Hereditary Hemorrhagic Telangiectasia Foundation International, Inc.
(Osler-Weber-Rendu Syndrome)

DIRECT CONNECTION

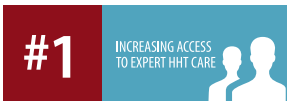
Volume 1 • 2014

For Family, Friends and Supporters

2014 IS THE YEAR OF ACTION FOR HHT DISEASE

Our medical experts tell us that we have never been closer to making disease-modifying progress in the fight against HHT disease. We know a cure is possible in our lifetime. We also know that too many people are exhibiting the signs and symptoms of HHT but yet are undiagnosed. And we know that more consistent quality of care for those who are diagnosed must be available for persons with HHT disease.

This year, the HHT Foundation International is taking bold steps. There has never been a more urgent time for you to stand with us as we aim to transform HHT disease.



We will continue to **increase the number of HHT Centers of Excellence throughout North America**. In addition to current application protocol, the HHT Foundation has established a mentorship program where proposed new Centers visit existing HHT Treatment Centers. The HHT Foundation ensures on-site evaluation with a full team of experts and facilities as part of the HHT Center certification.

We are also **launching a Physician Registry** which will be a searchable database of physicians that will appear on our website, www.hht.org, including their experience in caring for HHT patients. The registry will give patients the closest HHT center in addition to a list of doctors in their geographical area that are either medical professionals from current HHT Centers, specialists recommended by current HHT Center physicians, or physicians recommended by HHT patients. The database will include the physicians name, specialty, contact info, training history and procedural expertise. It will also include HHT research participation and experience. You will hear more about this in the coming months.

This year, the HHT Foundation is committed to developing relationships with **Bio/Pharma Companies** to either accelerate drug discovery efforts for HHT or repurposing their existing FDA approved drug therapies. Our goal is to develop and study new antiangiogenic therapy for HHT (reverse blood vessel growth). Through collaborations with Bio/Pharma companies, novel therapies are already under evaluation for proof of concept studies.

The HHT Foundation is looking for a long-term solution to improving the quality of life for HHT patients who suffer from moderate to severe nosebleeds. In 2014, the HHT Foundation will be announcing the results of its first sponsored Phase II Clinical Trial, **North American Study of Epistaxis** (NOSE Study), where three nasal spray agents and a placebo were tested on 120 HHT patients at six North American HHT Centers.

The HHT Foundation recognizes the critical importance of an HHT outcomes database. To this end, the HHT Foundation has solely funded this project and enlisted the expertise at the BioInformatics Service Center (BSC) at Dartmouth's Geisel School of Medicine to develop the **Outcomes Registry for HHT**. The purpose of this registry is to conduct natural history studies as a critical step in furthering research on HHT and to follow HHT patients on a long-term basis. Bio/Pharma companies have indicated that access to this **anonymous** patient data resource is a determining factor in their decision to invest financial resources in drug development for HHT.

HHT experts have been working collaboratively across North America to identify priority research questions to be answered via the Outcomes Registry for HHT. These questions relate to every aspect of disease (from nasal bleeding to stroke), with emphasis on determining the outcomes (severity of symptoms, quality of life, heart failure, stroke, severe bleeding, etc.), the personal factors that affect these outcomes (HHT genes and modifier genes, lifestyle, age, sex, etc.) as well as therapies. The answers to these questions can then assist in targeting clinical research studies of novel new therapies. The Outcomes Registry will include a module for patient entered data including HHT diagnosis, treatment, and quality of life. By linking all North American HHT Centers, the number of patients is increased twenty times over what any single HHT Center could gather and analyze which makes the Outcomes Registry a powerful research resource. The HHT Foundation will provide financial support to all participating HHT Centers in this endeavor.

In 2014, the HHT Foundation is committed to **launching a new awareness campaign** to increase outreach and public awareness of HHT as an un-recognized disease.

Look for several awareness building, advocacy and fundraising activities scheduled to take place during June's HHT Awareness Month. The HHT Foundation will also be providing HHT supporters tools to host their own fundraising event to showcase their involvement in the HHT movement.

We will continue to educate others by sponsoring Patient Conferences and hosting live Webinars on a variety of topics from medical treatments to psycho-social issues to current research.

Dennis Routledge
President

PRESIDENT'S MESSAGE

On February 22nd, I traveled from New York City to Cary, North Carolina to participate in an HHT fundraising gala hosted by Despina Curtis, a fellow board member, in memory of her mother who died as a result of HHT complications. In attendance were over 300 family, friends and businesses that came out to support Despina and her family in making this gala a success. The venue was beautiful and the guests thoroughly enjoyed themselves.

During the evening, Despina shared the poignant story of her mother's medical odyssey from doctor to doctor and the results of those many visits. Despina shared her frustration of not knowing what the medical diagnosis was or how to help her mother. That night, it struck me that I have heard similar stories from many of you over the years; misdiagnosis, no understanding of HHT by the medical community, who in the family inherited HHT and endless worry about your own medical situation. By Despina sharing her family's story to 300 (primarily non-HHT) guests she made a bold statement to this North Carolina community that HHT is a disease that needs recognition, diagnosis, more research and a cure!

Advocacy and education about HHT begins with us. Any opportunity to run an HHT event in your community, regardless of its size, will produce big results in education and support in your community. If you are not able to run an event in your community, please consider being a volunteer at an event or financially supporting someone who is hosting an HHT event or walk.

And, don't forget.... Our next Patient and Family Conference will be in Santa Clara, California on July 18-20, 2014. This conference is an amazing opportunity for you to educate yourself, your family and your local physicians about HHT while engaging with others who share your story. You are not alone! Come join us and talk one-on-one with over 25 HHT medical professionals and scientists to hear all that is changing (for the better) in the HHT community.

I hope to see you there.

Dennis Routledge

HHT Foundation MISSION

The HHT Board of Directors and Foundation staff live by this mission on a daily basis. Every project must meet at least one of our mission's objectives. This mission acts as a beacon of light that we are always striving to reach.

The mission of the HHT Foundation International is to find a cure for HHT while saving the lives and improving the well-being of individuals and families affected by HHT.

To achieve this mission, the HHT Foundation will:

- **FUND RESEARCH** to find better treatments and a cure.
- **EDUCATE FAMILIES AND PHYSICIANS** about HHT so that awareness of crucial diagnosis and available treatments prevents needless disability and death.
- **PROVIDE LINKAGES BETWEEN PEOPLE AFFECTED BY HHT.**
- **COLLABORATE WITH MULTIDISCIPLINARY HHT TREATMENT CENTERS WORLDWIDE** while advocating for patient access to these Centers.
- **ADVOCATE FOR AND SUPPORT THOSE WITH HHT** while increasing public, private, and governmental awareness of the disorder.
- **ENGAGE THE SCIENTIFIC AND MEDICAL COMMUNITY** so that talented individuals dedicate efforts toward advances in HHT screening, diagnosis, treatment, and research.

DIRECT CONNECTION

Published by:
HHT Foundation International, Inc.
P.O. Box 329, Monkton, MD 21111 U.S.A.
TEL: **410-357-9932**
FAX: **410-357-0655**
WEBSITE: www.hht.org
E-MAIL: hhtinfo@hht.org
Graphic Designer: Rachael Bajus

The HHT Foundation International, Inc. was formed to aid and support families with the genetic disorder Hereditary Hemorrhagic Telangiectasia (Osler-Weber-Rendu Syndrome).

Please feel free to copy this newsletter and share with family, friends and physicians.

FROM THE DESK OF THE **EXECUTIVE DIRECTOR**

Marianne S. Clancy
Executive Director

Your HHT Foundation has had many accomplishments since its founding in 1991. We are embarking on a bold new course and have many exciting changes planned this year! We are calling ***2014 The Year of Action!*** We are not content to rest on our laurels. There is much work to be done and we invite you to join us as we transform HHT as a disease and channel energy and resources into new discoveries. The HHT Foundation plans to make dramatic changes in our understanding of how HHT develops in order to advance new treatments for each and every organ affected so we can invest in game changing therapies that will impact all of our HHT families.

In order to make game changing progress, we will be launching an **Outcomes Registry for HHT** that will gather anonymous information on the way HHT affects an individual, from genetics to the effects of current treatments, in a platform that allows us to build upon this information so that we can launch important breakthroughs. We live in the information age and it's time that we make smart investments to create bold progress!

In addition, we will be launching a **Physician Forum** where medical experts can engage in HHT disease specific conversations. This will allow doctors to continue to grow their knowledge of HHT, to help educate other physicians and to work through more complicated HHT cases in an effort to improve care and quality of life for patients around the world. We will also continue to provide **webinars** on HHT "hot topics" and research breakthroughs. Our **website**, www.hht.org, will see major changes over the next year to provide all of these aforementioned resources along with fundraising and awareness toolkits.

And, let's not forget that in July 2014 the HHT Foundation will host the 17th National Patient and Family Conference in the heart of Silicon Valley. This year we will expand our Youth and Teen program and completely revise the adult program. We are excited to offer two lecture tracks on the opening day (a) for young adults and families and (b) for those of us who are a bit more "seasoned"; with subject and topic areas that are important to each group. In addition, we will have three panel discussions on (1) nosebleed management (2) current HHT research and (3) insurance and disability. We will have over 25 HHT experts on the program as well as staff from our newer HHT Centers of Excellence. There are fun filled activities planned for Saturday evening so we hope that many of you can join us, as a family, for the weekend!

I read recently a statement that reflects our current thinking: **"It Should Not Take a Generation for the Next Generation of Treatments and Advancements."** *I know that I am not content to wait another generation, are you?*

Join us as we transform 2014 into *The Year of Action!*

Marianne S. Clancy

FOLLOW US

twitter @hhtawareness

Find us on Facebook

Please note: Be careful dispensing or receiving medical information on Facebook or Twitter. The information contained on these sites should not replace necessary consultations with qualified health care professionals.

HHT FOUNDATION INTERNATIONAL

Marianne Clancy, Executive Director
Monkton, MD

James Gossage, MD, Medical Director
Augusta, GA

Marie Faughnan, MD, Scientific Research Director
Toronto, Canada

Robert Berkman, Member Emeritus
Bluffton, SC

BOARD OF DIRECTORS

Dennis Routledge
President
Garden City, NY

Terry Thompson, Jr.
Vice President
Summit, NJ

Chandra McMahon
Treasurer
Downington, PA

Beth Plahn
Secretary
Garretson, SD

Rachel Bishop
Fallbrook, CA

Angela Carlisle-Brown
Concord, CA

Marjan Chittaee
New York, NY

Despina Viniotis Curtis
Raleigh, NC

Roy Forey
Washington, DC

Ed Jurevic
Chesterfield, MO

Michael Lewis
Venice, CA

Jody Nissan
Canton, MI

Mike Nolan
Cork, Ireland

Scott Olitsky, MD
Leawood, KS

David Rinn
Seattle, WA

John Spencer, Jr.
Spring Hill, FL

INTERESTED IN SERVING ON OUR BOARD OF DIRECTORS?

Call the HHT office at
410-357-9932
or e-mail us at **hhtinfo@hht.org.**

IN **THIS ISSUE...**

OUTCOMES OF PREGNANCY IN WOMEN WITH HEREDITARY HEMORRHAGIC TELANGIECTASIA

by Els M. de Gussem, MD
Director Winnipeg HHT Clinic

March 2014, an article has been published in Obstetrics and Gynecology describing pregnancy outcomes in women with Hereditary Hemorrhagic Telangiectasia[1].

Women having been screened at the Toronto HHT Centre between January 1997 and June 2007 and who were aged 18-55 years in June 2007 were eligible for this study. These women were contacted by phone and asked for consent to participate in this study. Eighty-seven of the contacted women had had previous pregnancies (a total of 244 pregnancies) and a definite clinical or genetic diagnosis of HHT and were included.

Miscarriages occurred in 20% of pregnancies, which is similar to the general population. Most miscarriages occurred in the first trimester (78%).

In most pregnancies 172/185 (93%) women did not develop serious complications. Women had an increased frequency of nosebleeds during 31% of pregnancies and some women noticed more telangiectasia during pregnancy (14%).

Complications from a **pulmonary arteriovenous malformations** occurred in eleven women: one woman coughed up small amounts of blood and five women had a hemothorax (bleeding in the chest due to rupture of a pulmonary AVM). Another complication from an untreated pulmonary AVM can be a transient stroke (two women) or a myocardial infarction (two women). None of these women knew they had HHT before their pregnancy and none were screened for the presence of pulmonary AVMs before pregnancy.

Eleven women were screened for pulmonary AVMs before pregnancy. Three of these women had had treatment for pulmonary AVMs in the past. None of these women had a pulmonary AVM needing treatment during pregnancy. They did not develop complications from pulmonary AVMs during pregnancy and screening after pregnancy revealed that none of them needed treatment for pulmonary AVM.

Cerebral vascular malformations can occur in patients with HHT, leading to a stroke. Only one woman had a stroke due to a bleeding from a cerebral vascular malformation, after delivery. She had had a stroke several years before pregnancy as well and it is known that the risk of developing a bleed from a cerebral vascular malformation is higher once it has bled before. Seventeen women were screened for the presence of a brain vascular malformation before pregnancy and none had a brain vascular malformation detected during screening. None of these women developed a bleed in their brains during pregnancy.

Liver vascular malformations can lead to symptoms, usually later in life, but during pregnancy it can rarely lead to complications, like heart failure. In our study one woman developed heart failure due to liver vascular malformations. She was admitted to the ICU and treated with medication during her pregnancy. Unfortunately there are no preventative precautions possible for liver vascular malformations, but thankfully these only cause problems in pregnancy in extremely rare cases.

Most women delivered vaginally (70% of pregnancies) and only 30% of pregnancies required a cesarean section, similar to the general population. Fifty percent of women had epidural or spinal anesthesia (for 92 pregnancies) during labour without having been screened for spinal vascular malformations prior to delivery and none developed complications from the epidural. Sometimes anesthesists are concerned about the presence of a spinal vascular malformation and refuse an epidural. In literature to date there is no report of women with HHT developing complications from an epidural. There are case reports of women who do not have HHT developing complications after an epidural due to a vascular malformation. Spinal vascular malformations occur rarely in HHT and have mainly been reported in children.

Child outcomes were not affected by the diagnosis of HHT in the mother: children were averagely delivered after 39 weeks of pregnancy and averagely they weighed 3194 kilograms. Nonetheless, if a mother presented with a hemothorax, it affected the outcomes of the child: children had to be delivered prematurely by cesarean section and had a low birth weight at delivery.

In conclusion: Complications during pregnancy and delivery occurred in women who had not been screened or treated for AVMs prior to pregnancy. Screening following the International HHT Guidelines[2] is recommended to prevent complications.

Based on this study and our experience, we recommend screening for lung and brain AVMs prior to pregnancy, when at all possible.

1. Screening for pulmonary AVMs should be done with contrast echocardiography, before pregnancy in women with HHT or suspected HHT and, if present, significant pulmonary AVMs should be treated before pregnancy by embolization. If not done prior to pregnancy, screening for pulmonary AVM should be

performed in early second trimester, and if screening is positive low-dose CT without contrast can be done performed in the second trimester to confirm the diagnosis. If a significant pulmonary AVM is discovered, embolization should be considered during pregnancy, at an expert centre.

2. Screening for brain vascular malformations is recommended before pregnancy and if a vascular malformation is found, management should be discussed with an expert neurovascular team. If screening has not been done before pregnancy, an MRI of the brain can be considered during the second or third trimester or after delivery. Treatment is usually delayed until after delivery, in consultation with a neurovascular team. The presence of a vascular malformation in the brain should not prohibit a vaginal delivery, although a prolonged second stage should be avoided.
3. Risk of puncturing of a spinal AVM during an epidural is very low, without any evidence in our series. This is not surprising since the prevalence of spinal AVMs is very low in HHT. Consultation with an anesthesist prior to labour is recommended, so decisions can be made on a case-by-case basis, but HHT should not be considered an absolute contraindication to epidural anesthetic. If screening for a spinal AVM is considered, it should preferably be done before pregnancy with a MRI. If screening is considered during pregnancy, a MRI without contrast is recommended.

[1] *Outcomes of Pregnancy in Women With Hereditary Hemorrhagic Telangiectasia. EM de Gussem et al. Obstet Gynecol. 2014 Mar; 123(3): 514-20.*

[2] *International Guidelines for the Diagnosis and Management of Hereditary Hemorrhagic Telangiectasia. ME Faughnan et al. J Med Genet, 2011 Feb;48(2):73-87.*

The HHT Foundation wishes to acknowledge Dr. Els de Gussem for her collaboration with GRMAB members Marie Faughnan, M.D, MSc, Karel G. TerBrugge, M.D., FRCPC, and Johannes J. Mager, M.D., Ph.D. on this important research. Dr. de Gussem received the Robert I. White Young Clinician Award from the HHT Foundation in 2009.

BRAIN ARTERIOVENOUS MALFORMATIONS (BAVM)

HHT RESEARCH SUMMARY FROM 2013 SCIENTIFIC CONFERENCE

by Dr. James Gossage, M.D.
Medical Director, HHT Foundation

Cork, Ireland was host to the 10th HHT International Scientific Conference where over 200 medical professionals and researchers gathered to present their HHT specific research findings. Several members of the HHT Foundation's Global Research and Medical Advisory Board (GRMAB) were tasked with the responsibility of providing a summary of organ-specific research. Here is the summary of Brain AVM research presented in Cork. All other summaries will be presented by a Research Panel at the National Patient and Family conference in California this summer.

Brain AVM are seen in 5-10% of HHT patients and can cause serious complications such as bleeding (intracranial hemorrhage), seizures, and death. These serious complications can even occur before one year of age. Although brain AVM are treatable by various techniques (such as surgery or embolization), it is unclear which patients are at greatest risk for complications and which treatment modality is best. Additionally, 5-10% of patients have other types of vascular malformations that are not true AVM and therefore probably at less risk for complications.

Dr. Takeo Nishida and colleagues compared MRI and angiography to evaluate blood vessel architecture in small vascular malformations of the brain. They found that certain findings on brain MRI scan were only seen in true AVM, but these features were only seen in 29-44% of AVM, indicating that angiography may be needed in most patients to properly characterize the lesions as true AVM versus benign malformations. Dr. Karel TerBrugge, Toronto Western Hospital, and colleagues found that patients who had prior curative treatment of brain and spinal AVM were more likely to develop additional brain and spine AVM in the future. Dr. Jean Francis Bergerot, des Hôpitaux de Lyon, and colleagues did a careful analysis of brain MRI in HHT patients to look for a link between HHT and improper brain cortex development. In addition to the expected increase in AVM, they found aneurysms in 6%, polymicrogyria in 7%, and anatomic variants of the cerebral arteries in 13%.

These studies widen the spectrum of neurological complications in patients with HHT and suggest the need for long term monitoring of treated AVM patients. They also indicate that although MRI brain scans are very high tech, angiography is necessary in most patients to exclude the more serious AVM variety in patients with small vascular malformations, since they are more likely to bleed. However, we still have little data to help us determine which patients are at greatest risk for complications and which treatment modality is best. These questions might be partially answered with long term follow up of many HHT patients – such as will be possible with the North American Outcomes Registry for HHT. Ideally, these questions will be addressed in a multi-center research study comparing treatment versus no treatment, to insure that the treatments are both safe and effective.

Do YOU or YOUR CHILD have HHT and a Brain AVM?

If so, **WE NEED YOU!** As you know, one of the major health risks to patients who develop a brain AVM is the possibility that the blood vessels that make up the AVM will break or rupture and cause a stroke. "Cerebral (brain) Hemorrhage Risk in Hereditary Hemorrhagic Telangiectasia (HHT)" is the first large-scale study of brain arteriovenous malformations (AVMs) in HHT patients. This National Institutes of Health (NIH) funded research study aims to find out what genetic and clinical factors signal high risk for bleeding from brain AVMs. These risks have never been fully assessed for patients with HHT. The results of this study will help doctors make decisions about brain AVM treatment for individual patients and will drive further research in brain AVM therapies.

Brain Vascular Malformation Consortium (BVMC) investigators including the HHT Foundation Scientific Research Director, recently presented preliminary results at the International Stroke Conference in San Diego. They demonstrated for the first time in HHT patients, that previous hemorrhage from a brain AVM predicts increased risk of hemorrhage going forward. This is the first risk factor identified in HHT patients, predicting hemorrhage from brain AVM, and is an important step toward advancing our knowledge. The BVMC team continues to investigate more risk factors for hemorrhage from HHT brain AVMs which will to help guide decision making for doctors and patients.

The principal investigators of the Brain Vascular Malformation Consortium have committed to recruiting 875 HHT patients with BAVM's –To date we have 202 patients recruited so your help is urgently needed! We are nearing the end of the enrollment phase so we are anxious to enroll as many BAVMs as possible before the end of June 2014.

Participation is easy. . . All that is required is a telephone conversation, documentation (letter of diagnosis/genetic testing results, imaging, etc), consent to have information used by the principal investigator, and a saliva sample. That's it. You don't have to travel anywhere. There's no cost to you.

The only eligibility requirements are that the patient must:

- Have a documented clinical or genetic diagnosis of HHT
- Live in the United States, Canada or The Netherlands
- Be diagnosed with a Brain AVM, whether or not it's been treated
- Be 3 years of age of older

There are 13 North American sites (9 US; 4 Canada) and 1 international site participating in this HHT research. Ten of the thirteen research sites are now recruiting children (ages 3 and older). Contact the research coordinator directly if you or your child has been screened and/or treated at one of the participating HHT Centers. Otherwise, call the HHT Foundation at 800-448-6389 or email cathleen.kinnear@hht.org to begin making a difference in HHT research.

To learn more about this study, visit the Rare Diseases Clinical Research Network website at https://rarediseasesnetwork.epi.usf.edu/BVMC/studies/6203-HHT.htm.

United States

California

- **University of California, Los Angeles**
  Polly Kay, RN, CCCRP
  310-794-0376
  E-mail: pkay@mednet.ucla.edu
- University of California, San Francisco
  Liz Clain
  415-502-2151
  E-mail: Claine@anesthesia.ucsf.edu

Connecticut

- Yale University School of Medicine, New Haven
  Bob White, MD
  E-mail: bob.white@yale.edu

Georgia

- Georgia Health Sciences University, Augusta
  Melissa James, RN
  706-721-0470
  E-mail: mjames@georgiahealth.edu

Maryland

- Johns Hopkins University, Baltimore
  Doris Lin, M.D., Ph.D.
  443-287-3079
  E-mail: ddmlin@jhmi.edu

Minnesota

- Mayo Clinic, Rochester
  Cathy Bornhorst
  507-284-9259
  E-mail: bornhorst.cathy@mayo.edu

Missouri

- Washington University, St. Louis
  Stacy Schrum
  314-362-7507
  E-mail: sschrum@dom.wustl.edu

North Carolina

- University of North Carolina, Chapel Hill
  Karen L. Smith, RN
  919-966-2790
  E-mail: karens@med.unc.edu

Utah

- University of Utah, Salt Lake City
  Jamie McDonald, MS
  For Scheduling: 866-292-4HHT
  Direct Phone: 801-585-9702
  E-mail: jamie.mcdonald@hsc.utah.edu

International Centers

Alberta

- University of Alberta, Edmonton
  Debbie Boyko
  780-492-9714
  E-mail: debbie.boyko@ualberta.ca

British Columbia

- St. Paul's Hospital, Vancouver
  Leslie Love, BSc CCRP
  604 806-8117
  E-mail: llove@providencehealth.bc.ca

Ontario

- Hospital for Sick Children, Toronto
  Stephanie Jeanneret Manning
  416-813-4903
  E-mail: stephanie.jeanneretmanning@sickkids.ca
- St. Michael's Hospital/University of Toronto, Toronto
  Myra Slutsky, Study Coordinator
  416-864-6060 extension 2887
  E-mail: slutskym@smh.ca

Netherlands

- St. Antonius Hospital, Nieuwegein
  M. Nauta, RN
  + 31 (0) 30 609 3459
  E-mail: m.nauta@antoniusziekenhuis

Offering Dedicated Care for Children with HHT

The Pediatric HHT Center at Washington University and St Louis Children's Hospital

by Andrew White, M.D.
Co-Director, Washington University HHT Center

The HHT Center at Washington University is an internationally recognized program providing care to both children and adults with HHT. Founded over 20 years ago, the HHT Center has been accredited by the HHT Foundation International as a Center of Excellence. While it is one of only 16 centers in North America treating the disorder, it is perhaps the only center in the United States with a dedicated pediatric team with substantial experience managing and treating HHT in children. This pediatric team includes board certified specialists from pediatrics, pediatric pulmonology, pediatric otolaryngology, pediatric radiology (including pediatric interventional radiology), pediatric neurosurgery, pediatric genetics, pediatric interventional cardiology, pediatric gastroenterology, pediatric hematology, pediatric dermatology and dedicated pediatric HHT nurses. Our Pediatric Center has been treating children with HHT since 1999 (prior to that year "Adult" physicians saw these children, not Pediatricians!), and since that time, we have evaluated over 300 children and their families. Our entire team understands the special needs of children and will do everything we can to ensure that a child feels safe and relaxed during the office consultation, diagnostic procedures and testing

Dr. Andrew White (Pediatrics) and Dr Murali Chakinala (Adult Medicine) have been co-directors of our HHT Center since 2006. This sharing of directorship responsibilities is one reason our center is better able to coordinate our efforts. It is our goal that every family with HHT can have all their HHT related health care needs addressed in a single, well-coordinated, visit.

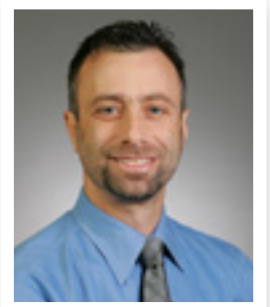
A visit to the Washington University and St. Louis Children's HHT Center begins with a telephone conference with a team member. We gather detailed family and patient health history and obtain all necessary medical records. Once this information is carefully reviewed by our multidisciplinary team, we will tailor the specific diagnostic tests and imaging necessary for your child. On the day of the visit, your child will undergo a thorough physical examination and imaging that may include an MRI of the brain, CT angiogram of the chest, or bubble echocardiogram. This testing and consultation can often be completed within one to two days. At the conclusion of the visit, results will be communicated to the family and a treatment plan provided.

The HHT Foundation is grateful for the excellent care and compassion shown to all children and adults at the HHT Center of Excellence at Washington University.

For additional information or to schedule an appointment: Physicians, patients and families may call 314-454-2479. St. Louis Children's Hospital at Washington University One Children's Place, Saint Louis, MO 63110

Mid-West ENT Surgeons Learn about HHT

by Scott Olitsky, M.D.
Member, HHT Foundation Board of Directors



On January 10, 2014 I was invited to speak at the Kansas City Otolaryngology and Ophthalmology Society's Annual Meeting. This regional meeting attracts ENT physicians from several Mid-West states to learn about clinical updates, new surgical procedure and other ENT related issues.

My presentation entitled, "Hereditary Hemorrhagic Telangiectasia- New Treatments for HHT Epistaxis and the Role of ENT Specialist beyond Nosebleeds" was well received. The 45 minute lecture discussed new treatments of nosebleeds and the systemic manifestations of HHT with special emphasis on the vital role ENT doctors play in helping to diagnose HHT patients and get them properly screened. Following my presentation, ENT surgeons had the opportunity to ask me questions.

Special thanks to Amy Sokol and Andrea Olitsky for staffing an HHT Information Booth during the meeting and helping to raise awareness amongst the Mid-West ENT's.

The HHT Foundation is grateful for Dr. Olitsky's dedication to educating other medical professionals about HHT so that patients can receive proper diagnosis and more effective treatments. Scott's volunteer service as an HHT Foundation board member and educator is very much appreciated.

cure hht team

Upcoming 5K Walks

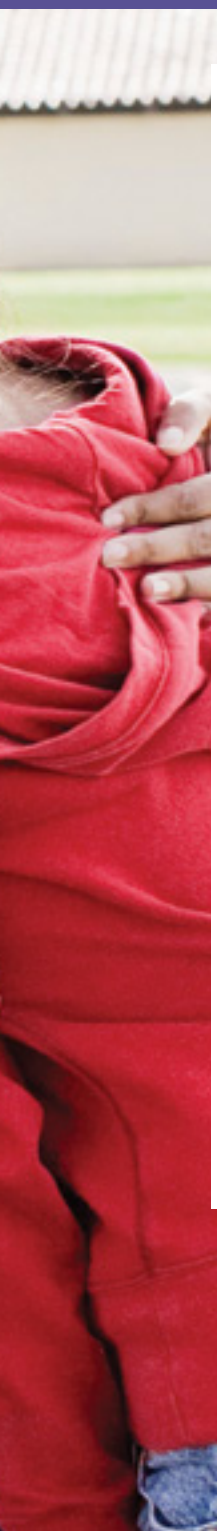
May

May 4 Cleveland, OH
May 18 Chapel Hill, NC
May 31 St. Paul, MN

June

June 7 Michigan City, IN
June 22 Washington, DC

Together, Cure HHT Team members are taking actions, raising awareness and funds to help cure HHT.



Join Cure HHT Team!

Become part of Cure HHT Team by joining and supporting events in your community or starting your own! You, your family and friends can support the HHT Foundation as part of the **Cure HHT Team** program by holding your own event or participating in an existing event.

The HHT Foundation provides **Cure HHT Team** members with a great fundraising platform for everything from lemonade sales, to hosting a 5k walk/run, to using an event entry you already have to raise fund for HHT.

As a Cure HHT Team member, your benefits include: Cure HHT Team branded marketing materials and a volunteer handbook that is filled with helpful tips to help you reach out to friends and family, raise funds and awareness to achieve your goals.

So whether you are an athlete or a community fundraiser, you are part of a nationwide team of individuals who are making their actions count and supporting the HHT Foundation.

It's easy to join in the fun!

Email events@hht.org to get started or visit www.HHT.org/Cure-HHT-Team for information.

GIFT PLANNING: THE LEGACY SOCIETY

The Legacy Society is an honorary association of men and women who make gifts to the HHT Foundation International in their **wills or living trusts**. Or they may have made the HHT Foundation a beneficiary of a life insurance policy, retirement account, gift annuity, or charitable trust. Legacy gifts come in many forms and amounts. Most of them are from donors who have ordinary means and resources.

Besides their importance to the Foundation, legacy gifts often reduce or eliminate income and estate taxes for donors. And some legacy gifts produce life-long income for donors – gift annuities and charitable trusts are examples.

Our Legacy Society members are truly extraordinary. In gratitude for their generosity, members in the Legacy Society will receive invitations to special events and seminars and a subscription to our newsletter. The most important benefit you will receive from joining The Legacy Society, however, is the satisfaction derived from making a lasting contribution to the HHT Foundation International's long-term strength.

If you would like to make a legacy gift, or if you have questions or ideas about your estate planning, please contact Marianne Clancy, Executive Director, at or 410-357-9932.

For more information, and to download our Legacy Society Confirmation Form, go to http://hht.org/donate/planned-giving.

Our Story

by Sarah Beckwith
HHT Foundation Member

MIGRAINES – A WARNING SIGN!

22 year-old Sarah Beckwith has a complex case of HHT and would like to spread the word about this disorder by sharing her story so that others to understand that migraines may be a warning sign of AVM's in HHT.

It was the day of my second grade line-dancing debut; I had been excited for this day for weeks. We were all lined up and ready to go, when I noticed a strange light show out of the corner of my eye. It got bigger and bigger until it blocked out most of my vision. When the light show ended I was hit with an enormous headache, it was so bad I started to get nauseous. My mom was called to the school and came to pick me up. As soon as I reached her car, the prop hat that I was supposed to wear for the dance became a bucket to be sick in. This was my first migraine.

> *"I am very lucky to have such an amazing and supportive family to help me through my journey and I don't know where I would be without them.*

I continued to get migraines for years, which progressively became worse and worse. As I got older I started to have issues with breathing, which some doctors "diagnosed" as asthma because they had no other explanation. Along with the migraines and breathing problems I would wake up in the morning with an occasional nosebleed.

When high school started my migraines had become such a problem that I would have to go the hospital to get them treated. They would come almost twice a week. So crippling that laying on a pillow felt like my head was expanding. They always started with the light show, followed by the immense head pain, followed by nausea, and at its worst, my migraines ended with the numbing of the right side of my body and tongue.

Needless to say my family and I were starting to get very concerned. We went from doctor to doctor without any real diagnosis or cause to my problems. We had known about HHT on my mothers' side of the family but didn't think much of it because we had thought that the most common sign of HHT was constant nosebleeds. In many cases this is true. But my nosebleeds were rare; my biggest problem was the headaches.

In my sophomore year of high school my mom decided it was time to get my sister and I checked for the disorder. We found the closest HHT center to us, which was The University of Utah Hospital in Salt Lake City. We went to the Utah HHT Center where they ran several different tests - blood tests, Echo bubble, CT scans, and MRI's.

Waiting for the results felt like it took a lifetime even though the results came extremely fast. We got a call from the geneticist. I remember watching my mom's face as she listened to the person on the other line. It was panic, followed by fear, followed by the "we need to take action" face.

She told me that I did indeed have HHT, but the good news was that my sister did not. From the tests, they had found an AVM in my brain and several (too many to count) in my lungs mostly in the left lung. I don't remember what I was feeling, just that I was in shock.

We scheduled the brain surgery and lung embolization a couple of weeks later. Both surgeries went extremely well and I can't thank the doctors and nurses at the University of Utah Hospital enough. They saved my life and kept me sane!

I have since been back to the hospital to continue to get more of the AVM's in my lungs treated. I am scheduled to go back again at the end of February 2014 for another procedure.

I now live in Seattle, WA but I will always go back to the University of Utah Hospital because of the bonds and relationships I have made there. I am very lucky to have such an amazing and supportive family to help me through my journey and I don't know where I would be without them.

THE AGAPI & HOPE GALA FOR HHT

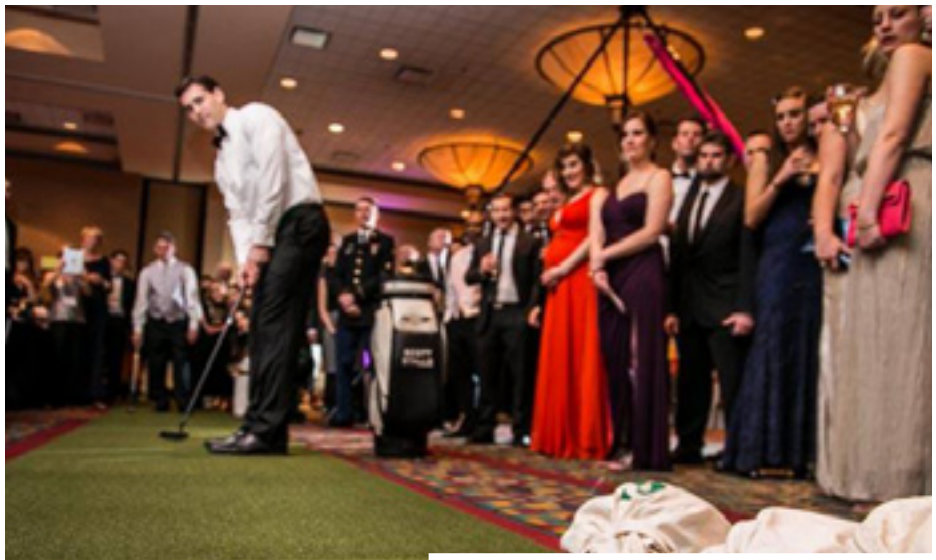
by Cathleen Kinnear
Manager, Education & Awareness Programs

On February 22nd, more than 300 people gathered in the Grand Carolina Ballroom of the Embassy Suites in Cary, North Carolina to enjoy a glamorous black-tie gala like no other. The event, which raised over $77,000, began with an eloquent cocktail hour where guests graced the red carpet and posed for pictures. Inside the ballroom, gala attendees looked on in amazement as an aerial artist performed some high flying acrobatics. The fun continued as a few lucky raffle winners putted for a chance to win $10,000. Guests also perused over 80 silent auction items that included everything from sports memorabilia, vacation getaways to signed celebrity memorabilia just to name a few.

The evening continued with a successful live auction that included one-of-a-kind auction items, such as as VIP LA Access & Tickets to the ESPY Awards in Los Angeles. Following the auction, guests danced the night away to the sounds of DJ Joe Bunn.

The evening had special meaning for Despina Viniotis Curtis, HHT Foundation Board Member and event chair who organized this gala in memory of her mother, Maria Viniotis, who died of HHT-related high output cardiac failure at the age of 53. Despina took the podium, surrounded by her sisters, and shared her mother's final days while choking back tears. Guests looked on as two moving videos; one showcasing the Vinitois family, the other honoring the lives of those who have battled with HHT, played on the big screens. Despina's moving tribute left guests with a better understanding of this genetic blood vessel disorder that affects 1 in 5000. She shared the need for continued research to help find a cure so no other family will endure the pain her family has experienced.

The HHT Foundation would like to thank Despina Curtis and her husband Simon, as well as her father, Yannis Viniotis and her sisters, Natalie and Nora for helping to plan this successful and moving event to benefit the HHT Foundation.



JOIN A **CURE HHT TEAM** WALK

Join in the Year of Action with HHT and get active with a Cure HHT Team event in your area! This year, volunteers in 5 states are planning 5K run/walks to raise awareness and funds for HHT.

Sunday, May 4th 9:30AM *Cleveland, OH*

Walk Event Chairs: Denise Sherman – denise.sherman4hht@gmail.com
Marijo McCune – marijo.mccune4hht@gmail.com

To Register/Donate: **curehhtteamwalkcleveland.causevox.com/**

Sunday, May 18TH 1PM *Chapel Hill, NC*

Walk Event Chair: Dr. Charles Burke

Contact Laurie Birdsong: 919-966-5469 or laurie_birdsong@med.unc.edu

To Register/Donate: **curehhtteam5krunwalkchapelhill.causevox.com**

Sunday, May 31ST 8AM-10:30AM *St. Paul, MN*

Walk Event Chairs: Colleen Libhardt – Libhardt66@gmail.com
Katy Green – Ktgreen813@gmail.com

To Register/Donate: **curehhtteamwalkstpaul.causevox.com/**

Saturday, June 7TH 10AM -12PM *Michigan City, IN*

Walk Event Chair: Stephanie Lee – stephanielee67@hotmail.com

To Register/Donate: **curehhtteamwalkmichigancity.causevox.com/**

Sunday, June 22ND 10AM *Washington, DC*

Plan a trip to Washington D.C., enjoy touring our Nations' Capitol, visit your Legislators and educate them on HHT, and join us on Sunday for Cure HHT Team Walk Event in DC.

Walk Event Chair: Roy Forey – royforey@comcast.net

To Register/Donate: **curehhtteamwalkdc.causevox.com/**

For Cure HHT Team 5k run/walk locations and details, visit the causevox.com registration/donation pages listed above. ***We need your support to move the needle together to cure HHT.***

Stroke Survivor Plans Cross-Country Journey to Raise Awareness for HHT

In 2005, at age 41, Dan Zimmerman suffered a stroke that paralyzed his right arm and leg and damaged the left side of his brain. A doctor told him that he would never walk or talk again. Angry, Dan knew he had to give up or fight.

Determined, it took eight weeks before he could hobble with a walker after the stroke. For six months he visited job sites in a wheelchair and pointed to communicate as his business wrapped up projects.

A year passed before he could barely talk, and three years before he could spell simple words. Although his gait is unsteady at times and his speech hesitant as he searches for a word, he realizes that recovery is a life-long process.

By 2008, Dan wanted more mobility than his wheelchair afforded. He began riding a recumbent trike with the "Bent Riders of Arizona" on weekly rides and weekend tours. He logs 500 to 700 miles a month in preparation for his cross-country journey.

Cycling, Dan says, has improved his health and more importantly, given him a purpose in life. He wants to raise awareness of HHT, the disease that killed his brother at age 19, and mother and is present in Dan and his younger son. He also wants to raise awareness of stroke prevention and inspire other stroke survivors to fight back against the disease.

Many individuals ride bicycles across the United States, but Zimmerman said few make the trip on a recumbent trike. He plans to raise awareness about recovery from strokes and offer hope of stroke survivors through television, radio and newspaper interviews.

The biggest challenge of Dan's life - **Spokes Fighting Strokes (SFS)** - will begin on July 1, 2014. A ride of more than 5,200 miles across the United States from the San Juan Islands in Washington state to Key West in Florida. Dan has set a goal to raise $100,000 for HHT. The HHT Foundation has established a fundraising page at curehhtteamspokesfightingstrokes.causevox.com/ if you would like to help Dan achieve his goal. For more information about the ride, and to see when and where Dan will be going, go to the bike tour website at www.spokesfightingstrokes.org.

DONATIONS TO THE *Foundation*

Our sincere and heartfelt thanks go out to everyone who as contributed to the HHT Foundation whether through research fund, membership, general, or tribute donations. Below we list the donors who have contributed to the HHT Foundation through a memorial or tribute gift from November 1, 2013 to February 28, 2014.

EDUCATION & RESEARCH FOUNDERS DONATING $100,000 + (over a lifetime)

Charles and Oranee Abbott
In loving memory of Patricia Abbott Rasmussen, Stephen Abbott and Rachel Abbott

Robert and Leora Berkman

Jennifer Blevins and Christopher Licata

Terence and Cynthia Brown

Armin and Esther Hirsch Foundation

The Estate of David and Clara Bartley Jordan

Anonymous Donor Family

Anonymous Donor Family

Marc and Diana Meiches

The Estate of Eileen Morrell

Clayton and Jody Nissan

Mike and June Nolan and the Grace Nolan Foundation
In loving memory of Grace Nolan

Dennis and Maureen Routledge

Stephen and Jane Silk

Mary Vann

TRIBUTE DONATIONS

DONATIONS THAT CELEBRATE *In Honor Of...*

All with HHT
Robert Garcia

The Olitsky Family
Mary Eisman

Elizabeth Acerra
Betty Acerra
Camille Toto
Martin and Tracey Toto
Peter and Christina Toto

Marla Akins
Richard Olitsky

Molly Alter
Yvonne Hettrich

Dr. Mary Barber
Frank and Judy Sublette

Bob and Lee Berkman
Leslie Harrison

Rachel Bishop
Tom McNeal

John and Cathy Bukauskas and sons
Charles Bukauskas

James Caillouet
Ashleigh Garner

Judy Daly
Sal and Priscilla Russo

Carey James Davis
Jay Davis

Jack Devizia
Linda DeVizia

Eakins Family
Janet Heatherly

Steve and Sue Eiredam
Charles Bukauskas

Brad, Holden, Jonathan and Ryan Estes
Trina and Gene Estes

Rupert Family
Karen Berg

The Cantrell Family
Karen Campanaro

The Goldberg Family
Amanda Madnick

The Olitsky Family
Paul and Arlene Frimark

Hans Forsch
Kristen Laine

Ryan Francis
Cathy Francis

Edith Garvey
Helen Leven

Lori Geittmann
Helen Leven

Sandy Haywood
Terry Haywood

HHT Foundation Staff
Art and Sandra Lurie
Frank and Trish Linke

Paul Hirsch
Joni Blackman

Nicholas Horowitz
Charles and Barbara Horowitz

Scarlet Howard
Anthony Bender

Sheila Hutman
Ted Hutman

Barbara Isenberg
Kimberly Isenberg

Bill Jukes
Georgia and Craig Hendricks

Jamie McDonald Kamm
Connor Kamm

Nish Karakashian
Dave and Heather Karakashian

Jean Klinghoffer
Diane Masseth-Jones

Lynn and Bill Kosson
Charles Bukauskas

Robert Lewis
Charles Hicks

Frank and Trish Linke
Marie White

Joe and Bonny Mack
Mr. Charles Bukauskas

James and Ellen McDonald
Jamie McDonald

Tina Mejia
Toni Hoyt

Clay Nissan and Family
Scott and Sarah Gilbert

Anna Pillow and Family
Keene's Pointe Community

Arlene Salkin
Rhea Geer Discount
Marcia & Sid Weinberg

Elizabeth Acerra and Savino Family
Gary and Nancy Toto

Gracie and Jack Schull
Anonymous

Jane Silk
Eli Bishop
Elizabeth Del Pozo
Haig and Susan Koshkarian
Nancy Polley-Aibel
Sally Shapiro
Elizabeth Suskind

Frank Smith
Nancy Stagnitta

Eva Stutz
Cristen Van Vleet

Jane Silk, Rachel Stutz, and Eva Stutz
Michael and Laura Coca

Terry Thompson
Peter and Jean Piecyk

The Thompsons
Michael Reed

Dr. Robert White
John Chirgwin
Stanley and Betty Weinberger

MEMORIAL DONATIONS

IN LOVING *Memory of...*

Marilyn Bell Allyn
Lizabeth Allyn

Julian "Monty" Adams
James Gossage, M.D.

Michael Andjelich
Nancy Margraff

Phyllis Benton
Odessa Lasalle

Lois Berdar
Michell Magerko

Everest Black
West Seattle Aux F.O.E. 2643

Mary K. Black
Brian and Barbara Vogel

Mary Blevins
Gayle Blakeslee

Sheila Bresnan-Bradley
Eileen Bresnan

Elsie Winn Brunjes
Erica Chalecki
Don and Gail Lyons
Chuck and Cristl Phillips

Emily Bukauskas
Charles Bukauskas

Mary Louise Buttimer
Mark Buttimer

Barbara Krames Cohen
Sanford Cohen
Matthew and Lisa Russman

James and John Cooley
William and Mary Novotny

Gordon and Arline Cooper
Donald Bjork

Jean Crockett
Kelly Crockett

Dorothy Deffenbaugh
Joan Kord
Judith Rohr

Barbara Feldman
Elissa Feldman

James Flora
James and Teresa Flora

Rosaline Amelia Forey
Roy Forey

Jane Gadjo
Eric Rowell
Nan Rowell
JoAnn Wishart

Gladys Gallagher
Gallagher Family

Barbara Brown Gardner
Laurie Boyce

Dreux, Earl, and Betty Grever
Kim Grever

James Noel Griffiths
Lynn Griffiths

Hyman Grossman
Barbara and Ron Isenberg

Sandra Haywood
Terry Haywood
Philip Jeffares

Sam Holsapple
Jeffrey and Danielle Holsapple

Rita Satloff Jacobs
Barbara and Ron Isenberg

Earl Johnson
Eric Johnson

Vivian (Kossow) Johnston
Walter and Linda Johnston

David and Clara Bartley Jordan
Rachel Jenkins

Betty Katanick
Isabelle Friedlieb
Don and Vicki Greene
Douglas and Rosemary Kaploe
Beverly Kikoler
Elaine Klapman
Ron and Joann Kramar
Wendy Madnick
Walter and Florence Marshall
Ellen Mayer
Samuel and Roberta Pepkowitz
Gary and Sue Thompson
Arlene Williams

Sol Katanick
Douglas and Rosemary Kaploe
Beverly Kikoler
Elaine Klapman
Ron and Joann Kramar
Wendy Madnick
Walter and Florence Marshall
Gary and Sue Thompson

Solomon and Betty Katanick
William and Marion Soady

Leonora Kloder
Mitchell and Katie Kloder

Doris Muriel Knowles
Philippa Burns
Nigel Knowles

Maria Lamus
Clemencia and Albert Hernandez

Lois Lawrence
Arlene Lewis

Elizabeth and Gregg Legler
Glen and Lisa Legler

Alfred Lewis
Madeline Knapp

Harold Lisonbee
Boyd Lisonbee

John Lownsbro
Gillian Duffy

Lois Marakovitz
Susan Marakovitz

Beverly Conrad and Anna Markow
Karen Sawyer

Evelyn and Martha
Anonymous

Kathlyn Roberta "Bobbie" Molale
Michael Brown

Patrick A. Murphy
Anonymous

Valerie Nelson
Cheryl Nelson

Sandra K. Notch
Diane and Tom Mulvany

Anthony Pagliuca
Randolph Bello
Patrice Carney
Vincent Hauser
Edward and Susan Mulvihill

Robert "Bob" Parsons
Herbert Parsons
Linda Parsons

Janetta Patterson
Fidelity Charitable

Claire Pero
Anonymous

Barbara G. Polstra
Tim and Cindy Burke

Trevelyn Ray
Brad and Kerry Dawkins

Jerry Reid
Susan Reid

Melissa Reigel
Sean Reigel

Peter Ribicoff
Stephen and Jane Silk

Harold Riekhof
Karen Carl

Lionel Roseff
Jane Chwick
Abby Cohen
Alan and Annette Florin
Anita Frankel
Goldman, Sachs & Co. Matching Gifts Program
Alvin and Phyllis Katz
Norman and Lisa Kurtz
Luftig/Paer Family
Anonymous
Jeff and Wendy Maurer
Albert and Marilyn Mittman
Jules and Helene Saunders
Stephen Sokolow
The Stecher Family
The Suzanne Nora Johnson & David G. Johnson Foundation

Noelle Ross
Denise Estes
Sandra Glatzle
Kathy Kuehling
Justine Shaver

Eric Rubin
Arlene Lewis

Arnie Ruskin
Lois Wonboy

Robert Santorelli
John and Marilyn Fidler
Anonymous

Yale Solomon
Jackie Hammes

David S. Stoner
Lambert Family

Barbara Underwood
Kay Holm

Luis Ramos Valentin
BJ and Julia Choi

Roberto Vera
Yasuo and Elyna Ito

Maria Viniotis
Rose Marie Connell
Anonymous
Jennifer Kraut
Ladies Philoptochos Society Holy Trinity Greek Orthodox Church
Arthur and Emma Paradeses

Bob Warren
Jackie Hammes

Daniel and Susan Way
Michael Way

Vivian Wherley
Jerry Wherley

Edwin George White
Linda Hierholzer

Kyle Wilfert
Jarrett and Maria Wilfert

Eloise Yeager
Kit Ryan

HHT Foundation International, Inc.
P.O. Box 329
Monkton, MD 21111
U.S.A.

ADULT & YOUTH PROGRAMS • LEARN ABOUT HHT • THE LATEST RESEARCH,TREATMENTS AND GUIDELINES • AND MUCH MORE...

Registration materials will be mailed and available online in late April.