
◆ THE COPPER CONNECTION ◆

Wilson's Disease Association, International Newsletter

November 1999

PRESIDENT'S MESSAGE

I anticipate great fun for all at our annual meeting in Orlando at Disney World this coming May 5-7. We will have a great program plus some exciting new developments to share. So rush out to buy your calendar and mark the date.

What's new? The Wilson's Disease web site is expanding. We now have information available in a variety of languages. Contacts are listed for many countries. The Italian page is the newest to be added with part of the French and German pages already up. For those of you who are interested, the copper contents of food are also listed on our web site. As you may know, it's far more important that you take your medication than it is that you worry about the content of your food. With a few exceptions like lobster, liver, organ meats, most other foods in moderate amounts are reasonable to eat. You probably know by now, that it is not the copper in your system that poisons you, but the free copper in your system. Perhaps you don't realize it, but no matter how much decoppering you go through you will always have enormous amounts still left in your liver; these will have no bearing on your health as long as you take your medication.

A new clinic has opened at the University of Michigan for patients with Wilson's Disease. Currently, it (See page 2, MESSAGE) (MESSAGE, continued)



Next Annual Meeting:

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

We look forward to seeing you at our next annual meeting. The meeting will be at the Hyatt Orlando in Kissimmee, Florida. The schedule will be as follows:

Friday, May 5
Hospitality Reception 5 -7 p.m.

Saturday, May 6
Meeting 8 a.m. - 5 p.m.
(Breakfast and lunch included)

Sunday, May 7
Meeting/elections 8 a.m. - 11 a.m.
(Breakfast included)

A mailing will be going out to members in January with more details of the meeting, as well as registration information.

Hotel reservations may be called in now to the Hyatt Orlando (407) 396-1234. Hotel rates are \$89.00 single/double; \$114.00 triple; \$164.00 quad (plus tax). This rate will also be extended to you if you choose to come in before or stay after the meeting. Simply identify yourself as attending the Wilson's Disease Association meeting May 5-7, 2000.

This meeting is a great source of information. It is also a fantastic opportunity to receive and lend support to other patients and relatives who are dealing with this disease on a daily basis.

Our Story Needs To Be Told

The mother who lost her teenage daughter, the twenty year old man who will live the rest of his life in a nursing home, and the woman that was committed to

a mental hospital are all tragic stories. These cases are all too common for



Wilson's Disease sufferers and their families. Most of us would jump at the opportunity to share our experience with the public hoping to prevent others from suffering this ill-fated disease. The opportunity has been given.

Several testimonies of lives that have been affected by Wilson's Disease will be sent to national syndicated news shows (20/20, Dateline, 60 Minutes, etc...) and other media types. This is an attempt to inform the public of Wilson's Disease and its tragic effects on people's lives. The awareness could prevent someone from long term suffering or perhaps death.

Recently, an editor and reporter with an Arizona newspaper was diagnosed with Wilson's Disease. During the reporter's treatment at the University of Michigan, he wrote a two part article about Wilson's Disease and how the disease had suddenly turned his life upside down. The articles from the local paper resulted in several people being properly (See page 2, STORY)

meets once a month and its future expansion is eagerly anticipated.

Dr. Michael Schilsky has moved his academic location to Mount Sinai Hospital in New York City along with the center of excellence for Wilson's Disease care in the Northeast.

Money is the primary limitation the association has in trying to expand its support, education and its patient relief activities. I sincerely hope that you, your family and friends will direct as much of your charitable giving as you possibly can to the Wilson's Disease Association. It is my hope in the near future we will be able to afford full time staff to direct their activities exclusively to the assistance and betterment of patient's with Wilson's Disease.

Please send your tax-deductible contribution to the Wilson's Disease Association. My Aunt and Uncle Gladys and Barney Goldstein have made a substantial contribution to the Association and have since made gifts on the occasions of relative's birthdays and other events to support the activities of the organization. These are acknowledged by a card to the recipient in honor and acknowledgment to the donor of the gift.

Those of you with whom I have been in touch, by phone and e-mail, please keep me aware of your progress.

All of you are encouraged to write letters requesting contributions to Wilson's Disease. Please send me copies of these letters that you send out or that we may send out on your behalf.

Congratulations to those of you undertaking an effort to awaken the media to the existence and problems of Wilson's Disease. This must be an on-going effort to keep this

disease in the public eye. For those of you interested in participating, please contact Jimmy Splawn at his e-mail address: JSplawn@pdq.net.

I also want to apologize to those of you who received your July 1999 newsletters late. It was our intent to e-mail our newsletters to approximately 100 members, however, this was never accomplished because of some technical difficulties. Until these difficulties are overcome, everyone will receive a newsletter by mail. If you do not choose to have your newsletter sent by E-mail once the glitches are straightened out, please let us know. Send a note to Stephanie Kaplan notifying us to mail you a hard copy of the newsletter instead of sending it by e-mail. Please bear in mind that the newsletters mailed cost more than 75 cents a piece, and by e-mail it costs nothing. Therefore, every time you get a newsletter by e-mail you will be making a contribution to the Association of just under \$1.00. Thank you and I wish you Happy Holidays.

Ascher Sellner, M.D.

(STORY, continued)

diagnosed. Just imagine what national attention would do for the awareness of Wilson's Disease. The national media receive hundreds of suggested stories to air, hopefully some day they will listen to our story.

(If you are interested in sharing your story, please e-mail Jimmy Splawn at Jsplawn@pdq.net).

Name Recognition

Wilson's Disease is currently treated with three FDA approved medications — Penicillamine, Trientine and Zinc Acetate. We are fortunate to have choices in our individual treatment plans and look forward to future research of new therapies. For now, here is a list of names associated with each medication.

- Penicillamine (Depen, Cuprimine)
- Trientine (Syprine)
- Zinc Acetate (Galzin)

TRIENTINE AVAILABILITY

Some of you have had trouble obtaining Trientine (Syprine). This has happened in the past. At the annual meeting of NORD (a National Organization for Rare Diseases), Dr. Sellner met with Congressmen and Senators to request that they require manufacturers of orphan drugs to maintain adequate supplies so that patients requiring the drugs will never run out of them.

Patient Care Fund

A Patient Care Fund was established at the Annual Meeting. Donations of \$1000 will be matched up to \$35,000. Donations can be made using the gift form in this issue.

Newsletter Volunteers

The Copper Connection Newsletter Editor organizes and compiles news information to include in each newsletter. The topics include articles and letters of interest to Wilson’s Disease patients and their families. We welcome your contributions. Please send them to the Editor, Delia Ruiz. Her address is located on page 7. Thank you.

Volunteers Needed

The WDA is a non-profit organization run by volunteers. Help is always needed. Please give us a call at:
1 (800) 399-0266.

How Does Zinc Acetate Work?

Zinc works by inducing a protein (metallothionein) in the intestine which in turn binds copper with great affinity and holds on to it, so copper does not get into the blood system. This protein and the copper bound to it eventually pass out of your body via the stool as intestinal cells slough off every 4-6 days. Furthermore, zinc induces this protein in other tissues such as the liver, which binds copper and prevents the excess copper from causing harm. The bulk of evidence indicates that the copper that is bound to this protein is not available to do harm.

Taking zinc acetate with regular food is a bad idea. You may not be absorbing any of it as many foods (such as most sources of carbohydrates and dairy foods) bind it and make it unavailable for absorption. Zinc acetate should not “just be taken”, but it should also be “delivered” in a way the body can use. It must be absorbed to do its job. Therefore, zinc acetate should be taken at least one hour away from food (before and after).

Wilson’s Disease patients should always be under the care of a medical physician.

DONATIONS

We gratefully acknowledge the following gifts to the Wilson’s Disease Association:

- Pat Pei \$3000.00
- Don Keller Fund of the Community Foundation of Collier County \$4000

Thank You!!

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:

Beginning March 2000, The Copper Connection will be mailed to updated and renewed members only. This is due to the high cost of mailing newsletters. If you have not yet mailed your application, please do so today. Thank you.

**Wilson's Disease Association
Membership/Yearly 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

MY LIFE WITH WILSON'S DISEASE

by Agnes Mattic

I was born in Vancouver, Canada on November 19, 1933, the youngest of thirteen children. My mother had four miscarriages. The first two died when they were young so I never knew them. My brother Albert died in 1942 at the age of 19. They said he had cirrhosis of the liver. He died very suddenly. Then my sister Gladys died at the age of 26, in 1945. She died in a mental hospital. We later learned they both had Wilson's Disease. I knew I was different, always a loner, and always sickly, but I didn't know why. My first symptoms showed up when I was 16 years old. I went blind for a month. I was working at that time. I quit school in grade 10 because it was too hard to walk the six miles to school and back.

I kept on getting sick, so my mother said I better go into the hospital. I went to Crease Clinic outside Vancouver. They gave me the B.A. L. shots, sixty of them, two a day. I remember every one of them. Boy were they painful! That is when they told my mother I had Wilson's Disease. When I went back to work, I met my husband and got married, January 7, 1954 and came to Michigan. Between getting sick, having eight miscarriages, and three children, I survived, but my marriage did not. In 1955 they gave me Iversinete in Grace Hospital in Detroit, then I went to Lafayette Clinic in 1956, also in Detroit. From there I went to the University of Michigan Hospital. I was pregnant so they told me I should be hospitalized full time. So I went to Northville, where my son was born on my birthday. He was beautiful. He is now 37 years old, lives in Hawaii, has a good job, and I am very proud of him.

In 1962 I had a total collapse; my legs and my speech went, and I couldn't feed or dress myself. So the U of M asked me if I would try Penicillamine. My family sent \$500 for the doctors to get the medicine from England. When I first took it, I went into a coma for two weeks. After I came out I got stronger, my speech came back, I could feed myself, but it took four months before I could walk. I was on Penicillamine until my body became immune to it in 1985. I returned to the University of Michigan and started taking zinc acetate, but it didn't agree with me. So Dr. LeWitt told me of another drug named Trientine. I tried it and have been taking it ever since. I had two more beautiful children. A daughter and a son. I am very proud of all my children.

By the way, I went back to school in 1986. I got my High School diploma with a 4.0 grade point average at the age of 54. I also went to college for three years. I am finally enjoying life. I have been going to the Salvation Army senior camp and look forward to going again this year. I also go to Bible Camp, because God is a very important part of my life. I love to cook, read, line dance and especially bike riding. I ride all over Ann Arbor, where I live.

It is 1999 now and I have moved to a senior citizen apartment. I have a beautiful two bedroom apartment and I also have a walker. I enjoy traveling with my son and picnicking with my girl friends Nancy and Alice. So as you can see, I am still getting up and about.

Wilson's Disease Facts

- 1 in 100 individuals in the general population carries the Wilson's disease gene. Carriers have one normal and one abnormal gene.
- More than 30 different mutations have been identified.

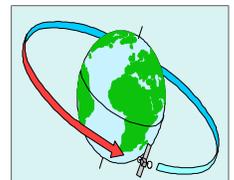
Contributors To This Issue

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Thank You!!

WDA INTERNATIONAL CHAPTERS

The WDA welcomes international members. We have received a great response from people around the world (see page 7 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



NORWOOD-DINGELL PATIENTS' BILL OF RIGHTS
PASSES HOUSE OF REPRESENTATIVES

On October 7, 1999, the U.S. House of Representatives passed the bi-partisan Patients' Bill of Rights sponsored by Representative Charles Norwood (R-GA) and Representative John Dingell (D-MI). NORWOOD has been a staunch supporter of this bill (H.R. 2723), which will provide critically important health care protections such as:

- Health plans must provide for an external appeals process when a patient is denied coverage for a service or product that his or her doctor feels is necessary, and the appeal panel's decision will be legally binding on the insurer.
- Access to specialist physicians.
- The right to sue health plans if insurer decisions hurt the patient, or if the plan does not abide by the ruling of an external appeals panel.
- Safeguards against frivolous lawsuits.
- Health plans must provide for patient choices of doctors and hospitals; availability of local health services; access to appropriate specialists; optional use of pediatricians and ob/gyns as primary care providers; continuity of care (patients not being forced to change doctors or hospitals mid-treatment); guaranteed coverage of emergency care at the nearest emergency room based on a reasonable determination by the patient that there is an emergency ("prudent layperson" standard).
- Health plans may not have the "gag rules" that prevent open communication between doctors and patients, nor financial or other incentives for providers to deny or limit care, or discriminatory practices that prevent certain licensed providers from participating in the plan.
- Insurers must obtain patient permission to release private medical information.
- Plans may not discourage or interfere with patient participation in clinical trials, and insurers must pay for routine medical costs during clinical trials if patients have a serious or life-threatening illness for which no standard treatment is effective, and the therapy being tested in the trial offers potential for benefit. The trial must be approved by the NIH, Veteran's Administration, or Department of Defense.
- Plans that cover prescription drugs and have a "formulary" must have participation of doctors and pharmacists in developing the formulary, must disclose the formulary restrictions to patients, and must provide for exceptions to the formulary where the doctor determines a certain drug is necessary for the patient.
- Health plans must provide, in layperson's terminology, good information to patients about what is covered and what is not, about the plan's appeals process, and about any other conditions that apply.

The right to obtain a pharmaceutical that is medically necessary even if it is not on your insurance company's formulary.

The bill contains many other important protections for patients. The major difference between the Norwood-Dingell bill and three other proposals that were debated by the House of Representatives was the right for patients to sue managed care insurers in state courts. Until now, health insurers have been shielded from most lawsuits because of a prohibition in federal law.

If you wish to read the Norwood-Dingell bill (or the rival defeated managed care reform bills), you can locate them at: (www.house.gov/rules/rule2723.htm), or you can access: www.familiesusa.org.

What Happens Next?

The Senate passed a managed care reform bill (S.1344) several months ago, which the President has threatened to veto. The Senate bill has been criticized for several reasons. It does not provide a mechanism for patients to sue a health plan in a case where the health plan's decision not to cover a medical treatment has had an adverse impact on the patient. This legislation would affect only those who are employed by large multi-state employers who are "self-insured." The House bill will affect all insured Americans.

Now the House and Senate must form a Conference Committee where they will try to formulate a compromise between the two bills. Republican leaders in the Senate have stated that they will not agree to any health insurance reform legislation that allows patients to sue health insurance companies. Therefore, the Conference committee could: 1)

Delete the provision in the House bill that allows patients to sue insurers.

2) Weaken the House bill so it more closely conforms to the Senate bill.

3) Come to no agreement, which means a compromise bill will not be brought to Congress for a final vote and the legislation will not be enacted, or 4) Adopt the House bill, H.R. 2723, as written or formulate an acceptable compromise that the President will sign into law.

President Clinton has supported the Norwood-Dingell bill, and, if it is left intact, he says he will sign it. He has asked Congress not to weaken it. However, Congress must feel pressure from constituents to enact a law this year that provides protections for all American patients. Representatives and Senators must be told that not passing a managed care reform bill this year is not an acceptable alternative.

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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

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NOTE:

If you have changed your address, please send us your current address so you will not miss the next edition of "THE COPPER CONNECTION".

NAME: _____ TELEPHONE __ (____) _____

ADDRESS: _____ E-mail: _____

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:

*Wilson's Disease Association
Annual Meeting: May 5-7, 2000*

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The Copper Connection, Editor
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(FORWARDING SERVICE REQUESTED)

TO:
