Neuro-Oncology Program
Annual Report 2010

Neurosciences
Hematology and Oncology Center

Cook Children’s
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From the Desk of Rick W. Merrill,
President and CEO,
Cook Children’s Health Care System

This report outlines the comprehensive care that is the hallmark of our Neuro-Oncology program. As part of Neurosciences and the Hematology and Oncology Center, this program highlights our multidisciplinary approach to care. As one of the largest pediatric Neuro-Oncology programs in the Southwest, we participate in national and international clinical research trials and continue to forge a collaborative approach to care both within Cook Children’s and through initiatives such as the Telemedicine Neuro-Oncology Tumor Board Conference with other centers in the region.

We are immensely proud of our Neuro-Oncology team and the breadth of care and services they provide to our patients and their families. Their distinguished reputation has been showcased through participation in national and international symposiums and again when they hosted the inaugural Pediatric Brain Tumor Symposium at Cook Children’s. They share a relentless commitment to providing patient-centered care while employing leading-edge technologies and best practices to ensure outstanding patient care.

They are truly redefining the future for our patients through research, engagement and collaboration.

Sincerely,

Rick W. Merrill
President and CEO
Cook Children’s Health Care System
From the Desk of Jeffrey C. Murray, M.D., Medical Director of Neuro-Oncology

Dear colleagues and friends,

Great progress has been made in the past decade in the understanding and treatment of central nervous system (CNS) tumors in youth. Advances in neurosurgical technologies and techniques, radiation therapy approaches and optimal chemotherapy delivery strategies have been significant. Furthermore, the gradual unraveling of previously unknown molecular biological features of many tumors is translating into novel classification categories and treatment. Tragically, despite this exciting progress, CNS tumors remain the leading cause of death in children and adolescents with neoplastic disease. With nearly 3,000 tumor cases diagnosed each year in the United States, this patient population deserves ongoing efforts by the pediatric neuro-oncology scientific community to further its understanding of the myriad of diagnoses and therapy mysteries that remain unsolved.

The Cook Children’s Neuro-Oncology program, part of both Neurosciences and the Hematology and Oncology Center, is excited to be positioned at the forefront of clinical research and patient-centered care in the treatment of children with CNS tumors. The 2010 program year was busy and rewarding. The program evaluated 85 new patients, the largest annual patient volume to date, positioning Cook Children’s Neuro-Oncology as one of the largest pediatric programs in the Southwest.

This year’s annual report highlights the complex medical care and support programs that have been established by a collaborative and multidisciplinary care team dedicated to the well-being of our young patients. The team continued and expanded on many successful programs in 2010, such as the *Evenings with Neuro-Oncology* series and *Telemedicine Neuro-Oncology Tumor Board* conferences and pursued meaningful scholarly activities. In addition to numerous national and international meeting presentations and publications, we have reviewed our 18 year experience in the diagnosis and management of patients with ependymoma; the patient care evaluation discussing this unique CNS tumor is summarized in this report.

The Neuro-Oncology program was excited to host the inaugural
*Pediatric Brain Tumor Symposium* at Cook Children’s Medical Center in November 2010. The continuing medical education meeting was attended by 120 physicians, nurses and other health care professionals from across the state and country. Eight renowned speakers shared the latest information on topics such as medulloblastoma, gliomas, germ cell tumors, radiotherapy and neuropsychology.

As Tarrant County and the surrounding communities of North Texas continue to grow with relatively youthful populations, the burden of childhood, adolescent and young adult CNS tumors will continue to increase. The Cook Children’s Neuro-Oncology program is well-positioned and committed to provide compassionate, technologically leading edge, patient-centered care to this very special population of children.

Jeffrey C. Murray, M.D.
Medical Director of Neuro-Oncology
Neurosciences
Hematology and Oncology Center
2010 Summary of CNS Tumor Registry

Neuro-Oncology program, new patient evaluations

Neuro-Oncology program, age at diagnosis of new patient evaluations

Neuro-Oncology program, new patient tumor location

Supratentorial brain/cerebral cortex
Posterior fossa (non-brainstem)
Other (spine, non-CNS, etc.)
Optic pathways/hypothalamus
Brainstem (intrinsic, e.g., DIPG)
Brainstem (non-intrinsic)
Pineal region
### Neuro-Oncology program, new patient tumor histology

<table>
<thead>
<tr>
<th>Tumor types</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tumors not biopsied (brainstem gliomas, optic pathway tumors, observation only patients)</td>
<td>24</td>
</tr>
<tr>
<td>Low-grade astrocytoma (W.H.O. grades I, II)</td>
<td>14</td>
</tr>
<tr>
<td>Neurofibromatosis (abnormal MRI, suspected tumor, other)</td>
<td>11</td>
</tr>
<tr>
<td>Glioneuronal tumors</td>
<td>8</td>
</tr>
<tr>
<td>Ependymoma</td>
<td>7</td>
</tr>
<tr>
<td>Other histologies (vascular malformations, dermoid lesions, others)</td>
<td>7</td>
</tr>
<tr>
<td>Craniopharyngioma</td>
<td>5</td>
</tr>
<tr>
<td>Medulloblastoma</td>
<td>2</td>
</tr>
<tr>
<td>Germ cell tumor</td>
<td>2</td>
</tr>
<tr>
<td>Pineoblastoma/supratentorial PNET</td>
<td>2</td>
</tr>
<tr>
<td>High-grade astrocytoma (W.H.O. grades III, IV)</td>
<td>2</td>
</tr>
<tr>
<td>Choroid plexus tumor</td>
<td>1</td>
</tr>
<tr>
<td><strong>TOTAL</strong></td>
<td><strong>85</strong></td>
</tr>
</tbody>
</table>
Distribution of new patient referrals
Pediatric Brain Tumor Symposium
The Neuro-Oncology program hosted the inaugural Pediatric Brain Tumor Symposium at Cook Children’s, Nov. 5-6, 2010. This continuing medical education symposium brought together 120 physicians, nurses and allied health professionals interested in the care of youth with CNS tumors, to learn from eight renowned speakers:

- Daniel C. Bowers, M.D., Children’s Medical Center, Dallas, Texas
- Murali Chintagumpala, M.D., Texas Children’s Hospital, Houston, Texas
- Stewart Goldman, M.D., Northwestern Fienberg School of Medicine, Chicago, Ill.
- Hayden W. Head, M.D., Cook Children’s, Fort Worth, Texas
- John Honeycutt, M.D., Cook Children’s, Fort Worth, Texas
- Craig Lustig, M.P.A., National Cancer Institute, Bethesda, Md.
- Donald W. (Will) Parsons, M.D., Ph.D., Texas Children’s Hospital, Houston, Texas
- Douglas Ris, Ph.D., Texas Children’s Hospital, Houston, Texas
- Shiao Y. Woo, M.D., University of Louisville, Louisville, Ky.

Topics discussed included medulloblastoma, gliomas, CNS germ cell tumors, brain tumor molecular biology, neuroimaging, radiation oncology, neuropsychology and patient advocacy.

Neuro-Oncology Tumor Board Conferences

The Neuro-Oncology Tumor Board Conference is a weekly multidisciplinary conference designed for the discussion of complicated CNS tumor patients, both new and existing, as well as didactic teaching. The board brings together representatives from Neuro-Oncology, Neurosurgery, Neurology, Radiation Oncology, Pathology, Neuroradiology, Nursing, Social Services, Pastoral Care, Rehabilitation and other disciplines, in an exciting conference format aimed at making the best patient care decisions. A continuing medical education-accredited conference, more than 100 patients were actively discussed at Tumor Board in 2010. Quarterly invited guest speakers, part of the ‘A View Into Their World’ series, included Joel Steelman, M.D., (Neuroendocrine Complications of CNS Tumor Therapy) and a presentation from the in-patient rehabilitation staff about matters pertaining to the rehabilitation of children with brain tumors.

Evenings with Neuro-Oncology

The program continued its quarterly Evenings with Neuro-Oncology events, a patient and family education series, in 2010. This free dinner program series is held at Cook Children’s Medical Center and hosted by Neuro-Oncology staff. Following an informal group dinner, patients and their siblings are treated to special activities provided by Child Life staff, while their
parents gather to learn more about CNS tumors. Presentation topics in 2010 included: *Hormones and Health After Brain Tumor Treatment*, Joel Steelman, M.D.; *Brain Tumors and Neurosurgery: A Picture Show*, John Honeycutt, M.D. and Jeffrey C. Murray, M.D.; *Siblings, School and So Much More: What Can I Do To Support My Child Through Treatment*, Katie Campbell, C.C.L.S.; *Surviving a Brain Tumor: I’m at Risk for What?*, Lisa Bashore, M.S., CPNP, CPON. Family feedback remains tremendously positive. An evaluation of the event was performed using feedback questionnaires from participating families. The results were accepted for presentation at the 2010 International Symposium on Pediatric Neuro-Oncology in Vienna, Austria.

**Scholarly Activity**
The program authored numerous scientific papers that were accepted for presentation and publication in 2010:

**Abstracts:**


Harrell LM, Marks WA, Murray JC, Colaluca B, Braly EZ. Duration of in-patient neurorehabilitation course and improvement in neurologic function for posterior fossa syndrome: Differences between


**Medical Journal Manuscripts:**

**Neuro-Oncology Comprehensive Clinic**
The Neuro-Oncology Comprehensive Clinic is a multidisciplinary clinic held twice monthly at the Cook Children’s Hematology and Oncology Center. Patients are evaluated by each member of the care team, which includes Neuro-oncology, Neurology, Radiation Oncology, Neuropsychology, Endocrinology, Nursing, Child Life, Social Services and the Life After Cancer Program. Treated patients vary from recently diagnosed children to those who have completed therapy for many years. Convenient to the patient and family, the clinic is an optimal way for patients to see all of their specialists simultaneously. Patients and families continue to provide positive feedback about the benefits of this special clinic.
**Neurofibromatosis Comprehensive Clinic**

Children with neurofibromatosis (NF), a rare genetic disorder leading to developmental disorders and various CNS tumors, require specialty care from an experienced team of care providers. The Neurosciences program conducts a formal multidisciplinary monthly clinic, with care provided by specialists in Neurology, Neuro-Oncology, Neuropsychology, Genetics, Social Services and a Patient Care support organization (Texas Neurofibromatosis Foundation). In 2010, the clinic evaluated more patients than ever before, with very positive patient and family feedback.

**Professional Growth, Learning and Outreach**

Staff members of Cook Children’s Neuro-Oncology program attended numerous local, regional and national professional meetings in 2010:

- Genetics & Biology of Childhood Cancer Meeting, San Antonio, Texas
- International Symposium on Pediatric Neuro-Oncology, Vienna, Austria
- Neurofibromatosis (NF1) Annual Conference, Baltimore, Md.
- Neurofibromatosis Type 2 State of the Art Meeting, Las Vegas, Nev.
- Pediatric Epilepsy and Movement Disorder Symposium, Fort Worth, Texas
- Pediatric Neuro-Oncology Symposium, Fort Worth, Texas
- Annual Association of Pediatric Hematology/Oncology Nurses Conference & Exhibit, Minneapolis, Minnesota
- Society for Neuro-Oncology Annual Meeting, Montreal, CA
- Medical trainee education and pre-medical trainee observation and research experiences are becoming an ever more important aspect of Cook Children’s. The Neuro-Oncology program hosted local high school, college pre-medical (Texas Christian University) and medical students (Texas College of Osteopathic Medicine) in 2010, as part of the summer research intern program.

The Neuro-Oncology program team participated in a team retreat in the summer of 2010 as an opportunity for team building, better understanding of the stresses of caring for extraordinarily sick children with brain tumors and having a day of fun together.

The Neuro-Oncology program presented Pediatric Grand Rounds at the University of Oklahoma, Tulsa Campus, Tulsa, Oklahoma, June 2010, on Neuro-Oncology & Neurosciences: New Frontiers for Pediatric Specialty Care. The program was also an invited presenter at the Amarillo Area 2010 Cancer Symposium, September 2010, on Brain Tumors in Children: Symptoms, Diagnostics and Treatment.

**Endowments and Community Partnerships**

The N.B. Carter Neuro-Oncology Endowment, established in 1994 at Cook Children’s Medical Center, continues to serve as an extraordinarily meaningful source of funds to permit growth and expansion of various special patient and family care programs, as well as staff education.

The Cancer Research Foundation of North Texas (CRFNT) is a well-established medical grant foundation which has been a special supporter of numerous Cook Children’s cancer research projects over the years. In 2010, the CRFNT and the Neuro-Oncology program partnered in the creation and hosting of the inaugural Pediatric Neuro-Oncology Symposium professional education meeting at Cook Children’s.

**L I V E S T R O N G**

The Neuro-Oncology Program was the proud recipient of a 2010 LIVESTRONG Community Impact Project grant award, to develop and implement a unique Artist in Residence program for brain tumor patients at Cook Children’s. The program will be modeled after the very successful ‘The Creative Center: Arts in Healthcare’ program in New York City, NY.

LIVESTRONG fights for the 28 million people worldwide living with cancer, giving people the resources and support they need to fight this disease head-on. LIVESTRONG raises awareness, funds research and ends the stigma that many survivors face. Anyone, anywhere can join the fight against cancer at LIVESTRONG.org.
Neuro-Oncology Weekly Team Meeting

All staff members of the Neuro-Oncology program, including Oncology, Neuropsychology, Neurology, Nursing, Social Services, Pastoral Care, Audiology, Physical Therapy, Clinical Research and Dietary Services meet weekly to review details on all neuro-oncology children hospitalized in the medical center and all patients to be seen in the outpatient clinic the following week. This is the ideal venue to assure efficient and organized patient care and adherence to clinical trial guidelines.

Neuro-Oncology Telemedicine Tumor Board Conference

The Cook Children’s and Texas Children’s Cancer Center (Texas Children’s Hospital, Houston) Neuro-Oncology programs continue to share a monthly Neuro-Oncology Telemedicine Tumor Board Conference series. This shared conference, made possible by sophisticated telecommunications technology, allows the creation of a ‘virtual room’ occupied by the multidisciplinary neuro-oncology teams from both Cook Children’s and Texas Children’s. Each institution presents several complicated CNS tumor patients, complete with imaging and pathology data, followed by discussion from the entire ‘virtual room.’ This conference continues to serve as a great teaching opportunity and results in improved patient care. Pediatric oncology colleagues from The Children’s Hospital at Saint Francis, Tulsa, Okla., began participating in the conference in September, making for a dynamic, three-institution collaborative monthly conference.

Neuro-Oncology Program Clinical Trials

The Neuro-Oncology Program actively participates in numerous clinical, biology and epidemiology trials via various research organizations, primarily the Children’s Oncology Group (COG). Examples of open trials that Cook Children’s participated in during 2010 include:

COG:
ACNS0221 — A Phase II Study of Conformal Radiotherapy in Patients with Low Grade Gliomas
ACNS0331 — A Study Evaluating Limited Target Volume Boost Irradiation and Reduced Dose Craniospinal Radiotherapy 18.00 Gy and Chemotherapy in Children with Newly Diagnosed Standard Risk Medulloblastoma: A Phase III Double Randomized Trial

ACNS0332 — Efficacy of Carboplatin Administered Concomitantly With Radiation and Isotretinoin as a Pro-Apoptotic Agent in Other Than Average Risk Medulloblastoma/PNET Patients

ACNS0333 — Treatment of Atypical Teratoid/Rhabdoid Tumors of the Central Nervous System with Surgery, Intensive Chemotherapy, and 3-D Conformal Radiation

ACNS0334 — A Phase III Randomized Trial for the Treatment of Newly Diagnosed Supratentorial PNET and High Risk Medulloblastoma in Children < 36 Months Old with Intensive Induction Chemotherapy with Methotrexate Followed by Consolidation with Stem Cell Rescue versus the Same Therapy Without Methotrexate

ACNS02B3 — A COG Protocol for Collecting and Banking Pediatric Brain Tumor Research Specimens

ACNS0831 — A Phase III Randomized Trial of Post-Radiation Chemotherapy in Patients with Newly Diagnosed Ependymoma Ages 1 to 21 Years

ALTE07C1 — Neuropsychological, Social, Emotional and Behavioral Outcomes in Children with Cancer

Texas-Oklahoma Pediatric Neuro-Oncology Consortium:

TOPNOC 001-08 — A Phase II Study of Valproic Acid and Radiation, followed by Maintenance Valproic Acid and Bevacizumab in Children with Newly Diagnosed High-grade Gliomas or Brainstem Gliomas

Other Trials:

TCCC H6650 – A Brain Tumor Tissue Banking Study, Texas Children’s Hospital and Cook Children’s Medical Center

Camp Neuro-Oncology Weekend

In September 2010, the program hosted its second annual, free-of-charge, three-day weekend camp for patients, siblings and parents. Camp Neuro-Oncology Weekend (Camp NOW) is held at the beautiful Camp John Marc facility in the Texas Hill Country. It provides a special opportunity for patients and their families to get away and enjoy time together and with other families. In addition to fun-filled activities, such as fishing, arts and crafts and a ropes challenge course, families are offered various education opportunities about brain tumors and emotional support. Returning with official Camp NOW T-shirts, other freebies, and many pictures and memories, Camp NOW remains a great event and will return in 2011.

Future Projects and Program Goals

• Participation in the 2012 International Symposium on Pediatric Neuro-Oncology
• Participation in a planned multi-institution trial studying the neurocognitive damaging effects of medulloblastoma therapy in the context of genetic polymorphisms
• Professional growth and enhanced patient care and research via education and partnerships with regional centers
• Prepare and submit research, via scientific abstracts, to various planned 2011 professional meetings
• Continue and enhance special patient care programs, including Evenings With Neuro-Oncology and Camp NOW

You are invited to participate in the Neuro-Oncology Family Camp at Camp John Marc, Texas Hill Country. It provides a special opportunity for patients and their families to get away and enjoy time together and with other families. In addition to fun-filled activities, such as fishing, arts and crafts and a ropes challenge course, families are offered various education opportunities about brain tumors and emotional support. Returning with official Camp NOW T-shirts, other freebies, and many pictures and memories, Camp NOW remains a great event and will return in 2011.
Ependymoma is a malignant glial neoplasm derived from the normal ependymal cells that line the surface of the brain's ventricles and the central canal of the spinal cord. It accounts for 8-10 percent of all primary central nervous system tumors in children. Intracranial ependymoma is the most common location, with 30 percent arising in the supratentorial compartment and 70 percent arising in the posterior fossa, most commonly in or near the 4th ventricle (Figure 1). The mean age at diagnosis is 4-6 years, but 25-40 percent of patients are less than two years old. Infratentorial ependymoma is predominant in children less than three years of age, while ependymoma of the spine is most common in the 15 to 40 year old population.

The incidence of ependymoma is two new cases per million healthy children per year, with less than 170 new cases diagnosed in the pediatric age range annually in the United States. There is a slight predominance of males, as with many childhood neoplasms. Metastases are uncommon, but if present portend a very poor prognosis.

Image source: Lippincott Williams & Wilkins

Figure 1
Sagittal view of the brain ventricles, the cavities that contain cerebrospinal fluid, serving to cushion the brain and spinal cord. There are two lateral ventricles split across the middle of the brain, the third ventricle in the center, and the fourth ventricle located near the cerebellum.
Clinical Presentation
Children with ependymoma typically present with a history of persistent headaches, nausea and vomiting, all signs of increased intracranial pressure. When ependymoma arises in the posterior fossa, ataxia, cranial nerve palsies and changes in overall mental status may be seen. Supratentorial ependymoma may present with mood and personality changes, cognitive problems, as well as seizures. Regardless of site, tumors may cause weakness. Most children will present with symptoms presenting or evolving over several weeks to months.

Diagnostic Imaging
Magnetic resonance imaging (MRI) is the preferred modality for imaging ependymoma tumors. Compared to computed tomography (CT), MRI scans show greater soft-tissue detail. Typically, comprehensive images are taken with T1 and T2 weighted techniques and gadolinium enhancement. Ependymoma appears hypointense (dark) on T1-weighted images, hyperintense (white) on T2-weighted images and gadolinium enhancement is typically prominent.

Histopathology
The microscopic features typical of ependymoma include perivascular pseudorosettes, dense cellularity, round tumor cells and increased mitotic activity (Figure 2).

The World Health Organization (W.H.O.) has developed a grading system (grades I-III) for ependymomas, which roughly correlate with clinical tumor behavior and prognosis.

Subependymomas are rare, slow growing, benign tumors, while myxopapillary ependymomas (grade I) are almost exclusively found in the spinal cord. The subcategories of classic ependymomas are differentiated as follows: cellular (grade II) are the most common, demonstrating significant cellularity without abnormal mitotic activity; papillary, in which the cells form linear structures near CSF surfaces; the clear cell variant are typically supratentorial; and tanycytic are the rarest subtype. Finally, anaplastic ependymomas (grade III) are the most malignant form, with greater mitotic activity and the poorest prognosis.

Prognosis
The 5-year overall survival rate and progression free survival rate of children with ependymoma is 50-64 percent and 23-45 percent, respectively. The 5-year survival rate of spinal ependymoma alone, however, is more promising, ranging from 70-100 percent.

The prognostic significance of gross
Figure 2
Photomicrograph of a W.H.O. grade II classic, cellular ependymoma. Tumor cells are densely populated around vascular structures, forming perivascular pseudorosettes.
total resection of the primary tumor mass (usually then followed by adjuvant radiation therapy), is critical. The 5-year survival rate following GTR is 67-80 percent compared to 22-47 percent with subtotal resection. Younger age at diagnosis, higher grade histology, lesser extent of resection, and disease dissemination are the most important factors associated with unfavorable outcomes.

The recurrence rate is greater in intracranial ependymomas than spinal, with survival following relapse being generally poor.

**Treatment**

The current standard of care for ependymoma is gross total resection (GTR) of the tumor, followed by conformal radiation therapy. GTR is assessed based on the description from the operating neurosurgeon, as well as postoperative imaging studies showing no evidence of residual disease. GTR is achieved in approximately 40-60 percent of patients.

Conformal radiation therapy, using photon or proton techniques, has become the standard adjuvant (i.e., after recovery from neurosurgery) therapy for the vast majority of ependymomas.

Chemotherapy has not proven curative, but is known to shrink tumors enough to delay the need for radiation therapy in infants and younger children or facilitate a second-look surgery for patients with residual tumors.

The primary problem for patients with recurrent ependymoma is that of local tumor control. Repeat neurosurgery and/or the delivery of further radiation therapy have yielded the best salvage results. Radiotherapeutic techniques

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**Table 1. Five-year overall survival probabilities for all children diagnosed with and/or treated for Ependymoma at Cook Children’s Medical Center, 1992-2009‡.**

<table>
<thead>
<tr>
<th></th>
<th>Living n (%)</th>
<th>Deceased n (%)</th>
<th>5-year OS ± SD (%)</th>
<th>p-value†</th>
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<tbody>
<tr>
<td><strong>Tumor Location</strong></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Infratentorial</td>
<td>9 (52.9)</td>
<td>7 (87.5)</td>
<td>48.8 ± 13.8</td>
<td>0.0110</td>
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<tr>
<td>Spinal</td>
<td>1 (5.88)</td>
<td>1 (12.5)</td>
<td>50.0 ± 35.4</td>
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<tr>
<td>Supratentorial</td>
<td>7 (41.2)</td>
<td>0</td>
<td>100.0 ± 0.0</td>
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<tr>
<td><strong>Metastases</strong></td>
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<td></td>
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<tr>
<td>No</td>
<td>12 (75.0)</td>
<td>4 (57.1)</td>
<td>68.9 ± 13.4</td>
<td>0.8918</td>
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<td>Yes</td>
<td>4 (25.0)</td>
<td>3 (42.9)</td>
<td>50.0 ± 20.4</td>
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<tr>
<td><strong>Surgical Resection</strong></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Partial</td>
<td>3 (18.8)</td>
<td>4 (50.0)</td>
<td>34.3 ± 19.5</td>
<td>0.1697</td>
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<tr>
<td>Full</td>
<td>13 (81.2)</td>
<td>4 (50.0)</td>
<td>66.6 ± 14.0</td>
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<tr>
<td><strong>Chemotherapy</strong></td>
<td></td>
<td></td>
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<tr>
<td>No</td>
<td>13 (76.5)</td>
<td>4 (44.4)</td>
<td>69.3 ± 17.0</td>
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<tr>
<td>Yes</td>
<td>4 (23.5)</td>
<td>5 (55.6)</td>
<td>50.0 ± 15.8</td>
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<tr>
<td><strong>Recurrence</strong></td>
<td></td>
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<tr>
<td>No</td>
<td>12 (70.6)</td>
<td>3 (37.5)</td>
<td>68.6 ± 13.6</td>
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<tr>
<td>Yes</td>
<td>5 (29.4)</td>
<td>5 (62.5)</td>
<td>44.4 ± 16.6</td>
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</tr>
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</table>

OS=Overall Survival; SD=Standard Deviation
‡ Partial Patient Data missing on 3 patients: 1 original diagnosed in 1992 with limited documentation, 2 originally treated outside of CCMC
† Log-Rank Statistical Test for difference between groups
commonly used are stereotactic radiosurgery, reirradiation to the locally recurrent tumor bed, or full craniospinal irradiation. The detailed biological features of ependymoma are not well-known. Recently, coexpression of the ERBB2 and ERBB4 genes was found in over 75 percent of pediatric ependymomas. These factors have prognostic implications in ependymomas, with ERBB pathway inhibitors currently under evaluation in a contemporary phase 1 clinical trial. Additionally, monosomy or translocations of chromosome 22 have been found in approximately 30 percent of tumors. Recent studies have suggested that an ependymoma tumor suppressor gene exists on chromosome 22. Ongoing studies are also examining the relationship between ependymomas and neurofibromatosis type 2, the causative gene of which is located on chromosome 22.

The Cook Children’s Experience
Between January 1992 and December 2009, Cook Children’s diagnosed and/or treated 26 patients with ependymoma. Following approval from the Institutional Review Board, a retrospective analysis of the medical records of this cohort of patients was conducted in order to gain insight into the patterns of disease, treatment strategies and outcomes, with intent to guide future ependymoma patient care at Cook Children’s.

Tumor location was found to have statistical significance within the population (p=0.011). The majority of patients had W.H.O. grade II ependymoma (76.9 percent), which also had the best survival probability (70.6 percent). This has been recognized in the medical literature. What is striking from tumor location data [Table 1] is that one-fourth of the patients had supratentorial lesions, and all are living at last follow-up. This data correlates well with published reports. Data for administration of chemotherapy seemingly demonstrates no beneficial effect; however, chemotherapy was administered to patients who were either infants, in order to prevent harmful radiation effects, or as adjuvant therapy when surgical resection and radiation therapy were not effective.

A curious observation from our patient set was that females had a better overall survival than males [Figure 3], although this was not found statistically significant.

The overall 5-year survival of our study population, as seen in Table 1, is similar to published reports. We conclude from this retrospective review that Cook Children’s patient population does not differ from national reviews and our Neuro-Oncology Program has been successful in providing ‘best care’ to this unique population with a rare CNS tumor.

Figure 3
Kaplan-Meier survival curve based on gender illustrates increased survival in females (n=12) vs. males (n=14); Log Rank X² = 2.42, p = 0.1195.
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www.cookchildrens.org/neuro