



THE
cholangiocarcinoma
FOUNDATION

spring 2008

*“The greater danger for most of us lies not in setting our aim too high and falling short;
but in setting our aim too low, and achieving our mark.”*

—Michelangelo

The vision of the Cholangiocarcinoma Foundation is, admittedly, focused upon a lofty goal: to become the primary resource for leadership in Collaboration, Understanding, Research, and Education directed towards a CURE for cholangiocarcinoma. With this singular vision guiding the Foundation’s efforts, much has already been accomplished. For instance, the Foundation has recently launched an expanded, state-of-the-art website (www.cholangiocarcinoma.org) to serve as a useful and informative resource for patients, friends, caregivers, and loved ones. Also, the Internal Revenue Service has just officially approved our status as a charitable foundation under Section 501(c)(3) of the Internal Revenue Code, enabling the Foundation to receive tax-deductible contributions to fund the battle against cholangiocarcinoma. You are reading the inaugural issue of the Foundation’s quarterly newsletter, which will provide regular updates on the Foundation’s efforts and notable developments in the understanding, diagnosis, and treatment of cholangiocarcinoma.

The vision of the Cholangiocarcinoma Foundation is lofty, primarily, out of necessity. For years, cholangiocarcinoma has largely been overlooked by the medical, surgical, research, philanthropic, and support communities because of a pervasive myth: that cholangiocarcinoma is a “rare” cancer, supposedly afflicting only a small percentage of elderly patients. But this myth is as dangerous as it is fictional, and it has led to a void of attention to cholangiocarcinoma that the Foundation intends to fill.

But if you are reading this newsletter, you likely already know the disturbing truth. In the United States alone, we can conservatively assume that at least 3,000 to 3,500 people are diagnosed with cholangiocarcinoma each year, and this figure does not include many patients who have been misdiagnosed but who are, in fact, afflicted with this disease. And that is just domestically. Abroad, cholangiocarcinoma is just as bad and, in some cases, worse. The examples are numerous: in Japan, researchers estimate that more than 7,000 people are diagnosed annually, while half way across the globe, in Israel, some 550 people receive the same terrible news each year. If you consider only those three countries—the United States, Japan, and Israel—more than 11,000 people are being diagnosed with cholangiocarcinoma every year. That is about 31 people every day. Do the disturbing math, and you quickly realize that one person is diagnosed with this cancer on every hour of every day of every week of every year; if we had reliable statistics for the entire international community, the results would be worse still.

By furthering public awareness of cholangiocarcinoma through, among other means, education, advocacy, patient support groups, physicians, researchers, and national conferences, the Foundation will be working hard

to dispel myths like this one, while underscoring the pressing need to devote meaningful attention, effort, and resources to this disease. While the Foundation is totally committed to its vision, we are also realists who appreciate that we cannot achieve our vision alone. We will need—and ask for—your help and assistance, and that of your families, friends, and communities.

For the Foundation, failure is not an option. We are thoroughly committed to our vision because it is a vision borne out of the most powerful, yet basic of all human emotions: love—love for our family and friends who have battled this disease. Because of our love for them and our compassion for those who currently fight cholangiocarcinoma or will do so in the future, the Foundation will work tirelessly to make our vision a reality. Please join us. Together, we can accomplish great things. ✨

the cholangiocarcinoma foundation

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What is Your Top 10?

Dr. Geret Giles

Newspapers, websites, and TV programs all list their top stories of the year. What would you list as your personal top stories of 2007? If you are reading this newsletter, cholangiocarcinoma probably figures prominently in your life. In fact, if you were to list your personal Top 10 Stories of 2007, cholangiocarcinoma might be mentioned more than once. But what other things belong in that Top 10? They can be positive or . . . less so. Get out your calendar and review the year month by month. You'll be surprised what you have forgotten. Here are some questions to ask yourself as you make up your list.

Where have I gone?

What have I seen?

What changed for me this year?

What/who did I lose?

What/who did I gain?

What experiences have I had?

Do you have ten, yet? Once you get started, you will probably have a hard time keeping your list to ten items. Don't be afraid to list the hard things. Sometimes those are the most influential--and ultimately the most growth-producing. After you have made your list, share it with loved ones and ask them to do the same. If there's one thing my friend's battle with cholangiocarcinoma has taught me, it's that the most valuable things we have are the connections we share with others.

Now, one more thing. After reflecting on the past, turn your thoughts to the future. A year from now--at the end of 2008--what would you like to be able to list as your personal Top 10 Stories of 2008? Write those down. Reflect on them often. Let the words inspire you. Our minds have a way of working things out when given reflective, meditative time but it is difficult to see opportunities when we are not looking for (or thinking about!) them. May this year be all you think it can be--and more! ✨



Dr. Geret Giles is a licensed psychologist with a private practice in Orem, Utah. He received a Masters Degree in Marriage and Family Therapy from Brigham Young University and a Ph.D. in Counseling Psychology from the Pennsylvania State University. Dr. Giles works with individuals, adolescents, children, and couples suffering from a wide range of issues including depression, anxiety, and relationship difficulties. He is often asked to address psychological issues for large groups in the community. His section, "Ask Dr. Giles," is one of the most popular features of cholangiocarcinoma.org. In January of 2007, he lost his best friend to cholangiocarcinoma.

cholangiocarcinoma.org

Stacie Lindsey

The first three months after Mark (my brother) was diagnosed I burst into tears many times as I had so completely immersed myself in research on his behalf that exhaustion, frustration and a desperate need to provide him with some hope often overtook me.

When Rick and I decided to do the discussion board, it was out of a need for us to connect with other families and patients that were suffering as we were, in the hope that perhaps we could find ways to share research and general information so that the burden could be spread and the load lightened.

The intent for this website was two-fold, that no one, having received a diagnosis of cholangiocarcinoma would have to feel alone and overwhelmed. And that we could gather all of the pertinent information and research into one place. That being said, we certainly won't be able to cover everything, and we will all still feel alone and overwhelmed at times, but we've made a good start and there is much more to come.

Both of those things have come to fruition and now we have additional goals as well, they are much bigger than any of us thought possible but I have no doubt that they will happen.

I wanted to thank everyone who has worked tirelessly on this project, especially Rick and Sara. The three of us are bonded for life over this labor of love. ✨



Medical Writer: Erin Grothey

Source: Abstract presented at 43rd Annual Meeting of the American Society of Clinical Oncologists in Chicago, IL, June 1-5, 2007

Reference: El-Khoueiry A, Rankin C, Lenz H, Philip P, Rivkin S, Blanke C. SWOG 0514: A phase II study of Sorafenib (BAY 43-9006) as single agent in patients (pts) with unresectable or metastatic gallbladder cancer or cholangiocarcinomas. *J Clin Oncol* 2007; 25(18S): Abstract 4639.

Medical Review: Dr. Jean-Nicolas Vauthey, MD Anderson Cancer Center, Houston, TX

Dr. Steven Alberts, Mayo Clinic, Rochester, MN

Dr. El-Khoueiry and a team of research clinicians led a clinical trial testing the drug sorafenib (Nexavar®, Bayer Pharmaceuticals) as possible treatment for patients with gall bladder cancer or cholangiocarcinoma, and results of this study were presented at the 2007 Annual Meeting of the American Society of Clinical Oncology.¹ In the study the authors concluded that sorafenib, when administered alone, was unable to induce shrinkage of the cancer in any of the 31 patients treated. In addition, El-Khoueiry and colleagues found that the median amount of time that sorafenib-treated patients survived and remained without progression or growth of the cancer was similar to that found with other chemotherapy regimens currently used by physicians. These results did not reach the goals set for proving a benefit to patients treated with sorafenib in regards to tumor shrinkage, therefore the clinical trial was closed early and did not move into the second stage of recruiting patients for treatment.

Survival of patients with locally advanced or metastatic biliary cancers (i.e. gallbladder cancer or cholangiocarcinoma) that cannot be resected is amongst the shortest of those who suffer from cancer. Chemotherapy drugs have had limited activity in stopping the growth of this cancer type. Gemcitabine is currently considered a standard chemotherapy drug for patients with this form of cancer. As conventional drugs, such as gemcitabine or capecitabine, that indiscriminately kill dividing cells have a limited benefit in improving the length and quality of life for patients with biliary cancers, interest and hope was placed in newer “targeted agents” that specifically shut down tumor cell machinery that allows them to grow out of control. Sorafenib, one such targeted agent, has improved survival for patients with kidney and liver cancer. Dr. El-Khoueiry and colleagues designed a clinical trial to evaluate whether sorafenib could also improve survival for patients with locally advanced or metastatic gall bladder cancer or cholangiocarcinoma.

Patients who had not yet received chemotherapy, and were therefore more likely to benefit from this experimental therapy, were treated with sorafenib twice a day, every day, in a pill form that patients could self-administer at home. Nineteen treated patients had diagnosis of cholangiocarcinoma and 12 of gallbladder cancer. Patients were seen by their physicians and re-evaluated every 2 months for growth or shrinkage of the cancer. After an average of 2 cycles (1 cycle=1 month) of treatment with sorafenib, Dr. Khoueiry and colleagues reported that no patients had shrinkage of the cancer, but in 10 patients the cancer had failed to grow. More than half of the patients remained progression-free for at least 3 months, but all progressed by 8 months of treatment. Among the 31 patients treated with sorafenib, half survived 9 months, and at the time of this analysis, the range of survival time after beginning treatment with sorafenib was 4 to 10 months.

Patients did not substantially benefit from sorafenib treatment, and they were likely to have experienced side effects while being treated. The more severe toxicities (NCI CTC grade 3 or 4) reported for patients in the study included 4 cases of hand-foot skin reaction, 3 cases of venous thrombosis/embolism, 3 cases of abdominal pain, and 1 case each of gastrointestinal bleeding and gastrointestinal perforation. One patient died while on treatment because of heart complication and venous thromboembolism.

References

1. El-Khoueiry A, Rankin C, Lenz H, Philip P, Rivkin S, Blanke C. SWOG 0514: A phase II study of Sorafenib (BAY 43-9006) as single agent in patients (pts) with unresectable or metastatic gallbladder cancer or cholangiocarcinomas. *J Clin Oncol.* 2007;25(18s):Abstract 4639.

Donations

Your donation supports a comprehensive approach to fighting cholangiocarcinoma through patient support, community outreach, research and advocacy for a cure.

You can send donations to:

The Cholangiocarcinoma Foundation, 10 West Broadway, Salt Lake City, Utah 84101
or online at www.cholangiocarcinoma.org.

Riding the Rockies for a Cause

The annual "Ride The Rockies" bicycle tour takes 2,000 cyclists on a 6-7 day ride through Colorado's Rocky Mountains. Ride the Rockies is not a ride for the faint of heart because it taxes even the most experienced competitors. From June 17-23, 2007, Sandy Fuller was "Riding the Rockies" as she had done for the past three years. However, this year she rode for an entirely different reason.

Sandy has maintained a close relationship with her cousin Peter Betts throughout their lives. Their relationship might be more akin to siblings than cousins. Although they live in different parts of the country, they have remained kindred spirits through the years. "Peter and I have the same values and priorities in life, which bonded us together. We have always been able to talk and be confidantes for each other," Sandy shared.

In the Fall of 2005, Sandy received some news from Peter. They took a long walk together, where Peter revealed that he had a rare form of liver cancer: cholangiocarcinoma. Sandy confided, "We were unsure at the time, how long he would be around." As they talked that autumn afternoon, Peter told her, "I don't know what is going to happen, but whatever it is, I'm going to do it well." Peter remains true to his word.

As Sandy pondered Peter's situation, "Ride the Rockies" came into her mind. She immediately called Amy Somerstein, Peter's wife. Amy thought it was a great idea, and enlisted the help of Peter's daughter, Mariah. Mariah sneaked into Peter's personal email and stealthily extracted his list of

contacts. Unknown to Peter, Mariah contacted Peter's friends, his Mom's friends, Sandy's friends and their entire family. "One of the benefits of organizing this ride for Peter was how nice it was to reconnect with so many family members and friends for a united cause," said Sandy.

On June 17, 2007, Sandy began the ride with 7 friends. At 57 years of age and slight of frame, Sandy said the ride was difficult, however, climbing was not the part of the ride that got to her. Sandy recalled, "The long, flat stretches were far more difficult, as the weather was extremely hot and windy. The ride was a personal challenge for me; riding for Peter enhanced the whole thing and made it really worthwhile. It was all plusses and I truly enjoyed doing it."

Sandy called Peter after the ride. Sandy asked Peter where he was, and he said, "I'm standing in the middle of a field." After she told him what she had done, Peter replied, "now I'm sitting in the middle of a field."

The emotions were overwhelming. Peter said, "I felt the love and support of so many people. It was very touching that Sandy would do this for me and that so many would support her efforts." Sandy raised over \$2,735.00! "It is easy to send \$10.00 or a card to someone you care about, but this has been so rewarding for me. I have never regretted or resented the many hours I spent organizing and riding because success requires that kind of effort," said Sandy. ✚



For more information about "Ride the Rockies" please visit www.ridetherockies.com



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