National Walk for WD
Lisa Simopoulos

When: September 19, 2015
Where: Various cities across the United States
Who: You and Everyone you know!
Why: To bring awareness and funding for WD
What: A walk- but not a standard, run-of-the-mill 5 or 10K

Our family was thrown into the world of WD as it happens with most people- one of our kids became gravely ill which lead us down a path of many doctors, possible diagnoses and eventually the diagnosis of this unheard of disease. After subsequently diagnosing two of our other children we were grappling with what we could do to help others and advance research etc. One of the biggest challenges we saw was the fact that it is so difficult to bring large numbers of people who are dealing with this together because of the rarity of the disease. Then the idea of a 'virtual walk' was born. Simply put, we thought that we could have individuals volunteer to organize a walk on the same day in different cities across the country. The idea would bring solidarity to an otherwise isolating disease while at the same time creating awareness and raising funds for research.

There are two ways to participate: 1) sign up for a walk in a city in your region or 2) sign up as an individual and have your own walk. There is no prescribed length for the walk and we wanted to keep it simple by holding it in places where permits were not needed which add to the complexity and the cost. For example, our family is holding one at a park near our house and we will be walking 1-2 miles culminating with a picnic afterwards. We also will be drinking La Croix sparkling water, which is generously donating their water to every venue!

The walk is, as far as we know, the first of its kind. Our website is www.thewilsonsbigwow.com and you can register directly on the site. There are 17 cities participating across the nation including Canada. At the end, we will be posting pictures from every venue, coast to coast for people to see. We would love to have a big turn out in support of this great cause, plus it will be a fun and exciting way to connect us all!

2015 Annual Conference: Saturday, September 26, 2015

This year’s annual conference, Blueprint for the Future, will be held on the east coast in the New York City area. Because of the higher cost of holding an event right in NYC, we chose instead to host the conference at The Sheraton Lincoln Harbor in Weehawken, New Jersey, http://www.sheratonlincolnharbor.com/.

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NEWSLETTER DEADLINE:
If you would like to submit an article to be published in the next printed edition of The Copper Connection the deadline for submission is December 1, 2015. Please e-mail your article to the WDA office at info@wilsonsdisease.org.

The Wilson Disease Association is a nonprofit 501 (c)(3) organization. The Copper Connection is a semi-annual newsletter published by the Association that informs members of findings in the area of Wilson Disease. There is no copyright. Newsletters and other publications can disseminate any information in The Copper Connection. Please cite attribution to the Association and the author.

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Our Mission Statement
The Wilson Disease Association funds research and facilitates and promotes the identification, education, treatment and support of patients and other individuals affected by Wilson Disease.
The Sheraton sits right on the banks of the Hudson River with beautiful views of the Manhattan skyline.

Should you desire to take a side trip in your spare time, the NY Waterway Ferry dock is adjacent to the hotel. The ferry is a 10 minute ride and takes you right to Pier 11, located at 38th Street & 12th Avenue, in Manhattan.

Now for the exciting program! The all day Saturday program will feature physician speakers from Yale, Baylor University, University of Michigan, University of Washington, and University of Florida. Educational presentations will be given in the morning and small round table discussions will be held in the afternoon. The day will conclude with the Conference Banquet in the evening.

**CONFERENCE REGISTRATION:** Registration for the conference is free for members, $60.00, a reduced price from actual cost, for non-members. The Conference Banquet is $30.00 per person for members, $50.00 for non-members.

To check your membership status, please login to your account at https://wda.donortools.com/. If you do not have an account established please create one. This is also a great way to keep track of your donation history. This year registration will be done completely online at http://2015wdaconference.eventbrite.com.

If your membership is not current, and you would like to renew, you will also be able to do that on the conference registration page. If you are unable to register online, please call the WDA office at 866-961-0533. All registrations must be received no later than September 12, 2015.

If you are flying in for the conference, we recommend flying in to Newark Liberty Airport. Visit https://www.panynj.gov/airports/ewr-ground-transportation.html for ground transportation options.
WDA Supports Doctoral Thesis, Copper-associated hepatitis in the Labrador retriever

Mary Graper

The WDA was pleased to offer partial support for Dr. Fieten's interesting research, in the amount of 500 Euro, which culminated in her thesis publication. Hille Fieten, DVM, PhD is a young, ambitious copper researcher at Utrecht University, The Netherlands. She was also an invited speaker at the WDA sponsored scientific workshop, Human Disorders of Copper Metabolism: Recent Advances and Main Challenges, held at Johns Hopkins University in June of 2013. There Dr. Fieten presented her preliminary research on new canine models of copper toxicity (the Labrador Retriever).

Copper toxicity in Bedlington Terriers has been well documented for many years. Beginning in 2008, Dr. Fieten and her Dutch colleagues began researching this disorder in the Labrador purebred dog breed. While we all know the gene responsible for Wilson disease is ATP7B, at that time the gene responsible for “Wilson disease” in Labradors was unknown. Female dogs are over-represented and in most cases clinical signs start between 3-7 years of age. Unlike Wilson disease in humans, only the liver disease is noted in dogs. Labradors do not develop neurologic symptoms. Treatment is the same as in humans.

Dr. Fieten believes that the results described in her thesis, “will have major implications for understanding the biological background of the disease, and may be valuable for development of new therapies for both humans and dogs.”

In the Acknowledgement section of her thesis publication Dr. Fieten wrote,

“Dear Mary Graper and other members of the Wilson Disease Association, your work is so important! Thank you for organizing a great workshop in Baltimore, granting me the possibility to present my research and the opportunity to meet fellow researchers. The get together with the patients made me realize what a great impact Wilson and Menkes disease have on the life of people and their loved ones and are a true inspiration for my research.”

I think I can speak for all of us when I say, thank you too Dr. Fieten for your interest and hard work in the field of copper metabolism disorders.
Patients are actively being recruited for a Phase 2, multi-center, open-label, study to evaluate the efficacy and safety of WTX101 administered for 24 weeks in newly diagnosed Wilson Disease patients. This study is being sponsored by Wilson Therapeutics to evaluate WTX101 (bis-choline tetrathiomolybdate) as a de-coppering agent for the treatment of Wilson Disease. The results of the Phase 2 study will allow Wilson Therapeutics to advance WTX101 into late-stage clinical trials in patients with Wilson Disease.

The study is being conducted at 9 Wilson Disease expert centers (University of Michigan Hospital, Ann Arbor, MI; Yale University Medical Center, New Haven, CT; UCLA Ronald Reagan Medical Center, Los Angeles, CA; Northwestern, Chicago, IL; Medical University of Vienna, Vienna, Austria; University Hospital, Heidelberg, Germany; Institute of Psychiatry and Neurology, Warsaw, Poland; City Hospital, Birmingham, UK; Royal Surrey County, Guildford, UK).

The ongoing study has recently been modified to allow for an optional 12 month treatment extension for subjects who successfully complete the initial 24 weeks of WTX101, with regulatory and ethics committee review and approval ongoing for this amendment.

WTX101 is the bis-choline salt of tetrathiomolybdate (TTM). TTM is a novel de-coppering agent with a unique mechanism of action that has demonstrated a more rapid and improved control of copper in Wilson Disease patients. Unlike other de-coppering agents currently available for the treatment of Wilson Disease that form unstable complexes with copper and other metals (e.g., iron and zinc) and are excreted via urine, TTM selectively forms high stability complexes with copper and proteins. These complexes are then primarily excreted via the bile, restoring the normal excretion route of copper that is impaired in patients with Wilson Disease. By rapidly binding and controlling copper in stable complexes, TTM may reduce the risk of mobilizing and transiently increasing the levels of free copper in patients starting de-coppering therapy. The rapid and improved control of copper is very important as high levels of copper cause tissue damage and transient increases in free copper after initiation of therapy is believed to be involved in causing additional tissue damage, especially in the central nervous system. As a result of the improved control of copper WTX101 is therefore expected to improve control of the disease as well as reduce the risk of neurological deterioration after initiation of treatment in Wilson Disease patients with neurological disease.

The improved salt formulation of TTM, WTX101, has also been tested in clinical trials in oncology and was found to be safe and tolerable while efficiently lowering copper levels with once daily dosing. A once daily dosing regimen is expected to translate into improved patient compliance in Wilson Disease patients and therefore fewer treatment failures.
Yale Hires New Biotech Associate

In May, Michael Schilsky MD, Director of The Wilson Disease Center of Excellence for Wilson Disease at Yale University, approached the WDA about financially assisting him in the hiring of a Biotech Associate to work at Yale University from July 1, 2015 to December 31, 2015. The position is intended for assistance in generating an NIH grant application that hopefully will lead to the creation of a national consortium, in partnership with the NIH and the Wilson Disease Association, on Wilson disease. The new Biotech Associate will be responsible for being liaison to the Association and to the medical centers that will participate in the proposed consortium. Dr. Schilsky and his assistant will create appropriate data forms for all proposed medical centers to participate, generate clinical research protocols and begin the process of creating case report forms and information needed to generate a patient registry and tissue, serum and DNA repository for Wilson disease.

Dr. Schilsky was able to generate partial funding from his research funds at Yale that are available for this purpose. We were very fortunate to have a donor step up and offer to help us fund the rest of the six month position. This is a very exciting step toward the WDA’s goal of establishing a Patient Registry to learn important new information for the benefit of current and future generations of families affected by Wilson disease!

Gordon Research Conferences

The Gordon Research Conferences provide an international forum for the presentation and discussion of frontier research in the biological and chemical sciences, and their related technologies, and for over 80 years, have been viewed as the world’s premier scientific meetings. This July one of our medical advisors, Dr. Dennis Thiele from Duke University in North Carolina, is co-organizing the Cell Biology of Metals Gordon Research Conference (GRC) in Mount Snow, Vermont.

The organizers have assembled a scientific program and a slate of speakers with an international collection of world-leading scientists engaged in research on the roles of metals in biology. Particularly relevant to Wilson Disease are speakers such as Michael Petris PhD, Svetlana Lutsenko PhD, Jonathan Gitlin MD, Roman Polishchuck PhD, Martina Ralle PhD, and others whose work is closely tied to understanding human copper metabolism.

The WDA was pleased to offer partial support, along with ten other organizations whose focus is on metals and disease, for this stimulating conference.
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*Vote*

If you are a current member of the WDA and will NOT be attending the Annual Conference in Weehawken, NJ, please vote by mailing your completed ballot to the WDA office before September 26, 2015.