Wilson's Disease Association - UK , Volume 2, Issue 1
March 2001

WELCOME

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.

A date for your diary!

The next WDSG-UK all day event/with buffet is on:
Saturday 8th September, 2001
Nottingham University Staff Club
11.30-3.30pm

See end of the newsletter for a booking form…

Wilson's Disease Support Group UK

Putting you in touch with each other…
Many of you have expressed the desire to be in contact with other people affected by WD. Please let us know your contact details that you want adding to our list, 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 at the address given. We have arranged our third meeting again in Nottingham. The date for this event will be Saturday 8th September , Nottingham University Staff Club1, 11.30—3.30 , buffet lunch by arrangement. Please return the booking form (see later) If you would like to attend . Further details of how to get to the venue and details of the 'fun programme' will be posted to you on request.

NOVEMBER 25th 2000 Nottingham - A great day was had by all

Though it was a wet day I think all who came found it a wholly good day. We had a great turnout (50 plus to count) and again it was nice to bring people together and give everyone the opportunity to meet others, see old friends, meet new friends and have a really good day (some fun too!).

The day itself began at 11.30am , with tea, coffee on arrival. I am told the food, was superb (especially the sausage plait!!!) even though I didn't get time to eat anything, I was enjoying meeting and chatting to people so much that I plain forgot!!!

We sincerely hope to see many of you at the next event, booked for September 8th at Nottingham University Staff Club. If you would like to come, (£5 per head including buffet!) please fill in the cut off form at the back of this newsletter and return with your name and address to Caroline (at the address given on the form).

More Info. on Water Filters…

Following the article on Brita Water Filters, Volume 1 issue 2 Sept 2000, Diane Dixon (the original inventor of the wooly mammoth) has kindly sent us additional info. regarding ‘‘ under the sink’’ water filtration systems which are available.
She has spoken in person to the company Clearwater Systems Northwest, who are based in the U.S.A. where Diana now lives. The company have said they will willingly ship products to the U.K if anyone desires them.

CLEAN WATER
Systems Northwest
16305 64th Street East, Sumner, WA 98390
Phone (USA) 800 525 7444 Fax (USA) 253 863 3408
e.mail pacrim@frugal.com

All KDF cartridges may be used for lead (Pb) and heavy metals (this includes copper).
Diana has also written an interesting story about her own experience, which shows how these systems can prove extremely useful in certain situations.

Description	Code	Use	Function of Cartridge	Approx Capacity	Cost
lead/ iron	CRC	2	lead and heavy metal remover	2500 ppm	$25.00
GAC+KDF	KCD+1	2	Premium GAC with 1-0lbs of KDF	10,000 gallons	$26.85

What’s in a cup of Coffee? by Diana Dixon

Seven Years ago I had a setback; all my symptoms returned. After weeks of testing, and adjusting to penicillamine and getting nowhere, I decided to do some detective work of my own. We had already got test results from the water department which were fine, However I drink a few cups of instant coffee, water straight from the tap, first thing in the morning. I took a sample of that water to be tested. The copper level was very high. Over the years, drinking those cups of coffee, with the water that had sat overnight and leached copper from the pipes, the copper level in my body had built up and I was almost back to square one. I called water treatment places that I had found in the yellow pages of the phone book. I had to call quite a few before I found one who seemed to know what I was trying to ask for; I wasn't too sure!

Clean Water Systems Northwest. I now have their filter installed on my sink. The filters are under the sink, the tap is mounted anywhere you like on the sink. It is easy to install, my husband did it!
I use the water from this tap for drinking and cooking Almost immediatley after getting the filter installed my copper levels went down, and I got back to normal (for me). I change the filters once a year. I called them yesterday and told them about your organisation, and your search for de-coppered water. The filter was designed, and is made exclusively for them. They are a small family type business, with an excellent reputation . If anyone is interested, please contact Caroline who can arrange getting the product shipped from the U.S.A, or alternatively fax, e mail or phone them directly at the given company details above

Wilson's Disease before diagnosis

by Maureen Adams

Up to the age of 21 I had led a relativley normal life apart from nosebleeds and irregular periods.Early in 1971 I began to swelling in my ankles and gradually over the weeks the swelling was getting worse untill the bottom two thirds of my body was affected. I was put on diuretics but these were not strong enough to have much effect.

By March of 1971 I was feeling very tired and depressed and pressure from the fluid build up was giving me backache and making it very diffuclt to breathe. In March I went into our local hospital, for a month, where numerous tests were done. Fluid was drained off my stomach,which made me feel abit better as the pressure was now off my lungs and breathing was much easier. The doctors said I had cirrhosis of the liver but couldn't find out why so they referred me to Addenbrooke's in Cambridge. I came out the hospital 2 stone lighter in weight than when I went in.I went into Addenbrooke's in June where more tests were done. A liver biopsy was taken.Although the doctors knew I was very ill ,they still couldn't make a diagnosis, so they referred me to London.

In October I went to the Royal Free Hospital in London where more tests were done and after afew days a diagnosis was made at last.I was then started on Penicillamine,lots of other different drugs and put on a high protien/low sodium/ copper free diet. After awhile I was getting back to normal and fit enough to go back to work.

I feel very fortunate that I had anormal childhood before getting ill and that when diagnosis was made,treatment worked for me.I can never remember feeling very ill and think it was a much more worrying time for my family.I always felt that whenever I was in hospital the other patients were a lot more ill than I was.

Had it not for the work and dedication of Dr Walshe,treatment wouldn't have been possible and I probably wouldn't be here today so a massive thank you to him. Had I seen Dr Walshe my treatment would have started 3 months earlier.
May he have a long and happy retirement.

The Green Man Stirs

poem by Steve Plowright

The Green Man stirs…
Through these wintery days of February,
Grey damp days, drops dripping
From barren boughs overhanging
Swollen streams fed by February rain.

There's something afoot in the tracery
Of leafless twig and twisted limb,
A spark to fire the fuddled brain,
To loose the passion, ease the pain,
A hint, a glance, a feeling
To set the senses reeling,
To stir the early draughts of March
And hasten on the Spring.

Look to the ground and see the earth's rebirth,
Behold the branch and note the brown bud swelling.
See the sap of Spring arriving,
Around each bush and briar entwining.
The energy is waiting there
To make Old Winter disappear,
Nuturing the new, quickening the change,
Dispelling darkness, doubt and fear.
Tis' time for Nature's healing.
The season's daily turning -
Can't you feel the Green Man stirring?

Joining forces with other WD groups in Europe
European Community funding

THE PROGRAMME OF COMMUNITY ACTION ON RARE DISEASES has been established by the European Commission and has been adopted for the period of 1st Jan 1999 to 31st December 2003

The aim of this progrramme is too contribute, in order with other community measures, towards ensuring a high level of health protection in relation to rare diseases. Specific attention is given to improving knowledge and to facilitate access to information about rare diseases. Wilson Disease falls within the category of being a rare disease and hence we are currently working with other WD groups throughout Europe to make an application for funding to help to facilitate the project goals.

Future generations could benefit from such funding and projects, better awareness , genetic screening programs, training, research etc…

As you may well be aware there are active WD groups in other country states within the EC. These countries include :

- GERMANY
- DENMARK
- AUSTRIA
- HOLLAND
- SPAIN

these are just ones we know of , there may be others.

By collaboration of group efforts towards common goals which will help promote a greater awareness and diagnosis of Wilson Disease.

We feel WD groups in Europe (and Worldwide!) are already making a difference, but… we want to make an even bigger difference. The awareness campaign needs to widen and become stronger for the future.

If we are successful in our application for monies this would be of immense benefit to the European WD community (and future generations who may be born with the condition)

ON THE NET..........

We have set up a web page for WDSG - UK

http://www.wilsons-disease.org.uk

there’s a mailing list attached so please sign up

Questions and Answers

Edited by Caroline Simms

Q1?/ Is it safe for someonw with WD to wear copper braclets for arthritus relief? (sent in by Jane Ridley - Peterborough)

A1/ No, this is not recommended!
Copper can be absorbed in small amounts through the skin from copper jewellery. As it is sensible to avoid excess intake of copper then it might be cautious to avoid wearing any copper jewellery which has direct contact with the skin.

Q2/ Is there a type of dark chocolate which is low in copper content? (sent in by Brian Waters)

A2/ No, unfortunately all dark chololate is quite high in copper content. Carob is supposed to be a chocolate substitute but personally I feel it tastes nothing like the real thing (i.e. Lindt's (Swiss) 70% cocoa !). A little occasionally probably won't hurt as long as you take your meds on a regular daily basis (just don't eat it a block at a time!).

Q3/ If you dug a hole to Austrailia, would you come out head first or feet first ?

A3/ We really don't know the answer to this but we have decided to give a prize to the best answer to be sent in (to be printed in the next issue!)

Do you have any specific questions regarding any aspects of Wilson's Disease which you would like us to answer or any aspects or information you would like us to find out about?

You may have a question which others might want answered so go on. Ask us and if we don't know we will do our very best to find out for you!!! More of your questions and answers will be included in the next issue of WDSG-UK.

The Story of Trientine - by John Walshe

As with the story of penicillamine in the previous issue of the News Letter it is not exactly clear when this story began.

The story certainly began with Robert. Robert had been diagnosed with presymptomatic Wilson Disease at the age of 6 following the conformation of the diagnosis in his elder sister. There was a reluctance on the part of local doctors to start prphylactic treatment at that age was delayed until he was 9 years old in 1961. All went well until 1967 when he developed severe kidney damage with swelling of the legs and abdomen from fluid retention.

This was the first serious side effect of penicillamine I had seen in 10 years. Attempts to desensitise Robert to penicillamine failed and the other drugs available at the time proved inefficient in getting out copper. So at that stage a search was started for an alternative compound able to mobilise and excrete excess body copper. Was this the start of the story when Robert first got into dificulties or was it a year later when the first trial of Trientine was started? I leave you to choose.

A year may seem a long time to have gone without any effective treatment but find a new drug and ascertain if it is safe is no easy task and only 16 months is remarkably quick—impossible now in the days of ethical committees, the committee of safety of medicine and the general public antipathy to any new venture by a doctor from the safe path of convention.

During that year many new compounds were tried, on rats. The animal rights activists would not approve of this either ! Nothing Kay Gibbs, my invaluable assistant, and I tried looked opeful until one of my colleagues in the University Department of Biochemistry in Cambridge suggested we look at triethylene tetramine , an oily corrosive liquid used in industry as an epoxy resin hardener ! It bore no chemical resemblance to penicillamine but had a structural formula which he predicted should bind copper. Before this could be proven, even in a rat, it had to be neutralised with a strong mineral acid, hydrochloric acid proved to be the best, and freeze dried. This we did and then administered to some rats. To our delight it caused a significant increase in copper in the urine without upsetting the rats. One problem which we had not yet overcome was the commercially available triethyene tetramine was not very pure. However as the impurities did not seem to harm the rats so it appeared safe to give Robert a trial dose and see what it did to his copper excretion. The trial can be described as nothing but a spectacular success. Copper simply poured out into his urine, even more than was mobilised by penicillamine. So August 1968 was a red letter day in the management of Robert's illness and has consequently proved to be the case, that of many other patients with Wilson Disease. However , as per usual, all was not straight forward .

The triethylene tetramine, or TETA as we called it for short, had to be prepared by Kay Gibbs in my laboratory, packed into capsules by hand and posted to Robert regularly. After a while other patients appeared from all over the country needing our new treatment and the physical load on our resources became a real problem, routenly cutting down on research efforts.

In 1974 our source of crude TETA ran out and we had to look around for another source. The new supply was our undoing: shortly after taking it into use two new patients arrived needing this drug and both rapidly developed severe kidney damage. Treatment was at once stopped and all the patients on our postal register were told to discontinue their treatment until we could identify and remove the impurity which was responsible. The best organic chemists in Cambridge could not find any difference between the new and old source of TETA so we never solved this problem.

But my collegue in Biochemistry, Dr Hal Dixon, came to our rescue and devised a method of purifying the crude freeze dried TETA by crystallising it from pure alcohol. After this we had no further problems, except for that of making enough of the drug for all the patients who were accumilating on our list. The then Addenbrooke's Hospital pharmacy came to our aid and took over production whilst I tried to find a commercial source of production. By this time I had also started negotiations with the Department of Health for a product licence from the Committee of Safety of Medicine. This was like fighting with cotton wool in the dark or playing pass the parcel. No one wanted to be responsible for saying YES, very risky, but equally nobody wanted to be responsible for saying NO, equally risky !!!

While all this bureaucracy was going on I tried to interest one of the Pharmaceutical companies to make TETA for us. However, as you may well guess, none were interested, but… A chemical company, called Adrich Chemicals, agreed to have a look and eventually decided that this was a project that was worth while I am extremely grateful to Aldrich Chemicals, as they then were, for all their subsequent help. Finally, triumph, the Ministry of Health decided to bite the bullet and issue a Product Licence for the new drug, with restrictions that it could only be given to named patients through a hospital pharmacy. That was way back in 1984, fifteen years after my initial tentative studies. What is more this just beat the U.S. Food and Drug Administration (FDA) by a short head. The initial prescribing restrictions have only recently been rescinded.

One interesting sideline. In the early animals studies, when we were still using the crude original product, we tried giving TETA to a rat by injection. The rat collapsed immediately after injection and microscopic examination of its tissues suggested it had died of shock. Looking at the known impurities in the TETA, the main one was triamino triethylamine, closely related to triethylene tetramine. This looked to me like a compound that could cause an immediate fall in blood pressure and this might well explain the collapse of the rat. One of my pharmacological colleagues investigated this for me and found my hypothesis was correct. Here I thought was a goldmine, a new treatment for high blood pressure - fame and fortune at last. A drug firm was interested and took out a provisional patent but this was just the time when a whole host of new blood pressure drugs came onto the market and my goldmine proved to be a mirage.

The interesting lesson of this story, as with that of penicillamine, is that two new drugs for treatment of Wilson Disease have been introduced to the market, having been shepherded through all the bureaucratic baffles, without the help of the Giant Multinational Pharmaceutical companies.

I doubt very much if this will be possible again

A Funny Day Out - by Helen

One fine September day I was fed up sitting in the house, so I decided to put on my good clothes and go out . So I donned my lovely long dress; where to go I thought , maybe Long Eaton. So off I went to the railway station; good the train for Long Eaton was in,so I boarded it. The train was quite full up, I couldn't get a window seat , but I got one next to the corridor,as I sat down a young man of about 28 with a pony tail said good morning; to me. I thought how nice and polite as the majority of folk don't say good morning. So I sat and laughed into myself at this and felt happy , though not for long !

In the distance the lady was pushing the refreshment trolley, as she passed by me I felt my dress being pulled and tugged , really tight too. Flashing through my mind, I thought what the dickens is happening! To my horror I realized, quick as a flash I shouted to her to stop, but she either did not hear me or understand my accent, she kept on going till she could go no further. My best dress had got caught in the trolley wheel as she had passed by! One man had seen what was happening and he shouted at her to stop.

Thank goodness no one laughed! I didn't know whether to laugh or cry, I felt so embarresed, I was so afraid to look at my dress. The refreshmrnt lady apologised. My beautiful dress had been ripped at the bottom. The refreshment lady remarked that this happens a lot, the trolley wheels catch onto things (so beware when on trains !) . Gosh it was like something from candid camera. Just think if I had been wearing a dress of thinner material or a skirt, the trolley wheel could have ripped it off and I could have been left sitting there in my bra and knickers!

I wrote to British Rail in Derby but never got compensation, as they promised.

WOOLY MAMMOTH WDSG-UK T-shirts available to order

For those of you who met Keith Pereria, the man with the Wooly Mammoth T-Shirt ; this was a really great idea for the group! Thanks to Keith for this.

If anyone wants to order T-shirts (see T-shirt design) please send your orders and payment in advance to WDSG-UK . The T-shirts will be available to collect at the September event and a limited number only to purchase on the day on a first come first serve basis. (Cheques or postal orders made out to WDSG-UK). T-shirts are available in size Medium or Large at £10 each (pounds sterling) and we can arrange postage and packaging if needed (overseas also).

My Skirmishes with “ MAVIS BEACON ”

By Linda Hart

I was hoping to be able to write about notifiable diseases and why WD isn’t one, but unfortunately as yet have been unable to find out any useful information . I have written to Nottingham Health Authority , who informed me that the Department of Health do collect data on notifiable diseases, but WD is not included !! They tell me that “the incidence of WD in Europe is one in 300,000” which would suggest to them that the incidence in Nottinghamshire “is statistically insignificant!” - not if your me it isn’t! I suppose the next step is to contact the Health Minister and ask why data on WD isn’t collected! We’ll keep you informed.

On a different note, this year I have been dragged screaming into the computer age. I have avoided computers like the plague—I lived in the 70’s—I liked it there! I was very happy there and I didn’t want to know about hard drives, floppy disks or software. Even if my nieces and nephews spoke in a language that I didn’t understand, I didn’t care, I just ignored it all! But, since the Support Group has started, it’s become obvious I needed to learn how to use a computer, or at very least, how to type! So my education began.

The first two weeks I spent mastering the art of making paper aeroplanes and playing Pac Man. This is easy I thought, then I thought I’d try letter writing—this was a different ‘kettle of fish’. My first effort, a letter to my friend Jane, took almost an hour and when I eventually got the printer to function and read my effort, I realised I had to learn to type properly. So along came the “Mavis Beacon Typing Course”. Mavis is a dragon. I think of her as a cross between Medusa and Mrs. Merton! Up to now, I haven’t yet learnt two rows of keys and it has been painful! Mavis puts little notes on the screen to tell me I’m not trying or my standards are dropping. If I dare to press the ‘escape key’, she says “Tut tut you’ll never learn that way…”, even if I’ve been sitting there for two hours! I have come across a game on her programme which I’m getting to like. It looks as though you are in the driving seat of a car looking through the windscreen. Words pass in front and you have to copy them. Every time you make a mistake an insect ’splatters’ on the screen. (I get more engrossed in splattering insects than typing, which isn’t quite the point of the game!) But, I shall persevere with my typing and hopefully this time next year I shall be an old hand at it.

Just one last word , if anyone gets a letter from me, I’ll warn you in advance there won’t be any Z’s, C’s, V’s, B’s or punctuation in it, can’t do them yet!

My New Friend!

By BigAl

Who am I? Not important! I've recently met someone with WD and, not knowing a lot about the disease (see Lynda's comments on page 2—I'm not the only one!) I am amazed at the neglect and lack of support that is (or isn't) available to those with WD.

These "Newsletters" should be circulated much more widely, but obviously cost is a problem—"copy 'em" and "give 'em out" to your friends, in the pub or wherever you get to! (nothing personal here!), I think it's called initiative?

Anyway, back to the script. My new friend has the most positive attitude that I've experienced and is determined to enjoy life to the full—a lesson to us all! We enjoy the odd walk together, which usually causes me to 'perspire' (that's how steeplejacks get paid?) but it's good fun and one of the many things in life to be enjoyed. And before I close, remember the famous definition:- BOOTY is in the eye of the deceiver!

Cheers

Some New Words for the 21st Century…….

- Blamestorming: Sitting around in a group, discussing why a project failed, and who was responsible.
- Uninstalled: Being fired.
- Umfriend: A sexual relation of dubious standing, or a concealed intimate relationship. For example: "This is Alan, my…...um…...friend".

and what about this:- If at first you don't succeed, skydiving is not for you!

More in the next Newsletter!

Coming next…..

We intend to keep you updated not just via the Newsletters so expect the occasional "newsflash" in between work, play, trying to have a holiday and coordinating the Support Group activities! These may well arrive strapped to a pigeon's leg (especially if we can't afford the stamps!)

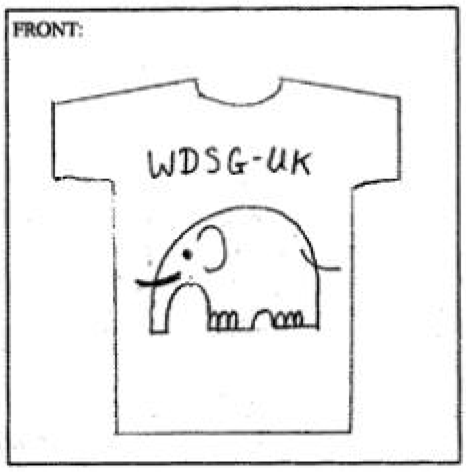
WOOLY MAMMOTH T-SHIRTS AVAILABLE TO ORDER !
WDSG-UG

Wooly Mammoth T-Shirts are now available to order for the price of £10 each

T-Shirts are White with Black Print
These will be available in Sizes Medium and Large

Full Payment to WDSG-UK must be sent with the order (as we have to prepay for the T-Shirts in advance!).

Please remember to let us know your name and full postal address and if you wish to have items posted to you! And here's what the design looks like……..



T-Shirt order form

Name…………………………….

Address…………………………………..……………….

…………………………………………………………….

I wish to order ……….. Wooly Mammoth T-shirt/s
I enclose full payment of £………….. for these items

Tick as Appropriate
I wish to receive the items by post ………
I shall collect T-shirts at WDSG-UK event in September 2001………

RETURN FORM FOR WDSG-UK EVENT

Name………………………………
Address……………………………………………………………………………
……………………………………………………………………………………
I/we would like to come to the WDSG-UK meeting on Saturday 8th September at Nottingham University Staff Club

I / we would like to book ……. seats for the meeting with other people for a day out in Nottingham (a central and relatively accessable location)

I/we require information regarding overnight accomodation
Yes / No

I/we would like details of how to get to the venue Yes / No

I/we shall be travelling by …………………………………………………....
……………………………………………………………………………………

I would like to order ……..WDSG-UK Wooly Mammoth T-Shirts
Size: Medium ……… Large ……….(state number of each!)
I enclose full payment of £…….. For the T-shirts ordered
(T-Shirts are £10 each) see p13 for design!

Please return this page along with your completed membership form/ or renewal form and dues to:

Dr Caroline Simms (Membership Coordinater)
Wilson's Disease Support Group UK
36, Sunningdale Drive, Woodborough,
Nottingham. NG14 6EQ.

Would you like to write an article to be included in the next issue of the WDSG -UK newsletter?

We would appreciate your feedback on this Newsletter.
It is you that counts and together we can help each other !

Please send your articles, views, opinions, experiences letting us know if you would like them included in the WDSG - UK Newsletter to:

Dr Caroline Simms36, Sunningdale Drive, Woodborough,
Nottingham. NG14 6EQ.
e.mail : carolinesimms @hotmail.com

Tell others about the WDSG-UK

Please tell others who you may know have WD, who might benefit from the Support Group and what we are doing.

Inform your family, friends, consultant physicians, GP surgery, local MP's about WDSG-UK

The more people who know about us , the more we can promote a better awareness of Wilson's Disease within the community and the better the chance of early diagnosis of WD for future WD patients.

Wilsons Disease Support Group UK

If more copies of this newsletter are required, please contact:
Dr Caroline Simms
36, Sunningdale Drive, Woodborough,
Nottingham. NG14 6EQ.
e.mail : carolinesimms @ hotmail.com

For membership, correspondance, and information
Contact:
Dr Caroline Simms (Group Secretary and coordinator of newsletter production)
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
Contact:
Linda Hart (Patient and family corresspondance coordinator)
36, Audley Drive,
Lenton Abbey, Beeston,
Nottingham. NG9 2SF

or

Mrs Valerie Wheater (Coordinator and group liason, interest in genealogy)
38, Grantchester Road,
Cambridge. CB3 9ED.

Dr John Walshe - Honorary President, medical expert and world authority on Wilson’s Disease
Dr Godfrey Gillett - Group advisor
Dr Alan Stevens - Group advisor
Professor Jim Lowe - Webperson