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# Promise

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# Promise

A publication of St. Jude Children's Research Hospital Autumn 2010



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# Highlights



# **Honored by Royal Society**

St. Jude virologist Robert Webster, PhD, who holds the Rose Marie Thomas Chair in Infectious Diseases, recently presented the 2010 Leeuwenhoek Prize Lecture, a prestigious recognition awarded by the Royal Society in London.

The prize lecture acknowledges Webster's many contributions to the field of virology.

"The Leeuwenhoek Lecture is a well-deserved acknowledgement for Dr. Webster, whose life's work has greatly progressed influenza research worldwide," said Dr. William E. Evans, St. Jude director and CEO. "The Royal Society honor is a reflection of an esteemed career that has provided significant advancements to the field of virology."

Webster's presentation provided an overview on pandemic influenza viruses and included a discussion of origins, global surveillance efforts as well as response to the 2009 H1N1 strain worldwide.

# St. Jude tops in academia

For the fifth consecutive year, St. Jude is ranked as one of the top 10 institutions in the annual "Best Places to Work in Academia" list by *The Scientist* magazine.

"The best way to accelerate progress toward our mission of advancing cure rates for childhood cancer and other catastrophic diseases is to have the best and brightest researchers collaborating in an environment where they can do their best work," said Dr. William E. Evans, St. Jude director and CEO. "We work hard to create an environment that facilitates innovation and teamwork, and this latest acknowledgement further validates that St. Jude is an exceptional place for scientists and clinicians to conduct research and make an impact on serious childhood illnesses."

Earlier this year, the hospital was also named the nation's top children's cancer hospital in the 2010–11 Best Children's Hospital rankings published in *U.S. News & World Report.* 

# Understanding origins of cancer subgroups

An international team led by St. Jude scientists has developed an approach using genomic information from various species to understand the biology that drives the formation of different cancer subtypes. The team used that strategy to identify new subtypes of a brain and spinal cancer based on the molecular fingerprint of each tumor.

The work focused on ependymomas, cancers of the brain and spine that strike both children and adults. Richard Gilbertson, MD, PhD, Developmental Neurobiology and Oncology, said the approach is now being used to identify the stem cells and genetic mutations that combine to launch other brain tumors and to search for new, more targeted drugs. Gilbertson is senior author of the study, which was published in the advance, online edition of the journal *Nature*.

The study also identified *EPHB2* as the first ependymoma oncogene. The research provides additional evidence that ependymomas in different regions of the nervous system result from specific mutations in the stem cells specific to those same areas. The work expanded the number of ependymoma subtypes to nine, identified the particular subpopulation of stem cells from which several subtypes likely develop and led to the first laboratory model of the cancer.



"These are the same tools and techniques that we are now using to find new targets and drugs to treat childhood cancer."

# Promising drug for medulloblastoma

Researchers from St. Jude and the Pediatric Brain Tumor Consortium recently announced the findings of a study using an experimental drug to target the underlying genetic makeup of a brain tumor called medulloblastoma. The research focused on a new way to attack the tumors by blocking the Hedgehog pathway that is linked to approximately 20 percent of medulloblastomas.

The study is the first to report that the drug, named GDC-0449, can be safely administered to children and is showing early signs of efficacy.

"The trend in treating children with these cancers is toward targeted therapies like this one, which block key signaling pathways and disable the cancer's ability to function or reproduce," said the trial's principal investigator, Amar Gajjar, MD, Oncology co-chair.

The Pediatric Brain Tumor Consortium is a federally funded, collaborative clinical trials group based at St. Jude. The consortium includes the National Cancer Institute and seven research institutions.

# Targeting a killer of children

An international team led by St. Jude investigators has released data detailing the effectiveness of nearly 310,000 chemicals against a malarial parasite that remains one of the world's leading killers of young children.

As a result of the research, which appeared in the journal *Nature*, investigators have identified almost two dozen families of molecules they consider possible candidates for drug development. St. Jude researchers have already used one of the molecules to stop the parasite's growth in the laboratory.

"Malaria causes roughly 8 percent of childhood deaths worldwide and remains among the greatest threats to children in the developing world," said the report's senior author, Kip Guy, PhD, Chemical Biology and Therapeutics chair. "At St. Jude, we focus on diseases that kill children, but lack good treatments. That is what drove us to start this work."

Added Dr. William E. Evans, St. Jude director and CEO: "These are the same tools and techniques that we are now using to find new targets and drugs to treat childhood cancer. This work illustrates their enormous power for drug discovery."



# This time, it's personal

Armand Guiguemde, PhD, who hails from Burkina Faso, Africa, nearly died from malaria when he was 9 years old. Recently, he was first author of a St. Jude malaria study that appeared in the journal *Nature* (see related story, above). "When I learned about the principles on which St. Jude was built, I agreed with Danny Thomas' philosophy that no child should die in the dawn of life," said Guiguemde, who joined St. Jude four years ago as a postdoctoral fellow. "Cancer and malaria are both catastrophic childhood diseases. I wanted to come to St. Jude to make a difference in a child's life and have the best opportunity to be involved in science with the potential for translation."

# Skin cancer may indicate future risk

Childhood cancer survivors later diagnosed with nonmelanoma skin cancer may be at increased risk for having a malignant tumor within 15 years, according to research led by St. Jude investigators.

Almost 20 percent of survivors in the Childhood Cancer Survivor Study (CCSS) who had basal or squamous cell skin cancer after radiation therapy developed another more aggressive cancer within 15 years, said Gregory Armstrong, MD, of Epidemiology and Cancer Control.

"These findings suggest non-melanoma skin cancers are a potential marker for survivors who are at risk for future invasive malignancies," he said.

The study involved 14,358 childhood cancer survivors whose cancer was diagnosed between 1970 and 1986. All are now participating in the CCSS, which is funded by the National Cancer Institute to track the health outcomes of childhood cancer survivors. Armstrong is project director for CCSS, which includes investigators at 30 institutions in the U.S. and Canada. St. Jude is the central coordinating institution. Les Robison, PhD, St. Jude Epidemiology and Cancer Control chair, is the CCSS principal investigator and the study's senior author.

# St. Jude in Washington

St. Jude was recently noted by the federal government as a leading example in health care cybersecurity when Clayton Naeve, PhD, senior vice president and chief information officer, was invited to Washington, D.C., to outline the hospital's accomplishments in safeguarding patient and research data.

Chaired by Howard Schmidt, U.S. cybersecurity coordinator, the meeting recognized the progress that has occurred in the field. Naeve was part of a five-member guest panel that discussed successes and challenges in the cybersecurity arena.

"In the past few years, St. Jude has increased our data security staff, increased funding to enhance our data security infrastructure, strengthened our policies and enhanced our employee training," Naeve said. "I was pleased to have the unique opportunity to serve on the panel to represent St. Jude and health care in general."

Prior to Naeve's presentation, President Barack Obama addressed the meeting's 100 participants to emphasize the importance of cybersecurity to the U.S. economy and the need for cooperation to combat cyber threats.



# A tale of two genes

Recent findings in the laboratory of J. Paul Taylor, MD, PhD, Developmental Neurobiology, link two neurodegenerative diseases with the same molecular misstep and trace the problem to two very different genes. The research was conducted using the fruit fly *Drosophila melanogaster.* The study adds a new chapter to the understanding of both amyotrophic lateral sclerosis (ALS) and a rare, inherited disorder known as IBMPFD, which involves widespread, progressive deterioration of muscles, bone, the brain and other tissues. The findings also raise hope that treatments would be effective against both disorders in the future. The work uncovered a previously unrecognized molecular partnership between the genes TDP-43 and VCP. A report on this work appeared in the *Journal of Neuroscience.* 

# Professors of Philanthropy

# BY JANICE HILL

## They taught powerful lessons by example. Today, their children carry on their work.

MARY AND PETER RUSSO SR. taught their children about philanthropy without saying a word.

"We saw our mother do so many things for others, from cooking food to helping a poor family in the neighborhood. And our father was unbelievably giving to family, friends and anyone who needed help," says Nancy Russo McDevitt, the youngest of the Russos' three adult children.

Peter owned an insurance and real estate company in the Lawrenceville, New Jersey, area where he had grown up. Nancy recalls times when some clients could barely pay their premiums.

"He'd say, 'That's OK; give me what you can now, and you can give me the rest later.' He never sat down and told us to help others, but they both led by example," Nancy says. "That was the foundation they created for us."

Mary and Peter are now gone, but they continue to set an example of generosity and compassion through their legacy to St. Jude Children's Research Hospital.

The couple created a Charitable Remainder Unitrust several years ago through the transfer of appreciated stock. Mary passed away in 2006; when Peter died in October of 2009, the trust was terminated, with St. Jude receiving 45 percent of the remaining principle.

Mary Russo loved the story of how Danny Thomas founded St. Jude, and she felt a very strong connection to him and the hospital. Like Thomas, both Mary and Peter grew up in poor immigrant families—Mary's parents were from Austria and Peter's from Italy.

"Our mother stood in ration lines as a child and never forgot where she came from. She would have given the

shirt off her back to help a child. She knew she was lucky to have three healthy children, and she wanted to give back to help others who were not as fortunate," Nancy says.

> Likewise, Peter began supporting his family when he was a young teen. Although that kept him from going to college, he later studied insurance and built a successful business. He related to Thomas' life experiences and his mission to provide hope to sick children regardless of their families' ability to pay.

All three Russo children live by the example of their parents and give back when they can. For instance, Nancy, who owns High Street Grill in Mount Holly, New Jersey, holds annual fundraisers supporting charities,

including St. Jude. Her brother, Peter Jr., who owns The Octopus' Garden, a restaurant in Mayetta, New Jersey, and her sister, Marianne Nahin, who has a real estate business in California, find similar ways to give back.

The Russo children are now fulfilling their parents' wishes to establish a family foundation with a lead trust that will continue benefiting St. Jude and other charities for years to come.

"They built their lives from the bottom up," Nancy says. "Now it is our job to see that our parents' wishes are carried out."

To learn more about charitable lead trusts and other options for leaving a legacy that will help the children of St. Jude, visit www.stjudelegacy.org or call (800) 395-1087.●

# Turning the Tide

Clinicians and researchers chart a course to find cures for high-grade gliomas.

By Elizabeth Jane Walker



s Noah Shumate bounded onto a blustery South Carolina beach last December, he beheld an expansive panorama of sand, surf and seagulls. To the excited 3-year-old, the possibilities seemed as endless as the horizon.

But two days later, his family's idyllic vacation plans collapsed like a sandcastle at high tide.

When Noah's right eye suddenly began to cross, Billie and Richard Shumate rushed him to a local hospital. A scan revealed a tumor deep within Noah's brain. After further testing at another hospital, a pediatric oncologist asked to speak privately with the couple.

"Our world shattered," recalls Billie, who is a nurse. "The doctor said that Noah had an inoperable brain tumor called a pontine glioma. She told us that there was no treatment for it and that Noah had about six months to live." The physician suggested that the Shumates make hospice arrangements for their younger son.

Reeling with shock, Billie spoke to her aunt, who encouraged the couple to have their physician obtain a referral to St. Jude Children's Research Hospital.

"She told me not to give up hope," Billie recalls.

The morning after requesting the referral, Billie phoned St. Jude.

"I didn't give my name or Noah's name to the lady who answered," she says, "But as soon as I mentioned that my son had a pontine glioma, she said, 'Is his name Noah?" My heart began to race, because I knew God was going to take care of us."

## **Turbulent seas**

Noah's brain tumor is a type of high-grade glioma known as diffuse intrinsic pontine glioma (DIPG). High-grade gliomas are malignant tumors that arise from glial cells in the brain or spine. When a high-grade glioma originates in the brainstem, the tumor is known as DIPG. Fewer than 30 percent of children with high-grade gliomas survive the disease. For patients with DIPG, survival rates are even more dismal, hovering below 10 percent.

"Survival is tied to how much of the tumor can be surgically removed," explains Alberto Broniscer, MD, of St. Jude Oncology. Unfortunately, DIPGs are entrenched in the middle of the brainstem and entangled among the nerves.

"If you have a high-grade glioma in another part of the brain, you can remove more of it," Broniscer says. "With a DIPG, you can remove little, if any, of it."

Despite the diagnosis, the Shumates were heartened after meeting Broniscer.

"He was positive and optimistic about the course we were going to take in treating this tumor," Billie recalls. "Dr. Broniscer said Noah would begin a six-week run of radiation



Noah Shumate (at left) is enrolled in a clinical trial led by Alberto Broniscer, MD, of St. Jude Oncology. The study marks the first time the particular drug combination has been tested with irradiation in children and adolescents with diffuse intrinsic pontine glioma. Scans performed earlier this year indicated that Noah's tumor had decreased dramatically.

treatments five days a week. He'd also take two forms of chemotherapy for two years. Hopefully, the radiation would shrink the tumor as much as possible and the chemo would kill off the remaining tumor cells."

# All hands on deck

St. Jude researchers and clinicians have been teaming up to battle high-grade gliomas for years.

"In the past, the standard treatment for DIPG was irradiation—that's all," Broniscer observes. But teams of St. Jude clinicians and laboratory scientists have been focusing their efforts on finding new and more effective treatments for DIPG and other high-grade gliomas. Investigators have been exploring different methods of killing the tumors. These techniques range from treatments that cut off the tumor's blood supply to medications that block the receptors instructing brain tumor cells to divide and grow.

Several years ago, St. Jude opened the world's first clinical trial to use irradiation and a drug called vandetanib to treat children with DIPG. Broniscer and Suzanne Baker, PhD, of St. Jude Developmental Neurobiology are also collaborating on a biologic study of DIPG tumors. The investigators have collected 43 samples of the tumor and are performing molecular analyses on the samples to gain a better understanding of DIPG's genetic make-up.

In another study, Baker, Broniscer and their colleagues recently published the most comprehensive

analysis to date of the genetic mutations driving pediatric high-grade gliomas.

The investigators evaluated DNA from 78 tumors, looking for additions or deletions in the genetic material. Using advanced technology, they scrutinized more than 500,000 points across the genome. For 53 of the 78 samples, scientists also evaluated gene activity and compared the results with data from adult tumors.

"We were hoping to find patterns of abnormalities that happen over and over again in multiple tumors," Baker explains. "That would tell us which genes, when disrupted in a certain way, can make the cell lose control and transform itself from a normal cell into a tumor cell."

# **Uncharted waters**

Although high-grade gliomas occur in both adults and children, investigators found that vastly different mutations drive the disease processes. A gene called *PDGFRA* is crucial in development of the childhood disease, whereas the *EGFR* gene is the most important culprit in the adult form. Baker and her colleagues were exhilarated when they found the link to *PDGFRA*, because a drug called dasatinib had already been developed to target that receptor.

"This is a perfect example of how exciting it is to work at St. Jude," Baker says. "In many places a basic researcher would have difficulty quickly translating a lab discovery into the clinic. But when we found that *PDGFRA* was important and that a drug was already available, I called Alberto Broniscer and Amar Gajjar [co-leader of the St. Jude Neurobiology and Brain Tumor Program] and told them about it. Before our paper was even published, Alberto Broniscer had a clinical trial open to test dasatinib. I don't know of anywhere else where that could happen so efficiently."

In the new DIPG clinical trial, St. Jude clinicians administer vandetanib and dasatinib along with radiation therapy. Tests conducted elsewhere on adults indicated that each of these medications can cause some types of cancer to shrink or stop growing. The hospital conducted a previous treatment combining irradiation and vandetanib in children with DIPG. However, the St. Jude protocol marks the first time this drug combination has been tested with irradiation in children and adolescents.

# Secrets of the deep

Noah's parents enrolled him in the new DIPG protocol in December 2009. Scans performed earlier this year showed that his tumor had decreased by 60

percent. "We were thrilled," says Billie, who realizes that challenges still lie ahead.

Meanwhile, clinicians and researchers at St. Jude continue their quest to better understand high-grade gliomas.

"Every time you answer one question, many new interesting ones come up," Baker observes. "For instance, there are a lot of things we don't know about drugs that target PDGFR: Are they optimal? Will they cross the blood/brain barrier efficiently? Do they need to do that to be effective for DIPG? If a tumor is driven by this abnormal protein, can we merely block the activity of that protein and stop the tumor, or will we need to combine that therapy with something else? What combinations are most likely to succeed?"

St. Jude researchers are now conducting functional studies for *PDGFRA*, which include developing markers to test how well the drug blocks the receptor in laboratory models.

grade gliomas. The rate that technology is moving may accelerate that discovery process.

"In the project we recently completed, we used technology that is still relatively new," Baker says. "This study couldn't have been done five years ago in the same way; it couldn't have been done even three years ago in the same way. We're currently doing a similar analysis on diffuse pontine glioma, and we're using the next generation of technology, which gives more than double the information we had for our previous high-grade glioma study."

In spite of the rapid technological advances, Broniscer emphasizes that progress is usually incremental.

"You know the story of leukemia treatment at St. Jude," he says. "They didn't move from a 10 percent to 90 percent survival rate in a single shot. With every study, we learn a little more about these tumors."

Each discovery provides the basis for another question and yet another discovery. As scientists and clinicians continue their research, they can gain inspiration from patients like 4-year-old Noah, who pursues his own important projects with energy and single-minded enthusiasm. With his condition currently stable, Noah spends his days romping with his dogs, riding his big

## On the horizon

Somewhere amid all those queries lie answers that may offer hope to children and adolescents with high-



St. Jude scientists recently published the most comprehensive analysis to date of the genetic mutations driving pediatric high-grade gliomas. Investigators working on that study included Suzanne Baker, PhD, of St. Jude Developmental Neurobiology (center) and colleagues (from left) Chunxu Qu, PhD, Junyuan Zhang and Barbara Paugh, PhD. "This is a perfect example of how exciting it is to work at St. Jude," Baker says. "Before our paper was even published, Alberto Broniscer had a clinical trial open to test dasatinib [the drug that targets the receptor the researchers identified]. I don't know of anywhere else where that could happen so efficiently."

brother's quarter horse, playing T-ball and swimming in his grandparents' pool. During the summer of 2010, he and his family also vacationed at another beach—a place of blue skies, shimmering sand and a horizon of boundless possibilities. ●

# The St. Jude Factor

# The hospital's Bleeding Disorders Program is an important resource for families and children.

KIDS FALL DOWN. It's a fact of life. Scoop them up, kiss their boo-boos, send them on their way unless the child has hemophilia, a rare disorder in which blood does not clot properly. Then all the rules change.

Just ask Andrea and Miguel Celis. Soon after their son was born, they discovered he had hemophilia. That means if he skins his knee, the flow will not stop on its own. So when 2-year-old Miguel Jr. hit his



Miguel Celis frolics in the spray at a water park. St. Jude staff provide counsel and treatment to help manage Miguel's severe hemophilia.

head last year, Andrea rushed him to a local hospital. A CT scan indicated no internal bleeding, and she breathed a sigh of relief.

A week later, the toddler began to vomit and walk erratically. Two clinic visits that day yielded vastly different diagnoses.

"He has an ear infection," one pediatrician said.

"He has a stomach virus," another physician said.

Hours later, Miguel's condition deteriorated further. Again, the Celises rushed to the hospital. They learned that despite the results of the earlier CT scan, their son had a lifethreatening hemorrhage in his brain.

"We need to do surgery now," the doctor said.

#### THERE IS NO CURE for

the inherited disorder known as hemophilia. Children like Miguel lack one of the essential enzymes necessary to make the blood clot. As a result, hemophilia patients may bleed for an extended time after injury or bleed internally, especially into the joints. Two-thirds of all cases of hemophilia occur when women carrying defective F8 or F9 genes pass the mutation along to their sons.

Approximately 1 in 4,000 to 5,000 males worldwide is born with the most common form of the disorder, hemophilia A. These boys lack sufficient amounts of clotting factor VIII for the blood to clot normally. About 1 in 20,000 boys worldwide lacks adequate levels of clotting factor IX, which results in hemophilia B.

An injury that might be minor for most people may be fatal to a child with hemophilia. Many hemophilia patients require regular transfusions of replacement clotting factor to prevent bleeding episodes. The physical and emotional toll of the disorder can be compounded by the financial burden of purchasing clotting factor: Care for a child with severe hemophilia can cost \$200,000 to \$400,000 per year.

Miguel is fortunate. At least twice a year, he and his family visit the hemophilia clinic at St. Jude Children's Research Hospital. There, he receives the specialized care he requires to lead a normal life. The St. Jude Bleeding Disorders Program is an important resource for families and children affected by the disorder. Directed by Ulrike Reiss, MD, the facility currently treats about 310 children, 70 of whom have hemophilia. The program is in the final steps of the approval process for becoming a Hemophilia Treatment Center (HTC) by the federal government. As one of 140 HTCs nationwide, the St. Jude facility will be eligible to participate in a federal drug pricing program that Reiss says may save the institution \$1 million per year on the purchase of clotting factors.

St. Jude hemophilia patients also have access to the latest clinical

trials, including a study evaluating the extent of joint disease in hemophilia patients and the safety of the nation's blood supply.

Another study explores the use of gene therapy for hemophilia B. Principal investigator of the gene therapy trial is Arthur Nienhuis, MD, of St. Jude Hematology. The approach was developed at St. Jude by Andrew Davidoff, MD, Surgery chair; John Gray of Hematology; and Amit Nathwani, MD, PhD, formerly a visiting scientist in St. Jude Hematology and now at University College, London. The treatment involves inserting the gene for the factor IX protein into liver cells by a one-time peripheral vein infusion. The goal is to restore bloodclotting function and eliminate the need for frequent infusions of factor IX for bleeding.

#### VIGILANCE IS CRUCIAL

for kids who suffer from hemophilia, a complex disease that can have serious complications.

"In children with hemophilia, bleeds can happen with injury, but they can also just happen spontaneously," Reiss explains. "Bleeding frequently occurs in the joints, such as ankles, knees and elbows, because they're exposed to pressure and injury."

Bleeding into a joint can set off a vicious cycle of swelling, pain and chronic inflammation that, in turn, leads to more bleeding. If unchecked, the result can be severe joint damage and a crippling arthritis in which patients have severely contracted joints. "It can happen quickly without treatment, even 6- or 8-yearolds can have knees contracted at 90 degree angles," Reiss says.

Another serious complication of hemophilia is intracranial bleeds. A brain hemorrhage can result in



Ulrike Reiss, MD, directs the Bleeding Disorders Program at St. Jude. Two-year-old Evan Wolfe is one of 70 children currently receiving treatment for hemophilia at the hospital.

debilitation or death. Miguel underwent two brain surgeries after his injury.

"At first, we thought he was going to die or be in a permanent coma," Andrea recalls. "Then we thought he might never walk or talk again. But he recovered quickly. It was a miracle. He's the same Miguel—he's perfect!"

#### A MULTIDISCIPLINARY

team of experienced staff at St. Jude provides the comprehensive care Miguel needs to manage his disease and prevent long-term complications.

Using a holistic approach, St. Jude helps each patient and family deal with the physical, emotional, psychological, educational and other issues associated with a hemophilia diagnosis. Hematologists, nurses and nutritionists implement a treatment plan, coordinate care and train parents in administering drugs and monitoring the child's health. A social worker provides a variety of services that include accessing community resources. Physical therapists provide preventive, rehabilitative and adaptive care. A teacher from the hospital's school program serves as a liaison with the child's school or educates the patient's teachers and classmates about hemophilia. A Child Life specialist uses age-appropriate techniques to minimize stress and anxiety for the patient and siblings. A genetic educator helps families

understand the inheritance patterns of hemophilia. Dental, gynecological and orthopedic services are also available, depending on the patient's specific needs.

"Whenever I have a problem or a question, I call St. Jude, and they help me immediately," says Andrea, who administers clotting factor to her son to prevent bleeding episodes.

Now an active 3-year-old, Miguel leads a normal life. He runs on the playground, pedals riding toys and chases his older sister, Fatima. "When I tell people he has hemophilia, they say, 'No! No, it's impossible,"" Andrea says.

The dark anguish of brain trauma now a distant memory, Andrea envisions a bright future for Miguel.

"I am very thankful," she says. "For me, he was born twice. I love him more and more and more each day. I know that he is special. He is living for a special reason." ●

# Still in the GALLE

# St. Jude helps football standout Shon Coleman tackle ALL.

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BY MIKE O'KELLY

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EAST

was one of the biggest days of 18-year-old Shon Coleman's life—the first Wednesday in February 2010, known to college football followers as national signing day. Flanked by five of his high school teammates, Shon sat in his high school's library at a long, wooden table, dapperly dressed in a suit and tie and proudly wearing an orange Auburn University cap.

Ecstatic coaches, beaming parents, supportive friends and media packed the room to witness the six players sign with their respective schools. With the sound of camera clicks filling his ears, the quiet and introspective high school senior made his decision official—he would continue a 13-year dream of playing football in college and maybe one day professionally. Shon had no reason to think that in less than two months, his football career would be on hold and he would be depending on the world-class treatment and care offered at St. Jude Children's Research Hospital to help him tackle the most common form of childhood cancer—acute lymphoblastic leukemia (ALL).

#### ALL-star talent

Shon's mother, DeKeishia Tunstall, says she never really worried about her son getting hurt from playing football. Always one of the bigger players, he had developed a stoic toughness that benefited him as he matured and as play became more aggressive.

Shon says he enjoys the contact and the knowledge gleaned from playing the game. A skilled left tackle on the offensive line, Shon was responsible for halting opposing defenders in their desperate attempts to bring down the quarterback from the blind side. Shon showed exponential improvement each year of his high school career. He grew to 6-foot-7, 285 pounds by his junior season and was highly sought by some of the nation's best college football teams. As a senior, he was the top-rated high school football player in Mississippi and the No. 3 offensive lineman in the nation. Shon was also selected to play in several prestigious high school all-star games.

During Shon's senior season, a mysterious lump appeared on his head. Accustomed to scrapes and bruises, he and his family dismissed it as a minor football injury.

"We thought he had hit someone or something wrong, and then another lump popped up," DeKeishia says. "We were thinking maybe he was just getting hit in practice, and it wasn't healing well."

#### A game changer

As his senior year of high school progressed, more lumps developed on Shon's neck, chest and torso. His pediatrician referred Shon to a dermatologist for cosmetic removal of what he believed to be harmless scar tissue. In January 2010—just weeks before signing with Auburn— Shon underwent an ultrasound. The lumps, which contained blood vessels, would need to be removed by a plastic surgeon to prevent scarring.

Shon's hectic schedule delayed the surgery until March. He and his family were told the operation would be a simple removal of benign tumors composed of fatty tissues. However, during the biopsy, the surgeon was slightly concerned about one of the masses. A week later, the surgeon's suspicions were confirmed—the lumps were malignant. Clinicians believed Shon had lymphoma, a cancer that originates in the lymphatic cells of the immune system.

"Shon's reaction was far better than my reaction," DeKeishia says. "He wasn't sad. He didn't cry or anything like that. It was like he immediately knew he'd have to fight. He knew he would get through it, and that's the way he comforted me."

While showing a determined resolve to fight the cancer, Shon admits he questioned the idea of whether or not he would ever suit up in a football uniform again.

"I was shocked," Shon says. "But at the same time, I was worried about everything—about my football career and everything else."

After being referred to St. Jude by the plastic surgeon, DeKeishia, Shon and his stepfather, Travis Tunstall, drove to the hospital's campus.

#### **Maintaining focus**

With only seven weeks elapsed since the celebratory day in early February, Shon found himself in a hospital room at St. Jude undergoing a blood test to confirm the diagnosis of lymphoma. However, within an hour of his arrival, the test revealed that he had acute lymphoblastic leukemia (ALL), a cancer of the white blood cells known as lymphocytes. The following day, a bone marrow aspiration, which involves removing bone marrow from the hip bone, and a lumbar puncture—also known as a spinal tap—confirmed the diagnosis and details about the subtype of ALL.

Shon's attending physician, Deepa Bhojwani, MD, of St. Jude Oncology recalls the first discussion she had with Shon, his mother and stepfather.

As a high school senior last year, Shon was the top-rated high school football player in his state and the No. 3 offensive lineman in the nation. Then doctors discovered that he had acute lymphoblastic leukemia.

"All three of them were focused. They wanted to figure out what the problem was and deal with it," Bhojwani says. "Shon didn't say much at the initial meeting, but he was definitely listening and processing what I was saying. I'm sure getting back to the football field was the biggest question on his mind, but he didn't bring it up at that first meeting."

Bhojwani brought up the subject of football, assuring Shon that treatment was a step-by-step process that would proceed depending on how he responded to chemotherapy. She also pointed out during the initial meeting that the treatment of ALL had advanced greatly in the last four decades thanks in large part to St. Jude. When the hospital opened its doors in 1962, the five-year survival rate for children with ALL was 4 percent. Today that number is 94 percent.

Shon was placed on the Total XVI clinical protocol for newly diagnosed leukemia patients up to age 18. Treatment involves three stages: induction, consolidation and continuation. The induction stage requires the most intensive chemotherapy.

"Changes are made according to how a person responds to therapy," Bhojwani says of the protocol. "We have sensitive methods of measuring this response by minimal residual disease." Minimal residual disease is the small number of leukemic cells that may survive remission induction therapy.

Shon responded remarkably well to treatment, entering remission with negative minimal residual disease after two weeks. He has completed the first few months of the last phase of treatment, which involves two years of weekly chemotherapy.

#### **Staying strong**

With his football career delayed, Shon maintains his strength by working out and by going through physical therapy exercises with the St. Jude Rehabilitation Services department. Because of the central line in his chest, he is unable to perform some upper body exercises, but he keeps fit through cardiovascular exercise and moderate strength training. The coaching staff at Auburn has saved Shon's scholarship, and his future teammates have reached out to him to show support.

"We have gotten thousands upon thousands of letters and e-mails," DeKeishia says. "People from all across the country have reached out just to say they are thinking about him."

Bolstered by the outpouring of support, Shon is again focused on returning to his dream of being involved in the sport he loves. Whether that results in playing professional football one day or coaching a budding group of new stars to their dreams, Shon is grateful to St. Jude for the opportunity to be in the game.

"I knew St. Jude would take care of me and everybody would do their best to help me get back to where I need to be," Shon says. "You've got so much hope here, and everybody here is so good to me." ●



Deepa Bhojwani, MD (at right), says Shon is responding well to treatment. With his disease in remission, he is currently undergoing the final phase of treatment, which involves two years of weekly chemotherapy.



Staff in St. Jude Rehabilitation Services work with Shon to ensure that he maintains his strength while undergoing chemotherapy. He keeps fit through cardiovascular exercise and moderate strength training.

# Savoring Success

## By Elizabeth Jane Walker

They've boosted AML survival to an all-time high, but St. Jude investigators hunger for even sweeter statistics.

Martin perched on uncomfortable plastic chairs. Cradling their feverish infant, the couple glanced upward at a movie on TV.

Through the din of conversations and clattering equipment, they listened as Forrest Gump shared his familiar park-bench philosophy.

"My momma always said, 'Life was like a box of chocolates,'" Gump drawled. "'You never know what you're gonna get."

Seemingly on cue, a doctor strode into the cubicle and informed the couple that their daughter did not have the urinary tract infection they originally suspected. She had leukemia.

Five months later, Lindsey still winces at the timing of the announcement. "It seemed so unreal," she recalls.

# Hard to stomach

During the preceding weekend, 3-month-old Adele Litchfield had exhibited a mild eye infection, a worrisome rash and a fever that would peak and then recede. "Every time her temperature went down, we'd think, 'OK, it's passing,'" Nate recalls.

In reality, it would be their first taste of an extremely serious illness.

Adele had acute myeloid leukemia (AML), a cancer of the blood and bone marrow. Although nearly 12,000 new cases of AML occur in the United States each year, only about 500 of those are in children and adolescents.

AML originates in cells that normally develop into white blood cells. Healthy infants have fewer than 15,000 white blood cells per microliter of blood. Adele had more than 250,000. "We need to act fast," the ER physician told the family. "We're going to get you in an ambulance and send you to St. Jude Children's Research Hospital."

Soon after Adele arrived in Memphis, the hospital announced



Eight-month-old Adele Litchfield listens happily as her parents chat with Jeffrey Rubnitz, MD, PhD, of Oncology, "The AML protocol is tough," Adele's dad admits, "But it has been like a miracle. St. Jude saved her life."

the results of its most recent AML clinical trial. In a study conducted at St. Jude and six other hospitals, investigators had pushed the pediatric AML survival rate to 71 percent three years after diagnosis—a rate 20 percent higher than previously reported U.S. rates.

During the preceding years, the overall survival of St. Jude AML patients had risen from 33 percent in the 1980s to 55 percent in the 1990s. In 2000, scientists began designing a clinical trial that they hoped would boost the rates even further.

Known as AML02, the study became the hospital's first multiinstitutional leukemia trial. In addition to obtaining large amounts of patient data, the collaboration allowed St. Jude clinicians and laboratory scientists to create and enhance relationships with their peers at other cancer centers.

## On the menu

One chief limitation in planning AML02 was the scarcity of new medications to fight AML. Drug development is a slow and laborious process—a fact that is exacerbated by the rarity of the disease in children.

"We were limited in what we

could do in terms of new therapies, because only one new agent had been developed that we felt comfortable using in children," recalls Jeffrey Rubnitz, MD, PhD, of St. Jude Oncology.

The team optimized the use of conventional drugs and added a new medication for certain patients who did not respond to traditional therapy. Investigators also evaluated genetic information to determine the risk factors for each child.

After each child completed the first or second course of chemotherapy, Dario Campana, MD, PhD, of St. Jude Oncology and Pathology and his colleagues assessed the patient's response to therapy based on the presence of minimal residual disease, or MRD. If scientists found even one leukemic cell among 1,000 normal bone marrow cells, clinicians immediately adjusted treatment in an effort to prevent relapse. In previous trials, clinicians had looked at MRD retrospectively, but in this study, samples were sent to Campana's lab and the results were used to make treatment decisions. As a result, this was the first AML trial to use MRD in real time.

Worldwide, many children have

succumbed because of the intensive therapy required to eradicate AML. In the St. Jude clinical trial. investigators focused on improving and optimizing supportive care in hopes of decreasing toxicity, infection rates and inpatient stays. One tactic they used was to administer antibiotics after each course of chemotherapy. In the past, patients had

received these medications only after developing fever or other signs of infection.

"This is the first time preventive antibiotics and antifungal agents have been used in a pediatric AML trial," Rubnitz says. "We found that it decreased admissions to the ICU, decreased infection rates and improved quality of life. We're very happy with how well it worked. As a result, we're also using this technique with some of our children after they undergo intensive rounds of chemotherapy for acute lymphoblastic leukemia."

# Insatiable investigators

Tasting success, St. Jude investigators immediately began hungering for even higher survival rates.

AML is difficult to eradicate because it begins with an immature cell known as a stem cell. Originating in the bone marrow, stem cells are designed to survive throughout a person's lifetime. Because the cells grow slowly, they are unaffected by drugs that disrupt the cell cycle and cell division. These hardy cells are also resistant to natural toxins. AML stem cells share these attributes;

In the most recent St. Jude AML clinical trial, investigators pushed the pediatric AML survival rate to 71 percent three years after diagnosis—a rate 20 percent higher than previously reported U.S. rates.

they divide slowly and are adept at pumping out toxins and drugs.

"It's hard to kill leukemic stem cells," Rubnitz says. "Intense therapy designed to kill them may also kill the healthy stem cells. So we need to devise ways to kill the leukemic stem cell but not the normal stem cell." Rubnitz says researchers are making progress.

The hospital's newest protocol, AML08, compares a new drug combination with the conventional medley of medications used to combat AML. Scientists hope the new chemotherapy regimen will enable them to avoid using one particular conventional chemotherapy agent that can cause heart damage. The new protocol is also exploring new markers and methods for detecting MRD.

AML is a disease with many different subtypes. Ideally, scientists would like to devise a specific therapy for each. For instance, some St. Jude patients have mutations in a gene called *FLT3*, which carries a survival rate of about 30 percent. The new St. Jude clinical trial uses a drug that has been shown to inhibit the FLT protein. "It's early, but thus far the drug seems to be effective for this particular subtype of leukemia," Rubnitz says.

# Delicious discovery

The AML08 protocol also incorporates a new form of cellular therapy. In the previous clinical trial, high-risk patients underwent stem cell transplantation. Although this treatment greatly reduces the risk of relapse, it is potentially toxic and depends on donor availability. Currently, St. Jude clinicians recommend transplant only for highrisk AML patients. For some other patients, investigators are now using another form of cellular therapy called natural killer (NK) cell therapy. Scientists have discovered that NK cells are particularly effective at killing AML cells.

St. Jude recently conducted a pilot study testing the safety of NK cell therapy. Clinicians used this treatment on 10 children who were in remission from AML. The therapy consisted of a mild conditioning regimen to suppress the immune system and an infusion of natural killer cells from a parent.

"There was very little toxicity; actually it was much milder therapy than a course of AML chemotherapy," Rubnitz says.

All 10 patients responded well and are still in remission. As a result, a standard-risk AML patient may now receive chemotherapy as well as an infusion of natural killer cells from a parent. Investigators hope this technique will eliminate any leukemia cells that might be resistant to chemotherapy.

## Just desserts

Rubnitz attributes the success of the hospital's AML studies to a seamless collaboration between the laboratory and the clinic, as well as the treatment team's expertise. "Everybody in this institution is an expert in taking care of children with cancer," he says. "From data managers to nurses and nurse practitioners, from postdocs to physicians, this hospital is devoted to pediatric cancer. Our supportive care here is excellent. Our transplant team is outstanding. We have great people and great resources."

Children like little Adele Litchfield are the direct beneficiaries of that specialization. As soon as she and her parents arrived at St. Jude, the staff leapt into action.

"Everything happened so fast," Nate recalls. "Almost immediately, they started doing amazing things replacing her entire blood count twice and starting treatment the next day."

As they contemplate the past few months—beginning with Forrest Gump's dogma and continuing through Adele's remission—Lindsey and Nate are thankful that their box of chocolates has included St. Jude.

"The AML protocol is tough," Nate admits, "But it has been like a miracle. St. Jude saved her life." ●



Thanks and Giving

Holiday shoppers. National corporate partners. Celebrities. They're all uniting to raise funds during this year's *Thanks and Giving* campaign.



National Outreach Director Marlo Thomas kicks off *Thanks and Giving* with the help of St. Jude patients (back row, from left) Christy Clarkson, Jason Kresge, Ana Mendez-Villalba, Lea Lawrence and (front row, from left) Anna Pike, Gloriuna Hamilton, Haley Hines, Zowie Kile and Clinton Milliken.

### By Amanda McGee Robbins

ore than seven years ago, Marlo, Terre and Tony Thomas, the children of St. Jude founder Danny Thomas, had an idea: Why not harness the power and generosity of consumers during the holiday season to create a unique fundraising campaign and share the St. Jude story nationwide?

Out of that idea came the *Thanks and Giving*<sup>®</sup> campaign, an unprecedented union of corporations, retailers, celebrities and the media that asks consumers to "Give thanks for the healthy kids in your life, and give to those who are not." Since 2004, caring consumers across the country have embraced this heartwarming call to

action, helping to make the campaign a holiday tradition.

This year marks the seventh year of *Thanks and Giving*, and more than 50 companies will be encouraging their customers to give to St. Jude through add-ons at the register or by purchasing specialty merchandise. These dedicated partners—including Kmart, CVS/pharmacy, Kay Jewelers, Target, Dick's Sporting Goods, Williams-Sonoma, AutoZone and many more—are ready to repeat the success of previous campaigns when *Thanks and Giving* kicks off November 22, the start of the country's busiest shopping season.

Among the new partners joining *Thanks and Giving* this year in the fight against childhood cancer and other

catastrophic diseases are Tumi, Old Navy, HomeGoods, Cornerstone Brands and Expedia.

# Sharing the mission

In addition to raising much-needed funds to help St. Jude find cures and save children, *Thanks and Giving* helps people nationwide learn about the hospital's cuttingedge research and treatment. The messages are shared through print, broadcast and Internet ads. Celebrity friends Jennifer Aniston, Robin Williams, George Lopez and Morgan Freeman will join Marlo Thomas and the biggest stars of all—St. Jude patients—in television spots and a movie trailer. This year, Olympic gold medalist Shaun White and NBA player Dwayne Wade join the St. Jude family of celebrities.

*Thanks and Giving* also continues to reach out to the Hispanic community. Popular Puerto Rican musician and actor Luis Fonsi is again sharing the St. Jude mission through the Spanish-language media. Joining him this year is Angelica Vale, a noted Mexican actress, singer and comedienne.

St. Jude National Outreach Director Marlo Thomas will kick off *Thanks and Giving* during Thanksgiving week, when she will appear for five consecutive days on NBC's *TODAY* show. Thomas will also share the lifesaving work of St. Jude through the national media, as she has during past campaigns, by appearing on *The View*, *Larry King Live, Bloomberg Night Talk* and *Nightline*. Adding to the excitement of these media appearances is the recent release of Thomas' new book, *Growing Up Laughing: My Story and the Story of Funny*, which is available in bookstores and online now.

Beginning in November, holiday shoppers will see the *Thanks and Giving* magnifying glass logo displayed at stores across the country and online, signaling retailers who are helping St. Jude in the battle to save children's lives. Employees at these companies will rally to raise as many donations as possible to support the hospital.

In 2009, Kmart associates asked for donations of \$1, \$5 and \$10 at the register at more than 1,300 locations in the United States, Puerto Rico and Guam. Their dedicated efforts paid off, and Kmart raised nearly \$6.9 million, setting a new record for *Thanks and Giving* fundraising.

On Saturday, November 20, thousands of St. Jude supporters in more than 60 communities nationwide will unite to participate in Give thanks. Walk.<sup>™</sup>, an exciting, family-friendly 5K walk. Participants are encouraged to form walk teams with their friends, family and co-workers and recruit sponsors to make donations in support of their efforts and St. Jude. Visit *www.givethankswalk.org* to find an event near you.

A new, interactive feature on the *Thanks and Giving* website encourages individuals to share virtual gifts with friends and families on the popular social media site Facebook while making a donation to support the children of St. Jude. Individuals can also share stories of their donations on Facebook while asking others to also support St. Jude.

# A beacon of hope

For families in communities everywhere, St. Jude is a place of hope, a place where pioneering science, medicine and patient care are making remarkable strides against deadly childhood diseases. During the holidays, through the *Thanks and Giving* campaign, more and more families become aware of how the work of St. Jude is changing the way the world treats childhood cancer.

"The *Thanks and Giving* campaign is an astonishing effort by so many people to help support St. Jude," says Richard C. Shadyac Jr., CEO of ALSAC, the fundraising organization of St. Jude. "This huge effort is made possible by the dedication of all those involved from sales clerks who ask for donations and corporate presidents who commit their companies to the St. Jude mission, to celebrity friends who give their time and talent. And at the heart are the holiday shoppers, whose amazing generosity and compassion reflect the true spirit of the season."

# How can you help?

To support the children of St. Jude, look for the *Thanks and Giving* magnifying glass logo wherever you shop this holiday season. *Thanks and Giving* partners include Kmart, CVS/pharmacy, Kay Jewelers, Target, Dick's Sporting Goods, Williams-Sonoma, AutoZone and many more.

Learn more about the *Thanks and Giving* campaign by visiting *www.stjude.org.* There you will find a complete list of *Thanks and Giving* partners, with details about where to shop and how to donate. You will learn how you can share your own donation with others via social media, and you can find out how to purchase special virtual gifts for the loved ones on your holiday lists.

Delta Air Lines is the premier travel partner of St. Jude Children's Research Hospital.

# BY THE **NUMbers**

She may be one in 3 million, but for **Ryan Christian** only three things really matter—being strong, healthy and loved.

orie Christian likes to tell people that her daughter is one in a million. Actually, that's an extremely conservative estimate—but who's counting?

Although her dad is the professional athlete in the family, 8-year-old Ryan Christian has amassed some impressive stats of her own. She's an award-winning artist, an accomplished ice skater and an avid animal lover. A fashion consultant, soccer player and social butterfly. A gymnast, honor student and basketball player. And she pulls it all off with uncommon grace and enthusiasm.

During a checkup at St. Jude Children's Research Hospital, Ryan and her parents rattle off the names of the medications she takes. Talk about numbers: The third-grader requires about 40 pills each day as part of treatment for an extremely rare form of cancer known as adrenocortical carcinoma (ACC). Although the overall incidence of the disease is one in a million, the vast majority of those patients are adults. Only 20 to 25 children in the U.S. are affected each year (an incidence of one in 3.3 million children under 14 years of age). But Ryan and her parents, Jeff and Dorie, have reason for optimism. After all, they have come to a hospital that is internationally known for the research and treatment of ACC.

## Eight long days

*Eight days.* That's how long it took staff at a hospital in Missouri to determine why an otherwise healthy and active girl had suddenly begun suffering from headaches, seizures and dangerously high blood pressure. "Don't even worry," a doctor told Ryan's parents in February 2010, when scans indicated a mass. "It would be rare for it to be cancer."

But the tumor that originated on Ryan's adrenal gland and extended up toward her heart and lungs was malignant. The medical staff's response did not inspire confidence. "We may be too late already," their surgeon admitted. "We saw one case of this cancer two years ago," another doctor said.

As Ryan recuperated from an operation to remove her tumor, one of Jeff's friends offered welcomed counsel.

"You have to go to St. Jude. It's the best place in the world for kids with cancer," he said.

Dorie, an attorney, and Jeff, a professional hockey player, immediately went online to look for information about the hospital.

# BY ELIZABETH JANE WALKER



Michaela Shurden (at left), Ryan Christian's St. Jude schoolteacher, helps the third-grader continue making academic progress while undergoing treatment for adrenocortical carcinoma. "Miss Michaela continues to encourage Ryan to achieve her potential," says Ryan's mom, Dorie. In addition to services such as regular school lessons, Dorie says her family appreciates "the positive attitude, the hope, the children running around having fun and the way that they make us feel that anything is possible."



"We have a long history of studying this disease in children," says Lisa McGregor, MD, PhD (pictured here with Ryan, Jeff and Dorie Christian). "As a result of our interest, this hospital is one of the international centers of expertise in this tumor in children."

"We learned that St. Jude had done a lot of research on the p53gene," Dorie explains. "Patients with p53 mutations are at risk for a variety of tumors, and one of those is ACC. We also learned that St. Jude has a special protocol they use for Ryan's form of cancer. Although we were told we could receive the same treatment in Missouri, we knew that it had been developed at St. Jude. 'We need to go where they specialize in this,' we said."

### Twenty years of experience

In Memphis, the Christian family felt an overwhelming sense of relief. "After a month of holding our breath, we felt like we could breathe," Jeff says. "Once we got here, we felt good about everything. They've beaten this cancer before, they're optimistic and we have an amazing doctor."

"Even our social worker, Judy Hicks, is amazing," Dorie adds. "She has been there every step of the way. This summer, Judy was awarded the national social worker of the year award. Since St. Jude was recently named the No. 1 children's cancer hospital by *U.S. News & World Report,* we tell people that we have the No. 1 social worker at the No. 1 children's cancer hospital in the world."

Lisa McGregor, MD, PhD, of Oncology says the level of experience available at St. Jude is unsurpassed.

"We have a long history of studying this disease in children," she says. "Dr. Raul Ribeiro and Dr. Gerard Zambetti have collaborated for many years with researchers in Brazil because this tumor occurs more frequently there. The International Pediatric Adrenocortical Tumor Registry is based right here at St. Jude. As a result of our interest, this hospital is one of the international centers of expertise in this tumor in children. Ryan is being treated on a national Children's Oncology Group study that is led by investigators from St. Jude and came

out of research that was done here." (See related sidebar on next page.)

Although most children with ACC have a mutation in the p53 gene, genetic testing has revealed that Ryan does not have such a mutation.

"This is very good news for Ryan's future," Dorie observes.

Children with a *p53* mutation have an increased risk of developing another cancer compared to children without a mutation. Since Ryan does not have the mutation, her risk of developing a second cancer is no greater than that of any other pediatric cancer survivor.

"We do understand that this disease has a dismal prognosis, but our Ryan seems to be in the positive minority," Dorie says, "and we couldn't be more grateful."

## Three simple words

During a regular clinic visit one summer afternoon, Ryan is clad in a flamboyant cowgirl costume, replete with white satin shirt, gold fringe and faux cowhide chaps. Swinging silvertipped leather boots over the side of an exam table, she removes her sparkly red cowgirl hat.

"I have some curl action going on over here," she announces to McGregor, proudly pointing out the wisps of strawberry blonde hair that have appeared since her last chemotherapy treatment. She then proceeds to compliment the oncologist on her choice of clothing.

"I have an 8-year-old fashion consultant," McGregor says, with a laugh.

The protocol for stage IV ACC is brutal, but Ryan endures it with courage and flair. On the day she is scheduled to begin her eighth and final round of chemotherapy, she chooses an ensemble befitting the momentous occasion—a shimmering fairy costume that features a filmy tunic, diaphanous wings and a garland of flowers encircling her head. When clinicians discover that her blood counts are not robust enough to begin chemo that day, she sheds a tear, knowing that the delay means she will not return home as soon as originally anticipated. A few days later, she finally begins the treatment.

Thus far, Ryan is doing well. "We see evidence that the tumor in her lungs is responding to treatment and that she's tolerating chemotherapy," McGregor says.

The Christians admit that excellent medical care is only one of the reasons they love St. Jude. "It's not just the medicine and the research," Dorie says. "It's the positive attitude, the hope, the children running around having fun and the way that they make us feel that anything is possible."

That optimism permeates every moment of the family's day. As predictable as a good-night kiss is their bedtime mantra. Each evening, Dorie and Jeff repeat three simple words to their little one-in-a-million girl: "Ryan," they tell her as she snuggles beneath the covers, "You are strong, healthy and loved." ●



Ryan Christian visits with FOX Sports on-air personalities Joe Buck (at left) and Troy Aikman while filming a public service announcement at St. Jude. "We feel so privileged to be asked to represent St. Jude in this way," Dorie says. The public service announcement will air throughout the football season.

# Global Collaborations

C linicians and scientists at St. Jude were studying adrenocortical carcinoma (ACC) long before Ryan Christian was born. This year marks the 20th anniversary of the International Pediatric Adrenocortical Tumor Registry, which was developed by St. Jude to provide a centralized repository for data and tumor samples.

"When we began studying this disease, oncology textbooks included about one paragraph on it," recalls Raul Ribeiro, MD, St. Jude International Outreach Program director. "Nobody else was interested in developing protocols because of the rarity of this tumor."

Ribeiro had previously identified an area in southern Brazil where the ACC incidence was 10 to 15 times higher than in other parts of the world. "We decided to create the registry so that we could collaborate with researchers in Brazil and other countries and start obtaining detailed information on this cancer," Ribeiro explains.

Since its creation in 1990, the registry has accrued more than 315 patients. Investigators use information gleaned from this repository to better understand the disease, its outcome and risk factors. Clinicians also use data from this registry to design studies and to determine the best treatment approaches.

St. Jude scientists use the tumor samples and related data to conduct molecular studies and pinpoint the genetic alterations that cause ACC. Researchers at St. Jude have identified specific mutations in the *p53* gene that increase the predisposition for ACC but not for other cancers. Gerard Zambetti, PhD, of Biochemistry has created an adrenal tumor model in the laboratory that scientists can use to test drugs for ACC. In addition to developing a gene array expression that predicts which patients will relapse, Zambetti and his colleagues are also trying to better understand the mechanisms that cause adrenal tumors to form in the first place.

Ninety-five percent of ACC patients in southern Brazil carry a germline mutation that increases their predisposition to cancer. St. Jude is currently partnering with a group in Brazil to look at the long-term side effects of therapy and to determine the incidence of secondary malignancies.

As a result of research conducted at St. Jude, the Children's Oncology Group has created a multi-institutional treatment program that studies the biology of the tumors and the incidence of the different types of *p53* mutations. Ryan Christian is participating in that ongoing study.

# Perspective

# By Don Germano

# Lessons in Empathy

"You think your life is complicated? Just imagine what these families go through."

This fall, many all-star companies will once again help St. Jude Children's Research Hospital raise money for treatment and research through the *Thanks and Giving* campaign. The customers who support this effort are absolutely amazing. Although many of them may be struggling financially, they're willing to give to help a great cause. I'll also be doing my part to make *Thanks and Giving* a success. For me—like many other Americans the decision is personal.

My mom fought an on-andoff battle with cancer for years.

Eventually, cancer won-a loss that

was devastating for my family. Because of that experience, I want to help in any way that I can to eradicate cancer. I also have three children, ranging in age from 11 to 18. As a dad, I listen to St. Jude moms and dads talk about their own battles and I think, "I want to do anything I can to help families and kids not have to go through this."

I've had the opportunity to visit St. Jude and to meet some of the children, the families, the researchers, the doctors. I'm absolutely blown away by the courage and the hope I see there. Several families have shared their stories with me. These are people whose lives were good; everything was going along great. Then cancer struck, and their worlds turned upside down. The families have told me how fortunate they feel that their doctors recommended St. Jude. And then they told me how they've been treated by the doctors and the researchers—



how the whole family, not just the sick child, is cared for during a very scary time of need.

St. Jude not only offers hope to the patients, but it helps the siblings as well. A cancer diagnosis is a big deal for siblings. While the focus is on the sick child, the siblings are suffering, too. Families have told me over and over how St. Jude does everything possible to make the experience as stress-free as possible for the entire family.

Stories like that are a real credit to the integrity of the people running the hospital and to Danny Thomas and his children for working so tirelessly to make his dream a reality. I can cite many compelling reasons to support this

hospital. There's Danny Thomas' mantra that no child should be turned away regardless of a family's ability to pay. There's the hospital's focus on the child and the family. There's the fact that St. Jude shares its research so that children around the world can benefit. And there's the knowledge that kids are our future.

I listen to these families' stories and I know it's my responsibility to pitch in and help out any way I can.

You think your life is complicated? Just imagine what these families go through...then do your part to help them.

Don Germano is senior vice president of operations at Dick's Sporting Goods Inc., which has raised more than \$7.8 million for St. Jude during the past three years. ●

GIVE THANKS FOR THE HEALTHY KIDS IN YOUR LIFE, AND GIVE TO THOSE WHO ARE NOT.

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# CHANGE SERVICE REQUESTED

# Joining forces against sickle cell disease

St. Jude supporters Darrell Byrd (left) and Jim Byrd discuss the latest progress in the treatment and research of sickle cell disease with Jane Hankins, MD, of St. Jude Hematology at a recent reception hosted by the two brothers. The Byrds are leading efforts to raise awareness and support for the hospital's research of the debilitating and potentially deadly genetic blood disorder that affects millions of children and adults worldwide. Hankins provided an update on the promising work underway at St. Jude, the largest pediatric sickle cell research center in the United States.



# Finding cures. Saving children.