tower.

The Section of Oncology

Section Chief-Alan Gamis, MD

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Director-Alan Gamis, MD

Bone Marrow Transplant Service

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The Section of Hematology

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Regional Hemophilia Center

Center Director-Brian Wicklund, MD

Children's Mercy is the region's largest childhood cancer center averaging nearly 150 new oncology diagnoses each year. In addition, Children's Mercy is the only free-standing hospital made just for kids from St. Louis to Denver treating children and adolescents from birth to 21 years of age.

Among the services offered by the Division are:

Case Management - Guided by a nurse case manager, usually an advanced practice nurse teamed with an oncologist, each child's case is overseen from diagnosis through follow-up care. The Division of Hematology/Oncology recognizes that each child's and family's needs are unique. Consequently, each child's care is designed and coordinated specifically for them. The care plan is based on his/her medical and supportive needs and their developmental level.

Pediatric Hematology/Oncology Fellowship Program - The Division of Hematology/Oncology at Children's Mercy Hospital supports six fellowship positions. Fellows complete a rigorous three year clinical and research training during their time at Children's Mercy.

Cancer Care Conferences - Weekly conferences, attended by a multi-disciplinary team, are held to discuss and deliberate patient diagnosis and care.

Children's Mercy Cancer Care Committee (CMCCC) - The CMCCC provides program oversight to the oncology services. The committee functions in accordance with the American College of Surgeons' accreditation standards. Quality improvement in all areas of patient care is a goal of the committee.



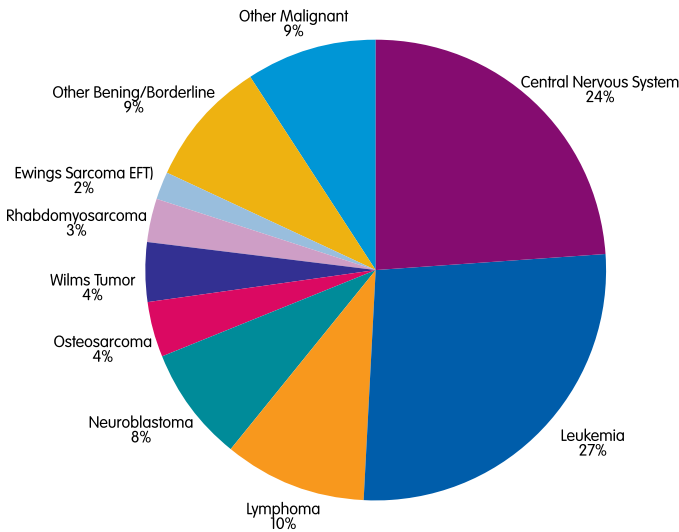
Matthew was diagnosed with a craniopharyngioma in 2006. Since then, this 9-year-old hasn't let his brain tumor slow him down. He takes his MRI in stride while listening to his favorite artist, Weird Al Yankovic.

The Cancer Registry at Children’s Mercy is a vital component of the Cancer Program. Under guidance from federal law, data is collected and maintained on all patients with malignancies and certain benign tumors. During 2006, 135 new cases were added to the cancer registry at Children’s Mercy Hospital. Eligible patients accessioned in the database now total 1,671. Twenty-four percent of the 2006 cases were diagnoses of the Central Nervous System, which is the medical focus of this year’s report

Highlights

The general distribution by diagnostic category is best shown in the pie chart in Figure 1. The top five diagnoses continue to include leukemia, brain tumors, lymphomas, neuroblastoma and osteosarcoma. Leukemia led the year with 27 percent of the caseload. (see Table I)

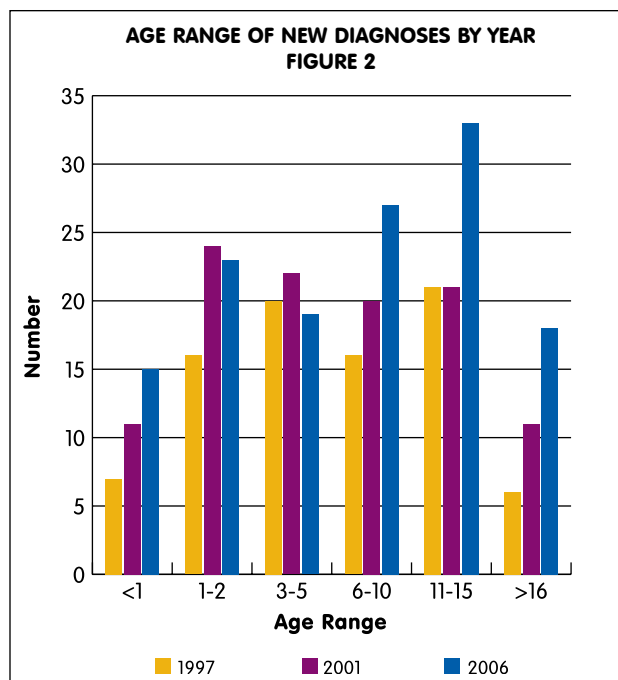
**2006 DIAGNOSIS FREQUENCY
FIGURE 1**



**CANCER REGISTRY
2006 FREQUENCY OF DIAGNOSIS
TABLE I**

| Diagnosis | Totals | Percentage |
|-------------------------------|-----------|------------|
| Central Nervous System | 33 | 24% |
| Astrocytoma | 12 | |
| Glioma | 5 | |
| Medulloblastoma | 3 | |
| Ependymoma | 1 | |
| N.G. Germ Cell | 3 | |
| Benign/Borderline | 9 | |
| Leukemia | 37 | 27% |
| ALL | 32 | |
| AML | 2 | |
| Other | 3 | |
| Lymphoma | 14 | 10% |
| Non-Hodgkins | 6 | |
| Hodgkins | 8 | |
| Neuroblastoma | 11 | 8% |
| Osteosarcoma | 5 | 4% |
| Wilms Tumor | 5 | 4% |
| Rhabdomyosarcoma | 4 | 3% |
| Ewings Sarcoma (EFT) | 2 | 2% |
| Other | 24 | 18% |
| Carcinomas | 5 | |
| Germ Cell Tumors | 2 | |
| Malignant Rhabdoid Tumor | 1 | |
| Melanoma | 2 | |
| Myelodysplastic Syndrome | 1 | |
| Retinoblastoma | 1 | |
| Misc. Reportable Conditions | 12 | |

Age and Sex at Diagnosis: The median age of patients diagnosed during 2006 was 7 years. The age distribution is shown in Figure 2 with comparison to past years. The male/female ratio was $73/62 = 1.18$ for 2006.



Race and Ethnicity: Race distribution during 2006 included 115 Caucasians, 18 African Americans and 2 others. Within the 115 Caucasian numbers, 14 patients were designated “Spanish/Hispanic” ethnicity.

Mortality: There were 24 cancer-related deaths during 2006. Six of these patients were newly diagnosed during 2006. Eleven of the patients had a diagnosis of leukemia and five deaths were related to brain tumors.

Analytic Patients: During 2006 there were 122 analytic cases. The analytic patient grouping is included in the statistical analysis of the database and meet reporting status. Patients diagnosed elsewhere but treated at Children’s Mercy made up 22 percent of the cases in 2006. Reportable by agreement cases are those benign or borderline disease processes which the Cancer Care Committee has decided should

be collected because of local interest. Please see Table II for further breakdown of patient groupings.

Active follow-up continues on 1,468 patients. The follow-up is maintained on analytic patients, including benign borderline CNS cases diagnosed after 1/1/2004. (see Table II)

By following our patients we contribute to medical surveillance and make improving patient outcomes possible.

2006 Survey of Program: The American College of Surgeons’ Commission on Cancer was initiated in the 1930s. The intent of the program was the establishment of benchmarks of high standards and systematic program reviews. Today, these standards continue to promote and

2006 ACCESSIONED CASES
TABLE II

| Class of Case | Definition | 2006 |
|-------------------------|--|------|
| (Analytic) | | |
| 0 | Diagnosed at CMH /Treated Elsewhere | 1 |
| 1 | Diagnosed/Treated at CMH | 94 |
| 2 | Diagnosed Elsewhere/Treated at CMH | 27 |
| (Non-Analytic) | | |
| 3 | Diagnosed/Treated Elsewhere (Treated at CMH for recurrence or disease progression) | 1 |
| Reportable by Agreement | Inclusion due to location of tumor or because of propensity to recur. | 12 |

support multidisciplinary care and improvements to overall quality. The Children’s Mercy Hospital Cancer Program has been approved by the Commission on Cancer since 1981.

In August of 2006 the Commission on Cancer surveyed Children’s Mercy Hospital. Children’s Mercy Hospital received the Approval Award with a three year commendation. Our program also achieved the Outstanding Achievement Award. This honor was awarded to only seven percent of surveyed programs.

Hannah's Heroism



On a typical school day, 17-year-old Hannah realized she could no longer read the projection screen at the front of the classroom. For a girl who had always had perfect vision, this was troubling. When her vision was still blurry the next day, her mother, Tina, brought her to an eye doctor. After being told by the eye doctor and a family care physician that nothing appeared wrong, Tina and Hannah were not satisfied.

"I had to cover one of my eyes so I could see straight," Hannah said. "I couldn't drive or read or do anything. I was at work with my hand over one eye!"

Obviously, something was wrong. Tina brought Hannah to another doctor, who performed an MRI. This physician referred Tina and Hannah to an oncologist at Children's Mercy. Before the visit, Hannah's parents wanted her to have a last worry-free weekend. "They wouldn't tell me what an oncologist was. They just said brain doctor."

That first visit was difficult. "I had always thought to myself, 'I have three healthy children,'" Tina said. "That day, I thought, 'Now I have two. We don't belong here.' I wanted to run."

Dr. Richard Shore told Hannah that it was, in fact, a brain

tumor. "My parents were on the floor, crying." Hannah recalls. "But I was kind of relieved because I knew what it was now and we just needed to get rid of it."

After that, things happened quickly. Hannah was in surgery two days after she was diagnosed. The inch-and-a-half tumor was in the center of her brain and could not be surgically removed. She woke up to her first 3-day chemo treatment. The experience was rather overwhelming for Hannah: "Two weeks later I just lay in bed. It was weird. I couldn't go to school anymore. I was just lying in my bed and I had cancer. Who would have thought that?"

On a **typical** school day, 17-year-old **Hannah** realized she could no longer read the projection screen at the front of the classroom.

After another two weeks, her long, brown hair started falling out.

"She couldn't comb it," Tina said. "She kept it pulled up in a pony tail because it was just falling out left and right."



Dr. Richard Shore takes a look at Hannah during a recent visit.

Although she had been advised against it, Hannah decided to shave it off and wear a wig. After her head was shaved, "I looked like a boy with makeup on!" she exclaimed.

The road to recovery was long and difficult with 18 weeks of chemo, 30 days of radiation and visits to the clinic between. They lived more than an hour away and had to come in four times each week.

Hannah recalled one particularly rough spot: "I had to lie on the couch because I felt like I was going to pass out if I lifted my head. My dad was freaking out. He had to carry me through the hospital's parking lot. I was there for three days and then a week later had to go back for chemo."

"I knew I was going to be okay from the beginning. The hardest part was just knowing everyone thinks you're

going to die and you don't feel that way."

In the midst of these trials were some bright spots.

Hannah was put on steroids to decrease the pressure in her brain. "Within the first week of steroids, my vision was back. I could see. I was reading. It was just really good. I knew if I could see my tumor must have shrunk," Hannah said.

Dr. Shore has been Hannah's attending physician. "He won't give you false hope. He was down to earth. He wasn't fake with me," Hannah said. "My parents were going crazy. They took my license, took my car. He just put me in a room and talked to me. He told me to milk it at Christmas because next year I'm not getting this!"

Hannah's two younger brothers wanted nothing at Christmas that year but for Hannah to get better. Their wish came true. The day after Christmas, the family found out that Hannah was cancer free.

Tina recalled several moments that stood out to her during Hannah's illness. "These are the things that I share with Hannah that make my heart melt and put a lump in my throat," she said.

Tina became a little teary during their first visit back to the clinic. "I remember looking around, seeing parents looking no different than how I had felt – tired and emotionally worn out. I looked at all the beautiful children playing and getting ready for chemo. My heart was just breaking for the children. When I looked at Hannah I was teary eyed and she said, 'Mom, don't cry. We are all chosen and blessed.' My child is truly amazing."

During one office visit, Hannah saw a boy she had seen before but who was now missing a leg. A few hours later, the radiation doctor told her that it would be longer than she had expected for her hair to grow back. Hannah cried until she remembered the boy. Then she told Tina, "It's okay, Mom. It's only hair." Tina said, "I am a very lucky mom to have such a strong and heartwarming child."

Hannah lost 30 pounds during her illness. Her parents were so concerned about how much weight she was losing that they made a

CHILDREN'S ONCOLOGY GROUP

Hannah was part of a trial study conducted by the Children's Oncology Group. The COG tries to answer the question: Can there be a better outcome? Hannah participated in a study that is looking at how chemotherapy given before the primary treatment (radiation) may decrease the time or dose of radiation needed. This is important because radiation to the brain can cause loss of memory or concentration. Hannah will be monitored periodically by the COG for ten years.

bargain with her. If she ate, they would buy her a brand new car. It worked.

"I started eating. I wanted a car!" she said. "May came, school was out and I went to the dealership with my parents. We got out and there was my car. A brand new 2007 Monte Carlo with balloons all over it and my dad said 'get in there.' I started crying. It was great."

Hannah is now a senior in high school and looking forward to attending college. She is also enjoying her new hair. "That hair was dyed. I get new hair, and it's really soft."

Hannah sums up her experience at Children's Mercy Hospital: "I don't think I could have gotten the same care anywhere else."



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Stewart walked on his first birthday. By 15 months, he was saying several words. His mother also began noticing him holding onto the wall as he walked. A few weeks later his walking seemed unsteady and he would keep heading to the right.

Emily was a good student and loved school. She especially liked to sit up front near the teacher. One morning when she woke up to get ready for school, she had a terrible headache and vomited. This continued for two weeks.

Roger was a very active young boy. He loved every sport and he was good at all of them. Roger was never sick, but one day on the soccer field he suddenly began seizing.



12-year-old Lane uses some of his energy on the playground in front of Children's Mercy and pauses to pose with his favorite neurosurgeon, Dr. Clarence Greene, Jr. Dr. Greene operated on Lane when he was diagnosed with an astrocytoma.

Joel was a moody child. His could change from sweet to angry quite suddenly. When his grades at school began to decline and he was seeming more aggressive, his teacher thought that he might have ADHD. After an evaluation he was started on medication, but several weeks later his anger seemed to be more intense and he fought more and more.

The next line in each of these stories is the same: A scan of the head was done and it showed that the cause of the problem was a brain tumor.

Of the many cancers that affect children, almost one third are tumors located in the central nervous system (CNS). The central nervous system is the part of the nervous system that includes the brain and the spinal cord. Sadly, in the US approximately sixteen hundred children are diagnosed each year with a brain tumor. In children, signs and symptoms of a CNS tumor may vary greatly in both intensity and length. It is not uncommon for there to be a very prolonged period of time (often months) between the onset of symptoms and the discovery of a brain tumor. Often the child has seen one or more health care providers along the way.

At Children' Mercy, children with CNS tumors are cared for through cooperative services of specialists from many different areas of the hospital. Included are the oncologist, but also surgeons, neurosurgeons, radiologists, radiation oncologists, physical and occupational therapists, nutritionists, pharmacist, psychologists, researchers, neurologists, nurses, social workers, child life therapists and clergy.

In this report, you will meet some of the real Emilys, Joels, Rogers and Stewarts. You will also learn about the many specialists here at Children's Mercy Hospital who are here to provide all aspects of care needed when the words, "Your child has a brain tumor" are uttered.

Making the Diagnosis

Radiology

The Department of Radiology is a vital link in the care of children with cancer of the central nervous system. The mission of the members of the radiology department is to provide the best possible diagnostic tools that will enable the Oncology and Neurosurgical teams to successfully treat these children. Currently, this is done by utilizing two state of the art 1.5 Tesla MRI units and 3 multi-slice CT scanners. These scanners are integral in brain and spine imaging.

Typically, a child with a tumor of the brain will be diagnosed after an initial MRI of the brain and spine. These pictures localize and characterize the tumor. They also can show if the tumor has spread. Once diagnosed with a tumor, additional advanced MR imaging may be performed. This includes diffusion-weighted imaging

and MR spectroscopy that allow for pre-operative characterization of the tumor, and better surgical planning. Radiology can also create images which guide the surgeon in the operating room.

In 2008 the hospital will open a new, state of the art MRI center. This center will house new 3 Tesla and 1.5 Tesla magnets. These units will include all available advanced imaging techniques including white matter tract mapping and functional localization of eloquent cortex, to further advance effective and safe surgical and radiosurgical planning.

The Department of Radiology commits itself to obtaining and utilizing the best technology for the care of children with central nervous system tumors. Their work is a vital link in the process of successful therapy.



Neurosurgical Oncology

Treating a brain tumor often starts with the neurosurgeon. Many of the children come to the hospital after being sick for weeks or months. Unlike in the adult population, brain tumors in children grow in the midline part of the brain. Problems in this part of the brain produce symptoms that could have several explanations.

The CMH Neurosurgery team includes experienced pediatric nurse practitioners, operating room nurses, RNFA's (surgical assistants), surgical ward RN's, and a dedicated outpatient clinic nurse team. All these individuals work as a unit to

make the children well again. They also strive to meet the emotional

and educational needs of the entire family. They understand that the course of treatment for a brain tumor and its associated problems is often a prolonged and arduous journey.

As tumors grow, more and harsher symptoms occur. One of these is hydrocephalus ("water on the brain"). Hydrocephalus creates increased pressure within the head (ICP). When this happens, the neurosurgeon is frequently called upon to intervene. The surgeon drains the CSF to relieve the ICP and stabilize the child. This important step is done before any thought can be given to the tumor mass itself.

The neurosurgical operating suite at Children's Mercy Hospital is outfitted with state-of-the-art equipment that aids the surgeon in attacking these problems.

A computer driven NeuroNavigational tool allows the brain imaging (MRI/CT) to be synchronized with the child's topographic anatomy. The surgeon can plan an approach that minimizes surgical exposure and risk to the normal brain.

Advancements in neurosurgery have done much to decrease the recovery time post-surgery. Miniaturized neuroendoscopic devices allow the surgeon to often biopsy tumors with only nickel-sized openings in the head. They can also often treat hydrocephalus without leaving in a drainage tube. Direct electric recordings (electrocorticography) on the surface of the brain can guide the removal of tumor-affected brain tissue. When this tissue is not removed it can cause epilepsy even after the tumor mass is removed.

Pathology

When a child with a CNS tumor has surgery, the pathologist uses the surgical material to make a diagnosis. Pediatric pathologists have extensive training that includes several years of specialization after completing medical school and experience in dealing with pediatric conditions that are relatively rare in the population.

Because current medical knowledge and technology offer many choices of treatment, clinical decisions in oncology are based on accurate information about a particular

disease. The pathologist provides critical information by examining the tissue, blood or tumor resections

in the laboratory. The pathologist's diagnosis provides information necessary to plan optimal treatment. Details from the pathologist

can also estimate the prognosis or possible outcome of a particular disease.

The pathologist has the opportunity to discuss and present his/her findings at clinical multidisciplinary conferences that involve professionals from different specialties. These conferences reflect the state of the art approach of malignant neoplasms. At the same time they promote collaboration and team effort in dealing with oncological challenges.

The pathologist's diagnoses are like road signals indicating the way to increase the possibilities of success in caring for children with cancer. The pathologists are happy to be part of a team that makes a positive contribution to children in need of their professional expertise.

The pathologist's diagnoses are like road signals indicating the way to increase the possibilities of success in caring for children with cancer...





A. Karyotype of a choroid plexus papilloma (CPP):
52<2n>,XX,+1,+8,+9,+12,+14,+20

B. Karyotype of a choroid plexus carcinoma (CPC):
32<1n>,XY,+1,+2,-4,+7,+add(12p),add(13p),+add(14q),-16,+add(18p),add(21q),+4mar

The CP papilloma can be distinguished from the malignant CP carcinoma by karyotype. The CP papilloma is commonly hyperdiploid (>46 chromosomes) while the CP carcinoma is often hyperhaploid (>23 chromosomes)

Cytogenetics

While central nervous system (CNS) tumors are the most common category of solid tumor in childhood, there are many different pathologic types. The cytogenetic laboratory investigates each child's tumor for the presence of genetic anomalies. Fresh tumor tissue from surgery is grown by the laboratory in tissue culture. Chromosomes, the genetic material, are obtained from the tumor cells and then analyzed under the microscope for abnormalities.

The chromosomes of brain tumors often show abnormalities. The abnormalities are often unique to a particular kind of brain tumor. By finding the genetic signature of a tumor cell, determining the diagnosis may be helped. It may also provide important information to the oncologist for therapy selection and family counseling.

In addition to conventional chromosome analysis, the cytogenetic laboratory often uses molecular cytogenetics or fluorescence in situ hybridization to detect genetic aberrations. This method utilizes

additional information with prognostic significance. New information regarding the genetics of brain tumors is always becoming available. The latest information is used in the care of the patient with a brain or CNS tumor.

Neurology

Children with central nervous system (CNS) tumors present with various signs and symptoms—ranging from seizures to motor-skill dysfunction to sensory losses to behavioral changes to headache. Many of these complaints are initially evaluated by child neurologists. Their workup can lead to neuroimaging (such as MRI scans) followed by the subsequent tests that make the accurate and specific diagnosis. At Children's Mercy Hospital, the Section of Child Neurology works closely with the neuro-oncology team to serve children with a range of tumors within the CNS.

The mission of the entire Section of Child Neurology is to improve the lives of children with all neurological disorders. This is achieved through the integration of evidence based



Our cover cutie, 10-year-old James (JT), looks at a scan of his brain with Dr. Craig Morriss. JT's parents listen as Dr. Morriss explains more from the scan. JT was diagnosed with an astrocytoma in 2000.

DNA probes to search for specific abnormalities. If present, a specific anomaly may further define the disease process and may provide

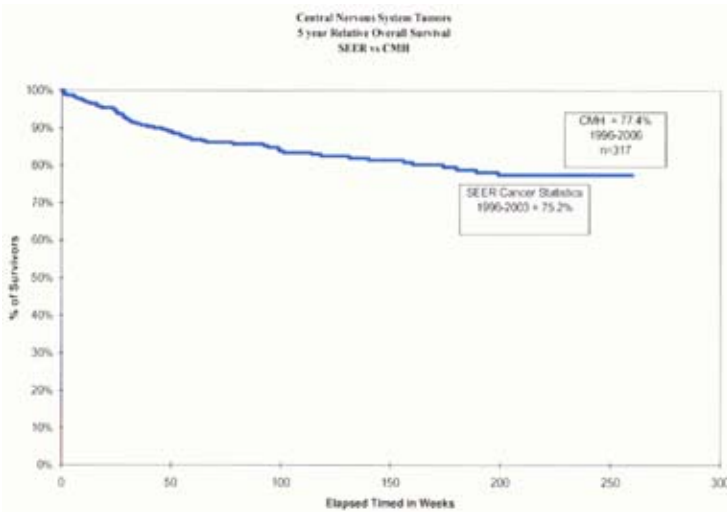
clinical care, education and research. For children diagnosed with tumors of the central nervous system, we work closely with other departments



as multidisciplinary team members dedicated to issues such as disability management. Individual treatment plans for each child depend on the needs of the child as well as the nature and extent of the condition. Our family centered and interdisciplinary approach is

an important part of the eye exam. In many cases, the first suspicion of a tumor is the result of a good

Our family centered and interdisciplinary approach is designed to encourage parents and families to make informed decisions while promoting health, optimal function, and quality of life.



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dilated exam which allows a thorough evaluation of the optic nerves. The initial eye visit provides a baseline from which to compare any changes that may occur. This exam includes a visual acuity assessment of each eye to detect any loss of vision. Pediatric ophthalmologists examine children of all ages and can assess vision even in the newborn baby.

Ophthalmology

Children with CNS tumors may present to the ophthalmologist in the eye clinic with symptoms of headache, blurred or decreased vision, a “lazy eye,” or double vision. Results from this exam may lead to the child being sent for further tumor workup.

Even when there are no visual symptoms, all children with CNS tumors receive an eye exam early in the course of treatment. These exams look for problems that may not even be noticeable to the patient or parents. Dilation of the eyes is

Other important aspects of the eye exam include assessing the eye alignment and movement. Children can have misaligned eyes because of a tumor. Eye alignment is measured and followed and treatment is provided when necessary. Treatment options can include patching to relieve double vision or to avoid developing a “lazy eye.” Eye movement can also be limited due to a tumor. Surgical intervention to realign the eyes is considered only if the movement deficit is longstanding and the tumor has been successfully treated.



A multidisciplinary team meets to discuss patients.

The Oncology Team

Children receiving oncological care for their CNS tumor meet their primary team during the early stages of their diagnosis. Time is taken to introduce the members of the team that will be guiding the care of the child. The child and family also are told what needs to happen before treatment can begin. This is a time to ask and answer questions. The family begins the process of understanding the care that is available for their child right here at Children's Mercy Hospital.

therapist as there may be deficits related to the tumor and or the surgical process.

The goal of the oncology team during this time is to help the patient and family get through the process of surgery and recovery. The child's oncologist follows up with the pathologists to confirm the diagnosis and reviews scans with the radiologist to evaluate objectively the completeness of surgery.

Once the pathology is confirmed and the tumor location and any spread is

of spreading or recurring. These types of tumors are often treated with surgery alone.

For the tumors classified as High Grade or malignant, there is a higher likelihood of spreading or recurring. These tumors often require modalities of treatment (radiation and/or chemotherapy) along with surgery. The overall success rate for these kinds of tumors is closer to 50 percent. Many are significantly poorer than this. It is for these tumors that we are continuing to strive to find better treatments. Many of these

Treatment

The oncologist and neurosurgeon work closely to coordinate getting all the testing done as quickly as possible. While not all brain tumors are treated the same way, a general principal is that whatever it is, if the surgeon can remove the entire tumor this is always preferable to only a biopsy. The location of the tumor, however, can sometimes preclude anything but a biopsy. Sometimes even a biopsy is considered too dangerous to do. Scans can help determine this, but sometimes what looks easy to remove on scans is anything but and what looks like something that will be very difficult to remove is not.

Patients usually spend sometime in the Pediatric Intensive Care Unit following surgery and once stable (usually only a couple of days) they transfer back out to the oncology inpatient unit. Post surgery, the patient may have a reason to interact with our physical and or occupational

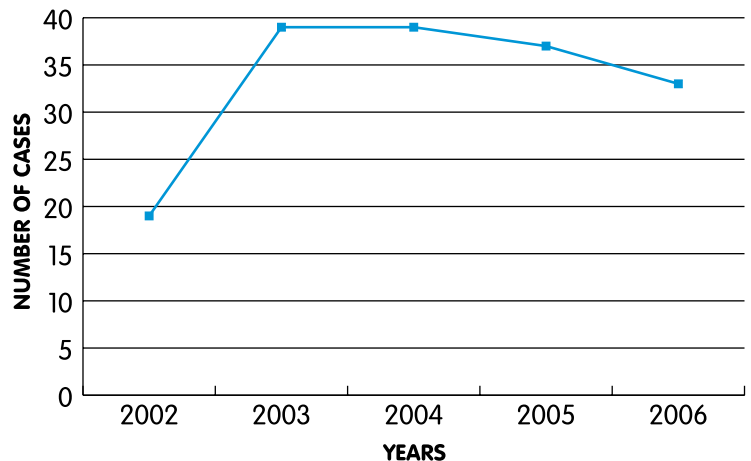
identified the oncologist and his/her team meets again with the family to review all the information, answer questions and discuss treatment options. Oftentimes, the child is a part of this discussion.

While overall success with treating brain tumors is around 70 percent, this number

is a somewhat misleading. This success reflects a large number of brain tumors that are described as being low grade or benign. These tumors do not have a high likelihood

improvements already seen have come from patients taking part in clinical trials. Many of these trials are

INCIDENCE OF CNS CASES AT CHILDREN'S MERCY HOSPITAL



sponsored by the Children's Oncology Group, the leading cooperative group doing pediatric research studies in oncology. Locally, all studies are also reviewed by our Children's Mercy



Hospital Institutional Review Board.

CNS Research and Clinical Trials

Research and clinical trials have had a substantial impact on childhood cancer for several decades. Through cooperative research efforts, cure rates for many childhood cancers have increased dramatically.

The majority of brain tumor research at Children's Mercy is done through a cooperative effort with the Children's Oncology Group (COG). COG is made up of other children's hospitals whose physicians are dedicated to improving pediatric cancer treatment and long term outcomes. Clinical trials approved for enrollment are designed for specific brain tumor types, such

Among these are the Head Start protocol which seeks to find less toxic treatment for younger patients, and protocols open through the Pediatric Blood and Marrow Transplant Consortium. CMH also enrolls patients on the national Atypical Teratoid/Rhabdoid tumor registry. Surgical specimens are also submitted to research banks for future research.

In order for patients to participate in any kind of research, the study must have IRB approval. The IRB is an institutional committee that works together with investigators to make sure that research is safe for human subjects. Once there is institutional approval, the coordinator works with the treating oncologists to screen

Current and future research will continue to answer questions and guide treatment for children with brain tumors.

Pharmacy

The CMH pharmacy staff is both knowledgeable about the special medication needs of pediatric patients and is ready to provide services to our oncology patients with CNS tumors. One of the ways that the pharmacist helps is by finding the best way to give children medications. Because some chemotherapy medications (e.g. temozolomide, lomustine & procarbazine) may only be available for oral use and in dosage forms not ideal for small children, special arrangement may need to be made.



Little 4-year-old Joshua has a little fun with Dr. Alan Gamis in the Hematology/Oncology Clinic. They give each other a "high-five!" Joshua's brain tumor was diagnosed in 2006.

as ependymoma, medulloblastomas, CNS germ cell tumors, and both low and high grade gliomas. Once a patient is enrolled in a research study, a research coordinator will ensure that accurate data is submitted to the research group regarding the child's response to the treatment, all of the side effects encountered and outcome data during long term follow up.

Along with COG trials, CMH also participates in studies in cooperation with other institutions and groups.

patients who are eligible for each trial. Many factors play a significant role to determine whether the child is able to enroll on the clinical trial. The research coordinator must ensure that all pieces of information from each of the other clinical areas as well as oncology have met the requirements laid out in the study design and implementation. All patient data submitted must be accurate, timely and abide by the federal privacy guidelines.

CNS tumors can also have unique treatment pathways that involve the pharmacist. Some brain tumor patients develop a condition after surgery or radiation which may require precise delivery of a hormone given into the nostrils. In each of these cases our pharmacists provide information and education for nurses and families that allow for safe and effective use of these drugs - even in infants! Another chemotherapy agent, bleomycin, is sometimes instilled directly onto the surface of a

rare tumor occurring in the brain. The pharmacy staff prepares a special mixture of this drug when needed for this purpose.

delivery, with our ability to better target the region of interest. In spite of improvements in radiation, we recognize that young patients often suffer devastating consequences

related to therapy. IMRT, an advanced method to focus radiation, is employed whenever indicated for the child. By working as a team, we can offer the best therapy for each patient, offering improved cures and optimal quality of life, as well. Children's Mercy has partnered with Kansas City Cancer Centers and The Gamma Knife

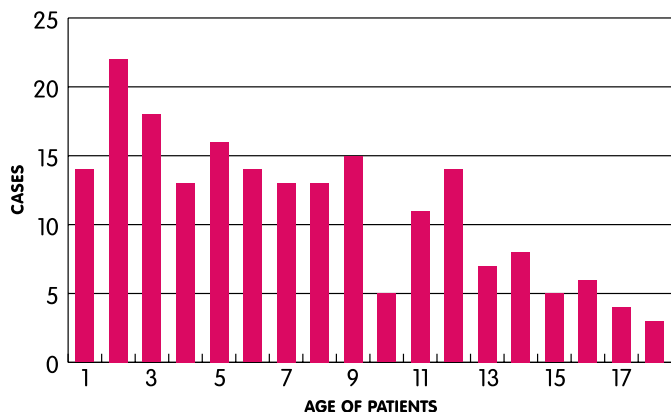
cases, radiation is added to kill even more cancer cells.

This kind of therapy also damages normal tissues. Bone marrow is affected the most. After this high dose therapy, the bone marrow may take a very long time to recover or it may never recover. A person cannot live without a working bone marrow.

A stem cell rescue or autologous transplant gives back saved healthy stem cells that can make the bone marrow work again. Stem cells are collected when the bone marrow is still healthy. These stem cells are frozen in the laboratory and saved until needed. Some children with CNS tumors have stem cells collected, but never have to use them.

When cells are needed, they are thawed and returned into a vein of the patient. The healthy stem cells then grow and the blood counts and immune system gradually return to normal.

AGE OF PATIENTS WITH CNS DIAGNOSIS AT CHILDREN'S MERCY HOSPITAL 1997-2006



Pharmacists dedicated to the oncology service are always available to provide advice on the use and side effects of any drugs.

to offer the most effective treatment options to patients.

Radiation Oncology

Radiation therapy has long been a standard treatment modality for brain tumors. The techniques, delivery precision, and doses have evolved over the years to tailor therapy to best suit the tumor histology, location of the tumor, and even the child's age. Indications for radiation first are determined by the cell type and extent of involvement. Combined modality treatments have altered the timing, and in some cases the doses of radiation used. The volumes treated by radiation therapy are determined by the location at diagnosis and histology, because the patterns of failure and spread are related to these directly. One of the most important improvements to occur has been the precision of

Transplant

Chemotherapy and radiation therapy have been used for many years to treat CNS tumors. For more than a decade, autologous stem cell rescue has also been used in treatment. Chemotherapy and radiation work by killing cancer cells. Higher doses of treatment are sometimes used so more cancer cells can be killed at once. One reason to do this is because cancer cells can become resistant to chemotherapy (chemotherapy drugs no longer kill the cells). The higher doses of the same chemotherapy drugs may be able to kill these resistant cells. In some

5 YEAR RELATIVE SURVIVAL RATES FOR CNS TUMORS NATIONAL / CMH COMPARISONS

| HISTOLOGY | ICDO Code (s) | SEER 1973-2002 | CMH 1990-2004 |
|-----------------------------------|---------------------------------------|----------------|---------------|
| Astrocytoma, NOS | 9400 | 76.6% | 86.5% (n=30) |
| Pilocytic Astrocytoma | 9421 | 94.2% | 97.3% (n=77) |
| Anaplastic Astrocytoma | 9401, 9411 | 50.7% | 33.3% (n=3) |
| Ependymoma/ anaplastic ependymoma | 9391, 9393 | 54.7% | 57.1% (n=23) |
| Embryonal /PNET/ Medulloblastoma | 8963, 9470-9473, 9502-9503 | 55.7% | 52.4% (n=59) |
| Glioma, malignant, NOS | 9380 | 47.3% | 61.3% (n=34) |
| Glioblastoma | 9440-9441 | 19% | 14.3% (n=7) |
| Germ Cell, CNS SEER 1996-2003 | 9064, 9080-9085, 9070-9071, 9100-9101 | 85.6% | 90.9% (n=11) |

Social Work

A variety of services are provided through our psychosocial team. Social workers are an integral part of the team as they demonstrate the importance we place on comprehensive, family centered care. Our social workers are Master's -prepared and each has expertise serving the needs of our children and their families. Each social worker works closely with a primary oncologist and advanced practice nurse to become the patient and family's 'primary team'.

This team approach is especially important for the child or adolescent with a CNS tumor. At diagnosis, the primary team sits down with the patient's family and patient (when

and emotionally from the diagnosis. In addition, when appropriate, they refer patients and families to other psychosocial team members such as child life, chaplaincy, and child psychology.

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

support group setting. The program also offers bereavement services to families. A network of parent volunteers who have lost children supports this service.

Referrals are also made to vocational and rehabilitation services as needed. These programs can help children keep up with daily tasks, as well as learn future skills. Necessary and accessible medical equipment is also obtained. At parents' request, social workers assist with the application for Social Security Disability benefits.

In addition to the Parent to Parent support, social workers also offer counseling and support to families who are grieving the loss of a child.

Support

age appropriate). As a group, they discuss the diagnosis, treatment plan and begin to answer questions. The social worker provides supportive counseling and assists the family in organizing their own resources. They also begin identifying additional resources that could aid the family in the journey ahead.

The social worker completes a psychosocial assessment on each patient. This forms the basis for the subsequent relationship with the child and family. The assessment aids the family and social worker in determining the resources and assistance needed as well as making preparations to begin treatment. Social workers provide counseling and active listening as patients and families begin to heal both physically

Social workers refer patients and their families to outside agencies for support and financial assistance. Families get assistance from such groups as the Pediatric Brain Tumor organization, American Cancer Society, Turning Point and Solace House. Referrals for lodging and transportation assistance, are also given when appropriate.

CMH offers an exciting Parent to Parent program, coordinated by one of our social workers. In this program, experienced parents of children with cancer volunteer their time and are paired with a family with newly diagnosed children. The volunteer shares their expertise and offers support. This unique team has been much more successful than the

Families are guided through the funeral process and continue to receive support once outside the hospital. Both internal bereavement groups and outside resources are offered in order to best meet the needs of each individual family.

Social workers realize that treatment is not merely a series of chemotherapy, clinic appointments, radiation therapy and inpatient hospitalizations. It is a process where the most important outcome is that the patient and family are able to define their 'new normal' after treatment ends. For many of our patients with CNS tumors, their lives will never be the same. They will require adjustment, assistance, equipment and opportunities to

compensate for lost functions. As part of an ongoing partnership with each patient and family, social workers consider it their privilege to accompany patients and families on their journey and bear witness to their determination, courage and resilience.

Developmental Medicine

No parent ever anticipates that they will ever hear the words “Your child has a brain tumor.” Families frequently experience fear, anger, anxiety, sadness and depression. At the same time they feel hope, determination and optimism. While there are few things more

unit services can be offered to the patient, siblings and parents. In addition to a designated psychologist who works closely with the Division of Hematology/Oncology, consultative services with a psychiatrist and family therapist are also available.

Along with the emotional impact on a child, brain tumors and their treatment can lead to changes in cognitive functioning. Children can have changes in how they problem solve, memory, attention, and processing speed. These changes can lead to problems in school. We routinely provide evaluations to children diagnosed

Child Life Specialists

Child Life specialists dedicated to the Hematology/Oncology Division are available in the outpatient clinic, on 4 Henson the inpatient floor and in the Bone Marrow Transplant unit. A Child Life specialist is trained to meet the developmental, emotional and psycho-social needs of children and their families. They accomplish this by advocating for children in all areas of the hospital.

Child Life specialists can be seen playing with children or “hanging out” with teens. They recognize “play” as a child’s “work” – allowing the building of rapport with children

Little Joseph is ready to play with Child Life Specialist Amanda Woelk in the Hematology/Oncology playroom. Joseph had surgery for an ependymoma in 2007.



stressful in life than having to deal with cancer in a child, a cancer discovered in the brain adds ever more challenges.

The Section of Developmental and Behavioral Sciences offers comprehensive psychological services to children and families affected by these tumors. Services are tailored to the needs of each child and family. Staff members counsel as well as facilitate coping with the stressors that accompany the diagnosis and treatment. Counseling can be done both on the inpatient unit, and in the Developmental and Behavioral Sciences Clinic. Individual or family

with brain tumors, and other CNS cancers. These evaluations help to identify any cognitive/learning areas that treatment might impact. This information is used to monitor progress over the years and to assist the child, parent and school in developing individualized educational plans that maximize the child’s learning potential and ease frustration. Staff can assist the family in communicating with the child’s school. They can relay information regarding the unique strengths and challenges of each child, as well as what the school, family and medical team can do to support that child.

and helping them to adjust to hospitalization, diagnosis, and understanding.

Child Life specialists aid in pain management and facilitate positive coping skills. They also are there to help the child adjust to their new diagnosis and the life changes that come with it. The Child Life specialist works to provide multiple opportunities for socialization and peer interaction. They also provide sibling support in their role.





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