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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Natural History of Hemangioblastomas

reporting articles by J.E. Wanebo, R.R. Lonser, JR. J. Weil, H.L. DeVroom, G.M. Glenn, and E.H. Oldfield from the Surgical Neurology Branch, National Institute of Neurological Disorders and Stroke, U.S. National Institutes of Health, Bethesda, Maryland

For the past several years physicians and researchers at the U.S. National Institutes of Health (NIH), led by Dr. Edward H. Oldfield, have been reviewing their very large set of case histories of hemangioblastomas of VHL. The goal of their study is to better understand the natural history and growth pattern of the hemangioblastomas of the central nervous system (CNS) that are associated with von Hippel-Lindau (VHL) disease. They would like to be able to learn what characteristics of hemangioblastomas may indicate that they will soon develop symptoms and require treatment.

As better imaging techniques become available, we are finding tumors at much earlier stages. But the existence of a tumor does not necessarily mean that it needs treatment. We know that some tumors may stay at the same small size for 20 or 30 years, while others grow quickly. Is there a way to predict the future growth pattern of a particular tumor? Stereotactic radiosurgery, for example, gives us a tool that might be used to stunt the growth of a tumor. But once we have treated that small tumor, we really do not know whether it failed to grow because of the treatment, or whether it is one of the tumors that was not ready to grow anyway. How do we know which of the several tumors we should treat? We do not want to treat every tumor, because radiation adds up in the body. We need to use each tool wisely and sparingly and to the best advantage for the patient.

The authors review clinical histories and a series of MRI images for 160 patient over as many as 15 years. For each image they measured the volume of each tumor and any associated cysts. A total of 650 hemangioblastomas were identified in these 160 patients: in the cerebellum (250 tumors), in the brainstem (64 tumors, all of which were located in the posterior medulla oblongata), in the spinal cord (331 tumors, 96% of which were located in the posterior half of the spinal cord), and in the supratentorial brain (10 tumors). (See Figure 1.) They mapped the growth of each tumor over the course of the series of MRI's.

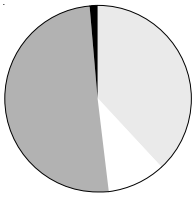

Figure 1:
Of 650 total hB there were
250 in the cerebellum
64 in the brainstem
331 in the spinal cord
10 supratentorial

As the patients progressed from no symptoms to increasingly severe symptoms and required surgery, the hemangioblastomas were seen to increase in size. In all cases, the symptoms were related to a mass effect, the effect of the pressure created by the volume of the tumor and / or the cyst. Twenty-one (72%) of 29 symptom-producing cerebellar tumors had an associated cyst, whereas only 28 (13%) of 221 nonsymptomatic cerebellar tumors had associated cysts, compared with only four (8%) of 52 nonsymptomatic brainstem lesions. (See Figure 2.)

Ninety-five percent of symptom-producing spinal hemangioblastomas had cysts, which are elongated and take the shape of a hotdog in the confined space of the spinal column and are called syrinx or syringomyelia. Symptoms were unpredictable. Among the 88 patients who had a series of MRIs over a period of 6 months or longer (an average of about 3 years), 164 (44%) of 373 hemangioblastomas and 37 (67%) of 55 tumor-associated cysts enlarged. None of the tumors or cysts got smaller. Symptomatic cerebellar and brainstem tumors grew at rates six and nine times

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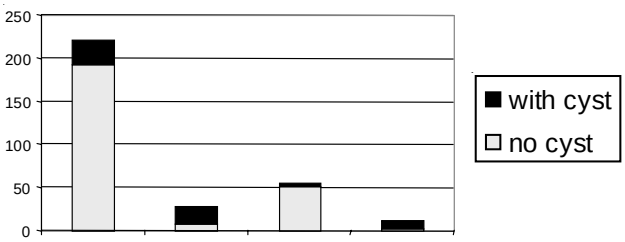

Figure 2: Influence of the cyst in causing symptoms

greater, respectively, than asymptomatic tumors in the same regions. Cysts enlarged seven (cerebellum) and 15 (brainstem) times faster than the hemangioblastomas causing them.

Hemangioblastomas frequently demonstrated a pattern of growth in which they would enlarge for a period of time (growth phase) and then stabilize in a period of arrested growth (quiescent phase). Of 69 patients with documented tumor growth, 18 (26%) had tumors with at least two such cycles. Of 160 patients with hemangioblastomas, 34 patients followed for an average of 4.25 years were found to have 115 new hemangioblastomas, and 15 patients had new cysts.

In conclusion, cysts were commonly associated with hemangioblastomas of the CNS. The pace of enlargement was faster for cysts than for hemangioblastomas. By the time symptoms appeared, the majority of symptoms were caused by mass-effect from the cyst, rather than from the tumor causing the cyst. These tumors often have multiple periods of tumor growth separated by periods of arrested growth, and many untreated tumors may remain the same size for several years. These characteristics must be considered when determining the optimal timing of screening for individual patients, and for evaluating the timing and results of treatment.

Implications for managing brainstem hemangioblastomas

There are no clear guidelines for optimal management of brainstem tumors. In an attempt to clarify some of the uncertainty about the operative treatment of these lesions and its outcome, the authors reviewed all cases of VHL in which resection of brainstem hemangioblastomas was performed at the NIH during a 10-year period.

Twelve patients with VHL, six male and six female, ranging in age from 15 to 46 years old, underwent a total of 13 operations to remove 17 brainstem hemangioblastomas. A series of examinations, hospital charts, MRI's, and operative records were reviewed. Clinical grades were assigned to each patient before and after surgery.

The best predictor of long-term outcome was the level of neurological function before the surgery. Patients who underwent CNS surgeries for hemangioblastomas were more likely to improve or to remain neurologically stable. Tumor or cyst size, the presence of a cyst, or the location of the tumor did not affect the outcome. No patient was neurologically worse after brainstem surgery. At long-term follow-up review only one patient had declined neurologically and this was due to the cumulative neurological effects caused by eight additional hemangioblastomas of the spinal cord and their surgical treatment.

The study concludes that hemangioblastomas can be removed safely. They generally should be resected when they become symptomatic, or when the tumor has reached a size such that further growth will increase the risks associated with surgery, or the presence of an enlarging cyst. MRI imaging is usually sufficient for preoperative evaluation, and presurgical embolization is not necessary. The goal of surgery is complete resection of the lesion before the patient experiences a disabling neurological deficit.

Implications for managing spinal cord hemangioblastomas

The authors set out to determine factors that predict which patients with spinal cord hemangioblastomas need surgery or what outcomes of this procedure should be expected. They reviewed a series of patients with VHL who underwent resection of spinal hemangioblastomas at one institution (NIH). Their goal is to identify features that might guide surgical management of these patients.

Forty-four patients with VHL (26 men and 18 women ranging in age from 20 to 58 years old) underwent 55 operations with resection of 86 spinal cord hemanbioblastomas. They were followed an average of four years following these surgeries. Patient examination, review of hospital charts, operative findings, and MRI imaging studies were used to analyze surgical management and its outcome. To evaluate the clinical course, clinical grades were assigned to patients before and after surgery. The best predictors of postoperative outcome were preoperative neurological status, tumor size, and tumor location. Patients with no or minimal preoperative neurological dysfunction, with lesions smaller than 500 mm3 (the size of a kernel of corn), and with dorsal lesions (on the back side of the spine) were more likely to have no or minimal neurological impairment. The syrinx (cyst) deflated when the tumor was removed, and was not influenced by whether the syrinx cavity was entered.

The study concludes that spinal cord hemangioblastomas can be safely removed in the majority of patients with VHL. Generally in these patients hemangioblastomas of the spinal cord should be removed when they produce symptoms or signs.

Anterior and posterior surgical approaches

Spinal hemangioblastomas arise mostly in the

posterior aspect (the back side) of the spinal cord and are often associated with an intraspinal cyst or syrinx. Rarely, the tumor develops in the anterior or ventral (belly) side of the spinal cord. Ventral spinal hemangioblastomas are a surgical challenge because of difficult access and because vessels feeding the tumor originate from the anterior spinal artery. The goal of the study was to clarify whether an anterior or posterior surgical approach is better for management of hemangioblastomas of the ventral spinal cord.

The authors reviewed cases of eight patients (two women and six men, ranging in age from 19 to 49) who underwent resection of ventral spinal hemangioblastomas (nine tumors: five cervical and four thoracic). Two surgical approaches were used to resect these tumors. A posterior approach was used for five patients; an anterior approach was selected to treat the remaining three patients. Immediately after surgery the ability to walk remained unchanged in patients in whom an anterior approach had been performed, but deteriorated significantly in patients in whom a posterior approach had been used, because of motor weakness (four of five patients) and/or loss of sensation in the muscles. While the patients in the posterior group improved significantly, there was still a significant difference between the two groups even six months after surgery. In all cases MR images showed that the tumors had been removed completely and five of the six cysts were gone.

The outcomes of these eight patients indicate that both immediate and long-term results are better when an anterior approach is selected for resection of hemangioblastomas of the anterior side of the spinal cord.

1. JE Wanebo, RR Lonser, GM Glenn, EH Oldfield, "The natural history of hemangioblastomas of the central nervous system in patients with von Hippel-Lindau disease." *J. Neurosurg,* 2003 Jan; 98(1):82-94. 2. RJ Weil, RR Lonser, HL DeVroom, JE Wanebo, EH Oldfield, "Surgical management of brainstem hemanbioblastomas in patients with von Hippel-Lindau disease." *J. Neurosurg* 2003 Jan; 98(1):95-105. 3. RR Lonser, RJ Weil, JE Wanebo, HL DeVroom, EH Oldfield, "Surgical management of spinal cord hemangioblastomas in patients with von Hippel-Lindau disease." *J. Neurosurg* 2003 Jan; 98(1): 106-16 4. R.M. Pluta, B. Iuliano, H.L. DeVroom, T. Nguyen, E.H. Oldfield, "Comparison of anterior and posterior surgical approaches in the treatment of ventral spinal hemanbioblastomas in patients with von Hippel-Lindau disease." *J. Neurosurg* 2003 Jan; 98(1):117-24.

ABTA Sharing Hope

The American Brain Tumor Association (ABTA) will hold their 6th biennial Sharing Hope Family Weekend on July 18-20, 2003, at the Lincolnshire Marriott Resort, Chicago. They are planning a special weekend of neuro-oncology expertise. To receive program and registration information visit our website at www.abta.org/events.htm, send an e-mail to info@abta.org or call ABTA at 800-886-2282.

New NSGC President

The National Society of Genetic Counselors (NSGC) elected Robin L. Bennett as President for 2003. Bennett is Senior Genetic Counselor and Clinic Manager for the University of Washington Medical Genetics Clinic in Seattle, and the coordinator of the VHL Clinical Care Center there.

As President of NSGC, Bennett is responsible for leading the association and serving as its chief spokesperson. In her first act as president, she has called for an "environmental scan" to examine how the profession is perceived by key individuals inside and outside of genetic counseling.

Bennett has been with the University of Washington since graduating from the Sarah Lawrence Human Genetics Program in 1984. She is certified in genetic counseling by the American Board of Medical Genetics and the American Board of Genetic Counseling. She has been a forerunner in the development of genetic counseling practice recommendations including developing criteria for pedigree nomenclature that are now the world standard. She has spoken and published extensively on topics related to her work in Huntington disease, consanguinity (when cousins have children together), neurogenetics, VHL, cancer genetics, inborn errors of metabolism, genetic family history, and ethical issues in genetic counseling and genetic testing. She has authored two books including the recent release, *The Practical Guide to the Genetic Family History*, published by John Wiley & Sons in 1999.

NSGC is a not-for-profit professional membership organization, consisting of more than 2,000 genetic counselors. Genetic counselors work as members of a health care team, providing information and support to families and individuals who have members with birth defects, genetic disorders, or may be at risk for inherited conditions.

Spanish Hotline

Hotline services in Spanish language are now open in the United States. People can dial 1-800-767-4VHL. The menu of choices, read in English and then in Spanish, direct speakers of English to press 1, and speakers of Spanish to press 2. There are options also to reach the Director of Volunteer Services in Mississippi (3), and the home office in Brookline, Massachusetts (4).

Please spread the word to speakers of Spanish that they can now get support in their native language.

We are grateful to hotline volunteers Alexandra & José M. of New Jersey for their service to the Spanish-speaking community.

Pioneering for Hope

by Tammy N., Mississippi

It was two years ago that I first began talking with Dr. Kaelin about possible participation in a clinical trial. I am forty-one years old. I have had six brain surgeries and one on my cervical spine. My right arm is partially paralyzed -- it moves, but I have no clear idea of where it is, and I can't trust its grip. My pancreas is completely displaced with cysts, causing me serious digestive issues. And of course I have a number of small brain and spinal tumors, and a couple of kidney lesions . . .

The most worrisome tumor is one the size of a grape in the ventricle area of my brain. It does not cause symptoms, and it has room enough to grow to the size of a pecan before it causes me any problems, but it's in the thinking area of the brain so I really would like it not to grow at all. I would really love to avoid any more surgery. I'd love to have a drug that would keep these tumors from growing. Shrinkage would be ideal, but I'd settle for keeping them just the way they are. But even if it didn't help me -- even if I could only help them learn the pros and cons of a new drug and develop a better one for my daughter and other young people coming along -- I would be glad to have contributed to our knowledge and hopefully helped to lay the groundwork for the right drug in the future.

It was in that spirit that I approached Dr. Kaelin and began the process of enrolling in his clinical trial. We began the interviewing process by phone and fax. And in the midst of it all the drug company cancelled the trial. They had discovered some potential long-term negative effects in their testing on dogs that they did not like, so they "pulled the plug" on the trial.

The sense of devastation was intense. It felt like shattered hope. But then in discussing it with friends I realized that they had done the right thing. As great as my disappointment was, how much worse would it have been to take a drug and have some awful side effect that worsened my health? I had to be grateful that they had the honesty to admit up front that this drug was not yet right. Dr. Kaelin assured me that there were other drugs "in the pipeline" -- it was as if they did not yet have quite the right recipe, but they were still working on a number of similar drugs, and were using this same technology to check for long-term effects before they got to human trials. It was disappointing, but in another sense also comforting.

In January 2003 Dr. Dan George of Dana-Farber Cancer Institute, working in collaboration with Dr. Kaelin, called me to say that the drug was now ready to go, and they were carefully approaching clinical trials. Additional studies suggested that the side-effects seen with long term administration in some animal species was unlikely to translate into humans. Phase I trials have gone well, and Phase II trials with prostate and kidney cancer are encouraging.

He asked if I would go through the evaluation process, to see if I would qualify. They had to make sure that any issues that needed surgery were taken care of first, and that I met all the other criteria. He needed copies of records and scans, family history, hours of detailed interviews by phone. In addition, I would need to go to Boston for two and a half weeks of onsite tests. If I qualified, they would send me home with the pills, but I would need to come to Boston once a month for a year.

It was a big commitment of time and money too. I talked with my family and my pastor, and prayed about it. I also spoke with my neurosurgeon. Dr. Sanford, a pediatric neurosurgeon, has done every one of my brain and spinal surgeries since I was nineteen. He read over the protocol and said he thought it was a good idea. If this drug could postpone another surgery, or keep me from needing it at all, it would be a blessing. He immediately wrote a letter to Dr. George, recommending me for the protocol and offering his cooperation.

My daughter clinched it. "What if this drug helps you to feel better and never have any more surgeries and live to be a great grandmother?"

With the backing of my family, my surgeon, and my friends, I began to tackle the logistics. Through the VHL Family Alliance I got the name of Mercy Airlift (see following article). They asked if this transport was needed for a life-threatening condition. We said yes. They organized free airplane tickets for myself and my father to travel to Boston for the evaluation. Mercy will help once every six months, and Southwest has low-priced fares to Hartford or Providence. My grandmother lives in Hartford, Connecticut, and we thought we could commute from there. VHLFA volunteers in Boston offered housing if we wanted it. So off we went to the big city.

We arrived February 6, and were greeted by a snowstorm. Commuting 100 miles from Mississippi to Memphis is one thing. Commuting 100 miles from Hartford to Boston in February is another thing altogether. We commuted from Hartford for three days and were totally exhausted before we accepted the offer of Boston-based housing during the week, and drove back to Hartford for the weekends. February broke snowfall records in Boston -- more than 30 inches of snow fell during my visit.

It was quite an adventure at the hospitals too.

There are five hospitals in the Harvard Cancer Center -- Dana Farber Cancer Research Institute, Joslin Diabetes Center, Brigham & Women's Hospital, Beth Israel Deaconess Hospital, and Massachusetts General Hospital (MGH). Each one is doing the detail work on one part of the research. I had tests at each. Four of the five are in the same part of town, just blocks apart. The MGH testing was done in Charlestown, not far from the U.S.S. Constitution, the oldest ship in the U.S. Navy, launched in 1812.

The testing was similar to the routine I went through at NIH. They did thorough tests on every part of my body, making sure they had a detailed record of every bit of VHL involvement. Most tests were the standard VHL clinical tests, but when they came to a tumor they intend to track for the study, they switched to the "research routine" which is a much more painstaking analysis of the tumor, its size and characteristics, and in particular the blood flow to and from the tumor. This meant longer MRIs. For the spinal MRIs they had a television mounted inside the tube showing a movie with subtitles to help pass the time.

I guess the most important thing it takes to be part of a clinical trial is patience. You have to be flexible, be on call, and roll with changes in the scheduling. It's a trial, after all, and I was the first patient in the trial, so they were learning as they went along. One day they called and asked me to come a day earlier than planned, another day they asked me to stay home and come the following day. I kept my senses of humor, curiosity, and adventure too, and chatted with the staff or whatever other patients were nearby. I met some lovely people. I learned a lot too. The radiologist went over the results with me immediately following the scans, and I got to watch the G.I. test on the monitor. Everyone was very nice and clearly highly skilled. Everyone from the nurses to the maintenance people assured me I was in "the best place." On Valentine's Day I found a card my husband had tucked into my suitcase, full of love and hope.

Half-way through the testing, they called to say we had hit a snag. My metanephrines were 291. According to the protocol, participants' metanephrines must be no higher than 290. I sat out the day waiting for them to get clearance from the drug company to proceed at this level. It seemed an insignificant deviation to me, but in order for the data from the study to be meaningful, they have to follow the protocol exactly. They did get approval, but it was an anxious day for me. To come all this way and be ruled out for one stinking point! But then it was resolved and we proceeded.

So at the end of two weeks of testing, I am officially enrolled in the study! I start the drug on Monday, have MRIs Tuesday and Wednesday, and then go home with the pills. Five pills a day, stay away from grapefruit, and come back in a month. They want my local doctors to do some blood tests in between and fax them the numbers, and they'll see me in March. It will take a full year to get the final results from this trial, and meanwhile there are other trials going on with other drugs. It seems a long time since our whole family sat with Dr. Zbar's researcher the 1990, drawing blood from everyone in the room to try to locate the genetic cause of VHL. But at last we are seeing some potential therapies emerge from all the research! I hope to have some very good news to share next year!

Dr. George is accepting VHL patients who have asymptomatic CNS hemangioblastomas (hB) and have had at least one prior neurosurgical procedure and do not currently require surgical intervention, OR have symptomatic CNS hB which can not be successfully treated with standard therapy, OR have retinal hB causing impairment in visual function (visual acuity, contrast sensitivity, visual field, or color vision), which cannot be successfully treated with standard therapy. If you are interested in exploring this further, please call Judy or Stephanie at 617-632-5068. Eligibility criteria are listed at
http://clinicaltrials.gov/ct/gui/show/NCT00052013?order=1

Patient Travel & Lodging

by Altheada Johnson, New York

There are lots of good people in the world. One of them is Edward R. Boyer, President and founder of the national charity ***Mercy Medical Airlift*** (MMA). The goal of MMA is to ensure access for all to distant specialized evaluation, diagnosis or treatment within the United States. Financial hardship should no longer deny access. MMA provides 70% of all charitable medical air transportation in the U.S. www.mercymedical.org

Modes of Medical Air Transportation include: Air Ambulance, Airline Travel (up to three weeks notice needed), and Angel Flight Travel (usually five days advanced notice needed.) A family member can accompany the patient.

To get more information about travel assistance call the National Patient Travel Helpline 1-800-296-1217, visit www.patienttravel.org or e-mail mercymedical@erols.com Advice and referral are available based on the specifics of the patient's need.

The ***National Association of Homes that Help and Heal*** (NAHHH) was established in 1986 to provide family-centered lodging and support services to families and their loved ones who are receiving medical treatment far from home. The NAHHH promotes and assists not-for-profit programs to provide lodging at little charge or no fee for a stay near treatment. There are 500 homes in the U.S. that are a part of this program, including Ronald McDonald House, Hope Lodge sponsored by the American Cancer Society, Fisher Houses on military bases, and Children's Inn at the NIH.

The Hospitality House Helpline 1-800-542-9730 or visit www.nahhh.org

The Gift of Friendship

by Jan C., Illinois

In 1997 my son Tony, then 16, was diagnosed with VHL. We were blessed to be in the care of a brilliant neurosurgeon, Dr. Tanadori Tomita, at Chicago Children's Memorial Hospital. Dr. Tomita removed two tumors from the cerebellum and one from the cervical spine. This was the first case of VHL at Children's to their knowledge, and we will be forever grateful to Dr. Tomita for diagnosing what we understand to be a very often misdiagnosed disease. This is also where we had the fortune to be under the care of Dr. Stewart Goldman, an outstanding pediatric oncologist whose sensitivity helped my son through a very difficult diagnosis. His knowledge, sense of humor and dedication to patient care and passion for helping families are unmatched.

VHL was a frightening unknown disease to all of us -- myself, my husband Scott, my daughter Gina (then 18), and most of all of course to Tony. While he was in intensive care, a colleague and friend of mine looked up von Hippel-Lindau on the internet and printed out invaluable information from the website www.vhl.org. Even since then the VHLFA has provided our family with information, knowledge, the questions to ask and the people to call. Before we even left the hospital we all had genetic testing which resulted in finding out that this was not an inherited gene in our family, but that Tony's gene "zigged when it should have zagged," to quote the geneticist. Tony is the first in our family to have VHL.

We have been through the roller-coaster that this type of disease presents, but all along the way we have been truly blessed by incredibly supportive friends, family and clergy. Through this support Tony and our family have been carried through another surgery this December, removing two more tumors in the cerebellum. This time, looking for a physician that Tony could continue with into adulthood, we went to the University of Chicago on the recommendation of friends and VHLFA. They had followed and had a working knowledge of several VHL cases.

Tony, now a sophomore at a university in Ohio, came home for Christmas break, had surgery, and returned in time for the spring semester. He has always been very pragmatic about his health, dealing with it when he has to, and enjoying everything he can in between. He tells us he takes nothing for granted and it bothers him when others do. His friends have been a terrific support system for him, and he has also been overwhelmed by the generosity of others.

I have struggled with how I might be able to make a difference in our fight against VHL and had spoken with my friend Nonie about fundraising. She spoke with another friend of ours who has extensive experience in fundraising, and she said that it's key to connect fundraising to a *celebration*. Our community was also rallying for funds and team participation for the American Cancer Society's Relay for Life. So we put our efforts there, feeling we were doing something to fight cancer.

At the same time, unbeknownst to me, friends were planning another event -- a 25th Wedding Anniversary surprise party. Our friends, knowing that we wanted to do something for VHL, turned this *celebration* into a small fundraiser. They knew that we would much rather have donations for VHL over anniversary gifts!

This was all so enlightening to me -- and that is why I am writing to you. We can all make a difference in the fight against VHL by raising research funds that go directly to our cause. We have so many reasons to celebrate life and to gather with friends and family. I am challenging myself to put on my thinking cap and get creative with Celebrations for VHL. Is it a golf outing, a day at the races, a picnic -- the possibilities are endless! Together we can make a real difference.

Our Relay for Life team was pleased to have raised almost $7,000. And our Anniversary team blessed us with a donation to VHL of over $2,700. Both of these efforts were very grass-roots, simple yet making an impact.

Again, our sincere thanks to VHLFA for everything you do for families.

Nominations Open

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

Nominations are now open for new members of the Board of Directors. If you would like to nominate yourself or another person, please send your nomination to Maria Shipton, Chairman of the Board, vhlpa@aol.com. It should include the nominee's name and address, work history including volunteer leadership positions, and the experience and talents the nominee would bring to the board.

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

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

June Meeting in Nashville

Please plan to join us at our Regional Meeting June 21 in Nashville, Tennessee. Nashville is the first in a new series of four regional meetings a year.

The VHL Family Alliance is always working to provide better support for our community. This year we are trying a new model for conferences in the United States. Instead of a single large two-day conference, we are dividing our meeting planning into three tracks:

For families: four regional meetings a year, designed for families. Healthcare professionals are welcome to attend.

For healthcare professionals: online continuing medical education events.

For physicians and researchers: Medical Symposia will continue as in the past, every two years, with the 2004 meeting planned for Kochi, outside Tokyo, Japan, May 20-22, 2004.

With more meetings closer to home, we hope that more of our members will be able to attend the Regional meetings. You will have an opportunity to meet other individuals and families affected by VHL, to share your stories, and learn together how best to manage your health.

The first Regional meeting will be held Saturday, June 21, 2003, in Nashville, Tennessee. The Annual business meeting of the Alliance will be held briefly at lunch, and new board members will be elected at that time. A second Regional Meeting is being planned in Boston in the fall, and other meetings will be announced as plans mature.

Featured speakers at the Nashville meeting include:

Dr. Gladys M. Glenn, Medical Officer, Genetic Epidemiology Branch, National Cancer Institute. Dr. Glenn has been a clinical investigator at the National Cancer Institute since 1984. She holds concurrent appointments in Urologic Oncology, Genetic Epidemiology, and Genetics. She received a B.A. in Chemistry from Cheyney University in Pennsylvania, and completed residency and post-graduate training at Thomas Jefferson University Hospital in Philadelphia and Johns Hopkins Oncology Center in Baltimore. Since the beginning of the NCI Clinical Research Program for VHL and other forms of familial kidney cancer in 1988, Dr. Glenn has served as the primary physician for clinical evaluations, diagnoses, treatment recommendations, referrals and genetic analyses and counseling of more than 700 at-risk and affected family members.

Dr. Peter L. Choyke, Chief of MRI, Diagnostic Radiology Branch, Clinical Center, National Institutes of Health. Dr. Choyke trained at Yale-New Haven Hospital in Diagnostic Radiology and completed a fellowship in cross-sectional imaging at the University of Pennsylvania. He joined NIH in 1987 where he has worked closely with the Urologic Oncology Branch under the direction of Dr. W. Marston Linehan. Dr. Choyke has a special interest in the imaging of von Hippel-Lindau disease along with other hereditary conditions that affect the kidneys. He is especially interested in the abdominal manifestations of VHL and the impact imaging studies have on organ-preserving treatments.

Dr. Lewis Blevins, Founder and Director of the Pituitary Center at Vanderbilt University in Nashville. Dr. Blevins received a B.S. in Chemistry from East Tennessee State University in Johnson City in December 1982. He attended East Tennessee State University College of Medicine and received his M.D. degree in May 1987. Dr. Blevins was Assistant Professor of Endocrinology and Metabolism at the Emory University School of Medicine and is a member of the Endocrine Society. He has worked with VHL in the adrenals both at Emory and at Vanderbilt. In 1997, he received the Golden Apple award for excellence in teaching.

We are making every effort to keep costs down for these Regional meetings. The modest registration fee of $25 covers lunch and breaks on Saturday.

Information is power, especially in living with VHL. The information you will gather from these presenters will help you maintain your health. The experience of hearing others tell their VHL story and learning from each other is not only touching, but empowering. Through these regional meetings you will make VHL Connections with other families, and with experts in the field.

Hotel space has been reserved at the Residence Inn by Marriott, 206 Ward Circle, Brentwood, Tennessee, 1-615-371-0100. Rooms must be booked by May 30 to be eligible for the special conference rate of $99 per suite for up to four people. Attendees are eligible for this rate Friday and Saturday nights. If you would like to stay and enjoy Nashville, they will accommodate you on Sunday night for only $75.

A welcome reception with cash bar will be held Friday evening in the courtyard of the Marriott. The meetings will be held all day on Saturday, June 21, in the Fellowship Hall at St. Paul's Episcopal Church, 510 West Main Street, Franklin, Tennessee.

-- Peggy Marshall, Director of Volunteer Services, and Kathy Braden, Director of Chapters

Y'All Come, now, y'hear?

Do I Want to Know?

by Ken C., Indiana, age 14

Several years ago, my dad's mom had to have an eye removed because of a tumor. She later found out that she had several cysts on her spinal cord and even more tumors in her kidneys. She died a couple of years after giving birth to my dad. She had no idea that she had a rare inherited disease. Unfortunately, my dad inherited the disease and there was no way of knowing, until recently, if I had inherited it also.

The disease that my dad could pass on to his children is called von Hippel-Lindau, or VHL. VHL is an autosomal dominant disease, which means, each child has a 50-50 chance of inheriting it from a parent. VHL can cause cysts and / or tumors to form in the eyes, brain, spine, kidneys, pancreas, endolymphatic sac and adrenal glands. There is currently no cure for VHL, so the best way to fight this disease is to closely monitor all of the areas that it can affect. My parents sent me for testing on an annual basis, and as I got older they wanted me to add an MRI of the brain and spine and a CAT scan of the abdomen, along with blood and urine tests. These tests can be very costly when done every year or two for your entire life, not to mention stressful. An alternative to the yearly monitoring is to have a DNA test to determine if you have inherited the mutated gene. If not, there is no need for monitoring.

In order to have the DNA test done, my dad had to have the test completed first. My dad had a blood sample taken and it was sent to the laboratory at the University of Pennsylvania. Once they found the mutated gene, I could submit a sample to the same lab and they could compare my DNA to his. They would look at the same place on the DNA where they found his mutated gene and see if my DNA also contained the mutation.

The decision to have the DNA test was a difficult one. I do not like to have my blood drawn, so I really did not want to have a sample drawn. I had heard on television about DNA tests being done from a cheek swab and had my parents find out if it was possible to have a sample taken that way.

After asking many people, my parents found out that the doctors could still read the DNA, and tell if I had the disease, from a cheek swab. (See also following article.) Once we found this out, I had to decide if I really wanted to know if I had the disease or not. I debated in my head if I wanted to know, or if I wanted to continue having the screenings done every year. I also had to consider how I would feel if I found out that I didn't have VHL, but my sister did; or how I would feel if I did have it and my sister didn't. After awhile, I decided to have the test done. I had my parents schedule an appointment.

Early one January morning, my mother drove me to the Indiana University (I.U.) Department of Genetics downtown. This is where I met with the genetic counselor, Cindy Hunter. I had to meet with the counselor so she could explain what VHL is, how it is inherited, how they would obtain the sample, how they would test the sample, and how the results would be returned. She asked me if I had any questions about VHL, the test method, or what the test results would say. Once I had all my questions answered, she called the geneticist, Dr. Gail Vance.

When Dr. Vance arrived, she gave me a more in-depth explanation of the test. She also informed me of what utensils would be used to extract the tissue sample. She showed me an example of the swab that would be used. She explained that she would send the swab to a laboratory in Pennsylvania that would perform the test on the tissue sample to find out if I had the disease. When she finished, she called my mother into the room and told us both that we would need to come back at a later date to have the test performed. Dr. Vance explained that they prefer to have the patient return after they have a chance to think about the information that they have been given. Sometimes people will decide not to return to have the test done.

After a few months, we went back to the I.U. Department of Genetics to have the test done. Cindy Hunter called me back to the exam room and asked if I had any questions before we did anything. I did not have any questions, so she opened the test kit. She handed me the swab and told me to scrape the side of my cheek with the brush-like end of the swab. Cindy reminded me that I really needed to scrape hard or there would not be enough tissue for the doctors in Pennsylvania to get the results. I scraped my cheek as hard as I could with the swab, without breaking it. After I was done scraping, I gave the swab back to her. She put it in a tube and told me she would mail it to the lab. Once the kit was sealed, she called my mother in and told us that the results should be returned to them in about two to three weeks.

About three months after the sample was mailed, we finally got a call from the genetic counselor saying that the results were in. My mother drove me downtown one more time. Cindy was waiting on us and she called both of us into the room. The doctor and the counselor both were in the room. They asked us if we had any questions and asked if I still wanted to hear the results. I was pretty anxious, so all I wanted

to hear was whether I had the disease or not. Since neither of us had any questions, the doctor got out the paper and started to read. The paper stated that the laboratory did not find the VHL genetic mutation in the sample that was sent to the lab. In other words, it said the result was negative.

Knowing that I do not have VHL is a huge relief. Going to the genetic counselor and having the test done have really taught me a lot. It helped me understand the disease that my dad has a lot more than anything else before. It also helped me understand just a little bit better what my dad goes through every time he has to wait for the results on a CAT scan or an MRI. It helped me the most though in that it made me realize that I'm really happy I don't have to deal with VHL. I'm very pleased that I decided to have the DNA testing done.

Cheek Swab DNA Test

by Catherine Stolle, Ph.D., DNA Testing Laboratory, Children's Hospital of Philadelphia

Cheek brush samples can not be used for all mutations. They are fine for single base (point) mutations detected by DNA sequence analysis, but are not useful for partial or complete deletions that are detected by Southern blot analysis. Cheek brush samples do not yield enough DNA, nor do they yield the large pieces of DNA needed to perform Southern blot analysis. In the future, we may be implementing a PCR based assay for deletions (much like that reported by the French group at the meeting in Padua) which will get around our current need for large amounts of big DNA fragments.

Meanwhile, it is safe to assume that the first analysis in a family will require bloods drawn, and can not be done with only a cheek swab. Once that first mutation has been identified, depending on the nature of the mutation found, cheek swabs may be sufficient for subsequent tests in a family. Please inquire to determine what is required for your family.

A 'Normal" in Genetics Test can bring new problems

By Carey Goldberg, The Boston Globe, *17 February 2003*

Samantha Sylvia Powers had steeled herself well for the test results.

She had watched her mother, grandmother, and sister battle breast cancer, and she knew she had a 50 percent chance of carrying the same vicious gene mutation. And she had already faced the possible implications: no more children, prophylactic removal of her ovaries and breasts, lifelong medication, and extra screening.

"I'm really ready to fight," she said just before entering the small office of Kristin Baker Niendorf, a genetic counselor at Massachusetts General Hospital, to get the news.

There was just one thing she hadn't figured on: She tested normal, no added risk.

"Initially, I was so excited," Powers said afterward. But then, within moments, "It was weird -- I just started to cry, and I felt so bad for my sister, thinking, 'I have everything ahead of me, and she's still fighting.'" In all her preparation, she said, "I just didn't count on that. It was an unexpected emotion."

As more at-risk Americans take advantage of a growing array of new tests to check their genes for disease proclivities, many are finding out that testing normal can carry a surprising downside: guilt, sorrow, family problems, even identity crisis.

Just 20 years ago, there were no genetic tests for adult-onset diseases, and members of families riddled with cancer or neurological disorders had no way to find out if they were in line to be next. Today, there are more than 50 tests for various kinds of cancer alone, and the company that makes the tests for two breast cancer genes, BRCA1 and BRCA2, checks more than 10,000 women a year. As researchers link more genes to diseases, such tests are expected to multiply further.

Kathy Schneider, former president of the National Society of Genetic Counselors, predicts that in the next five years, the number of adults choosing to take genetic tests will rise into the hundreds of thousands.

The rise in genetic testing has spurred the growth of a new kind of professional, the genetic counselor, to help patients decide whether to get tested, and to cope with the results once they arrive.

One might expect that the easiest part of their job would be delivering the good news, but in fact, in their accumulating experience, it is not so simple.

Research suggests that "there are just as many people that have problems with the good news as with the bad news," said Robin Bennett, president of the National Society of Genetic Counselors.

"One of the major problems is that it doesn't take away the disease in your family," she said. "It's still there and still affecting your parent or sister, or niece or nephew, or your future relatives who might be born.

"One patient of mine said, 'I feel like I'm standing outside a burning house, and my whole family's inside and I'm outside,'" Bennett said.

Good news can even bring family alienation, said Schneider, who is also a senior genetic counselor at Dana-Farber Cancer Institute in Boston.

"One woman described it very eloquently: She was no longer in the club," she said.

The woman, who had tested negative for a gene mutation associated with von Hippel-Lindau, belonged to a family that heavily addressed cancer issues, and she no longer had the same risks or same worries about her children, Schneider said.

Another problem that can arise, said Nathalie McIntosh, codirector of the genetic counseling program at Brandeis University in Waltham, is the jarring destruction of long-held assumptions.

"If you feel you're very much at high risk, and you organized your life around that premise, and then maybe later in life you've discovered that premise was wrong, that can be devastating," she said.

She said she has seen that mainly in Huntington's disease, a neurological disorder that usually hits in middle age and progresses inexorably to mental impairment and death.

"People decided not to marry or not to have children, and then they have testing and it's negative, and it's, 'What did I do with my life? I lived it the wrong way!'" she said. Many worry about not having saved for retirement, she added.

For some people, the seemingly heartening discovery that they are free of a dangerous gene can prove strangely disheartening. Counselors compared it to the disappointment experienced by some dieters who expect their successful weight loss to change everything wrong with their lives.

"For people who've lived under this cloud for a long time, suddenly they don't have that cloud anymore, and they expect everything to be perfect, and of course life is not," Schneider said.

Counselors also described patients going through a disconcerting readjustment of their identity, because many had seen their high risk as a defining aspect of their selves. The authors of a study published in 2000 in the journal *Research in Nursing and Health* on this topic even titled it "Redefinition," referring to how participants had to redefine themselves.

They followed 10 people who came from families with a history of Huntington's disease but had tested normal themselves. They found "paradoxical emotions" among the 10 in the six months following their tests, and a prevailing sense of the bittersweet. Some had trouble even believing that the results were correct, and others seemed to have difficulty reimagining the lives that they had always expected to end prematurely.

By the six-month mark, one of its authors, Debra L. Schutte, said, "We didn't feel like the process was done yet." The people were still working through their new visions of their lives, she said.

A handful of other articles have also looked at the effect of normal results on genetic tests, but as a research topic, it has barely been scratched.

"There is a need for us to understand what the impact of genetic testing is, in general, and in all types of groups and diseases and populations," Schutte said.

The popularity of gene testing will probably grow further if, as expected, companies begin direct-marketing tests to consumers.

Schneider and other genetic counselors emphasized that counseling can help people who choose genetic testing to prepare for the possible emotional consequences of their results, good or bad.

"It's important for people to recognize the emotional land mines that might be there," she said, "but also, they're not going to be relevant for everybody," so counseling must be highly tailored to the individual. Counselors will often refer a patient for therapy or to others who have been through similar processes.

Samantha Sylvia Powers said that her genetic counselor, Niendorf, had helped her with open empathy and by telling her, "This is not uncommon to feel this way." She was "very comforting and kind of let me cry."

And though Powers, 33, may have reacted with ambivalence, her 38-year-old sister with cancer, Pamela Sylvia, did not. She has undergone a hysterectomy, double mastectomy, chemotherapy, and radiation.

Powers's sister told her: "Don't look at it like that at all.

"This is how it went for me," she told her, "and if someone has to have cancer, I'm just glad that it's not you."

Canada meeting postponed

The Canadian meeting, previously announced for Ontario, May 2003, has been cancelled. Efforts are under way to plan a more modest regional meeting late in 2003. Volunteers are needed to assist with planning and organizing the meeting. Please contact Jill or Sue at canada@vhl.org or 519-268-1567 to offer your help.

Health Disparities

A conference on Health Disparities and Disability among African Americans was held at the Harvard School of Public Health (HSPH) in Boston February 5, 2003, co-sponsored by HSPH and the Massachusetts Department of Public Health (MDPH). The lecture honored the memory of Dr. William A. Hinton, one of the first African American graduates of Harvard Medical School (1912), who founded one of the first schools for laboratory medical techniques. During the 1920's he developed and perfected the Hinton test for the diagnosis of syphilis used throughout the world for over four decades. In 1959 Dr. Hinton's estate began funding scholarships for graduate students in memory of his parents who "although born into slavery and without formal education, nevertheless recognized not only the highest ideals in their personal conduct, but also the true democratic principle of equality for all."

Presenters described the differences in health care among various ethnicities in the United States. Part of the problem is economic, and another part is in the attitudes of the health care system. Dalene Basden of the Parent Professional Advocacy League (PAL) is the Parent Support Coordinator of the City of Lynn. As the mother of two sons with special needs, she has seen first hand the differences in treatment received by her own two African American sons versus others with the same disease.

Ethel Briggs, Executive Director of the National Council on Disability, shared a number of statistics: 12% of Americans are African American. The life expectancy of African Americans is 6 years shorter than for white Americans. By age 50 African Americans have a higher rate of chronic disease. 30% are uninsured (versus 20% of whites). 50% have job-based insurance (versus 70% of whites). 28% have no regular doctor (versus 19% of whites). 22% have no choice of where to go for health care and are less likely to receive preventive care. Rates of heart disease, cancer, and diabetes are all higher among African Americans. The rate of prostate cancer is three times other men. The rate of HIV is six times the rate among whites, and two times the rate among Hispanics. The rate of HIV among child-rearing women is 15 times that of whites. These conditions tend to be identified later, with more morbid consequences.

In Washington, D.C., alone, black mothers have the highest rate of death in childbirth, mostly due to the lack of prenatal care and distrust of the health care system. One effort being tried there is to provide free transportation to appointments, and to give women points when they keep their appointments, which they can cash in for cribs and baby supplies.

The rate of disability among African Americans is also disproportionately high. 17.8% of people with disabilities are African American. 19% are in special education classes. The school drop-out rate is 13.6% in the general population. Among people with disabilities, the drop-out rate is 28%. Among African-Americans with disabilities, the drop-out rate is 44%. 72% of African Americans with disabilities are unemployed. They are less likely to receive rehabilitation services, job training, financial aid, private funding, or health care services. Surveys reveal that African Americans are less likely to receive aggressive treatment or therapy.

Lisa Sinclair, MPH, is a Health Science Policy Analyst with the Center for Disease Control (CDC) in Atlanta. Over the past nine years she has been involved in developing the "Healthy People 2010" documents and was instrumental in creating a chapter on Disability and Secondary Conditions.

Ms. Sinclair described her analysis of the need for public health interventions targeting minorities with disabilities. The highest rate of disability is among Native Americans (20%), followed by Blacks (14%), Whites (12%), Hispanics (10%), and Asian / Pacific Islanders (7%). Among people with disabilities in general, 82.4% are white, 13.4% are Black, 2.1% are Native Americans, and 1.1% are Asian / Pacific Islanders.

People with disabilities are often very healthy, but they live with limitations, impairments, and / or chronic disease. Among ethnic communities, chronic conditions tend to emerge earlier. Among children with Down's syndrome, for example, the life expectancy for a black child with Downs is 25 years, compared to 50 years for a white child with Downs. The difference is reflective of the frequency of aggressive treatment for the morbid heart defects of these children.

Health differences are influenced by the majority behavior of the society, the institution, the individual, and the health behaviors and resources of the minority itself. For example, traditional values and beliefs among Native Americans preclude vocational rehabilitation (Mille and Joe, 1993). The other five key factors found to influence a person's ability to find the necessary health resources needed are the level of education achieved, whether people are employed, whether they have health insurance, whether they use a computer, and whether they are married or have a close partner.

The NIH and the CDC are working to address these various factors for all Americans, and in particular to identify and resolve the barriers of language, culture, economics, transportation, and literacy that make it particularly difficult for some minorities to obtain the preventive health care and rehabilitation they need to maximize health and minimize disability.

Crucial Conversations in Hospitals

”Research has made it clear that many of the 90,000 fatal mistakes made each year in U.S. hospitals are the result of staff inability to candidly and directly challenge physicians,” says Joseph Grenny, co-author of the *New York Times* bestselling book *Crucial Conversations: Tools for Talking when Stakes are High.* “We were contacted by one hospital after a woman had her foot amputated when she was admitted for a tonsillectomy. Hospitals call this a ‘wrong-site surgery.’ We call it an unforgivable mistake” because any of seven people could have averted that disaster by doing one simple thing: speaking up.”

Grenny explains that, “Hospital staffers are so intimidated by physicians that they often avoid speaking up even when they notice things that look risky. We will never reach our potential for high quality patient care until we have the full engagement and insight of the entire care giving team. And that means all have to be willing and able to speak up at any time.”

The Joint Commission on Accreditation of Healthcare Organizations (JCAHO), that gives safety certification to 80% of American health facilities, seems to agree. In January 2004, the JCAHO standards will include a mandatory provision to “Improve the effectiveness of communication among caregivers.” It turns out, if you want a healthy “outcome” from your hospital visit, you better choose a provider that trains and educates its employees in modern and effective communication.

Grenny adds, “We have known for years that the ability to handle crucial conversations well, determines an organization’s performance, efficiency and the quality of its products. Now, we have hard evidence these same skills may make the difference between being healed and being seriously hurt, when you visit your local hospital. We’ve seen a direct correlation between staff candor about patient-related issues with physicians and almost every measure of hospital performance “from quality, to profitability, to nursing turnover.”

“Commit to a culture that supports analysis of why an error has occurred and that rewards such behavior,” is the recommendation from JCAHO. Peggy Troy, the current president of LeBonheur Children’s Medical Center in Memphis, Tennessee, couldn’t agree more. “I have learned that great health care can only be delivered if the caregiver is a skilled communicator as well as a skilled healer. At the heart of our ability to heal is our capacity to challenge anyone at any time about patient-related issues.”

After 20 years of teaching communication excellence and researching over 20,000 individuals, Grenny agrees. “The essential lever within any organization is the ability to handle crucial conversations well. “

Next time you check into a hospital or visit your local health care provider, ask them how they are doing with JCAHO standard 2(a). Alternatively, if you want to be sure you are going to get the best care possible, begin by having a conversation with your caregiver. If they stay healthy in their conversation and don’t become angry or offended, even when the subject turns crucial, you are probably in good hands!

Joseph Grenny is one of the authors of the *New York Times* best-selling book, *Crucial Conversations, Tools for Talking when Stakes are High.* Over the past seventeen years, he has designed and implemented major change initiatives for numerous healthcare clients.

A Man’s Guide to Coping

Written to fill the void in the literature regarding the special needs of men with disabilities, this book includes information about men’s responses to disability, with a special emphasis on the values men place on independence, occupational achievement, and physical activitiy. Information on finding local services, self-help groups, laws that affect men with disabilities, sports and recreation, and employment is applicable to men with any type of disability or chronic condition. The disabilities that are most prevalent in men or that affect men’s special roles in society are included. Chapters on spinal cord injury and stroke include information about the disease or condition, psychological aspects, sexual functioning, where to find services, environmental adaptations, and annotated entries of organizations, publications and tapes, and resources for assistive devices. Includes internet resources. From Resources for Rehabilitation, Winchester, Massachusetts, +1-781-368-9094 or www.rfr.org E-mail: orders@rfr.org. Third edition, February 2003, ISBN 0-929718-32-1 $46.95, or ask your library to order it for their collection.

A companion guide, *A Woman’s Guide to Coping with Disabilities*, includes information on pregnancy, childrearing, caregiving and employment. Special attention is paid to ways in which women can advocate for their rights with the health care and rehabilitation systems. ISBN 0-929718-26-7

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First Meeting in Japan

by Hanako Suzuki, Tokyo, Japan

Our first VHL meeting was held in Osaka, February 15, 2003. It was chaired by Dr. Taro Shuin of Kochi Medical School, who heads VHL research in Japan.

My husband is actually the VHL patient, but unfortunately he was on a business trip. Happily, he is very well now. Most of time he is able to forget about his disease. Anyway I went to Osaka instead of him. I arrived at Osaka at 11:30. It took 2 hours and a half by Shinkansen (the bullet train). Osaka is the second largest city in Japan and famous as the city of laughter. Most of the famous Japanese comedians are from Osaka.

Fourteen patients and family members gathered in a conference room of the Pharmaceutical Division of Kirin beer.

As you know, it was our first meeting, so first of all we asked everyone about their experiences and their wishes for this group. Then we decided to propose our actions for the first fiscal year.

Dr. Kanno made a presentation about symptoms and treatments of CNS hemangioblastomas. Dr. Ito talked about his experience with a group which he supports for another rare disease called ADSL. Dr. Shuin described his VHL research in Japan. Most Japanese VHL patients have CNS hemangioblastomas. The peak ages are age 16-20 and age 26-30. It seems it's younger than in western countries. However we seldom have pheochromocytomas. I don't know if this kind of data is available for other Asian countries and for Japanese who emigrate to another country.

Ms.Hikosaka and Ms. Akagi, Kochi Medical School staff.

After the meeting we had a small party. I talked to an interesting guy from Kyoto who has two other people with VHL in the family. They own a family business. When they were diagnosed with VHL, they all had tumors and their surgeon recommended that three of them have surgery immediately. They couldn't close their store, so they argued to decide who would have surgery first. In 50 days he is going to have brain surgery again. He brought his MRI films to the meeting and was given advice from doctors who are familiar with VHL. It seems everyone really enjoyed the conversation, foods and drinks.

We are delighted with the many patients and families with VHL who now belong to our new group!

We are looking forward to the International Medical Symposium on VHL scheduled in Japan May 20-22, 2004.

For VHLFA Japan see http://www1.odn.ne.jp/vhl_japan/
For Kirin Pharmaceuticals, see http://www.kirin.co.jp/english/r_d/pharma/020630.html

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