

To Scan or Not to Scan?

Editor's Note: There is a great deal of conflicting information in the press these days about the use, overuse, and underuse of scans, contrast media, and radiation. While there are certainly valid concerns, as with so many areas of life we have to balance the need to know versus the cost of not knowing – the risks attached to the imaging versus the risks of not doing the imaging – the need for a good picture of what is going on, balanced against the potential side effects of the procedure.

When they talk about the overuse of imaging in the press, they are usually talking about a child who comes in with a skinned knee and they do 20 or more scans to check that leg. Were those really necessary? Or could the doctor have applied some hands-on evaluation and common sense? Especially in children, we need to be careful not to over-use radiation.

Having lived with VHL for 45 years, I can tell you that the single most dramatic beneficial difference between managing VHL today and managing VHL in 1970 is the existence of medical imaging. Thanks to scans, we can see VHL issues brewing, make a good plan of action in a calm manner, and truly *manage* VHL. The benefits in most cases far outweigh the risks. Nonetheless it is important for you to understand the risks so that you can manage them appropriately in your own case and keep them in proportion to the benefit.

We are privileged to have two excellent articles from the University of Michigan to help you understand two important issues.

First, Dr. N. Reed Dunnick, one of the leading radiologists in the country, and Dr. Mitchell M. Goodsitt, a radiation physicist on his team, explains how the amount of radiation is calculated, and how to understand the amount of radiation you are subjected to – both in daily life and in medical scanning.

Second Dr. Richard Cohan, a member of Dr. Dunnick's department who specializes in abdominal radiology, helps us calculate our risk of developing a rare but nasty complication -- nephrogenic systemic

fibrosis (NSF).

There are no simple answers to either of these questions. We hope that this additional information will help you have a constructive conversation with your physician and your imaging department about the best approach for you. *Articles begin on page 2*

Lots of Exciting News Inside this issue!

Important Information for you and your doctor about the pros and cons of imaging. See pages 1-3

Time to **Renew your Membership** and **Vote** for five new members of the Board of Directors. We need your vote! See pages 10-11

Hear from two of our younger members about their efforts to raise awareness of VHL and find a cure! pp. 6-9

Participate in one or more of the exciting events happening on **International VHL Day May 9, 2009** – see page 9

Plan to come to the Annual Meeting of the VHLFA Saturday, June 27, in Anaheim, California See page 16

Consider our new Green Membership, to help conserve funds, cure VHL, and save the Earth all at the same time!

Everyone whose membership is paid in full for 2009 can receive one free copy of the new Children's Book on VHL -- to be presented for the first time in Anaheim! -- See page 12

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It Takes Us All!

Radiology Examinations – Weighing the radiation risk

By Mitchell M. Goodsitt, Ph.D., Professor of Radiology, and N. Reed Dunnick, M.D., Chair, Dept of Radiology, University of Michigan, Ann Arbor, Michigan USA. Dr. Dunnick is also a member of the Academy of Radiology Research.

When deciding whether to obtain a radiology examination, the potential benefit of the diagnostic information must be weighed against the risks of the study. For many radiology exams, there is a very small increased risk of cancer due to x-ray exposure.

All of us are exposed to x-rays every day of our lives. This exposure is due to external radiation from sunshine, cosmic rays and radioactive isotopes that are in the water and soil, and due to internal radiation from radioisotopes within our bodies.

The dose we receive from x-ray examinations can be related to the time it would take to receive the same dose from natural background radiation. A table of these values for some common x-ray studies appears below (See Figure 1).

Figure 1:

X-ray study	Time to get same dose from nature
Dental, intra-oral	2 days
Chest x-ray	12 days
Lumbar spine	3 months
Head CT	8 months
Abdomen CT	1.5 years

As you can see, the doses from most exams are low and equivalent to short times of exposure to background radiation. Even the highest value in the table (1.5 years) would be equal to 0.15 years or 1.8 months in other areas of the world where background radiation is 10 times greater. [See note 1.]

The risks of ionizing radiation are of greater concern to people (such as those with VHL) who get repeated CT examinations. To put these risks into perspective, consider the following: From population studies, it has been determined that 1 in 5 or 20% of us will eventually die from cancer due to non-radiation sources. There is very little data available for risk estimates at the CT dose levels, and we do not know if there is a radiation

dose threshold for cancer. Very conservative risk estimates are obtained by 1) disregarding any threshold, and 2) linearly extrapolating cancer risks down to the CT dose levels from the risks observed at much greater dose levels for survivors of the WWII atomic bombs. According to those estimates, the additional risk of cancer from a CT exam of the abdomen is 0.02%. Thus, if you had one CT scan of your abdomen every year for 30 years, your added risk of eventually dying from cancer due to x-rays would be 0.6%. Your total risk of dying from cancer would be 20.6% compared with the population average of 20%. In summary, even conservative estimates indicate the added risk of multiple CT scans of the abdomen is small and almost

“... if you had one CT scan of your abdomen every year for 30 years, your added risk of eventually dying from cancer due to x-rays would be 0.6%, or one-half of one percent.”

insignificant.

There are several things you can do to further minimize your risk. First, talk with your physician to make sure each examination is needed. Second ask if the CT examination is being supervised by a board certified radiologist. These physicians design the examination to use the least amount of radiation necessary. Third, you may want to consider an examination that does not use ionizing radiation such as ultrasound or magnetic resonance imaging (MRI). Ultrasound does not usually provide the detail needed to examine the kidneys of VHL patients. MRI is more expensive, often less readily available and may require sedation for claustrophobic patients, but is as useful as CT in detecting small renal lesions.

In summary, x-ray examinations including CT are generally safe. Any small risk is outweighed by the tremendous benefit of the diagnostic information that is obtained.

Notes: 1. The level of natural background radiation varies depending on location, and in some areas the level is significantly higher than average. Areas of high concentration include Ramsar, Iran; Guarapari in Brazil, Kerala in India, the northern Flinders Ranges in Australia and Yangjiang in China. In Ramsar a peak yearly dose of 260 mSv has been reported. Source: Wikipedia: “Background Radiation.” Additional information is available at <http://www.radiologyinfo.org/>

Issues in Imaging the Kidney

Compiled with the kind assistance of Richard Cohan, M.D., Abdominal Radiology, University of Michigan, Ann Arbor.

Introduction

People with VHL are at increased risk for kidney cancer. With our worldwide advisors, we have worked out a system of “watchful waiting,” monitoring the growth of kidney tumors, and ablating or removing them before they reach the 3 cm level. While size is not a precise measure of the metastatic potential of a tumor, it is the best indicator we currently have, and it has proved to be very reliable.

In order to do that “watchful waiting,” it is important to obtain images of the kidney on a regular basis (with many recommending that this imaging be performed once a year) to check on the size and rate of growth of the kidney tumor(s) being watched. Until a few years ago, we were using both CT and MRI relatively freely.

VHL tumors of the kidney are highly vascular, and are best seen when a contrast medium has been administered. The contrast is a substance injected into your bloodstream that colors the blood vessels and gives the radiologist a better outline and view of the tumor, and a more accurate measurement of its size.

Contrast medium for CT and MRI

Although the contrast medium used for CT (iodinated contrast medium) is well tolerated in people whose kidneys are functioning normally, it has been known for many years that this contrast can damage the kidneys in people who already have compromised renal function. Due to this problem, many physicians turned to the use of contrast-enhanced MRI, as an alternative imaging study to evaluate the kidneys in people with kidney disease. (MRI contrast medium, which is completely unrelated in structure to iodinated contrast material, does not have this same potentially damaging effect on the kidneys as does iodinated CT contrast, when the MRI contrast medium is used at standard doses).

Nephrogenic Systemic Fibrosis

Beginning in about 2003, reports began to emerge of a nasty complication that occurred among people with poor creatinine clearance, a complication called nephrogenic systemic fibrosis (NSF). In April 2006, it was first noticed that most patients who developed NSF had received an injection of contrast medium for MRI (which are gadolinium-based contrast material) within weeks to several months of their developing symptoms. The association between bad kidney function, MRI contrast-medium, and NSF has since been confirmed in many other studies.

NSF produces stiffening of the skin and can

become so severe that affected individuals lose their ability to move their joints and may even become immobile. NSF can sometimes affect internal organs as well.

VHL and NSF

The question for people with VHL is: how can we get the best possible images of the kidneys without putting the kidney at risk, and without causing NSF? Since the kidneys can only be evaluated well if some type of contrast material is administered, the option of doing an unenhanced CT or an unenhanced MRI, as an alternative, is not a very good one.

It is important to emphasize that people with VHL who have normal kidney function are not at any greater risk of NSF than anyone in the general population. VHL is not a “renal disease” in that it does not erode kidney function. You can have multiple kidney surgeries and still have perfectly good renal function. But there are some people with VHL who will be at increased risk for developing NSF, and these people should confer with their own urologist and radiologist about how best to proceed. These are people who have lost so much kidney tissue, due to prior surgeries, that their remaining kidney function is severely limited.

How is kidney function measured?

There are two ways in which kidney function is now measured. The older method involves obtaining a serum creatinine level. A normal creatinine level is usually about 1.4 mg/dl or lower. A higher creatinine level indicates that your kidney is not functioning normally. Many physicians believe that a measurement of creatinine clearance or glomerular filtration is more accurate. This is not easily obtained directly. Instead, formulae have been developed that estimate glomerular filtration rate (eGFR). These formulae use several variables, including the serum creatinine level, but also age, gender, and race. Normally, the calculated eGFR should be over 90 ml/min. A lower eGFR indicates that your kidney is not functioning normally.

As far as we know at this time, all MRI contrast media can be used safely, without concern for NSF, in patients who have normal or even mildly elevated serum creatinine levels or eGFR levels of 30 ml/min or higher. It is only those patients with severe renal insufficiency/failure who are at increased risk (GFR < 30 ml/min).

Does the type of MRI contrast medium matter?

The incidence of NSF in high risk patients also may vary depending on which gadolinium-

based contrast medium was used. The chemical composition of each brand of MRI contrast media is different. To our knowledge, there are several brands that do not yet have a single case of NSF definitely associated only with them, so use of one of these agents may lower one's risk.

So what to do?

At the present time, many experts have made a number of specific recommendations. First of all, if you have any concerns about your kidney function, you can ask your physician to get a blood test so that your serum creatinine level can be determined and your eGFR calculated.

1. If your kidneys are working normally:

If you have normal kidney function, you can be imaged with either contrast-enhanced CT or MRI without fear of damaging your kidneys or of risking NSF.

2. If you are already on dialysis and further kidney damage is not a concern:

If your kidney function is damaged so badly that you are already on permanent dialysis AND there is no concern about making your kidneys function more poorly, your kidneys can (and probably should) be imaged by CT rather than MRI, with iodinated contrast medium administered.

3. If you have mild or moderate kidney damage:

If you have mild kidney damage (with an eGFR of 30 ml/min or more) a contrast-enhanced MRI is probably preferable to a contrast-enhanced CT. The risk of NSF here is minimal.

4. If you have severe kidney damage and are not yet on dialysis:

If, on the other hand, you have severe kidney damage (with an eGFR < 30 ml/min) and are not yet on permanent dialysis, then the decision is more difficult and should be made after carefully consulting with your physicians. If a decision is made to perform a contrast-enhanced MRI, it is suggested that one of the contrast agents that has not been observed to produce NSF be used and that the standard recommended doses of the MRI contrast-agents not be exceeded. This means not having magnetic resonance angiography (MRA), which may use higher doses of MRI contrast medium.

What is the actual risk in high-risk populations?

It should be remembered, however, that even in high-risk patients, and even when using high doses of the brand of contrast medium with the highest reported association to NSF, the risk is no more than 5-10%. More than 90% of patients injected with this contrast medium did not develop NSF. We now believe that when alternative MRI contrast-agents

are utilized at lower doses, the risk of developing NSF is probably much lower.

For example, the radiology department at Johns Hopkins University (JHU) School of Medicine reviewed all their patient cases (not just VHL) from 2003 through 2006 and worked to reduce the risk of NSF. They published the recommendations shown in Figure 1. Using these guidelines, they reduced the rate of NSF at JHU from 36.5 cases per 100,000 gadolinium-enhanced procedures to four cases per 100,000, "suggesting that the new policy was associated with a significantly lower incidence of NSF." Using the precautions mentioned above, at the University of Michigan, no cases of NSF have been reported since late 2006, even though contrast-enhanced MRI is still performed in some patients with severe kidney disease.

Again, there is no pat formula that works for everyone, and we will continue to learn as new information becomes available. We encourage you to have a conversation about this with your urologist and with your radiologist. You and your team should make the decision together as to how to proceed.

Questions to ask your doctor:

- What is my estimated glomerular filtration rate?
- Do you recommend a CT or an MRI?
- Am I at increased risk of developing NSF?
- What precautions do you recommend I take to make sure that my risk of further damaging my kidneys or of developing NSF is minimized while getting the best possible images for my urologist?
- Would it be helpful for me to drink more water before or after the imaging procedure? (this is not a good idea for everyone, so be sure to ask your doctor)

We should also add that this article is not intended as an absolute recommendation, but rather as mechanism for providing you with information to assist you in having an informed discussion with your own doctors, so that you can participate in determining the best course of action for you.

References:

1. Wayne Forrest, "JHU Study: Screening patients prior to MRI lowers NSF risk", <http://www.auntminnie.com/index.asp?Sec=sup&Sub=mri&Pag=dis&ItemId=84478>
- Perez-Rodriguez et al, "Nephrogenic systemic fibrosis: incidence, associations, and effect of risk factor assessment-report of 33 cases." <http://www.ncbi.nlm.nih.gov/pubmed/19188312>
- American College of Radiology, Manual on Contrast Media, see esp. the chapters on "Contrast Nephrotoxicity" and "Nephrogenic Systemic Fibrosis". http://www.acr.org/SecondaryMainMenuCategories/quality_safety/contrast_manual.aspx

Figure 1: JHU School of Medicine policy for the prevention of NSF

Creatinine/estimated glomerular filtration rate (eGFR) is to be obtained for patients at risk for reduced renal function, including people who have:

Age \geq 65 years

Diabetes

History of renal disease*/renal transplantation

History of liver transplantation, hepatorenal syndrome

Other medical conditions as determined by attending radiologist [In acute renal failure, eGFR may be inaccurate and gadolinium use should be avoided.]

Patient on dialysis, or estimated GFR < 30 mL/min/1.73 m²:

Radiologist to determine if gadolinium use is essential for diagnosis. Confirm that alternative tests are not available.

Patient consent for gadolinium is obtained.

Maximum recommended dose is 0.1 mmol/kg gadolinium.

If patient is on hemodialysis: dialysis to be scheduled same day. Dialysis to be repeated 24 hours later. For patients who are on dialysis, there must be verification that the patient will receive dialysis as soon as possible after the MRI. Two dialysis sessions separated by one day are recommended.

If patient is on peritoneal dialysis, use of gadolinium contrast is strongly discouraged unless highly necessary for diagnosis. Nephrology should be consulted to determine if hemodialysis can be performed.

* **Editor's Note:** VHL is not a "renal disease" in that it does not in itself diminish kidney function. Repeated kidney surgeries may diminish renal function. eGFR is a measure of renal function that will tell you and your doctor whether your kidney function is normal or diminished. See article for details.

Notes on eGFR:

eGFR should be current (e.g., within the last month). eGFR by the Modification of Diet in Renal Disease (MDRD) formula is based on serum creatinine, age, gender, and ethnicity.

Source: Radiology (February 2009, 250:2, pp 371-377) and JHU School of Medicine

Anaheim, Here we Come!

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See page 16 for details

Why be a member of VHLFA?

We need YOU. There are four bubble people in our logo. They represent the

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AND

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working together to diagnose, manage, and ultimately cure VHL.

Besides, there are two books coming out within the coming year for which members will enjoy discount prices. You can will be able to buy them on Amazon.com, or you can buy them half-price from VHLFA with your member discount.

VHL for Children

VHL Handbook, next edition

Ariel's Story

by Ariel R., New Jersey

About six years ago, I had just started high school. My brother, Daniel, was a senior at the same school, and my sister, Shelby, was ten years old. Daniel was in the middle of college applications, I was introduced to the woes of a bottom locker, and Shelby was happily in fifth grade. We were a very typical family; our biggest heartbreaks were when our great-grandparents passed away. We were close, and we had two loving parents to prove it. However, when Shelby started complaining of seeing "black swirly things," our lives changed forever.

Daniel always wore glasses, so my mom took Shelby and my brother to the optometrist for a checkup. I was home doing schoolwork when Daniel called the house. "Shelby has a tumor in her eye. She needs to go to another doctor." I thought he was joking, and I even laughed; I didn't believe him. How could my ten-year-old sister, the one with whom I shared a room and played Barbies with not so many years ago, have a tumor? My mother rushed Shelby to a retinal specialist, and from there, she went to Will's Eye Hospital In Philadelphia. Shelby had a collection of leaking tumors in her left eye, and they were causing her retina to detach. She needed surgery right away, in order to save her left eye's sight.

The formation of Shelby's tumors were suggestive of a rare genetic disease called von Hippel-Lindau Syndrome (VHL). A person with VHL is lacking the von Hippel-Lindau tumor suppressor gene; this gene produces a protein that helps to control cellular growth. Without this gene, and hence, without the protein, tumors are predisposed to grow in places rich with blood vessels, like the brain, eyes, spine, pancreas, adrenal glands, and kidneys. In the kidneys, these tumors are cancerous. If Shelby had VHL, then she had most likely inherited it from one of my parents. If one of my parents had VHL, then it was possible that Daniel and I could have it; the chance of passing on the disease to a child is fifty percent.

After Shelby's suspected diagnosis, a blood test confirmed that she had VHL. The rest of my family underwent genetic testing, and it turned out that my Mom, who was 42, also had VHL. Even more rare, she was a new mutation; she was the first person in our family to have VHL, and none of her parents had passed it on to her. This was so much news for my family to swallow; the only positive note was that Daniel and I had not inherited the disease, and my father did not have it, either.

The VHL diagnosis left my family with a lot of

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questions. What other problems might Shelby have besides these eye tumors? If my mom has gone 42 years not knowing she had this disease, is it really that serious? How do we find out if there are any other problems? Can we just wait and see until a problem crops up? How come we never heard about this disease before? What in the world is going on?

We did get some of the answers we wanted. After consulting the VHL Family Alliance (VHLFA) and doing some of her own intense research, my mom consulted the National Institutes of Health (NIH) in Bethesda, MD (none of our doctors at home knew what to tell her). Shelby and my mom were lucky enough to be accepted into a clinical study of VHL at NIH. Every six months, they go for neurological scans. My mom also goes for kidney and abdominal scans at the NIH, while my sister gets those done at Sloan-Kettering in New York City. My dad and I often accompany them to provide emotional support.

A typical visit to the NIH involves getting blood work, two brain and spine MRIs (one for my mom, one for my sister, both over an hour long), the corresponding neurological exams (can you walk in a straight line? can you touch your finger to your nose with your eyes closed?), a 24-hour urine test for my mom (to check for catecholamines, which are secreted by tumors in the adrenal glands called pheochromocytomas), sometimes a hearing test (some VHL tumors can result in hearing loss), and an abdominal MRI or CT scan or ultrasound (depending on how long it has been from the previous abdominal scan) to check for growths in the kidneys or pancreas.

These tests are essential for all VHL patients, not just the ones whose tumors are being tracked by renowned doctors for a clinical study. When a tumor or problem arises, it is important to either nip that growth in the bud, or track it to see if it becomes a problem. A growth that might be nipped in the bud is a retinal tumor; likely, doctors will want to treat this tumor before it leaks and causes the retina to detach. Meanwhile, doctors might decide for a kidney tumor to be tracked. For example, if there is a kidney tumor, once it reaches the size of three centimeters, it should be removed; once past this size, it has the greatest potential to metastasize. However, the doctors have to be careful to treat each kidney carefully, to maintain its function. For reasons such as these, it is important for VHL patients to make sure their medical team is knowledgeable about VHL.

I am now a junior in college. Come spring, Daniel will graduate from the University of Virginia with a master's degree in English education. Shelby is the typical high school junior; she just took her SATs, she would rather not think about college, and she is very involved both in and out of school, in dance, singing, and theater. She is also blind in her left

eye. Since the initial VHL diagnosis, she has had numerous eye surgeries. I will explain some of them: photodynamic therapy to get rid of recurring tumors, vitrectomies (removal of the eye's vitreous humor) to help treat glaucoma (a vicious complication from the many surgeries) and repair a detached retina, removal of her eye's natural lens, cryotherapy (freezing of tumors to destroy them), and the insertion of a shunt to further help treat glaucoma. Unfortunately, all of these surgical efforts failed to save her eye; eventually, Shelby will need this eye removed and a prosthetic put in its place. Her eye has become so weak and damaged from the tumor and its complications, and though she feels no pain now (just occasional stings and aches), it will one day become very painful. People sometimes ask her why her eye is so red, but it is not easy to very briefly explain what has happened in the past six years.

Shelby has not been deterred by these challenges, though; she had a starring role in our dance school's yearly production this past spring, and she just got accepted into the National Honor Society. I aspire to have as much charisma and strength as she does.

Some people say Shelby saved my mother's life. My mother went 42 years not knowing she had VHL. After my mom went for her first set of scans, her doctors found a few brain tumors, numerous growths on her pancreas, and too many growths to count on her kidneys. Shelby also has a few brain tumors, but thus far, nothing on her kidneys. To this day, both my mom's and sister's brain tumors are being watched until they cause neurological symptoms (dizziness, nausea, difficulty with balance, etc) and need to be removed.

When I began my senior year in high school, the doctors became concerned with one of my mother's kidney tumors. It was nearing three centimeters and would soon have to be taken out. On June 6, 2006 (known as 06-06-06 by some, the supposed day of the Apocalypse), my mother underwent a laparoscopic partial nephrectomy at the NIH. The doctors removed the offending tumor and got rid of as many of the smaller tumors as they could. My mom came home from the hospital, back to New Jersey, after three days, just in time for my very last dance recital. The surgery was not a piece of cake to go through, but my family could not believe how soon my mom was able to come home. Even more shocking was that if we had never learned my mom had VHL, she would have never known she had these growing tumors on her kidneys until after the cancer had metastasized. Truly, in spite of the difficulties, my family has been extremely lucky.

My family's experience with VHL has changed everyone's lives. For one thing, it brought us all closer, especially as our loved ones were taken



Ariel, Lisa, and Shelby

away from us; my grandfather passed away from melanoma when I was sophomore in high school, and three years later, during my freshman year of college, my grandmother passed away. My grandmother was very ill, and she succumbed to a combination of lupus, strokes, and kidney trouble. I was very unhappy with how her doctors treated her, especially compared to the compassionate care my mother and sister received at the NIH.

The following summer, when my sister officially lost vision in one eye and learned that it would have to be removed, I started to rethink my career aspirations. I was a sophomore studying industrial engineering, but I was not entirely satisfied. Interested in learning more about medicine, I applied to be a summer research intern at the NIH. I wrote to one of the doctors who participated in my mother's kidney surgery, and I was so excited and grateful when he invited me to work with him over the summer. Through this experience, I was able to learn more about the field of medicine. Plus, I learned a lot more about VHL, as well as other rare inherited diseases that can cause kidney cancer. From the two months I spent at the NIH, I realized I wanted to be a doctor. I have some time ahead of me before I reach medical school, but the path will be well worth taking.

Outside of my academics, I have been very active in VHL awareness. When my mother and sister were first diagnosed, telling people about the disease helped me to deal with what was happening to my family. I also thought I could change the horrors that had already occurred; if all eye doctors knew to dilate a baby's eyes to check for signs of disease (eye disease can be indicative of all sorts of problems, not just VHL), perhaps Shelby's eye tumors could have been treated before they ever caused such a huge problem! I realized how powerful a tool awareness was; increased knowledge would create better care and maybe even help find a cure! Advocacy could prevent

people from being turned away from their insurance because they have a genetic disease! Awareness would help people find others dealing with the same problem, and make the experience that much easier to swallow! During high school, I sold VHL awareness bracelets in school, and I maintained an online website to raise funds for the VHLFA., which supports the VHL cancer research fund.

When I started college, I was introduced to the world of social networking, via Facebook. Through online social networking, I could interact with many people, from fellow classmates to friends I had not spoken to in years! Facebook has a Groups feature, through which people of a common interest, like TV show fans or members of a school sports team, can come together and share information. Inspired by this idea, I created a VHL awareness group called "Spreading Awareness of Von Hippel-Lindau disease (VHL)." I invited all of my friends to join, and I asked them to invite their friends to join, too. To date, the group has 987 people, including people who have VHL, family members and friends of VHL patients, and people who just want to spread the word.

I update the group's page with relevant news articles, and I send messages to its members with awareness opportunities. I also have listed relevant websites for learning more about VHL and where to go for clinical care and genetic testing. I am so excited that there are 1331 members! I never anticipated the group getting this large. There is a corresponding group on the Myspace website.

These groups offer the VHL community, including its advocates, an opportunity to network with each other and spread the word about the disease. Even if an Internet user sees the group on a friend's profile, takes a look at it, but decides not to join, that user is one more person who has heard of the disease. With each extra person who hears of VHL, visibility is further increased, and with increased visibility, a cure comes even closer.

My goal is to get the entire country to know what VHL is, not just the people who have it and their family, friends, and medical support team. It is not easy to get people to care about something or donate to a cause that they have never heard about, especially if it does not readily affect them. Plus, it tears at my heart to know that because so few doctors fully understand VHL, there are many VHL patients who cannot receive excellent care.

My mother and sister go to NIH next week for their next round of testing. They are lucky to be under the care of very cognizant doctors; I wish everyone was so lucky. We all hope for the best, and whatever happens, we realize how fortunate we are to have each other.

Alex's Story

by Alex A., New Jersey

I am Alex Anderson, and I was unusually young when I was diagnosed at age four, with von Hippel-Lindau disease. I'm twelve now. Here's my story:

At age four I was lying on my left side on my mom's bed. She said to me ... well ... I'm not sure what she said, and never will be. That's because I couldn't hear from my right ear. When my mom asked me why I couldn't hear, I said, very matter-of-factly, "This is the ear I hear out of," pointing to my left ear.

My mom realized that something was wrong. She took me to our pediatrician, who said to go to Childrens Hospital of Philadelphia. After a biopsy, we were informed that I had an endolymphatic sac tumor (ELST). That is how I was diagnosed with VHL. I had the ELST removed the following week, in a surgery that took 14 hours. Since then, I have devoted much of my time to the epic battle against VHL disease, trying to raise money and awareness to help find a cure.

My life is a bit different from that of other kids. I am constantly monitored, to make sure nothing is wrong. My mother researches all the literature on diet for cancer prevention, and I do my best to follow those guidelines. My friends and family are all so great, though! They help me with fundraisers, and support me prior to operations! I've had four eye laser surgeries to eliminate about 24 retinal hemangioblastomas. I don't know where I'd be without the support of my friends and family at home, and my "other family," the VHL Family Alliance, a network of VHL families all around the world.

And now, with my fundraiser, Alex Jay VHL Day, in progress, they are better than ever! One of my main goals for AJVHLD is to get kids involved in fundraising. I have often felt that kids feel powerless in today's society. Giving them an opportunity to participate in helping to cure cancer also gives them a feeling of significance. When I'm fundraising, I always feel more significant in the world, like I really matter. I have been lucky to be able to pull together a fantastic team of middle-schoolers to help me with the fundraising stands in my small community on May 9th. Each student is excited to help in any way they can, and the teachers have been so supportive! We all feel like we can make a difference.

Another one of my visions for the big day would be to have all participants in the VHL Family Alliance USA do the same fundraiser from coast to coast. Other countries have other cultures and may have other fundraising ideas which will be just as

fantastic, but since we will all be working together, I know it will be a huge success. That's my mantra. "Working together is the one true path..."

It will be a truly inspiring day, when the world can unite for a cause, despite wars and disagreements. We are all on this Earth, so we are all family. And family helps family. We can be an incredible force when united.

Bob, Karen,
Alex, and
Amy



***Help us make Alex's and Ariel's Vision a Reality
Join in one of these events,
or help to create another event near you!***

Raise Awareness ...

Raise Money for Research ...

Find a Cure for VHL ...

May is VHL Awareness Month

International VHL Day, May 9, 2009

See the complete list of activities at <http://vhl.org/announce/09aware> or <http://vhl.org/meetings>
The following listing is current as of April 10, but is expected to grow...

Missed the event? You can still be part of it by donating. Want to help? Call Anita at 800-767-4845, ext 4

California

Manteca, California (an hour from Sacramento or San Francisco), event being planned
contact Elyse, vday09-cl@vhl.org
VHL Annual Meeting, Anaheim, CA
June 27, 2009, see page 11 or vhl.org/meetings

Florida

VHL Pancake Breakfast, May 9, Seffner, FL
8-11 AM, 1310 Kingsway Rd, Seffner FL
(near Tampa), contact pancake@vhl.org

Indiana, being planned

contact Michelle, vday09iIN@vhl.org

Maryland, crabfest being planned,

contact Kelley, us-md@vhl.org

Massachusetts, contact MaryLou, us-ma@vhl.org

State House Proclamation, May 11
Celebration on Boston Common, May 23, 11-3
featuring Keith Richards, Jennifer Behnke
other performers. contact anita@vhl.org
See also Rhode Island

New Jersey,

Jennifer Behnke concert, March 21, Hoboken NJ
"Reflections, Renewal, Rebirth"
see also Massachusetts May 23
Cheeburger Cheeburger, May 4, Malapan NJ
(30 miles from Trenton)
contact Ariel, alr2143@columbia.edu
Dine and Donate for VHL, May 9, Margate NJ
(5 miles from Atlantic City) bodydr@aol.com
contact Karen, bodydr@aol.com

Rhode Island,

One-act play about VHL
"The Thing That Ate My Brain ... Almost"
Perishable Theater, May 9, with refreshments
and dialogue with the author, Amy Lynn Budd
For tickets call +1-401-331-2695
or vhl.org/meetings to buy online

Washington State

Kim Hall competes in triathlons
May 30 and
August 16 Danskin SheRox Triathlon in Seattle
see <http://www.firstgiving.com/kimbrehall>
or contact kimbrehall@comcast.net

Canada, contact info@LTgraphics.ca

Bowl-a-thon, April 11, Windsor, Ontario
Sue Lamb, phone +1-519-735-0236

England,

VHL Contact Group annual meeting
Manchester, England, May 9
see vhl.org/meetings or contact uk@vhl.org
Mary Weetman, phone +44 (1204)886112

Germany

Regional meeting in Neufahrn, north of Munich
May 9, see vhl.org/meetings
Gerhard Alsmeier, phone +49-5931-929552
g.alsmeier@hippel-lindau.de

India,

Introduction to VHL for Doctors
Mumbai, India, by Dr. Shutish Patel

We Need Your Vote! - See Ballot, p. 15

The Nominating Committee submits this slate of candidates for your approval.

Your vote counts! To be confirmed, each candidate needs the votes of 10% of the members.

❑ Michelle Cieslak, California

Michelle Cieslak first became aware of Von Hippel-Lindau in 2004, when her 12-year old son reported a problem with his eyesight. After successful retinal angioma surgery, he was screened and subsequently diagnosed with VHL. With no family history of VHL, and knowing nothing of the disease, she became acutely interested in learning more, as much as she could about VHL and spontaneous mutations in particular. Michelle is tenacious and proactive and has always looked to the VHL Family Alliance as an incredible resource.

Michelle is an accountant with over 25 years of work experience in various industries. She and her husband enjoy listening to their son (now 17) play music and watching their daughter (14) play soccer. Over the years Michelle has enjoyed working with her local kid's schools and within her church to organize and lead different group activities as well as assist in financial matters. Michelle is looking forward to working alongside others within the Alliance to help find a permanent cure for VHL.



❑ Altheada Laverne Johnson, New York

Altheada Johnson is a retired Registered Dietitian. She has previously served twelve years on the VHLFA Board of Directors. She is the Chairman of the Hotline Committee, overseeing the volunteers who respond to Hotline calls. She has represented VHLFA as a speaker at a number of meetings, including the first meeting of VHLFA South Africa in March 2008.

She focuses in particular on Outreach to the medically underserved. She is a tireless letter writer, and has appeared on agendas at meetings on Minorities and Cancer (D.C.); Women and Genetics (Illinois), Minorities and Cancer (Alabama). Altheada is our in-house consultant on nutrition, and heads an informal alternative medicine interest group. She is in touch with doctors and researchers on the topics of pain management and pre-implantation testing.



❑ Camron King, California, candidate for re-election

Camron King is Program Director for the California Association of Winegrape Growers, a non-profit trade association. He tracks state legislative and regulatory actions related to environmental and natural resource issues.

His experiences include staffing non-profit boards and committees, fundraising for non-profit organizations and management and oversight of paid and volunteers staffs. He serves on a number of community boards and committees. He has a Master's in Public Policy and Administration. He has served on the VHLFA Board for three years, and is currently Chairman of the Board of the VHL Family Alliance.

Diagnosed with von Hippel-Lindau (VHL) in 1996, he has endured a number of surgeries to remove tumors from his eyes, both adrenal glands, kidney, liver and spinal cord. This experience has moved him to become more involved in advocating for greater cancer research funding, accessibility and a desire to educate the community about VHL. Camron and his wife Alicia have two children, Brayden and Addison.





□ **Robert T. Kramer, D.M.D., Pennsylvania and Arizona**

Robert Kramer is a dentist and dental educator. His dental practice was voted "Simply the best dental practice" in 2003.

"I currently coach boys travel soccer, and enjoy biking, fishing and hiking. I completed the Steamtown Marathon in 2000 and enjoy competitive sports. My wife and I have restored our current residence, Springfield Farm, to its original 18th century splendor over the past 15 years. This has spurred my interest in both historic preservation and early American antiques with an emphasis on Pennsylvania German folk items.

Our family enjoys a very active lifestyle. My wife is a triathlete, competitive marathon runner and avid horse person. She has completed an ultra marathon (50 miles) and currently holds the state record for it in her age group. She operates a personal training business. Gunnar, our 8 year old son, plays tennis, soccer and currently holds his high red belt in Hapkido.



□ **Thomas D. Rodenberg, Esq., Missouri**

Thomas D. Rodenberg is an attorney who currently serves as General Counsel and Director of Legal Services for the Blue Springs R-IV School District. The District consists of 14,000 students and 3,000 full and part time staff.

Tom previously served on the VHL Family Alliance Board of Directors for nine years, two of those years serving as Chairman of the Board. Tom has a particular expertise in insurance issues as they relate to VHL and genetic conditions and has made numerous presentations on this subject. Tom and his wife, Katherine, have four boys ages 20, 18, 16, and 15. They live in Blue Springs, Missouri.

Please renew your membership today, and Vote!

In order to certify the election, we need the votes of 10% of the membership.

This is YOUR Alliance. We exist to serve you and your family. Please participate by paying your annual dues, and making sure we hear your voice.

We have held the Annual Dues to \$25 since 1993. We provide information to everyone, whether or not they are able to pay. But if everyone paid at least this modest dues amount, we would more than meet our goals, and we would have more money for research.

Please make a contribution -- whatever amount is right for your budget is fine with us. But your participation is important to us. Please let us know that we are being helpful to you, but sending in whatever donation you can. Coupon on page 15.

Thank you!

Green Membership!

VHLFA introduces the Green Membership, a new way for you to make a long-term impact on VHL and the environment. Green members receive benefits and communications electronically, which helps shrink your carbon footprint, plus an automatic annual renewal feature means less paper in your mailbox.

Green Memberships start at just \$25 annually and your automatically renewed contribution will help sustain the programs of the VHL Family Alliance. Sign up on page 15.

Thank you for supporting VHLFA while doing your part for the earth!

***Join us in Anaheim!
Annual Meeting, June 27, 2009
For more information, see page 16***

Our thanks for donations from:



New!

***VHL Handbook,
Children's Edition!***

Coming this summer!

Gayun Chan-Smutko, M.S. and Melissa Kruger and their wonderful committee of parents and professionals has assembled a book on VHL for children, to help them understand what's happening and learn to take responsibility for managing their health.

***Full color,
Photos of real families
with VHL.
Illustrations by real kids
with VHL***

***List price \$12.99
Member price: \$6.50
will be available on
Amazon.com***

The Thank-You List appears only in the print edition



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It is springtime, time for renewal.

It is less expensive for us to make all annual subscriptions renewable in May.

Membership & Orders

If you have already paid your dues in 2009, thank you!

Name: _____

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Ballot: *vote for five*

☐ I vote for all five candidates as proposed by the Nominating Committee
or you may vote individually if you prefer:

☐ Michelle Cieslak ☐ Altheada Johnson ☐ Camron King ☐ Robert Kramer ☐ Tom Rodenberg

Preferences:

☐ I would like to receive occasional alerts via e-mail ☐ I want to Go Green!

Please Renew My Membership

Annual dues: \$25 per mailing address (\$35 outside U.S.* credit card preferred for international payments) \$ _____

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

☐ Please charge \$ _____ to my card monthly for _____ months

☐ Please renew my membership annually until I change it

☐ Children's book full color, illustrated - member price \$6.50, non-member price \$12.99 \$ _____

☐ Go Green! Please send all materials by e-mail to reduce costs and save the earth too!

Join us in Anaheim, California, for the 16th Annual VHLFA meeting! June 27, 2009

Please register _____ total people for Anaheim (includes Saturday meetings, lunch and snacks)

First person @ \$100 before May 15 (after May 15, price goes up to \$130) \$ _____

Others in your party @ \$75 before May 15 (after May 15, price rises to \$130 per person) \$ _____

You will need to book your own room with the Hyatt Regency Orange County at +1-714-750-1234
or <http://orangecounty.hyatt.com> - special rate for the VHL meeting, \$139 for a standard room
Be sure to book before May 15 to obtain this special rate.

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

Special gifts total \$ _____

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: _____

Total Amount Enclosed \$ _____

☐ My check is enclosed ☐ Please charge my Visa or Master Card, Amex or Discover

Name as it appears on the card: _____

Card number: _____ Expiration: _____

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Thank you!

Please vote!



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Join us in Anaheim!

VHLFA Annual Meeting 2009, Anaheim, California

Saturday, June 27, 2009, 8 AM to 5 PM, Hyatt Regency Orange County

The 16th Annual Meeting of the VHL Family Alliance will be held at the Hyatt Regency Orange County in Anaheim, California, near Disneyland and other theme parks.

The meeting will be held Saturday, June 27, 2009, from 8 AM to 5 PM. Discount registration prices are in effect until May 15, and hotel room rates will be lowest before May 15.

Remember - *Prices go up after May 15!* Reserve early to get the best prices!

Registration includes a day of fascinating meeting, lunch, and snacks throughout the day. This is information you can't get any other way, and fellowship with other families with VHL.

Between now and May 15 we are offering the first registration for \$100, and a 25% discount for additional members of your party. After May 15 all registrations will be \$139 each.

Hotel

Make your own hotel reservations by May 15 to get our special conference rate of \$139 for a standard room, for up to four adults in the room. Call +1-714-750-1234 or go to <http://orangecounty.hyatt.com>

Hyatt Regency Orange County
11999 Harbor Blvd.,
Garden Grove, California, USA 92840

Tel: +1 714 750 1234 Fax: +1 714 740 0465

Worldwide events
See page 9

Convention attendees can receive great discounts to Disney Theme Parks. Just show your convention badges to the Disney Parks Office in the Hyatt! 1 day 1 park pass \$65, 1 day park hopper pass \$90, Twilight pass from 4pm until close \$48, 2-day pass \$143

Agenda

The agenda will include presentations on

Advances in treatment of eye lesions

Advances in diagnosis and treatment of pheochromocytoma

Latest news on nutrition and cancer prevention, including a trial you can help with, to evaluate a new food product that may inhibit tumor growth!

News of a new clinical trial -- more specific, fewer side effects

Announcement of two new clinical care centers -- in California and Florida

Register Now on page 15