



Dr. Eric Gou Protect the Future's Newest Doctor *The Protect the Future (PTF) program is one of the most important programs the APF. To explain, the present porphyria experts have been the major source of porphyria expertise for over thirty years. Having men and women of this caliber is an extraordinary treasure, however, all of them are approaching retirement age. When this occurs, their expertise will be lost if we do not train young experts as a means to protect the level of knowledge about porphyria already attained, as well as protect our future health. Thus, the APF initiated the PTF program to have the present expert train the next generation of experts. These young men and women not only learn from mentors in the US, they are also included in the international conferences where they receive in depth knowledge from some of the most esteemed porphyria experts in Europe. The PTF doctors and their mentors diagnose and treat patients together, as well as collaborate on research projects, medical journals and medical texts.*



We are proud to announce that our newest future expert is **Dr. Eric Gou**. A native of Houston, Dr. Gou moved to Austin at age twelve. He spent a few years outside Texas for undergraduate

studies at Johns Hopkins University in Baltimore, Maryland where he studied biology and psychology and graduated *cum laude*. Next he attended the University of Texas Southwestern Medical School in Dallas, Texas and met Michelle, his "significant other" of four years. Michelle is currently a second year resident in Pathology at the University of Texas Medical Branch in Galveston, Texas where Dr. Gou is pursuing porphyria research with expert, Dr. Karl Anderson.

Dr. Gou loves road cycling, swimming and watching cooking shows and trying to replicate the dishes at home. He combined his love of cycling with a challenging volunteer effort for the benefit of cancer research. Dr. Gou biked from Baltimore to San Francisco to raise money for the American Cancer Society while teaching communities across the country about cancer awareness and prevention. In addition, he organized transportation and lodging in the communities involved. He also worked with officers of the Asian Pacific American Medical Student Association to organize the Hepatitis B Awareness Forum, as well as organizing transportation and lodging for regional and national conferences. We are fortunate that young doctors of Dr. Gou's experience are part of our PTF program.



Dr. Ashwani Singal is another PTF Doctor on our team. We are very proud that an article about his research project, which began as Dr. Singal's MS thesis, *Low-Dose Hydroxychloroquine is as Effective as Phlebotomy in Treatment of Patients with Porphyria Cutanea Tarda (PCT)* has recently been accepted by the prestigious journal, *Clinical Gastroenterology and Hepatology*. The paper describes initial findings from the study that will continue as a research project. PCT is an iron-related disorder caused by reduced activity of hepatic uroporphyrinogen decarboxylase (UROD); it can be treated by phlebotomy or low doses of hydroxychloroquine. The research team, Csilla Kormos-Hallberg, M.D.; Chul Lee, Ph.D.; V.M. Sadagoparamanujam, Ph.D., James J. Grady,

Ph.D.; Daniel H. Freeman Jr., Ph.D.; and Karl E. Anderson, M.D. also shares authorship on the journal article and will perform the prospective pilot study to compare the efficacy and safety of both therapies. The aim was to show that time to remission, as defined by a normal plasma porphyrin concentration, is noninferior for HCQ when compared to phlebotomy. Phlebotomy is the more accepted treatment in the U.S. The data in the paper indicates they are equivalent, but statistical significance for noninferiority was not achieved. The researchers analyzed data from 48 consecutive patients with well-documented PCT to characterize susceptibility factors; patients were treated with phlebotomy (450 mL, every 2 weeks until they had serum ferritin levels of 20 ng/mL) or low-dose hydroxychloroquine (100 mg orally, twice weekly, until at least 1 month after they had normal plasma levels of porphyrin). The research team compared the time required to achieve a normal plasma porphyrin concentration (remission, the primary outcome) for 17 patients treated with phlebotomy and 13 treated with hydroxychloroquine. The result was that the time to remission was a median 6.9 months for patients that received phlebotomy and 6.1 months for patients treated with hydroxychloroquine treatment, a difference that was not significant, 100 mg twice weekly, is as effective and safe as phlebotomy in patients with PCT, although noninferiority was not established. Given these results, higher-dose regimens of hydroxychloroquine, which have more side effects, do not seem justified. Compliance was better and projected costs were lower for hydroxychloroquine than phlebotomy treatment. Long-term comparative studies are needed.

Support the PTF Program You may ask how does the APF facilitate training almost 20 young doctors, which is a very costly program. Like anything else the APF accomplishes, it is through your help donations and those of your family, friends, industry, donors and all who recognize that without training young doctors, porphyria expertise will be lost. We need your help please. Future experts are essential for your future health. They are the ones who will be writing the text books your doctors will be reading to know how to treat you. They will be performing the life saving research. They will be the ones consulting with your physicians. Please make a special contribution to this program and mark your special donation PTF. *Thank you!!!*

More Porphyrin Research



The American Porphyrin Foundation (APF) is very active in recruiting patients for research, as well as raising funds from members, private donors and industry to support life changing research. Below are a few of the recent research projects that need our members support.

Dr. Herbert Bonkovsky and his research team at the Carolinas Health Care Systems in Charlotte, NC, plan to discover how heme exerts its beneficial, as well as its toxic effects, in order to improve their knowledge of the manifold effects of this primordial molecule and in order to improve the health and well-being of patients with porphyria. Heme is a molecule formed from iron and protoporphyrin upon which most life on earth depends. Heme makes our blood red and helps our hemoglobin to carry oxygen to our tissues and carbon dioxide away from our tissues. Normally, levels of heme in

our tissues are closely regulated: Excesses of heme cause decreases in new hemesynthesis and increases in heme breakdown. How heme does this is not well understood as in the porphyrias in which heme synthesis is deficient and/or in which potentially toxic precursors of heme accumulate. Some of these diseases can be treated by giving heme intravenously, but excess heme, too, may be toxic, so there is much to learn in this new project.

Dr. Karl Anderson's research/laboratory center also has a longstanding interest in the human porphyrias. Their research focuses on developing new therapies and strengthening the evidence for existing therapies. They primarily design and conduct prospective studies, rather than retrospective studies of existing records. This research requires significant numbers of well-characterized patients with whom they have positive ongoing relationships. Many patients with porphyria are inadequately diagnosed and managed, because there are few specialists with adequate experience to oversee the complexities of porphyria diagnosis and treatment. To engage enough patients countrywide, the research team needs to provide advice on what they need in terms of more accurate diagnosis and better treatment. Other centers have mostly emphasized nonhuman studies or molecular research that does not require prospective relationships. Engagement with the extended community of porphyria patients also highlights issues that the community thinks need further research. To find and establish with enough patients for research, Dr. Anderson's team interacts frequently with patients, family members and physicians at a distance and respond to their requests to provide advice on diagnosis and treatment. They also maintain the only laboratory in an academic institution in the US that is certified to accept samples for diagnostic biochemical testing and issue reports that include detailed interpretations of the results. For information about other clinical trials, see www.clinicaltrials.gov. ClinicalTrials.gov is a [registry](#) and [database](#) of federally and privately supported clinical trials conducted in the United States and around the world. ClinicalTrials.gov gives you information about a trial's purpose, who may participate, locations, and phone numbers for more details. This information should be used in conjunction with advice from health care professionals. **IF YOU HAVE AN IDENTIFIED MUTATION, PLEASE CONTACT THE APF AS THE CONSORTIUM NEEDS YOUR INPUT asap!!**



PANHEMATIN®, a therapy for the Acute Porphyrias, is used effectively in treating attacks and preventing attacks. Approved over 25 years ago as the first Orphan Drug, some doctors are still not familiar with treatment. The APF continues to educate physicians on the benefit of Panhematin® and provides comprehensive information and instruction on its use. If you would like your physician to receive this physician education packet, please contact the APF. It is very important for all doctors to know about the clinical benefit of Panhematin®, because attacks of the Acute Porphyrias can progress to a point where irreversible neuronal damage can occur. Panhematin® is intended to prevent an attack from reaching the critical stage of neuronal degeneration. The clinical benefit depends on prompt administration of Panhematin®. Unfortunately, Panhematin® is not effective in repairing neuronal damage. The exact mechanism by which hematin therapy produces symptomatic improvement in patients with acute attacks has not been determined. However, your input on the Longitudinal Study and other studies being conducted by the researchers should prove very helpful to answer these and other questions.

If you want to visit an outstanding site featuring information for you and your physician on the Diagnosis and Treatment of the Acute Porphyrias, see: www.aiporphyria.com. This website is filled with [videos](#), [images](#) pertinent to the [acute porphyrias](#). Superb animations illustrating the normal and disease heme biosynthetic process, as well as the role of Panhematin® can be found on the same website at: www.aiporphyria.com/heme-biosynthetic-pathway.html. You can also view a video of APF member, Tracy Yelen, as she describes her experiences during attacks and after treatment with Panhematin®. Take a look at: www.aiporphyria.com/living-with-aip/real-patients-real-stories.html. Don't forget that the APF website also has a Panhematin® section that includes the stories of patients who have been prescribed Panhematin® treatment. Their stories will help answer many of your questions about the treatment.



The Rare Diseases Clinical Research Network (RDCRN) is composed up of 18 distinctive consortia that are working in concert to improve availability of rare disease information, treatment, clinical studies,

and general awareness for both patients and the medical community. The RDCRN also aims to provide up-to-date information for patients and to assist in connecting patients with advocacy groups, expert doctors, and clinical research opportunities. The Porphyria Consortium is one of these important research groups. Despite their rarity, many fundamental advances in medicine have come from the study of rare diseases and these have benefited common diseases. The consortia will meet October 2, 2012 in Rockville, Maryland. Attendees will meet key leaders in rare diseases research, engage in formal and informal networking and idea generation with trainees and investigators, discuss career development and strategic planning for academic success and explore research collaborations.

The Porphyrias Consortium includes six of the leading porphyria centers in the United States. These centers provide expertise and experience in the diagnosis and treatment of patients with porphyria. The staff in each Center includes porphyria physicians, researchers, research coordinators, and technical/laboratory staff. Together with the American Porphyria Foundation, the Porphyrias Consortium enables a large scale collaborative effort to develop new strategies and methods for diagnosis, treatment, and prevention of illness and disability resulting from these rare disorders. The Consortium website is a forum for disseminating information to porphyria patients and their families, healthcare professionals, and other interested parties. The website also provides a list of currently active studies with a description of each and contact information for those who are interested. Of particular importance is the Longitudinal Study with the objective of characterizing the long term course and outcomes of each porphyria. Clinical trials are being conducted to establish the safety and efficacy of new therapies leading to approval by the US Food and Drug Administration. Our future health depends on the work of the Consortium. To learn more or to join their research efforts, see the following website, <http://rarediseasesnetwork.epi.usf.edu/porphyrias/index.htm>.

Normosang comes to Canada. Medunik Canada, is an orphan drug company dedicated to helping Canadians with rare diseases access therapies currently unavailable in Canada. Medunik Canada recently announced that it has signed an exclusive distribution agreement with Orphan Europe Recordati Group. Orphan Europe, a pharmaceutical company dedicated to the development and distribution of orphan drugs for the treatment of rare diseases, was founded in 1990. It was a privately owned company until December 2007, when it was acquired by Recordati, a European pharmaceutical group. Under their agreement with Orphan Europe, Medunik Canada receives the exclusive Canadian rights to market and distribute Normosang (heme arginate). Normosang, which is a heme therapy similar to Panhematin®, is registered as a standard of care for acute porphyrias in most European countries. It is still not available in the USA due to FDA requirements relative to the blood bank in Finland where Normosang is manufactured.



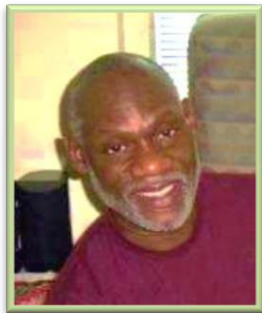
Another Research Project is underway at five porphyria centers. If you live within driving distance of any of the following five Porphyria Centers, you are needed to participate in this important research: University of Texas Medical Branch, Galveston, TX; University of Alabama, Birmingham, AL; University of Utah, Salt Lake City, UT; University of California San Francisco, CA; Mount Sinai Hospital, New York City, NY.

The official title of the study is *Quantification of the Effects of Isoniazid Treatment on Erythrocyte and Plasma Protoporphyrin IX Concentration and Plasma Aminolevulinic Acid in Patients With Erythropoietic Protoporphyrin (EPP)*. In EPP there is an accumulation of protoporphyrin IX in the plasma and liver. The reason it builds up is either the last step to make heme, insertion of iron into protoporphyrin IX, is rate limiting or there is an increase in activity in the first step in the heme pathway. It may be possible to decrease the amount of protoporphyrin IX made and see a decrease in symptoms. The first step to make heme is the key step in the pathway and it uses vitamin B6 as a cofactor. If the investigators can limit the amount of vitamin B6 the investigators can possibly reduce the activity of this rate limiting step. With decreased activity of the enzyme it may be possible for the body to utilize all the protoporphyrin IX that is made so that none builds up.

The researchers, Drs. Karl Anderson, Joseph Bloomer, Montgomery Bissell, Herbert Bonkovsky, and Robert Desnick and John Phillips, Ph.D. who devised the study, will be studying the use of ISONIAZIDE, which is a drug currently used for Tuberculosis but in this case will be used for EPP. Patient volunteers will return to the center every two weeks for monitoring. All EPP patients are eligible for the study except those who are currently in the present Phase III clinical trials with *Scenesse*. Since *RESEARCH IS THE KEY TO OUR CURE*, everyone who participates in research as a patient volunteer is extremely valuable to the future of every person with porphyria. If you are interested in participating, please contact the APF at porphyrus@aol.com or call 866 APF 3635.

Nathan Wayne Carr has quite the story to tell. He was born in Germany, lived all around the US and now resides in Charlotte, NC. You will see from his story that despite his worldwide search for a correct diagnosis, he was not diagnosed properly until he was almost 40 years old.

At an early age, Nathan says his skin looked like it had been eaten by bugs and was infected with sores. This continued throughout high school and beyond that prevented him from being in the sun without burning terribly. Then new symptoms were added, labored breathing, searing abdominal and leg pain, nausea, and a host of other symptoms all of which required medication and frequent lengthy hospitalizations. During his years at Howard University, Nathan was thought to have Sickle Cell, which proved false. From ages 30 to 40, he had numerable surgeries, none of which helped his symptoms. Finally, Dr. Marjorie Smith, Anchorage, Alaska discovered he had Hereditary Coproporphryia (HCP). Life was still challenging, especially while trying to raise his children by himself and not miss work due to his many attacks. His doctor tried to talk him into early retirement, he was proud to be working with two major companies. Eventually, he had to retire because of his frequent attacks and severe photosensitivity. Sadly, Nathan's misadventures, while wearing sun protective gear, included being mistaken as a robber at the bank ATM and chased by police in the car. Such incidences have happened so often that Nathan has contacted the local police department to advise them of his circumstances and notifying businesses that he is coming that day. Despite carrying his necessary paperwork and medical alert bracelet and calling the North Carolina Disabilities Rights Office, he continues to be harassed so much he now only goes out if necessary. Seven family members have had the genetics test and 5 are positive. His mother, 2 sisters, 1 niece and he, all suffer from this illness in their own way. For the past 2 years he can no longer tolerate sunlight, UVA/UVB rays. They did not understand at first because there was so little information. *See his full story on the APF website.*



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Hollyanna Cougar Tracks de Coteau Pinkham is a power packed name for a power packed woman.



Hollyanna is a Native American who hails from the Yamaka nation and is a dynamo for the people who need health care. Hollyanna discovered the APF when she joined the APF's EPP FACEBOOK group while seeking to learn more about the disease. After enduring four bouts of Melanoma, she was diagnosed with EPP after a lengthy series of misdiagnoses. Not wanting to let her photosensitivity and melanoma dictate her life, Hollyanna decided she would "work at living and not live at work." After time at the fire and police departments, she tired of having to work the graveyard shift to keep out of the sun and made some major life changing steps. First, she decided to finish college and subsequently took on a position as the Homeland Security Emergency Management Planner. At the same time, she began advocacy efforts to improve healthcare on the reservation. Her efforts have been so successful, she has been asked to testify for Congress on the lack of health care in rural areas, a fact she feels jeopardized her life after her melanoma and EPP diagnoses were missed. She feels her

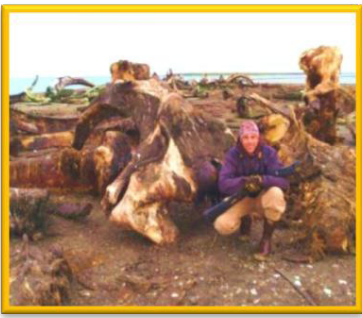
health advocacy and Homeland Security work are ensuring better life for her three sons and one daughter.

For pleasure, Hollyanna loves to hunt, fish, play golf and build double plateau saddles, which is a dying art form. In fact, Hollyanna is the last known traditional saddlemaker among Native Americans and is publically recognized for her talent. These saddles are taller than regular saddles and have horns on both ends and were used by the plains and plateau tribes for packing three "rawhide packs" on each side of the horse. Although her life seems to be one of teaching, she says she learned a lot from her FACEBOOK friends like how to make her own photoprotective clothes with a Ritz sun block product and how to use prosthetic contact lenses. She has even met other Native Americans on the Facebook group as well. *Welcome to the APF!*

Margarita Cianchini will be helping the APF as a Spanish translator. During the year, we have quite a few



people from Mexico, and Central and South America who contact the APF for help, but they speak Spanish only. Elizabeth Pettit, whom many of you know from the APF office, has done an excellent job translating but sometimes the patient's requests are too complex to understand. We are very excited that Margarita has volunteered to translate for us. Margarita has AIP and lives in Miami, Florida but is from San Juan, Puerto Rica and speaks Spanish. Margarita is also promoting porphyria awareness within the Mount Sinai Hospital in Miami. The APF has repeatedly said that our members can change the world of porphyria one patient at a time. Not only can YOU teach your doctor about your type of porphyria, but you can take a tip from Margarita and solicit your hospital's help to teach their entire medical staff. *Thanks Margarita!*



TYLER ZOLLINGER says, *"I am wired for Shadow Jumping"* and have never known what it is like to live without EPP. With four out of my seven brothers having EPP, too, sun protection was a way of life for us. I live covered up and take precautions like a neoprene face mask, protective clothing and whatever it takes to protect myself, because I don't let EPP slow me down. To me, coping with EPP is like having a "sun bank account." Tyler described his coping mechanism with EPP as having X number of "sun credits," which he uses sparingly as he spends time in the sun. "If you use too many, you will pay dearly, but if you use them wisely, you can spend more time in the sun. Basically, you live with the sun in the middle of your life." That system and

his protective gear enable Tyler to do the things he loves, like rafting, the ocean, and even camps by creating a "black-out" tent. Tyler has a perfect job for an outdoorsman. He is District Manager of Total Safety, US, a company that provides security for oil workers, environmentalists and scientists who are working in Alaska. The above photo is of Tyler sitting by whale bones. He is armed to protect the environmental researchers from the big bears who lived near their research spot. He is also thrilled with his new opportunity to participate in the *Scenesse* clinical trials. Despite the arduous 4 1/2 hour flight one way from Anchorage to the Salt Lake City Porphyria Center, Tyler volunteered to join the clinical trials. He received his first implant, which he says felt more like a small stick being implanted than a the usual grain of rice description. Dr. Charles Parker inserted the implant and within a few days, Tyler felt his ability to stay in the sun has quadrupled and expects it to increase more in the next six months. He is grateful for the opportunity to be a part of a trial that hopefully will lead to a new treatment for EPP. Perhaps one day soon, Tyler will be walking in the full sun all day long instead of jumping from shadow to shadow as he has done his entire life.



Carrie Hunter was diagnosed with genetic PCT last year at age 33. Until December 2010, she had no signs of PCT. Soon she began to notice her hands were getting cuts and blisters and were overly sensitive, but she didn't know why and neither did the many doctors she visited, including a dermatologist who ran many tests and biopsies. Still there was no answer. Even after being sent to the Comprehensive Cancer Center, it still took six months and four more doctors to diagnose her correctly. That occurred only after a trip to the Caribbean and a subsequent hospitalization that her diagnosis was discovered and a treatment plan started. According to Carrie, *"From September to December I had a phlebotomy every week to regulate my ferritin and porphyrin numbers. My doctor consults with Dr. Karl Anderson to help keep my levels normal, especially since I still have major issues with sensitivity to iron and foods. I have made big*

changes. I don't drink alcohol, eat any meat or iron rich foods or eat things cooked in iron and I avoid the sun. I once heard someone describe it as shadow jumping... and that is exactly what I do." Despite her many ongoing challenges with PCT, Carrie volunteers at the Children's Hospital near her home in Denver. Carrie wants to give back because she says, *"I have a beautiful husband and three children to be grateful for, and I live a fulfilling life. Porphyria has been difficult to adjust to but has not ruined my life!! I am grateful to Desiree Lyon and all the wonderful folks at the APF and my APF Facebook friends, who also have made this all easier to deal with--I finally don't feel so alone!"*



FACEBOOK If you have not joined one of the APF FACEBOOK groups, you have missed the pleasure of getting to know people who have your type of porphyria and hearing their experiences while sharing your own. We have almost 1000 FACEBOOK members now, who participate in groups that represent every type of porphyria; AIP, VP, HCP, ALAD, EPP and PCT. The main APF group is an

"open" group, which means you can enter the group and see what is being said. This is a good option for people who have never been involved with FACEBOOK. The porphyria specific type groups, however, are closed, which means you must join the group to view what is being shared among the members and to post your own comments. This option is an excellent means of protecting your privacy while still sharing your experiences in a friendly, atmosphere. The Banner was designed by Aletta Longari, a graphic artist, who shared her amazing talent to create our FACEBOOK banner.

Friends Around the Globe contact the APF for help--Argentina, Australia, Austria, Belgium, Brazil, Canada, Chile, Czech Republic, China, Columbia, Costa Rica, Denmark, England, Egypt, El Salvador, Estonia, Finland, France, Georgia, Germany, Greece, Guatemala, Guam, Hungary, India, Ireland, Israel, Italy, Japan, Mexico, Netherlands, Norway, Pakistan, Paraguay, Peru, Philippines, Poland, Portugal, Qatar, Romania, Saudi Arabia, Scotland, Spain, South Africa, Sweden, Switzerland, and Wales. The APF has helped many international friends when they were ill and also when they wanted to create an organization like the APF by sharing our information, organizational structure and tips to locate members and experienced doctors.





Aletta Longari, an active member of our APF VP/ FACEBOOK group. She hails from South Africa where Variegate Porphyria is the most common genetic disease and has kindly shared her porphyria story below and in full on the APF website. "Like a typical youngster, I rolled my eyes when my mom gave me 'that look' and said; *'Jy moet yourself laat toets my kind! Van lekker lag kom lekker huil.'* Translated: *'You have to get yourself tested, my child! From too much laughter, comes crying.'* I grew up in a loving home. My mother was a retired nurse with a passion for the medical field and a relentless urge to help people. My father loved teaching, literature, fishing, rugby and a Sunday braai (barbeque) with friends. I knew he had a blood condition that made him sick and he wore gloves when driving.

I finished school a few days before my 18th birthday and was full of hope that my new life would be everything my teenage years weren't, as they were riddled with depression, failure, pain and the unfounded thought I was dying of cancer. Debilitating headaches, flank aches, constant nausea, fatigue, dizziness, anxiety, an itchy forehead and depression made up an average day. My pride, fear of judgement and simple teenage ignorance kept me from seeking help. Despite my symptoms, I set out at 18 for a new life in the big city. Again, I found myself alone and trying to live life as an adult while thinking like a child. Stress, lack of sleep, alcohol, poor diet and too much sun turned my life into a misery. The worse I felt, the harder I partied. The harder I partied, the worse I felt.

At 20 years old, I had a root canal that resulted in blinding pain. The doctor promptly prescribed an anti-depressant which I took without thinking. That night, something that my mother said kept milling in my head, 'Always take half of the prescribed dose!' Even with half a pill, my world spiralled out of control again. My third full-blown Porphyria attack was triggered by a bout of flu and the medication a geneticist who was writing a paper Porphyria prescribed for me." Because of her health issues, Aletta was forced to resign her job and start her own business. Soon, she ran into a business colleague, Roby, who became her husband. At the birth of their son and daughter, she suffered more attacks and post natal depression, both of which were hard on Roby, too, but he has remained her "emotional rock."

When asked how Porphyria affected her life, Aletta replied,

~ **Personally;** *I lost my self-worth and confidence and guilt is an overwhelming reality.*

~ **Spiritually;** *I lost my creativity and my passion and lived in self-conviction.*

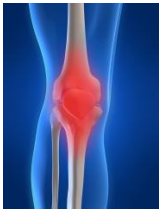
~ **Romantically;** *I lost simple moments to make lifelong memories that I could never re-claim.*

~ **Socially;** *I lost the respect of many through the years, and still have to tolerate naïve comments.*

~ **Physically;** *I lost my strength and became all I feared, such as being fat and full of scars.*

~ **Financially;** *I lost jobs, business, and still loose clients and lack of money is a constant worry.*

But I gained a deep appreciation for life and a better understanding of the "Golden Rule"; the gift to help others which is a blessing itself; the ability to see beauty and celebrate it in everything and compassion and patience for the matters that matter. Above all, I regained my self-worth by sharing my story." (Aletta also created the FACEBOOK banner.)



APF Pain Project The APF has undertaken an enormous project—advancing awareness and understanding of porphyria pain among physicians. As individual states have tightened their requirement regarding pain medication, porphyria patients are paying a terrible price for the misuse by other people. Patients contact the APF daily to report yet another sad story about treatment denials or under-treatment for pain. To be sure, there are many drug abusers, but they should not be compared with people who are very ill with porphyria and suffering severe pain. Preeminent pain expert and internationally recognized neuro-oncologist, Dr. Kathleen Foley, is passionate about her work as a pioneer in the battle to avert needless pain and suffering and management of acute pain during the treatment process. Dr. Foley is recognized worldwide for her extraordinary pain relief work and her focus that governments have. The APF will be using much of her research and educational materials in the development of our PORPHYRIA PAIN PROJECT FOR PHYSICIANS. She and other pain experts have addressed the issue that drug abuse has hindered the need of people who are suffering badly. It is not infrequent that the APF receives a call with a person crying from the indignity of accusations that they were drug seekers. The APF will be distributing pain materials for physicians and hospitals, contacting state and national Health Departments, advancing awareness of porphyria pain at medical meetings and conventions and other national efforts.

Testing Questions There are certain questions we hear every day and one is "What preservative is in the urine collection for PBG and what if it is not in the bottle?" When you are given a brown or opaque bottle for a urine collection, it often has sodium carbonate— Na₂CO₃ in the bottom of the bottle. If it is not in the bottle, that is OK, too. Many people have thought that their tests were no good, because they didn't have the Na₂CO₃ in the bottle. Don't worry, the urine sample/test is good without it and your test results will be fine. If you have other testing questions, call the APF or email: porphyrus@aol.com.



EPP Hurts, Too! With EPP, a few hours in the sun can cause terrible swelling and burning as seen on Ryan's arm. Adam is so photosensitive he has to use a body suit when he must go in the sun. Though rare, Adam's case does illustrate the wide scope of photosensitivity among patients. Despite the level of photosensitivity, EPP patients anxiously await the result of the new EPP Phase III clinical trials with Afamelanotide/SCENESSE and subsequent FDA approval to make it available to all.



When Your Doctor Won't Treat You

Many of our members have reported that they have not been able to convince their physicians to prescribe treatment even though they suffer lengthy, severe attacks or painful bouts from sun and light exposure. Some members tell us that doctors have refused to treat porphyria patients. The APF believes that building better rapport and in turn, a good relationship will help gain access to better treatment. The physician/patient relationship is central to the practice of medicine and is the subject of many articles in prestigious journal articles, like *JAMA* and *Annals of Internal Medicine*. Most universities teach medical students before they enter hospitals to maintain a professional rapport with patients, uphold patients' dignity, and respect their privacy. They contend that the better the relationship in terms of mutual respect, knowledge, trust, shared values and perspectives about disease and life, and time available, the better will be the amount and quality of information about the patient's disease transferred in both directions, enhancing accuracy of diagnosis and increasing the patient's knowledge about the disease. Below are a few steps that will help the visit with your doctor:

- ***Research** your physician so that you choose the right physician for you. Generally, doctors have never had a porphyria patient but sometimes you may find one who has some experience. The APF has a list of experts and also a list of doctors who are knowledgeable about porphyria. Just call, and we will share our list of doctors.
- ***Be prepared** ahead of time with your list of questions and keep them very concise. This is the best way to show your doctor that you understand and respect his/her time constraints.
- ***Be direct** about your health situation and not chat aimlessly. Rather, briefly mention something friendly to lighten the visit. Doctors spend their whole day under stress and time limits, so breaking that tension might be just what it takes to lighten the mood and free up your doctor to show his friendly side.
- ***Ask questions** and don't be afraid to speak up and let your doctor know what you don't understand.
- ***Take notes** and bring them home. Write down the medication instructions and ask for brochures or other educational materials if they are available.
- ***Do not bring** materials printed from the internet. Most doctors do want to read prestigious medical journals and are usually very willing to read those that the APF provides. If you have not asked the APF to send your doctor a physician packet, please call or email the APF with your doctor's contact information.
- ***Advise the doctor** of the APF physician packet and the Health Professional Section on the APF website. Be sure to note that the educational material is written by experts.
- ***Tell your doctor** about any current and past health care issues, concerns, past health history, your recent tests and x-rays, important personal information, a list of your current medications, vitamins and supplements.
- ***Thank your doctor** if you feel as if you've experienced that compassion and friendliness you've needed.



In Memory

We are saddened to hear of the passing of dear family members and friends. Some of you have chosen to honor a life by making a gift to the APF. Your gifts will perpetuate the mission of the APF to enhance research, physician and patient education and provide patient support. Many of your loved ones have also become personal friends of our APF staff over the years. We have enjoyed the camaraderie we developed after first talking about porphyria and then sharing our personal lives with each other. Please join us in thanking: Oksanna Wolowec, Yara North America, Inc., George Simpson, Jr., Carlos and Linda Smith, Jerry and Martha Osborn, Mr. and Mrs. J.C. Cunningham, Valley Metal Services, LLC, LeRoy and Marlene Merkle, Wesley B Johnson, Jason Woulfin, Jane and Mark Goldman, Ann Underwood, Anthony J Haut for *Terri Easterwood*; Donald Johnson for *Peggy Lewis Johnson*; Janice Desilva-Aqui, Bill and Marge Clark for *Norma Faggs-Fout*; Randolph and Loretta Childs for *Clara Gayle Mitchell*; Rosalie Nielsen for *Stephen Kancsar, R.Ph.*; Kathleen Toelkes for *Donna Pagano*; Barbara Brown Witter for *David Brown*; George A Cortina for *James Howell*; Bob and Sandra Schuette for *Robert Sauntmon*; Fred L Story for *Lavonne A Story*; Roy and Joy Connell, Catherine and Robert Braddock, Amy Haussmann for *Velma Key*. Desiree Lyon Howe for *Alan Hunter*.



In Honor

We also thank those who donated to the APF in honor of a friend or family members: Carmille W Atwood for *Cason and Caul Cook*; Santa Sophia Academy for *Megan McCarthy*; Jerry Benedict, Rosalie Nielsen, Andrew Nielsen for *Lisa Kancsar*; Mary Ida for *Jill Gould*; Sharon I Koch for *Debbie Koch*; Eric S Gray for *Ralph M Gray*.



The information contained on the American Porphyria Foundation (APF) Web site or in the APF newsletter is provided for your general information only.

The APF does not give medical advice or engage in the practice of medicine. The APF under no circumstances recommends particular treatments for specific individuals, and in all cases recommends that you consult your physician or local treatment center before pursuing any course of treatment.

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What's New at the APF **www.porphyrifoundation.com**

Is Your Membership Up to Date? The APF is able to maintain our physician and patient education programs and many other services because of your support. We do not receive government funding to run the APF, rather we receive donations from you, your friends, your family and people interested in the porphyrias. Now we need your support for several programs that are very special.

First, our ***Protect the Future*** program to train future experts is important to our future health. Without experts, our primary doctors have nowhere to turn for advice and to learn about porphyria. This is a serious problem that we are trying to prevent by training young doctors, but we need your help funding this program.

We also have an enormous physician education program that distributes exceptional educational materials to doctors. Please help us produce these materials.

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