VHL FAMILY FORUM

Membership Renewal Issue!

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

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Therapy of Retinal Angiomas of VHL

by Emily Y. Chew, M.D., Deputy Clinical Director, National Eye Institute, National Institutes of Health, Bethesda, Maryland, USA

Retinal angioma in the eye may be one of the earliest manifestations of VHL disease. As many as 60% of patients in some studies of large kindreds may have eye involvement. The clinical appearance of these angiomas is very typical and diagnostic of VHL. The initial appearance of a retinal angioma is a subtle red or grayish dot no larger than a few hundred microns. (See Figure 1.) As the proliferation of the vascular tumors (mostly of capillaries) progresses, secondary alterations occur to produce a distinctive clinical appearance. (See Figure 2.) The blood vessels leading to and away from the tumor become characteristically dilated with marked enlargement. This tumor can lead to leakage of fluid and fatty deposits both around the tumor and in the central important area of the retina, the macula, which is responsible for the fine vision needed for reading, driving, etc. If the angiomas enlarge to an extent that the retina can be detached, hemorrhaging and scarring can occur. These can all lead to decrease in visual acuity of the affected individual. Rarely can these tumors regress spontaneously.

Often patients do not have symptoms as these lesions tend to progress slowly. The tumors can be detected in children to adults in the eighth decade of life on a routine exam. Symptoms such as decreased vision or a turned-in eye (crossed-eye) may result in the detection of VHL in children. Decreased visual acuity can also cause adults to seek medical help and subsequent detection of the disease.

The treatment of the retinal angiomas will depend on the location and size of the lesions. Small lesions are easy to treat successfully while large lesions are notoriously difficult to treat. Laser photocoagulation can eradicate small retinal angiomas in most locations. However, for those tumors too large or located in the very periphery of the retina, cryotherapy (freezing treatment) may be indicated.

If the tumor is located on the optic nerve, the nerve that connects the eye to the brain, treatment is fraught with difficulties. (See Figure 3.) Marked adverse side-effects are associated with treatment of such tumors with laser photocoagulation. Fortunately, these tumors may remain asymptomatic for long periods of time. For patients with the more severe changes such as retinal detachment, hemorrhage and scarring, the procedure called vitrectomy can be performed. This involves the introduction of microinstruments under the guidance of a microscope to remove the areas of scarring and to flatten out the retina. Other treatments that have had some limited success include radiotherapy. However, experience with this modality is somewhat limited. Other therapies have also included photodynamic therapy which has been reported to have beneficial results in a few cases. (See article by Dr. Singh, page 3)

More recently, ten cases of severe optic nerve tumors or multiple tumors of the retina were treated with two different types of anti-Vascular Endothelial Growth Factor (VEGF) drugs (Macugen and Lucentis) with very limited success at the National Eye Institute (NEI) of the U.S. National Institutes of Health (NIH). We are now testing the use of oral Sutent (sunitinib malate) in cases of severe optic nerve tumors or multiple tumors of the retina that are causing vision loss. This drug is FDA approved for the treatment of metastatic renal cell carcinoma. Because of its ability to attack the tumor through different mechanisms of action, it might be important in the therapy of these retinal angiomas that are not

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Inside this issue!

Dr. Chew

amenable to other treatment. This is an open label study in which all patients will receive the therapy for at least 9 months. The study is now open for recruitment.

The importance of maintaining good visual function in patients affected with VHL depends on regular dilated (the opening of the pupil of the eye with drops) eye examination. For patients who are at risk of developing VHL, an annual dilated eye exam will provide important information and help maintain good vision. It is very important that all patients affected or who are at risk be examined annually through DILATED pupils. Good vision can be achieved and maintained in many affected individuals, especially if the lesions are detected and treated early in the course of the disease.

For those individuals who are interested in the NEI/NIH trial of Sutent for optic nerve tumors associated with VHL, please contact Katherine Shimel, RN at Katherine.Shimel@nih.gov or by telephone: +1 (301) 402 2863.

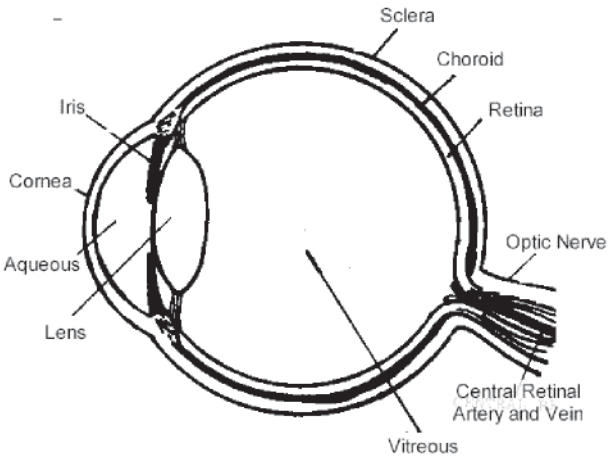

Figure 3: Illustration of the human eye, showing the optic nerve. The periphery is the part of the retina farthest away from the optic nerve.

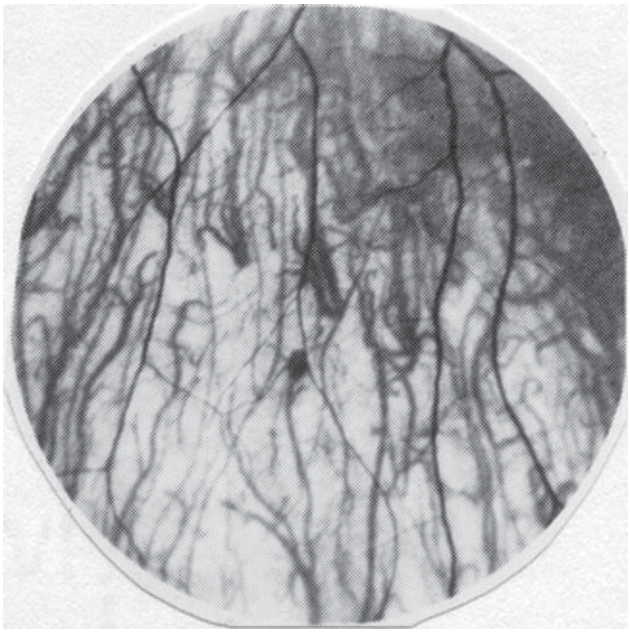

Figure 1: A small, preclassical angioma is located slightly below the center of the photograph. This small lesion can easily be overlooked during an eye examination.

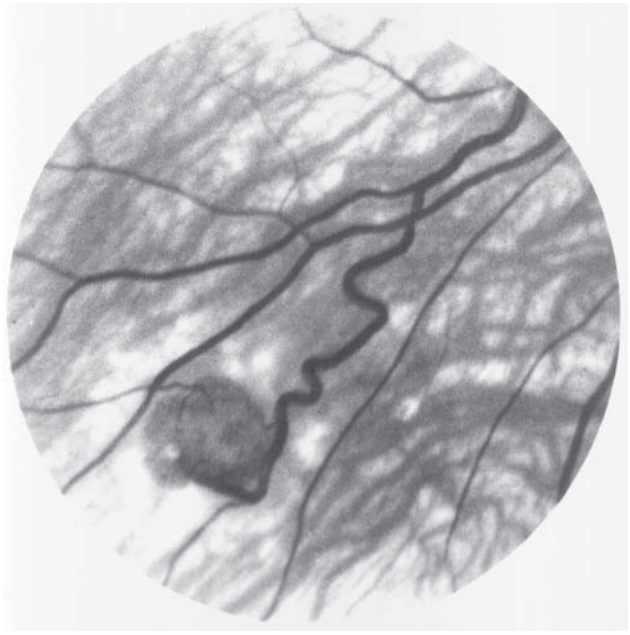

Figure 2: This angioma shows a large feeder vessel leading to the angioma with a large vein as a draining vessel.

These illustrations are taken from an article "Von Hippel-Lindau: Ocular Complications," by Lloyd M. Aiello, M.D., and Jerry Cavallerano, O.D., Ph.D., published in the *VHL Family Forum*, 1994. See http://www.vhl.org/newsletter/vhl1994/94ceeye.php

Photo-Dynamic Therapy

-- Arun D. Singh, M.D., Director, Department of Ophthalmic Oncology, Cole Eye Institute, Cleveland Clinic Foundation, Cleveland, Ohio, gave a presentation on treatment of VHL eye lesions in Sandusky, Ohio, October 2007. Joyce Graff asked some follow-up questions about photodynamic therapy.

Dr. Singh

Joyce: Let me ask you one more question about the tumors on the optic nerve, since those seem to be the most problematic. You said that we should wait and not treat them until there is some visual disturbance?

Singh: Absolutely. I have no hesitation in saying that, because the tumors on the optic disc tend to be inactive for many, many years. So number one, if they are causing no problems whatsoever with the vision, you are better off leaving them alone. Because many times they may just stay that way for a long time. That's number two. And number three is the fact that if you were to treat them, chances are you would end up losing some vision because of the treatment related complications.

Joyce: So if the tumor begins to grow and begins to disturb the vision, then what is your preferred way of dealing with them?

Singh: Well, the preferred treatment for optic disc hemangioma presently is photodynamic therapy (PDT), where in a dye is injected that specifically locates itself in the tumor, and with the a special wave length laser one can preferentially destroy the area where the dye pools, which is the hemangioma. So the aim is to achieve selective ablation or destruction of the hemangioma while attempting to spare the surrounding blood vessels, retina, and the optic disc.

Joyce: So this dye goes and hooks onto the tumor cells in the hemangioma, and not the healthy cells, and then this special laser light goes in and essentially kills off those tumor cells?

Singh: Yes. The laser light is picked up by the dye, so wherever the dye is, that's where the energy will be taken up. The normal blood vessels don't have the dye, so they will not be affected by the laser. And this laser is not heat-dependent. It triggers a chemical reaction in the dye and that chemical reaction kills the cells. The tumor will shrink with time. The end point or objective here is not necessarily the total ablation of the tumor, but enough scarring or fibrosis within the tumor that it stops leaking and stops causing the exudation around it.

Joyce: So it makes it less active, and less likely to secrete the fluid that causes the visual problems.

Singh: Exactly. So the end point is resolution of the fluid and the symptoms and improvement of vision, and not necessarily complete ablation of the tumor.

Joyce: So if this tumor is made up of capillaries, though, and you kill off these capillaries, isn't that going to cause some bleeding?

Singh: There can be small hemorrhages around the tumor from the laser uptake. That is an indication that you did the laser alright, and there has been laser energy uptake. Those hemorrhages typically will go away in a few weeks and should not cause any permanent problems.

Joyce: OK. It does take a few weeks, though, for that blood to clear out of the eyeball.

Singh: Yes. These hemorrhages are typically in the retina, not in the vitreous cavity. These are small hemorrhages and they will go away. The response to laser treatment is not immediate, though. It may take anything up to six weeks or so before you see the tumor shrinking away, and many months for the lipid exudation to go away. And it may take a few weeks for the fluid to go away. So the response is not immediate. But if one has to repeat the treatment you almost always end up waiting as much as three months because you want to give that much time for the response to begin to show.

Joyce: But the point here really is that we are not damaging the healthy retinal tissue by using PDT, right?

Singh: That is the aim, but there is some damage to the optic nerve, one would imagine, because the tumor is right on the optic nerve. So there will be some spill-over collateral damage, although PDT would still be the best method that we have out there right now.

Joyce: Right. There is no perfect method for this.

Singh: No. There is no perfect method.

For the link to Dr. Singh's presentation with his original slides, please see the online version of this article at http://vhl.org

Parent / Patient Counseling

by Markus Jansen van Vuuren, Chairman, VHL South Africa

As part of the preparation process to formally structure the VHL Association of South Africa (VHLSA) in support of its founding purpose, a Parent/Patient counseling course was organized for VHL patients and their caregivers in the Johannesburg area.

A group of 13 volunteers attended this counselor training course held at the Sappi Head Offices in Braamfontein, a suburb of Johannesburg.

The course was held over a 4 day period split over 2 weekends: 26-27 July 2008 and 2-3 August 2008 and was presented by Merlyn Glass from the South African National Health and Laboratory Services (NHLS) and Julie Lampret, a Counselor in private practice.

The fundamental goal of the course was to equip the parents, patients and caregivers of the VHLSA with the basic techniques and principles of counseling. Although the main purpose of this course was to create support counselors for the Support Group that can apply their skills and knowledge towards helping others in need, these same principles can be applied within our own lives by doing self exploration and working on those things to make us better people.

We do acknowledge the fact that not all of us are counselors and do not want to apply these counseling techniques to others in need, but at least training equips us with basic techniques and principles in order to help ourselves in our own day-to-day lives.

The group of people who attended the course were challenged through constant role-play in order to practice the newly introduced tools and techniques. The practical examples used by the trainers created a relaxed atmosphere whereby personal stories could be shared amongst the attendees.

One such role-play was to have a conversation with a partner whilst pursing a marshmallow between your lips. (See Figure 1.) This was done to illustrate only one of the many barriers that may exist between people in a counseling session. One such barrier in South Africa is the 11 official languages. Although English is the universally accepted language, most of the elderly people in the rural areas cannot read or write, let alone communicate in the universal language.

Another major barrier to overcome is cultural beliefs. Although role play is different from and most of the time more difficult than real life situations, some attendees shared real stories, experiences and situations with their counselors. Some of the attendees noted that they received valuable advice during these role-play sessions.

A practical brainstorming group session was held whereby the groups developed a process to follow when arranging a counseling session with someone in need. (See Figure 2.)

Each group had to present their process and ideas to the other groups. Discussions were then held around these ideas. Each group came up with excellent ideas which will be incorporated into the action plans of the organization which we believe will add tremendous value to others to be reached by the Association.

> “... only 7% of our communication is verbal”

This revealed various aspects, not part of the initial design, that are vitally important and must be considered when arranging a meeting with a person in need.

In order to practice the newly introduced techniques on counseling, practice sessions were held where each person had the chance to be alternately the counselor, and then the person being counseled.

During the feedback session, after each role play, experiences were shared and discussed within the group. Mistakes were pointed out and each person shared their failures and successes with the group.

Some of the counseling skills surfaced naturally depending on personal life experiences, but others skills were new to us. We learned many new skills in this course.

These skills and techniques were grouped together in a logical way whereby every person can apply them in their day-to-day lives, not only to make a difference in the lives of others but also for themself and their loved ones.

It is my belief that everybody should attend a

Figure 1: A marshmallow serves as an example of one barrier to communication.

course which teaches you the techniques of applying the counseling tools. Continuous self exploration, constantly working towards bettering yourself, acknowledging your weaknesses and strengths, it becomes a way of life so that your world can be a better place filled with happiness and joy.

Before you realize it, this will have a positive effect on those people around you: at your work, among family and friends, and will start to spread rapidly to others. An old saying states that birds of a feather flock together. The law of physics clearly teaches us that: Positivity attracts positivity and negativity attracts negativity.

Although most of the attendees had already met at the conference held in March 2008 and VHL workgroup meetings, relationships were deepened as they started to know their colleagues better. The lunch-table proved to be no different as the attendees enjoyed the relaxed atmosphere with a hot meal prepared by Sodexho.

These counselors were trained to assist people to tell their full story and assign priorities to their needs. The various values of a good and bad counselor were discussed and put to practice after group discussions were held.

Listed below are a few good Counselor attributes:

1. Helps you to focus
2. Is constructive
3. Summarizes
4. Is trustworthy
5. Is discreet and confidential
6. Is approachable
7. Is open
8. Makes him or herself available
9. Takes you seriously
10. Is self-aware
12. Is gentle

Some of the most common errors made by counselors are:

1. Satisfying one's own needs
2. Talking more than listening
3. Concentrating on facts, not feelings
4. Asking too many questions and too many closed questions
5. Being judgmental
6. Ignoring discrepancies between verbal and non-verbal
7. Breaking confidentiality
8. Not being open to feelings
9. Making assumptions about the client's situation
10. Generalizing – not recognizing the individuality of each person's situation.

Grief is one of the most effective killers in any relationship due to insufficient communication and saying things in a reactive manner. When communicating about things that are not according

Figure 2: Brainstorming session.

to our own liking, we have to change our sentence and tone of voice to be constructive instead of provoking anger and grief.

There are three major components to communication: verbal (7%), tone and word selection (23%), and body language (70%). Although only 7% of our communication is verbal, it causes the most harm due to the selection of words and the tones used. A major part of self-awareness is to be constantly aware of the body language we use, which speaks far more than any spoken word.

By practicing to create a perfect harmony among these three methods of speech, you will get the best results not only when dealing with a person in need but also through the vibrations you create that affect people around you.

By understanding the various stages of grief, you will be able to categorize a person's grief and guide him or her through the stages of grief to healing. These stages are: Denial, Anger, Bargaining, Depression and Acceptance. The lessons learnt through this course were most valuable, and we are now equipped with powerful and essential tools to make a difference where needed -- not only within ourselves and the organization but also with our loved ones and the people around us.

A special word of thank to our sponsors who made the training possible:

- Sappi Limited for the use of the Auditorium and facilities,

- Sodexho for the catering and refreshments,

- South Africa Inherited Disorder Association (SAIDA) for covering the catering costs,

- Merlyn and Julie for offering their weekends to empower others.

Sponsors' web sites:

www.sappi.com

saida.org.za

A similar Counseling course is being planned for the VHL people and their caregivers in the Western Cape province, South Africa. The dates will be published once finalized. Bookings to be made with Markus van Vuuren.

For more information on VHLSA, its purpose and programs, you may contact Markus Jansen van Vuuren on +27(0)82 779 5019 or e-mail to: southafrica@vhl.org

Europe: Cross-border health care

The EC adopts a proposal for a directive aimed at improving healthcare cooperation

Accessing health care is particularly pertinent to rare disease patients in the European Union (EU), who often have limited resources of care and expertise in their region.

With over 7,000 rare diseases identified to date, it is not possible for each EU country to develop expert care and treatment services for every disorder. Thus, the rare disease patient – perhaps more than any other EU citizen – needs to be able to access medical expertise where it exists.

In July the European Commission (EC) adopted a proposal for a directive on the rights of patients accessing cross-border healthcare. The proposal tackles the complex issues of reimbursement for care obtained in member states and the coordination of social security schemes; mutual recognition of professional qualifications; discrimination; and the need for a community framework for the protection of personal data.

The proposal next has to go through a co-decision process involving the Council of Ministers and the European Parliament. If the readings are not completed before June 2009, the process will have to recommence upon the election of the new parliament rapporteurs.

Once a decision is taken, the member states will have one year to implement the directives of the proposal.

from OrphaNews Europe, the newsletter of the Rare Diseases Task Force, OrphaNet, August 2008.

Nutrition and Cancer

It sounds like a simple question: Can what we eat influence our risk of cancer? In fact, finding the answer is a complex puzzle worthy of Sherlock Holmes.

Do you eat the exact same things every day? Of course not. Most people's diets are complex, and they change over weeks, months and years. To pinpoint associations between diet and cancer, researchers must dig through this massive menu to isolate the specific effects of individual foods.

Each scientific study provides another clue to the evolving mystery of how diet affects cancer. But as with any good mystery, some clues hold more weight than others. Since cancer isn't going to confess to what caused it, no single piece of evidence gives us all the answers. The "body of evidence" formed by many studies must be considered as a whole as we investigate the mystery of the diet-cancer connection.

Click the link below to help you to understand the studies.

http://www.aicr.org/site/PageServer?pagename=res_studies_home

This is a great website to help you understand how to interpret theinformation you read about diet and its impact on cancer.

U.S. Medical Costs

High Medical Costs Are Weighing Heavily on Both the Insured and Uninsured

According to a survey by the Commonwealth Fund, "Americans are struggling to pay medical bills and are accumulating medical debt at an increasing rate" due to health care costs that are rising faster than incomes and the soaring prices of food, gas and other items, reports the *Washington Post*.

"Two-thirds of the working-age population was uninsured, underinsured, reported a medical bill problem or did not get needed health care because of cost in 2007," the study found. Among those struggling to pay their medical bills, "39 percent used all their savings, 30 percent incurred large credit card debt, and 29 percent said medical bills left them unable to pay for basic necessities such as food, heat or rent," according to the article. The survey showed that 28 percent of working-age adults were uninsured at some point in 2007, compared to 24 percent in 2001. But the problems dealing with medical costs were not limited to those without insurance. In fact, "61 percent of those with medical debt or bill problems were insured at the time they needed medical attention."

While half of people with incomes below $20,000 were uninsured in 2007, up one percentage point from 2001, the increase among those earning between $20,000 and $40,000 was much higher--from 28 percent to 41 percent. Eighteen percent of those earning between $40,000 and $60,000 were uninsured, up five percentage points from 2001, and the uninsured rate among those earning more than $60,000 rose from 4 percent to 8 percent, reports the *Post*.

According to the study, a universal health care system "is key to improving health care." Karen Davis, president of the Commonwealth Fund, said: "The U.S. stands out for being the only country… that reports significant fractions of the population not getting needed care."

Source(s): Joshi, *Washington Post,* 8/20/08

VHL Radio Show

Powerful Patient tops 30,000 users a week!

The Powerful Patient radio show produced by the VHL Family Alliance is now being heard by more than 30,000 listeners each week.

That's thirty thousand people each week who are hearing about VHL, learning some of the skills we have learned in dealing with a complex medical condition, and gaining an appreciation for the breadth of topics you have to deal with in managing your health.

When is it on?

PowerfulPatient is available on-demand on the internet whenever you want it to be on. Just go to PowerfulPatient.org and select the program you want to hear.

It does run on "terrestrial radio" periodically. The schedule is on the network website, http://www.webtalkradio.net You can help by suggesting to your favorite station that it carry the Powerful Patient.

Why does it talk about more than VHL?

When we select a guest, we always ask: What can we learn that would be helpful to people with VHL? What can those of us with VHL teach others? In speaking to a broader audience, it is also serving a role in public awareness of VHL.

What topics are covered?

Here are just a few of the topics that have recently appeared:

- **Nutrition** to keep that second copy of the VHL gene strong so it can naturally suppress tumors
- The importance of **sleep** in supporting the body's own natural defenses
- Two people **living with cancer** (one with VHL) talk about how they are managing their risks
- A discussion of the importance of the **health of both parents** in making good copies of their DNA when having a baby. Dr. Jeanne Wilson adds her expertise from Chinese medicine in partnership with a number of fertility clinics in California.
- Important insights on the mistakes that can happen in hospitals, the steps that hospitals are taking to reduce errors, and how we as patients can help to safeguard our own patient safety.
- The booming new industry of **Medical Tourism**. In 2007, 750,000 Americans went outside the U.S. for medical treatment. That number is expected to reach 1.5 million by the end of 2008, and 4.5 million by 2010. We speak with one U.S. health care provider network that is offering its members the option of surgery in New Zealand with no co-pay.

Please join us!

Go to http://powerfulpatient.org

Conference Recordings

Recordings of the outstanding professional presentations at the last three major meetings are now online:

Orlando 2008

Houston 2007

Boston 2007

It's a bargain!

It cost us at extra $3000 per meeting to make these DVD recordings for you. For that reason, there is a charge to obtain these recordings, either on the web or as a DVD/CD set sent to you. It is no more than people paid to register for the meeting itself -- and you have no travel or hotel costs! Where else can you find such outstanding information on VHL? When you order the DVD you also receive a CD with the handouts from the meeting. See page 15 or order on the net at vhl.impactlearning.org

We need your feedback

Please give us your feedback on this program. We need to hear from you about the convenience, the content, and any other comments.

Webinars this fall!

We are embarking on a series of Webinars this fall.

What's a Webinar?

It's a seminar, on the web, with audio and slides. The original "live" program will be open to attendance by registration only. Your only cost will be the cost of the telephone call.

After the original "live" presentation, we will have a recording of the meeting available on the website for you to replay for free.

How is this different from the radio show?

The radio show is a more free-ranging conversation with one or more experts.

A Webinar allows us to have slides to accompany the conversation. It will usually be a presentation, with some opportunity for questions and answers during the "live" program only (not during the replay).

What's the line-up?

24 Sept - VHL 101 with Joyce Graff

16 Oct - Pre-implantation Genetic Diagnosis (PGD) with Dorothy Twinney, M.S., of Genesis Genetics

23 Oct - Report from Copenhagen

20 Nov - Report from Kidney Cancer Symposium

Watch vhl.org/meetings for announcements, registration for the live shows, and the archive of replays

Is Your Family from Germany?

Black Forest families sought

Professor Hartmut Neumann, a nephrologist at Albert-Ludwigs-University of Freiburg, Germany, is working on pedigrees of patients with the Black Forest mutation, which may be written

c. 505 T>C (Y98H) (old style) or

c.292T>C (p.Tyr98His) in standard nomenclature.

He is especially interested in patients with this mutation living outside of Germany. He knows that there are many people from this lineage in the USA, whose ancestors moved to Pennsylvania in the 1600's from the Black Forest area of Germany. There may also be people from this lineage in South Africa, Australia, or other countries.

While most people with VHL are not related to one another and have many different kinds of changes in the VHL gene, this one large extended family was identified in 1995 by Dr. Neumann in Germany and Dr. Berton Zbar in the United States. Because they share a common mutation and have a fairly well-recorded family history, Neumann and Zbar were able to determine that VHL has existed in this lineage since before some members left for Pennsylvania in the 17th Century.

This phenomenon is called the Founder Effect -- we can identify a lineage where there is one Founder prior to the exodus of the Pennsylvania members of the family from Germany.

If you have this particular mutation type, he would like very much to be in touch with you. He is at the University of Freiburg in Germany, and serves on the medical advisory board of the VHL Family Alliance. If you are willing to share some information with him, please contact him directly at DrNeumann@vhl.org, or by mail at

Dr. Hartmut Neumann
University Medical Clinic
Hugstetter Strasse 55
Freiberg im Breisgau
D-79106 Germany

Reference: Brauch et al, "Von Hippel-Lindau (VHL) disease with pheochromocytoma in the Black Forest region of Germany: evidence for a founder effect Human Genetics, 1995 May;95(5):551-6.

Abstract: We identified a germline missense mutation at nucleotide 505 (T to C) of the VHL tumor suppressor gene in 14, apparently unrelated, VHL type 2A families from the Black Forest region of Germany. This mutation was previously identified in two VHL 2A families living in Pennsylvania (USA). All affected individuals in the 16 families shared the same VHL haplotype indicating a founder effect. This missense mutation at codon 169 (Tyr to His) would probably cause an alteration in the structure of the putative VHL protein. The association of this distinct mutation with the pheochromocytoma phenotype in VHL may help to elucidate the genetic mechanism of carcinogenesis in this multi tumor cancer syndrome.

Ask the Experts

Wearing Glasses for Protection

Question: If someone is blind in one eye, should they wear glasses for protection to the good eye? About a year ago my daughter was seen by her opthalomologist and while taking a family history the doctor was adamant that I should be wearing glasses to protect my good eye. Then just last week I was talking with another VHL patient who has lost the vision in one of her eyes and she asked me why I didn't wear glasses. She wears glasses to protect her good remaining eye.

If we should be wearing glasses, I wonder how many people are just ignorant like me and didn't even think about that as being important. My own Doctor kind of shrugged it off as not being important so I never followed through on it. What's the concensus on this issue with the experts?
-- Donna B., Oregon

Response: We polled our Medical Advisors, and everyone agrees that people with only one eye should wear glasses for protection.

"We strongly recommend that all patients who have only a single eye that is providing functional vision should use eye protection (glasses) at all times. We have all seen cases in which people have had freak accidents which resulted in blinding of their remaining eye. Polycarbonate lenses offer the best protection at a reasonable price. I make this one of my "checklist items" for all of my monocular patients." – *Michael B. Gorin, M.D., Ophthalmology and vision sciences, University of California, Los Angeles*

"The American Academy of Ophthalmology recommends and I quote 'Patients who are functionally monocular should wear proper protective eyewear full time, even if they do not require corrective lenses.'" – *Arun Singh, M.D., Ophthalmology, Cleveland Clinic, Cleveland, Ohio.*

What's a Webinar?

See page 7

Cell Phones? Use a headset or speakerphone

By Andrew Weil, M.D., from Ask Dr. Weil

Question: I was alarmed to hear about the cancer researcher who warned his staff to limit cell phone use because of a potential cancer risk. This is after I heard neurosurgeons on "Larry King Live" warn against holding cell phones to your ears. This is scary. Your thoughts?

Response: I wish I knew more about what motivated the July 23, 2008 warning from Ronald B. Herberman, M.D., director of the University of Pittsburgh Cancer Institute. According to the Associated Press he based his warning to his faculty and staff on "early unpublished data."

In a letter sent to 3,000 faculty and staff on July 23, Dr. Herberman said we should "err on the side of being safe rather than sorry later." He urged that children use cell phones only for emergencies (because their brains are still developing) and that adults use headsets or the speakerphone rather than holding the phones to their ears. He also cautioned against using cell phones on buses and in other public places because you might expose those around you to the instrument's electromagnetic field.

In the discussion about cell phone safety on "Larry King Live" in May 2008, three neurosurgeons -- one from Cedars-Sinai Medical Center in Los Angeles, CNN's chief medical correspondent Dr. Sanjay Gupta, and one from Australia -- all said they observed widespread dangers from cell phone use now that millions of people worldwide spend so much time on them.

So far, results of formal studies have been contradictory: some find a link between cell phone use and brain tumors and some do not. The difficulty is that brain tumors can take 30 to 40 years to develop, so it could be a long time before we know for sure whether cell phone use is safe or, if not, how great the risks may be. Until then, the big worry is that children may be more vulnerable than adults (just as they're more susceptible to risks posed by exposure to cigarette smoke, lead, and radiation). For this reason, even before Dr. Herberman's warning, experts in the United States and Britain advised that youngsters' use be limited. [See Note 1 below.]

I understand that two of the physicians who appeared on "Larry King Live" said that they always use earpieces with their cell phones to keep the energy from the microwave antenna away from the brain. The third said he uses his cell phone in speakerphone mode. I agree with them and with Dr. Herberman on this. In fact, I've been recommending the use of earpieces or headsets for years. Until we know whether or not long-term use of cell phones increases the risk of brain tumors, I think we should be careful.

I've also been asked whether the type of brain tumor that Sen. Edward Kennedy (D-MA) has (a glioma) has been associated with cell phone use. Yes, it is, [See Note 2] but we have no hard evidence showing that this (or any other kind of tumor) is directly caused by using cell phones and, of course, no way of knowing what caused Sen. Kennedy's tumor. Gliomas were around long before there were cell phones. Some research has linked cell phone use with salivary gland cancer and with acoustic neuroma, a benign but troublesome tumor of a cranial nerve.

In addition to using a headset or earpiece, I suggest taking these precautions:

- Save long conversations for conventional phones.
- In your car, use a cell phone that has a remote antenna outside the vehicle.
- Find out how much radiofrequency energy your cell phone emits. (This measurement is called the Specific Absorption Rate or SAR; find the SAR for your cell phone via the FCC. [See Note 3.] The SAR permitted in the United States is 1.6 watts per kilogram.)

Published August 22, 2008, on *Ask Dr. Weil*, www.drweil.com. Reprinted with permission. Dr. Andrew Weil is not an expert on cell phones, he is a medical doctor and professor of integrative medicine at the University of Arizona who answers consumers' questions on his website and on public television. VHLFF editor add the following notes:

Note 1: His recommendations are entirely consistent with the report from the National Cancer Institute (NCI): *Do cell phones cause brain tumors?* "There is no evidence to date that they do. However, the National Cancer Institute does recommend caution since there may be long-term effects which we do not yet understand." http://www.cancer.gov/cancertopics/factsheet/Risk/cellphones

Note 2: Most of the studies published to date do not show a link between glioma and cell phone use. According to the NCI, "Overall, research has not consistently demonstrated a link between cellular telephone use and cancer or any other adverse health effect."

Note 3: The Specific Absorption Rate for your telephone can be found in the literature that came with your phone, or on the Federal Communications Commission (FCC) website: http://www.fcc.gov/oet/rfsafety/

Start Near to Home!
Help your local Chapter!
Raise Awareness?
See page 10 for ideas

VHLstock!

-- by Calvin Cieslak, California

As I've struggled with the challenges of Von-Hippel Lindau in my life these past few years (major surgeries each of the last two summers), I've also wondered what I could do to more proactively battle the disease.

During a discussion with my parents and college counselor last year, we talked about the possibility of putting on a fundraiser that would benefit the VHL Family Alliance.

At first, the plan was to have a small fundraiser/ concert for friends and family – featuring me on guitar, playing some of the music I have written. Over time, however, plans for the event grew into something much larger.

Enlisting the help of my immediate family, grandparents, aunts, uncles and cousins, we invited several hundred people to join us for an evening of music, wine tasting and dessert under the stars. I called the evening VHLstock in the spirit of rock & roll. And instead of just me on guitar, we pulled together a whole band, featuring my uncle and me playing guitar, my dad on drums and my cousin playing keyboards.

Calvin Cieslak

Calvin and Brian Cieslak

Over 150 people attended and the event was a tremendous success! The event is something that has finally made me feel as though I'm doing something progressive for VHL.

Calvin and his family raised more than $10,000 for VHL.

Watch the vhl.org website for a link to a YouTube video from this concert.

Congratulations and thanks to Calvin, his family, friends, and supporters!

Bloomsday Run!

-- Jenny M. and Kim H., Washington

Jenny M. and Kim H. joined 50,000 trekkers in May 2008 for the annual Bloomsday walk in Spokane, WA. Here is Jenny's description.

"We were amazed at the number of people, both participants and spectators alike, that asked us about VHL. There were 50,000 participants and many more spectators. People were cheering us on saying "Way to go VHL!" and radio stations positioned throughout the 7.46 mile course always asked what VHL stood for and then gave us a plug over their loud speakers. We decided my sister, Conny, should be our official spokesperson because she did such a good job explaining VHL to everyone.

"KXLY TV interviewed Kim and me and the interview will air today (thanks to Sallie Rowe). It was such a great experience I encourage more people to do something similar to just get the VHL name out there -- if they see it, they will ask about it! We decided next year we will hand out cards with info to those who ask about VHL and once again we will put VHL.org on our shirts."

Jenny and Kim and friends raised over $2000!

PoWow Triathlon

-- Beth S. and her training buddies, Janet and Alicia, Massachusetts

Thank you for visiting our fundraising page for VHL Family Alliance - Cancer Research Fund!

We did it!!!! On Saturday, July 12, 2008, we competed in the Powow Triathlon in Amesbury. We swam .5 miles in Lake Gardner, biked 12.7 miles and ran 3.2 miles.

In June our training definitely been stepped up a notch. We began swimming 4,650 yards each week, that is about 3 hours in the pool. We biked 45 miles a week in addition to running 12 miles. It was tough, but now that we are in better shape, it is much easier.

The actual event was such a blast!!! We plan to continue our training and compete in future triathlons this summer.

Thank you to all of our friends and family for your support throughout the training process as well as all of your generous support in our fundraising efforts. We had set a goal of $2500. So far we have raised $4,175!! If you'd like to contribute, please see our web page, http://www.firstgiving.com/powowtriathlon

How can you help?

- Clinical Care Committee
- Membership Committee
- Help develop the local group in your area

Please let us know -- see page 15.

"Stealth" exercise

Add a little exercise to your day

Regular exercise can lower your risk for heart disease, diabetes, and even some forms of cancer. Yet many people still aren't making physical activity a priority. In its first-ever single-topic special issue, the September 2008 *Harvard Health Letter* offers 27 tips to get your heart rate up without going to the gym. Its recommendations include these:

1. Take the faraway spot. Walking from the farthest corner of the parking lot will burn a few calories. If it's a parking garage, go to the roof and use the stairs.

2. Get into the swing of it. Swinging your arms when you walk will help you reach the brisk pace of 3 to 4 miles per hour that is the most healthful.

3. Be part of the fun. Adults shouldn't miss a chance to jump into the fray if kids are playing on a playground or splashing around in the water. Playing along will strengthen muscles and bones and set a good example.

4. Clean house. Even if you have a cleaning service, you can take responsibility for vacuuming a couple of rooms yourself. Fifteen minutes burns around 80 calories.

5. Adopt someone as your walking, jogging, or biking buddy... Adding a social element to exercise helps many people stick with it.

6. ... even a buddy with four legs. Several studies have shown that dog owners get more exercise than the canineless.

7. Be a stair master. Taking the stairs is good for your legs, knees, and cardiovascular system. Don't overdo; take one flight at a time.

Also in this special exercise-themed issue:

- Why we should exercise—and why we don't
- Exercise gadgets
- Tracking exercise in metabolic equivalents (METs)
- Benefits of yoga
- Knee injuries in female athletes
- Discussion with I-Min Lee, Harvard activity and health expert

The *Harvard Health Letter* is available from Harvard Health Publications, the publishing division of Harvard Medical School, at www.health.harvard.edu/health or by calling 877-649-9457 (toll-free).

The thank-you list appears only in the print version

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Membership & Orders

If you have already paid your dues in 2008, thank you!

Name: _____

Address: _____

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

Home phone: _____ Work phone: _____

E-mail: _____

Preferences:

☐ I would like to receive occasional alerts via e-mail ☐ Please send the newsletter on paper only

Yes, I want to help!

☐ I can help in my local area

☐ I have the following talents to share: _____

_____

Public Awareness:

☐ Please send me a press kit

☐ Please send me a copy of Adam Linn's film, Smell the Light (9 minutes) @ $12 $ _____

Conference Recordings - a set: DVD of the talks plus a CD with the handouts

__***# Boston 2007 -*** Research, Imaging & Contrast, Feeling Beautiful, Diagnosis & Treatment of Pheochromocytoma, Biomarkers for Kidney Cancer, RFA & kidney, Pancreatic lesions @ $50 $ _____

__***# Houston 2007 -*** Radiofrequency Ablation (RFA), Neurosurgery & VHL, Sutent Trial, HIPAA and VHL, Pheochromocytoma, Minimally Invasive Surgery @ $50 $ _____

__***# Orlando 2008 -*** Genetics of VHL, Robotic Surgery, Brain & Spinal Tumors, State of VHL Research, Nutrition & VHL, Understanding and managing stress @ $50 $ _____

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

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

☐ in Honor of ☐ in Memory of _____

Acknowledgement card to be sent to: _____

_____

Payment information:

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

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

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

This is a ☐ one-time payment

☐ Monthly pledge to be charged to my card for ___ months.

Name as it appears on the card: _____

Card number: _____ Expiration: _____

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It Takes Us All!

by Camron King, Chairman of the Board

The VHL Family Alliance celebrated 15 years of success in Orlando, Florida this past June. The group gathered for the Annual Meeting and celebration dinner was excited and enthusiastic about where the Alliance has been, where we are today and what the future holds before us. The strength and reputation of the VHL Family Alliance has been fostered by fantastic leadership and volunteers, along with an incredible staff to support our goals and efforts, but as we look to the future we need to be conscious that it will take us all playing an active role in promoting the Alliance, supporting its efforts and moving the search for a cure forward.

We all have different issues in our lives: family, work, school, other activities, but the tie that binds us all is VHL and our Family Alliance. You are receiving this newsletter because of your support and involvement with the Alliance and the efforts it undertakes on our behalf. We all need to continue to support the VHLFA and help it move forward!

As the Alliance continues to grow and progress and we get better and better research proposals to fund, we need to make some strategic investments in the search for a cure. The staff has made sure that they are prudent with the funds generated by the generosity of members and supporters of the VHLFA, but there is always a demand for more: more research needs funding, more education needs to be done, and more advances towards a cure!

This is where each and every one of us comes in! These are tough economic times and for non-profit organizations that rely upon the generosity of people's discretionary support, these times are even tougher. That being said, with a minimal investment of time and request from each one of us, we could easily help the VHL Family Alliance thrive and move forward in a highly sustainable manner. Just think, if we each contributed or raised a small $100, the possibilities of what the Alliance could do would seem endless in comparison to where we are today!

Let's break it down:

There are some 15,000 members of the VHL Family Alliance in the world, half of whom are in the U.S. If each person either contributed or asked friends family and colleagues to help raise a total of $100, we would have a whopping $1.5 million in the bank to fund research, education and promote the VHL Family Alliance! This is almost six times what VHLFA brings in annually! What a great leap forward!

Now, we don't all have $100 to contribute, but if we could each ask 10 people to donate $10 or 5 people to donate $20, we could easily reach our goal and in doing so reach out to many more people and better get the word out about VHL – what a perfect scenario!

This is a simple and effective way to help support the VHL Family Alliance. Should you need further assistance, the fantastic staff is always at the ready to help! Let's all work together and build upon 15 years of success and take the VHL Family Alliance to another level in the future!

See exciting projects from our members, p. 10

See great new learning opportunities, p. 7

Volunteer to add your talent to the team!

Write to chairman@vhl.org -- ***Thank you!***

VHL FAMILY FORUM

Newsletter of the VHL Family Alliance
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