VHL Family Forum

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First in the Family: VHL Mosaicism

by Lindsay A. Middelton, RN, CGC, and Gladys Glenn, MD, PhD, National Institutes of Health (NIH)[1]

VHL is generally inherited as an autosomal dominant trait. In other words, most people with von Hippel-Lindau have a parent who also has VHL. It is currently believed that if a person has VHL, the specific genetic mutation will be found in the laboratory. However these general rules do not apply to all VHL families. There are families in which a child with VHL has parents who do **not** have VHL. Some people with VHL do not have a VHL genetic mutation. And some unaffected parents are known to have more than one affected child. All these situations can be explained by a concept known as mosaicism (mo-ZAY-eh-sizz-em).

In order to understand the concept of mosaicism one needs a basic understanding of several biologic processes. It is necessary to understand a little bit about chromosomes, genes and the nature of genetic mutations. You need also to know how genes are passed from one generation to another at the time of conception; a process known as fertilization. After the father's sperm (with his genes) fertilizes a mother's egg (with her genes), an embryo carrying both sets of the parent's genes develops through a fascinating and complex series of events.

Chromosomes and Genes

Our bodies are made up of trillions and trillions of cells. Each cell (see Figure 1) has a nucleus inside, which contains the chromosomes. The chromosomes carry the genes, made of DNA units strung together. We have 46 chromosomes, or 23 pairs of chromosomes. One chromosome of each pair is inherited from our father and the other is inherited from our mother at the time of conception. Each chromosome is made up of genes; therefore, we have two copies of every gene; one from our father and one from our mother.

Each chromosome is one long strand of DNA: a DNA helix, containing hundreds, if not thousands, of genes. Genes are the basic units of heredity, and line up on a chromosome like beads on a string. We each have around 80,000-100,000 genes that determine what we look like and how our bodies work. A gene is like a recipe; it is a list of instructions that tell the cell how to make proteins. A gene is comprised of a series of 3-letter words and each word refers to a specific protein building block. Some proteins are structural, in that they help to form tissue like the heart or bones. Other types of proteins are critical in the thousands of chemical processes that occur in our cells to make our bodies function properly.

Genetic Alteration / Mutation

All plants, animals and humans have genes that don't work as well as they should. They have been altered or changed by a process known as mutation. The term "genetic mutation" is often thought of as negative or bad, but mutations are the basis of normal human variation, such as long fingers vs. short fingers, red hair vs. black hair, blood type A vs. blood type O, etc. However, some mutations are harmful and cause disease.

Spontaneous mutations occur in everyone, but usually enzymes repair them. Also, normal everyday life exposes us to things that are known to be *mutagenic*, something that alters or changes the spelling of DNA. There are some chemicals that we know are mutagenic. We know that high levels of radiation cause DNA mutations. Very rarely are we able to determine the reason or cause of the genetic mutation in a family. Alterations, or changes, in our DNA can be of several types: a spelling mistake in a single letter (point mutation), a portion of the gene can be missing

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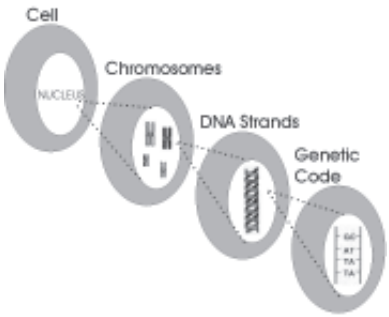


Figure 1: Inside the Cell

We have 46 chromosomes, or 23 pairs of chromosomes. The first 22 pairs are similar in males and females and are called autosomes. The 23rd pair are the sex chromosomes which determine whether we are male or female. Males have an X and a Y chromosome and females have two X chromosomes.

(partial deletion) or an entire gene can be missing (complete deletion).

We all have 10-15 genes that don't work as well as they should; they are altered or mutated. Mutated genes function differently; either they don't work as well as they should or they work too hard. Once DNA has changed in a family, that change may be passed from parent to child in the sperm or egg (germline) during conception.

Pattern of inheritance in VHL

Von Hippel-Lindau is an autosomal dominant genetic condition caused by a non-working gene on the top of chromosome #3. Autosomal dominant conditions usually occur when just one copy of a pair of genes does not work correctly. Because the VHL gene is a tumor suppressor gene, tumors and cysts arise after the second copy of the VHL gene becomes altered or mutated within a cell.

Each offspring of a parent with VHL has a 50% chance, or 1 chance in 2, of inheriting the non-working dominant gene, putting them at risk for the health problems associated with VHL. Males and females can equally inherit the mutated VHL gene. A child who does not inherit the mutated gene will not develop VHL and cannot pass VHL on to the grandchildren.

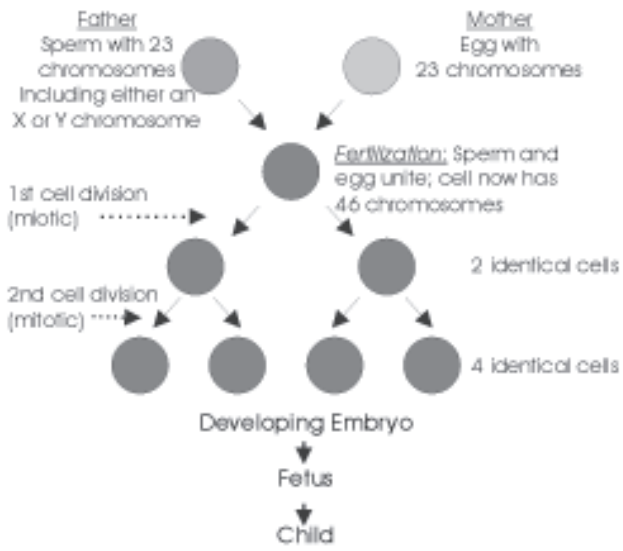


Figure 2: Fertilization

Fertilization and Early Development

Egg and sperm cells (see Figure 2) each contain one-half of the chromosomes through a process called *meiosis*, during which egg and sperm cells receive one copy of each chromosomal pair. Fertilization occurs when a father's sperm and mother's egg unite at the time of conception.

A short time later, within the fertilized cell, the DNA copies itself and the cell divides, becoming two cells, each with identical chromosomes, identical DNA and genes. This is the process of *mitosis* or cell division. The single cell that is formed in fertilization becomes two cells, then four cells; then eight cells, and continues until a baby is completely formed.

Cell Differentiation

In the first few days after fertilization each cell of the early embryo is capable of becoming any type of cell (a blood cell, a nerve cell, a bone cell, etc.). Four days after fertilization the embryo contains around 30 cells; at this point the cells begin to specialize. For example, one of the cells develops into the nervous system; another specializes to become the skin; others become the circulatory system (blood vessels); still others become the kidneys. This is the process of *cell differentiation,* which will have more meaning when mosaicism is discussed below.

Inherited mutation

Most people who have VHL inherited the VHL gene from a parent who is also affected with VHL. We would expect *every* cell of the offspring to have one chromosome #3 that carries the VHL mutation and one chromosome #3 from the unaffected parent who has a correct "spelling" of the VHL gene. At the time of fertilization, the child received the mutated VHL gene from the affected parent, and every cell thereafter will have the same VHL mutation.

New Mutation

In every VHL family the gene mutation began as a brand new event in someone; a grandparent, a great grandparent or a parent. From the time the first heritable VHL mutation occurs, the family from that point on has an autosomal dominant genetic condition that may be passed down through the generations. It is often difficult to determine in a family exactly where, when or why the VHL gene mutated.

Mosaicism

Sometimes we identify people who have two types of body cells. In this case, as it relates to the VHL, there would be some cells with the VHL mutation and some cells without the mutation. This is what we call "mosaicism". The word mosaicism basically means the person has two types of cells, or *cell lines.* Most, if not all, autosomal dominant conditions are known to have some people who are "mosaics" for that condition. If you test many different cells of their bodies, you will find some cells with the mutation and some cells without the mutation. We do not know precisely,

because it is often difficult to be certain a particular person does not a have a second cell line.

To understand mosaicism, let us go back to fertilization. Let us presume that a mother's egg and father's sperm each have two working copies of the VHL gene (that is, the gene is "spelled" correctly). They do not have VHL. Fertilization occurs and the first few cell divisions result in identical cells containing two VHL genes that are spelled correctly. At this point there are 8 cells and each of these cells can become any type of cell; differentiation has not yet occurred. We are now ready to learn what happens when a mutation occurs at this stage.

Somatic Mosaicism

A somatic (so-MAT-ik) mutation (see Figure 3) is a gene mutation that occurs in a cell of the body that is not a germ cell (not eggs or sperm). Because somatic mutations do not come from the eggs or sperm, they were not passed down from a previous generation.

Let us presume that when the pre-embryo is eight cells in number (differentiation has not occurred), the VHL gene in one of these cells is altered or mutated. One cell now has one #3 chromosome that has a VHL mutation, and one copy of the VHL gene that is spelled correctly. The VHL genes in the other 7 cells remain unchanged; they work well. The eight cells continue to divide.

All cells that are derived from the one cell in which the mutation occurred will also have the VHL mutation. All cells that derive from the 7 non-altered cells will be identical; they will not have the VHL mutation. This person now has two cell lines: some cells with the VHL mutation and some cells that do not have the VHL mutation. These cells will go on to differentiate into the various organs in the body.

For example, let us presume the VHL gene mutation occurred after the cells became specialized. Many of the cells are now specialized; let us presume

Figure 3: Somatic Mosaicism. A portion of developing tissue will have the mutated VHL gene. Thus VHL may develop in some, but not all tissue sites.

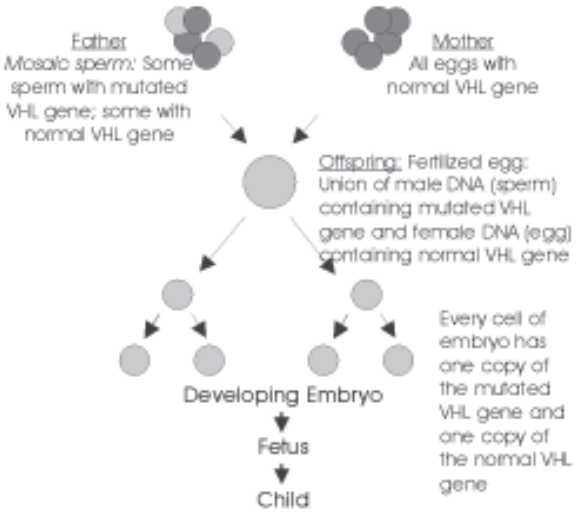


Figure 4: Germline or Gonadal Mosaicism. Some of the egg or sperm cells have a VHL gene mutation.

few cells that become the central nervous system (brain and spinal cord). The cells that become the kidney do not have a VHL mutation, and as such, will not develop VHL-related health problems. However, the brain and spinal cord that have the mutation may very well develop VHL-related problems. In this situation, analysis of a blood sample may not find a mutation, because the mutation occurred in the cells that became the brain and spinal cord.

Gonadal (germline) mosaicism

Another example of mosaicism (see Figure 4) is when a somatic VHL mutation occurs in the germline tissue, creating two types of germ cells. In other words, some egg and sperm cells have the mutation; others do not. This is called germline or gonadal mosaicism. In this situation, the person does not have a VHL mutation in any other cell in the body. Therefore, he or she does not have VHL-related problems, andthere is no family history of VHL. But because the gonads are affected, a portion of this person's eggs or sperm have a mutated copy of the VHL gene. When this person has a child, there is a chance that the child will have VHL.

Possible scenarios:

For the purpose of this discussion, the terms "unaffected" and "affected" will be used. It used to be that a person was determined to have VHL based on "test results". There was only one type of testing; these were the clinical tests such as X-rays, MRI's and CT scans that are done to monitor the organs affected by the altered VHL gene.

However, around 5 years ago, the VHL gene test became available. Now we can "test" a person who is at risk of inheriting the VHL mutation by checking the DNA to see if he or she carries the known gene mutation in the family. When we use the word "test for VHL", it can be confusing. Does this mean "clinical test" or "DNA test"? In this next section, the term

"unaffected" means that both DNA and clinical testing do not find evidence of VHL.

Parents do not have VHL, but have one or more children with VHL:

There are two possibilities. The most likely scenario is that the VHL gene mutated in a cell that gave rise to an egg or sperm, such that the fertilized egg (the very first cell) contained the VHL mutation. The child that is created now has a VHL mutation in every cell. Because the parents do not have the VHL mutation in any other cell, they do not have VHL, will not develop VHL and cannot pass a VHL mutation to another child they may have. A new mutation occurred in a single cell that gave rise to an egg or sperm. That one germ cell conceived that child, but all the other germ cells are free of the mutation. Therefore, the risk of VHL in another child is extremely low.

> "Mosaicism may explain why a DNA mutation can not be detected in a person who has VHL tumors and cysts, or why unaffected parents may have one or more affected children."

Another possibility is that one parent has gonadal mosaicism; some germ cells (eggs or sperm) have the mutation; some do not. In this scenario, both parents are not affected by VHL because the VHL mutation they have is found only in their germ cells. There is no evidence of VHL by clinical tests and no DNA mutation can be found in a blood sample. However, it is possible for this parent to have more than one affected child with VHL. If their child is conceived by a sperm or egg that has the mutation, that child will be affected. If the child is conceived by a sperm or egg without the mutation, the child will not have VHL and cannot pass VHL to his or her offspring. The risk of having multiple affected children (recurrence risk) depends on the degree to which the germ cells are affected.

Some features of VHL, but no DNA mutation detected:

In this situation, we presume that a somatic mutation occurred early in the development of the embryo. Therefore, multiple tissues may carry the mutation, such as the kidney, central nervous system, eye and in some cases including germ cells (eggs and sperm). The DNA test is generally done with a blood sample. If the blood DNA test does not demonstrate a VHL mutation in a person who has VHL, we presume the cell that was destined to become the blood system did not carry the mutation. Because some systems in the body have the mutation, we can expect some features of VHL. But the gene mutation may not be detected in a blood test.

The tissues that are affected depends on where, in the process of development, the mutation first occurred. It is possible for one tissue or many tissues to be affected.

How mosaicism can be detected

If a person has VHL symptoms but a negative DNA test using DNA extracted from blood, other types of tissue can be analyzed. Skin tissue is an easy tissue to obtain and analyze in a research lab. A small sample of skin can be taken by a skin biopsy, which is a simple procedure done under local anesthesia in an office setting. Generally no stitches are needed. Another easily obtained cell type are those found in cheek cells which are obtained by swishing mouthwash for a few minutes and spitting into a cup. Cells from the skin and cheek cells may demonstrate two different cell origins.

It would be helpful in many situations to know whether mosaicism is present. In order to be certain that a person is *mosaic* for the VHL mutation, one would have to test cells in all body systems, which is not realistic. It may be possible in a specialized research laboratory to test sperm for the presence of a VHL mutation, but the sperm cell is destroyed by the analysis.

It is difficult to guarantee that VHL mosaicism in a single individual is not present. Estimates about the frequency of mosaicism in specific autosomal dominant conditions are based on observation of disease occurrence in families and by laboratory analysis of available tissues. We currently estimate the incidence of mosaicism in VHL to be approximately 5%. Currently 23% of the families we are seeing have one person with VHL, with no VHL previously in the family.

Genetic Counseling and medical screening in these cases requires a step-wise approach using DNA testing, and adding clinical examinations as indicated.

Conclusions:

If you are the first in your family to have VHL:

1. If a mutation is found, then there may be VHL mosacism in one of your parents, perhaps at a low level without symptoms.

2. If a mutation is not found, and if the test was done at a lab with a 99% hit rate (e.g. Pennsylvania or Padua),[2] it is possible that you, yourself, are mosaic for VHL, and your parents are not expected to be at risk for VHL. Consult a geneticist to explore this possibility.

4. In either case, whether the mutation is found or not, your children are considered to be at risk until their own DNA test is shown to be negative. See the *VHL Handbook* for suggested screening for children.

5. NIH plans further studies of VHL mosaicism.

1. The authors: Lindsay A. Middelton, RN, CGC, Urologic Oncology Branch, National Cancer Institute (NCI), U.S. National Institutes of Health (NIH), Bethesda, Maryland; and Gladys Glenn, MD, PhD, Genetic Epidemiology Branch, NCI, NIH.

2. When there is no prior history of VHL in the family, it is usually best to ask your geneticist to submit the sample directly to a testing lab with a particularly high "hit rate." University of Pennsylvania, Dr. Catherline Stolle, 215-573-3030 or +1 800 669-2172, Fax: +1-215-573-5892; University of Padua, Dr. Alessandra Murgia, E-mail: alessm@child.pedi.unipd.it, Fax: +39-49-821-3502

Ask the Family

vhlfa@egroups.com is a discussion group among 125 people with VHL in the English language. There are discussions also in Spanish, German, and French, with smaller numbers. Won't you join us? After introducing herself, one member received the following reply from another member:

Welcome to "the family". We are sorry you have to be here but since we all are faced with VHL this is the best place to be. It is so nice to know that in moments of stress and confusion you can go to a site and get information you need, lots of good doctors to turn to, and a host of friends that know what you are going through, because we are all going through it ourselves. Also, knowing that others suffer the same plight sort of lessens the load on your shoulders just by knowing that others know what you are feeling at that moment. On top of that! We have Gale and Maria and others that provide us with factual info, and of course the families that provide you with their own experiences that helps you deal with yours. I am thankful for this site and the people that are involved.

Now you take care, and remember that we are all here for you and your family.

Good Luck. God speed. -- *Linda D., Alabama*

A new member wrote to the Editor: One thing I was concerned about when I started reading these messages was that I may find it depressing reading all the situations everyone is dealing with in their day to day lives.

I was wrong, I find it inspiring seeing all the courageous stories and the support and friendship for each other. Also, on days when I may be feeling down, these stories stop me from feeling sorry for myself.

-- *Neil W., Massachusetts*

Slow Recovery from Brain Surgery

Question: My husband had headaches from VHL hemangioblastomas for many years. His cyst grew quickly over the last year, and his neurosurgeon decided that it was time for brain surgery. He was back on his feet in weeks, and felt great at the 5-week checkup. There is a leak, which the surgeon feels will go away in six months. Meanwhile, however, it causes periodic swelling which causes a lack of well-being, making him generally unable to work. Is this normal? -- *Concerned*

Answer: Just a few thoughts to share. The surgery might have been 'routine' from the doctors perspective, but quite a trauma for your husband's body -- it takes a long time to heal. The removal of the tumor or cyst marks the beginning of the healing process. I found recovery a process of expanding my limits and gradually easing back into a healthy lifestyle. Taking small steps, pushing little by little, trying to do more everyday -- letting my body tell me when limit has been exceeded. Then there is the emotional healing. Considering that your husband is young and otherwise healthy, and probably in good physical condition with his outdoors job, the surgery might have prematurely brought several philosophical and psychological issues to the surface.

Regarding the leaking -- I question everything the doctors tell me. What do they think is causing the leak? Why wait 6 months? What do they expect to happen in the 6 months? What will they do if the symptoms haven't changed in 6 months? Is it common???? Keep asking question. --- *Rob A., Ohio*

Ask the Experts

Mutation Rate

Question: I am doing an oral report on VHL for a genetics class. What is the specific mutation rate for the VHL gene? I realize that the prevalence is about one in 36,000, but for tumors to occur, both genes must become inactive. Is there a published rate for the mutation of the VHL gene? -- *David V.*

Answer: According to our paper in 1991 the new mutation rate in VHL is similar to that in retinoblastoma. Genetic aspects of von Hippel-Lindau (VHL) disease were studied in familial and isolated cases. Complex segregation analysis with pointers was performed in 38 kindreds with two or more affected members. Dominant inheritance with almost complete penetrance in the highest age classes (0.96 at 51 to 60 and 0.99 at 61 to 70 years) was confirmed and there was no evidence of heterogeneity between families ascertained through complete and incomplete selection. The point prevalence of heterozygotes in East Anglia, England, was 1.89/100,000 (1/53,000) persons with an estimated birth incidence of 2.73/100,000 (1/36,000) live births. Reproductive fitness was 0.83. Direct and indirect estimates of the mutation rate were 4.4 (95% CI 0.9 to 7.9) x 10(-6)/gene/generation and 2.32 x 10(-6)/gene/generation respectively. There was no significant association between parental age or birth order and new mutations for VHL disease.

-- *Eamonn R. Maher, Professor of Genetics, University of Birmingham, England.*

ER Maher, L Iselius, JR Yates, M Littler, C Benjamin, R Harris, J Sampson, A Williams, MA Ferguson-Smith and N Morton Cambridge University, Department of Pathology, "Von Hippel-Lindau disease: a genetic study." *Journal of Medical Genetics* 28 (1991) 443-447.

I have a 17-year-old grandson, John, who has VHL. He has had two eye surgeries and one brain surgery. He has tumors on his eye, brain stem, and pancreas. I hope your research will progress in time to save my grandson and all others with VHL from further trauma. I am enclosing a donation. I wish I could do more. -- *Jeanette K, Indiana.*

The Spouse

by Richard H., Copenhagen, Denmark

Some facts

A month or so ago I picked up my wife, Vibeke (say VEE-be-keh), from Copenhagen airport. She had been away for a few days on a business trip.

On the way home in the car I asked her, "Do you remember how many years we've been married?" In some marriages this would be considered a dangerous question. In this case it was just a silly question. Vibeke knows to the day just how long we've lived together, or used a particular brand of toothpaste, and when it was that I last cooked dinner.

Vibeke had the answer on hand. "Just about 18 years!" A reply void of excessive detail so as not to give me the impression that she resented me not knowing.

"Hmm, you get less time than that for murder in most countries." I replied.

Vibeke and I don't always share the same sense of humor. This was one of those occasions. Vibeke has grown used to the fact that I only have a rough idea how many years we've lived together, and subsequently how many years we've been married. It's not that I don't care, I just find it irrelevant.

What caused me to ask the question in the first place was that Joyce Graff had asked me to write a few words on the subject of being a VHL spouse. Therefore I had to do some research. So here are some facts: I have lived with Vibeke for about 20 years. I have been married to her for about 18 years. We have known that she has VHL for about 15 years. I too have lived with VHL for about 15 years.

Fifteen years ago we thought we might have children. We knew Vibeke's mother died as a result of VHL. We knew nothing of genetics but, fortunately, had the presence of mind to meet with a geneticist. The geneticist told us what you already know. We were also told that Vibeke should be checked for symptoms immediately. That year Vibeke had her first operation.

The hospital chair

At the time I was a self-employed photographer. My business was new, and I had to work pretty hard in order to get customers. The day Vibeke had her first operation I went to work as usual. I tried to keep my mind off the operation and what it really meant. At around five in the afternoon I closed the studio and drove to the hospital.

In retrospect I shouldn't have driven at all. I parked the car in the hospital car park and tried to release my safety belt. It jammed. I pulled furiously at it and it jammed even more. By the time I freed myself from the belt the car windows were completely fogged on the inside. I struggled out of the car clutching a plastic bag containing two bottles of wine, gifts for Vibeke from friends.

I made it onto the ward, looking a bit wild with the bottles making plenty of noise inside the bag -- but was stopped by a nurse. She was not about to let a sweaty, dishevelled alcoholic anywhere near her patients! Finally I calmed down and explained who I was and what I wanted. So instead of being escorted to the exit, I was shown into Vibeke's room.

Whilst I may not be able to remember how many years we've been married, I remember far too much of what I saw in that hospital. The sights, smells and sounds have never escaped me – nor has the horror I felt when I saw Vibeke with all those tubes and bags and bottles. I sat down on a chair next to her bed -- and fainted.

This, then, is an article describing what it feels like to be me: a man married to a woman who has VHL. I agreed to write about my experiences because I felt that too few of us, whose loved ones have VHL, ever have the courage to ask for help. I have spent weeks writing and rewriting what you are now reading. It has been surprisingly difficult to convey my feelings without, I hope, giving the impression that I wallow in self-pity. The truth is, I have no self-pity. I have, however, suffered considerably through the years, and I expect many of you reading this will recognize this suffering as being similar to your own experience. I also know that very many of you will have experienced first the hospital chair, and then the hospital bed, as is the case with Vibeke.

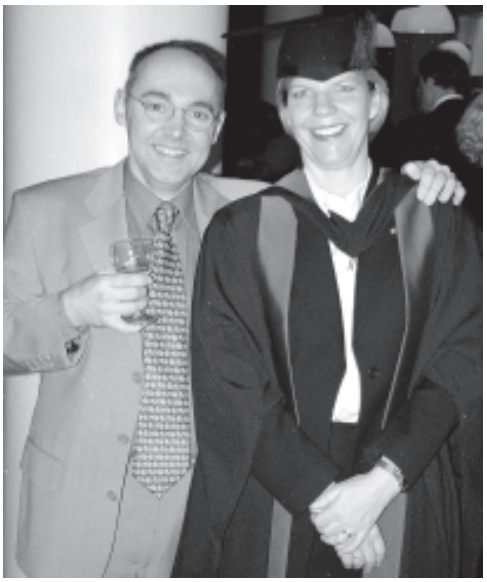
I have drawn exclusively upon events in my life over the past few years and observations I have made of others. I have made an effort to be honest about myself, as well as an effort to respect the feelings of others.

After the operation

For a long time after my first visit to the hospital I was in a condition of shock. I have absolutely no recollection of how I got through those first days. Nor do I ever wish to remember. What I didn't realise at the time was that this was only the beginning of months, maybe years, of despair and depression.

After a week or two Vibeke came home from hospital. She lay in bed for much of the day. I couldn't touch her. I was afraid that anything I did would hurt her. When she got out of bed to go to the bathroom I stood there helpless. I think I could almost feel the pain. When the time came to have the stitches removed, a friend of hers, a nurse, came by and took them out. I was called into the bedroom to see the superficial result of the operation.

Two things crossed my mind. First I was relieved that the scar was not as frightening as I had imagined. Secondly I was heartbroken that Vibeke should have to go through life with a scar from one side of her stomach to the other. I could not imagine that life would ever be normal again.



Richard and Vibeke H. as she received her Master's degree in Business Administration (MBA), Copenhagen, Denmark

I got it all wrong, of course. It has been years since I noticed the scarring at all. Honestly! Incidentally, we discovered that Elizabeth Arden's "Eight Hour Cream" really helps scars to heal well. I remember now that it smells slightly of pepper. It tingles too, I know; I tried it.

I made other mistakes. The biggest mistake being the assumption that I had no right to suffer and to say to others that I was suffering. This is what I told myself: "I wasn't the one with this disease. It wasn't me who had just undergone major surgery, so what did I have to complain about?" I spoke to no one about my worries. It turned out to be a very serious mistake.

Being an Englishman, and therefore completely unable to confide in others about my personal problems, I became withdrawn and emotionally cold. I told nobody, and especially not Vibeke, how I felt. So what you are reading is in some way an historical document. This is the first time I have shared my thoughts and memories of that time, in any detail, with anyone.

Had I been able to talk to another person, things might have not gone so badly. Those first weeks I went to work and tried to look after Vibeke as best as I could. At weekends I would drive her to the country and take her for a walk. We actually called it "taking Vibeke for a walk." I probably needed the fresh air and exercise more than she did. I was suffering from severe stress. I felt that I was experiencing the world through a filter. I remember that even the birdsong sounded alien.

Eventually Vibeke became well enough to return to work. On the surface things appeared to be OK. I knew Vibeke was deeply troubled but I don't recall us talking about it. I expect both of us were thinking about her mother's death, and if Vibeke would also share the same fate. Within months things began to go very wrong. We were not very considerate towards each other. Panic describes it well enough. So we came close to the destruction of our marriage. We had a choice between marriage counseling and divorce.

We visit a psychologist

Before I go on, let me offer you some advice. Finding a psychologist is in itself something of a challenge. I think most of us would rather be seen leaving a strip club in the company of known criminals than be recognized at a psychologist's clinic.

Vibeke and I visited a psychologist recommended by a business friend. Had I stopped for a moment to consider the state of my friend's marriage, I might have chosen someone else. So here is my advice: If you and your partner ever decide to visit a psychologist in order to rescue your relationship, try to choose one who actually believes in marriage.

As I recall, one of the first things he asked about was the state of our finances – a not so subtle way of making sure we could afford him. Then he stated that he did not believe in repairing damaged relationships.

Looking back on it all, it was really funny. I have never discovered if it was just a ploy on his part, to pull us back together, or whether he was just hopeless at his job. The only thing I remember well is his statement that "normal people are boring." That was alright with me. I have been called a lot of things in the past, but I've never been accused of being boring.

Anyway the three of us sat there for forty-five minutes every week for a couple of months playing mind games with each other. Afterwards Vibeke and I would go shopping as if nothing had happened. In the end we grew tired of visiting him. We told him so, and he pronounced us healed. He had the good taste not to send his bill immediately.

As unintentionally amusing as they were, the visits to the psychologist only provided temporary relief. A dark cloud settled over my mind and remained there for a long time. I suppose any competent psychologist could have told me what I was wrong. On reflection, I think it was the same thing that I noticed Vibeke suffered from, a long time after her mother died. What I saw then was a person devoid of joy. She rarely smiled. She was distant. She merely functioned. And then one day, slowly, very slowly, she started living again.

We are different

One of the major differences between Vibeke and myself is her ability to see ahead. It was this that kept her alive and, I believe, saved her. She has always set goals for herself, and I have never known her not to reach them, sooner or later. To improve her ability to cope with physical stress, Vibeke started running. At first she ran or walked four kilometers. She also decided to study to improve her career choices. Today she runs ten to fifteen kilometers several times a week. Two years ago she got her MBA. Her business trips have taken her to every continent on earth, apart from Antarctica. Anyway, if you've seen one penguin, you've seen them all.

I am different. I have always been inclined to worry,

often for no good reason. From the very first day, when we were told that Vibeke had to have surgery, I entered a state of despair. My wife was seriously ill. My business suffered from a combination of economic and mental depression. I was alone. On the surface I must have appeared normal. Yet inside I had absolutely no idea what to do, or where to get help.

I also experienced some astonishing, and perhaps predictable, behaviour from other people. Quite a few asked if there wasn't a cure for VHL, so I had the laborious task of explaining that genes were genes and couldn't be favourably altered with a bottle of medicine. Vibeke's father tried to pretend that Vibeke couldn't be ill because, as he said, she did not have the same personality as her mother. He was thinking of the effects of a brain tumour. I remember him drinking heavily when his wife died of VHL and we often got phone calls at night from bars. We would then have to go and pick him up and drive him home.

After a while he stopped getting drunk. He developed a latent interest in sailing and surprised us all when he took lessons in seamanship, bought a boat, and turned out to be an excellent sailor. He is currently learning how to use a computer at the age of 65. The most remarkable thing about him is that he now regularly visits one of our VHL association members, a woman who is confined to bed and dependent upon a respirator.

Another person I knew stopped me in a shop one day and said that as a photographer I shouldn't be so "negative" and that I should be more "positive." He actually thought he was amusing. It's true, it really happened. I think it was one of the few occasions in my life where I would have been justified in striking another person. As it was I was so shocked by what he said I stood there dumb struck.

Then came Vibeke's second operation. At least I didn't faint when I visited her in hospital that time.

Adapt

I am either a slow learner, or I have seldom found good teachers. Following Vibeke's first operation, our near divorce and then her second operation, I seemed to spend years just existing. It took me quite some time to understand that I really could have some influence on my own life and that it was me who had to take the first step.

It also took me some time to be honest with myself about my abilities. I sold my business and spent a couple of years working for others and trying to pay off my debt to the bank. Fortunately for me things didn't work out too well and I ended up unemployed. I know it sounds strange to use the word "fortunately" – and I certainly wasn't happy about it at the time – but I can't imagine how boring life would have been if everything had gone well.

At that time I had been working for twenty years and had never been without a job. Unless you have tried it you cannot believe the damage unemployment can do to your self-esteem. Following Vibeke's illness, being unemployed was the second great crisis in my life. And there is something else. I think I was jealous of Vibeke. Despite having VHL, and the near break-up of our marriage, Vibeke was doing very well. She was studying, passing exams and pursuing her career. Yet when I looked at myself I saw a middle-aged, unemployed business failure. It took me quite some time to change my attitude about myself and my abilities.

I decided to study computer graphics. I had seen an interesting brochure advertising a course and decided to take a chance -- perhaps I had learnt something from Vibeke, after all? What I did was to take the first step towards taking control of my life – although I didn't understand it at the time. In fact, I was in the process of learning the simple Darwinian law: Adapt or die.

Whilst I was studying, Vibeke was offered an exciting job in Copenhagen. At the time we were living way out in the county in another part of Denmark. We sold our house and made enough profit on it to pay off my bank debts. Eight long months later I had a job. It sounds so easy now, but in reality I had just come through some of the worst years of my life. I had to do it on my own, too.

Choose life

I wonder how other cultures react to death and disease? The culture that I have been brought up in is quite simply hopeless at dealing with it. These two phenomena are, apart from sex and birth, the most common occurrences in life -- and yet we react as if they were entirely alien to us. I can only recall one occasion when I was asked how I felt. I replied, as far as I remember, with a lie.

During recent years I have become aware of another fact of life, which has greatly influenced me: I too could die young. I know it sounds like a platitude, but life is far too short to waste. So consider this instead:

Five men I know are dead. They were all relatively young men, in my age group. The printer who had his office next to my studio died of cancer. Two colleagues, both photographers, are dead. One died from cancer, the other from a brain hemorrhage. Another man I knew, from Iran, died of a heart attack. I remember him telling me that, all things considered, he preferred being beaten by the Shah's men, to being tortured by the Ayatollah's men. This kind man, with a dark and wonderful sense of humor, just dropped dead one day. Recently a graphic designer I had worked with some years ago, died one evening, also from a heart attack. He is buried in a country churchyard overlooking the fields, hills and valleys that he loved so much. Yet I am still alive and so is Vibeke.

I am not one of those insufferable optimists who go around telling others to "cheer up and look on the

bright side." I certainly don't try to fool myself with artificial optimism any more, and I wouldn't recommend it as a solution to anyone else. What I do recommend is that when you find yourself sitting in the hospital chair you should say "I feel dreadful, and I make no apologies for saying so!" And if your marriage is suffering as a result of the unbearable cruelty of fate, then say so! Do something about it! Things won't improve overnight – they never do – but you will have taken one step in the right direction. You will start living again. It is also very likely that you will avoid the mistakes that I made, which caused me to waste so many years of my life with bitterness and depression.

Today Vibeke and I live well together. We both enjoy interesting careers and we are able to support each other, financially as well as emotionally. Sure, we fight on occasion – who doesn't? We also plan for the future, sometimes years ahead – who doesn't? We are also well aware that one day Vibeke may have another operation – or maybe this time it will be me who becomes ill. Maybe one of us will even die. Who doesn't?

Resources

Hot Braille. At the HotBraille.com website, a small group of tech-oriented young men are doing good online. The service allows people to send letters, notes and other printed information to visually-impaired friends or family members who read Braille. You send e-mail and the HotBraille team converts it to Braille and mails it out, all for free. http://www.hotbraille.com

The Gift of Pain. I have been reading a book called "The Gift of Pain" by Dr. Paul Brand, who has worked with Leprosy patients in India. He also has worked with patients with RSD, a syndrome which causes a person to over-respond to trauma, by having a stiff and painful foot or hand. He has had some interesting observations on pain....He tells of a hospital in India where there was a shortage of nurses, so patients were asked to help each other. They all asked for less pain medicine..then when the staffing was worked out, the patients had nothing to do but to focus on themselves. They all asked for more pain med again!!!! Food for thought!!! I know that when I am in a lot of pain and I go to work and care for those who have similar pain, I forget about mine and focus on helping them. Good medicine is helping others. -- *Deb H., Massachusetts*

Capitol Hill. To see what's happening in the U.S. Federal government, to find a senator or congressman, see http://www.fedbuzz.com You can file health discrimination complaints online, get information on Occupational Health and Safety (OSHA), posters, free literature, and access to all branches of the US Government.

Happy Father's Day!

About 25 years ago I was placed up for adoption in the state of Pennsylvania. My birth parents must have made the hardest decision of their lives in doing this, but today I thank them. For any of you who believe in a higher power, please continue to read this.

A couple of months after I had been placed with my adoptive family, my birth father added a medical note in my file telling me that his family had been part of a study on VHL at Duke University. Little did we know at that time that he would become my saving angel. Last month I started having "brain to feet" problems. After going through four doctors, I finally found someone who knew about VHL and took me under his wing. After a full MRI this week, I should know whether or not his assumption of a VHL-related growth on my spine is the cause.

Long story, short message. Even if I never get to meet this incredible man, I need to publicly thank him for being such a wonderful human being. I thank him for my life, both in June of 1973 and in April of 2000. Without his foresight in 1975 when he placed the note in my file, I would still think I was going crazy. Thank you, Dad, and I love you, whoever and wherever you are. -- *Heather Ann, DOB 6-29-73, Lancaster, Pennsylvania.*

Dear VHLFA,

Last fall our seven-year-old son was diagnosed with a retinal hemangioma. We were told that he might have von Hippel-Lindau disease. He has since had the hemangioma treated by laser surgery. Four weeks ago a blood sample was drawn and sent to Philadelphia. We learned today that the laboratory found no sign of the VHL genetic flaw.

I write this letter to you and enclose a small contribution to say how much we appreciated your web page. I also enclose a check from my mother who shares our support for the work you are doing. I can say without hesitation that your web site gave my family all the information we needed and answered all of our questions. The information both enlightened me and through greater knowledge comforted my wife and me. The information helped us make more informed decisions on how to proceed with the treatment of our son's condition, and whether we should use genetic testing.

We are much relieved that it is almost certain that my son does not have VHL. However, for the short time between discovery of the hemangioma and the receipt of the test results, I think we experienced a little of what many families have gone through who have dealt with VHL. My heart goes out to everyone affected by VHL. It has been a long four months.

We hope this small donation can be used to support your work and we pray for the continued progress in finding a cure for this disease. ***Thank you!***

-- Frank R., Wisconsin

Getting Help with Depression

by Dan Kavanaugh, M.S., social worker, National Institute for Neurological Disorders and Stroke (NINDS), Bethesda, Maryland, one of the U.S. National Institutes of Health

Last summer my youngest daughter broke her leg in a playground accident when another child ran into her. We took her to the physician and the X-rays confirmed a break. The healing process began when the cast was placed on her leg. What if I had taken my daughter to the physician and instead of putting a cast on her leg he had just said "That's understandable that she broke her leg." Suppose the physician had left it at that — no further medical intervention, no cast, just the words "That's understandable." Wouldn't that be a fairly shocking response? Yet when a person who is diagnosed with a chronic illness begins to express feelings of depression, they are often met with the words "That's understandable" and are not offered the opportunity to have the depression treated.

Prevalence of depression

Depression is one of the most common complications of chronic illness. According to one survey it is particularly common in those with recent heart attacks (45%), hospitalized cancer patients (42%), recent stroke survivors (40%), and diabetes (33%). In fact it is not unreasonable to view chronic illness and depression as a two-way street — a diagnosis of a chronic illness can be depressing and the increase in depressive feelings can exacerbate the illness. The risk of depression increases in proportion to the severity of the illness and the life disruption it causes. My daughter's broken leg was certainly sad and it was disturbing to see her in pain, but being diagnosed with a life-threatening tumor can be severely depressing.

Risk of depression

The risk of depression also increases with the possible complications of a chronic illness. A person may be successfully managing a chronic illness, but the illness may have so many possible complications (frequent hospitalizations, depletion of financial resources, decreased ability to maintain gainful employment) that even one who is in control of the disease may have a high rate of depression.

Signs and symptoms of depression

What are some of the signs and symptoms of depression?

1) Depressed mood most of the day, nearly every day

2) Markedly diminished interest or pleasure in all, or almost all, activities most of the day nearly every day.

3) Significant weight loss or gain when not dieting or decrease/increase in appetite nearly every day

4) Insomnia or hypersomnia nearly every day

5) Fatigue or loss of energy nearly every day

6) Feelings of worthlessness, or excessive or inappropriate guilt, nearly every day

7) Diminished ability to think or concentrate

8) Recurrent thoughts of death (not just fear of dying) or of suicide without a specific plan, or a suicide attempt or specific plan for committing suicide

As you read the list of symptoms above it is important to keep in mind that many physical illnesses produce some of the symptoms of depression. Cancer, for example, is associated with weight loss, sleep disturbance, and low energy.

That is why it is imperative that a person who is otherwise known to be healthy get a medical workup when they are experiencing these types of symptoms. Often once the presenting physical ailment is treated, then the depression is also eliminated.

Deciding whether or not to seek professional help

There are times when a person who is experiencing depression will get better without treatment. How does a person decide whether to seek professional help? Some questions for the person to consider are:

1) Is the distress level intense enough that they want to do something about it?

2) Do they feel that they are no longer able to problem-solve on their own? Do they feel the need for more support?

3) Is the level of distress such that it is negatively affecting their relationships, usual activities, or work? and

4) Are they contemplating suicide?

A person who answers yes to one or more of these questions may benefit from entering a counseling relationship with a mental health professional.

Who treats depression?

What types of professionals treat depression? Psychiatrists, clinical social workers, psychologists, and psychiatric nurse specialists are the primary treatment providers for depression. In addition, there's a wide range of professionals who can also help people with depression, including members of the clergy and school guidance counselors who are trained to detect the disorder, and can provide referrals.

Treatment options

There is a wide range of treatment options for depression, but they can generally be divided into three categories: Antidepressant medication alone, psychotherapy alone, or a combination of antidepressant medication and psychotherapy.

There are a variety of antidepressant medications available, but they can be separated into three main categories: Tricyclics (TCA's), Monoamine oxidase

inhibitors (MAOI's), and Selective serotonin reuptake inhibitors (SSRI). Medications within these categories work differently on the brain and have different side effects. Unfortunately, there is not a definitive way of knowing beforehand which medications will be most effective. A depressed person may have to try a few different medications before finding one that is effective. This is not to say that prescribing medications is just guesswork. Certain depressive conditions may respond better to particular antidepressants. It is important that the physician and patient work closely together in order to determine an appropriate regimen.

The term "Psychotherapy" refers to psychological treatment for depression. Under the broad category of psychotherapy there are over a hundred schools of "talk therapy." The two main categories within these different schools can be classified as insight-oriented therapy and cognitive-behavioral therapy. Insight-oriented therapy is focused on helping the patient to gain a greater understanding of their unconscious motivations and increase insight into the root of the problem. It is focused on reviewing the past for clues into current behavior. Cognitive-behavioral therapy is more focused on the present; looking at current behavior and thought processes, and how to change behavior and thinking that may be contributing to depressive feelings.

"How do I know which practitioner to see and which type of therapy will work for me?"

Basically, it is impossible to say that any one specialty is superior to another. The main difference is that only psychiatrists can prescribe medications. However, these professions often work collaboratively to treat a person. For example, a clinical social worker may work in conjunction with a psychiatrist where the social worker provides the psychotherapy (or counseling) and the psychiatrist takes responsibility for prescribing the appropriate anti-depressant.

It is difficult to predict which type of therapy will work for any one person. The most important factor to consider is that you are working with a person who is willing to explore a variety of treatment options. A person may encounter health professionals who will say "medication only" and others who might say "psychotherapy only." My own view is that practitioners ought to tailor the treatment to the needs of the patient and not enter into the healing process with an "either/or" view.

How do I find a mental health professional in my community?

A good place to start may be with the physician who knows you best. You are probably not the first person to ask this question, and he or she may have a few names to recommend. If you have health insurance, check to see if mental health services are covered. For a person who is taking antidepressants and engaging in psychotherapy they may find that the medication costs are covered or reimbursed, but that the psychotherapy is not. While there are moves at the state and federal level to achieve greater parity between treatment for physical and mental health it is safe to say that coverage for mental health services such as psychotherapy are more restrictive than medical treatment for illness. Some policies may cover individual therapy, but not group or family therapy. The bottom line is to find out what your insurance will pay for or will not pay for. If cost is not an issue a person may choose the professional they believe will be most effective in treating them and agree to self-pay and not submit for insurance reimbursement.

I have found that there are many agencies across the country that provide excellent services to people coping with depression as it relates to chronic illness. These agencies are usually a mixture of private and public funding and have a sliding-fee scale. That is, the amount the patient pays is based upon income and is not a flat fee. While there is not enough space to list all the agencies that I have referred people to over the years, I find Jewish Social Services and Catholic Charities to provide high quality counseling to patients who could not otherwise afford the usual fees charged for counseling (up to $100 an hour or more in some parts of the country). Even individuals in private practice may be willing to consider a fee lower than their established rate, but you will not know unless you ask.

One can also contact the following professional organizations than can assist in finding a mental health professional in the United States:

- American Psychological Association 1-800-964-2000
- American Psychiatric Association 202-682-6000
- American Psychiatric Nurses Association 202-857-1133
- National Association of Social Workers 1-800-638-8799
- Cancer Care 1-800-813-HOPE

Similar resources exist in other countries.

Dan Kavanaugh, LCSW-C, BCD, is a clinical social worker at the National Institutes of Health Clinical Center, Bethesda, Maryland.

Sleep

by Rob W., Minnesota

Thick liquid slumber lumbers
as my conscious mind tumbles.
In a chaos of colors and lights
my soul slowly, silently slips into flight.

My soul is free to be all it wants
but is limited by the memories of the body it haunts.
My past is its master and controls my soul's fate
awaiting freedom from my body, its mate.

My body is frozen, as if by God's will,
although its essence controls my dreams still.
Muscles and skin are steeled to the bone
while my soul, in its own world, is set free to roam.

When morning comes and my body awakes
it recaptures my soul from its evening escape.
My body and soul, again, unite
for my soul has been grounded from evening flight.

To the School Nurse

by Camille M. Wendekier, R.N., Pennsylvania

Ms. Wendekier, a school nurse herself, wrote this paper for a degree program. There are a number of VHL families in her local area. The amount of time that the school nurse is able to devote to these activities varies greatly from one community to another, but parents should look to the school nurse as a possible resource.

The school nurse has an essential impact in helping students with VHL by tending to the medical needs of the student, educating the student and their families, and acting as the liaison for the student to the school system and the community.

Since VHL is not being diagnosed until adolescence in the local community, interventions in the educational arena will be limited to the high school student body.

The school nurse must first meet with the student and the parents to determine the level of involvement they would like the nurse to have in the student's care, and the level of privacy they wish to maintain. Disclosure of the student's medical status to teachers or other members of the school is not appropriate without the agreement of the student and the parents.

The school nurse can assess for any clinical symptoms of disease progression. This will include tracking complaints of headaches, muscular weakness, gait disturbances, back pain, visual disturbances, profuse sweating, etc. This can be achieved through student interviews, teacher interviews (if appropriate), and monitoring office logs. Because of the tension resulting from academic achievement and organized sports, it is imperative to routinely screen blood pressure and monitor the student after challenging events to assess for the presence of pheochromocytomas. Should recurrent symptoms of VHL persist at school, the nurse will advise the family to follow up with their doctor.

The school nurse also acts as the liaison for the student to the school system when environmental changes are necessary to accommodate physical disabilities. Whether it is rearranging seating in the classrooms, installing adaptive equipment, or acquiring a learning support aide, the nurse fosters a smooth transition for the child's adaptation to his or her disabilities. If necessary, the nurse initiates an Individualized Educational Plan (IEP) meeting to accommodate these needs.

In addition to managing the medical needs of students with VHL, the school nurse can assist to neutralize any potential psychosocial quandaries. Erickson described the primary task of adolescents as the formation of an identity and overcoming a state of role confusion (Ormrod, 1998). A diagnosis of a chronic condition such as VHL can impede the process necessary for proper identity formation in the following ways. First, students at this age are very sensitive to body image. Any resultant disabilities or the diagnosis itself can foster a negative self-concept (Kaluger & Kaluger, 1984). Also, if not properly informed on VHL, the teachers may form negative attitudes toward the student. Not only would this be a source of tribulation in school, but may induce negative judgments of peers toward the student (Wallace, Patrick, & Parcel, 1992). Second, teen years provide the opportunity to prepare for a productive role in society. The possible disabilities resulting from VHL may intimidate these children when preparing for future careers. Third, in the state of role confusion, adolescents develop an emotional independence from their parents and the presence of a chronic disease can impede this process (Broadwell Jackson & Saunders, 1993). Fourth, teenagers are developing physical and emotional control of their sexuality (Kaluger & Kaluger, 1984). A genetically transmitted condition can have a negative influence on the child's sexual identity and foster unfounded fears for these teens.

Because students learn more effectively in a supportive classroom, the school nurse may be called upon to help the teachers form constructive and supportive attitudes toward VHL. (Ormrod, 1998). For example, if a student were exhibiting inappropriate behavior in the classroom, the nurse might intervent to encourage the student to vent fears and frustrations. If this failed, and if the parents agreed, the nurse might share with the teacher some insights into the issues the student is coping with, in an effort to facilitate confident and constructive interactions between the teachers and the affected student. Care must be taken to assess the teacher's own prejudices toward medical situations, to ensure that the teacher not lower expectations of the student, or not encourage a talented student to pursue a career in law or medicine. It is imperative to promote the student to establish and strive toward appropriate goals. It is also important to note that a diagnosis of VHL does not necessarily imply that this person will ever have physical deficits. The school nurse can assist the student in making informed decisions when preparing for his or her future.

The school nurse will also assist the student and family cope with the ramifications of VHL through education and community referrals. When possible, the nurse will assist the student and family to find authoritative information about VHL and supplement this education by providing handouts such as materials from the VHL Family Alliance. The school nurse will also encourage both the student and family to work with their doctor. This will help to ensure that the student receives appropriate treatment to ward off disabilities. Referrals to agencies such as the VHL

Family Alliance will provide the accurate information and support needed for these families.

Armed with the necessary knowledge, the student and family know their risks and can take actions to minimize the impacts on the life of the student. This proactive tactic can prevent the secondary disabilities that may result from untreated VHL.

If necessary, the school nurse will initiate a referral to the school psychologist. This will help the student in two ways. First, the school psychologist will promote the emotional development necessary to discern the student's identity from the family. Second, it will allow the student to vent fears and frustrations produced from this genetic disease. This counseling can positively influence the student's personal, sexual, and social identity. Consequently, the student will develop healthy coping mechanisms and will be less likely to engage in risk-taking behaviors.

The diagnosis of VHL permanently changes the lives of the affected individual and their family. These families need to convert their fears and apprehensions into positive coping mechanisms. The school nurse can facilitate this through apporopriate education and referrals. By managing the physical and emotional needs of affected students, the school will be viewed as a supportive, warm, and inspiring environment. Consequently, students affected by VHL will experience increased academic success that will enhance their future contributions to society.

This issue is dedicated to: *Mary Louise Mahan of Tacoma, Washington, who passed away in April 2000. She is remembered with great fondness by her family and friends.*

Thank You for contributions from...

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R. C. Williams, Oregon
Dr. Gary Wood, Florida
Beverly Yoder, Illinois

Pansies for Hope

Hope sees the invisible, feels the intangible and achieves the impossible.

Help achieve the impossible!

Give to advance research on VHL! see page 15

"Whenever I'm upset, I invite my very close friends to attend my "planned hysterical moments." Then I pick myself up, dust myself off, and go dancing or something. -- *L.M., Penn.*"

In Honor Of . . .

Matt & Monica Andjus by Mr. & Mrs. Floyd Adams, Foust Radiator Company
Our daughters Erika and Amanda, by John & Marilyn Barcus
John Barcus, by Ellen & Frank Barcus
Kelly Blazich, by Karen Frame
Scott & Emma Borg, by Mr. & Mrs. Floyd Adams, Foust Radiator Company
Andy Braden, by Paul & Virginia Braden
Sharry Brydon's birthday, by Mary Frances Duval
Susan Burgoon, by Barbara Smith
Chastity Carroll, by Debbie & Jeffrey Free
Mary Cawley, by Patricia Foye, Susan Ryan
Alice Coday, by Stephanie Armacost
John Conarpe, by Mr. & Mrs. J. Ernest Starmer
Theresa Fruecht, by Paul & Virginia Braden
Damon Graff, by Don & Phyllis Freed, Karen Freed, Marian Hege
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Charles Hall, Jr., by Colleen Venture
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Thank you for contributions . . . in Honor Of . . . (continued from page 13)

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Jay Platt, by Karen & Doug Murray
Gladys Rodriguez by Lissette Rodriguez
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Tim Walter, by Joseph Chapko
Lindsey Webster, by Ellen & Frank Barcus
Dr. & Mrs. Harry Wilcox, by Sally Pope Overman
Regina Willey, by Audrey Clifton
Rob Wright, by Ellen & William Wright

In Memory Of . . .

Rondo Anderson, on his birthday, by Amy Anderson, Willie & Anne Morillon
Anne Barry, by Denisa Kelliher, Henry Ploski, Albert Varnerin
Fred Barry, by Kathy & Paul Neimann
Brian Beaulieu, by Constance Fiala
Edna M. Bentz, by H. W. Maier
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Maria Tatro, by Dresser Masonelian
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Chuck Mayo, by Dawn Cerf
Meg Irvin Moody by Mary Karys, Nancy & Tom Lusk
Carl Nelson II, by Jeffrey Bain, Elaine Boudette, Neil & Jeanne Coughlin, Don Fleischer, Georgine Lyons, Kathleen O'Brien, Keith Staples
Lorna Newell, by G. Baker
Raymond O. Puller, Sr. & Jr., by Marguerite Golding
Dr. Leo Rauch, by Gila Ramras Rauch
Brenda Ray, by Arthur Adams, Paul & Gloria Buckel, Dolores Gramling, J.W. & June Harper, Yushin Lee, Dan Osolinski, Gregory & Lisa Ray, Barbara Siebert, Steven Stoner, Sherry Wichmer
Mathew Rider, by Rita Miller
William Ronald Roche, Sr., by Patricia Brennan, Jane Brewer, J. Calvin Jenkins, William Jenkins
Denise Rouleau, by James & Sheila Lunny
Theodore Strom, by Norbert & Jean Reig
Dr. Samuel E. Tisherman, Pennsylvania
Elnora Voth, by Donald Voth
Joseph & Craig Warnick, by Karen & Doug Murray
Thomas Werner, by Joan Drake
Keith Willie, by Trudy Willie Woods, Australia

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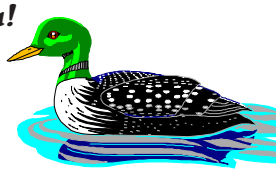
Register before June 28 to get the Early Bird Discount!

Late? Don't let that stop you! Please join us in Minnesota!

The complete program is posted at http://www.vhl.org/conf2000



DONATION & MEMBERSHIP INFORMATION

Please check one: I am a ❑ Person with VHL ❑ VHL Family Member ❑ Supporting Friend
❑ Health Care Professional (physicians nurse, social worker, etc.)
Occupation, or medical specialty: _____ _____
All members receive the VHL Family Forum. Check here ❑ if audio version is needed

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Membership:

❑ New member ❑ Renewal (Dues $25 per household, $35 per professional) $ _____
❑ I cannot afford dues at this time, but please accept my donation (any amount)
❑ Please send me ____ brochures about VHL Fund for Cancer Research to share with others

Donation:

Tax deductible donation of . . . *(amount)* $ _____
❑ In Honor of _____
❑ In Memory of _____
❑ Please send a card to: _____
_____

Purchase merchandise: (proceeds help the VHLFA!)

❑ #___Copies of *How Are You?* by Patricia Foote @ $17 each $ _____
❑ #___Copies of *Sacred Process* by Karen Koenig @ $15 each $ _____
❑ #___Copies of the *VHL Video* @ $20 each, (specify __ US or __PAL format) $ _____
❑ #___VHL Teamwork Pin in ❑ silver ($17 each) or ❑ gold filled ($27 each) $ _____
❑ #___Copies of Pierre Jacomet's CD of Bach's Goldberg Variations @ $12 each $ _____

VHL Symposium registration (see page 16 for information)

Prices include breakfast and lunch on session days, dinner Friday evening, and handout sets.

❑ #___Basic Science Day only @ $70 per person $ _____
❑ #___Track 1: Full Symposium Thursday thru Sunday @ $225 per person $ _____
❑ #___Track 2: Physician Track Friday thru Sunday @ $ 180 per person $ _____
❑ #___Track 3: Family Track Friday thru Sunday @ $ 170 per person $ _____
❑ #___Companion dinner tickets Friday evening for people not attending the sessions + $ _____
❑ #___Add $35 CME certificate fee if credits are desired + $ _____
❑ #___Subtract $20 per person if ordered before June 28 - $ _____
❑ #___Subtract $20 for second person in family ("spouse discount") - $ _____

Payment Method:

❑ Enclosed check, payable to the VHL Family Alliance ***TOTAL:*** $ _____
❑ Master Card/Visa Card #_____
Expiration date: _____ Name as it appears on the card: _____
Signature _____

Send this form to VHL Family Alliance, 171 Clinton Road, Brookline, MA 02445 USA
Canadians, send donations to Canadian VHL Family Alliance, RR 1, Renfrew, ON K7V 3Z4 Canada

Chapter News

Fit Fest! Maria Shipton represented VHL at a booth at the Philadelphia Fit Fest, sponsored by NBC TV News, Channel 10 Philadelphia. Maria gave out brochures and chatted with hundreds of people. It was a wonderful opportunity to raise consciousness about VHL. Among those she met were three celebrities: Seth Peterson, who is the son on *Providence*; Maureen O'Boyle, from *Extra*; and Chris Meloni, from *Special Victims Unit*.

New England. Fred Turner, chairman of the New Hampshire chapter, delivered a talk to a group of genetics professionals in Providence, Rhode Island, on a consumer's view of DNA testing. His presentation was very well received, and stimulated much discussion. Most of the attendees had never had the chance to talk with an affected family before this.

Wisconsin Chapter

Mary Kay S., Chapter Chair for Wisconsin, is very excited about her duties and responsibilities. She is 41 and lives in Cottage Grove, Wisconsin. "VHL was diagnosed in 1997 in my left eye. In May 1998 other cysts and tumors were discovered. In struggling to accept further complications of this disease, I reached out and found the VHL Family Alliance. I don't know what I would have done without this group! After getting back on my feet, I wanted to become involved to help others.

"My father had VHL and unfortunately died at the young age of 33 in September 1967. With my work in helping others in Wisconsin, I am doing it all in honor and memory of my Dad. He missed the technology we have today that enables us to live somewhat normal lives. I want to lengthen people's lives and reduce the need for surgeries. My dream is to see a cure in my lifetime.

"I am looking forward to meeting everyone with VHL in the state of Wisconsin, and working with you to make more contacts in the medical community, raising their awareness of VHL, and helping to create an outstanding supportive environment for people with VHL in Wisconsin. We have some wonderful doctors here. Let's make sure they know about VHL, and about how to help us manage our health and enjoy life!" Please write to her at marykay@terracom.net, or reach her via the hotline, 1-800-767-4845.

Conference 2000!

The early-bird deadline for registration is June 28! Be sure to register and book your hotel rooms before this date to get your discount. You are can even register onsite at the conference, but pre-registrations help us plan food and chairs.

This year's Medical Symposium, to be held at Phillips Hall on the campus of the Mayo Clinic, July 20-23, 2000, promises to be a very exciting meeting. Mayo will award 25 CME's to registered physicians who attend all sessions.

We have designed the meeting in three "tracks" to meet the needs of attendees with varied backgrounds in medical terminology and genetics.

Thursday, July 20: Basic Science Day for those with a genetics background.

Friday thru Sunday: Sessions for physicians and families on clinical aspects of VHL.

Hotel space is reserved at the Kahler Grand (1-507-282-2581, $75/$85), and the Rochester Marriott (1-507-280-6000, $119/$129). Reservations must be made by June 28 to qualify for these conference rates, which are about half the normal rates at these hotels. Call 1-800-767-4VHL or 617-277-5667 for a conference brochure, or see http://www.vhl.org/conf2000 ***See you there***!

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Brookline, MA 02445-5815

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