VHL Family Forum

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Retired Marine Fights for our Lives

My name is Joseph A. "Jay" Platt. I am 32 years old, recently retired from the U.S. Marines after 14 years of faithful service. I am hiking the Appalachian Trail from Mt. Katahdin, Maine, to Springer Mountain, Georgia -- 2,160 miles of wilderness trails

I have a condition called Von Hippel-Lindau, or VHL. The disease affects various organs. I have had my left eye and left testicle removed as a result. I also had brain surgery in July 1997, in which three tumors were removed. Next, I had a partial nephrectomy performed on my left kidney in August. I currently have more tumors on both kidneys, a tumor on my cerebellum, multiple cysts in my pancreas, and several cysts in my spinal cord, all of which will probably have to be removed eventually. With this prognosis, it was decided by a Medical Board that my career as a Marine was to be ended.

I understand and respect that decision. A Marine must be able to be deployed anywhere in the world at a moment's notice, and with my current health status I no longer meet that requirement. The Physical Evaluation Board, or PEB, that makes the decisions concerning discharge for medical purposes found me "Unfit For Duty" and instructed that I be discharged without any benefits other than a small amount of severance pay. This would have meant that I would not receive any medical care, disability payments, etc. Under the current guidelines set down by the Department of Defense, the PEB decided that my having VHL was not caused or aggravated by my military service.

VHL is considered to be a "genetic" disease, as are many others, including diabetes, heart disease, breast cancer, colon cancer, etc. This simply means that one has a predisposition toward a particular disease. Certain diseases, and I would learn that VHL is one of them, are for some reason placed in a category that is considered to be "Not the fault of the service." According to the DOD policy, these certain disorders constitute a pre-existing condition. Technically, it is termed "Existed Prior To Entry," or EPTE. This is the case regardless of whether the military member had any idea that they carried this faulty gene or not upon entry into the military.

I happen to fall into that situation. No one else in my family has ever been diagnosed with this disease and I showed no symptoms of it until I had been in the Marine Corps for about three years. Upon my diagnosis in 1987 (when I became suddenly blind in my left eye*) I was allowed to continue to serve because I was able to perform all the duties of a Marine. After serving my country for my entire adult life, I was shocked when I received the PEB's decision.

Fortunately, there is an appeals process. I was able to find evidence that supported my claim, that at a minimum, my service in the Marine Corps aggravated my condition and could have even caused it to appear in certain organs. I was able to show evidence that VHL is a "two hit model disease." Someone with VHL is born with one "hit," one change in the genetic code. However, this is not enough to initiate the disease process. For tumors to form, a second "hit" must occur. This second "hit" is possibly caused by something in the environment. Researchers still don't

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Jay Platt has been given the Trail Name "Patch" by his fellow hikers.

know for certain. Therefore, it was my contention that, since I had no symptoms of VHL upon my enlistment, I must have been exposed to something during my military service that caused the second "hit." The Board that I appealed to found in my favor. I was obviously thrilled at the appeal board's decision.

When decisions arise around employment and medical coverage, it takes time and resources to appeal such a decision. I was fortunate that upon reconsideration, the Marines granted me full retirement with medical benefits. While that is an important victory in the U.S. health care system -- keeping my health care coverage -- it will seem difficult for people in other countries to understand. And in the grander scheme of things, it is only one small skirmish. The real battles are in wrestling one by one with the tumors themselves, and dealing with each one as constructively as possible, without losing our health. What we really need are better weapons in the war on VHL.

Through these last several years -- as I have come to learn this diagnosis, and begun to understand how to manage my own health -- the VHL Family Alliance has been a true source of support for me. This community of survivors has helped to support my spirit through the medical battles and the challenges I encountered with the terms of my retirement and health care coverage.

I would like to give back something in return. After much thought and research, I decided on a project that could be of benefit to all of us in our VHL "Family." I have set a goal to walk the entire length of the Appalachian Trail. The walk began the first of August at Mt. Katahdin, Maine, and will end 2160 miles away at Springer Mountain in Georgia. Why? Because it challenges your mental and physical toughness. Not only Marines, but persons with VHL and their families must have perseverance and courage. We are challenged to manage this disease, until all of us working together find the cure for VHL and cancer.

My goal is to raise $100,000 for the VHL Family Alliance during this walk through fourteen states. I want to raise that money to help find a cure for VHL -- to help people get an earlier diagnosis and to provide better ways of controlling tumors until one day we can stop the tumors from coming at all. I believe that people will be generous when they understand the way this disease hits families like theirs. If it means walking this trail when I'm tired, I will do it to help those who have supported me.

I'll need your help to do this! Why not join my team and help me reach this target, please send your donation to the North Carolina Chapter of the VHL Family Alliance at:

VHLFA-NC
102C Commonwealth Ct.
Cary, NC, 27511

In this first month, I have already hiked 300 miles -- to the top of Mt. Katahdin with a 60-pound pack, through woods and swamps, in dry weather and wet. I'm hiking an average of 15 miles a day, with a one or two day rest after 5 days of hiking.

I'll do my part -- you do yours. With your help we can cure VHL!

Follow Jay's journey on http://www.vhl.org/hike

* Editor's Note-- VHL causes angiomas of the retina. It does not usually cause sudden blindness but in Jay's case the angiomas went undiagnosed until they were quite large and had caused a retinal detachment.

A native of Valdosta Georgia, currently residing in Camp LeJune, North Carolina, Platt joined the Marine Corps in 1984 and served in various roles, including ammunitions and explosive technician when the 1987 diagnosis exploded much of his world. Over the years, the genetic disease has required surgeries to remove tumors on his brain and in several organs. In the next year he is facing surgeries to remove tumors in his spine and on both kidneys.

The Appalachian Trail Conference estimates that only about 300 people in 70 years have completed a thru-hike southbound. Different segments of the trail offer a variety of scenic terrain and views as they connect with mountains, lakes, ponds, streams and forests. Extremely rugged in many sections, the wooded, unlit trail offers small lean-to's for night shelter. Established in the 1920's and 30's by hikers, it was made part of the National Trail System by Congress in 1968. The trail is maintained by volunteers. ❑

Vegetables: Count on Three to Five Servings

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Just imagine Italian food without tomato sauce, Mexican food without salsa, or a stir-fry without broccoli! Vegetables add taste, color, texture, and nutritional value to many favorite meals. Many people think that they are too busy to eat enough vegetables or that vegetables just don't taste very good. But there are lots of easy ways to enjoy great-tasting vegetables without spending hours shopping and chopping!

The Food Guide Pyramid recommends eating three to five servings of vegetables a day. Vegetables contain very little fat, no cholesterol, plenty of dietary fiber, and lots of vitamins, minerals, and phytochemicals. Phytochemicals are the chemicals found in plants that may protect our cells from the damaging effects of toxic substances, which can result in cancer and heart disease.

Three to five servings is easier than you think

Three to five servings may sound like a lot of vegetables, but 1 cup of raw leafy vegetables or salad or 1/2 cup of cooked, frozen, or canned vegetables counts as a serving.

Other examples of one serving are:

3 to 5 spears of broccoli
7 or 8 baby carrots or carrot sticks
1 ear of corn on the cob
6 spears of asparagus
1 medium stewed or fresh tomato
1 medium baked potato
4 dark green lettuce leaves

Some people may find preparing fresh vegetables an inconvenience. So it's good to know there is little difference in the amount of nutrients in fresh, frozen, or canned vegetables. Frozen and canned vegetables are processed within hours of being harvested and are often fresher than "fresh" vegetables that may have been in transit or storage before you eat them. Also, frozen and canned vegetables are available year round, whereas some fresh vegetables may be available only seasonally. Eating both frozen and canned vegetables makes it easy to include those three to five servings each day for good health.

Whatever type of vegetables you choose, be careful not to overcook them. Cooking vegetables just right retains nutrients and preserves their appealing colors and flavors. Usually frozen and canned vegetables need only to be heated, while fresh vegetables should be cooked until tender but still crisp.

Whether eating at home, in restaurants, or on the run, you can find many easy ways to eat more vegetables every day. Remember you don't have to eat three to five different vegetables every day--you could eat a whole cup of broccoli to count as two servings. Here are some ideas to help include more vegetables into a healthful eating plan.

At home or for sack lunches:

Combine one can each of kidney beans, garbanzo beans (chickpeas), green beans, and wax beans to make a multibean salad. Toss with low-fat or fat-free Italian dressing.

Top a microwaved baked potato with heated frozen broccoli or cauliflower in low-fat cheese sauce--a quick meal in just 10 to 15 minutes.

Add frozen or canned corn to salsa. Or, when making nachos, sprinkle some corn on tortilla chips with low-fat cheese. Olé!

Add vegetables to soup, rice mixes, or pasta dishes. Try adding one package of frozen pasta and vegetables to two cans of lower-sodium or reduced-fat chicken or beef broth.

Stir-fry vegetables using 1 teaspoon of oil per person. Season with ginger and soy sauce and serve over rice.

Stuff baby carrots, celery, broccoli, and cauliflower florets into a pita pocket with low-fat cheese for lunch.

Stir peas into macaroni and cheese, tuna noodle, and other casseroles.

Roll up canned kidney or pinto beans, salsa, and low-fat cheese in a tortilla.

Here is a hearty sandwich containing vegetables--a meal in itself!

Turkey Veggie Melt

1 (1-lb.) pkg. frozen broccoli, cauliflower, and carrots
1/4 cup creamy Parmesan salad dressing
1 (8-oz.) loaf French bread
1/2 lb. thinly sliced cooked turkey breast
3 oz. (3/4 cup) shredded reduced fat Cheddar cheese

Heat oven to 400° F. In medium microwave-safe bowl, microwave frozen vegetables 3 to 7 minutes or until thawed; drain well. Stir in salad dressing.

Split French bread horizontally; place on ungreased cookie sheet. Arrange turkey evenly on bread halves. Spoon vegetable mixture over turkey. Sprinkle with cheese.

Bake for 7 to 10 minutes in oven or until thoroughly heated and cheese is melted.

Makes 4 servings.

Nutrition information per serving: 400 calories; 65 mg cholesterol; 14 grams total fat; 4 grams saturated fat; 700 milligrams sodium; 5 grams fiber.

For more information, contact the American Dietetic Association/ National Center for Nutrition and Dietetics Consumer Nutrition Hotline. For food and nutrition information or for a referral to a registered dietitian in your area, call 800/366-1655 of www.eatright.org/find.html. For customized answers to your food and nutrition questions by a registered dietitian, call 900/CALL-AN-RD (900/225-5267). The cost of the call will be $1.95 for the first minute and $.95 for each additional minute.

For information about Green Giant® vegetables, call 800/998-9996. Or write to Green Giant®, 2866 Pillsbury Center, Minneapolis, MN 55402-1464. This fact sheet is supported by a grant from Green Giant ®.

Hastening the Road to Diagnosis

The Role of the Broad Ligament Cystadenoma in Early Detection of VHL

Excerpted from a working paper by G. P. James, Ohio

Editor's introduction: This article highlights a little known benign lesion sometimes found in women with VHL. We want to share with you what is currently known, and enlist your help in learning more. If you have experienced something that sounds like this, we would appreciate your sharing your experience with us. The only way we learn is to assemble the experiences of a large number of people. Part of the problem is the widely varied terminology used to describe them. They occur among structures with names women usually do not recognize. We will explain some of this terminology and provide a list of terms you may hear.

In the *VHL Handbook* you will find these lesions referred to as "broad ligament cysts". In fact they are not cysts (simple fluid-filled sacs) but are cystadenomas (a benign epithelial tumor containing one or more cysts). In other words, they look more like a tumor on radiographic screening, but they are not cancer and will not invade other tissues. They do not pose a danger to the patient, but they can be helpful in diagnosing VHL. They are not all located in the broad ligament. A more appropriate name for them is *adnexal papillary cystadenoma of probable mesonephric origin,* or APMO (See Note 1).

More importantly, APMOs may easily be mistaken for a number of more dangerous tumors in the female reproductive tract. Labeling of an APMO as one of these more dangerous tumors might result in a recommendation for more treatment than is actually required. It is important that we learn more about APMOs.

Five women in the United States have been reported to have the unusual — and benign — APMO found to date only in women with von Hippel-Lindau disease (VHL).[1] (See Figure 2.)

The good news in these small numbers is that four of the five women were diagnosed very recently, during a short six-year period beginning in 1988, so hopefully the diagnosis rate is rising.

The even better news is that accurate diagnosis of APMO in two of the patients prompted timely VHL screening and VHL diagnosis before the more typical lesions of VHL caused serious problems.

It has been known since the mid-1960's that when the comparable epididymal cystadenoma (EC) occurs in men, it can be an important finding that may indicate that the person has VHL. Lindau himself was the first to recognize this cystadenoma in 1927 in a man already diagnosed with VHL. Important advances were made during the 1950's and 60's in identifying this benign cystadenoma and distinguishing it from others that occur along the male reproductive tract. Today, with routine screening and technological advances in our ability to detect small tumors, as many as 60% of men with VHL will be found to have one or more ECs.[2]

Until quite recently, physicians knowledgeable about VHL have not been in agreement about whether women with VHL could even have APMOs. Some doctors thought that it was an anatomical impossibility. Others believed that APMOs were occurring in women but that they were either not being reported or correctly diagnosed. In any case, APMOs were labeled "exceedingly rare" as recently as 1995 and hopes seemed slim that a finding of an APMO would assist in VHL diagnosis.

Through the late 1980s APMOs were reported only in women already diagnosed with VHL. The first two patients ever diagnosed with an APMO in the United States were women known to have VHL. But in 1990 and again in 1994, for the first time, two women were diagnosed with VHL after — and in one case because — an APMO was correctly identified.[3]

Note 1: APMO: A New Name for the Broad Ligament Cyst

The papillary cystadenoma of mesonephric origin is commonly called the "broad ligament cyst" because most of them form in remnant mesonephric duct tissue that happens to be embedded in the broad ligament. However the cystadenomas that are important in VHL are not all in the broad ligament (some are below it), and they are cystadenomas, not cysts.

The broad ligament is a large area of tissue that lays on top of the reproductive organs (see Figure 1). The broad ligament looks like drapery material, lying in folds and creases on top of both ovaries and uterine tubes, connecting these structures to the larger body of the uterus.

As discussed in this article, some papillary cystadenoma of mesonephric duct origin that can help in diagnosing VHL will be found attached to adnexal (adjoining) tissue that is not part of the broad ligament.

Gaffey et al (1994) coined the more appropriate name *adnexal papillary cystadenoma of probable mesonephric origin (APMO).*

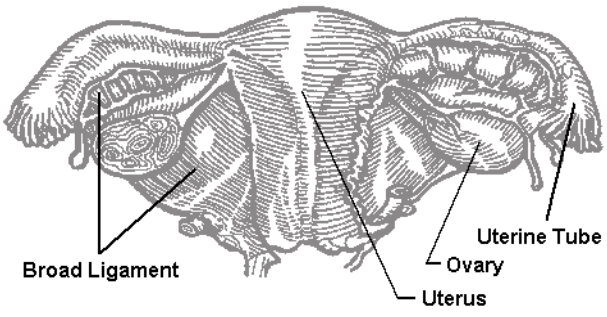

Figure 1: Broad Ligament

Patient #	Reported	Ages of Patient		Cyst Location	
		at APMO finding	at VHL dx	Side	Location
1	1978	na	na	R&L	BL
2	1988	46	20's	L	BL/btw T & O
3	1990	35	36	R&L	BL/over both Ts
4	1991	41	8	R	T/ through T
5	1994	22	23	R	V/near vaginal fornix

Figure 2: Five Reported Cases of APMO in VHL Diagnosed Women (United States)
na=Not Available; R=Right; L=left; BL=Broad Ligament; O=Ovary; T=Uterine Tube; V=Vagina; dx=diagnosis.

VHL diagnosis for patients #3 and #5 followed soon after the APMO finding. Age at VHL diagnosis for these two cases was set to "age at APMO finding plus one year" to indicate this diagnostic sequence. Detailed information on patients #1 and #4 is unpublished. Not included here is information on a sixth patient, a member of a Netherlands family with VHL, who was diagnosed with a broad ligament papillary cystadenoma (Karsdorp et al., 1994).

The route to diagnosis for these two women began with visits to their doctors for health problems that seemed to be unrelated to VHL. One woman told her physician that she had a pain in her lower right abdomen. Radiologic exam showed a calcified mass in the right pelvis. During surgery, two cystic areas were found near each of the uterine (or Fallopian) tubes. When the cystadenomas were removed and the tissue analyzed, the diagnosis was "papillary cystadenoma…highly suggestive of von Hippel-Lindau disease."[4]

The physician reporting on this patient writes, "The classic lesions of von Hippel-Lindau disease were clinically silent in this case." This patient was apparently experiencing none of the neurological or ocular symptoms that typically lead a physician to consider a diagnosis of VHL. Yet screening revealed pancreatic cysts and lesions of the cerebellum and kidney; renal cell carcinoma was diagnosed during follow-up surgery.

The second woman had been suffering for ten years with chronic ear problems. A year and a half after surgery for a lesion in the bony area behind her right ear (which we now understand was an endolymphatic sac tumor or ELST), a "golf-ball sized" pelvic mass was discovered during a routine physical exam. Exploratory surgery located a small tumor in the vaginal fornix, an area at the top of the vagina near the uterus. The tumor was removed and diagnosed as a "benign papillary cystadenoma." During subsequent screening, cysts of the pancreas and kidneys were found; renal cell carcinoma was later diagnosed.

The more typical lesions of VHL were not truly silent in this second patient. A "classic" cerebellar lesion had been found two years earlier, at the time the ear tumor was identified. The woman had no family history of VHL, and a diagnosis of VHL had evidently not been entertained.

Traditional guidelines for VHL diagnosis rely heavily on Central Nervous System (CNS) findings (eye, brain, spinal cord), often requiring documentation of one or more CNS hemangioblastoma in either the patient or a relative. Physicians have known since the 1960's, however, that a different set of diagnostic criteria is necessary when an unusual cystadenoma such as the EC in men is encountered. Melmon and Rosen, in their important study of a VHL kindred, wrote in 1964, "in one of our patients…palpation of an epididymal cyst was the key to subsequent diagnosis of Lindau's disease."[5] This patient had none of the typical signs of VHL yet diagnostic screening was begun as soon as the ECs were found. Screening revealed both a CNS tumor and an eye lesion. Melmon and Rosen understood the significance of this departure from traditional diagnostic procedure. "This is the first reported case in which a diagnosis of Lindau's disease was established before the appearance of symptoms. Decision of vigorous investigation of the patient was based on the finding of epididymal cysts and the knowledge that he had one chance in two of having inherited Lindau's disease."[6]

The diagnostic guidelines developed by Melmon and Rosen remain one of the most commonly used classifications in clinical practice today. Those criteria recognize that many patients will not have symptoms from a CNS lesion and broaden the definition of VHL to include only a single major lesion of VHL in a person who has a family history of VHL. Reproductive tract cysts in men have been considered, since that time, an important presymptomatic finding, alerting physicians to begin careful screening for VHL when such cysts are found in a male patient. Could the APMO serve the same role in women?

Where do APMOs occur?

The early warning bell that sounded for men in the 1960's has echoed for women in the early 1990's. Reasons for the delay in activating this important warning system for women can be found buried in our anatomy, in developmental differences that begin to take shape before we are born.

The technical name for the benign reproductive tract cyst that we have been talking about is "benign

papillary cystadenoma of probable mesonephric (duct) origin." This rather long diagnosis has three easy-to-understand meanings:

Benign means noncancerous. "Papillary cystadenoma" is simply any tumor that has both nipple-like projections and a cystic component. They can occur in a number of places in our bodies. When this cystadenoma is found along the reproductive tract, it can be made up of a unique type of tissue called "mesonephric duct" tissue.

Mesonephric duct is a term used by embryologists, scientists who study the development of the human being before birth. It is the name given to a pair of long ducts — and a smaller set of tubules attached to each duct — that will, by the time we are born, form important parts of the reproductive system in both males and females.

During the 38-week period before birth, while the embryo and fetus are developing, the mesonephric duct undergoes a lot of changes. Some parts or segments of this embryonic duct evolve into other structures that are given different names at the time of birth. Other parts "regress" or fall away.

The transformation process is different in men and in women and it is this embryological difference that becomes important as we try to understand why papillary cystadenoma has been easier to detect in men than in women. (See Figure 3.)

In men, the mesonephric duct stays intact and functional. It becomes the viable, tightly-coiled routing system upon which procreation depends and upon which a VHL diagnosis can be grounded. The duct itself becomes two ducts, the *duct of epididymis* and the *ductus deferens*. A set of small tubules, the *efferent ductules*, connects the head, or beginning of the epididymis to each testis. Together these structures measure over 20 feet in length and perform the important function of carrying sperm from the testes through the spermatic cord for ejaculation. Papillary cystadenoma in men are found throughout this duct system.

In women, the mesonephric duct system is a remnant system. Although it is made up of the same

Figure 3: Comparable Mesonephric Structures in Males and Females.

Embryonic Structure	At Birth	
	Males	Females
Mesonephric Duct	Duct of Epididymis Ductus Deferens	Duct of Epoophoron Duct of Gartner
Mesonephric Tubules	Efferent Ductules	Epoophoron

Source: Moore, *The Developing Human: Clinically Oriented Embryology,* 2nd ed,1977, p. 237.

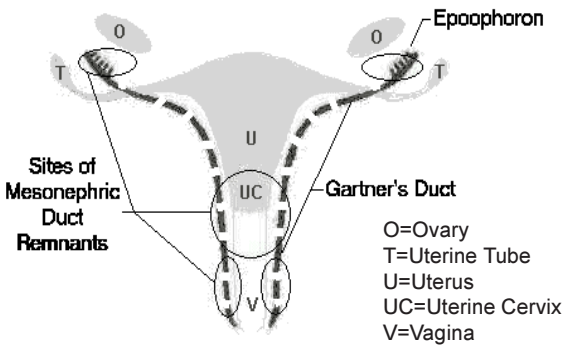

Figure 4: Sites of Mesonephric Duct Remnants in Females

Sources: Moore, *The Developing Human: Clinically Oriented Embryology*, p. 234; Skandalakis, *Embryology for Surgeons,* 2nd ed., 1994, p. 826.

set of tubules and ducts that are found in men (see Figure 3), only short segments of the embryonic system, called "remnants", remain by the time a female is born. None of these segments performs any function. It is in tissue associated with these parts of the old mesonephric duct that APMOs can be found.

The first segment of the old mesonephric duct found in the adult female is called the *epoophoron*. The epoophoron is a cluster of eight to thirteen small tubes, each connected to a very short duct called the *duct of epoophoron*. In the diagram (see Figure 4) these tubules and the short duct to which they are attached look a bit like the head and bristles of a toothbrush, laying in broad ligament tissue between each uterine tube (T) and ovary (O).

All women are born with these small tubules and short duct and it is in this segment of the mesonephric duct that most APMOs have so far been found. APMOs are reported in broad ligament tissue located between the ovary and uterine tube (patient #2) and in tissue close to each uterine tube (patients #3 and #4).

The second segment of the old mesonephric duct is a longer duct called the *duct of Gartner*. Although it is a "duct," it does not conduct anything. In most women, only pieces of Gartner's duct tissue remain, scattered along an almost invisible highway where the old mesonephric duct used to be.

Only the top of the Gartner's duct is located in broad ligament tissue. The bottom end is embedded in other types of tissue.

Only about 20% of adult women are estimated to have Gartner's duct remnants. Because not all women even have such tissue remnants, fewer APMOs will be found in this segment of the old mesonephric duct.

Surgeons find mesonephric tissue remnants clustered in two "hot spots" along the Gartner's duct :

(a) along the side (or lateral) wall of the uterus close to the cervix. The round circle in the middle of Figure 4 indicates this remnant area, the second site for papillary cystadenoma.

The pathologists who examined tissue from the papillary cystadenoma found in patient #5 wrote that this APMO most likely came from a "mesonephric duct remnant." The specific location of the cystadenoma, "lateral to the posterior vaginal fornix," is an area that corresponds to this Gartner duct remnant area.

(b) along the walls of the vagina and vulva. A pair of small ovals at the bottom of Figure 4 mark this remnant area. It is not known at this time whether APMOs do not form in this area, or whether they are simply not being reported in women with VHL.

Several other infrequently reported cysts (e.g. "the Gartner's duct cyst," the "mesonephric cyst") as well as "mesonephric remnants" also occur in this area of the reproductive tract. To date, these other mesonephric cysts and tumors do not appear to have a role in diagnosing women with VHL. (See Figure 5.) Nonetheless the variety of names by which mesonephric cysts may be called, and the difficulties in differential diagnosis, compound the problems of compiling case histories.

Careful work by radiologists and pathologists has firmly established that the reproductive tract cystadenomas that can help in diagnosing VHL occur — in both women and men — along all persisting structures and remnant tissue of the embryonic mesonephric duct system.

Changes in VHL Screening Guidelines.

In 1994, two unusual cystadenomas that can herald the onset of VHL — the APMO of the female reproductive tract and the endolymphatic sac tumor (ELST), a tumor of the middle ear — were both added to a new set of screening guidelines for VHL.[7]

These new guidelines, developed by pathologists at the University of Virginia Health Sciences Center, suggest that "strong consideration…be given to the diagnosis of VHL" in a person who has:

(1) *either* the mesonephric cystadenoma *or* the ELST *and* a relative with VHL or

(2) *either* the mesonephric cystadenoma or the ELST and one other documented VHL lesion, if there is no relative with VHL.

Next Steps

The finding of an APMO can serve as an early warning bell, signaling both patient and doctor that a diagnosis of VHL should be considered. Yet the number of women who enter VHL screening via this route remains alarmingly low.

Hastening the road to diagnosis for a portion of women with VHL will require changes in our screening protocols, in our diagnostic and reporting procedures, and in the awareness of local-level physicians who stand in a pivotal position to make the critical care decisions that can lead to VHL diagnosis.

Equally important is the development of precise tumor identification guidelines. Clear diagnostic guidelines are still lacking. Tumor identification guidelines will need to specify sonographic appearance, histological features, and common tumor sites. These will need to be developed by consensus of gynecologists and pathologists.

Structure/Tumor	Location
Ovarian tumor of probable Wolffian origin	ovary broad ligament retroperitoneum
Adnexal tumor of probable Wolffian origin	broad ligament vagina
Papillary cystadenoma in VHL	broad ligament vaginal fornix
Paratubal and Paraovarian cysts	various
Mesonephric remnants	uterine cervix
Mesonephric hyperplasia	uterine cervix
Mesonephric adenocarcinoma	uterine cervix vagina
Mesonephric cyst	vagina
Mesonephric adenocarcinoma	vagina
Mesonephric cyst (Wolffian duct cyst, Gartner duct cyst)	vulva

Figure 5: Mesonephric cysts and tumors found along the female reproductive tract.

Wolffian is another name for mesonephric. Information for this chart assembled from Kurman (ed), Blaustein's Pathology of the Female Genital Tract (NY 1994); Scully, Bonfiglio, et al, Histological Typing of Femals Genital Tract Tumors, 2nd ed.,(NY, 1994); Rosai, Ackerman's Surgical Pathology, 8th ed., (St. Louis, Missouri, Mosby-Year Book, 1996).

Accurate diagnosis of APMOs in women currently requires:

- surgery with sampling of the lateral walls of the cervix, a common site for mesonephric remnants
- careful excision of the mass
- a clear description of the APMO's location along the persisting segments of the embryonic mesonephric duct, and
- precise histological identification by pathologists familiar with the unique structural characteristics of mesonephric tissue.

Thus in cases where surgery is not warranted, accurate diagnosis will not be possible until better guidelines are developed.

Simple palpation and ultrasound — methods which successfully locate the majority of ECs in men — are less useful in locating and diagnosing APMOs in women. Physicians can sometimes palpate a cystic area during a routine pelvic examination. Follow-up ultrasound and abdominal CT will help pinpoint the general location of the cyst. Sonographic findings for APMOs in women to date have shown areas of "curvilinear calcifications" and masses that contain "both water attenuation and soft tissue attenuation." ***Radiologists caution that CT alone will not differentiate papillary cystadenoma from other types of benign and cancerous tumors that occur along a woman's reproductive tract.***

Pathology and sonographic records with clinical histories are available on at least six VHL women who have been diagnosed with APMO. An independent review would be a helpful next step in furthering our understanding of this unusual and diagnostically significant manifestation of VHL.

Women with VHL who are discussing with their physicians a cyst or tumor of the reproductive tract, should be sure to point out to the physician that benign APMOs have been found in women with VHL, and request that care be taken to differentiate an APMO from a possibly more dangerous kind of tumor, to ensure against unnecessarily aggressive treatment. We hasten to point out that this differential diagnosis is extremely difficult at this time, and may require analysis of the genes in the tumor cells. Very few hospitals in the United States can make this determination.

It will be necessary to remove the tumor and send it for an expert analysis before a final determination can be made. If appropriate, the patient and physician together might discuss the appropriateness of a "staged" approach to treatment.

Tumor tissue may be sent for analysis to: Dr. Tanya Tavassoli, Dept of Gynecology and Breast Pathology, Armed Forces Institute of Pathology, Washington, DC 20306. Tel: 202-782-1600; Fax: 202-782-3939.

Tissue previously removed can also be analyzed. The pathology department will likely still have slides, or a paraffin block, that could be used to determine whether it was an APMO. This will be statistically significant in determining the prevalence of APMOs in women with VHL. With your help, we hope to see better diagnostic guidelines in the future that can be more easily applied at all hospitals.

If you are willing to contribute your own experience to such a study, please contact us at +1 617 232-5946 or info@vhl.org, or send samples directly to Dr. Tavassoli along with a patient history and the local pathology report.

1. Patient cases: #1: Scully et al, eds., Case 1, *NEJM* 298 (1978) 2:95-101; #2: Gersell et al, Papillary Cystadenoma... *Am J Surg Path* 12 (1988) 2:145-149 and Funk & Heiken, "Papillary Cystadenoma..." *Am J Radiology* (1989) 153:527-528; #3 Korn et al, "Papillary Cystadenoma..." *Am J Obstet Gynecol* (1990) 163: 596-598 and operative records from the diagnosing physician; #4 pathology report provided by the patient; #5: Gaffey et al, "Aggressive Papillary Tumor of Middle Ear...and Adnexal Papillary Cystadenoma", *Am J Surg Path* 18 (1994) 12:1254-1260. Outside the U.S., see Karsdorp "VHL: New Strategies*..." Am J Med* (Aug 1994), 97:158-168. The earliest reference to a possible APMO in a woman already diagnosed with VHL dates to a 1931 article published in Virchows and referenced by Melmon & Rosen (1964).
2. Choyke et al (1997) *Urology* 49, 6:926.
3. Clearly in the Korn paper the APMO findings prompted VHL screening and diagnosis.
4. Information about these two patients was provided by the patients' physicians in published reports. Women reading this article who have been diagnosed with any APMO are encouraged to contact the VHL Family Alliance and share with us their experiences.
5. Palpation means touching with pressure in order to diagnose.
6. Melmon & Rosen, "Lindau's Disease," *Am J Med* (1964) 36:608-609.
7. Gaffey et al, "Aggressive Papillary Tumor…", *Am J Surg Pathology* (1994) 18, 12:1259. Manski et al (1997) and the June 1997 issue of the *VHL Family Forum* provide detailed information on the endolymphatic sac tumor.

There You Are!

Boston cabaret performers Carol O'Shaughnessy, Jan Peters, and John O'Neil gave a special performance of their smash-hit musicale ***There You Are!*** for benefit of the VHL Family Alliance Fund for Cancer Research. Carol, an actress and comedienne, had everyone in stitches with her sketch of an Italian grandmother. Jan Peters performed a varied program including a delightful medley of alto parts to Broadway hits. John charmed us with his one-of-a-kind blend of campy humor and compelling ballads. It was a delightful evening of fun and frolic, and raised money for research!

Our thanks to the performers and to the Lyric Stage Theater in Boston for their generous donations, and for a marvelously memorable evening.

Ask the Experts

Draining Cysts in the Brain

Question: What is the best way to deal with a hemangioblastoma with a cystic part? My doctor keeps draining it, but it keeps coming back. I've had eight brain surgeries to drain cysts and it looks like I'll need another. Is there an end to this? -- *T.P., Ohio*

Answer: Draining the cyst only relieves the immediate symptoms; it does not get to the basis of the problem. The cystic fluid is generated by the tumor itself. As long as the tumor is in place, the cyst will continue to re-fill. Thus, the tumor should be removed. With advances in surgical techniques, a neurosurgeon up-to-date in microsurgical techniques and familiar with hemangioblastomas should be able to remove this vascular tumor. They can be tedious and it requires certain surgical techniques that are not needed for most other tumors of the brain or spinal cord, so not all neurosurgeons deal with them.

In the rare cases where the patient is pregnant, or if the patient has already undergone so many surgeries that an additional surgery is not advisable at the time, then draining, shunting, or otherwise managing only the cyst may be the right short-term strategy, but it is not a long-term solution and is rarely advisable, or needed. For the same operative stress on the patient, the tumor could be removed and a longer-term solution reached.

The NIH will be happy to consult with patients and physicians facing this difficult question. -- *Edward H. Oldfield, M.D., Chief, Surgical Neurology Branch, National Institute of Neurological Disorders and Stroke, Bethesda, Maryland*

Concerned about Eyestrain and VHL

Question: I have multiple retinal angiomas in my left eye. Despite several laser treatments, I have lost central vision in that left eye. I am concerned about what might happen to my right eye. Will the strain of using my right eye to do the job of two eyes make my condition worse? -- *Brenda R., South Africa*

Answer: In response to your question, you will not do any harm to your good eye by performing any visual task. You should feel free to read, write, use computers, etc.

Several considerations:

1) We recommend that you use "protective eye wear" (such as polycarbonate lenses, goggles when playing sports such as tennis or when performing tasks that might injure your eyes) to protect your right eye physically

2) Be cautious, especially when driving or in unfamiliar surroundings, since loss of significant vision in one eye can result in loss of depth perception and stereoscopic vision

3) Be cautious when skiing, playing tennis, etc. since you will have loss of depth perception and may have difficulty judging distances.

Please be assured that you should feel free to perform any visual task you desire.

-- *Jerry Cavallerano, OD, PhD for Lloyd M. Aiello, MD, Beetham Eye Institute, Joslin Diabetes Center, Boston, Massachusetts*

Tuning Out Tinnitus

Question: I found an advertisement for a device that "tunes out" tinnitus. Is there any reason why someone with VHL should not use one of these?

Answer: In general, I don't see any harm in these devices as long as the volume or intensity of the masking is not loud enough to cause further damage to the hair cells of the cochlea. They work about as well as any of the other techniques for tinnitus. Biofeedback, hearing aides, etc. .... all have about the same efficacy. The causes of tinnitus are too numerous to count. So it makes sense that not all patients would respond to the same treatment. I wouldn't recommend someone just using something for tinnitus without figuring out why they have the tinnitus in the first place. It could range from anything like a tumor, to too high a medication, low thyroid, high thyroid, infection, aspirin, Motrin, kidney failure, aneurysm, arteriovenous malformation, post-trauma, etc...just to name a few. Too commonly though, a real cause for tinnitus cannot be pinpointed. But its important to make sure its not a treatable, identifiable cause. As a caveat, most tinnitus devices claim amazingly high success rates. Unfortunately, I have not seen a single one that lives up to its claims.

-- *Daniel Choo, M.D., Neuro-otology Branch, NIDCD, National Institutes of Health, Bethesda, Maryland*

Ophthalmic Ultrasound

Question: I heard that they can now detect retinal involvement early with ultrasound. They can see the blood vessels curl before they turn into angiomas. Is this true? -- *Emily S., New Jersey*

Answer: Ophthalmic Ultrasound is extremely useful and suggested in the evaluation of retinal angiomas. This is a technique that is highly dependent upon having the most sophisticated equipment and a highly experienced examiner. It is a method best used to follow existing angiomas to note change. The Joslin Vision Network and digital imaging is terrific for the initial evaluation of retinal vessels and adjunct to ophthalmic ultrasound. Should you need additional information, feel free to call me. -- *Dr. Richard M. Calderon, Associate Chief, Clinical Practice, Beetham Eye Institute, Joslin Diabetes Center, Boston, Massachusetts*

In Your Own Back Yard

-- Amy A., Louisiana

I wanted to do something to raise money for research. I have two small children who are at risk for VHL. I wanted to do it for them, for the memory of their father, Rondo, who died last year, and to give back some of the caring support the VHL Hotline has given me over the last few years.

I set out to have a yard sale. I set a goal of raising $500. I asked a few close friends if they had anything I could sell and donate the proceeds to the VHL Family Alliance. As people learned what I was doing and why, more friends and neighbors brought things over and offered to help.

By the time we counted up the receipts, I could scarcely believe it! I am thrilled to send the enclosed check for $1026.15 in memory of Rondo. Best of all, it was a labor of love -- not just from me, but to me as well. It was a wonderful outpouring of love and support from my community.

The Sky is Falling

-- Patricia F., California

My husband sat opposite me sipping on a delicious double Starbuck's cappuccino. We sat on those small fanciful wrought-iron chairs at a round table in the middle of the Chevy Chase Pavilion, a few miles from the National Institutes of Health. We were completely surrounded by the wonderful shops: Pottery Barn, Hold Everything, the Limited, Express, a sun glass shop, and a party shop to name a few. On the upper levels were the rooms of the Embassy Suites Hotel.

Immediately overhead was a giant, light teal green ornamental truss system in the shape of a gazebo that seemed to be holding up the sky. Through the clear skylight and truss system the irregular shape of the clouds were clearly visible: racing past on this sunny, blustery day. A short distance away, the cherry tree blossoms around the Tidal Basin were a day away from their glorious peak.

I gingerly tasted a few sips of my husband's aromatic coffee, afraid that too much might aggravate the diarrhea I experienced as a side effect of the gastroview I had drunk the day before for my CT scan. I peered up at the sky and was reminded of the folk tale, *Chicken Little*, and the repeated phrase, "the sky is falling." Supportive armature, like the trusses, would be necessary to keep my own will power from collapsing.

Two hours earlier, Dr. Phillips entered the consulting room cheerfully enough and began our consultation with the words, "only a slight change," This is what my husband and I heard and remembered. I, however, wanted more details – specifics, numbers, how much was this slight change? On which of the two small malignant tumors, one in each kidney, was the change?

I was told that according to the CT scans the tumor in the left kidney had gone from 0.7 centimeters to 1.5 centimeters. A malignant tumor of 1.5 centimeters in the kidneys is not in itself something to be too concerned about for VHL patients. Experienced urologic oncologists have decided through research that there is little risk of metastasis before tumors reach the threatening size of 3 centimeters. Because of the frequency of tumors over a patient's lifetime they are comfortable postponing surgery until that time. Then surgeons do nephron sparing surgery, when they take out the smallest amount possible of the kidney, thus preserving a patient's own kidney function as long as possible. What seemed worrisome about the increase in growth rate of my tumor in the left kidney was that it had doubled in size in one year. I mentally calculated that if it doubled again in the next 12 months, I'd need renal sparing surgery this time next year or shortly thereafter. Not a happy thought.

I felt angry. Not violently angry; just in a bad mood. I had hoped to beat the averages and get a 100% "no changes" report. I had tried to live healthily by eating lots of fruits and vegetables and drinking green tea and eating soy foods! My surroundings in the impersonal mall with all the people shopping looked surreal. I felt as if they were of another species – the cancer-free. I tried to feel part of that world, but this afternoon I did not. I felt like an alien surrounded by lovely, attractively displayed things to buy that all seemed superfluous. What I wanted was not for sale. I wanted a new pair of genes (VHL genes, not blue jeans). I wanted a magic pill to kill my tumors. Among all the reproductions for sale immediately before me I couldn't find an artificial kidney. None of the world's most prestigious shops that adjoin the Pavilion had any of these items. Not even Saks, Neiman Marcus, Tiffanys, Jaeger, Gianni Versace nor Cartier.

I was keenly aware, as I was brooding on my latest diagnosis, that I'd have to get out of my surreal way of looking at things and get back into the swing of life promptly. For with or without me, life would go on. People would go to the Pavilion shops and purchase what they wanted and derive pleasure just from being there. They would share a coffee or a beverage with a loved one and enjoy idle chit-chat. I needed to feel I could, and would want to, do the same. From past experiences with other tumors I took comfort in knowing that my anger and surreal lenses were temporary. I knew that I'd find more strength. Someday, I hoped, I would find the silver lining in those clouds above, which for now was completely hidden. ❑

How Are You?

Manage Your Own Medical Journey

review of a new book by Patricia Foote, California

One in 10 people will go to the hospital this year. Yet we receive no training for how to be a patient or how to navigate the professional medical world. The book *How are You? Manage Your Own Medical Journey* empowers the reader to be informed, proactive, and at peace with a serious medical crisis. The fascinating role and complex issues of genetics, life with a chronic illness and relationships are described in an honest, thought-provoking account of one woman's journey with von Hippel-Lindau disease.

Ms. Foote tells her own story in a very moving way. Based on her personal experience with VHL, she provides guidelines for people with any chronic or rare disease -- strong lessons in being a savvy consumer. Any serious illness deeply shakes our sense of self, and indeed the meaning of life. She shares her journey from diagnosis through multiple procedures to the new reality of living -- yes, happily! -- with a chronic illness. After reading this book, you may never use the question, 'How are you?' in the same casual way again.

"This book is a milestone in the arena of self-help and patient advocacy," says Dr. William Kintner of the Pacific Southwest Regional Genetics Network. "It is a wellness handbook for every person."

"Rare diseases are often the last to receive attention and funding on scientific levels, and even less likely to focus on survivorship issues. This book offers a beacon of hope to people living with von Hippel-Lindau (VHL) or other rare diseases as it shines light on an often misunderstood disorder. By sharing an intensely personal journey, the author offers information and inspiration that will no doubt decrease the suffering of other survivors," says Susan Leigh, R.N., Past President of the National Coalition for Cancer Survivorship.

A series of "How to" sections offer practical advice:

How to find a good general practitioner
How to deal with a serious diagnosis
How to get ready for hospital
How to make those first steps towards recovery
How to find a specialist
How to deal with medical incompetence
How a patient can help relationships
How to be a caring caregiver
How to locate a support group
How to locate a genetic counselor
Questions you may have for your Genetic Specialist
How to evaluate a health insurance company
How to accept the new reality of life after a serious illness (How to live life with a chronic disease)
How to find clinical trials

Ordering information is on page 15. For each book sold through VHLFA, $5 will be donated to the Alliance. ❑

Kathy & Andy B., Indiana; Renée R, California, at the Seattle meeting.

Ask the Family

Question: Is there an issue of the VHL news letter where people discuss pros and cons they have experienced from having their children undergo DNA testing?

Answers: There could be, with input from you! Please share your own thoughts and feelings about choosing DNA testing for yourself and your children.

- What are your reasons for choosing to test?
- What are your reasons for not choosing the test?
- Tell us where you are confused or undecided.
- If you have already made your decision, are you glad or not that you made the decision you did?
- What are your observations of your child's reaction?
- We would be very glad to hear directly from the young people involved too!

Please share your thoughts and feelings -- as anonymously as you wish. Mail or fax to VHLFA, or e-mail to joyceg@pipeline.com.

Ellen and Rob W., Minnesota; Altheada J., New York.

Seattle Meeting, June 1998

The Fifth International Patient/Provider Conference on VHL (Seattle) took place June 5-7, 1998, in Bellevue, Washington, outside Seattle. The conference was wonderful! Alice Coday, VHLFA Washington chapter chair and conference co-chairperson, designed an outstanding meeting that covered lots of important issues, was well-paced and pleasant, and offered lots of time for interaction among the attendees. Robin L. Bennett, M.S., conference co-chair from the University of Washington, and several other genetic counselors, were with us throughout the meeting, and a number of physicians from the Washington area attended. We had four outstanding speakers from the National Institutes of Health. One family drove all the way from Minnesota. Altogether we had eighty-four people from 22 U.S. states and 2 Canadian provinces -- Arizona, British Columbia, California, Colorado, Georgia, Idaho, Indiana, Iowa, Kansas, Maryland, Massachusetts, Minnesota, Mississippi, Nebraska, Nevada, New York, North Carolina, Ontario, Oregon, Pennsylvania, Tennessee, Utah, Virginia, Wisconsin, and of course Washington!

Friday's presentations were on employment rights and legislative concerns related to genetic factors in health. Friday evening at the patient panel included a particularly rich discussion of men's feelings, both as patients and as spouses.

Saturday Dr. Roberta Pagon of Seattle gave an overview of VHL, and explained how to go about getting DNA testing. Dr. Gladys Glenn of the U.S. National Cancer Institute in Maryland followed with a discussion of the "state of the art" in the VHL clinic there, and the usefulness of DNA testing information in clinical practice. Dr. Peter Choyke, also of the National Institutes of Health, presented NIH's findings on organ preservation. After nearly ten years of investigation, their findings support treatments that preserve normal organ function, especially kidney and pancreas, but including preservation of the adrenal cortex when pheos are removed. There were reviews of kidney, eye, ear, and central nervous system tumors and their treatments, and Robin Bennett shared her thoughts on improving communications between patients and professionals.

Patti and Ken K., California; Dr. Gladys M. Glenn, Maryland, at the Seattle meeting.

At lunch on Saturday Susan McGuire and Kathy Braden were duly elected to the Board of Directors, and two new research awards were announced: to Dr. James Gnarra of Louisiana State University and to Dr. Robert Burk of Albert Einstein School of Medicine in New York. Both will work on understanding different facets of the role of the VHL protein.

On Sunday we heard from a naturopath who specializes in co-treatment for cancer patients, an acupuncturist, and a nutritionist. All gave us excellent insights into how we can keep our bodies strong alongside the stresses and scans and procedures that go with managing VHL. Dr. Paul Reilly shared some vitamin suggestions for before and after surgery.

Many thanks to co-chairs Alice Coday and Robin Bennett for their superlative planning and execution, and to each and every presenter and attendee.

"I was so impressed and elated with the Seattle Conference. It was such a pleasure to meet you and the others. I am so glad that I attended." -- *Maria S., Pennsylvania*

"I enjoyed the conference. As usual it brought together some interesting people and showed some real problems with the health care delivery system. I am always humbled by the courage and perseverence the VHL Alliance members show." -- *Peter Choyke, M.D., National Institutes of Health* ❑

Paris Conference

The Third International Symposium on von Hippel-Lindau will be held in Paris, September 16-18. The programme will include three plenary lectures: Inherited kidney carcinomas by Berton Zbar; Ethical problems in genetics by Josué Feingold, Paris; and Tumoral angiogenesis pending acceptance by a collaborator of Judah Folkman, Boston. Mini-symposia will be held on various aspects of clinical management of VHL, and genetic research. Attendees are expected from Japan to Poland.

"Love Life, and Live Life Loving!" -- Donna Masters, Australia

submitted in memory of Donna Masters, by her family and friends, with fond memories of her laughter and sense of humour, courage and unconditional love that she so willingly gave. And most of all for touching all our lives in such a special and unforgetable way. -- *Julie M.*

Chapter News

New Zealand, by Val J.

Things here are moving along slowly but surely Jon, myself and Martin (when ever he is in Auckland) have had several meetings with our genetic department people. To date we have established a list of doctors to help advise and guide patients in New Zealand. The response so far has been positive..

Once we get this all together, we hope to have some sort of meeting with our ever increasing group next year in Auckland for everyone to meet. With plenty of notice we are hopeful for a good turn out. My own family from the South Island will be up this way for a wedding so we will be trying to set dates about the same time.

As to my health - things are stable at present with regular checks continuing. Jon and I are disappointed that we can't make the conference in Paris but wish all the best. We have fond memories of Hawaii.

New York, by Altheada J.

VHLFA honored Mr. Wilbert Cooper for his work and help in creating our famous T-shirts, sweatshirts and golf shirts, and in helping to promote the VHL Family Alliance. The plaque was presented by Altheada at the annual Cooper Family Reunion in Dover, Delaware, in July.

I'd like to dedicate this poem to Jay for his continued love and support, my husband and friend, -- Tania D., Ontario, Canada.

The Friend Who Just Stands By

When trouble comes your soul to try,
You love the friend who just "stands by."
Perhaps there's nothing he can do --
The thing is strictly up to you;
For there are troubles all your own,
And paths the soul must tread alone;
Times when love cannot smooth the road
Nor friendship lift the heavy load,
But just to know you have a friend
who will "stand by" until the end,
whose sympathy through all endures,
Whose warm handclasp is always yours--
It helps, someway to pull you through,
Although there's nothing he can do.
and so with frevent heart you cry,
"God bless the friend who just 'stands by'!"

Written by B.Y Williams, published in *Poems That Touch The Heart* by A.L. Alexander

Research Grants Awarded

Thanks to *your generosity*, we were able to award a record $80,000 in research grants this year. This total includes donations, memorials, yard sales, raffles, a concert, a benefit cabaret, and a major gift from the Rasmussen family in Minnesota. We are hopeful that these two grants will further our knowledge of the function of the VHL gene so that we will understand better how to intervene in the process and make a difference in the outcome.

Let's do it again! Give what you can, and send brochures to others. Join Jay in going that extra mile. Together we will find a way to control VHL.

VHL Control of VEGF Expression

Dr. James R. Gnarra, Louisiana State University Medical School, New Orleans, $40,000

In this project, Dr. Gnarra proposes to continue his studies on how the VHL protein regulates the expression of the vascular endothelial growth factor (VEGF) and the growth of blood vessels. The proposed studies are a continuation of those presented last year, for which he was awarded funding. His hypothesis, which is shared by many researchers within the VHL scientific community, is that altered regulation of VEGF expression in individuals with a mutated form of the VHL gene is critical for the development of VHL disease. Therefore, understanding how VHL regulates VEGF is very important, since it may allow the identification of targets for therapeutic intervention. Modulating the activity of these targets could prevent or lower VEGF expression, and hence the development of VHL tumors. The regulation of VEGF expression by VHL is believed to be carried out through binding of certain proteins to the mRNA that encodes VEGF. He seeks to identify these factors and study how they work.

Studies on the VHLp18(MEA) protein

Dr. Robert D. Burk, Albert Einstein School of Medicine, New York, $40,000

Dr. Burk proposes to study how VHL functions normally in the cell. Based on the type of proteins that interact with VHL, it has recently been postulated that VHL may participate in the elimination by degradation ('proteolysis') of certain cellular proteins. Dr. Burk plans to investigate this possibility further by determining the precise location of VHL within the cell, identifying the proteins that interact with VHL and establishing the cellular structures that VHL associates with. In addition, he has reported that another protein, identical to VHL but slightly smaller, is in fact the most abundant form of VHL in the cell. He plans to compare the function and subcellular distribution of each form of the VHL protein.

This issue is dedicated to the memory of Madge Hall, *a recently retired member of the Board of Directors, who passed away August 13 in Oklahoma City. We are grateful for her years of devoted service, and most of all for her friendship and partnership.*

Our thanks for Contributions . . .

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In Honor of . . .

Andy Braden, by Paul Braden
Sharry Brydon, by John S. Duval
Alice Coday, by Jack & Marilyn Hoover
Lori Davids, by Lisa Hunnicutt
Stewart Duval on his birthday, by Sharry Brydon
her new grandson Ricardo Louis Feinstein, by Nadine Chase
Janie Kelting, by Beverly Johnson
Jean Lima, by Becky & Peter Lima
Jay Platt, by Jim Cathcart, Joyce Graff, Brian & Susan McGuire
Cathy Sladke, by Sandra & Kenneth Noel

In Loving Memory of . . .

Rondo Anderson, by Amy Anderson, Helen Dufrene, Marion Jones
Sue Ando, by Kit & Barbara Sugai
Jason Berry, grandson of Fred & Linda Turner, by Pamela Columbia, Kristin Littlefield, Telephone Pioneers of America
James Woodson Bray, by Harry & June Wilcox
Bernard Christensen by Jean Christensen
Gustave Gertz, by Grace Washburn
Frank Graff, by George & Michele Kelly
Charles Hall, by Marilyn Fannin, Gary Galey
Madge Hall, by Brian & Susan McGuire
C. Michael Kruse, by Susan Adamson, James Reilly, Witch Hill Ski Association
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Toll-free in the United States and Canada: 1-800-767-4VHL
Editor: Joyce Wilcox Graff, 1-617-232-5946 (eve)
Adviser: Debra L. Collins, M.S., U. Kansas Med. Center, 1-913-588-6043
Internet website http://www.vhl.org
171 Clinton Road, Brookline, Massachusetts 02445-5815 U.S.A.

VHL Video Now Available!

We are delighted to announce the availability of ***VHL: Quest for a Cure***. This 15-minute video presentation depicts life with VHL, its hopes and challenges, and the importance to everyone of finding a cure.

This project was originally conceived in 1996, and is now complete thanks to the hard work, vision and dedication of a large team of people:

- Filmed and directed by Fred Simon of Fred Simon, Productions, in Wellesley, Massachusetts
- Script editing by Amy Gray Light, Arkansas
- Narrated by Nelson Martinez, anchorman, KOAT-TV, Albuquerque, New Mexico
- Editing organized and supervised by Cary Schwanitz, KOAT-TV
- Titles and Editing services donated by the management and staff of KOAT-TV, Mary Lynn Roper, General Manager
- Interviews with members across the country

It includes a montage of photos of VHL families, photos submitted by members from as far away as Belgium.

We are grateful for the participation of every person who worked on the video, who helped to inspire it, craft it, or review the script, and who helped to bring it at last to fruition.

We are very pleased to present to you ***VHL: Quest for a Cure***. We hope that you will show it to groups in your community, and that it will help to introduce VHL, to raise consciousness in the medical community and among the general public, and that it will bring hope and a sense of community to people with VHL throughout the world.

See below for ordering information.

DONATION & MEMBERSHIP INFORMATION

Please check one: I am a ❑ Individual with VHL ❑ VHL Family Member ❑ Supporting Friend
❑ Professional (physicians nurse, social worker, etc.)
Occupation: _____ _____

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

Name: _____

Address: _____

City: _____ State: _____ Zip/Postcode: _____

Country (if not USA):_____ Fax: _____

Phone (home): _____ Phone (work): _____

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)
Yes! I want to join Jay's team!
❑ Please send me ____ brochures about Jay's hike for me to share with others.

Donation:

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

Purchase merchandise:

❑ #___Copies of Pierre Jacomet's tape of classical piano @ $12 each $ _____
❑ #___Copies of *How Are You?* (see review, page 11) @ $17 each $ _____
❑ #___Copies of the *VHL Video* @ $20 each, (specify __ US or __PAL format) $ _____

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

VHL: Quest for a Cure
Raising Research Funds

Even for a Marine of sound body, conquering the 2160 miles of southbound wilderness known as the Appalachian Trail would be a remarkable feat, something only about 300 people have accomplished. But 32-year-old Jay Platt who is now marching down the trail's lush footpaths in New Hampshire has undertaken the grueling five-month journey despite surgeries to remove his left eye and part of his kidney. Platt is determined to complete the tour, heading the VHL Family Alliance's annual drive to raise money for education and research. Platt's goal is to raise $100,000 for programming and cancer research.

We need your help! Brochures are available to share with friends and family. When Jay completes his walk atop Springer Mountain, Georgia, in December, let's welcome him with an impressive total in research funds.

"Please tell Jay there is somebody in Malaysia who says 'Sybas!' In Malay this means 'Well done!'"
-- *Babienne R., Malaysia*

Have you seen the video?

"I think the VHL video is outstanding!!! I watched it with someone unfamiliar with the disease and he found it clear and instructional without being boring. Congrats!!!!" -- *Paula S., Florida*

"I recently received your video about VHL and share it with some friends. We thought it was great. Thank you. We hope to share it with many more." -- *Terry W., South Carolina*

See page 15 for details

VHL Family Forum
Newsletter of the VHL Family Alliance
171 Clinton Road
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