



**You're Invited
to our
Birthday
Party!**

**The VHL Family Alliance
turns 15 years old!**

In January 1993 we published the first issue of the VHL Family Forum.

In 1994 we held our first national meeting in Kansas City, Kansas.

This year, in celebration of our fifteenth anniversary, we are planning to meet in Orlando!

Meetings will be held all day on Saturday, June 28, followed by the birthday dinner celebration.

Sunday the Board will meet while everyone else is on their own for fun in the parks.

The theme of this meeting is Staying Strong -- both mentally and physically. We will have some presentations designed for younger attendees. Some speakers will focus on complementary medicine -- vitamins and herbs that can help to soften the course of VHL.

You can register for the meeting right now, on page 15 of this newsletter or on the internet at vhl.org. Prices go up after April 30, so please make your reservations early!



You will need to make your own reservations with the hotel by April 30 to get the special room rate for the VHL meeting:

Courtyard Orlando Lake Buena Vista
8501 Palm Parkway,
Lake Buena Vista, FL, 32836,

(407) 239-6900 or 1 (800) 321-2211

\$99 per room per night

You will need to arrange transportation from the

airport to the hotel. Once you are at the hotel, however, there is a free shuttle from the hotel to the parks.

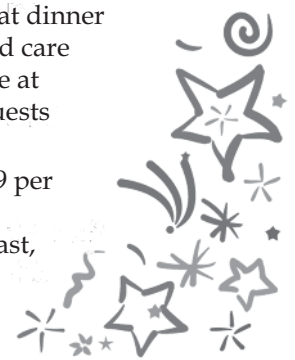
Meetings will be held 9 to 5 on Saturday, June 28, so you will want to arrive on Friday night.

The Birthday Dinner on Saturday night is included in the all-day conference rate. For those not attending the conference, extra dinner tickets will be \$20 per person (adult or child).

Children are welcome at dinner or meetings. Optional child care arrangements are available at additional cost. Send requests to orlando@vhl.org

Registration will be \$99 per person for the full day of meetings (includes breakfast, lunch, and festive dinner to celebrate our birthday!)

We encourage you to come with a spouse or



Inside this issue!

- | | |
|---|--|
| Nominations open for Board of Directors | Online Learning presentations Edbassmaster and Maureen |
| Intro to VHL for Children | Memory Quilt |
| Radiofrequency Ablation and Cryotherapy | Symposium in Denmark |
| CT and Radiation | Powerful Patient Radio Show |
| New Clinical Trial | Meetings in LA, Manchester, Johannesburg, Boston, |
| Managing Your Team | Symposium in Denmark |

significant other person, to make sure that your team is all on the same page! We are offering a discount for a second attendee from the same family -- take \$20 off the second registration.

AND if you register before April 30, you get \$20 off each registration (in other words, two people for \$79 + \$59 instead of \$99 +\$79). WHAT A DEAL!

We have left Sunday free so that you can enjoy the attractions in Orlando. You can arrange park tickets in advance, or purchase them on site. There are several websites on the internet where you can plan your journey and reserve tickets. A few examples:

<http://www.planorlando.com/>

<http://www.universalorlando.com/>

Disney offers a Free Vacation Planning DVD that includes a tour of the parks at

<http://www.waltdisneyworld.com/>

It is generally more economical to choose one park per day, rather than to "park-hop".

Orlando is particularly good for visitors with special needs. Check on the websites or call the hotel for advice.

Nominations Open

New Board members will be elected at the Annual Meeting in Orlando in June.

Nominations are now open for four new members of the Board of Directors. If you would like to nominate yourself or another person, please send a message to the address below. Your message should include the nominee's name and address, work history including volunteer leadership positions, and the experience and talents the nominee would bring to the board.

We are glad to have a diverse set of skills and talents and perspectives on the Board. In particular, we would like to put greater focus on the Clinical Care Centers and on enhancing Member Services.

The Board of Directors of VHLFA are all volunteers, citizens of the United States, governing the US-based corporation. Our affiliates in other countries are also looking for volunteers to add strength to their own organizations. If you would like to volunteer in another country, please send information to that country's contact person, or to nominations@vhl.org.

Thank you for adding your energy, enthusiasm, and talent to the VHL Family Alliance.

Send nomination messages to:

2001 Beacon St, Ste 208, Boston, MA 02135
nominations@vhl.org

Introduction to VHL for Children - Reviewers needed!

*by Gayun Chan-Smutko, M.S., Genetic Counselor,
Massachusetts General Hospital, Boston*

For the past year a committee of parents and professionals has been working on an introduction to VHL for children.

We are seeing more and more children who are diagnosed through DNA testing at early ages and have questions about what this diagnosis means to them. In some cases, they may have seen a parent or other relative go through some pretty difficult experiences with VHL, but there is much more hope for young people starting out now.

We wanted to provide accurate information in age-appropriate language, and help our children take responsibility for healthy choices and adherence to screening protocols.

Illustrator Needed

We are also still in the process of selecting an illustrator for the book. If you would like to submit samples of your work, please send them to the Children's Book Project, VHL Family Alliance, 2001 Beacon Street, Suite 208, Boston, MA 02135, or e-mail them to kidsbook@vhl.org

We are seeking volunteers to review pre-publication copies of this book. Please help us make sure the book is on target and will be helpful to you and your children.

Partial funding for the illustration and production of the book has been received from the estate of Deborah L. Jones of Philadelphia.



*Illustration by Jeffrey Byrd, or FAP and Me, copyright 2004
National Society of Genetic Counselors.*

Questions to Ask Your Doctor about Radio Frequency Ablation or Cryotherapy for Renal Cell Carcinoma

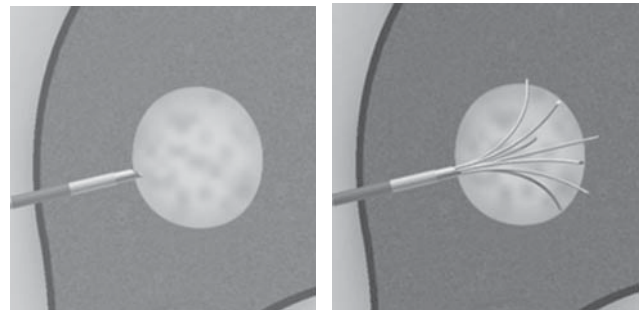
Radio Frequency Ablation is a radiological procedure where a heat probe is inserted through the skin and into the tumor. Then the tip of the probe is heated, and “cooks” the tumor.

Cryotherapy is similar, except that the probe is a freezing device. Once the probe is in the tumor, the tip of the probe is cooled to form an ice ball, freezing the tumor to kill the cells.

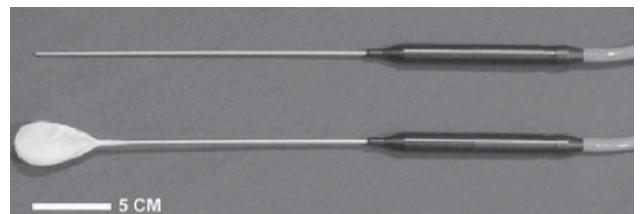
In both cases, the heat or cold energy must kill cells around the tumor in order to insure that all the tumor cells will be killed. Both procedures are attractive in that they are not open surgery, and the recovery is generally very quick. However, they are nevertheless both surgical procedures with risks of their own.

New treatments and new machines are being developed all the time that may further improve kidney treatment. The goal is to kill or remove only the tumor cells, and leave all the working nephrons doing the work of the kidney. A new DaVinci machine, for example, allows the surgeon to drive a robotic machine that can do very precise laparoscopic surgeries. As with all these options, the experience of the physician operating the machine and the treatment plan are critically important.

We have prepared the following list of Questions to Ask Your Doctor, in order to help you have a complete conversation before the procedure.



Radiofrequency is a type of electrical energy used to create heat. During a radiofrequency procedure, an ablation probe is placed directly into the target tissue. An array of several small, curved electrodes are deployed from the end of the probe into the tissue. The generator is turned on and target temperatures are input. The radiofrequency energy flows through the electrodes, causing ionic agitation, and therefore friction, in the nearby tissue. This friction creates heat, and once sufficient temperatures have been reached, the heat kills the target tissue within a few minutes. Source: University of Southern California Liver Transplant program.



A cryoprobe about 8 inches long, designed to be used under MR guidance, before (top) and after (bottom) creation of an ice ball. It took 10 minutes to create this 1 inch ice ball in water in the lab. Source: Silverman et al., Renal Tumors: MR Imaging-guided Percutaneous Cryotherapy, Radiology, 2005;236:716-724

Considering Radio Frequency Ablation or Cryotherapy for Kidney Tumors? Questions to Ask Your Doctor

1. Is the size of this tumor appropriate for this treatment -- or is it too big? How does size effect success rates?

2. How difficult will it be to get to the tumor?

3. Are there any structures adjacent to the tumor that might accidentally be injured by the treatment? (blood vessels, urine collection system, nerves or bowel)

4. Is there a way to protect those structures and still ensure complete destruction of the tumor? (e.g., injection of fluid or gas)

5. Are there other considerations that might recommend against using this therapy on this tumor?

6. Who might I see for a second opinion on other minimally invasive or image guided treatment options?

7. What do you feel are the chances of killing all the tumor cells in this tumor? or will some be left behind to grow back?

8. How long does it take to realize that a tumor was not completely treated?

9. How much normal kidney will be destroyed by the treatment?

10. How is the treatment likely to affect my kidney function?

CT and Radiation

In the November 29, 2007, issue of the *New England Journal of Medicine*, Drs. Brenner and Hall of the Center for Radiological Research, Columbia University Medical Center, New York, discuss the implications of the rising use of Computed Tomography (CT) scanning in the United States. "The widespread use of CT represents probably the single most important advance in diagnostic radiology." Nonetheless, CT does involve radiation, so it should be used with caution.

They point in particular to the sharp rise in the use of CT for diagnosing children, and for full-body scanning of asymptomatic adults. These two applications in particular are causes for concern. Some doctors are suggesting to use full-body CT instead of an annual office exam, which the authors feel is an unwarranted risk.

"The situation is even clearer for children, who are at greater risk than adults from a given dose of radiation, both because they are inherently more radiosensitive and because they have more remaining years of life during which a radiation-induced cancer could develop."

According to this article, for a 30-year-old person, the lifetime risk of developing a cancer as a result of a single CT scan is estimated to be 0.02% (two hundredths of one percent). Risks are higher for children, lower for older people.

In 1996 it was estimated that approximately 0.4% (less than half of one percent) of all cancers in the United States occurred as a result of CT use. With the increased use of CT scanning we see today, and factoring in estimates of future use heavily influenced by new programs to do full-body scans yearly, they estimate that in the future, as much as 2% of all cancer in the U.S. may be attributable to CT.

The press, of course, has focused on this 2%, misquoting the article widely as having declared that 2% of all cancers are now caused by CT. That is NOT what this article is saying. The authors are trying to limit the unnecessary use of CT. At the same time, they applaud the use of CT as a diagnostic tool for good medical reasons, including the preventive scanning of individuals at risk, as recommended in the VHLFA screening protocol.

The authors point out that many doctors were unaware, or at least not thoughtful about the fact that CT involves radiation exposure. Problems also arise "when CT scans are requested in the practice of defensive medicine, or when a CT scan, justified in itself, is repeated as the patient passes through the medical system, often simply because of a lack of

communication. Tellingly, a straw poll of pediatric radiologists suggested that perhaps one-third of CT studies could be replaced by alternative approaches or not performed at all." [p.2282].

"From an individual standpoint, when a CT scan is justified by medical need, the associated risk is small relative to the diagnostic information obtained. However, if it is true that as many as one-third of all CT scans are not justified by medical need, and it appears to be likely, perhaps 20 million adults and, crucially, more than one million children per year in the United States are being irradiated unnecessarily." [p.2283]

The bottom line is that the guidelines in the *VHL Handbook* are still on target. For children, we recommend screening methods that avoid radiation. Once you have a diagnosed problem that requires careful following, MRI would generally be preferred unless CT is deemed medically necessary.

We asked two radiologists to comment on this story:

– from Peter Choyke, M.D., U.S. National Cancer Institute, Bethesda, Maryland

We speculate about the effects of radiation at the doses/frequency of clinical imaging. But no one can put a number to it, even in the general population, let alone in an individual patient. At the same time, there are real, quantifiable problems from not making a proper diagnosis in a VHL patient. This is the same dilemma we have always had since we started advocating routine imaging. I think the balance still comes down clearly in favor of imaging rather than holding back in VHL patients.

– from Richard Cohan, M.D., Abdominal Radiology, University of Michigan, Ann Arbor, Michigan:

I don't think that this changes anything for the VHL patients. As per my lecture this past June, there are some risks related to radiation (although the risks being quoted may be a little overstated). There are radiation exposure risks to airplane flight, too.

In a VHL patient it is tremendously more important to use CT to detect and monitor small tumors. The risk of not imaging someone and allowing a renal or pancreatic tumor to grow uncontrollably is not acceptable. The likelihood of an unmonitored VHL tumor developing into a cancer is very high. I would encourage you to encourage your readers to relax. The message in all this is to eliminate unnecessary imaging.

As for gadolinium contrast in MRI, we are still learning; however, the news is encouraging. Just by being more cautious and by altering some of the agents we use in some instances, many centers have drastically reduced their number of cases of Nephrogenic Systemic Fibrosis (NSF). Some (including ours and one of the hospitals in Denmark that first reported the association) have virtually stopped seeing NSF cases in the past year.

For following tumors, either CT or MRI is acceptable, although I personally would skew toward MRI in younger patients (under the age of 40, say), if they have good kidney function. CT or MRI are fine to image older patients.

In young or older patients with severe renal failure, MRI is still probably safer than CT (which can damage the kidneys further if a patient is not on chronic dialysis); however, it might be reasonable to consider MRI without contrast first or to use one of the gadolinium agents that has not yet been seen to cause NSF.

Young or old patients on long-term dialysis should probably be followed with CT instead of MRI.

References: Brenner D and Hall E, Computed Tomography: An Increasing Source of Radiation Exposure, *New England Journal of Medicine* 2007; 357:2277-84.

Dr. Cohan's talk from the Boston meeting is reported in the September newsletter. The full DVD and handout are available at <http://vhl.impactlearning.org>

Dr. Libutti's Pancreas Article

The article we foreshadowed in the September newsletter has now been published.

"Clinical, genetic and radiographic analysis of 108 patients with von Hippel-Lindau disease (VHL) manifested by pancreatic neuroendocrine neoplasms (PNETs)" by Blansfield JA, Choyke L, ... Glenn GM, Middleton L, Linehan WM, Libutti SK. *Surgery*. 2007 Dec;142(6):814-8.

New Clinical Trial

Dr. W. Marston Linehan's urologic oncology group at the U.S. National Institutes of Health has opened a new clinical trial with a drug called AZ6474 from Astra Zeneca, another angiogenesis inhibitor. AZ6474 inhibits VEGF and EGFR.*

The goal of this trial is to determine whether kidney tumors in VHL respond to this drug. Ideally, tumors would shrink or disappear.

In order to qualify for this trial, you must have at least one measurable kidney tumor, but not one that needs immediate surgery. If your tumor is at or greater than 3 cm, you would be advised to have it treated surgically first, as response to this drug would not be expected to be swift enough to prevent the spread of the cancer.

It is assumed that people with VHL will have more than just one tumor, and probably in multiple organ systems. The team will watch all tumors to see if they respond to the drug, but the primary focus of the trial is on kidney tumors.

If you are interested in applying for this trial, please contact Sally Fowler, RN, at +1-301-435-6255.

This group's earlier trial with 17AAG (Geldanamycin) has slowed down because of problems getting sufficient quantities of the drug, and also because of the difficulty of administering the drug. 17AAG has to be infused; AZ6474 is a pill

Dr. Linehan expects to try a series of drugs alone and in combination, in hopes of finding an effective treatment with minimal side effects.

Further details will be posted on vhl.org/trials and clinicaltrials.gov.

* Vascular endothelial growth factor (VEGF) and epithelial growth factor receptor (EGFR) are two of the elements in the angiogenesis pathway of which VHL is a key component. While early studies seemed to indicate that inhibiting VEGF would be sufficient to constrain tumors, it seems now that it will be necessary to attack the problem from multiple directions at the same time.

Ready to Surf for a Good Cause?



Sign up as a shopper at www.iGive.com/vhl and VHLFA receives \$5.

Visit one of their stores, and VHLFA gets a chance to win a \$1000 prize, once a month through July 2008. No purchase necessary to enter this drawing, and each visit to one of their stores counts as one entry, with a maximum of 10 entries per person per day.

Purchase items through iGive, and VHLFA receives a small commision!

Managing Your Health Care Team

By June D., Wisconsin, june@vhl.org

The VHL Handbook (3rd International Edition, 2005) urges us to manage this disease with our "medical team". Having lived with VHL for nearly 30 years (I was born long before the days of genetic testing), I thought that I maybe had missed something. Although I'd received some training in team functions and management through my previous job, I had never thought about applying the principles I learned to the management of my disease. I had assumed that I was on my own in this realm. That is, until I contacted the VHLFA about this. In response to my inquiry, Joyce G. set up an email account, asking people to reply to my team questions: june@vhl.org

We would love to hear from you -- your questions and your suggestions. How have you approached gathering the specialists you rely on? Would you describe them as a "team"? What role do you play in getting them to communicate with one another?

Since many of our "team" members are highly-trained, very busy specialists, can we rely on them to take the time to communicate with all our other doctors? Does it make sense to have one person as the "hub" for these communications? In your case, is that you? or your general practitioner? or someone else?

What is the proper role of the patient, who probably is not medically trained but often is required to deal with complex, medical issues? Are there models out there where people living with other chronic conditions have developed successful, long-term management plans?

What about non-conventional care? For instance, I regularly see an acupuncturist to help me achieve optimal health while living with VHL. What complementary therapies do you use to maximize your health?

Finally, what have you learned about interacting successfully with your insurance company? What financial pressures have you met, and how have you dealt with them?

These are a few of the questions I have about living with this chronic illness. I will look forward to hearing from you, hearing your own questions, and learning from your own experiences and ideas. Please write to me at June@vhl.org, or respond to the Discussion we have started in the VHL Online discussion group, under the Topic "Health Management Issues".

Thank you! Let's pool our knowledge and learn from one another.

June@vhl.org or vhl.clinicahealth.com

Wish you were there? You can hear it ALL!

The presentations in Boston and Houston were excellent. We would like to share them with you.

A list of the Videos of the Boston and Houston meetings (2007) is available at

<http://vhl.impactlearning.org>.

You can watch them online, or order a DVD.

A DVD or audio CD containing all the presentations from each meeting is \$50 (the same price you would have paid if you had gone to the meeting). Handouts are included on a companion CD in either case.

Or choose selected presentations for \$10 each and watch them online.

As an introduction, Joyce's welcome presentation can be viewed at no cost to give you an idea of how it works.

No internet? No computer? See page 15 to order by mail, and play the DVD on your home television DVD player.

We will appreciate your feedback on this project.

Online Learning

Please try out the online learning at
<http://vhl.impactlearning.org>

You can sign up for *DVDs* or *podcasts*

Presentations from the Boston and Houston meetings from 2007 are available now!

Free Sample!

To give you the taste of it, you can watch Joyce Graff's presentation online for free.

We will be doing more of this kind of event. What would you particularly like to see?

Videos?

Powerpoint slides with audio tracks?

Audio only?

Send feedback to Joyce, director@vhl.org or paper mail in the envelope provided.

Be a Powerful Patient

by Joyce Wilcox Graff

More than once I have heard one of our members say to me that they helped a friend who had a serious illness. "Even though I know nothing about that condition medically, I was able to help with strategy, with managing a group of medical specialists, with managing the emotions and stresses that go with any illness."

Since 2006 I have worked with public relations advisers to shape the message of the VHL Family Alliance in a way that resonates with people outside of our specific community.

What has come out of that effort is a radio show called the Powerful Patient on webtalkradio.net. This one-hour show is carried weekly on webtalkradio.net on the internet. Shows are maintained in the Archive at <http://powerfulpatient.org>. You can sign up for the podcasts, or play an MP3 file over the internet.

The theme of the show is taking charge of your health, and creating teamwork with the medical professionals you need to "win the game" of keeping you healthy. It is not limited to VHL -- it applies equally well to any complex medical condition, which we define as being any condition where you are dealing with more than one medical specialist.

The Powerful Patient radio show is a service of the Cancer Resea

rch Fund of the VHL Family Alliance, raising public awareness of VHL and sharing the "Eight Ways You Can Help Your Doctor Save Your Life", which are helpful strategies for people with any complex health condition. You can get a free copy of this publication by signing up at the website <http://powerfulpatient.org>

The article on Edbassmaster and his sister Maureen, on pages 8-9 of this issue, is taken from that show. You can hear the full interview at powerfulpatient.org, which includes clips from some of Ed's skits on YouTube. The program guide that accompanies the interview includes the pointers to the skits quoted in the show.

There is an interview with Joyce Graff about how the theory of the Powerful Patient has evolved over the past fifteen years. In learning how to manage our health with VHL, we have learned valuable lessons about life and about health management that apply to any complex medical condition.

We interviewed Clenton Winford II about the diagnosis and treatment of his pheochromocytoma, and played excerpts from his album, *Wayfaring Stranger*. Bits of this album are featured as theme music for the show.

Ahead in the next few months are shows about

8th International Medical Symposium

September 4-6, Roskilde, Denmark

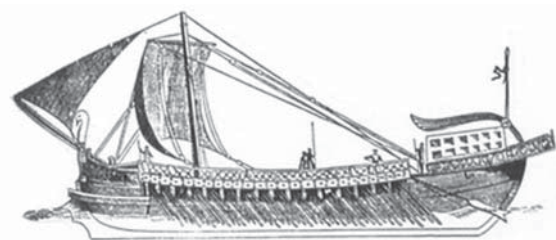
The International Medical Symposium on VHL has been held every two years since 1994. This year, the 8th symposium will be held in historic Roskilde, Denmark, the ancient capital of the Vikings.

The Scientific Chairman, Dr. Marie-Louise Bisgaard, invites scientists and physicians to submit abstracts for papers to be presented at the conference. See the website for guidelines for submission. <http://vhl.org/conf2008>

This is primarily an opportunity for clinicians and researchers to share their work, find collaborators, and move the pace of VHL research forward. Presentations and posters will cover molecular research, clinical research, and strategies for improving quality of life.

The language of the meeting will be English. Patients and family members are invited to attend, on the understanding that the medical professionals will be primarily talking to one another, not to patients, so the language may be somewhat technical. Even so, this is a marvelous opportunity to see the progress being made and the level of interest in the research community, and to take back to your local physicians news from the very best of VHL knowledge.

The symposium will be held at the Comwell conference center, which overlooks the fjord. Accommodation is available at the Comwell conference center, as well as two other hotels in town. To book accommodation please refer to the registration section. Social and shopping events are planned and will be published in the agenda section. For those of you interested in history we can show you real Viking ships as well as full size replicas which sail on the fjord. Danes became Christian about 1000 years ago and since then there has been a church or cathedral in the center of Roskilde, which is well worth a visit. If you prefer power shopping then Copenhagen is less than an hour away. Finally watch out for cyclists. They are everywhere and they will probably be too busy talking on their phones to see you crossing the road.



Edbassmaster and his sister Maureen

From a radio interview with Joyce Graff at <http://powerfulpatient.org/>

Edbassmaster is one of the comedy stars of YouTube, the video sharing community on the internet. He makes very clever comedy skits—short videos, some of which have been viewed millions of times. Nearly 2200 people subscribe to his videos.

Earlier in 2007 one of the members of our French discussion group posted a pointer in the online discussion to a video praising Ed for agreeing to donate a kidney to his sister Maureen, who has been dealing with VHL since her teenage years. In December, Joyce interviewed Ed and Maureen for her radio show, the Powerful Patient, on webtalkradio.net.

In December 2007, according to the United Network for Organ Sharing (UNOS), there are 98,000 people waiting for a transplant. In 2007 only 21,000 transplants were performed—one fifth of the number of people waiting on the list. That means that most people will wait for five years for a transplant.

If one of these people is someone you care about, how can you shorten that wait time? A living related donor has the best chance of giving a healthy organ to a loved one. Even if the blood type doesn't match, there is the possibility of doing a "designated donation"—you give an organ to the person at the top of the list in your blood type group, and your loved one gets the next organ available in the correct blood type group.

Maureen began to have VHL issues at the age of 4, when she was diagnosed with VHL tumors in her eyes. She lost vision in one eye at the age of 4, and in the second eye at 17. She had her left eye removed last August at the age of 26. She has had multiple eye treatments, 3 brain surgeries and 8 partial nephrectomies, and a pho in her bladder, so she has had 27 surgeries.

"I had one too!" quipped Ed. Ed stepped up readily as a kidney donor. "There wasn't really a decision. When she needed one, I just knew I was going to do it. I knew that she would do it for me too. It wasn't a hard decision. Everyone on my side—my wife and everyone—were all for it as well.

"She's been through so many surgeries, along with my mother and brother, it has been hard to stand there and not be able to do anything. It has made me feel better to be able to contribute something to help with her pain and suffering."

The transplant was done October 23. By mid-December, both Ed and Maureen were doing well, and feeling great. Maureen had spent seven months on dialysis, during which time she was quite sick the whole time. She had many problems with her blood

pressure and severe itching. Like most people with VHL, she lost her kidney function abruptly.

Maureen had a very difficult time on dialysis. She was sick for the entire seven months of dialysis.

How did she feel after the transplant? "I felt so much better! I can't even put into words how much better I felt."

"I really didn't feel that way," interjected Ed. Maureen giggled.

"I was up and eating and walking and doing well," Maureen said, "and he was still in bed across the hall." The first day she was still recovering from the procedure, but by the third day she was feeling great. Her new kidney was working wonderfully, cleaning out all the toxins that had built up during her time on dialysis.

Ed too had relatively little discomfort. Certainly the first and second day he was recovering from the surgery, but on the third day they sent him home with pain medication, but he didn't feel the need of any pain meds at home. "It's really, really, worth it if anyone out there is considering donating a kidney. It felt so good to be able to help Maureen, it was well worth the couple of days of discomfort."

The donor and the recipient each have their own surgeon and surgical team, each dedicated to the health and well-being of their own patient. The transplant teams always take very good care of the donor. It's truly a team effort.

Ed has a number of fascinating characters, mostly based on real people he sees every day. He has been on YouTube for a year. Before that, he had done some stand-up comedy and impersonation. A couple of his videos have been on national television already. His video "Dog Drives Car!" will be on Fox this winter. It has been viewed nearly 2.5 million times on YouTube.

Doing comedy has become a family affair. "It helps us deal with the pain through laughter. With all these surgeries, I basically try to keep everybody's spirits up, keep them laughing."

"He keeps us laughing all the time," says Maureen. "Before every surgery, we're the only ones in the waiting area hysterical, laughing at Eddie. My Mom was going in for emergency brain surgery, and we were in the intensive care unit, all laughing so hard we actually got told we had to be quiet."

"When my surgeon came in the day after I had my kidney taken out," Ed reported, "my Mom was in the room. I winked at her and told her not to say anything. When he came in I started moaning. The look on his face was priceless. 'What's wrong?' he asked me, 'what's the matter?' and then in a normal tone of voice I said, 'Nothing, I feel fine.' He busted

out laughing. It was so funny. It was fun to see a surgeon so nervously laughing. He didn't know how to handle it—he had never seen anything like that before. That's how we deal with it."

Joyce mentioned that she had heard some families say that when they make weird jokes about their VHL experience, sometimes medical staff will say they don't seem to be taking this as seriously as it is. "Believe me," says Ed, "we understand completely how serious it is, we just don't want to dwell there. We prefer to keep laughing."

The "Dog Drives Car!" video was a spur-of-the-moment idea, shot in half an hour. Ed adopted a pug dog named Richard. One day Ed got the idea of the dog taking his human for a ride in the car, so he switched roles with his dog. In the video Richard the dog is driving the car, and Ed is in the back seat panting and jumping and hanging out the car window like a dog would do.

"I had my Dad put Richard on his lap and I sat in the back acting like the dog. The car next to us was driven by my Mom, and my cousin Eric filmed us. With a little bit of photography and editing, we almost pulled it off as the dog driving. It's humorous—you'd have to see it. That got about 2.5 million views and close to 10,000 comments. 99% of the comments are people trying to break down the video, figuring that it's not real and that we tried to pass it off as real. Obviously dogs can't drive, but thousands of people think I was trying to pull that off and fool the public. I just did it for fun, and the majority of people understood that and got a kick out of it."

So this was a whole family project. Ed manages to enroll the whole family in some of his projects. "I like to involve everybody—it's fun. A lot of my family members, cousins, like to get involved in it, especially now that it's gaining popularity. Each one of my videos is getting a minimum of 1000 views, so they're getting seen by a lot of people. It gets people excited and interested and they want to be part of it. It's fun—I love doing it."

Maureen agreed that it is great to have a brother doing comic relief for the family. They all have a good sense of humor, which they get from their mother. "Ed is not only a very funny comedian, he is a great, great husband, a great father, and obviously a more than great brother to do what he did for me. He's one of my best friends, definitely."

"I can say the same," Ed agreed. "We're very close. We talk on the phone at least six times a day."

"I feel very, very blessed to have a brother like him."

There's another brother as well, who has severe learning disabilities as well as VHL. "I like to call my mother and my brother VHL wannabees," Maureen says, "because they have only had two surgeries. They definitely have it, but not as severe.

It's been good for me because my mother has been able to be there for me all the time, taking me to the hospital and helping me. She would never have been able to do that if she had had it as severely as I do. Everything happens for a reason."

It sounds like the family is dealing with VHL in a wonderful way.

"Yes," says Maureen, "that's just who we are."

"Yes, Joyce," says Ed, "That's how we do. You've just got to joke about it. This will probably get edited out, but this is an idea of how we deal with the disease and try to laugh it off. One day I called my Mom and I said, 'Hey, Mom, I just heard on the radio that they found a new form of VHL and it affects women in their breast.' And she said, 'O my God, get out of here!' And I said, 'Yeah, and it's called von Nipple.' And she lost it for about ten minutes. She just couldn't stop laughing.

"That's just an idea of how we deal with things around here. It's terrible, but otherwise we would just mope about it and get depressed about it. And that wouldn't be good for anyone. It helps us—it helps me, I know that."

When you have a serious condition in the family, you have to find a way to incorporate that into your life. It is a part of your life. You can't just keep pretending it's not there. Humor is a wonderful way to be dealing with it.

Maureen's advice to others with VHL: "Don't let it affect you negatively, don't let it affect who you are."

"If anyone is thinking about donating a kidney," says Ed, "it's definitely well worth it. It's a once-in-a-lifetime experience—you can only do it one time—and it's definitely the right things to do. If you're contemplating it, definitely go for it, and you'll feel great about it afterwards."

We hope to see lots more of Edbassmaster on YouTube and on television.

"It still amazes me," says Maureen, "how many doctors know nothing about VHL. We need to fix that." Ed hopes to be part of the solution, bringing more public attention on VHL.

See <http://powerfulpatient.org/edbassmaster> to hear the radio broadcast, which includes clips from his skits. The program guide that accompanies the interview includes the pointers to the skits quoted in the show at YouTube. Page 9



Ed and Maureen, clowning around at the park – as usual.

Wrapped in love: Homemade quilts keep memories of mother close

Stepmom helps children embrace past, future

by Billy Watkins, bwatkins@clarionledger.com, Jackson, Mississippi, December 25, 2007

She was taken from her mother's womb during an emergency C-section March 31, 2003, a precarious three months shy of her scheduled arrival.

Anna Catherine Hutchison weighed 2 pounds, 2 ounces, had a head full of dark hair and even managed to breathe on her own.

Though she would spend two months in the Intensive Care Unit at River Oaks Hospital in Flowood, Anna would be fine. Her mother, Robin, would not. She died the day after the surgery at the age of 33 without ever laying eyes or hands on her daughter.

But Christmas is a time for good deeds, even miracles.

Today, Anna and her 5-year-old brother, Jimmy, of Madison can literally feel their mother wrapped around them, thanks to an idea conceived by their stepmom, Amanda.

Last week, each was given a memory quilt made of Robin's dresses and maternity clothes. The quilts also include pictures of Robin at various stages in her life - from 6 weeks to adulthood.

"It started when Anna came to (husband) Mark and me crying one day back in May," Amanda says. "She said, 'I miss my Mama Robin. I never got to say goodbye. I never got to touch her.' That's a lot for a 4-year-old to cope with.

"I was talking to a friend on the phone in Memphis about it the next day, and she and I started talking about what I could do to fill this void. That's when we started talking about the memory quilts."

Over the next two months, Amanda duck-taped together a rough design of the quilts. Diann Loper, a professional quilter in Purvis, followed Amanda's basic suggestions but added her own touch. Kathy Bizzarri of Jackson embroidered the words "my arms around you" at the top, and the children's names at the bottom.

They were made just right for kids their age - 60 inches by 50 inches. Anna and Jimmy can completely wrap themselves in them.

"Maybe it won't have a major impact on them right now," Amanda says, "but when they're teenagers trying to find their way, it'll be more of a comfort to them. And this is something they can pass down for generations. Mama Robin will never be forgotten."

Mark Hutchison had his life planned out to the tiniest detail. He laughs about that now.

"You plan, you plan ... life throws things at you

that you don't see coming," he says, shaking his head.

He and Robin met in 1993 and dated for three years before marrying. An attorney who has handled numerous divorce cases, "The last thing I wanted was to ever go through a divorce ... I wanted to make sure I was marrying someone forever. Robin was that person."

They waited six years to have children "because I wanted to make sure we could afford them," he says.

Jimmy was born in February 2002. Robin had no problems during the pregnancy or delivery.

"But later that year, Robin started having headaches," Mark says. "And when I say headaches, I mean severe headaches."

As a teenager, Robin underwent surgery to remove a benign brain tumor. "The only way you'd ever know it was a tiny scar at the base of her skull," Mark says. "She seemed fine."

But the headaches got worse. Six months pregnant with Anna, Robin went in for a CT scan. Something that looked like a tumor showed up, and Robin was scheduled to see a specialist the next day.

"That night, she had a seizure," Mark recalls. An ambulance rushed her to River Oaks. Tests revealed four brain tumors. Surgery was scheduled to remove the largest one.

"But they told me that even if the surgery went well, she could be paralyzed ... they basically told me that she wouldn't be the same person I brought to the hospital," Mark says.

But there was even more for Mark to ponder: Doctors diagnosed Robin with VHL, a genetically transmitted disorder that involves abnormal growth of blood vessels. Which meant that Jimmy and the baby Robin was carrying faced a 50 percent chance of also having VHL.

Only once in the hospital did Robin wake up. She told Mark she had decided on a name for the baby:

Bring the Whole Family!

The Annual Meeting this year is in Orlando. See page 1 for information, and page 15 to sign up!

(Can you keep a secret? We will have a professional Clown at the meeting!)

Anna Catherine.

Less than 48 hours later, Robin was gone.

Almost immediately, Mark began attending a Parents Without Partners Sunday school class at Colonial Heights Baptist Church in Jackson. He was lonely, he needed somebody to talk with. He shared his story that first Sunday.

Amanda, who had gone through a divorce and survived the 2001 tornado that ripped through the Fairfield subdivision in Madison, was a member of the class. Every week, a half dozen members would meet for lunch.

Amanda and Mark struck up a friendship.

"I felt sorry for him," Amanda says. "He looked a little lost."

Their friendship grew, but Amanda finally told Mark that she and her son, Colton Comans, now 14, were moving to Fayetteville, Arkansas, where she wanted to pursue new job opportunities and be near her sister. Amanda had already purchased a house there.

"He told me, 'Go ahead and move, but you'll be back,'" Amanda says, smiling. "And he was right."

They married Dec. 22, 2003, a little more than eight months after Robin's death.

"People thought we were crazy," Amanda says. "And, at first, I wanted to make sure Mark had some time ... but it just felt right."

"I think about this a lot," Mark says. "Amanda married me, realizing she might have to take care of two children with VHL for the rest of her life. But she never made it an issue. I feel like I'm so lucky to have met two wonderful women in one lifetime."

Thirty-nine days after Mark and Amanda said their vows, a letter arrived. Test results were in.

Neither Jimmy nor Anna had VHL.

Just a couple of weeks into their marriage, Amanda had the house to herself one day - the same house Mark and Robin moved into in 2002, off Bozeman Road in Madison, where they still reside.

Mark was at lunch. Colton was at school. Anna and Jimmy were napping. Amanda stood in the



Anna wrapped in her quilt. Photo by Barbara Gannett.

den, staring out toward the lake, when she noticed something in the backyard.

While all else was still, the baby swing moved back and forth, back and forth ... and then stopped.

It did it over and over.

"I couldn't take my eyes off of it, and I honestly felt like Robin was out there," Amanda says.

Amanda didn't share the story with Mark for a few days.

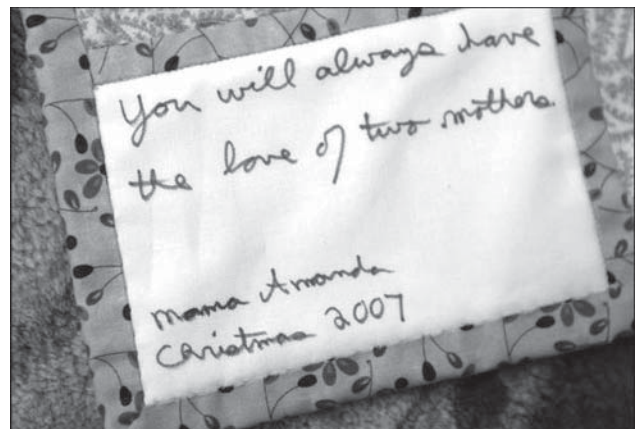
"But when I told him, he said that's the last thing Robin did before she went to the hospital - she pushed Jimmy in that swing," she says.

Mark also told Amanda that beneath that swing, shortly after Robin's death, is where he had found a diamond locket Robin had lost.

"It all sort of made sense to me then," Amanda says. "I just remember staring at the swing, knowing it was Robin. And I remember saying out loud, 'Go have fun. I'll take care of things here.'"

Two little children wrapped in their mother's clothing would attest to that.

To comment on this story, call Billy Watkins at (601) 961-7282, or write to Amanda at us-ms@vhl.org



Quilt label by Amanda. Photo by Barbara Gannett

Amanda is the Chairman of the Mississippi chapter of the VHL Family Alliance. You can write to her at us-ms@vhl.org

Our thanks for donations from...

Michelle Andjus

***The Thank-You List appears only in the print
version of the newsletter.***

***The Thank-You List appears only in the print
version of the newsletter.***

**The Thank-You List appears only in the print
version of the newsletter.**

Board of Directors, Staff

Joyce Wilcox Graff, +1 617 277-5667 ext 4
Executive Director, Editor VHLFF
Hannah Costa, +1 617 277-5667 ext 4
Administrative Assistant
Robert Cochrane, *Publication Services*
Jessica Costa, *Research Services*
MaryLou Linn, *Member Services*
Dieu-My Phan, *Children's Services, Grants*

Bruce Weinberg, Esq., +1 (323) 854 6664
Chairman of Board
Camron King, +1-916-549-6568
Vice Chairman of the Board
Christol Sorrell, +1 518-570-5064
Treasurer
Linda Berk, +1-858-866-0669
Secretary
Sunny Greene, +1-703-578-1181
Director

Robert Lydon (*died 12/07*)
Director
Jeanne McCoy, +1-864-292-3488
Director
Tom Rath, +1-202-715-3030
Director
Bill B. Scheitler, +1-712-546-6840
Director
Robert E. Schoenhals, +1-248-576-8070

International Leadership

Hélène Sultan, France, +33 1 49 59 67 29
VHL France, france@vhl.org
Gerhard Alsmeyer, +49-5931-929552
Chair, Verein für VHL Erkrankung b.F., Germany
M. Luisa Guerra, Italy, +39 (143) 643220
Chair, Alleanza VHL
Vibeke Harbud, Denmark, +45 46 75 70 33
Chair, Foreningen af Von Hippel Lindau Patienter
Chris Hendrickx, Belgium, +32-3-658-0158
Chair, VHLFA Belgium

Pierre Jacomet, +56 (32) 824461
Chair, Chile & Latin America
Helga Suli-Vargha, +36 (20) 34485 88
Chair, VHLFA, Hungary
Valerie & Jon Johnson, +64 (9) 534 8098
Chairs, VHLFA, New Zealand
Jennifer Kingston, Paul & Gay Verco
VHLFA, Australia +61 (2) 9475-1441

Susan Lamb, +1 (519) 735-0236
Jill Shields, +1 (519) 268-1567
Co-Chairs, Canada, canada@vhl.org
Hanako Suzuki, japan@vhl.org
Co-Chair, Japan
Markus Jansen van Vuuren, +27-11-407-4047
Chair, South Africa, SouthAfrica@vhl.org
Erika Trutmann, info-ch@vhl-europa.org
Chair, Switzerland

Medical Advisory Board

Lloyd M. Aiello, M.D., *Beetham Eye Inst
Joslin Diabetes Center, Boston, Mass.*
Debra L. Collins, M.S., *Division of Genetics
Univ. of Kansas Med Center, Kansas City, KS*
Eric Jonasch, M.D., *Genitourinary Med. Oncology
M.D. Anderson Cancer Center, Houston, TX*
Col. James M. Lamiell, M.D., *Clinical Invest. Reg
U.A. AMEDD Cntr & School, Fort Sam Houston, TX*
Richard A. Lewis, M.D., M.S., *Ophthalmology
Baylor College of Medicine, Houston, TX*
John A. Libertino, M.D., *Institute of Urology
Lahey Clinic Medical Center, Burlington, MA*

Joseph A. Locala, M.D., *Psychiatry & Psychology
Cleveland Clinic Foundation, Ohio*
Eamonn R. Maher, M.D., *Genetics
University of Birmingham,
Edgbaston, Birmingham, England*
Virginia V. Michels, M.D., *Medical Genetics,
Mayo Clinic, Rochester, Minnesota*
Haring J. W. Nauta, M.D., *Chief of Neurosurgery
Univ. Texas Medical Branch, Galveston, Texas*
Hartmut P. H. Neumann, M.D., *Nephrology
Albert-Ludwigs University, Freiburg, Germany*

Andrew C. Novick, M.D., *Chair, Urology
The Cleveland Clinic, Cleveland, Ohio*
Edward H. Oldfield, M.D., *Prof. Neurological Surgery
University of Virginia Med Center, Charlottesville, Virginia*
Stéphane Richard, M.D., *Neuro-Oncologie
Hôpital Kremlin-Bicêtre, Paris, France*
Armand Rodriguez, M.D., *Internist, Florida*
R. Neil Schimke, M.D., *Dir. of Genetics
University of Kansas Med Center, Kansas City, Kansas*
Robert B. Welch, M.D., *Chair Emer,
Ophthalmology, Greater Baltimore Medical Center, Baltimore,
Maryland*

VHL Family Forum, Newsletter of the VHL Family Alliance and the Cancer Research Fund / VHL

Volume 16, Number 1, January/February 2008, ISSN 1066-4130

Editor: Joyce Wilcox Graff, 1-617-277-5667, extension 4

E-mail: info@vhl.org; Tel: 1-617-277-5667; Fax: 1-858-712-8712

Internet website <http://www.vhl.org>

Toll-free in the United States and Canada: 1-800-767-4VHL

2001 Beacon Street, Suite 208, Boston, MA 02135-7787 U.S.A.

©2008 by the VHL Family Alliance. All rights reserved. Photocopy of the entire issue is permitted. Reproduction for publication requires written permission in advance. Distributed to members of the VHL Family Alliance, supported by dues and fund-raising. Advertising is accepted. We welcome your comments, suggestions, ideas and submissions. Copyrighted works or their modifications must be accompanied by the copyright notice. Opinions expressed by the authors are not necessarily those of VHLFA.

Postmaster: Please send address changes to VHL Family Forum, 2001 Beacon Street, Suite 208, Boston, MA 02135-7787

Membership & Registration

Name: _____

Address: _____

City: _____ State: _____ Zip: _____ Country: _____

Home phone: _____ Work phone: _____

E-mail: _____

Preferences: We can send you more timely information at a much lower cost via e-mail. We will continue to send a paper newsletter every three months. |

Please be sure to send us any address corrections, as returns cost us \$2 each.

I would like to receive periodic e-mail alerts Please send the paper copy as well



Orlando, here we come! Prices go up \$20 per person after April 30.

Please register ___ people for Orlando at \$79 each for conference and banquet (\$99 after April 30)..... \$ _____

Discount for second person in family (spouse/partner/friend/etc) subtract \$20 - \$ _____

Extra banquet tickets for those not attending the conference #___ at \$20 each \$ _____

Videos and Wristbands: Wear your wristband proudly! Hope ... Strength ... Vhl.org



Call the office at 800-767-4845 to make arrangements for bulk orders of wristbands.

Please send me ___ # wristbands. I enclose my donation of at least \$5 for each..... \$ _____

Send me the DVD & handouts from ___Boston 2007 ___Houston 2007 at \$50 each..... \$ _____

A Gift to the VHL Family Alliance is a living gift of love - Sent with Hope for a Cure!

Remember VHLFA when you want to celebrate an occasion or a loved one (minimum \$10 per honoree)

in Honor of in Memory of _____

Acknowledgement card to be sent to: _____

Payment information:

Annual dues: \$25 per mailing address (\$35 outside U.S.**)..... \$ _____

** Please send checks in US funds, or pay via credit card. For checks in funds other than US, please add \$15 for bank fees.

Yes, I want to help fund another research grant! My additional contribution: \$ _____

My check is enclosed Please charge my Visa or Master Card: \$ _____

This is a one-time payment Monthly pledge to be charged to my card for ___ months.

Name as it appears on the card: _____

Card number: _____ Expiration: _____



Hold the Date!

It's our
15th Birthday!

You're Invited



... to our Birthday Celebration Meetings!

**U.S. Annual Meeting,
Orlando, Florida,**

Saturday, June 28, 2008

**with Birthday Banquet Saturday 6:30 pm,
funtime Sunday**

make your reservations early - prices go up April 30

**International Medical Symposium,
Roskilde, Denmark**

September 4-6, 2008

**abstracts are being accepted now
see vhl.org/conf2008 or vhl2008.dk**

explore the ancient capital of the Vikings



watch vhl.org/meetings for other
regional meetings, including:

Los Angeles, March 1, 2008

Johannesburg, S.A., March 15, 2008

Manchester, England, UK, April 12, 2008

Boston Marathon Party, April 21, 2008

**Celebrate with us by creating your own
fundraising page at**

www.firstgiving.com/vhl



Newsletter of the VHL Family Alliance
2001 Beacon Street, Suite 208
Boston, MA 02135-7787

ADDRESS SERVICE REQUESTED

NONPROFIT ORG.
U S POSTAGE
PAID
Tampa, FL
Permit No. 2397