
◆ THE COPPER CONNECTION ◆

Wilson's Disease Association, International Newsletter

July 1999

WDA Annual Meeting Highlights

President's Note: I am providing the following highlights of the annual meeting for all the people who were unable to attend. I believe it was extremely informative. I personally look forward to seeing as many of you as possible at next years meeting.

The Annual Wilson's Disease meeting began Friday, May 7th with a two hour hospitality session at the Sheraton, Ann Arbor, Michigan. Refreshment were provided and many people had an opportunity to chat with other patients with Wilson's Disease. The WDA Board meeting was held afterwards that evening with 18 people present.

Saturday morning began with an informal continental breakfast followed by a full day of activities and speakers. A total of 98 people were in attendance.

We had a guest from Germany, Dr. Catherin Smolarek, who practices in Heidelberg Germany with Dr. Stremmel. Her practice to a great extent involves the care of Wilson's Disease patients.

Attorney William Crawford

The topic was Medicare/Medicaid and disability insurance along with the process by which patients become eligible for those services. Eligibility usually requires no less than five months. Applications for eligibility are frequently denied on the first round and often require appeals and even legal actions all the way to the courts, in order for success to be achieved. It is my opinion that generally, patients requiring these services would be best to speak to an attorney with experience in this field.

Dr. George Brewer

Dr. Brewer spoke on the current state of

Next Annual Meeting:

**May 5,6,7, 2000
Orlando, Florida**

(Details provided later)

Wilson's Disease diagnosis and therapy. He reported that he has a total of 225 patients, of which 197 are active. His treatment of patients with zinc acetate include thirty-seven patients who have been treated by him for over 10 years and 90 who have been treated for five years. He has treated 24 children and 22 pregnant patients. Thirty-one are pre-symptomatic patients.

Dr. Brewer and Dr. Michael L. Schilsky are currently conducting a trial of Tetrathiomolybdate (an investigational drug) and Trientine for the initial therapy of neurological Wilson's Disease patients. Newly diagnosed neurological WD patients interested in taking part in this study should contact Dr. Brewer at (313) 764-3784.

Dr. John Fink

Dr. John Fink is a Neurologist who described his experience at the University of Michigan treating the neurological symptoms of Wilson's Disease patients. He pointed out that what is unusual about WD patients is that they have a variety of different pathologies in different parts of their brains because of copper being deposited in different parts of the brain. As a result, they have various combinations of spasticity which is the tightening of muscles causing flexion or

IN APPRECIATION

The WDA would like to thank all the guest speakers who provided us with such informative presentations at this year's Annual Meeting in Michigan. In addition, thanks to Drs. Schilsky and Brewer for their medical expertise and support. A special thanks is extended to Len Pytlak for organizing and coordinating this year's meeting!!

bending of the arms as in making a fist or bringing your fist to your shoulder. They also develop rigidity (the inability to bend arms or legs in various positions).

In addition, they develop dysphonia difficulty with moving their tongue, lips and throats in order to form speech and to swallow. And lastly, they develop chorea, which is random slow motions in various directions. This is distinctly different from tremor, which is generally repetitive motion in the same directions.

Dr. Fink points out that each of these different types of symptoms can be treated with various medications, some of which take 5 or 6 months to demonstrate an improvement. It is, therefore, necessary for a long period of time to be used in many trials and much patience before seeing considerable response. (see page 2, HIGHLIGHTS)
(HIGHLIGHTS, continued)

Dr. Bob Luit

Dr. Luit is a neurologist specializing in movement disorders. He is organizing a meeting in the year 2001 of moving disorder specialists. A portion of the meeting will focus on Wilson's Disease.

He discussed some details of what will transpire at the meeting and how the WDA can participate.

Karen Kluin

Ms. Kluin is a speech and language pathologist at the University of Michigan. She discussed the various problems WD patients have with speech, language and swallowing. She demonstrated various different techniques patients could use to improve their speech. She had the whole group go through some of these exercises which included posture, deep breathing, slow speech, and carefully breathing out. It was quite interesting to the entire group to see the way in which she approached speech pathology.

Dr. Michael Schilsky

Dr. Schilsky discussed the indications for liver transplant. He pointed out that except in the acute phase, most patients would escape a liver transplant if medicated properly and if compliance is good. There is no reason to believe that patients with WD ultimately will require a liver transplant. He did point out that statistics show a 20% mortality rate in the first year, but that newer statistics suggest that it is at little as 10%, however more than half of patients waiting for liver transplants die while waiting for an organ.

Dr. Schilsky then discussed ways in which scientists are investigating the possibility of transplanting normal liver cells to abnormal livers in order to get WD patients to carry out normal metabolism of copper. Some of these experiments have been successful at this point in animals.

Afternoon Sessions

After lunch, three consecutive support activities were conducted. One was for relatives of patients, one was for teenagers and one was for the balance of the group. Many people found these activities of enormous benefit to them because they found themselves understood by people regarding issues that no one else previously seemed to understand. It also improved their insight into the situations with which they have had to deal.

University of Michigan Clinical Research Tour

After the afternoon sessions, the group took three buses to tour the Clinical Research Center at the University. The group was divided into five groups. They went to various rooms to hear talks on different aspects of the diagnosis, dietary treatment, speech treatment, swallowing treatments, diagnostic procedures and other issues involving care in the Clinical Research Center.

Special Reception For Mr. and Mrs. Morton Sellner

Mr. and Mrs. Morton Sellner (Ascher Sellner's parents) were acknowledged for their generous gift of \$1,000,000 to the University of Michigan Department of Human Genetics. A special reception was held after the tour at the University. Their gift was matched with \$750,000 by the University of Michigan, thereby creating The Morton and Henrietta Sellner Professorship in Human Genetics. This means that there will always be a physician providing medical care and research for Wilson's Disease patients! Mr. and Mrs. Morton Sellner received as a token of appreciation a Tiffany's engraved crystal piece. A similar gift was presented to their son.

Kid's/Teen Group

A Kid's/Teen Group was formed at the WDA annual meeting in May. Nicole Russell aged 17, and Molly Gilcrest, age 14, headed up a discussion/support group of 10 teenagers (7 Wilson's Disease patients and 3 friends/family of WD patients). Nicole commented that "it was beneficial for everyone of us to find kids our age with Wilson's Disease. By the end of the day, were so tight no one could tear us apart. We all wished the convention had lasted longer. It was the first time for each of us to meet someone else with Wilson's. It gave us so much comfort and hope. I can't begin to tell you how special each of them was to me. I hope more kids join us next year."

Nicole can be contacted by e-mail at the following address:
princesspussycat@hotmail.com

She would love to hear from other kids/teenagers aged mid 20's and younger who have Wilson's Disease or are in close contact with someone who has Wilson's.

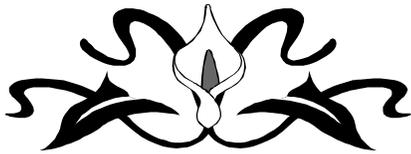
Patient Care Fund

A Patient Care Fund was established at the Annual Meeting. Donations of \$1000 will be matched with \$1000 from the \$35,000 gift received last year from Morton and Henrietta Sellner. Thanks to Barbara Fox, Len Pytlak, Ascher Sellner and Luke Chung for getting the ball rolling. Donations can be made using the

**Welcome Aboard
New WDA Volunteers**

**Diane Laurain
Lúcio Mazza
Barbara Noci**

We need assistance with fundraising. Anyone interested in this or other volunteer areas, please give Dr. Sellner a call at 1 (800) 399-0266.



WDA — In Action!

The WDA recently participated in NORD's exhibit booth at the recent American Academy of Family Physicians in San Francisco. Exposure to the 10,000 participants in attendance helped the WDA continue its mission of relaying valuable information about Wilson's Disease to healthcare professionals.

1999 Federal Budget- Orphan Disease Research Funding

The FDA's Orphan Products Grants Program will receive only \$12 million, the same amount as in previous years. However, FDA administrators have reduced this sum by 8% during the past few years, so that FDA could use the money elsewhere. This is the only federal program reserved for rare disease research and \$12 million is hardly enough to support investigations on just a few disorders.

Since initiation in 1983, the FDA Orphan Products Research Grant Program has provided funds for the development of 20 new orphan drugs and 3 medical devices which have been approved by the FDA for marketing.

*Orphan Drug Act
Enacted in 1983*

*Created financial
incentives to
entice drug
companies into
making drugs for
rare diseases.*

How you can help: Congress needs to understand how important this lifesaving research program is to people with rare disorders. Ask your Congressman and Senators to **increase the FY2000 appropriation for the FDA Orphan Products Research Grant Program to \$20 million.**

(NORD - National Organization For Rare Disorders, Inc)

Mrs. Wobble Goes To School

In the previous issue of The Copper Connection, information on a children's book written by Susan Kuiper (a WD patient) entitled "Mrs. Wobble Goes To School" was included. Copies are now available through the WDA. To purchase a book, send a check for \$11.00 and your name and address to Dr. Sellner, 4 Navajo Drive, Brookfield, CT 06804. This cost includes postage and handling.

DONATIONS TO THE WILSON'S DISEASE ASSOCIATION

A donation to the Wilson's Disease Association in honor/memory of a family member or friend could be the best present of all to receive and give. The Association will send a card notifying the person or family named below of your contribution.

Enclosed is my contribution of \$ _____. (Please check honor/memory/Patient Care Fund below)

In honor ____ of _____ (PLEASE PRINT NAME).

In memory ____ of _____

Patient Care Fund _____

Send acknowledgment of gift to: _____

(Please Print Complete Name and Address)

Donor's name & address: _____

(Please mail your donation to: Stefanie F. Kaplan, 869 Kallin Avenue, Long Beach, CA 90815.) Thank you.

ATTENTION ALL WDA MEMBERS:

Thank You!! We have received many renewal applications from WDA members!! If you have not yet mailed your application, please help us update the WDA database by sending this form today.

**Wilson's Disease Association
Membership/Renewal Application**

Membership (Check one): New ____ Renewing ____

Name: _____ Sex: _____

Street Address: _____

City _____ State: _____ Zip Code: _____ Country: _____

Home Phone: _(____)_____ Work Phone: _(____)_____

E-Mail: _____

Relationship to Wilson's Disease: (circle one)

Patient	Parent	Grandparent	Sibling	Child	Grandchild
Aunt	Uncle	Cousin	Friend	Other _____	

FOR PATIENTS:

Birthdate: _____

Current Treatment: (circle all that apply)

Penicillamine, Trientine, Zinc, Tetrathiomolybdate, Other: _____

Initial Symptoms: _____

Diagnosis Date: _____ Initial Treatment: _____

Other Treatments: _____

If you are unable to pay dues, please send back your form. Any amount, regardless of how small, helps. Amounts of \$50 or more will be acknowledged in the newsletter unless otherwise requested.

Basic \$35 - Sustaining \$50 - Endowing \$100 - Life \$1000 - Other \$ _____

Would you like your name placed on a list with others willing to give each other support by writing, calling or meeting? Yes ____ No ____

Please mail with your check or money order to:

Wilson's Disease Association
c/o Stefanie F. Kaplan
869 Kallin Avenue
Long Beach, CA 90815

WDA Board Meeting Summary

The WDA Board met on Friday, May 7, 1999 in Michigan. The Association has provided considerable patient services during the past year, including referrals to physicians and centers of excellence, information to patients, public organizations, individuals in schools and interested parties.

E-mail has been provided and answered and an Internet site has been established and broadly expanded to include information, references, contacts and various other services. Additional support was provided to patients in the form of transportation expenses for care.

Support groups were organized in New Zealand, Australia, Denmark, Spain, and Brazil as well as throughout the United States.

The membership has considerably increased in size. The mailing list is currently being up-dated with e-mail addresses so that members can be contacted and/or receive the WDA newsletter by e-mail. This will save considerably on postage and printing.

The By-Laws will be revised to increase the number of board members, thus increasing the skills represented on the board.

Thanks to all the WDA volunteers, membership, services and dissemination of WD information has increased internationally.

Volunteers are still needed in the following areas:

- * coordinator of Internet services
- * chairperson to increase physician awareness of WD
- * development of patient materials
- * contributors to the Copper Connection newsletter
- * coordinator of public education
- * coordinator for fundraising
- * coordinating local support groups

All volunteers receive direction for the specific areas. Please consider sharing your expertise in support for the WDA. Contact Dr. Sellner at 1 (800) 399-0266 or e-mail at hasellner@worldnet.att.net.

The Board agreed to allocate \$35,000 in the president's discretionary fund to create a matching fund to which \$1000 contributions will be matched by \$1000 from the \$35,000 matching fund in order to create a \$70,000 fund. This fund will be designated as an endowment of which the income will be expended entirely on patient care, such as diagnostic, therapeutic, medication and transportation services.

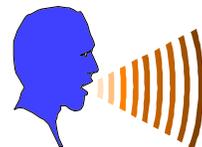
Dr. Sellner has become a member of the Board of Directors of NORD, The National Organization for Rare Disorders.

DONATIONS

We gratefully acknowledge the following gifts of \$50.00 or more:

Teresa Earley Bowen
 Luke Chung
 Barbara Fox
 Ruth Hill
 Joanne Doades
 Angela Doyle
 George Feldman
 Gladys and Barney Goldstein - In memory of Mitchell Lane-Price

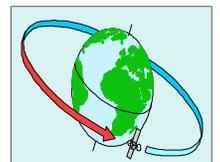
James and Iva Jean
 Larry Irwin
 Virgil and Mary Ann Laurain
 Diane Laurain
 Judy Martin
 Penny and Fetzer Mills
 Mr. and Mrs. Fetzer Mills, Jr.
 Jeffrey Nehal
 Len Pytlak
 Susan Rust
 Genevieve Roberts
 Ascher Sellner
 Ruthann Siebert
 Mr. and Mrs. Daniel Slavin
 Carol and Sparky Terry
 Charlene Walker
 Frank and Bernadine Wall
 Clara and David Watson
 Julia Whittier



Remind a Buddy to
take his/her meds!!

WDA INTERNATIONAL CHAPTERS

The WDA welcomes international members. We have received a great response from people around the world (see page 6 for a list of formed international chapters). If you are interested in forming such a group, please fax Dr. Sellner at 1 (203) 743-6196 or e-mail hasellner@worldnet.att.net



My Mother's Experience

My mother, Helena, died at the age of 58 of complications associated with Wilson's Disease. She was diagnosed two weeks prior to her death.

The first time any of us ever heard of Wilson's Disease was in December 1998, one month prior to her death.

She had had a brief hospitalization in December for tests because she had noticed some blood in her stools. This was the first obvious symptom she had ever present with. At the time, her physician ran several tests including a liver biopsy. Wilson's Disease was one of several conditions they were trying to rule out. They had discovered at that point that she did have high copper levels. She was placed on propranolol and sent home with a plan to follow up with a liver specialist as soon as possible. She scheduled that appointment for January 13th, after the holidays because it was such a busy time of year for her. She loved to decorate and bake and make a huge deal of Christmas, for her family, friends, the homebound, everyone.

Sometime between her episode in December and her appointment scheduled in January, she stopped taking her propranolol due to side effects. She chose not to start any other treatment or therapy until her upcoming appointment with her specialist. She also took high doses of vitamins daily, thinking they would help boost her energy level. Little did she realize in doing so she was taking a minimum of 1000 mcg a day of copper in addition to what she would normally consume in her diet.

On January 11, 1999, her 58th birthday, (2 days prior to her appointment with her new specialist) she had an overwhelming wave of nausea while preparing to go out for a birthday luncheon with my sister. She ran to the bathroom and began vomiting a significant amount of bright red blood. She called an ambulance and was admitted to the hospital. That evening, my mother's new liver specialist met with all of us outside my

mother's room. He told us she had Wilson's Disease. He was placing her on the organ donor list for a new liver, but said that she was not a good candidate for transplant at that point due to her new bleeding episode. It appeared that her liver was severely damaged and swollen, causing varices on her esophagus and possibly her stomach as well. He spoke to us about encouraging her to have a "Tips" procedure done to her liver. This would expedite the flow of blood through her liver and decrease the size of the varices, to reduce her chances of further bleeds. This procedure was still elective at that point because her bleeding episode of earlier that day had ceased. Her doctor also stated that should the bleeding re-start, this procedure would be done on an emergent basis.

The hospital called us the next morning at 3 a.m., my mother had started vomiting blood again, in copious amounts, and we were needed at the hospital. When my father and I got there my mother was still vomiting blood, but we were allowed to see her briefly before she was sedated, intubated, and sent down to surgery. It was the last time she was ever awake and able to speak, she was very frightened. She wasn't sure she would survive the procedure. She told my father she loved him.

Her "Tips" procedure took longer than expected, but was a success. Initially my mother did seem to improve after a few days. She continued to have gastric bleeding for a day or two after the procedure. She began to respond to our voices and touch. We thought she would get better.

Then her other vital organs began to fail. Her lungs first (pneumonia) and then her stomach. She became severely jaundiced. She was putting out very little urine and retaining a lot of fluid. She was placed on full life support and Morphine for pain.

After speaking with her physician, we all decided together to let her go. We sat with her all day as her life support was gradually discontinued. We watched her leave us. Fifteen days after

she was diagnosed with Wilson's Disease, my mother died.

After the fact, we can all look back over the last few years of her life and see the other subtle symptoms that we had passed off as being post menopausal or part of normal aging, or just part of the whole make-up of her personality which made her her.

She never had any of the classic signs of Wilson's disease, such as "Kayser-Fleisher rings around her irises. She did however become fatigued very easily, have increasingly frequent bouts of depression, and irrational mood swings. She had difficulty eating, unable to consume a normal meal and subtle significant weight loss over the past couple of years.

Anyone with these symptoms should check with their primary care physicians. If Wilson's Disease is suspected, see a specialist immediately. Time is of the essence!

If you know you have Wilson's, I beg you please, STAY ON your medication, it's so simple to do. Your families and loved ones need you. Irreversible damage and death can occur so quickly if you don't. I know, I lost my mother at age 58. She was too young to die.

** LMLM **



Support Group Meeting

Saturday, October 2, 1999
Washington, DC.

For more information, contact Jim McDonough (301) 294-7741 (leave message) or Dr. Ascher Sellner at (203) 775-9666.

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Wilson's Disease Listserv**Discussion Group**

Click on "discussion" in the WDA website
or send e-mail directly to: www.acsu.buffalo.edu/~drstall/

Wilson's Disease Patient Histories

www.gourmandizer.com/wilsons

National Organization for Rare Disorders® NORD®

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www.NORD-RDB.com/~orphan>
P.O. Box 8923, New Fairfield, CT 06812
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If you have changed your address, please send us your current address so you will not miss the next edition of "THE COPPER CONNECTION".

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ADDRESS: _____

CITY: _____ STATE: _____ ZIP CODE: _____ COUNTRY: _____

Please mail your change of address to: Stefanie F. Kaplan 869 Kallin Ave., Long Beach, CA 90815

Inside This Issue:

- * *WDA Annual Meeting Highlights*
 - * *Next WDA Meeting: May 5,6,7, 2000*
 - * *Support Group Meeting Oct. 2, 1999*
-

WILSON DISEASE ASSOCIATION, International

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(FORWARDING SERVICE REQUESTED)

TO:
