Hello Again From Seattle

Hello again from Seattle and thank you. Many of you called or emailed us since you received our first newsletter, and we appreciate your comments and suggestions. This second newsletter has been designed to provide some advice, explain some of the mailings we send to you, and introduce you to two members of staff many of you have talked with on the phone. Sarah Christie has interviewed many of you regarding pregnancies. Bobbi Benson talked with several hundred participants and family members during an interview study funded by the American Cancer Society. These important interviews resulted in an article described in the enclosed article by Dr. Judy Felgenhauer, the principal investigator for the project. The information you provide to us by mail or phone has and will result in important findings.

So, thank you for the contributions you make through your participation. The information you provide will help children diagnosed in the future with Wilms tumor to benefit from findings about improvements in treatment and long-term health. They will owe you immeasurable thanks, as do we.

B

etween 1995 and 1997, we interviewed 296 families to determine if members of the families of Wilms tumor patients have a greater than expected incidence of cancer before the age of 55. We appreciate the participation of many of you who were randomly selected to be contacted for this study and we also appreciate the participation of your relatives who willingly shared their medical histories and records with us to advance our knowledge of Wilms tumor.

We attempted to contact 530 patients (and their families) treated on National Wilms tumor studies (NWTS) between 1970 and 1992. We received permission to contact these families from their doctors at the hospitals where they were treated for Wilms tumor. We then sent letters to the families inviting them to participate in our study. Only 64 families either refused to participate or never responded to our attempts to contact them.

A telephone interview was conducted with the patient or most commonly, the patient’s mother. We requested information on the NWTS patient and the patient’s parents, offspring, siblings, half-siblings, grandparents, aunts, and uncles. We asked about dates of birth and death, birth defects, and occurrence of any cancer prior to the age of 55 in each of these family members. If a cancer was reported, we asked to speak with that relative. A separate interview was then carried out with this person and, with their permission, the cancer was verified with medical records.

Using this approach we were able to verify there were 99 cancers in the 4,258 members of the 296 families we interviewed. Using information from a national cancer registry called Surveillance, Epidemiology and End Results (SEER), we could predict the number of cancers we should have seen in this population, based on the ages of all the relatives.

We evaluated the results for the whole group and then for specific subgroups. In the group as a whole we would have expected to find 127 cancers. Since we only found 99, there were fewer cancers than expected. This

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Study of Cancer Risk in Other Family Members of Wilms Tumor Patients (continued from page 1)

suggests that extended families of Wilms tumor patients are not at a higher risk than other people for cancer. However, there were some subgroups that were difficult to evaluate because of their small numbers. These included the parents of former Wilms patients. They had more cancers than expected but because of the small numbers the difference is within the limits of chance fluctuations. Your continued participation in the National Wilms Tumor Long Term Follow-up Study will allow us someday to better answer questions such as this.

If you would like to read the paper discussing this study, it is entitled “No Excess of Early Onset Cancer in Family Members of Wilms Tumor Patients” and was published in the September 15, 2001 issue of Cancer, Vol. 92, No. 6 and a summary is available on the NWTS web site: http://www.nwtsg.org.

Please find below an article from our colleagues at the Minnesota Long Term Follow-Up Study (LTFU.) Some of you are participating in both our study and theirs, and we are pleased to have this opportunity to provide all our participants with information about them. We know that your time is extremely valuable and want you to understand why you might be asked to participate in more than one study. We want to emphasize that the studies cooperate to use as little of your time as possible.

Our study focuses exclusively on patients with Wilms tumor and includes all survivors of the National Wilms Tumor Study from North America who agree to participate in it. It uses the largest, most comprehensive cohort of patients ever assembled, over 8000 since 1969. The Minnesota Long Term Follow-Up Study includes patients from selected institutions in North America with all types of childhood cancer. It studies a much smaller number of patients with Wilms tumor.

Minnesota Long Term Follow-Up Study

By: Catherine Moen, LTFU Newsletter Editor

With current therapies, over 70% of children diagnosed with cancer and similar illnesses can be expected to become long-term survivors. The Long Term Follow-Up study was begun to address the needs of these individuals, many of whom have grown to adulthood and now face the likelihood of experiencing delayed effects from the treatment of their illness.

The LTFU is a study of children and adolescents who survived five years following the treatment for cancer or similar illnesses. Participants were diagnosed between 1970 and 1986 at one of 25 collaborating research centers. The National Cancer Institute has funded the LTFU study since it started in 1994. The study is coordinated by the University of Minnesota.

More than 14,000 survivors and about 3,500 siblings of survivors actively participate in the LTFU study. The siblings serve as the study’s comparison group. (Of interest to readers of the NWTS newsletter, the study includes 1,213 participants who were diagnosed with kidney cancer.) Participants provide information to study researchers about their health and health-related behaviors through written and telephone questionnaires. They are also asked to give permission for the study to obtain medical records of their diagnosis and treatment for their original illness. In addition, the LTFU study collects certain biologic specimens, which are used to study genetic factors of cancer treatment.

The mission of the LTFU is to investigate the long-term effects of treatment and to provide health-related education to its participants. Study investigators have reported on several important topics, including early and excess mortality among survivors, the occurrence of second cancers, and the psychological health of study participants. A recent paper reported on the finding that cancer survivors lack specific knowledge about their treatment for their illness. (This can make it hard for survivors to obtain appropriate follow-up medical care.) The study provides research updates and other health information to participants in a newsletter that is published twice a year. Additional information about the LTFU study, including copies of the newsletters and the study questionnaires, can be found at the following web site: www.cancer.umn.edu/ltfu.
My son is only 14, why should I return the Pregnancy Survey?

Many of you have asked us this question. Some parents do not understand why we are asking for this information for children under 18 years of age. Pregnancies are occurring at ever younger ages. Also, some young men think this is a question only for young women, although adolescent males have become fathers. Others think that there is no reason to return a survey when they have never been pregnant or fathered a child. Some parents believe it is irrelevant because their child is a young teenager. Have you perhaps not returned the survey because you had similar thoughts? If yes, please reconsider.

For our study of pregnancies it is important that we hear from as many participants as possible, both men and women. If we send out 4000 surveys and only 2000 are returned, we do not know what it means if 500 pregnancies are reported. The simple calculation is 500/2000=25%. Should we then believe that 25% of participants have had a pregnancy? If we did, we would be wrong. It is just as important for us to hear from female participants who have not become pregnant and male participants who have not fathered a child.

Preliminary information showed us that almost all survivors of Wilms tumor are capable of becoming parents. We are currently updating this study and need everyone’s help in order to confirm that the earlier information continues to be true. Many of you have contacted us with questions about fertility and pregnancy. We very much want to answer these questions, so we need everyone’s help to gather that important information. If you haven’t returned the pregnancy survey, please do so as soon as possible. If you have lost your survey please contact us on our message line, 1-800-553-4878, and we would be happy to send you another one. Your participation in this aspect of the study will be invaluable. Thank you to those of you who have returned the survey. Your help is greatly appreciated.

What To Do If You’re Uninsured

By: Gib Smith, JD – Director, Childhood Cancer Ombudsman Program
and Grace Monaco, JD – Director, Managed Care Ombudsman Program

Access to care is compromised when the family is uninsured. What can the medical care team do to help cancer patients/survivors and their families find health insurance? This is a real and constant problem in the cancer community. It is tough enough trying to recover from radiotherapy and chemotherapy without having to worry about how to pay for these necessary treatments. This article attempts to provide a brief overview on some of the resources and protections available to those affected by a diagnosis of cancer.

If you don’t have, have lost or are looking for insurance coverage we can suggest the following resources/options:

- Call your County Welfare Office and ask for information about applying for Aged, Blind, Disabled Medically-Needy Medicaid for your child. If you meet the income and asset limits your child can be put on Medicaid.
- While on the line with the welfare office they should check on the State Children’s Health Insurance Program (SCHIP). In 1997 the SCHIP Program gave the states new federal funds to expand children’s health coverage programs, which led to a dramatic extensions of coverage for low-income children through Medicaid and separate SCHIP programs. There is some form of SCHIP in the 50 states and the District of Columbia.
- An underutilized, but excellent source of information regarding local resources is the social worker in the pediatric oncology clinic. For example, in some facilities Social Security employees come to the hospital periodically to help families apply for benefits. The social worker should have contact information so you can apply for the right welfare programs.
- Check on resources of your State’s Childrens Medical Services Program. Some families who don’t qualify for Medicaid may qualify for assistance to pay for needed care under that program if the family income level is below their coverage income level.
- Consider the Health Insurance Portability and Accountability Act (HIPAA), which allows those with preexisting conditions to secure comprehensive health insurance coverage. From the parents’ perspective, HIPAA also helps people maintain their coverage if they need to change insurance or jobs. For more information regarding HIPPA visit http://cms.hhs.gov.
- Finally, the Consolidated Omnibus Budget Reconciliation Act (COBRA) mandates that both

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This list is intended to be a reference for you and your medical care team. Just having this information and the willingness to share it may save patients and their families hours of anguish, additional out-of-pocket costs, and perhaps lives if successful in obtaining some form of coverage for expensive cancer treatments that would otherwise not be covered.

NOTE TO OUR CANADIAN PARTICIPANTS: Similar information is being gathered for our Canadian participants and will be posted on our web site prior to the publication of the next newsletter.

Meet the Late Effects Study Staff

Bobbi Benson began her work with the Data and Statistical Center (DSC) seven years ago. She first conducted interviews for a study of cancer in families funded by the American Cancer Society. Results of this study were released just last year. Bobbi then coordinated registration for the Risk Factor Study, a study that evaluates the relationship between occupational, environmental and other suspected risk factors and the occurrence of Wilms tumor. Currently she works to document “targeted” late conditions, an important aspect of the Late Effects Study.

Targeted conditions identify certain late conditions former Wilms patients may have had as a result of treatment for Wilms tumor. These conditions are second malignant tumors, congestive heart failure or cardiomyopathy, respiratory failure and/or renal failure. Bobbi ensures all steps in documenting the onset of a targeted condition are met. This process can take some time as well as follow-up calls and letters to participants, their families and to doctors and hospitals. The information we gather on these conditions will enable us to advise people who were treated for Wilms tumor about what tests to have done so they can be treated at an early stage, if they develop.

What inspires Bobbi most about working with the Late Effects Study is “The possibility that the results of my work will benefit children.” Bobbi says, “I really enjoy talking with participants on the phone and I hope that my work, combined with the work of all my colleagues, will contribute towards learning more about what causes Wilms tumor, so one day, we might know more about how to prevent it.”

Bobbi’s favorite activity is traveling with her family. Several times they have had the opportunity to travel for a couple of months and because her children were only 3 and 5 years old when they started traveling, they would rent cottages and try to live as much as possible like local families. One time, this included having both of her children come down with chicken pox in England. During these trips they love to hike, be it through Capadocia in Turkey or the fjords of Norway. Most recently Bobbi was in Vietnam where she especially loved walking in the mountains around Sapa, a small village near the Chinese border. Bobbi also enjoys cross country skiing, cooking and reading.

Sarah Christie has been working for Fred Hutchinson Cancer Research Center (FHCRC) for three years and in March, 2002 joined the DSC. She coordinates all of the pregnancy correspondence for the Late Effects Study and contacts participants to conduct phone interviews. Previously with Collaborative Data Services (CDS) at FHCRC, she interviewed participants on various cancer prevention studies and then moved on to supervising other CDS interviewers.

Sarah says one of her favorite things about working for the Wilms Tumor Study is speaking with participants on the phone. “It’s great to hear the excitement in a mother’s or father’s voice as they tell me about their new baby. I also hear some sad stories and am always amazed by the strength and positive attitudes these participants have despite going through some very hard times.”

Although she loves Seattle’s energy, water and mountains, she still maintains a very close tie to her hometown Milwaukee, Wisconsin, where she tries to visit once or twice a year to catch up with family and friends.

Sarah enjoys spending her free time practicing yoga, rollerblading and camping (weather permitting). She is also a great fan of the movies and actively sings in a choir. Recently she even recorded a song that was featured at her friend’s wedding. Perhaps when you speak with her on the phone you can guess which 1972 Seals and Croft hit it is!
Interview with Chris Bridge, Co-Author of Andrew’s Story

In early 2002 we received a copy of a delightful book, Andrew’s Story. The authors, Andrew Bridge and his mother Chris, describe their experiences after Andrew was diagnosed with and treated for Wilms tumor in 1991 at three years of age. Chris Bridge agreed to share some of their story with us.

Q: You and Andrew wrote Andrew’s Story about his experience after being diagnosed with Wilms tumor. What motivated you to do this?

CB: The process started when Andrew was in the hospital getting tests. He was getting bored, and his grandmother had brought a camera. She gave him the camera to keep busy. I wrote little notes about what was going on, and he took pictures about what was happening around the hospital. We put these together in a book. After Andrew physically recovered we used the book to work on his emotional recovery.

At first it was just a book for him. Later we were watching Marlo Thomas on a program about St. Jude (Children’s Research Hospital). There was a little boy with her, Zach, who had Wilms tumor and was three years old. I said “Andrew, Zach is going through what you did when you were three. I wish we had a copy of your story to send him.” That’s when we came up with the idea to develop his book and send it to a publisher.

Q: You said you had published another book. Did you take Andrew’s Story to your publisher?

CB: No, we needed a publisher who was interested in health issues. A friend learned of Lerner Publication’s Company in Minneapolis. They had published a series of books for children. This is the “Meeting the Challenge” series, and it looked like a good fit for us.

Q: What is this series about?

CB: The “Meeting the Challenge” series is for children who are going through special situations from adoption to undergoing surgery. There are twelve titles that meet the needs of children with special needs.

Q: How did Andrew do after treatment ended?

CB: Andrew was three, and at that age children are learning to go beyond family and caregivers and learning to trust adults more and more. He was trying to set some personal boundaries. When he was being treated if he said “No, don’t touch me” they touched him anyway. It was difficult for him to try to keep control of his space and for it not to be respected. So he needed to work on his trust in adults.

We went back and talked with the doctors and nurses to have normal conversations without being treated. He was able to separate the experiences between being sick and being well. That was the beginning of recovery for him. He shared his story with others, and this helped him understand what the people did to help him even though it hurt him.

Q: How is Andrew doing now?

CB: Andrew is doing great. He’s involved in skiing, swimming and tae kwon do. He is also on the honor roll.

Q: Andrew has an older brother, Tyler. What was this experience like for him?

CB: Tyler was five at the time and just getting ready to start kindergarten. He was busy with his own activities. But, he did know that Andrew was sick and needed extra help. He wanted to help Andrew get better, and he wanted to be involved. He would spend time with Andrew playing, reading or running errands.

Q: What resources do you think would be most helpful for families experiencing all of this?

CB: In the past five years there have been more and more resources for families that weren’t available when Andrew was being treated. Now people can go to their computers, type in “Wilms tumor” and find out all kinds of information. However, a book I would like to see is one on how parents can be better observers of their children at home. I am not a nurse and didn’t know everything the doctors were talking about. Sometimes I would get home and not remember what the doctor had said. Yet I had to be a nurse for my child when I got home. A book about this would have been a great help.

Q: How can someone get a copy of your book?

CB: We have a limited number of books left that we can give out free. Local school kids did fund raising to pay for publishing so that the books could be given to kids in hospitals around the country. If anyone would like a copy they can email me at Chbridge@aol.com or order one through their local bookstore.
Treatment of childhood cancer has made great progress. However, your family doctor may not have experience with survivors of childhood cancer. She or he may not be aware of what follow-up exams are suggested given your medical history. If you have been off treatment for five or more years and you are not returning to your treating institution, the following guidelines should be of interest to your physician.

**RECOMMENDED ANNUAL FOLLOW-UP**

- Complete Physical Examination
- Blood pressure measurement
- Laboratory tests:
  - CBC
  - WBC Differential
  - Liver function tests
  - Renal function tests
  - Urinalysis

**Why are these tests suggested?**

Some of the treatment(s) you may have received can affect the body many years after your therapy is finished. It is important to see if any of these effects are present so that your doctor can treat them, if necessary.

**CBC and WBC differential.** These tests measure the number and type of cells present in the blood: these are white blood cells, red blood cells, and platelets.

Some chemotherapy drugs can affect how well the bone marrow works to make normal blood cells.

Sometimes kidney dysfunction can affect the production of red blood cells or hemoglobin (Hb).

Radiation therapy, if it is to a large area such as the whole abdomen, can make the bone marrow work less well and produce fewer blood cells than normal.

**Liver function blood tests (usually AST, ALT, AlkPhos, Bilirubin).** These tests measure how “irritated” the liver is as well as how well it is making bile to help with digestion of food.

Some chemotherapy drugs can irritate the liver. Most of these effects occur during or shortly after therapy, but some can last for a long time and affect how well your liver works.

**Renal function blood tests (Usually BUN and plasma Creatinine and GFR).** These tests measure how well your kidneys are working. If you have had one or part of your kidneys removed, the one (or parts) that are left behind have to work harder to filter the blood.

**Urinalysis + 24-hour urine collection.** This test measures the amounts of protein and sugar that pass through the kidneys and are excreted in the urine. Normally there will be very little of either.

**Blood pressure measurement.** Sometimes high blood pressure can happen when the kidneys are not working well and as early signs of renal failure. High blood pressure is associated with a higher risk for developing atherosclerotic heart disease and strokes.

**If you received Ifosfamide (or Cisplatin):**

Laboratory tests such as pH of the blood and urine, electrolyte plasma and urine level (K, P, bicarbonate and uric acid) help with early detection of Fanconi Syndrome (generalized dysfunction of proximal tubule).

**If you received Adriamycin, also called Doxorubicin:**

Adriamycin can make the heart muscle weak. This is a cumulative effect, that is, the more medicine you received, the greater your chances of having a problem.

The echocardiogram or MUGA scan will look to see how well your heart muscle is working.

Pregnancy and sudden vigorous exercise can place strain on the heart. It is important to know, before entering into these activities, how well your heart is working. It will be important for your doctors to be aware and continue to monitor for this potential problem. A heart specialist (cardiologist) should be consulted and tests done such as MUGA scan or echocardiogram prior to engaging in any vigorous sports, planning pregnancy and during the last trimester of pregnancy.

**If you had chest irradiation as a girl:**

You may be at an increased risk for developing breast cancer. Mammography is one way to screen for early tumors. Mammograms should be done annually in mature women beginning ten years after chest irradiation. (Boys are not at increased risk of breast cancer if they received lung irradiation.)

**If you were irradiated or treated for CCSK:**

Yearly skeletal survey and/or bone scan (x-ray) until you are fully grown, then every 5 years indefinitely. The bone scan and skeletal survey are done to diagnose bone irregularities. The NWTS recommends that both be done as each picks up conditions the other doesn’t.
Heart and Blood Vessel Disease after Treatment for Wilms Tumor

By: Melissa M. Hudson, MD, Director, After Completion of Therapy Clinic
St. Jude Children's Research Hospital

The success in treating children with Wilms tumor has made it possible to study the long-term side effects of cancer therapy. These studies have shown that some of the lifesaving therapies may affect the health of the heart and blood vessel system. These problems result from two treatments used in children with advanced stage Wilms tumor: anthracycline chemotherapy and radiation therapy.

Doxorubicin (also known as Adriamycin) is a type of anthracycline chemotherapy used in some Wilms tumor treatments. High doses of Adriamycin weaken the pumping muscles of the heart and may lead to a condition known as cardiomyopathy. In severe cases, cardiomyopathy can lead to heart failure. The risk of heart failure is related to the dose of Adriamycin. When doses are kept below 300 mg/m² in children, the risk of developing heart failure is less than 5%. But sometimes lower doses of Adriamycin may cause heart damage that cannot be detected by heart screening tests like an echocardiogram. These “subclinical” changes in heart function may increase the risk of heart problems in long-term survivors of Wilms tumor as they get older.

Radiation therapy can also injure the heart muscle, valves or blood vessels. In Wilms tumor patients, the left ventricle, the main pumping chamber of the heart, is likely to be in the radiation treatment field when radiation is delivered to the lungs or the left side of the abdomen. As a result, patients with Wilms tumor involving the lungs or the left kidney usually have some radiation to the left ventricle. Higher radiation doses (more than 3000 cGy) may cause the heart muscle to become stiff, which makes it difficult to pump blood through the heart chambers and into the bloodstream. Radiation may also lead to heart failure by causing the heart valves to become leaky or stiff. Finally, radiation may cause scarring in the coronary arteries, the blood vessels that feed the heart. This increases the risk of atherosclerosis, or blockage by fatty deposits circulating in the blood stream. Once blood flow to an area of the heart is blocked, severe chest pain (angina) may occur. If blood flow is not restored, the heart muscle tissue in the blocked area may die. This condition is known as a myocardial infarction (heart attack).

Compared to adults, children’s hearts are more sensitive to the effects of Adriamycin and radiation therapy. To avoid heart and blood vessel damage, Adriamycin and radiation doses are kept as low as possible in modern treatments for children with Wilms tumor. In patients who received older treatments, heart failure is uncommon unless therapy included high doses of Adriamycin, for example, after a relapse. Combining Adriamycin with lung or left abdominal radiation also increases the risk of heart failure. Some studies show that female patients have a higher risk of heart failure compared to male patients. The reason for this is not clear, but it may be due to a difference in how girls break down (metabolize) the Adriamycin.

There are several steps Wilms tumor survivors should take to keep their hearts healthy.

1. Know your cancer history. Ask the doctors at your cancer center to review the details of your treatment and tell you if you received any treatment that affects heart health.
2. Share your cancer history with your primary care doctor. Be sure your doctor knows about the specific health risks related to your cancer treatment.
3. Have regular check-ups and periodic heart tests if you were treated with anthracycline chemotherapy or radiation to the chest or left upper abdomen.
4. Remember that most survivors of Wilms tumor have normal heart function on examination and screening tests such as an echocardiogram or electrocardiogram. Subclinical injury that cannot be detected may increase the risk of heart problems as you get older.
5. Report persistent symptoms (like chest pain, rapid or erratic heart beats) to your doctor, even though serious heart disease is not a common occurrence.
6. Talk to your doctor about other medical conditions that can affect your heart and blood vessels. Keep blood pressure, cholesterol, and blood sugar in good control with diet or medication as recommended by your doctor.
7. Since pregnancy and childbirth can stress a heart that has been weakened by cancer treatment, be sure to get your doctor’s advice before becoming pregnant. Notify your doctor if you become pregnant so you can be monitored more closely for complications related to your cancer treatment.
8. Review your health habits and practice healthy behaviors that reduce the risk of heart disease. The following advice is a prescription for a heart healthy lifestyle:
   - Do not smoke. If you smoke, quit. Avoid smoke filled rooms.
   - Maintain a healthy weight. Reduce if you are overweight.
   - Exercise regularly for 20-30 minutes at least 3 times a week—every day is even better. Be sure to get your doctor’s advice before starting an exercise program.
   - Eat a heart healthy diet including a variety of fruits and vegetables—5 or more servings a day. Limit fat in your diet. Get protein from fish, beans, fat-free and low-fat milk products, skinless poultry, and lean meats.
   - Do not use cocaine or other recreational drugs.
Participation is Voluntary!

Once a year, Hal collects his National Wilms Tumor Study mail and takes it to be recycled. What Hal doesn’t know is if he doesn’t want to receive any more mail from NWTS, all he has to do is call 1-800-553-4878 and let them know he’s no longer interested in participating.

Feeling burdened by our requests? Your participation is voluntary. If you wish to withdraw, simply call us at 1-800-553-4878.

A Reader Asks . . .

A newsletter reader has asked us if the NWTS has a recommendation concerning whether or not Wilms tumor survivors should have smallpox vaccinations. Unfortunately we have no research supported data to make an informed recommendation about your vaccination.

However, we do recommend that you discuss your medical history carefully with your family physician before you decide whether or not to have the vaccination.