Building Hope Year by Year
by Manuel Greco, Vice President of the VHLA Board of Directors

Dear fellow VHL patients, families and VHLA friends,

As you know, VHL is a genetic condition. In my case, it is primarily seen in my brain. It doesn’t matter how hard I wish for the tumors to stop growing; they just keep coming back. So far I have had 15 brain surgeries. Of course, I have also had surgeries in my spine, kidney, and eyes—you know, it’s VHL! The only thing to do is to hope that one day a “magic pill” will surface that will eradicate the effects of this condition.

As a #VHLWARRIOR, I know that hope alone will not create a cure. I also know that I need to actively do something about it. I am not a researcher, so the only realistic way that I can work towards a cure is to raise money to help the researchers find that “magic pill”.

That is the reason that I decided to start an annual fundraising dinner in New York City. I am extremely fortunate to have many friends and colleagues working with me in the financial industry. With their assistance, I have been able to raise a substantial amount of money.

In January 2014, we held our first NYC Gala where we raised nearly $130,000 which, through VHLA’s competitive research grant program, now supports two exciting new projects. This year’s NYC event was even more successful! Over $300,000 was raised! This is in part thanks to Christy and John Mack (my former boss and former CEO & Chairman of the Board at Morgan Stanley). I am sure that the Willem Dafoe video, created for VHLA at no cost thanks to the request of our common friends, aided our ability to raise this sum of money.

Needless to say, NYC Gala 2016 is already in the works!

So, my fellow VHLA friends, please join me in this quest for a cure. While I know that not everyone has the connections to raise this type money, every dollar counts. No matter how small or large the amount of money raised, no matter what type and scale of event you are able to organize, know that by helping to raise money, you are doing so to increase your chances of a longer life. If each of us does our share, I am absolutely certain that we will be able to control the uncontrollable VHL. We need to be optimistic and believe that, with our help, it will be any day now!

All the best, Manuel

The Power of the Internet
by Ilene Sussman, Executive Director, VHLA

Recently, I received an upsetting call. Apparently, my cousin’s wife, a vibrant 30-year-old mother of two, had suddenly become violently ill from an unknown illness. While we all hoped and prayed for the best, she took a turn for the worse and passed away 24 hours later.

While her illness is unrelated to VHL, there are a number of relevant parallels I took away from this tragedy. This situation reminded me of the value of friends and family, the importance of living life to its fullest, and the significance of being appreciative for the “gifts” given. Oddly enough, I also learned the true power of the Internet and Viral Marketing.

In this situation, the Internet was instrumental in providing critical updates to friends and family regarding my cousin’s wife’s condition. At first, only a handful of people knew what was happening. However, within a few hours, word had spread to over 500 people. As I watched the updates and countless messages of support, somehow I drew solace following the website. By the time my cousin buried his wife three days later, nearly 1,000 people had heard the story.

Last summer’s Ice Bucket Challenge was another example of the power of viral marketing. The success behind this campaign was spurred on by one patient’s desire to spread the word about an important cause.

Since May is VHL Awareness Month, there is no time like the present for us to work together to spread the word about VHL. Imagine the impact we would have if each person impacted by VHL posted information on social media outlets, shared the Willem Dafoe video, or changed their profile picture. Together we could create a media buzz about VHL!

Now is the time to let everyone know that:

• VHL is a genetic form of cancer causing tumors throughout the body.
• With no known cure, VHL is a lifelong disease patients battle their entire lives.
• The VHL gene is involved in many other forms of cancer; finding a cure for VHL will play a vital role in curing other forms of cancer!

Spread the word about VHL! See firsthand how powerful the Internet can be!
Living with Chronic Pain

Pain is normally a warning signal to help protect your body. There are people born with a rare genetic disorder that makes them unable to feel pain. The gene mutation responsible for this disorder stops a protein that is part of the pain receptor pathway from “allowing electrically charged particles to cross the surface of nerve cells.” By studying this pathway, scientists hope to develop a new class of painkiller with fewer side effects. Researchers at the University of North Carolina School of Medicine recently found an enzyme that “controls the activity of cellular receptors that signal pain...and a compound that could dampen the activity [of a lipid that is present in higher levels in pain-sensing neurons located at the beginning of the pain pathway]. ... This compound... could lead to a new kind of pain reliever for the more than 100 million people who suffer from chronic pain in the United States alone.”

Pain management generally begins with medication, followed by targeted procedures, including surgery in some cases. Counseling and alternative medicine, such as acupuncture, are also effective approaches that may be added.

If you, a friend, or family member is living with chronic pain, it is important to see a physician who is a pain specialist. It often takes more than one medication or other therapy to solve chronic pain. The brain, spine, and nervous system all interact to create the sensation of pain. Neurotransmitters, chemical messengers that pass nerve signals along to the brain, and brain chemicals that affect both emotions and the feeling of pain, can be targeted by medications. Physicians who are pain specialists have the knowledge and experience to prescribe the combination of medications and other treatments that will best manage chronic pain. It is better to see a pain specialist early rather than later allowing for timely investigation and possible treatment of the source of the pain. For example, a broken arm needs to be set, but the associated pain also needs to be addressed. “If initial pain from an injury is not adequately treated, those pain signals are sent repeatedly which leads to changes in the central nervous system, making it more and more sensitive. Over time, even the gentlest touch can become very painful;” this is known as “central sensitization.”

While pain medicine is not yet recognized as an independent residency program in the United States, a number of different specialists may have completed a fellowship or equivalent training to become board-certified in Pain Medicine. These clinicians include anesthesiologists, neurologists, physiatrists (rehabilitation), neurosurgeons, orthopedic surgeons, rheumatologists, and psychiatrists. “… Be sure to find someone with the proper credentials, such as... certification from the American Board of Pain Medicine” (http://imis.abpm.org/abpmimis/abpm/directory.aspx). If possible, it is best to go to a pain management center where care can be coordinated between the different specialists you may need. Pain management generally begins with medication, followed by targeted procedures, including surgery in some cases. Counseling and alternative medicine, such as acupuncture, are also effective approaches that may be added.

The first step, which is usually taken before a patient is seen by a pain specialist, is to find the source of the pain. For example, in VHL, if a treatable source can be found, such as a VHL spinal tumor, the source of the pain needs to be treated or removed. The next step in pain management usually begins with medication. This is a trial-and-error process where the experience of a pain specialist is very valuable. “… Doctors typically start with oral painkillers like acetaminophen and NSAIDs (nonsteroidal anti-inflammatory drugs)... These reduce inflammation and relieve pain, especially related to arthritis, tendonitis, nerve injury, mild to moderate cancer pain, and other forms of chronic pain.” Stronger medications that may be tried include anticonvulsants for nerve pain, antidepressants (lower doses than used for depression), and narcotics. Some medications can be administered topically instead of orally in cream or patch form. An example is a new 72-hour narcotic transdermal skin patch. Drugs may also be administered via an implanted pump that sends a painkiller to the spinal cord on demand, controlled by the patient.

Targeted procedures are another method that may be used to treat chronic pain. For example, a local anesthetic can be injected as a nerve block to suppress pain enough to allow participation in physical therapy. Radiofrequency ablation can be used to decrease pain signals from a small area of nerve tissue. Muscle pain may be treated with transcutaneous electrical nerve stimulation (TENS) therapy, with two electrodes transmitting low voltage current through the skin along a selected nerve path, or trigger point therapy, an injection of local anesthetic often combined with a steroid. The spinal cord can be stimulated to block pain signals from reaching the brain using an implanted “pain pacemaker” controlled by the patient.

It is also important to address the psychological response to chronic pain as it can affect perception of pain and the ability to break the cycle of chronic pain. “In some people, anxiety or depression may have existed before the pain, while in others it comes afterward. For example, people who are chronically depressed may have a heightened awareness of their pain. Or pain can cause or worsen depression or anxiety especially when it is chronic or inhibits a person’s ability to pursue favorite activities.” Another reason to consider a psychological or counseling approach for chronic pain management is that “many patients with chronic pain have been in the traditional medical system for a long time. The expectation of doctor and patient is that the history, examination, and investigation will lead to a diagnosis which will result in a specific treatment and cure. The traditional disease method works for acute pain, but not for chronic pain, and relief is not forthcoming.”

Alternative medicine provides a number of therapies that can help patients manage chronic pain. These include stress management techniques, relaxation...
Have you Heard about the CGIP Databank?

The VHL Alliance launched its groundbreaking online research study, the Cancer in our Genes International Patient Databank (CGIP, www.vhl.org/databank) last spring with announcements in the April-June 2014 newsletter (including a centerfold brochure), email communications, and a featured placement on our website home page with a link “button” and participation “thermometer.” Articles from users and praise from the FDA director followed. Growth in the number of participants was rapid at first, but has slowed considerably over the winter and into the spring.

In order to learn more about possible barriers to participation, a short survey was posted on Facebook and Inspire and emailed to constituents who have not yet registered. So far, there have been three surprising results:

- Non-participants said they had not heard about CGIP
- Most participants found out about CGIP from Facebook or Inspire, not the VHLA website, or other forms of VHLA communications
- Approximately 30% of respondents are interested in a paper option for taking the surveys.

In order to find answers for many of your questions about VHL, we must be able to point research in the right direction and connect potential research subjects with researchers. This means that it is important for everyone connected with VHL to participate. Information can be entered for any family members with VHL as well as for yourself.

We need your help to make CGIP a success and shorten the time to find effective treatments for VHL.
How Can I Help Promote CGIP?

Survey Monkey questions (we’d love to hear your response)—follow the link or print and complete the questions below and mail it to the VHL Alliance:

www.surveymonkey.com/s/WGNWVVP

1) Please enter your name, phone number, and email so we can contact you to follow up on your interest in promoting the CGIP Databank.
   Name: ___________________________
   Phone Number: ____________________
   Email: ___________________________

2) I have VHL (please select all that apply):
   a. I will enter my own data
   b. I have already entered my data
   c. I have already entered family member data
   d. I will enter family member data
   e. I do not have any relatives with VHL

3) I do not have VHL:
   a. I have already entered family member data
   b. I will enter family member data
   c. I do not have any relatives with VHL

4) I am interested in helping others participate in the CGIP Databank:
   a. yes
   b. not sure
   c. no

5) I would like to help others participate in CGIP by (please select all that apply):
   a. phoning non-participants
   b. helping non-participants use the online CGIP software
   c. working in person with one of the VHL Clinical Care Centers
   d. other (see next question)

6) I have another idea on how to increase CGIP participation and/or how I can help:
   Please write on a separate sheet of paper

What is “Shared Decision-Making” in Healthcare?

You may have heard the term, “shared decision-making,” from one of your doctors, seen it mentioned in an article, or encountered it when taking a survey on care you received at a doctor’s office, hospital, or through your health plan. The concept of “patient-centered care,” was defined by the Institute of Medicine in 2001 as “care that is respectful of and responsive to individual patient needs, preferences, and values and that ensures that patient values guide all clinical decisions.”

The US federal government states that “shared decision-making is a model of patient-centered care that enables people to play a role in the management of their own health. It operates under the basis that, armed with good information, consumers can and will participate in the medical decision-making process by asking informed questions about their conditions and treatment options.”

The federal government considers shared decision-making to be an “intervention” in patient-physician communication and requires that it be measured annually for Medicare and Medicaid health plans. It is also measured as part of accreditation and performance ratings for commercial health plans, hospitals, and surveys on patient satisfaction with physicians.

Research conducted on the benefits of the shared decision-making approach found patients had better health outcomes, increased compliance with treatments, and lower demand for health care. This measurement is also increasingly tied to reimbursement for hospitals and health plans and compensation of doctors.

Shared decision-making becomes more important in management of complex, lifelong conditions such as VHL. “In many situations, there is no single ‘right’ health care decision because choices around treatment, medical tests, and health issues come with pros and cons. Shared decision making is especially important in these types of situations: when there is more than one reasonable option, such as for screening or a treatment decision; when no one option has a clear advantage; and when the possible benefits and harms of each option affect patients differently.”

As a patient, you now have more responsibility to be an active member of your healthcare team. Unlike your grandparents and possibly your parents, you should not assume a one-sided conversation where the doctor tells you what the health problem is and the treatment that will be prescribed; instead, there should be a conversation where both the diagnosis and treatment options are discussed. The treatment that is finally prescribed is a result of an agreement between you and the doctor. In order for shared decision-making to work, it is important to be knowledgeable about your health condition, treatment history, and the options available. The VHL Alliance makes knowledge about VHL and treatments for it easily accessible to help you prepare for each of your appointments.

Many doctors are still working on the best ways to incorporate shared decision-making into their day-to-day office routine—they may have gone through medical school and residency when a one-sided conversation was an accepted practice. Today, doctors are frustrated by time pressures to see higher numbers of patients, some have experience with patients who don’t know a lot about their medical condition, or with patients who “come from cultural backgrounds that lack a tradition of individuals making autonomous (their own, independent) decisions.” However, in today’s environment, most doctors should welcome your detailed knowledge of VHL and your research into the treatment options. Your preparation prior to your appointments may even include a list of questions to refer to so that you do not miss asking something important in person. This will help to have a shared decision-making process take place. When a decision on medical treatment must be made, “patients need to be involved in determining...
May is VHL Awareness Month!

Below are some of the events happening in May:

- Phoenix, Arizona, May 9: Cocktails for a Cause
- Ava, Missouri, May 9: Bake Sale
- New York City, May 14: Dance to Find a Cure
- Bronx, New York, May 21: Van Cortlandt Track Club Race. Contact arafael13.vhl@gmail.com

May is VHL Awareness Month

How will you help raise awareness?

JOIN US FOR THE MONTHLY TELEPHONE DISCUSSION GROUP

http://tinyurl.com/telephone-discussions • Call the VHLA office

HAPPENINGS AT VHLA

Hatfield-McCoy Marathon

Join Team VHL at the Hatfield McCoy Marathon and raise money for VHLA.

WHEN: Saturday, June 13, 2015
WHERE: Kentucky and West Virginia

There are a few openings left! You can also join as a virtual runner. See http://tinyurl.com/hatfield-mccoy-marathon-vhl for all the details.

Dance to Find a Cure

On Thursday, May 14th, VHLA Board Members Manuel Greco and Bettina Micheli are hosting “Bless this Mess—do the #vhlwarrior dance”.

Place: The Panther Room, 74 Wythe Ave, Brooklyn, NY Time: 10:00 PM - 4:00 AM
Contact: manuel@vhl.org

LET US KNOW WHAT YOU PLAN TO DO FOR MAY!

Mom’s Day Run 2015

Come join us for the 5th annual Mom’s Day Run to benefit the VHL Alliance. This is a Fun Run or Walk along the beach with your mom, in honor of your mom, or in memory of your mom.

8 am (check in at 7 am), 901 San Pedro St, Ventura, CA

www.tinyurl.com/MomsDayRun-Registration15

the management strategy most consistent with their preferences and values.\(^5\)

Shared decision-making “might seem like common sense: each patient has different priorities and preferences; what is right for one patient may be wrong for another. Of course patients should weigh in. But many aren’t accustomed to speaking up. Even the most engaged or educated patients may defer to their doctors because they are scared, they don’t want to be seen as difficult, or they think the doctor knows best.”\(^6\) A program to help patients with shared decision-making at the University of California at San Francisco pairs patients with volunteer note-takers. The volunteers help the patients to put together a question list prior to each physician appointment and take notes for the patient during the appointment. This helps both the patients and the doctors. In addition to bringing your own notes in with you to appointments, you may want to consider having a family member or friend with you to take notes. It can be very difficult to remember what was said or to make your own notes when you are dealing with the emotions as well as the facts being discussed.

As a patient with a rare disorder, you are probably comfortable with explaining VHL to doctors and may even have educated a doctor about some of the possible treatment options. Now that you have a better understanding of the doctor’s requirements and point of view, you should feel even better about the important role you play in managing your own healthcare.

5. Anna Gorman; “Letting Patients Call the Shots”; The Atlantic; March 2015; accessed online 3/23/15
EVENTS OF OCTOBER 16-18, 2015 • CHICAGOLAND, IL

FUNDRAISING DINNER & AUCTION
October 16, 2015: 6:30-10:00 pm • Meridian Banquets and Conference Center, 1701 W Algonquin Rd, Rolling Meadows, IL 60008
See: vhl.org/meeting/dinner2015

Honoring
Sarah Nielsen, MS, CGC
VHL Alliance Board Member, Genetic Counselor, University of Chicago

Jan & Scott Capinegro
Supporters of the VHL Alliance

2015 VHL ANNUAL FAMILY MEETING
October 17, 2015: Registration: 8:00 AM • Meeting: 8:45-4:45 PM • Meridian Banquets and Conference Center
See: vhl.org/meeting/meeting2015

AGENDA
• Introduction and Welcome: Raymon Grogan, MD, FACS: Assistant Professor of Surgery, Univ. of Chicago
Ilene Sussman: Executive Director, VHL Alliance
• VHL: 101: Sarah Nielsen, MS, CGC: Genetic Counselor, Univ. of Chicago
• Retinal Angiomas: Different Techniques for Treatment: Michael Blair, MD: Clinical Associate of Ophthalmology & Visual Science, Univ. of Chicago
• You, Your VHL, and Your Urologist: Scott Eggener, MD: Associate Professor of Surgery, Univ. of Chicago
• Exocrine Pancreatic Insufficiency: Andres Gelrud, MD, MMSc: Associate Professor of Medicine, Univ. of Chicago
• Talking to Children about Genetic Disorders: Ken Onel, MD: Assistant Professor of Pediatrics, University of Chicago.
Lindsay Rhodes, MS, CGC: Genetic Counselor, Univ. of Chicago
• The Emotional Roller Coaster: Separate Break-out Sessions: Patients: Amy Siston, PhD: Psychologist, Univ. of Chicago
Caregivers: Nancy Beckman, PhD: Psychologist, Univ. of Chicago
• Brain and Spinal Lesions: Not Just a Science: Rimas Lukas, MD: Assistant Professor of Neurology, Univ. of Chicago
• State of VHLA, 2015: Karen Ramsey: VHLA Board of Directors
• Update on VHLA-Funded Research: Eric Jonasch, MD: Associate Professor, MD Anderson Cancer Center, Houston, TX
• Adrenals and Management of Hypertension: Raymon Grogan, MD, FACS: Assistant Professor of Surgery, Univ. of Chicago
Colleen Majewski, MD: Assistant Professor of Surgery, Univ. of Chicago
• ELSTs and Surgical Techniques: Michael Gluth, MD: Assistant Professor in the Dept of Otolaryngology, Univ. of Chicago

2015 TEAM VHL 5K RUN/WALK
October 18, 2015: Check in: 8:00-9:00 AM • Run/Walk: 9:00 AM
Hyatt Regency Schaumburg, 1800 E Golf Rd, Schaumburg, IL 60173 (register in lobby)
See: www.crowdrise.com/teamvhl2015 (virtual runners/walkers welcome)

If you’d like to donate an item for the auction, please contact us at 617-277-5667 ext. 4.

REGISTER NOW!
See all three events and register on-line at http://www.vhl.org/meeting
or mail in Registration Form below to:
The VHL Alliance, 2001 Beacon St, Suite 208, Boston, MA 02135

MAIL-IN REGISTRATION FORM

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DONATION □ $ ______________

SPONSOR A RUNNER □ $ ______________

Show your support! Put a message in the program:
□ $25.00—your name among the well-wishers
□ $50.00—3-line message (enclose your text—maximum 100 characters)
For larger ad options, write to director@vhl.org

Total Enclosed $____________
Donations are seen on the printed version only.

Your help is greatly needed

We also need volunteers. Call 1-800-767-4845 ext. 4
2015 TEAM VHL 5K RUN/WALK
Sunday Morning, October 18, 2015
Hyatt Regency Schaumburg, Illinois
www.crowdrise.com/teamvhl2015

2015 VHL ANNUAL FAMILY MEETING
Saturday, October 17, 2015
Chicagoland, Illinois
www.vhl.org/meeting/meeting2015

2015 FUNDRAISING DINNER & AUCTION
Friday Evening, October 16, 2015
Chicagoland, Illinois
HONORING: Sarah Nielson, MS, CGC, University of Chicago, IL
Janice and Scott Capinegro, Barrington, IL
www.vhl.org/meeting/dinner2015

REGISTER NOW!

HONORING: Sarah Nielson, MS, CGC, University of Chicago, IL
Janice and Scott Capinegro, Barrington, IL

Chicagoland

www.crowdrise.com/teamvhl2015