VHL FAMILY FORUM

Family, Friends, Physicians, & Researchers dedicated to improving diagnosis, treatment, and quality of life for people affected by von Hippel-Lindau.

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Learning about Pheochromocytomas

The First International Symposium on Pheochromocytoma was held in Bethesda, Maryland, October 20-23, 2005. It is hard to believe that in 2005, this is the first international symposium ever held on this topic. [See Note 1] Drs. Graeme Eisenhofer and Karel Pacak of the U.S. National Institutes of Health co-chaired the meeting, bringing together 180 participants from all over the world. Forty patients and family members came from as far away as Texas, Michigan, and Oregon to be part of the experience.

Dr. William Manger of New York University opened the conference with this sobering thought: "Tragically up to 50% of pheochromocytomas are still discovered at autopsy, mainly because the diagnosis of this neuroendocrine tumor was not considered." Families that know they may be at risk of a pheo should know the symptoms of a pheo so that they can help lead their doctors to this diagnosis.

"Pheochromocytoma is still the most treacherous, deceptive tumor on the planet," said Dr. Manger. "Missing the diagnosis almost invariably results in devastating cardiovascular complications or death. Clinicians must always think of pheochromocytoma whenever evaluating a patient with sustained or episodic hypertension or any manifestations suggesting elevated catecholamines. Very rarely, familial pheochromocytoma may cause no hypertension, symptoms or signs. But biochemical testing can always establish the presence or absence of a pheochromocytoma, and localization with MRI, CT, or 131 or 123 I-MIBG is almost always possible."[2]

"The very large variety of symptoms and signs encountered in patients with pheochromocytoma frequently suggests a number of other conditions and may mislead and confuse the physician and cause an erroneous diagnosis. This treacherous tumor has correctly earned the title of the "Great Masquerader." Above all, it is essential when confronted with any manifestation of these conditions that the clinician be forever alert and *think* of pheochromocytoma!" It is also important for a family with VHL to suggest to their physicians that a pheo should be considered,

Dr. William Manger of New York University and the National Hypertension Association, giving the keynote address. Photo by Debra Harlander

and be prepared to give literature to their doctors to convey the best possible information – better than they remember from medical school. Alison's story in the following article is a good demonstration of how a mother saved her children by pressing for the best answers and the best treatment.

In order to raise the rate of diagnosis, the conference focused on:

- Understanding the many ways pheos may present, to raise consciousness of these symptoms
- Agreeing on diagnostic criteria, especially agreeing on the tests needed to diagnose a pheo

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- Agreeing on which biochemical tests are the most specific for pheo, and the most sensitive in finding even smaller tumors
- Agreeing on which imaging studies are the most effective in finding extra-adrenal pheos
- Agreeing on when patients should be screened for one or more of the genetic alterations that can result in a pheo.

Familial pheochromocytomas may occur in people with any one of six known genetic syndromes:

- Von Hippel-Lindau (VHL)
- Multiple Endocrine Neoplasia (MEN, especially type 1)
- Neurofibromatosis Type 1 (NF1)
- A newly recognized panel of genes referred to as SDHB, SDHC, and SDHD which cause head and neck paragangliomas

And at least 70% of pheos occur at random in the general population.

Pheochromocytomas (called "pheos" for short) were first described by Frankel in 1886. Dr. Von Euler won the 1946 Nobel prize for proving that norepinephrine (NE) was a neurotransmitter. But diagnosing and localizing or finding pheo within the body has been extremely difficult until recent advances in imaging and chemical diagnosis.

Pheos arise from chromaffin tissue in the sympathetic nervous system. 85% of pheos occur in the adrenal glands, but 15% occur outside the adrenals. These "extra-adrenal" pheos are called paragangliomas. For simplicity, the term "pheo" is here used to include these neuroendocrine tumors, wherever they occur in the body.

The genetic factors involved in pheochromocytoma/paraganglioma (Pheo/PGL) syndromes represent a perfect example for these exciting advances in modern biomedicine (4-7). Until recently, more than 90% of these rare but clinically important catecholamine-producing tumors were thought to occur as sporadic nonhereditary entities that should not need any genetic screening unless there was a family history of a certain form of multiple endocrine neoplasia.

Debra Harlander, Pheochromocytoma Support

The last three years have seen mounting evidence that a substantial proportion of patients with these tumors have a definable genetic defect with potentially important clinical implications.

They are usually described as exhibiting a classic triad of symptoms: headache, palpitations, and sweating. But not everyone exhibits all three symptoms. Dr. Debbie Cohen of the University of Pennsylvania and others shared statistics for their patients that showed that approximately 60% of patients complain of headaches, about 50% have palpitations or panic attacks, and about 35% have sweating. Some have all three, but more often people exhibit one or two of these symptoms. And some have none of these three symptoms at all. Nearly 50% have normal blood pressure.

Pheos in pregnancy are often misdiagnosed as pre-eclampsia. Failure to recognize a pheo in pregnancy carries a high mortality rate for both the mother and the fetus. As long as the doctor knows about the pheo, especially early in the pregnancy, precautions can be taken to protect the mother and the child.

In studies of "incidental" findings of pheos (found while looking for something else), the pheos found in this way are generally large -- 5-11 cm (2-4.5 inches in diameter). Such a large tumor has likely been growing for a number of years, and has probably been causing symptoms for some years.

Dr. Marta Barontini from Argentina shared her study spanning her 40-year career in pediatric medicine. She finds that pheos in children have more severe and more sustained symptoms than in adults. It is especially important to do genetic studies in children with pheos, as a higher percentage of them have a genetic syndrome. She has stayed in touch with all her patients, and has recalled them for genetic screening as additional genetic information has become available.

Dr. Henri Timmers of the Netherlands did a review of 141 papers in the medical literature on pheos in pregnancy, reporting a total of 174 cases. Only 7% of these had previously been treated for pheo, and only 17% of them had any history of high blood pressure. The initial diagnosis was wrong in 31% of these cases. The risks to the mother were highest in cases of incorrect diagnosis. The outcome for both mother and child were largely dependent upon timely recognition of the problem, and appropriate treatment. Things tended to go best when the pheo was removed earlier than 24 weeks

of pregnancy, or if the pregnancy was carefully managed to term and the pheo was removed after delivery. It is critically important to inform the obstetrician if the patient might be at risk of a pheo so that the mother and child can be diagnosed early and treated carefully.

How is it that six different genes can all cause a pheo? Dr. William Kaelin presented the work he published in Cancer Cell. Mutations in VHL, RET, NF1, SDHB, SDHC, and SDHD can give rise to pheochromocytoma/paraganglioma. These different genetic lesions may all act by decreasing the activity of a 2-oxoglutarate-dependent oxygenase, SM-20/EglN3/PHD3, resulting in reduced apoptosis of neural crest cells during development." [3] There are a number of checks and balances in the cell that verify that cells are proceeding properly. If they are badly formed, the body's normal protective mechanisms kill off the badly formed cell and make a new one. This is a process called apoptosis. In the case of pheos, cells that would normally have been killed off go on to become tumors. These genes interfere with the normal process of apoptosis and allow the cell to become a pheo or paraganglioma.

The most important messages for people with familial syndromes that may cause pheos is to keep strong, to monitor for early detection, and do what you have to do to get appropriate treatment. Read, learn, and help to bring your doctors up to speed on pheos.

Remember that you can't protect yourself if you don't know you are at risk. Make it your business to find out whether you may be at risk, and take responsibility for your own health and that of your children.

Since knowledge about pheochromocytoma/paraganglioma syndromes is so scarce among the vast majority of physicians and health practitioners, the conference participants asked that recommendations and updated treatment guidelines should be posted on the PRESSOR website (www.pressor.org) along with a list of centers with a satisfactory level of experience in treating pheochromocytoma. This information will be developed over the coming year. The formal Proceedings of this conference will be published in the *Annals of the New York Academy of Sciences*, forecasted for October 2006.

Notes: Our thanks to Prof Stefan R. Bornstein, University of Dresden, Germany, Debra Harlander, and Jo Ann Monroe of the Pheochromocytoma Support Board for their assistance in the preparation of this report.

1. An earlier meeting broke ground for this effort atCold Spring Harbor NY in November 2003. For the results of that conference, see *Endocrine-Related Cancer* (2004) 11: 423-436.

2. Finding a pheo or paraganglioma is best done with CT or MRI, or an MIBG. An MIBG test is a nuclear medicine procedure using a radioactive isotope or tracer, which is absorbed by pheochromocytoma tissue. Meta-Iodo-Benzyl-Guanidine (MIBG) is injected into the patient before the scan is performed, making the pheo stand out clearly on the diagnostic pictures. There are two radioisotopes used for this purpose, 131-MIBG or 123-MIBG. 123-MIBG is far superior, but has a shorter half-life and must be used within hours of being made up. Nonetheless it is becoming more available because of its greater ability to seek out pheos and paragangliomas wherever they are hiding in the body.

3. S. Lee, W. Kaelin, et al., Neuronal apoptosis linked to EglN3 prolyl hydroxylase and familial pheochromocytoma genes: developmental culling and cancer. *Cancer Cell.* 2005 Aug;8(2):155-67. The quote is from the commentary in the same issue by Dr. Patrick Maxwell, A common pathway for genetic events leading to pheochromocytoma. Cancer Cell. 2005 Aug;8(2):91-3.

Dr. Hartmut Neumann of Germany played the violin at dinner, a tradition at VHL meetings.

Yer Comin', Eh?

Meetings in 2006

Salt Lake City, Annual Meeting
June 24, 2006
see page 6

London, Ontario, Canada
Medical Symposium on VHL
October 26-28, 2006
see page 16

Pheos and Children

By Alison E., Texas

My husband Troy and I recently attended the First International Pheochromocytoma Symposium held in Bethesda, Maryland. It was exciting to be among all of the top pheo experts in the world but also a little daunting. I found myself in absolute awe of these wonderful doctors and researchers. My husband works in the music business, and we are around celebrities all of the time. However these masters of science had rock star status in my eyes! I, too, have done quite a bit of research, so I was familiar with the current protocol on how to diagnose and treat pheos. It was a privilege to have the opportunity to discuss and share with these dedicated men and women

I wanted to share my personal experiences with pheos from a Mother's perspective. I have two children that have experienced the wrath of pheos.

My husband Troy was diagnosed with VHL as a teenager (we have been together since we were teenagers). Once the location of the gene was discovered, we had our daughter Chloe tested at the age of 4. We started the screening process right away wuth Dr. Sharon Plon, a geneticist at Texas Children's hospital.

Chloe was six when her urine catecholamines started climbing. Her primary symptom was sweating, especially at night. She also looked very tired most of the time. She had headaches as well, but didn't complain about them too much. I remember visiting her second grade class and thinking that she just looked so tired and pale compared to all the other kids. That was kind of an eye-opening experience because I had gotten so used to seeing her look that way. My husband had had pheos as a child, and we knew from DNA testing that Chloe carried the VHL alteration, so we suspected a pheo.

With a positive DNA test and family history of pheos at a young age - what else do you need to start imaging? Yet during this time period, she was seen by the chief of pediatric endocrinology at a major hospital. He told us that there was "no way" that she had a pheo at that time despite her urine testing results, because she did not have *sustained* hyperte nsion. He said that children with pheos always have sustained hypertension. While it is true some kids will have sustained high blood pressure, there are still children that will not. There are a number of myths about pheos, and this is one of them. At the conference we learned that 50% of people with hereditary pheos have normal blood pressure.

A year later her urine catecholamines increased even more, and that is when they realized this indeed could be a pheo. It was during this time that I contacted Dr. Graeme Eisenhofer at the NIH. He told me about a new test that he and others had developed called the "Plasma Free Metanephrine" test. This test is around 97% accurate compared to the 24 hour urine test which is only about 80% accurate. [1]

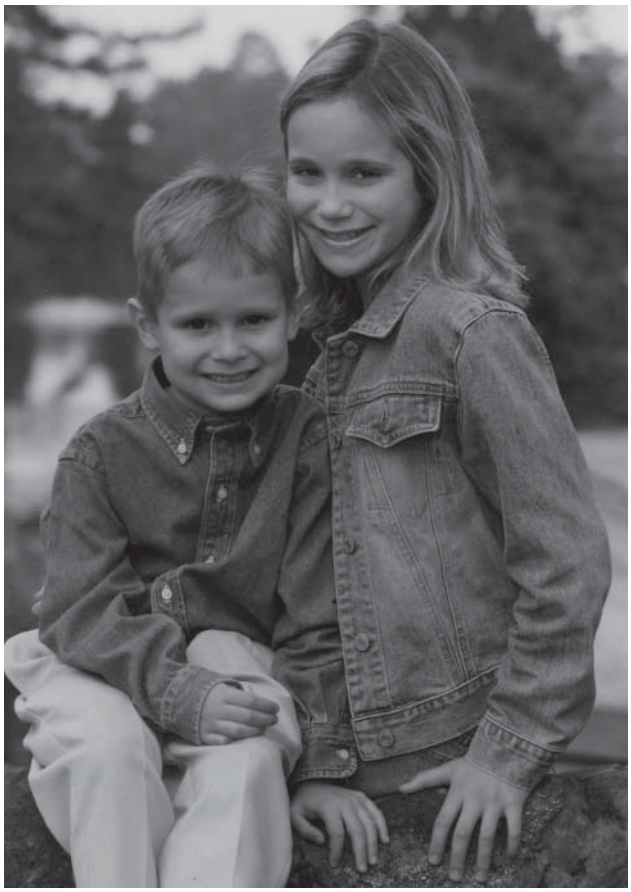
While Chloe was getting an IV for her MRI sedation, her blood pressure went through the roof. She is very dramatic and would rather be dismembered than have a needle coming at her. It was a high stress situation as it always is with Chloe and needles. Nurses ignored the dangerous high BP readings, claiming that "there must be something wrong with the cuffs." I tried to tell them that these tumors cause high blood pressure, but my words fell on deaf ears. I was mild and soft spoken back then and believed the doctors and nurses had to know more than me. How could I possibly know more about these tumors than doctors?

After Chloe was sedated, blood was taken and sent off to the NIH for the Plasma Free Metanephrine test. Dr. Eisenhofer called me and said she had the highest numbers ever seen with this test. Because of all the stress with the IV, it was thought that she had an "episode" while in the MRI chamber. When they rolled her out of the machine, she was drenched with sweat. It took a lot of medicine to sedate her because of all the excess stress hormones in her system. After we got her home, she would not wake up from being sedated at the hospital. We called her doctor who said to bring her into the ER [emergency room]. She finally began waking up very late that night.

Surgery was scheduled, and Chloe was placed on an alpha-blockade. This drug has to be ramped up slowly over 1-2 weeks. It has the potential to turn the patient into a "noodle-like" state which requires hospitalization and close observation. Chloe reacted strongly and was hospitalized after several days on the drug. The doctor had written orders on Chloe's chart that she was not to be "poked" at all prior to surgery because of her hypertension and extreme anxiety response with needles.

However, the night before surgery, because it is standard to draw the necessary labs, our pleas were ignored and blood was taken. (Today, I would ask the doctors to give me a written copy of their orders, so that I would be able to show the orders to the medical staff if another similar situation arose.)

Her surgeon was able to remove her three centimeter pheo laproscopically, and she recovered fully. Again, our excellent anesthesiologist, Dr. Nancy Glass, had to give her a generous amount of anesthesia because of the excessive catecholamines circulating in her system. The doctors could not believe that she was still wide awake and alert after all of the drugs they



Caleb and Chloe

administered. They all clamored around her not believing what they were seeing. Chloe finally went to sleep, but only after a lot of anesthesia.

Thankfully she came through very well. She is thirteen now and very active in competitive cheerleading and theater. She is my expert in making PowerPoint presentations. Currently, she has three eye tumors that are being watched closely.

Two years after my daughter's surgery, our son, Caleb, who is now eight, developed three paragangliomas (extra-adrenal pheos) on his aorta at the age of four. He is one of the youngest known cases world-wide. His biochemical testing revealed that his levels were two and a half times the upper limit of normal.

Since he was so young, we didn't notice a lot of symptoms. When he was only two, he complained of headaches. We took him to a neurologist and told him of his DNA diagnosis of VHL with a predominance toward pheos, but he didn't think we had anything to worry about. Caleb also had some night sweats, but that was pretty much it.

An MRI revealed two pheos. A follow-up MIBG confirmed it, and surgery was scheduled. Our nephrologist, Dr. Daniel Feig, admitted Caleb to the renal floor of the hospital where we were told that he needed to be on a high salt diet because of the alpha blocker -- phenoxybenzamine -- he was taking. But, because most renal patients are on a low salt diet, the nursing staff would not listen to us. We had to contact Dr. Feig so he could inform the nurses that this was correct and not a typo. Again, this wasn't the norm, and it required us to be on our toes to make sure that the staff understood the reasoning behind every decision.

An "open" surgery was deemed best because of the location of his pheos. While they were in surgery, our surgeon found another pheo close to the other two that did not show up on any of the previous imaging. Needless to say, his surgery was a delicate one, and ended up taking eight hours. All was well until the day after surgery; he had a code-blue because of a problem with his epidural line. The epidural was threaded high up to ease the pain of his rather large abdominal incision and there was some excess epidural anesthesia in the line that affected his respiratory rate. Caleb turned completely blue as his oxygen saturation dropped to 9% (normal being 95 to 100%.) So far I had handled everything else fine until this happened. I still have nightmares about it. It's something I don't think I will ever get over.

Despite my knowledge of pheochromocytomas and the Mama Bear instinct to protect her young, this experience has made me realize I cannot control everything. That truth has been very difficult for me to accept. My personal way of dealing with the stress of this disease is to educate myself as much as possible. I want to know it all, the good, the bad, and the ugly. I want to know numbers, statistics, ratios -- you name it. Because pheos are rare and even more so in children, you have to become an expert.

Not only do I search out the absolute best doctors locally, but we also have a team at the National Institutes of Health (NIH). After attending the pheo symposium [see page 1] there are a few more doctors I'd like to add to my team -- so now we can have a local, national and international team! My husband laughs at this notion, but he knows I am dead serious. With such a rare disorder, why not seek out the experience of as many experts as you can? They are few, and you have to seek them out.

The biggest blessing we have had during this whole journey is the knowledge of our DNA testing results. Had we not elected to have our kids tested at a very young age, our victorious outcome might have been different. We know that 50% of all pheos are discovered at autopsy. And, yes, I truly feel victorious, because when and if another pheo appears, we will conquer it will the same faith and determination that has gotten our family through it before. Ultimately, I believe my children are in God's hands and, in the meantime, He has blessed me by

surrounding my family with excellent doctors and resources to learn more about VHL.

What about my children's outlook on their experiences? Both of them recently asked when they could go back and have more surgeries at Texas Children's hospital. They asked in a happy Disney-World like tone, without fear. An ever-protective Mother's heart smiles when she hears questions like that, even though my jaw fell to the ground. I think it is extremely important to remain positive, especially when you are in this type of situation, because your kids will instinctively adopt your outlook. This is not difficult for me, because it is truly how I feel.

My biggest hope is that by sharing our personal experience with pediatric pheochromocytomas, Troy and I may be able to shed light on a rather dark area of medicine and hopefully help other families going through a similar situation. I got most of my knowledge from the pheo support board [2] as well as correspondence to all of the known pheo experts. We learn so much from each other!

Notes:

1. The Fractionated Plasma Free Metanephrines is the best test for a pheo. This lab test is only performed in a small number of labs, preferably at the Mayo Clinic, where many hospitals send blood for processing.

2. See www.vhl.org/pheo for pointers to the pheo support board and other pheo resources.

Camron and Brayden have raised $1625 so far for VHL by creating a Personal Fundraising Page.

Thank you for your kindness and generosity!

I want to encourage you to support and organization that is very close to my heart - an organization that has been so helpful and supportive through my diagnosis and treatments - the von Hippel-Lindau Family Alliance.

When I was diagnosed with VHL in 1996, it came as a true shock. I was told that I had a genetic cancer that could impact so mydifferent parts of my body and would change my life as I knew it. To this day, I have had approximately 10 procedures and surgeries to remove tumors and repair damage from complications of those tumors in my right eye, adrenal glands, right kidney, liver, and spinal column.

I seek to support the research and treatment efforts of the VHL Family Alliance in hopes that no one else would ever have to endure what I have and continue to go through.

I ask you for your support of this worthwhile organization and I thank you for your kindness and

Personal Fundraising Pages

see www.justgiving.com/vhl

Maybe you can't run a Triathlon, but you can definitely help in your own way, among your own friends and community. Here's one idea.

You can create an online Personal Fundraising Page for any occasion:

- In memoriam of a loved one
- Personal occasions like weddings or anniversaries
- Any social event you'd like to make more meaningful
- Marathons, walkathons, bike rides, or any sporting event

You will have a web page that you can e-mail to your friends, asking their support. Credit card donations come to the VHL Family Alliance. If donors prefer, they can give by phone at 800-767-4845, or mail a check to the office.

Join us in Salt Lake City!

June 24, 2006

Featured Speakers include:

Dr. Russell Lonser from the NIH will speak about endolymphatic sac tumors

Tom Rath, author of the best-selling book "How Full is Your Bucket?" will talk with us about his philosophy and his experiences with VHL.

Utah Chapter co-chairs Susan Sorenson and Mary Lynn Pearson will be our hosts. Call 800-767-4845, or see vhl.org/meetings

A Patient's Perspective

by Altheada LaVerne Johnson, RD, Brooklyn, New York

Altheada LaVerne Johnson

When I was finally diagnosed with VHL [in 1988], I had already had an EMG [electromyogram], 8 weeks of chiropractic treatments, countless CT's, MRI's, a myelogram, [1] an angiogram, and extensive spinal cord surgery. A hemangioblastoma and the associated cyst (a jelly-like substance causing a "sausage" in my spinal column, according to the neurosurgeon [2]) had been surgically removed and sent to pathology.

The doctor told me, "Ms. Johnson, you have a genetic disease called von Hippel-Lindau. You will need to have your eyes and kidneys looked at as well." All the time I'm wondering what he is talking about. Where could this have come from? While I felt devastated by the news then, I have since realized how very lucky I was: My surgeon knew enough about VHL to look beyond my spinal cord. I had a retinal examination, and a small angioma was found and treated. Eventually I had a craniotomy to remove a hemangioblastoma from my cerebellum.

Family tree

So many people with VHL go for decades without knowing what is wrong with them. I have talked with many who have had a number of different manifestations of VHL throughout their lives, never knowing that all these things were related to one disease.

My grandfather suffered severe headaches. One day he came home from work, lay down, and never got up again. That was about 1939. My father had several brain surgeries in the early 1960's. My siblings and I watched him taken away by ambulance in 1966 for a surgery from which he never recovered. Three years later, my 19-year-old brother died in the hospital after his second brain surgery.

Three generations, undiagnosed. At the time of my brother's death in 1969, doctors told my mother there was a tumor condition in our family, found only in males. We now know that it affects females equally. Not knowing anything about genetics, my three sisters and I felt there was no need to worry.

It was nearly 20 years before there were any other VHL manifestations. During my sister KJ's first pregnancy, she had vision problems. Her doctor told her she had a bleed as a result of high blood pressure. The only treatment she received was blood pressure medication. I know now that it is unusual for an ophthalmologist not to recognize VHL when he or she sees it. I have talked to many VHL patients, newly diagnosed by their eye doctor.

When my back pain started, it was so severe I had difficulty walking. When the VHL diagnosis came, we were finally able to make the pieces fit. We had an answer.

We have since learned that, among the sisters, only KJ and I have VHL. She has had two kidney surgeries and a craniotomy and is blind in her eye that did not receive any treatment. She has two children, both of whom have VHL. Her son Keith has had a craniotomy, but so far her daughter is symptom-free. I have had four spinal cord surgeries. My sister's first kidney surgery resulted in removal of her kidney. There is obviously lots more research needed to figure out just what role your environment, lifestyle, and actual genetic mutation play in which manifestations you will have. Right now, we just work on the assumption that if you have VHL, you are at risk for any or all possible manifestations.

A Way of Life

VHL is a familial cancer, an autosomal dominant genetic condition. A parent has a 50% chance of passing the VHL mutation on to his or her children. VHL is family-related in that it is passed on to you by one of your parents. But it is family-related in another sense: your family can pull you through those crises that are bound to pop up from time to time. My husband, mother, sisters, other family members, and friends were there for me. I could not have made it through without them. I spent about 3 months in the hospital with my first hospitalization. I had visitors every day. I had home-cooked food on a regular basis, and my room was filled with reminders of home. All this went extraordinarily far to ensure my survival.

Now, routine screening is a way of life for me: annual MRI's, CT scans and eye exams. It is so important to find the tumors early so that treatment, if necessary, is provided early. I believe that had I had early diagnosis and treatment, I would not be a

wheelchair user right now.

Because VHL is such an unusual disorder, patients have to be their own advocates. At the VHL Family Alliance, we encourage people to join a support/education group. Patients can offer each other invaluable information that can help them avoid repeating the same mistakes all over again. When I first learned I had VHL, I felt very alone. Now, I've met others with my condition. I know what to look and listen for. I'm active in the support group for VHL, and I have met many other health-care professionals who have the interest of VHL patients at heart.

While VHL is a serious disease, it does not mean your life is over. You learn how to deal with the bumps in the road VHL is to cause; you get over them and go on with your life until the next bump.

Notes: Altheada L. Johnson serves as New York Chapter Chair of the VHL Family Alliance, Chairman of the Hotline Committee, and Secretary of the Board. She can be reached at altheada@vhl.org. This article was published in *Community Oncology,* 2004 1(4):241 Reprinted with permission.

1. A myelogram is an x-ray test of the spinal cord with contrast. Before CT and MRI were widely available, this was the best test to look for areas of compression. Today MRI's are more often used for this purpose. Similarly, an angiogram maps the vessels of other tissues.

2. A cyst is a fluid-filled sac, like a balloon filled with fluid. When a cyst occurs in the spinal cord, it is constrained by the bony spinal column, so it spreads out long like a sausage within the spinal column, compressing the cord.

Acronym: ***Keeping All Rhymes True Original Obsolete Never***
Also Known As "***Just Sayin'***"

KARTOON is a lively, animated, fun-loving brother from Rosedale, Queens by way of Brooklyn. As an Emcee, the music he creates and performances he delivers are a direct reflection of his emotion and passion.

But don't get it twisted. It isn't all about fun and jokes. KARTOON is a brother with knowledge who knows who he is. He is a direct product and representative of The Golden Era of Hip Hop striving to create timeless contributions to music and global culture.

A man of achievement, KARTOON is a living example of the motto, "work = play." On stage, Kartoon is a free-spirited, wild, animated showman. Off stage, Keith Julian Richards is a focused, driven, business-minded individual who thrives on excellence and an incessant need to learn, create, and achieve. His music is a testament to the human spirit and experiences that everyone can relate to.

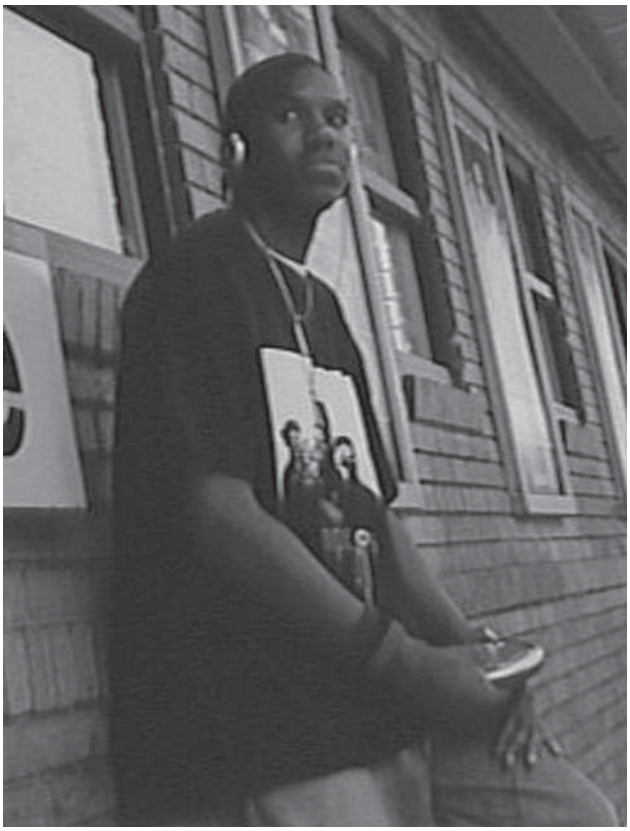
We asked him to share his experiences with VHL.

I first learned about Von Hippel-Lindau disease through my Aunt LaVerne's experience with the illness [See above]. When VHL began manifesting itself in her I was much younger. At this time I did not understand the ramifications that this disease could possibly have on my family. A few years later VHL hit closer to home when my mother [KJ] started to have manifestations. I remember praying many nights that my Mom would be ok. Even then I still did not know much about VHL.

In 1998 after I completed my sophomore year of college it really hit home. I remember it like it was yesterday. It was my 19th birthday and I started having dizzy feelings that evolved to constant fatigue and nausea. I tried to be strong for my family, especially for my mother because I know she worries about my sister and me even when we're feeling fine. But all of my symptoms just became worse and worse and it never dawned on me that I might have VHL until my sister said to me, "Mommy's crying because she thinks you might have VHL."

Things really came to a head at a family barbeque that we had at the house that summer. I usually play the music at our family functions but an hour or so into the party my father must have had a feeling because he came to me and asked if I was ok. I just shook my head 'no' and he walked me to the house. I spent the rest of the barbeque upstairs in my bed. The one thing I did not want to happen happened. I drew all sorts of attention to myself and the fact that I wasn't feeling well. And the last thing I wanted to do was spoil everybody's fun.

That night after all the guests had left, my Dad came to me and said "we're going to the emergency room." I was in the hospital for about a week where the doctors ran test after test and couldn't find anything. At this point I started to become very angry saying things like, "Isn't this a hospital?! Why the hell haven't they figured out what's wrong with me?!"



Keith Richards., Rosedale, New York
a.k.a. KARTOON or Just Sayin'

The doctors were about to release me and deal with me on an outpatient basis. But my mother spoke up and demanded they conduct an MRI of my brain and spinal cord. That's when the hemangioblastoma was found on my cerebellum. This is when I really became afraid because I immediately started thinking about the uncle and grandfather I never met who died of basically the exact same thing.

"...Never doubting the word of a physician.
But if I gotta choose I'll always go
with mother's intuition."

My Aunt Tena was the first family member I saw after I got the news. She helped me calm down and make sense of the situation and really gave me the feeling that I would be okay. Two days later I had my surgery, and about six days later I came back home. I decided to take that fall semester off from school to recuperate. In the Spring of 2001, through study, patience, and prayer, I received my Bachelor of Arts degree in Psychology from the University of Delaware. I'm currently working as a Case Planner for a non-profit agency in Jamaica, Queens, New York, while pursing a professional music career.

My mission and purpose in life is to use the power of music as a springboard into activism and spreading public awareness about VHL among many other happenings in my community, the African-American community, as well as the world community.

Through it all I knew that God would keep me and bring me through it as he did for my mother and my aunt. It's true my uncle and grandfather passed away, but I knew that God had a plan for me and they live through me. My Aunt LaVerne and my mother are my biggest inspirations when it comes to dealing with VHL. The two of them have endured much more hardship than I have at the hands of VHL, but they've never complained, so why should I?! We all have a cross to bear, and this happens to be mine.

My experience has definitely made me appreciate life more and every day I have with family and loved ones. My walk with God has become much more intimate also. I get regular checkups and urge everyone I know to do the same. I maintain a fresh haircut and my scar just adds a special uniqueness to my already good looks, if I do say so myself! So I'm still fly!!!

"...And I can't complain.
Life's been good to me thus far.
And I keep a fresh cut around my surgical scar."

Notes: "I'm still fly!" is slang for "I'm still fine, looking really good!" Look for a sample of Keith's CD on the vhl website.

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How Can You Help?
Pick a Project - Help Raise Visibility
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See www.vhl.org/help

A Gift of Love

Editor's Note: In 2004, a family in California lost a brother to a massive stroke. Even though he lay brain dead, they wanted to help him make the ultimate gift of life to others. They helped arrange the donation of all his organs, designating one of his kidneys for someone with VHL. We polled our membership through the online distribution lists, and within hours we had four candidates. Through the Designated Donor program of the United Network for Organ Sharing (UNOS), the kidney was sent to New York state, and was successfully transplanted into a gentleman with VHL who had spent five years on dialysis.

Living donors give the best chance for someone who needs a kidney, serving the recipient as much as 30 years. The following story tells of one woman's decision to be a living kidney donor for someone she had never met.

No Greater Love

by Bob Horning [1]

Two women lay waiting in adjacent operating rooms in St. John Hospital in Detroit on Dec. 4, 2002. Neither knew the other. They had nothing in common.

One was 55 years old, the other 43.

One is white and lives in Howell, Michigan, the other African-American and lives in Southfield.

One is Roman Catholic, the other Pentecostal.

For two years, the first woman – Suzanne Fales, a parishioner at St. Augustine in Deerfield Township – had been thinking, praying, preparing for the moment she would donate a kidney to someone who needed one. It felt good and right to her, and was something she wanted to do.

The second woman, Grace Drake, had had eight rough years prior to the surgery. "My daily life was subdued," she said. "I had lupus and hypertension; then my kidneys started failing. I had a decreased appetite; my diet and fluid intake were restricted because my kidneys couldn't filter correctly; I couldn't work because I was so drained from having dialysis treatments three times a week."

Chris, her husband, was struggling, too. Concern for Grace's health was taking its toll. He was constantly wondering if someone who was the right match would be willing to donate. He and Grace's brother had been tested but were not compatible donors. "It was trying my faith," he says. "I needed to learn to be a supporter, an encourager, a motivator."

Grace quoted Romans 8:28 to herself often: "For all things work together for good for those who love God and are called according to his purposes." Three weeks before Christmas 2002, that verse was fulfilled for Grace. In Howell, Suzanne was eager to go ahead, but wanted the go-ahead from her family. Looking back, Suzanne's husband, Tom, says the whole thing "hit me by surprise. I didn't even want to talk about it." However, Suzanne's unwavering attitude and a lot of reading about the transplant process broke down his reserve and, after a year, he became a supporter.

With Suzanne's mother, it just took some salesmanship to overcome her immediate "No." "But, Mother, you are the one who taught me about sacrifice," Suzanne said to her. "And you know that if you could do it, you would be the first one in line." Her mother was trapped, and after a silence, the only thing she said was, "Make sure you drink a lot of water."

Suzanne's daughter, Michelle, responded immediately, "Go for it."

That left Catelyn, Suzanne's granddaughter, who was 8 at the time. For Tom and Suzanne, she had to give the final OK. After the situation was explained to Catelyn, she agreed. "And if my brother or mother need a kidney some day, I will give them one of mine," she added.

"We received her response like a message from God sent through an angel, a selfless angel," Suzanne said.

Throughout the preparation time, Suzanne said fear and emotion were not the most difficult things to cope with. "It was all the tests – the EKGs, blood tests, psychiatric work-up. And along the way, the medical people kept giving me every opportunity to back out, like I didn't know what I was doing. I guess they wanted to be certain that I was serious." At one point, shortly before the surgery, Suzanne did begin to fear going under the knife. [But prayer renewed her courage.] Her excitement and commitment returned.

During a three-hour laparoscopic surgery by Dr. Abdelkader Hawasli, Suzanne's kidney was removed. It was then placed into Grace by Dr. Henry Oh, chief transplant surgeon at St. John's, during a slightly longer surgery. Dr. Oh said it was major, but not dangerous. "Anyone can undergo it with minimal discomfort," he said.

He noted later that, "We had screened two dozen potential donors who had the right physical match, but Suzanne seemed the best as far as overall willingness and psychological makeup." He pointed out that the rigid screening and testing beforehand is essential because there are some people who donate

organs for ulterior motives – like a lonely person looking for attention, or someone who might do it in order to get money from the recipient afterward. "Suzanne deserves applause for what she did – giving the ultimate gift of life, part of her body to someone else," Dr. Oh said. "She carried out what the Bible teaches."

As a result, Grace not only had a new kidney, but she and her family would soon have a new life. Not everyone is so fortunate. According to the Gift of Life Michigan, a non-profit full-service organ and tissue recovery agency in Ann Arbor, there are 2,075 persons in the state waiting for a kidney transplant. Last year, 663 of them received a transplant. One hundred fifty died while waiting. Suzanne worked for Gift of Life as a coordination specialist until recently, and still speaks for them at some functions. Within a week of the operation, Suzanne was driving a car. She was back to work in four weeks. "God has given me incredible health all of my life," she said, "and within six weeks I was feeling as good as ever. When I look at all that Grace had gone through compared to me, I consider her a hero. I am grateful that God showed me one of the major purposes of my life – why I am here – to donate a kidney."

The second woman? Her life was totally transformed. Today, Grace states that she is 100 percent better. "I have much more energy, eat whatever I want, the hypertension is gone, the lupus in remission. I am grateful to God, and have an increased faith in him." Her career, not surprisingly, is taking a new path. A registered nurse, she now intends to focus on working with kidney patients. "My heart bleeds for them," she says. "Though I'm not sure exactly what yet, there is a work for me to do." Even her attitude has changed. "I see how precious life is, and I have more compassion and tolerance for others."

Fifteen days after the transplant, at Grace's request, she was able to meet Suzanne. Suzanne had never intended to learn who the recipient was because it was "a personal, private thing for me." But her husband and others convinced her that meeting Grace could lead to some positive publicity for organ transplants, and be an opportunity to educate the public and reduce fears about the procedure. This was especially true since Grace's transplant was the first one in Michigan by a live donor to someone who was not a friend or relative.

The two of them hit it off immediately. Grace calls Suzanne her angelic hero. "I love her. Her priceless, selfless act of mercy gave me a second chance at life," she says.

Tom Fales, who has been married to Suzanne for 25 years, says, "She is a saint in my mind. She answered the age-old question of why God gave us two kidneys. She is the most selfless person I have ever met. I admire and respect her for what she did. I wish I could be more like her."

Note 1: Adapted from *FaithMag.com*, the magazine of the Catholic Diocese of Lansing, Michigan, 24 October 2005. Reprinted with permission.

Summary Statements of Various Religious Groups About Organ and Tissue Donation

adapted from the United Network for Organ Sharing, www.unos.org

The following summary statements concerning various religious groups' positions on organ and tissue donation and transplantation may be of help to you. Perhaps you can help your religious group adopt a more clearly defined position. A pro-active position does, indeed, help clarify a group's attitude on the subject. Your knowledge and action may help alleviate the suffering of the thousands of people who die annually for lack of available donor organs and tissues while a multitude of healthy organs are being buried every day. This dilemma is within itself an ethical issue.

AME & AME ZION (African Methodist Episcopal) Organ and tissue donation is viewed as an act of neighborly love and charity by these denominations. They encourage all members to support donation as a way of helping others.

AMISH The Amish will consent to transplantation if they believe it is for the well-being of the transplant recipient. John Hostetler, world renowned authority on Amish religion and professor of anthropology at Temple University in Philadelphia, says in his book, Amish Society, "The Amish believe that since God created the human body, it is God who heals. However, nothing in the Amish understanding of the Bible forbids them from using modern medical services, including surgery, hospitalization, dental work, anesthesia, blood transfusions or immunization."

BAPTIST Baptist groups have supported organ and tissue donation as an act of charity and leave the decision to donate up to the individual.

BUDDHISM Buddhists believe that organ/tissue donation is a matter of individual conscience and place high value on acts of compassion. Reverend Gyomay Masao, president and founder of the Buddhist Temple of Chicago says, "We honor those people who donate their bodies and organs to the advancement of medical science and to saving lives." The importance of letting loved ones know your wishes is stressed.

CATHOLICISM Catholics view organ/tissue donation as an act of charity and love. Transplants are morally and ethically acceptable to the Vatican. According to Father Leroy Wickowski, Director of the Office of Health Affairs of the Archdiocese of Chicago, "We encourage donation as an act of charity. It is something good that can result from tragedy and a way for families to find comfort by helping others." Pope John Paul II has stated, "The Catholic Church would promote the fact that there is a need for organ donors and that Christians should accept this as a 'challenge to their generosity and fraternal love' so long as ethical principles are followed."

CHRISTIAN SCIENCE The Church of Christ Scientist does not have a specific position regarding organ donation.

According to the First Church of Christ Scientist in Boston, Christian Scientists normally rely on spiritual instead of medical means of healing. They are free, however, to choose whatever form of medical treatment they desire - including a transplant. The question of organ/tissue donation is an individual decision.

EPISCOPAL The Episcopal Church passed a resolution in 1982 that recognizes the life-giving benefits of organ, blood and tissue donation. All Christians are encouraged to become organ, blood and tissue donors "as part of their ministry to others in the name of Christ, who gave His life that we may have life in its fullness."

GREEK ORTHODOX According to Reverend Dr. Milton Efthimiou, Director of the Department of Church and Society for the Greek Orthodox Church of North and South America, "the Greek Orthodox Church is not opposed to organ donation as long as the organs and tissue in questions are used to better human life, i.e., for transplantation or for research that will lead to improvements in the treatment and prevention of disease."

The **ROMA**, also referred to as "gypsies," are a people of different ethnic groups without a formalized religion. They share common folk beliefs and tend to be opposed to organ donation. Their opposition is connected with their beliefs about the afterlife. Traditional belief contends that for one year after death the soul retraces its steps. Thus, the body must remain intact because the soul maintains its physical shape.

HINDUISM According to the Hindu Temple Society of North America, Hindus are not prohibited by religious law from donating their organs. This act is an individual's decision. H. L. Trivedi, in Transplantation Proceedings, stated that, "Hindu mythology has stories in which the parts of the human body are used for the benefit of other humans and society. There is nothing in the Hindu religion indicating that parts of humans, dead or alive, cannot be used to alleviate the suffering of other humans."

INDEPENDENT CONSERVATIVE EVANGELICAL Generally, Evangelicals have no opposition to organ and tissue donation. Each church is autonomous and leaves the decision to donate up to the individual.

ISLAM The religion of Islam believes in the principle of saving human lives. According to A. Sachedina in his Transplantation Proceedings' (1990) article, Islamic Views on Organ Transplantation, "the majority of the Muslim scholars belonging to various schools of Islamic law have invoked the principle of priority of saving human life and have permitted the organ transplant as a necessity to procure that noble end."

JEHOVAH'S WITNESSES According to the Watch Tower Society, Jehovah's Witnesses believe donation is a matter of individual decision. Jehovah's Witnesses are often assumed to be opposed to donation because of their belief against blood transfusion. However, this merely means that all blood must be removed from the organs and tissues before being transplanted.

JUDAISM All four branches of Judaism (Orthodox, Conservative, Reform and Reconstructionist) support and encourage donation. According to Orthodox Rabbi Moses Tendler, Chairman of the Biology Department of Yeshiva University in New York City and Chairman of the Bioethics Commission of the Rabbinical Council of America, "If one is in the position to donate an organ to save another's life, it's obligatory to do so, even if the donor never knows who the beneficiary will be. The basic principle of Jewish ethics - `the infinite worth of the human being' - also includes donation of corneas, since eyesight restoration is considered a life-saving operation." In 1991, the Rabbinical Council of America (Orthodox) approved organ donations as permissible, and even required, from brain-dead patients. The Reform movement looks upon the transplant program favorably and Rabbi Richard Address, Director of the Union of American Hebrew Congregations Bio-Ethics Committee and Committee on Older Adults, states that "Judaic Responsa materials provide a positive approach and by and large the North American Reform Jewish community approves of transplantation."

In 1984, the **LUTHERAN** Church in America passed a resolution stating that donation contributes to the well-being of humanity and can be "an expression of sacrificial love for a neighbor in need." They call on members to consider donating organs and to make any necessary family and legal arrangements, including the use of a signed donor card.

MENNONITES have no formal position on donation, but are not opposed to it. They believe the decision to donate is up to the individual and/or his or her family.

MORAVIAN Robert E. Sawyer, President, Provincial Elders Conference, Moravian Church of America, Southern Province, states, "There is nothing in our doctrine or policy that would prevent a Moravian pastor from assisting a family in making a decision to donate or not to donate an organ." It is, therefore, a matter of individual choice.

The **MORMONS** (Church of Jesus Christ of Latter-Day Saints) believes that the decision to donate is an individual one made in conjunction with family, medical personnel and prayer. They do not oppose donation.

PENTECOSTALS believe that the decision to donate should be left up to the individual.

PRESBYTERIANS encourage and support donation. They respect a person's right to make decisions regarding his or her own body.

SEVENTH-DAY ADVENTISTS strongly encourage donation and transplantation. They have many transplant hospitals, including Loma Linda in California, which specializes in pediatric heart transplantation.

In **SHINTO**, the dead body is considered to be impure and dangerous, and thus quite powerful. "In folk belief context, injuring a dead body is a serious crime. . .," according to E. Namihira in his article, Shinto Concept Concerning the Dead Human Body. "To this day it is difficult to obtain consent from bereaved families for organ donation or dissection for medical education or pathological anatomy. . . the Japanese regard them all in the sense of injuring a dead body." Families are often concerned that they not injure the itai, the relationship between the dead person and the bereaved people.

UNITED CHURCH OF CHRIST is very supportive of organ sharing.

UNITED METHODIST Church in 1992 states that "Donation is to be encouraged, assuming appropriate safeguards against hastening death and determination of death by reliable criteria. ... Pastoral-care persons should be willing to explore these options as a normal part of conversation with patients and their families."

Our thanks for donations from:

The complete thank-you list appears only in the paper edition.

cont'd from page 13

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Continuing Medical Education credits will be available to attendees.

Scientists and medical professionals are invited to submit papers and posters on the following topics:

Thursday, October 26:

Understanding the basic science of VHL and how a drug might be designed to help

Friday, October 27:

Improving diagnosis and treatment of

- Hemangioblastomas of the retina, brain, and spinal cord
- Kidney Cancer of VHL and other hereditary kidney cancer syndromes
- Pancreatic tumors, determining which tumors need attention when -- finding the balance point to maintain health
- Pheochromocytomas and Paragangliomas of VHL and other hereditary Pheo/PGL syndromes

Saturday, October 28:

Living with VHL

- When, why, and how to do DNA testing
- Dealing with the psychological pressures of knowing you have a medical syndrome
- Family dynamics, relatives who don't want to know, managing caregiver stress
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For further information and specific submission instructions, see www.vhl.org/conf2006

The city of London is located in the heart of Southwestern Ontario, Canada. It is home to one of the country's leading univertisities, the University of Western Ontario, the Robarts Research Institute, and the Canadian Medical Hall of Fame and the Canadian Baseball Hall of Fame. London is the seat of Middlesex County, at the forks of the Thames River, almost exactly halfway between Toronto and Detroit. It is served by an airport (code YXU).

The language of the meeting will be English. The intent of the symposium is for the physicians and scientists to meet and collaborate. Patients and family members are certainly welcome, on the understanding that you will be listening in on a medical meeting. We will not ask the presenters to constrain the scientific language of their presentations. There will be opportunities for the patients and family members to play a more active role in the events of Friday and Saturday.

VHL FAMILY FORUM

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