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Highlights from Newsletter Vol 1 Issue 2

Newsletter September 2000

We are growing...

Since the launch of the Wilson's Disease Support Group we have had an amazing response from both WD patients, their families, other organisations such as the British Liver Trust, CLIMB (formerly known as RTMDC), and many more and most importantly we've had so much positive encouragment from everyone!

We have also forged links with other chapters around the world, including America, Australia, Brazil, Denmark, Holland, Germany, Israel, Romania, South Africa, Spain and New Zealand. It's truly great to know that so many people affected by WD can benefit from such groups!

We've been very busy sending information about WD (pamphlets, WDSG - UK group awareness posters, and newsletters) to hospitals throughout Great Britain and we are looking into doing a medical article for one of the national newspapers.

WDSG - UK is growing steadily as a group, providing a network of contacts, forging links with other organisations, providing information, spreading a greater awareness of WD and keeping abreast of current developments and research into WD. We will do our very best to keep you informed of our progress, via our newsletter and bulletin, and via our electronic mailing list. If you have internet access please add your name to the mailing list.

Thank you to everyone who has helped to make this group a reality! Patients, relatives, friends. Firstly thanks for joining us. Together we can make a difference! Thanks for all the letters you have sent in, articles you have written for our newsletters (please keep sending them), for all the questions you've asked, and for all your overwhelming encouragment, feedback and appreciation. Patients,Thank you for getting in touch

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Lift Off...The First Six Months

The Wilson's Disease Support Group UK {WDSG-UK} is an all volunteer organisation which strives to promote the well-being of patients with Wilson's Disease and their families and friends. The organisation aims to provide informative articles about the nature of the disease, articles written by patients, families and friends about their experiences of the disease, recent progress in treatment and much more, by way of a biannual newsletter. The organisation also aims to promote

networking of WD patients and their families by helping and encouraging them to correspond with each other. Finally, the organisation strives to generate a wider awareness of Wilson's Disease and its diagnosis within the medical profession.

Wilsons Disease Supprt Group - UK - some notes of thanks...

Dr Alan Stevens ..... for giving advice and encouragment, proofreading documents, arranging meetings to discuss ideas for the group, entertaining us over lunch, and entertaining us and providing links to other people

Prof. Jim Lowe..for tutorials in operating a computer properly, and for setting up our web pages

All the staff at Self Help Nottingham, for advice, encouragment and provision of photocopying services for production of newsletters

Staff at the British Liver Trust and CLIMB (formerly RTMDC).

Top Tap Tips

A leaflet which I picked up in a local Boots store whilst in the kitchenware department, caught my attention. On the first page it said,"TAP WATER ISN'T TO EVERYONE'S TASTE."..it went on to comment about chlorine.....and more interestingly from a WD perspective the fact that lead, copper and aluminium are often present in drinking water After contacting Brita, asking for information regarding the removal of heavy metals (specifically copper) by their water filter systems and speaking to their communications manager, they have confirmed the following points:

- The Brita water filter will indeed remove the majority of copper which may be present in tap water. 99% copper removal has been recorded in laboratory tests.
- Once the copper is removed it will not leach back into the water resulting in higher concentrations of copper. Once the cartridge has stopped working effectively the tap water will simply pass through the cartridge unfiltered.
- As all tap water is different Brita cannot guarantee removal rates or give a definitive lifespan of a cartridge for copper removal. We normally recommend changing cartridges every 4 weeks but your readers may prefer to change the filter after every 2-3 weeks as an added precaution.
- The Brita water filter is not sold as a medical product and although filtered water may be beneficial to certain groups Brita make no health claims for this product.

In general if you live in a soft water area (you can tell if soap forms a lather easily !) , there tends to be more dissolved copper present in the tap water. In hard water areas there tends to be less dissolved copper. I f

you are interested in knowing the copper content in your local drinking water supply, you can contact your local water authority who will give you this information.

You may already have or may wish to consider investing in a water filter system (particularly if you live in an area with higher dissolved copper in local supply). Brita filters are the only company who have confirmed copper removal.

On a last note, if you have a water jug filter, change the cartridge on a regular basis.

Always run the tap for a few minutes, before consuming tap water, by doing this you will remove standing water (water which has been sitting within copper pipes and which may have more dissolved copper in!)

Brita filters are available from larger branches of Boots and major department and kitchenware stores. Replacement filters are available from Boots at a reasonable price.

If you need more info. get in touch with Caroline Simms.

The Story Of Penicillamine

Dr John Walshe

When you have a story to tell you should, so it is said, begin at the beginning and go on until you come to the end. Sounds simple, the problem is that the beginning is not always obvious even though the end may be. Where does my story begin? Was it when Dr. Wilson first described the illness that now bears his name, way back in 1911? Was it when Professor Cumings, working at the same hospital as Dr. Wilson (The National Hospital for Neurology, Queens Square, London) identified copper as the toxic factor underlying the disease years later in 1948? No. Was it when the Oxford scientists, researching the structure of penicillin, isolated penicillamine as a major building block of penicillin? Possibly. Was it when I first noticed the appearance of a compound, not hitherto seen in human urine, when studying a patient who had just undergone major liver surgery? Well now we are getting warm. Was it when I succeeded in unravelling the structure of this new compound and showed that its chemical name was BB dimethyl cysteine? Or perhaps when my then chief , Professor Dent, pointed out to me that this compound was in fact penicillamine? This led me on to my next observation that if I took penicillin myself or gave it to any of my colleagues prepared to volunteer, that I could identify penicillamine in the ensuing urine specimen. The story may have started on one or more of these occasions but it was certainly given a kick start when it dawned on me that this compound had the right structural formula to bind copper and might , therefore, promote its excretion in the urine. If this could be demonstrated penicillamine might well have a place in helping to off load the excess copper causing the symptoms of Wilson Disease.

Let me expand a little on this defining moment which has proved so important to the lives and well being of so many patients with Wilson Disease. It is generally believed, by and large correctly, that all new drugs are introduced by multinational pharmaceutical companies at a cost of perhaps half a billion pounds and many years of research and testing. How came it then that this "do it yourself" idea of penicillamine ever got off the drawing board? First, things were different in the early 1950's. There were no ethical committees to block progress, patients still trusted their doctors and medical research was still a respectable pursuit to follow.

In 1954 I did what every aspiring young doctor had to do to climb the promotional ladder, I set sail (Yes! in 1954 one still travelled by sea!) for the United States to study and learn at one of the leading centres of liver research at the Boston City Hospital where they were making exciting observations on the causes of liver failure. Towards the end of my stay we were asked by the Professor of Neurology to see one of his patients with Wilson Disease, Joe G. Joe had very severe tremor but, in addition, he had recently shown signs of liver damage and we were asked to advise on this aspect of his illness. On our way back from the neurological wards to the liver unit it suddenly occurred to me that my old friend penicillamine should bind copper and promote its excretion in the urine. I immediately put this idea to the department head, Dr Charles Davidson, and after I had explained to him what penicillamine was and why it should work he agreed to try and find some for me. This he was able to do, first from the drug company, Lederle, and later from a colleague, Professor John Sheehan, professor of chemistry at the Massachusetts Institute of Technology. When the first two grams arrived from Lederle I decided it was safe because penicillamine could be found in the urine of every patient taking penicillin. I divided the sample in two and took one gram myself. As I was still alive the next morning I decide it was safe to give Joe the other gram. There were no ethical committees to stop me in those far off days - fortunately.

As predicted Joe excreted excess copper and, like me, was not poisoned by the experiment. It did look, from this simple experiment if I might have stumbled on a real treatment for Wilson Disease. Progress was nearly stifled when Leered sent a further supply of what they claimed was penicillamine that did not promote copper excretion: fortunately I was able to show that this was from a batch which had oxidised on standing and lost the vital group necessary for copper binding.But there were other stumbling blocks- further reliable supplies being the greatest. Before I left the States I bought up all the available penicillamine, 50 grams of a compound from which the active part could easily easily be freed and crystallised. Not a lot but enough to enable me to prove to my professor in London that the idea really did work chemically though I still had to prove that this new treatment would actually help the patients. To be able to demonstrate this I needed two things, a reliable and adequate supply for at least one year and a patient or patients who would join the trial. My first attempt was to make my own supply of penicillamine from penicillin by slitting the molecule with strong acid. This resulted in the production of a splendid chewing gum like substance but no penicillamine. After several failures I turned to industry.

In those days penicillin was still made by fermentation and, unlikely though it may sound, The Distillers company were the main producers. Here I was lucky - they sent their medical adviser, Dr Kennedy, to see me. By the time he had climbed the four flights of steps to my laboratory he was so breathless (he was an asthmatic) that I think he would have accepted any idea! He certainly accepted mine and agreed to supply me with the drug for the necessary clinical trial. But I still had to weigh out and pack into capsules myself for all the doses needed, a time consuming occupation which resulted in a rather suboptimal dose schedule. Next I had to find a patient or patients for the trial. Here again I was lucky! My father, Sir Francis Walshe, was Europe's leading neurologist and by asking amongst his colleagues he found me three patients. All showed the expected increase in copper excretion and one, Shirley, stayed on for the trial. After some early ups and downs she started to improve and by the end of the year she was better. Shirley has now been taking penicillamine since the end of 1955, perhaps 15 kilograms of penicillamine in all !

This is clearly not the end of the story of penicillamine but it has taken it to the point at which proof of effectiveness was achieved. In the next issue I hope to narrate the story of trientine and the long drawn out struggle against the forces of bureaucracy.

Putting you in touch with each other...

Many of you have expressed the desire to be in contact or perhaps even meet other people affected by WD. If you would like to be added to our patient and family contact list, please let us know. Let us know which of your contact details you want adding to the list, e.g. address, e.mail, phone, so we can put appropriate details on our list. Confidentiality is a priority and we will only pass on details if you give us permission! Write to Linda or Caroline at the addresses given. We have arranged our second meeting again in Nottingham, as this seems to be a central location for all to get to. The date for this event will be Saturday November 25th at around 12.30 lunchtime, buffet lunch at a suitable venue. Please email if you would like details...

Strong Need for the continuation of the Wilsons Disease Clinic

Since Dr John Walshe's retirement in April of this year the WD outpatient clinic (Middlesex Hospital, London) which he has run for some years appears to be on hold awaiting decisions about what will happen, and whether the clinic will continue. As a body of people affected by WD we felt that Dr Walshe's retirement would leave a vacuum in WD specialist consultation, we were somewhat relieved to hear that Dr Gillett might continue Dr Walshe's good work. Dr Gillett has shadowed Dr Walshe's WD clinic for the past few years , he is an excellent Doctor and he has always shown a particularly keen interest in WD, its management and most importantly, us the patients . We sincerely hope he will be able to continue in Dr Walshe's footsteps and

lead the WD clinic...

on behalf of the concerned patients

Our First Meeting - a real sucess!
Linda Hart

Well, it was very good to meet those of you who came to the informal get together at The Peacock Inn, Nottingham, back in June.I reckon the afternoon was very successful and the feedback we got from you was very positive. So much so that we are organising a second event , probably at the same venue, in November. I hope those of you who came enjoyed the event and found it useful. THE DAY...

I had arrived early (as I often do when I'm nervous) and so decided to sit in a quiet corner of the lounge downstairs, with a coffee and hide behind my newspaper, until I saw someone I recognised! People came and went and I remained hidden, then in came Valerie, there was no hiding place!

Valerie and I hadn't met for many years. I think I was 15 or 16. We were both at a college in Cambridge where I vaguely remember us being on a stage, being talked about and asked questions. It's one of those foggy memories, like walking through Downing College grounds to Dr Walshes lab, armed with a 24hour urine collection botttle (until I was old enough to get embarrasssed about it and stick it in a Sainsburys carrier bag. Anyway I seem to have gone off on a ramble....back to the Peacock in June. Caroline arrived, late as usual (she'd been for some retail therapy). Soon after this, people began to arrive and the afternoon got under way. I am sure new friendships and contacts were made that afternoon and I hope they wil continue. I do sincerely feel that the event has already been helpful to people and has made a difference and we are committed to making a difference. I have been lucky enough over the years to meet quite a few people with Wilson's Disease. It must be very isolating not to have met anyone else.

Many years ago when Dr Walshe had two or three patients on Ward D5 at Addenbrooke's Cambridge, we would always make a beeline for each other and end up talking in the Day Room. Dr Walshe often commented on this. I feel we could probably empathise with each other and understand each other's problems. Sometimes our friends back home couldn't, perhaps because they hadn't experienced them. We are currently organising another event for late November. We hope many of you will come. Please email us if you would like further details !!!

Great News about Sponsorship forWDSG-UK

We have just heard that UNIVAR, who are the UK manufactures of TRIENTINE are keen to sponsor our newsletter. This is excellent news as it means that this will guarantee that we will be able to afford to continue to produce the newsletter on a biannual basis and distribute it

to people affected by Wilsons Disease.

We are looking forward to meeting their product manager, and colleagues in September.

On behalf of all the people involved with WDSG-UK we would like to express our sincerest thanks to UNIVAR who recognise our aims as a support group and for acknowledge the needs of the WD community, offering us their support and encouragement to our efforts...

THANK YOU

MEMBERSHIP

Our regularly updated mailing list and database enable us to keep record of very important data. We will be able to keep track of you so that :

- We can keep you informed about new drugs, new side effects to drugs, and developments regarding Wilson's Disease
- We can keep track of relatives to keep them informed and be sure they are properly screened to promote early diagnosis and treatment
- We can keep track of people with a special interest in the disease
- We can keep track of organisations and research centres interested in the disease and keep them aware of the association

In order to accomplish these goals, we would like you to send us an application form

Wilson's Disease Support Group UK is an all-volunteer association and no one is paid for any service to the group.

The Annual basic membership fee is just £5 Please send for a form to:

Dr Caroline Simms (Membership Coordinator)
Wilson's Disease Support Group UK
36, Sunningdale Drive, Woodborough,
Nottingham. NG14 6EQ.
Any information given to us will be treated with strictest confidence !!!

Q&A

Since many of you have sent in lots of questions we thought it might be useful to produce a questions and answer page. So here goes...

Q1/ In 1971 my close family members were tested for WD and were told they hadn't got WD. Would the test at this time have shown whether they were carriers of WD as this was not mentioned at the time?

A1/The test at the time would not have determined whether your relatives were genetic carriers of WD. The gene responsible for WD was discovered in 1993 and since then over 30 different mutations (forms) of the WD gene have been found (there are probably many more too!). Unless the familial WD gene has been isolated and identified from a WD patient it is not possible to determine whether other siblings or close relations are carriers of the same WD. It is down to probability and statistics. The reason a standard genetic screening test for WD is not yet available is due to the number of different gene mutations.

Q2/ How many people are there in the United Kingdom who have WD?

A2/ At present, there is no way of knowing an exact figure. As far as we are aware , no studies have been done to determine numbers of people with WD. We estimate that there may be upto 500 but we do not know.

Q3/ Where does WD get it's name from?

A3/ WD was named after Dr Wilson, the person who first described the illness back in 1911.

Contact Us

For membership, correspondence, and information

Group secretary and coordinator, newsletter production, information and awareness Contact :
Dr Caroline Simms
36, Sunningdale Drive, Woodborough,
Nottingham. NG14 6EQ.
e.mail : carolinesimms@hotmail.com
If you would like to be put in touch with others with WD
Patient and family corresspondance coordinator Contact :
Linda Hart
36, Audley Drive,
Lenton Abbey, Beeston,
Nottingham. NG9 2SF.

Coordinator and group liason, interest in genealogy: Contact :
Mrs Valerie Wheater
e.mail: valerie@wheater7.freeserve.co.uk Please tell others about the WDSG-UK

Dr John Walshe - honorary president, medical expert and world authority on Wilson's Disease
Dr Godfrey Gillett - group adviser
Dr Alan Stevens - group adviser
Prof. Jim Lowe - web pages (http://www.wilsons-disease.org.uk)

The next WDSG-UK Event

Due to popular demand and the previous success of our first meeting/event we have organised another similar event which is scheduled for Saturday November 25th.

This event will be held at the staff recreation club at Nottingham University. (Buffet/ drinks and the opportunity to meet others, chat and make friends/contacts)

We are very pleased to let you know that Dr John Walshe and Dr Godfrey Gillett will be joining us for this event.

We hope many of you will be able to come !!!

If you would like further details regarding this event, please send an email to Caroline and ask for details.

LIVING A HEALTHY LIFE...SELF-MANAGEMENT COURSE

This course is available to people with long term health conditions who want to make changes to their lives.

One of the most important things that you learn on the course is because of similarities among long-term illnesses, the tasks and skills you need to live with different illnesses are similarÑwhether you have liver disease, arthritis, heart disease, asthma, multiple sclerosis, fibromyalgia, diabetes or any other disease.

Perhaps the most important skill of all is learning to respond to your illness on a day to day basis and try to solve problems as they arise.

The course offers you the opportunity to learn skills to cope with your illness, to help you continue your daily activities and to help you manage any changing emotions brought about by your condition.

You decide what you want to accomplish. The course enables you to do so at your own pace with the support of other people who are facing similar problems.

The course is about learning how to:

- Take care of your condition Ñ such as taking your medication, exercising, changing your diet
- Carry out your normal daily activities, for example household chores, work and social life
- Manage your emotional changes Ñ changes brought about by your illness, such as anger, uncertainty, depression, anxiety, including changes in your relationships with family and friends.
- Manage your symptoms, whatever they are
- Deal with pain and tiredness

- Relax, eat healthily and exercise gently
- Communicate more closely with people involved in your care
- Plan for the future

The course has been developed by an experienced team at Stanford University in America. It is based on over 20 years of research and development in patient education.

A course runs for 2 1/2 hours per week for 6 weeks and there is a small charge of £2 per session.

The course is facilitated by two lay leaders, both of whom have experience of living with chronic illness. They have attended a Living a Healthy Life Course and then completed a further training programme, and have continuous support and supervision.

For more information please contact

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