Wilsons Disease Support Group UK

For membership, correspondance, and information

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Dr John Walshe - honoury president, medical expert and world authority on Wilson's Disease

Dr Godfrey Gillett - group advisor
Dr Alan Stevens - group advisor
Professor Jim Lowe - webmaster

Wilsons Disease Support Group UK

W D S G U K

Tell others about the WDSG-UK

Please tell others who you may know who 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.

If more copies of this newsletter are required, please contact:
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Visit our website at http://www.wilsons-disease.org.uk

Wilson's Disease Support Group – UK

- WDSG—UK applies to become a registered charity
- Tales from Leipzig, Germany
- Caroline Barr shares her story
- Scientific Research—comments from the 8th International Conference
- Notes on health supplements

Inside this issue:

Application for Charity Status...

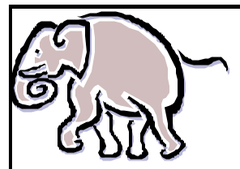
Now WDSG-UK has been established for nearly two years, the organisers (after much discussion) have decided to make application to the Charities Commission to become a registered charity.

After various meetings and discussions about the pros and cons of registering, we have decided that in the interests of the group's progress for the future that this is perhaps the best way forward.

If WDSG-UK is granted official charity status, we believe that the group could benefit immensely, gaining more credibility. Charity status would also enable us to apply to various funding bodies with the hope of generating significant monies to further the longer term aims of WDSG-UK, promoting awareness of WD within the community and throughout the medical profession, supporting academic research and expertise in management of WD.

The official governing document was drafted and signed by the four trustees (and witnessed by one of our group advisors) at our 3rd Meeting which was held in Nottingham on September 8th. After this has been returned, it will be sent with the application form to the Charities Commission for their consideration.

We are hopeful that we will be granted Charity Status and will of course keep you posted on this.



WDSG-UK

Linda Hart gets stuck in to a few college courses

written by Caroline Simms

After Linda's success in teaching herself to type (see Vol. 2, Issue 1 of this newsletter, Mavis Beacon), I am delighted to say that she is going to classes at night school to do a bookkeeping course and a computer course. She is finding the bookkeeping somewhat challenging, but then again the lady she sat next to at the first class whizzed through the questions which the tutor set (she later discovered that the lady worked as a bookkeeper for a firm in Nottingham so its hardly surprising that Linda may have felt a little left behind). Needless to say "Never compare yourself to others, as we are all unique!". John, Linda's partner, is as usual very encouraging which is great to see. And I, too, am confident that she will do well as she is a very determined person.

8th International Conference on Wilson Disease, Leipzig, April 16th—18th , 2001

Written by Ursula Mott

Having been invited to the International Conference in Leipzig I was delighted to accept. It presented an opportunity to update myself on the current scientific position regarding W.D. and I was also able to meet the experts and representatives from other W.D. support groups (Caroline and Linda were very good at networking!).

As W.D. " sufferers " we were warmly welcomed and before the conference officially began we had a special invitation to meet the professors at lunch. We spoke with Diane Cox, Frieder Berr, Irmin Sternlieb, Peter Ferenci, Hans Kuhn, Herbert Scheinberg and others. It was good to see Drs. Walshe and Gillett (known familiar faces) attending the conference and

I was glad that Dr Dooley (Royal Free Hospital where I attend clinic) was also present. I found the amount of debate and discussion taking place at sessions refreshing and reassuring.

Dr. Kazic from Yugoslavia pointed out the benefit of having more than one effective treatment for W.D. (during the conflict in his country it was impossible to get penicillamine, but zinc was obtainable which was used temporarily and with success).

Our evenings were spent mixing socially and enjoying historical Leipzig. On one occasion, after a reception at the Bach museum and an organ recital, we were having supper at the Paulanus Restaurant where Dr Hefter (an eminent neurologist from Dusselldorf) could hardly believe his luck when he ended up with 5 crayfish (very high in copper!) on his dinner plate which we had offloaded on him.

Not only were we (as patients) warmly received by the attending doctors, academics, and professors, but it was publicly stated that it was good to have W.D. patients at the conference. To meet W.D. experts from all over the world and to interact with them and their respective spouses socially, having constructive dialogue, making friends, forging links worldwide and also having fun made the trip to Leipzig a memorable experience for me.

Left to right: Dr Rupert Purchase, Ursula , Stefan Sandler, Linda, Simon, Marie, and Caroline

QUESTIONS AND ANSWERS :

edited by Caroline Simms

(please keep sending your questions to us so we can print them in the next issue WDSG - UK !!!)

Q1 Is WD an inherited disorder (i.e. is it passed on by reproduction)?

A1 Yes, for people to have WD they must inherit two WD genes (they inherit one from the mother and one from the father). In theory, if WD is in a family then the genes can stay in the gene pool for generations and the disease may reappear in a later generation.

Q2 How many people in the general population carry the Wilson Disease gene?

A2 The current estimate for the number of people who carry the WD gene in the general population is 1 person in every 90 people.

Q3 I have just been diagnosed with WD. Should the rest of my family be tested?

A3 This is definitely advisable! It is recommended that all other siblings are tested, cousins, aunts and uncles. If early and correct diagnosis of WD is made, before symptoms arise and a patient becomes seriously ill, the chances of irreversible damage to health occurring are much lower. WD is a very treatable condition if diagnosed early enough.

HERE'S A LITTLE SOMETHING TO MAKE YOU CHUCKLE…..

These , it is claimed , are genuine medical notes taken from hospital charts :

- Patient has chest pain if she lies on her side for over a year.
- On the second day the knee was better, on the third it had gone.
- The patient has been depressed since she began seeing me in 1993.
- Patient was alert and unresponsive.
- Rectal examination revealed normal sized thyroid.
- The patient refused autopsy.
- Patient has left white blood cells at another hospital.
- I saw your patient today who is still under our car for physiotherapy.

NOTES ON VITAMIN AND HEALTH SUPPLEMENTS

INTERESTING REPORTS ON VITAMIN E
...may be beneficial in Liver Disease
We are currently trying to find out about the potential applications of Vitamin E in liver recovery and WD.
We hope to include an article on this subject in the next issue!

CAUTIONARY NOTES - **natural supplements**

MILK THISTLE (Silymarin)
Milk thistle tincture is ***NOT SUITABLE*** for those with **chronic liver problems such as hepatitis or cirrhosis**

Vitamins and Minerals Handbook, by A. Ursell, B.Sc. Nutrition, DIP Dietetics. Published by Dorling Kindersley Ltd, London, 2001.

8TH INTERNATIONAL CONFERENCE ON WILSON DISEASE AND MENKES DISEASE LEIPZIG, *APRIL, 2001*

THE FOLLOWING AREAS OF RESEARCH WERE DISCUSSED AT THE CONFERENCE SESSIONS

- Regulation and function of genes in copper homeostasis - regulation and function of eukaryotic copper transporters, transcriptional control of genes by copper and other heavy metals……..
- Intracellular Processing of copper and copper transport proteins / genes - location and functions of ATP7B in hepatocytes , induction of metalothionenes for copper toxicity; protection and causation, copper chaperones……..
- Experimental Models and therapy - experimental approach to gene therapy for WD, prospects of gene repair for monogenic diseases, hepatocyte transplantation for WD, animal models for new therapeutic strategies
- Menkes disease and other disorders of copper metabolism
- Neurodegenerative processes in WD - copper proteins and metabolism, cerebral metal deposition in liver disease, modern tools to assess brain function, particularly interesting was a presentation on different patterns of brain glucose consumption in WD
- Epidemiology of ATP7B mutations and phenotype of WD - prevalence of WD throughout the world, frequency of WD in different populations, some examples of different gene mutations in WD were discussed and some correlations between common mutations were made, type of disease, mean age of onset were mentioned
- Clinical manifestations of WD - some patient case histories were discussed, presymptomatic cases were mentioned
- Clinical management of WD - the different drugs used to treat WD were discussed

If anyone wishes to read the scientific poster abstracts from the conference, please send a stamped self addressed A4 sized envelope to Caroline at WDSG - UK. We are trying to find out if the conference proceedings have been published and we will let you know in the next edition of the newsletter.

Though the conference was a scientific meeting, we as patients were welcomed by most. As the chairperson and group coordinator of WDSG - UK , the opportunity to attend a meeting like this was golden. As well as getting to meet and talk to many of the pioneering names in WD research and disease management, it gave me the opportunity to do some networking with other WD groups / associations and to represent and promote our group. Caroline

A Group Photo of the Participants Attending the 8th International Conference on W.D.

(photograph taken outside the Leipzig Opera House)

Caroline's Story

By Caroline Barr

I have had Wilson's Disease symptoms since I was thirteen, probably a bit earlier than that. My first symptoms were pins and needles in my hands and legs, and a problem controlling my saliva. After a painful lumbar puncture, numerous blood tests and a psychiatrist's report , I was diagnosed as having school phobia. I couldn't walk straight and was dribbling better than a one year old.

By then, because my life was becoming increasingly more difficult, I thought I was going mad. The psychiatrist that I saw based his whole diagnosis on the answer to one question, which was what do you think of school? He gave me three answers:

A/ Do you hate it

B/ Do you love it

C/ or Is it a necessary evil.

I picked the answer that I thought my sane friends would give, which was that it is a necessary evil. I loved school. I knew that the diagnosis was partly my own fault. But for a psychiatrist to base a diagnosis of a child on one answer is outrageous.

The psychiatrist that I saw based his whole diagnosis on the answer to one question, which was what do you think of school?

Before I could blink, I was transferred to the Victoria Hospital in Kirkcaldy to Stratheden Hospital where I found myself in a young persons unit. They didn't inform my parents, I got my money taken away from me and despite repeated requests I was not allowed to phone my family. I remember feeling very scared—I was only thirteen at this time. When Mum found out where I was she came and got me thank goodness!

Mum has a relative who was a doctor in Edinburgh. Through him I was admitted to Sick Kids Edinburgh, where I was diagnosed as having Motor Neurone Disease. But before I left, Dr John Livingston took a 24 hour urine sample and sent it off to the Philadelphia institute. I was most upset that I couldn't deliver it in person!

By the time the Wilson's Disease finally came through on the 5th February 1981, I couldn't walk or speak and I was living on Coca-Cola ice-cubes. I was admitted to Sick Kids the next day. I was put on penicillamine. My white cell count dropped very low. I was taken off it, it went up. By this time I was being fed by a nasal gastric tube, bed-bound and using

an alphabet board to communicate— which was made worse by the fact that I can't spell!

After about three months of me on and off treatment and not getting any better, Mum took the decision, against all the doctor's wishes, to take me to Addenbrooks Hospital, Cambridge, and Dr Walshe. My treatment since then has been Trientine. It was there I met Linda Hart. She has been an important friend to me to me ever since. I was no longer the only person with this condition.

After I got better enough to cope with some education, I got home tuition for two years. Meanwhile my peer group all moved on without me. So I had very little daily contact with people my own age. Even now I get on better with people who are a bit older than myself. I have had a few friends who tried to maintain contact but communication was still hard. School friends move on, have boyfriends, go to university, get married, have kids and they forget.

After home tuition I went to Glenrothes Technical College Special Needs department, which didn't really teach me anything, apart from socialising with my peer group, who all had learning difficulties. I was still using a wheelchair at this point. I did pass the Anatomy, Physiology and Health exam, but I had to do that through flexible learning though I managed to negotiate some tutor support.

I was eighteen walking with an awkward gait after a tendon lengthening operation. Communication at this point was and still is difficult, but I can now eat everything, including things that I'm not really supposed to!!

After three months at Queen Elizabeth Training College Leatherhead which was my first time away from home, I discovered the food was awful and the courses were inappropriate for my needs. I was also terribly homesick but I stuck it out, as it was another one of these experiences I had to go through. It also made leaving home for good easier.

From then on the hunt for a residential college was underway. We found the National Star Centre for Disabled Youth, in Cheltenham. I was 21 by this time. There I was taught basic computer skills and

CROCODILES

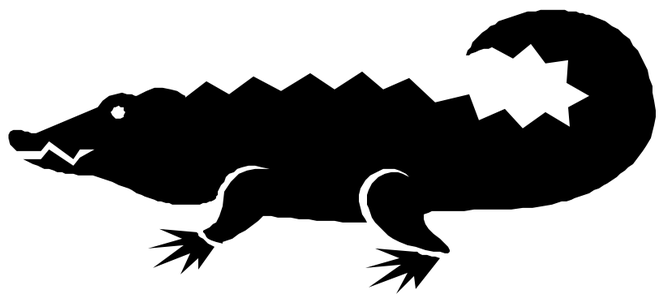
By Hans

Some people say -
" Beware the smiles upon the face of Crocodiles! "

But how they judge I cannot tell,
Until they really know them well.

Of course all Crocodiles have teeth,
And they are scaly, hard beneath.

But those whose features cause alarm,
May hide a nature full of charm!



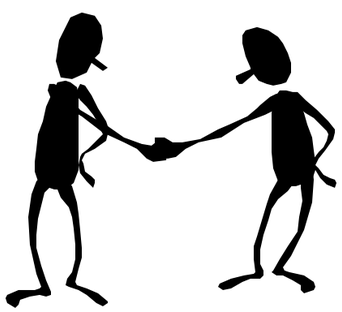
The 3rd Meeting of WDSG - UK Saturday 8th September, 2001

The venue, Nottingham University Staff Club, the weather , sunny and bright, the people (again lots of new faces!) who came made the day's event a great success and we only hope that everyone at least gained something from coming along to the event. We had in attendance about 50 people and it was great to meet you all. Like previous events we have held, it was a very enjoyable day.

People arrived from 11am onwards, and over the next hour and a half people chatted to each other over tea/ coffee. Dr Alan Stevens gave a short welcoming speech and summarised what would be in store for the rest of the day. Lunch was a little disappointing, too much spicy food (next time we shall ensure that the buffet includes some edible food!).
Dr Rupert Purchase gave a talk on copper chelators, quite technical but very interesting (an outline of this talk will be printed in the March 2002 newsletter).
The raffle was drawn (next time we shall have just a few superior prizes so it doesn't drag on quite as long) and I think most people ended up winning something. We can assure you that Ursula had not in fact bought up all the raffle tickets, though seven of her tickets were actually drawn as winners, she kindly donated six of these prizes back into the draw. Dr Walshe, Dr Gillett and Dr Purchase were awarded honorary membership of the group for their dedication to the WD community.

WDSG - UK T-shirts sold like hot cakes. We still have some left if people wish to order these (£10 each payable to WDSG - UK).
The events are really geared towards giving you the opportunity to meet others, be they other patients, family members, perhaps to meet the Drs involved with the group, meet old friends from Addenbrookes (those of you who used to go there), for you to ask questions, find out information about what we are striving to achieve from the group perspective. It was all quite informal really and for us as organisers of the group, we thank you for making the effort to come and hope you will be able to come to the next event which will be sometime next September, 2002. We also hope many more of you will perhaps come next year.

Special thanks to Rita Leatherbarrow and John Newman for being a tremendous help and making a major contribution to the days success.
And of course to Linda, Valerie, Ursula



I left college, moved in with my boyfriend, got engaged, then got disengaged, not before having a nasty accident while cooking our tea while he was at work which resulted in bad body burns.

Coming home to Fife, my confidence was low, my relationship had failed, my body was scarred, my sisters had both gone to University and I was sitting at home feeling sorry for myself. So Mum, Dad and myself started looking around for a place for me to live. An old student from the Star Centre was living in supported accommodation in Edinburgh so we looked into it. They thought they could provide an outreach service to me, which is what they did. They provided the physical support as well as emotional support. I

A turning point came when I had been ill longer than I had been well. I thought to myself, well then, this is it. There is going to be no miracles here. I am just going to have to get on with life.

came through some rough times under their care, including the death of my best friend who I had met at the Star Centre and my sister's wedding, which I felt at the time wasn't fair because she was younger than me and these are the things that make you feel a bit left behind.I joined a group called Women Unlimited, which was a woman's health project. This then led me on to writing poetry. I have since then published two books.

I have gone on to unsupported accommodation and now live totally independently in the community. I have done three flexible learning courses– communication 3, introduction to literature, and child development, also various HNC computer certificates and next year I am going to start one day a week in college.

Throughout this condition I have never felt ill. I have felt tired and frustrated, Ay! Have I felt frustrated!!! Tearing up rolls of paper hankies and shedding buckets full of tears for the life I might have had. I have now got some super friends who give me a lot of support and I wouldn't be here if it weren't for my family.
A turning point came when I had been ill longer than I had been well. I thought to myself, well then, this is it. There is going to be no miracle here, I am just going to have to get on with life.
Which is what I have done and plan to continue doing.

I would also like to thank Dr Walshe for saving my life. May you live long and have many hours reading "Fred Basset" to your grandchildren

Despite having been left disabled I am relatively happy with my life. But one thing that does frustrate me is the constant battles to get basic things like ramps on and off pavements and an education equal to my able-bodied peer group..

Caroline performs her poetry at the Edinburgh Festival

Would you like to write a story for a future issue?

A Trip to Germany

Written by Linda Hart

I had not really been abroad before save for the odd day trips to Calais or Boulogne, and the least said about those the better! So I was really looking forward to going to the Wilson's Disease Conference in Leipzig.
I was a bit worried about flying, I'd never really had any desire to fly, quite an unnatural means of transportation if you think about it! So when the plane hit some turbulence, I asked Caroline what was happening, don't worry she replied "if we hit anything it'll be so quick you won't know a thing about it" . Going through customs turned out to be a bit of an ordeal also. I was a bit worried that I would make the alarm sound, as I have a metal plate in my leg. Fortunately that didn't happen, but I must have one of those guilty looking faces because I was frisked on the other side. The full contents of my bag was searched. Horror of horrors, I was smuggling a cheese sandwich into Germany (and this was at the height of the foot and mouth epidemic)! We arrived in Leipzig, after almost getting on the wrong train in Berlin, which would have whisked us off through the night to Budapest. That would have been a good story! We were both so tired we could barely be bothered to talk.
The hotel looked wonderful! It had a pool, a Jacuzzi and sauna, oh heaven! I think the hotel staff wondered what had just arrived, as we sauntered in after taking ten minutes or so to negotiate the revolving doors.
All was going well now, we got our key card, found the room, managed to get in unaided, (key cards were a new experience to us) but getting the lights to work presented another problem. We turned every switch, and nothing- (except I kept getting shocks off everything I touched, much to Caroline's amusement!). Then I saw what I thought was a switch halfway up the wall and stuck my finger in an electric socket, much to my alarm! It was at this point we decided to go to reception to say our lights didn't work,. I shut the door, got a shock from the handle, and pressed the lift button , sparks flew this time, Caroline couldn't believe her eyes ! After this I took to jabbing the lift button with a biro.

Wine just £4.50 per glass

HOW MUCH !!!

When we told the man on reception our problem, he just looked at us with disbelief and informed us that we should put the card into the slot inside the door, and then the lights would work. We tried, they did, we felt very foolish.
While I was at reception I enquired if they provided rubber gloves, as I was getting shocks from everything I touched. I don't know if he misunderstood, or just thought we were slightly unhinged as he just stared at us blankly, until we could contain our laughter no longer. "Oh, I see." he said, "A joke." (the English and the German sense of humour just isn't the same.)
After a very arduous day we sat in the lounge bar and ordered two glasses of white wine, had quite a shock when we got the bill, Caroline and I agreed that this was the most expensive glass of wine we had ever paid for but I think we enjoyed it nevertheless.
The next day we went to explore Leipzig with Stefan, who had also arrived for the conference. It was bitterly cold and sleeting as we walked to the big indoor shopping center at the train station. As it was the Easter weekend, everything was decorated so beautifully. There were large pens with baby rabbits and chicks in them, bare tree branches decorated with lovely painted eggs and floral displays everywhere. So much effort had been made!

The next three days revolved around the conference, which I enjoyed very much, even though I didn't understand a lot of it. It was reassuring to be told by a doctor, "Don't worry many of us don't either."
The doctors and academics we met and spoke to were all very helpful and friendly and took time to answer any questions we had. I was especially delighted to meet doctors Scheinberg and Sternlieb from America, most eminent doctors and colleagues of Dr Walshe.

It was very good to meet old friends and to make many new ones, particularly Ashish and his daughter Ambuja. They had travelled all the way from India to be there, so a big **''HELLO''** to Ambuja and I hope we can meet again before too long.
All too soon it was Wednesday evening and the conference finished. Everyone went out for a meal that evening and a good time was had by all.

Caroline and I were planning to spend the next two days in Berlin before travelling home, but the best laid plans of mice and men.....
Early the next morning Caroline was taken ill with food poisoning. She'd eaten a salad (vegetarian) something different to everyone else and she ended up in the Krankenhaus (hospital) for the next four days on a drip, she was very poorly. Luckily Dr Gillett and Dr Walshe were still at the Hotel and got her admitted to the Hospital (under the supervision of Dr Caca, who had also been at the conference and specialized in WD).
We both learnt quite a bit more German in those four days. Caroline, to communicate with the nurses and other patients, me, just to get around and make myself understood. Very few people speak English in Leipzig.
In the next bed to Caroline was an elderly German lady who was quite senile. Apart from trying to escape from the hospital several times, she took a great liking to Caroline's shoes (possibly because the nurses had locked her own shoes away to prevent the woman from escaping!)

At every opportunity she would pinch them and hide them elsewhere. At one point I think there was a tug of war between them. Whenever I visited she would stand about a foot away from me and just stare; it was very unnerving, but it was hard to keep a straight face sometimes.
In the end Caroline ended up locking her shoes away!

Because Caroline was too poorly to travel back as scheduled, we had to miss our flights home. A very nice lady visited us from the British Embassy. She told us not to worry as she was here to help us in any way she could. She booked flights from Leipzig to Heathrow for the following Monday, (we were supposed to travel back to England the previous Saturday from Berlin till things went A.W.O.L.). I was staying in a place called the Battenhaus in the university grounds, a doctor had very kindly arranged a family room there for me. No one there spoke any English at all. I was given a key at reception and it was explained to me that "When you are here, you are a key, when you not here you give key, you become card". Some days I got so confused, I didn't know whether I was a key, a card or an umbrella stand!
Whilst Caroline was in hospital I decided not to eat any more food I didn't know about so I lived on bananas, muesli bars and bottled water for four days, well it seemed sensible at the time! I had walked into Leipzig on the Saturday evening to do shopping and was looking for a tram to take me back as it was a fair walk , I spotted one destined for Colditz. The only Colditz I could recall was the film of the same name, I had visions of ending up in Colditz Castle and decided to walk back! I think the hunger was addling my brain.
The next afternoon Caroline left hospital, the first thing we did was head to town and eat the most enormous pizza's I had ever seen. She was feeling much better, if not a stone lighter, and was busily practicing her German on anyone who'd listen , telling them "ich bin krank".
The next afternoon we headed for home and were very pleased to arrive, but we certainly won't forget Leipzig in a hurry!