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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VHL and Oxygen Regulation

VHL plays a role in regulating several key functions in the cell, binding with other proteins and marking them for degradation – essentially serving as an "off" switch for the function of these other proteins. You have heard a great deal about VHL's role in regulating VEGF which causes the growth of new blood vessels. Even more important is its role in the detection of low oxygen levels (hypoxia), regulating HIF-1alpha (hypoxia-inducible factor), leading to increased production of HIF-controlled genes, including erythropoietin (Epo) a hormone which stimulates the production of red blood cells, VEGF, and many other proteins. Without sufficient Epo, you can become severely anemic. With too much Epo, you can have too many red blood cells, a condition which is called polycythemia. Some rare patients with VHL syndrome have polycythemia, which has been attributed to Epo production by a tumor.

In the Chuvash Republic, a mid-Volga region of European Russia, there is a relatively high concentration of polycythemia which was clearly seen to be hereditary. Recently the genetic cause was mapped to the short arm of chromosome 3, the VHL gene. This Chuvash Polycythemia (CP) behaves differently from other VHL mutations. The most striking difference is that you only get CP if you have two altered copies of the VHL gene.

In the VHL mutations we know as causing the tumor-causing condition we know as VHL disease, inheritance is dominant, meaning that you need only one altered copy of the VHL gene to have a predisposition for development of VHL disease. In fact, a fetus with two altered copies of the VHL gene will normally die in utero. But the CP mutation is in a position where one copy seems to cause no symptoms, and only when two CP mutations are inherited does the individual show signs of polycythemia. This is helping us understand the functional importance of the various parts of the gene. Its position is near one of the mutations currently associated with a VHL pheo family.

The Chuvash converted to Christianity, and were surrounded by peoples who embraced Islam. They kept to themselves, and rarely intermarried with the surrounding peoples, keeping the gene pool relatively distinct. Among the Chuvash, the CP mutation is fairly common, leading researchers to wonder whether having one gene with the CP mutation has some beneficial effect. One copy of the sickle cell mutation, for example, protects that person from getting malaria, which in Africa is a very good thing. Two copies of the sickle cell mutation, however, cause sickle cell disease, which is a painful and crippling condition. But the beneficial effects of the single mutation are enough to ensure that this mutation will be preserved in the population.

VHL mutations have subsequently been found in a number of children with congenital polycythemia who are outside the Chuvash ethnic group – two Danish siblings, several US Caucasians, and two Pakistani and one Bangladesh families living in the U.K. Only one of these has any of the tumors normally associated with VHL disease. All are children, however, so it is possible that some VHL-related tumors may occur in later years. An ongoing epidemiological study of the Chuvash population has so far failed to identify an increased risk of VHL tumors. This may be due to other genetic or environmental factors such as carcinogen exposure or modifier genes. Learning more about these factors might also provide strategies for dealing with VHL disease.

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Inside this issue!

About half the patients with apparent congenital polycythemia and increased serum Epo have VHL mutations. The multi-national research team headed by Dr. Josef Prchal at Baylor College of Medicine in Houston, Texas, now believes that "VHL mutations are the most frequent cause for congenital polycythemia and define a new class of polycythemic disorder, polycythemias due to augmented hypoxia sensing," as recently published in the American Journal of Human Genetics (Pastore et al, August 2003).

References: Y.D. Pastore et al., Mutations in the VHL gene in sporadic apparently congenital polycythemia, Blood (United States), Feb 15 2003, 101(4) p1591-5

S.O. Ang et al, "Disruption of oxygen homeostasis underlies congenital Chuvash polycythemia", Nat Genet (United States), Dec 2002, 32(4) p614-21

Y.D. Pastore et al., "Mutations of von Hippel-Lindau tumor suppressor gene and congenital polycythemia," Am J Hum Genet (United States), Aug 2003, 73(2) p412-9

A woman in traditional Chuvash dress, beautifully embroidered.

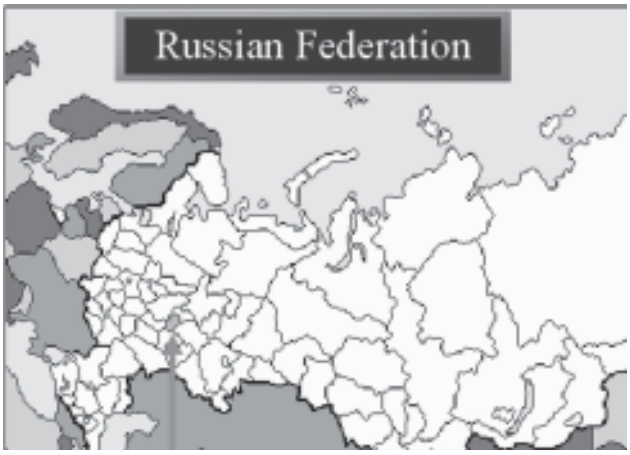
Can You Help?

This team would appreciate the help and participation of any family with pheochromocytomas that also has a member with polycythemia, or whose VHL mutation has not yet been found.

Please call Dr. Prchal or Dr. Pastore directly, or call Joyce Graff to discuss this further.

Dr. Prchal or Dr. Pastore:
+1(713)798-7520

Joyce Graff in the VHL Office:
+1 (617) 277-5667 or
1-800-767-4VHL, option 4



The Chuvash Republic [ch$\overline{oo}$väsh´] or **Chuvashia,** (1990 pop. 1,326,000), 7,066 sq. mi. (18,301 sq km), is a member state of the Russian Federation, in East central European Russia, in the middle Volga valley. Cheboksary is the capital. The region, consisting largely of the Chuvash plateau, is wooded steppe with some peat bogs. Grain, potatoes, flax, hemp, fruit, and sugar beets are grown, and livestock is raised. With about one third of the area in forests, both lumbering and woodworking are important occupations. Among the republic's other industries are oil and natural gas refining, metalworking, railway repair, and food and flax processing. The Trans-Siberian Railroad crosses the republic, and secondary lines from the main track service many towns. The Chuvash ethnic group makes up some 70% of the population and Russians (who are mostly urban) around 25%; there are Mordovian, Tatar, and Ukrainian minorities. The Chuvash, descendants of the medieval Bulgars, represent a mixture of Finnish and Mongolian peoples. They speak a Turkic language and adhere to Orthodox Christianity. Conquered by the Mongols in the 13th and 14th centuries, the Chuvash came under Russian rule in 1552. The Chuvash Autonomous Region was established in 1920; it became an autonomous republic in 1925. A revival of Chuvash culture began in 1968. Chuvashia was a signatory to the 1992 treaty that created the Russian Federation. It has a 200-member parliament.

Chuvashia is sometimes called the land of one hundred thousand embroideries and songs. Preserving national traditions, Chuvashes painstakingly protect their folklore and national crafts. Since olden times, Chuvash embroidery and woodcarving have been famous in the neighboring regions; this ancient trade has grown into a factory production, and nowadays art works receive recognition at international exhibitions, and are exported. The brilliant, beautiful dances of Chuvashes and their fine musical melodies affect the hearts of all those who see and hear them.

Adapted from Chuvash Republic: Invitation to Cooperate, http://www.cap.ru/cap/main.asp?id=103

Important New Information for Physicians

In the June issue of *Lancet*, a team of physicians from the U.S. National Institutes of Health has published an overview of von Hippel-Lindau — the clinical aspects, management, and treatment options for successful management of VHL.

The article summarizes the current state of learning on VHL, including a survey of the literature published over the past ten years. It updates many of the statistics and recommendations that have been seen in earlier papers.

"Before comprehensive screening surveys became routine, median survival of patients with the disease was less than 50 years of age [due especially to] complications linked to renal cell carcinomas and CNS hemangioblastomas. Improved surveillance, earlier diagnosis of lesions by modern imaging and laboratory studies, improvements in treatment, and increased knowledge of this disease have improved prognosis and reduced the complications related to these tumors." (p. 2059)

They include the following recommended intervals for screening in at-risk individuals, which begin at much younger ages than previous recommendations:

- Ophthalmoscopy: start in infancy, repeat yearly
- Plasma or 24 hour urinary catecholamines and metanephrines: start at 2 years of age, repeat yearly and when blood pressure is raised.
- MRI of craniospinal axis: begin at 11 years of age and repeat yearly
- CT and MRI of internal auditory canals: begin at onset of symptoms (hearing loss, tinnitus, vertigo, or unexplained difficulties of balance)
- Ultrasound of abdomen: begin at 8 years of age, and repeat yearly or substitute MRI as clinically indicated.
- Audiological function tests: begin when clinically indicated

While VHL is usually found to have been inherited in the family, it occurs for the first time in as many as 20% of families studied. Sometimes the first mutation arises as a VHL mosaicism, with the VHL gene mutation occurring in only some of the cells of the body. In such cases, "patients might have clinical signs of the disease, but test negative genetically" because the VHL mutation is not in all cells. If the mutation involves the sperm or egg cells, however, the condition can be passed to offspring, so children should be considered to be potentially at risk for VHL.

While the average age for first occurrence of retinal tumors is 25 years, "5% of retinal angiomas present in patients younger than 10 years of age." For this reason they now recommend beginning screening the retinas in infancy. "Early diagnosis and treatment can prevent visual loss or blindness."

Endolymphatic sac tumors (ELST) are rare in the general population, but are frequently associated with VHL. MRI studies found evidence of ELST in 11% of patients. "VHL is the only condition associated with bilateral ELST." Additional patients may have microscopic tumors smaller than can currently be seen on imaging, because "patients often have vestibulocochlear [inner ear] symptoms with no CT or MRI evidence of such a tumor." (p. 2062).

"The new insights into the underlying mechanisms of tumor formation, greater knowledge of the natural history of the various lesions associated with von Hippel-Lindau disease, and more precise diagnostic studies (laboratory and imaging) should lead to an improved quality of life and extend the life expectancy of affected individuals. The diverse multisystem effects of this disease need careful, selective, and coordinated planning to determine the treatment of individual lesions that will provide the best long-term management of these patients." (p. 2066)

This is an excellent overview for physicians of this complex multi-system disorder. We recommend that all specialists, general practitioners and other health professionals working with any person with VHL obtain and read this important article. The *Lancet* is one of the most prestigious medical journals in the world, and should be readily available through any hospital library. Patients are encouraged to share this information with their health care teams.

Reference: "Von Hippel-Lindau disease," by Lonser, Glenn, Walther, Chew, Libutti, Linehan, and Oldfield of the U.S. National Institutes of Health, Bethesda, Maryland. *Lancet* 361 (2003): 2059-67.

Regional Meetings --
Coming Soon to a location near you!

Indianapolis, October 25
Boston, November 1
Kochi, Japan, May 20-22
New York, June 19

See page 13 for details

A Century-Old Exercise Becomes Popular Again…

And Proves Success Doesn't Have to Mean Being Sweaty!

by Amy G. L., Arkansas

A little background…I had my first brain tumor removed in 1984 and was officially diagnosed with VHL in 1992 after a second brain surgery. I've had two partial nephrectomies in 1993 and 1994, one eye laser in 1996, spinal surgery in 1997, and a third brain surgery in 2002. The brain tumors left me with ataxia, and after the spinal surgery I had more difficulty walking and developed RSD of the right leg (reflex sympathetic dystrophy). I've undergone physical therapy off and on since 1992. Today, I walk with a cane, or weave around without one on 'better days.' I use a wheel chair when traveling in an airport to conserve energy. I work out two or three times a week: 45 minutes on a stationary bike or elliptical machine, and another 30 minutes on machines and/or light weights. In the future, I plan to be back in a therapy pool every day and exercising, mostly using Pilates…

When I first read about Pilates (puh-LAH-teez), the exercise system that strengthens and stretches the body's core muscles (abdominal, lower back, hips, and buttocks), I had high hopes that it would help me because the exercises were specifically designed for people with disabilities. I've now taken 11 hour-long lessons since May, and noticed differences almost immediately. After three lessons, an ankle that for 6 years has always swelled by the end of the day hadn't swollen once, even after a long plane trip or a day spent mostly sitting, which I consider a minor miracle and attribute to Pilates. The exercises not only strengthen and tone the body, but also encourage more circulation. I'm walking better, standing straighter than ever before, and feel stronger and more balanced.

I cannot recommend this exercise enough. I've taken yoga off and on for years, and while some of the exercises remind me of yoga, Pilates makes me feel much better overall.

I think it important to note, however, that I'm touting private lessons using the Pilates equipment. While there are group classes available where you learn different floor exercises and 'mat' work, as well as books and tapes available for home use, Pilates takes concentration, control, and practiced breathing. You really need to know you are doing it properly from the first. I don't discount these other ways to learn, in fact, once I'm confident I'm performing the exercises correctly I will probably go these routes, too, but this is an exercise routine that is tricky at first: there are subtle movements you must learn to make in your breathing in conjunction with your posture and abdominal muscles, and if done incorrectly, you can end up with little results, or worse, inadvertently hurting yourself. Start by investing in some private lessons; the one-on-one coaching could make the difference between failure and success.[1]

Doing a little web research on the subject, I learned that most instructors agreed: if you aren't strong enough to control your muscles, you should begin by using the machines first, building towards strengthening the body. Learning Pilates takes time; my instructor calls it the "thinking person's exercise," because you are engaging your body to work in unison with your muscles, and this takes time to learn (at least, it does for me). But it's well worth the effort. By lengthening joints and strengthening muscles, you gradually strengthen the body to deal with daily activity and sports. Once you learn to engage core muscles, you automatically learn to hold in your abdomen all the time. This alone helps greatly with chronic back pain.

Joseph Pilates was a German with an interest in the human body. Weak and sickly as a child, he taught himself exercise techniques that were so successful he was encouraged to begin working with disabled soldiers in World War I, strengthening their muscles with mild resistance exercises they could perform in their beds using bedsprings. He eventually moved to New York, where Ballanchine dancers began using his techniques to rehabilitate their bodies. Pilates wanted people to learn to focus on their bodies and not on their injuries – thinking of themselves as healthy, normal people who happened to have injuries that needed correcting. He felt that this way of thinking enables one to take responsibility for their body's health and overall well-being.

If Pilates intrigues you, call the National Pilates Studio (1-800-4PILATES) to ask for a master list of instructors near you, and give it a try. On the Internet, WebMD has articles on the subject. They will warn you to find an instructor with at least a year or two of solid instruction – this is such a controlled discipline that a teacher having only a week or weekend course will not suffice!

1. Note: Some pilates movements involve sudden or prolonged hyperextension of the neck and back that could pose a problem for persons with lower brainstem or spinal cord hemangioblastomas or previous spine surgeries. Also, holding some of the positions can involve a Valsalva maneuver that can increase Central Nervous System (CNS) pressure. Anyone with CNS hemangioblastomas or prior surgeries should check with their physician before starting a program, and, as Amy recommends, learn with a personal instructor.

DNA Testing for Young Children

– A conversation from our online support group, vhlfa@yahoogroups.com

Question: I am pregnant with our first child and have decided against testing for VHL with amniocentesis because the result would make no difference to us. We have been told, though, that we will have to wait until he is 5 years old before they can test once he's been born. Is that right? It doesn't seem to make any sense to us. – *Anne B., England*

Answer #1: Technically, genetic testing is possible at any age — from the womb (via amniocentesis) through childhood (via blood draw) and adulthood (via blood draw). In fact, preimplantation genetic diagnosis has also been performed for VHL and other genetic disorders — by testing an embryo before it is even implanted into a woman's uterus.

The philosophy for testing kids for many autosomal dominant disorders including VHL really seems to vary by institution. Our clinic's philosophy is not averse to VHL testing in children of any age provided it is for the benefit of the child (not the parent) and is in the context of careful genetic counseling. In VHL, DNA testing can be used to determine if a child can be spared regular screening, or should be monitored more closely for possible early signs of VHL.

In regards to the age cutoff of 5 years that you mention here, we can't think of any biological or ethical reason to use this age as a minimum.

– *Gayun Chan-Smutko, M.S., and Kristen Mahoney Shannon, M.S., genetic counselors, Massachusetts General Hospital, Boston, Massachusetts.*

Answer #2: My child was one of those that developed three pheos before age 5 — he was diagnosed at the age of 4, had the genetic test at the age of two. Thank goodness we did it early! And I think you wonder and worry before you ever have the test, so why put it off? My daughter also had a pheo removed at the age of 8. Yes, it's hard at times. But I wouldn't trade them for the world. I would have more children in a heartbeat.

Only a small percentage of very young children have tumors, but it is still worth mentioning. My kids are doing great now! And just my two cents - everybody has something, you're just lucky to know what to look for early before it's too much of a problem. That's how we feel about it. It's all in God's hands, and he doesn't make mistakes! Good luck to you and your family. – *Alison E., Texas*

Answer #3: We had our oldest daughter tested at 3 months and with our second daughter we had cord blood taken during the delivery which was used for the DNA testing. The genetics dept at the Children's Hospital made arrangements with the obstetrician. – *Amy B., Connecticut.*

Answer #4: Testing kids, eh? I'd test right away, though the decision is personal, I understand.

Here's why. My only child was 3 when I was diagnosed at 31 years of age (June 1987) and not very much was known about VHL, at least compared to today. There is no history of VHL anywhere in my family before me. It seems I'm a mutant, according to the geneticist I talk to. She says we're 'all mutants', but some mutations are as benign as a freckle on your face that neither you mom nor your dad had. My mutation got me membership into Club VHL, where all the nicest people hang out.

My daughter had a brain tumor (exactly like mine, we have the same scar today) removed when she was 13 years old. That was in 1997. Up to that point, my "VHL doctor" told my family that there was no need to test anyone under 12 years of age. Indeed, no blood testing was available, and what could be done involved all the familiar screening. She was never tested.

When my daughter began to manifest symptoms of a brain tumor, all of her doctors dismissed the chance that she might have a tumor, despite my history. Honestly, the most brilliant people can overlook the most obvious things. Einstein forgot to tie his shoes, or so goes the story. They blamed her symptoms on allergies, puberty, migraines, etc. Finally, when she complained of double vision one day, my very alert wife took her to an ophthalmologist immediately, who confirmed the increased interocular pressure. The tumor was removed successfully the next day.

I sure wish we could have tested her a long time ago. That tumor almost killed her. Advance notice would have been good.

Life is. –*Bob D., California*

The U.S. National Institutes of Health now recommends DNA testing by age 1, and screening of young children at risk beginning in the first year of life.

Early diagnosis and appropriate treatment are the keys to maintaining health.

See new recommendations from the U.S. National Institutes of Health, reported on page 3 of this issue.

Pain versus Promises

By Abbey Meyers, Connecticut

Editor's note: While we do not yet have an approved drug for VHL, we are hopeful that within the next five years there will be at least one. It is important that we watch legislative activity around similar drugs in each of our local areas, and let our representatives know how we feel, in order to ensure that when drugs are approved, they will be accessible.

Twenty years ago, when President Ronald Reagan signed the Orphan Drug Act into law, he said, "I only wish with the stroke of this pen I could also decree that the pain and heartache of people who suffer from these diseases would cease."

This is a promise that has been kept, with remarkable success.

In the decade before the Orphan Drug Act was enacted, only 10 new treatments for orphan diseases were developed. In the 20 years since it was passed, more than 240 new treatments for rare disorders have been approved by the Food and Drug Administration. An additional 900 potential new treatments are in various stages of development.

People once dying from rare diseases are now living with them. People who were incapacitated by the pain that disease inflicts on the human body are now leading productive, happy and pain-free lives.

Unfortunately, an important part of the ongoing legacy of the Orphan Drug Act is now in jeopardy. The federal Centers for Medicare and Medicaid Services (CMS), responsible for Medicare and Medicaid, has recently made most injectable and intravenous orphan drugs less available for Medicare patients. While the CMS regulation is arcane and complex, the effect is likely to be tangible and devastating for those suffering from rare diseases.

Last January, CMS made drastic cuts -- an average of 35 percent -- to Medicare reimbursement rates for drugs covered by the Hospital Outpatient Prospective Payment System (HOPPS). Hospitals must choose between losing money on every injection or living up to their responsibility to alleviate pain and suffering. They should not have to make these choices.

Those with rare diseases are particularly at risk when these decisions are being made. Orphan drugs sustained cuts at least as deep as the average. Hospitals are less likely to stock orphan drugs because they are associated with a small number of patients. A number of orphan drugs are bundled with high-volume, lower-cost drugs, assuring a wide gap between the cost of the orphan drug and the Medicare reimbursement. This didn't need to happen.

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Within the HOPPS program, CMS has the authority to exclude all drugs designated as orphans by the Food and Drug Administration. Of the 85 to 90 orphan drugs used by the Medicare HOPPS program, CMS excluded only four.

People with rare diseases are re-experiencing pain -- acute and immediate -- as well as pain from knowing that a bureaucratic decision is keeping them from cutting-edge drug therapies. CMS is backtracking on the covenant that Congress and the federal government has had with the orphan disease community to encourage development of new "orphan" treatments and make them accessible to patients.

Dean Cole, 53, of Little Rock, Arkansas, is living proof of the medical progress CMS is reversing through this rule. Suffering from dystonia, a rare neurological movement disorder, Mr. Cole experiences recurring severe pain all day, every day, from the cramping the disease causes in his arm and shoulder muscles. Four years ago, he found some relief when his doctor injected an orphan drug into his muscles. Three shots every three months, and much of Mr. Cole's pain from spastic muscle cramping was alleviated.

In January, because of the Medicare reimbursement cuts in the HOPPS rule, the hospital told Mr. Cole that it would no longer provide his orphan drug treatments. Spastic muscle contractions returned, along with horrific pain. Mr. Cole has called CMS, dystonia support groups, and legislators for answers. Recently, he told the Arkansas *Democrat-Gazette* that he doesn't care about Medicare reimbursements or orphan drug legislation. "All I want," he said, "is for the pain to stop."

So what has to happen for the pain to stop for Mr. Cole and thousands like him?

Further action by CMS is unlikely. Intervention by Congress is, therefore, necessary so that Medicare will pay for injectable and intravenous orphan drugs in outpatient hospital settings. The upcoming package of Medicare enhancements and reforms presents an ideal vehicle.

Congress must quickly legislate this fix so that patient access isn't denied; patient pain and suffering is alleviated and the progress that began in 1983 with the stroke of President Reagan's pen isn't reversed.

Abbey Meyers is President of the National Organization for Rare Disorders (NORD).

Britain plans a genetic census

How do nature and nurture combine to cause diseases?

adapted from an article by Gwen Kinkead, The New York Times, *Thursday, January 2, 2003*

Note: Projects like this one are being proposed in many countries, including the U.S. The potential for learning is vast; the ethical concerns of such a project are enormous. For example, the U.S. Department of Defense maintains a DNA tissue bank of all military personnel – for identification of remains, if necessary. There have been several requests to use this DNA bank for criminal investigations: rapes, the Washington sniper, etc. These requests have often, but not always, been blocked. Who should have access to DNA information, and for what?

In 2003, Britain plans to undertake the world's most ambitious study of the origins of disease. Looking forward to the day when people will know their genetic makeups and request a precise picture of their risks of developing various diseases, the study organizers plan to assemble a database of medical information about 500,000 Britons, including their DNA.

The goal is to sort out over the next 10 to 20 years the way that genes and the environment combine to cause common diseases.

A few countries -- including Estonia, Sweden and Latvia--are considering similar databases, in essence genetic censuses. Iceland, the pioneer, has collected medical data and DNA samples from 80,000 related people to hunt genes that touch off disease.

It will also develop a national database from patient records in its health care system. DNA-based diagnostics and drugs could result from the effort.

Britain's will be the largest of the databases proposed by governments or their private partners. It has the same goals as Iceland's but with a critical difference: It will try to quantify for the first time the roles of genes and such environmental influences as smoking, alcohol, viruses, pollution, exercise and diet in unrelated people for all common diseases. Studying a huge selection of diverse people could make its discoveries applicable worldwide. Iceland's population, by contrast, is extraordinarily homogenous.

If the $120 million project, called U.K. Biobank, goes forward, and enough people volunteer for pilot studies, 1.2 million healthy Britons from 45 to 69 will give blood samples to the Biobank. From their blood, DNA will be purified and frozen. Ninety percent of the donors will be white. The rest will roughly reflect Britain's demographics. From these, 500,000 will be chosen for the project by 2008.

When they sign up, volunteers will get brief health examinations and will answer 10-page questionnaires about their socioeconomic and psychological status, reproductive history, exercise, cell phone use and beverage preferences. They will note their diets for a week.

For 10 years, they will be followed through their national health care records, which will be copied into the Biobank. The data will be anonymous, but not completely, to allow for updates by doctors or new questionnaires. By 2014, 40,175 are expected to fall ill with diabetes, heart disease, stroke or cancer, and 6,200 are expected to have Parkinson's disease, dementia, rheumatoid arthritis or hip fractures.

The DNA of these people will be read and compared, and any normal gene variants, the one-nucleotide differences in DNA that make one person's biology different from another's, will be tagged for study. "Then you will be able to see patterns: X number have this sort of genetic makeup and this kind of lifestyle, and Y has that, and you can start analyzing, if you like, the nature-nurture, environment-genes secret," said George Radda, a molecular cardiologist who heads the Medical Research Council, a sponsor of the Biobank.

So far, opposition has been muted and polite. But a significant minority of British doctors oppose the project as unnecessary and too costly.

American geneticists are also split on the value of huge medical and DNA databases. Some argue that existing ones, like the Framingham Heart Study, which is gathering DNA from descendants of its original subjects, are enough. Others question their design.

David Altshuler, a geneticist at the Whitehead/MIT Center for Genome Research and Harvard Medical School, said: "I am not sure that a one-size-fits-all gene bank is what we want, but it is absolutely necessary to do prospective population studies if we are going to give any valuable information to the average patient who walks into their doctor and says, 'Does this genetic discovery I read about in the paper apply to me?' or 'What does it mean if I have a certain gene variant?'"

Health officials in the United States are beginning to discuss a large database for research. Smaller databases, gathering DNA from volunteers, are being started by the Mayo Clinic, among others.

Some people worry that they will be exploited and their privacy invaded. Critics are asking that it be suspended, saying it has yet to answer crucial questions about access by the courts and pharmaceutical companies and the type of studies that will be allowed.

Biobank backers are consulting with ethicists, doctors, scientists, drug companies and the public before issuing final plans. That, they hope, will rally Britain around the plan and prevent the outrage that greeted Iceland's database and doomed another in Tonga.

UK Welcomes Mary Weetman

– Mary Weetman, Chairman, VHLFA U.K., Manchester

I consented to take on leadership following Dan Whitmore's term as Chairman.

My interest in VHL stems from 26 years' work as a genetic counselor in the Department of Genetics in Manchester. During this time I was in contact with several families with VHL, and learned of their many problems, and the great courage and resilience they show in their every day lives.

I am therefore pleased to take part in the reinvigorating of the group.

Our first meeting last year was well attended, and all were keen for the group to continue. We need to formalise this through the Charities Commission, and my frequent contacts with them have been very time consuming, but I will get there in the end! We will then form a committee, etc. to help with the running of the group.

In the meantime I am establishing a web-site for the U.K. In the course of so doing I have contacted all regional centres in the U.K. and all have been keen to be featured on the site and have been very encouraging. Several have expressed an interest in forming contact groups in their areas, so I see our initiative spreading across the U.K. This is essential as I receive phone calls and e-mails from people from all parts of the U.K. and would love to be able to put them in contact with families geographically closer to themselves.

Our next meeting is to be in Manchester in September. This will be open to all people I have had contact with in the U.K.

I myself am mother of three now adult sons, and have three grand daughters and a grandson. So like most women I have experienced all the joys and sorrows that come with close and loving relationships.

Since retiring I have embarked on an Open University course on world religions. This is fascinating, and hopefully will keep me mentally alert!

My husband is also retired, and active on the local arts scene, doing water colouring and play producing.

Our German wire haired pointer ensures that we get plenty of exercise each day. So life is busy, but I would not have it any other way.

I look forward to meeting current and new members of the UK group. People can reach me at +44 (0)1706 358055, or through the VHL answerphone number in London +44 (0)20 7681-1796 or by e-mail to uk@vhl.org

Mary Weetman, Chairman, VHLFA UK

Members are asked to support efforts in their own country first.

Individuals and country affiliates are also invited to contribute to the international research fund, VHLFA, 171 Clinton Road, Brookline, MA 02445 USA.

VHL Family Day at Guy's Hospital

Von Hippel Lindau – Care in the UK

– Peter Risby, Sally Watts, and Mary Weetman

Anyone with a diagnosis of VHL or at risk of developing VHL, can be referred by their general practitioner (GP) or specialist to their local genetics centre. Because VHL is an inherited genetic syndrome, care for VHL is coordinated through the various genetics services.

The UK is divided up into 13 regions, each of which has a Regional Clinical Genetics Centre. For further information on each centre please go to the web address: http://www.bshg.org.uk/Directory/UKdirectory.htm The majority of the centres will be able to coordinate VHL screening for individuals either in a regional one-stop clinic or in conjunction with local specialists.

The three regional centres featured here have the greatest amount of experience and expertise with VHL in the U.K., and often serve in a consulting or supplementary capacity with other local teams.

Guy's Hospital, London

A one-stop multidisciplinary screening clinic, held 3 monthly, was established in January 1998. The disciplines represented at the clinic are Clinical Genetics, Neurology, Ophthalmology and Renal Medicine. Adults and children requiring VHL screening are offered an abdominal ultrasound scan, eye examination and neurological examination during an afternoon. Results are available the same day. A 24-hour urine collection is also arranged. MRI scans are arranged prior to these appointments so that results can be discussed. There is also the opportunity to discuss genetic issues if required.

Currently about 45 families are known to the clinic from the catchment area which covers South East London, Kent and West Sussex (population approximately 4 million).

In November 2002 we held a VHL family day for our families, which proved very popular. Following a short presentation explaining the genetics of VHL and screening protocols we were able to answer questions and families had a chance to meet each other over an informal lunch.

Sally Watts, Genetic Counsellor/VHL screening clinic coordinator, Guy's Hospital, London SE1 9RT Tel: 020 7955 4648 sally.watts@gstt.sthames.nhs.uk

Churchill Hospital, Oxford

The Oxford service is led by Dr Susan Huson and covers a catchment area of 2.9 million people over four counties. We are treating about 30 known VHL patients (with proven mutations) and 40 more who are on long term follow up due to either a possibly VHL related tumour having been treated in their past or an affected family member.

The approach we take is to run a register which involves yearly contact to ensure that our records remain up to date as well as a one-stop clinic where patients can meet with the genetic team (doctors and nurses), as well as undergo eye examinations, MRI scans of the abdomen and blood screens. Brain and spine MRI scans are arranged if the person is symptomatic, but are most commonly arranged by the neurological service as part of their routine follow up post brain/spine surgery.

The one-stop clinic is now held 6 times per year, and has proven to be extremely popular. Results are given (as much as possible) on the day and all scans are reviewed one week later. Blood screen results are collated, and patients are then sent a final clinic letter to summarize the findings.

Peter Risby, Cancer Genetics CNS, Department of Clinical Genetics, Churchill Hospital. Old Road, Headington, Oxford OX3 7LJ England, U.K. Tel: +44 (0)1865 226019; Fax: +44 (0)1865 226011; Peter.risby@orh.nhs.uk

St. Mary's Hospital, Manchester

The protocol followed by the Manchester Genetic Centre is almost identical to that followed by Guy's and Oxford. We serve about 50 VHL families in a catchment area of 4.5 million people.

Families are seen on an annual basis, and all the screening tests are carried out in the Eye Hospital, and Manchester Royal Infirmary, which are all part of the same complex, and therefore convenient for the family.

Families can of course be seen at any time between their annual appointments if they so wish.

All the families are on the genetic register and so are easy to follow-up.

St. Mary's Hospital, Hathersage Rd, Manchester M13 0JH. Dr. Fiona Lalloo, Consultant Clinical Geneticist, and Mrs. Carol Giblin, Genetic Counsellor, Department of Clinical Genetics. Tel: +44 (0)161 2766322; Fax: +44 (0)161 2766145. carol.giblin@cmmc.nhs.uk

See the call for papers on page 12
or http://www.vhl.org/conf2004

Nashville was Great!

The first in our new series of Regional Meetings was held in Nashville June 21. To be more precise, it was held at St. Paul's Episcopal Church in Franklin, Tennessee, a charming old town south of Nashville.

Fifty-two people from as far away as Vancouver, Canada, met in Franklin to visit with other VHL families, and to hear excellent presentations from Dr. Gladys Glenn and Dr. Peter Choyke of the U.S. National Institutes of Health.

Most attendees were from the local region, as we had intended, including people from Tennessee, Mississippi, North Carolina, and Indiana. There was lots of time to share our stories and get acquainted, and a festive country lunch of fried chicken, Tennessee barbecue, and Matt Painter's famous baked beans.

In addition, most of the VHLFA Board members were there for the Annual Meeting of the VHL Family Alliance. The election of new board members was completed at the Annual Meeting, and the following people were installed:

- David Forsell, President, Keep Indianapolis Beautiful
- Thomas D. Rodenberg, esq., attorney, Kansas City, Kansas (re-elected)
- Joseph Verdi, Ph.D., Maine Medical Center
- Bruce Weinberg, JD, Kaplan, Los Angeles

Watch for more information about these great additions to the Board of Directors in the Annual Report

Southeast Regional Meeting chairman Don Marshall had arranged for a cake in celebration of the tenth anniversary of the founding of the VHL Family Alliance in 1993.

At this meeting we announced that the Board has engaged Joyce Graff to be the first Executive Director of the VHL Family Alliance, to lead us into a new era of growth and an expansion of services and grants.

Pictured left to right: members of the Board: Kathy Braden (Indiana), Matt Painter (Tennessee), Joyce Graff (Massachusetts), Patricia Rasmussen (Minnesota), Altheada Johnson (seated, New York), David Forsell (Indiana), Peggy Marshall (Mississippi), and Maria Shipton (Pennsylvania).

By-Law Changes

Adopted June 21, 2003

After ten years of successful operation, the Board felt that some changes were needed to the By-Laws. We have grown from three families in 1993 to serving a population of 14,000 people with VHL in 72 countries of the world. The core team needed to support this expanded organization and keep it growing, is quite different than what was needed ten years ago. We have done a remarkable amount in ten years with all volunteer staffing. At this point, in order to grow, we know that we need to strengthen the home office: to expand our funding base, to obtain corporate and foundation grants, and to provide our many wonderful volunteers with the support they need to continue doing their good work.

We had two primary motivations for reviewing the By-Laws:

(1) in reviewing the Chapter structure, we wanted to group states into Regions. We reviewed the By-Laws with this in mind.

(2) at this point in our growth, we wanted to hire an Executive Director, and to specify the roles and relationships between the Executive Director and the Board. In general, the title of Chairperson of the Alliance has been replaced throughout with Executive Director.

The following is a summary of the changes approved and adopted by the Board of Directors on June 21, 2003.

Article III: Membership

Opening Paragraph: Language was deleted to make it consistent with the membership requirements set forth in Article III.

Section 1: Language was added making it a requirement of membership to complete an initial application and pay dues on an annual basis. Language was added requiring families who are affected by VHL but are unable to pay dues to notify the Alliance of their desire to be members and complete an initial application.

Section 2: Language was added regarding family memberships to require the completion of an initial application and the payment of dues on an annual basis. Language was deleted that gave family memberships two (2) votes and language was inserted to provide only one vote per family. Language was added requiring families who are affected by VHL but are unable to pay dues to notify the Alliance of their desire to be members and to complete an initial application.

Section 3: Language was added making it a requirement of professional membership to complete an initial application and pay dues on an annual basis.

Section 5: The section was deleted as it was repetitive of provisions in Article V, Board of Directors.

Article IV: Membership meetings

Section 1: Language was added requiring that the annual meeting of the membership be held in conjunction with the annual meeting of the Board of Directors.

The title "Chairperson of the Alliance" was deleted and replaced with "Executive Director".

Section 2: Language was added requiring that the notice of a special meeting of the membership include the purpose(s) for which the meeting was called. The title "Chairperson of the Alliance" was deleted.

Section 5: Language was added requiring that proxies be mailed to the membership at least 45 days prior to the Annual Meeting, and must be returned at least 10 days before the meeting.

Article V: Board of Directors.

Section 2.a.: Language was added requiring that not less than fifty percent of the Directors be representatives of families affected by VHL.

Section 2.b.: The section was deleted as sentence one was redundant and by incorporating sentence two into Section 2.a.

Section 2.c.: Language was added allowing a Board officer to extend his or her term beyond six consecutive years in order to complete service as an officer.

Section 3: Language on chapter representatives on the Board was deleted. Provision regarding ex-officio members of the Board was deleted.

Section 4: Language was added providing that members of a director's immediately family may not serve during the term of a director with immediate family being defined as parents, spouse, children, brothers, sisters, aunts, uncles, domestic partners or members of the household.

Section 6: Language added regarding resignation of board members requiring notice in writing and acceptance by a simple majority.

Article VI: Board Meetings

Section 1: Language added permitting the Board by majority vote to close meetings to persons other than directors.

Section 2: Language was added requiring at least 48 hours notice of special meetings of the board. Language was deleted that allowed the Chairperson of the Alliance to call a special meeting of the board.

Section 6: Added that meetings of the board be governed by Roberts Rules of Order.

Article VII: Officers

Section 1: Deleted reference to "Chairperson of the Alliance" as an officer of the board. Language added permitting board to designate additional officers. Language was deleted which permitted Board to appoint officers.

Section 4: Language was added permitting the Board to employ persons as it deems necessary. Language was deleted stating that agents hold office during the pleasure of the Board.

Section 7: Deleted (replaced with Section 11. Executive Director.)

Section 8: "Vice Chairperson of the Alliance" changed to "Vice Chairperson of the Board".

Section 11: Added to provide for Executive Director.

Article VII: Committees

Section 1: "Fund Raising" changed to "Development". "Legislative" deleted.

Dr. Gladys Glenn, speaker, with Patricia Rasmussen, Director of Clinical Care

Section 2: Changed the composition of the Executive Committee to include officers of the corporation and one or more other directors selected by the Board.

Section 3: Changed the composition of the Nominating Committee to include the Chairperson of the Board, Vice Chairperson(s) and two other members of the Board elected by the Board. Deleted language requiring the Nominating Committee to present a slate of officers for election by the membership. Officers are elected by the Board.

Section 8: "Public Relations and Education Committee" changed to "Public Awareness Committee".

Section 9: Deleted (Legislative Committee).

Section 10: Language added requiring the By-laws Committee to provide opinions to the Board as to the interpretation of the By-laws.

Article IX: Advisory and Honorary Boards

Section 1: Language added giving the Medical Advisory Board authority to make recommendations and provide opinions on Clinical Care Centers.

Section 4: Language changed making the Chapter Council an option. Language added providing guidelines as to forming Chapter Council. Language deleted regarding specifics of Chapter Council function. Language deleted as to officers of Chapter Council.

Article XI: Contracts and Commitments.

Section 1: Language added giving authority to contract based on Board policy.

A copy of the By-Laws showing all the changes adopted, and a clean copy of the revised By-Laws as adopted, are available on the website, or can be supplied to you by paper mail upon request. For questions regarding the revised Bylaws, you may contact Thomas D. Rodenberg, Esq, Chair of the Bylaws Committee, Maria Shipton, Chairperson of the Board, or Joyce Graff, Executive Director.

Yard Sale Fundraiser

by Lynn G., Tennessee

On June 5th and 6th, 2003, a yard sale to raise money for the VHL Family Alliance was held in Manchester, Tennessee, by Juanelle S. and Helen V. Juanelle is a member of the VHLFA and is affected by VHL. Juanelle and Helen raised over $1000 through the yard sale. Money and items for sale were donated from members of the community, including a $200 donation from the Coffee County Visually Impaired Persons (VIP).

L to R: Juanelle and Helen

Juanelle is my aunt, and four members of our family are affected by VHL. My aunt has not been able to walk without using a walker for several years due to multiple surgeries for spinal tumors. Other members of the family have also been severely affected by VHL.

With limited financial resources and physical ability, it is easy to say that there's nothing we can do to help in the fight to find a cure for VHL. The efforts of Juanelle and Helen prove that's not true. Through word of mouth and very little local advertising, they were able to enlist the help of concerned friends and family to raise this money. If only 50 people put forth the same effort, then they would raise $50,000 for VHL research and education!

I hope that Juanelle and Helen will challenge and inspire others to help.

Call for Papers

The next **VHL Medical Symposium** will be held May 20-22, 2004 in Japan, hosted by Dr. Taro Shuin of the Department of Urology, Kochi University Medical School.

This meeting is designed as a forum for experts in VHL — clinicians and researchers — to share their latest findings and accelerate progress in management of VHL. The language of the Symposium will be English.

Health care professionals who would like to present their work on VHL at this meeting should submit abstracts to Dr. Shuin at shuint@kochi-ms.ac.jp

So far we know that there will be speakers and attendees from U.S., Canada, Australia, New Zealand, Denmark, France, Germany, and a number of Asian nations.

See www.vhl.org/conf2004 for abstract format requirements.

VBS Supports VHL

— from the Cumberland United Methodist Church newsletter, North Carolina

It's a big order for children to learn about von Hippel-Lindau (VHL) as they learn to reach out in care and compassion for others and let the Light of Christ shine through them. Then again, God works in amazing ways. At Vacation Bible School (VBS) this summer, the 80-100 participants who came together as students and volunteers for Cumberland United Methodist Church in Fayetteville, North Carolina, were learning about God's Light and they were learning about how they could help make a difference for families dealing with VHL by contributing to the VHL research fund.

They learned that "Ms. Audrey," one of their VBS leaders, battles this disease. They learned that VHL causes tumors and that no one has figured out a way to cure it. They learned that they can be part of the solution and make a real difference in the lives of people like their "Ms. Audrey." They learned that God gives us ways to let God's Light shine through us as we help each other and that the VHL Family Alliance is one organization that gives family support and education all over our country.

When the week of VBS finished at Cumberland United Methodist Church, children and their families had contributed $124.76 as the VBS Mission Project for 2003.

Ms. Audrey was with the VBS team from the planning and set up stages all the way through the last day that ended VBS with an Ice Cream Social. Over the course of preparation and VBS week, Ms. Audrey came to a new appreciation of the work that now goes into VBS events and a new love and enjoyment for her young friends in her church family. The VBS Mission Project makes its contribution in honor of Audrey Clifton, and extends its thanks to her and the VHL Family Alliance for the opportunity to learn and serve together in this important work.

Please Join us at a Regional meeting

Midwest: Indianapolis

October 25, 2003

The U.S. Midwestern Regional Meeting of the VHL Family Alliance will be held in Carmel, near Indianapolis, Indiana, October 25, 2003.

Hotel reservations can be made at the Comfort Suites, 15131 Thatcher Lane, Carmel, Indiana, 46033, by calling 317-575-0000. Ask for the VHL meeting rate of $69.99/night including breakfast.

Registration will be $25, which includes lunch.

Speakers will include

- Cindy Hunter, M.S., certified genetic counselor and coordinator of the Indiana University Familial Cancer Clinic; and
- Dr. Ramana S. Moorthy, Opthamologist, Associated Vitreoretinal & Uveitis Consultants, Indianapolis, Indiana.

For more information, call Kathy Braden at 317-894-3909 or Shari Lipp-Levine at 317-575-9073. Please call to let us know if you plan to attend.

Annual Meeting New York/LaGuardia

June 19, 2004

The **Annual Membership meeting** of the VHL Family Alliance will be held Saturday, June 19, 2004, 9 am - 5 pm, in conjunction with a Northeast Regional Meeting.

It will be held at Crowne Plaza/LaGuardia Airport, 104-04 Ditmars Blvd., East Elmhurst, NY 11369. Hotel space is available at that hotel for only $99/night. Be sure to say you are coming for the VHL meeting 718-457-6300

Presenters will include Dr. Gladys Glenn and Dr. Emily Chew from the U.S. National Institutes of Health. Dr. Glenn will speak on VHL Screening and treatment. Dr. Chew will talk about eye issues. Dr. Wendy Chung, Clinical Care Center coordinator at Columbia-Presbyterian Medical Center, will present information on DNA testing. All will be available to answer your questions.

The Program will include **VHL Connections** - an opportunity for us just plain folks to share experiences and stories.

Conference Registration is $25 per VHL patient or family member and $50 for genetic counselors, nurses, doctors, social workers, and other health care professionals.

CEUs will be available for genetic counselors.

Northeast: Boston

November 1, 2003

The Boston meeting will be held Saturday, November 1, 2003, 9 am to 5 pm, in the Auditorium of Lahey Clinic, 41 Mall Road, Burlington, MA. Registration of $25 per person includes lunch and breaks

Hotel space has been reserved for the VHL meeting at the Marriott Burlington. Attendees should make their own reservations by calling 1 (800) 228-9290 or (781) 229-6565. The VHL conference rate is $79 per night for up to four people in one room. CEU credits will be available for genetic counselors and nurses.

Presentations on Kidney, Neurology, research advances, and DNA testing will feature physicians and researchers from the three Clinical Care Centers in Boston.

VHL Connections in the afternoon is a time to share our stories, connect with others traveling the same road, and learn from one another.

It's a beautiful time to enjoy New England -- see you there! For more information call 617-277-5667.

Our Thanks for Donations from . . .

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Jenny & Brent Mathison
Sarah Simpson & Mark McClure
Tammy & Bill Menk
Joseph Monaco, with thanks for the newsletter
Mary E. Mulqueen
Dorothy Nelson

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Sally Pope Overman
Robyn & Mattison C. Painter
Violetta F. Patrick
Kim & Darrin Peach
Scott Rea
Willa Mae Rice
Cheryl & Rob Risser
Sunanda Rodrigues
Joyce Reed Rosenberg
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Dr. R. Neil Schimke
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Hyman & Marjorie Weinberg Foundation
Michael & Lisa Whitten
Harry & June Wilcox
Susan Winkler
Dr. Gary Wood
York Electric Motors

In Honor of . . .

Rosemary Bonacci's 26th birthday, by Joseph & Margaret Flaherty
Dr. Hilde Boehme, by Evelyn & Dr. Friedrich Port
Susan Burgoon, by Mary Swanston
Tony Capinegro, by Jean Mulkerin
Liza Christian, by Sandra & Jerry Parker
Cindy Clark, by Deanna & Robert Hebert, Mack Ed Swindle; Whitaker, Chalk, Swindle & Sawyer
Audrey Clifton, by Cumberland United Methodist Church
Alice Coday, by Stephanie Armacost, Albert Young
Irma Colen's birthday, by Joyce Reed Rosenberg
Lori Davids, by Louise Hennicutt
Damon Graff's birthday, by Joyce Graff
Joyce Graff, by Rob Batchelder, Susan & Don Leonard
Charles Hall Jr on his birthday, by Michael & Tondra Bolinger
Karen Johnson, by Virginia & John Fritz
Kathleen Kane, by Glen & Kathleen Helme
Robert & Gary Keesler, by Audrey & Thomas Keesler
Becky & Jean Lima, by Tony & Carva Berkompas
Our daughter-in-law, by James & Sheila Lunny
Wedding of Diane Garbowski & Michael Massi, by Halina Garbowski
Erich Moan, by Margrit Moan-Nachreiner
Rachael Morgenstern's birthday, by June & Harry Wilcox
Jenny Proby, Ireland, by her friend Julie Dunlop
The Roche Family, and the wedding of Kelley Roche and Ross Mackesey, by Nancy & Shane Bolan, Brenda Dandy, Janice Frederick, Jane & William Love, Susan & Jim Pinnione, Nancy & Nicholas Tsottles, Joan & Thomas Ward, Mary Ellen & William Whitty, Phyllis Schott, Carl Wright
Nancy Southern, by Luann & Mathew Mascorro
Evabeth Thomas, by Ray & Patricia Larson
Kelly Randall Thomas, by Priscilla Randall
June Wilcox's birthday, by Virginia Worthington

In Memory of . . .

Al Bean, by Evabeth Thomas
Mrs. Otillie Blumer, by Allen & Linda Stone
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Rosamary Buck, by Paul L. Buck
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Lynn Ehret's Mom, by Evabeth Thomas
Dr. Alexander Fedinec, by Vlasta Fedinec
Carolyn Flanagan, by Jennie Hedberg
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William, Betsy, Russ, Glenn, and Jerome Fuller, by Jocelyn Fuller
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Ruth Harper, by Susan A. Burgoon
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Linnie Elizabeth (Beth) Lesley, by Eileen Abbott, Rita Anderson, Donna Baumann, Dr. Carolyn Beard, Dulcie Couillard, Beth Delano, Frenchtown Deli, Corene Gieskieng, Joseph Green, Danna Hardt, Hazelwood Middle School, Genell Henderson, Historic Frenchtown Association, Jerry Jones, Josephine Kelly, Shirley Lawing, Leading Ladies, Sue Leslie, Beth O'Callaghan, Pete & Sandy Seadeek, Norma Stahlhuth, Paul & Wanda Vipond, Madeline Williams, Gladys Wyas
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Mary Lombard, by Katherine Conway
Mrs. Charles (Peggy) Lutz, mother of Pam Lutz, by Joseph Bowditch, Wilbert Leopold, Charles Lutz, Patricia Slaman
Pam Mayville, by Wilbert Leopold
William Minster, by Alice Coday
Gerald Osborn, by Georgia Osborn
John Rainier, by Ilene Senoff, Helen Guttman, Ellen F. Rainier
Bruce Reifman, by Eric & Bridget Cooper
Dennis Roche, by Lynne Hart
Dude & Ann Rottger, by Mary & Jim Rottger
Rob Wright, by Ellen & William Robert Wright

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All members receive the VHL Family Forum quarterly. Check here ❑ *if audio version is needed*

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❑ Please send me ____ brochures about VHL Research Fund to share with others

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Registration:

___ people for the Regional Meeting in _____ @ $25 each $ _____

Great Gifts!

___ *Sacred Process, by Karen Koenig* -- inspirational poems about life & death with VHL @15.00 $ _____

___ *A Time to Walk,* by Jay Platt -- Lessons learned along the Appalachian Trail @14.95 ea $ _____

___ *How Are You?* by Patricia Foote -- Managing Your Health @17.00 ea $ _____

___ *VHL Lap blanket, all-cotton woven thermal* throw @ $40 each (brown & tan) $ _____

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Canadians, please send donations to: Canadian VHL Family Alliance,
4227 Hamilton Road, Dorchester, Ontario, N0L 1G3 Canada

Recognizing Caregivers

— Gary Barg, Editor, caregiver.com

I had just finished my second speech in as many states in as many days. My favorite kind of week. I was talking to caregivers and signing books in the lobby when one of the program hosts approached the table and made the pronouncement, "oh my gosh, during your speech, it finally dawned on me that I'm a caregiver, too!"

I have to admit that at first this was a surprising announcement since her job entailed dealing with family caregivers on a daily basis. Intrigued, I asked her to elaborate. It turns out that she spent the past five years helping her dad care for her stepmother who was living with a myriad of chronic illnesses. She would take them to the grocery store and the doctors' offices, make midnight runs to the emergency room and stay on the phone trying to straighten out problems with the insurance company. To top it all off, she would spend her 40 hour work week supporting families in the area going through the exact same challenges as her own family.

This shouldn't have been such a surprise, as I do know that self-identification is one of the primary barriers to being able to support family caregivers. In fact, at the Question and Answer session in our conference the day before, many of the questions being pondered by the professional caregivers had to do with their own family issues.

This realization also brings to light something I've been talking about for quite some time that these professionals who spend their days helping family caregivers, often leave work and step into their own role as family caregiver. And I truly believe that for these dedicated souls who give qualified and compassionate advice to other families while being able to maintain their professional detachment, the walls crumble quickly when they get their own phone calls in the middle of the night. So, let's stop for a moment to celebrate those whose caring knows no bounds and remember that more often than not, the professional in the white coat is also probably worrying about his or her mom, too.

Volunteer of the Year

Cari was diagnosed with VHL less than two years ago. After contacting the VHL Family Alliance and learning about the ongoing research for VHL, she felt encouraged and hopeful. She realized the need for public awareness and began a vigorous effort to spread the word. She wrote to friends, family, neighbors and businesses to educate and raise funds for research and awareness of VHL. An all day activities event was held at her Dad's business with coverage by local newspapers and TV station. Her Dad's employees also held a car wash, all to bring VHL awareness to the public. Cari is a dedicated Volunteer giving generously of her time, energy and talents.

VHL FAMILY FORUM

Newsletter of the VHL Family Alliance
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