Wilson's Disease Support Group-UK

Newsletter, volume 5, issue 1 **June 2004**

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, 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 one another. The organisation also strives to promote a wider awareness of Wilson's Disease within the medical profession If you have any questions regarding any aspect of WD please contact WDSG-UK at 36, Sunningdale Drive Woodborough Nottingham NG14 6EQ. We will do our very best to answer them as swiftly as possible

Greetings

WDSG - UK

I hope you will enjoy reading the 9th edition of our newsletter, my apologies for the lateness of it.

As many of you are probably aware, Caroline is in the Queen Elizabeth Hospital recovering from her liver transplant, I'm sure you will all join me in wishing her a speedy recovery and hope that she will be back with us before too long.

It has been an eventful year up to now, our meeting at King's went off extremely well; our web site is now functioning again (thanks to James) and we have more new members and friends.

Should anyone like a support group T shirt we have just had a new batch printed, they are high quality heavy cotton and available in black or white with our logo back and front, in S,M or L at a cost of £10 each.

Best Wishes to everyone and enjoy the rest of the summer.

Linda

Wilson's Disease Association. *Volume 5 issue 1*

A BEAUTIFUL SETTING
Webb's court at King's

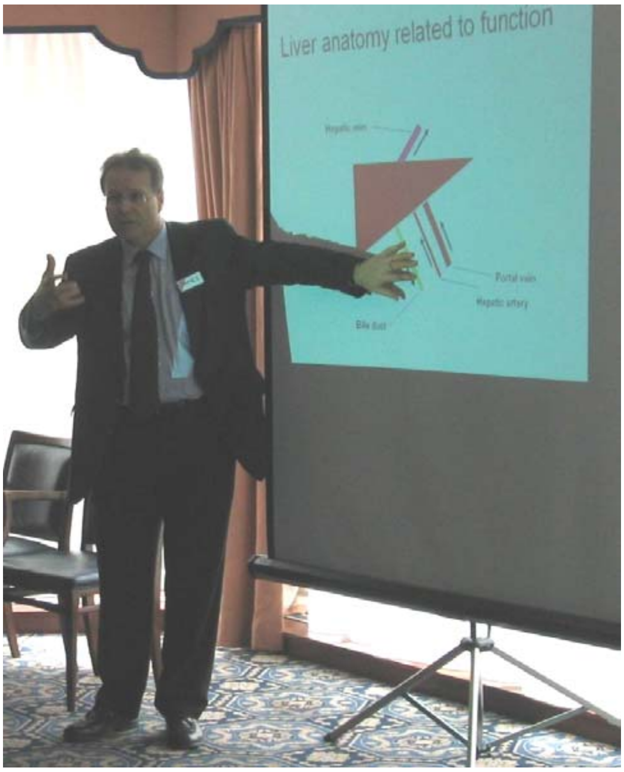
OUR MEETING AT KING'S

by Valerie

The 5th meeting of the WDSG-UK took place on Saturday, 8th May 2004 in the lovely surroundings of King's College, Cambridge, which is only a stone's throw away from Old Addenbrooke's Hospital, a building that many of us once knew rather too well!

Around 56 people attended and, as always, we were pleased to welcome new faces. There was a strong medical and academic representation including Dr John Walshe and Kay Gibbs, Dr Hal Dixon (a fellow of King's and former collaborator of Dr Walshe) Dr Godfrey Gillett of the Middlesex, Dr David Cartwright, of King's College Hospital, and our speaker, Dr James Dooley, of the Royal Free Hospital, London.
We were delighted again to see Dr Rupert Purchase, also Dr Jeremy Drummond and Nazia Ali, representatives from Univar, (the drug company which produces Trientine), who very generously presented us with another cheque towards group funds.

Friends chat in the Bevis room



Dr James Dooley's Talk

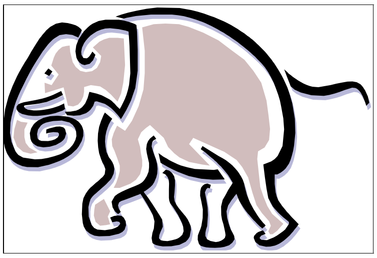
Unfortunately, Caroline was unable to attend as she is recovering from her liver transplant. Both Linda and Dr Walshe brought a ‘Get Well’ card for us to sign; Dr Walshe’s effort reflecting a not insignificant achievement in computer wizardry!

Coffee was available on our arrival, over which we had plenty of time to chat.

Over lunch Dr Dooley gave us a most interesting and informative talk on general aspects of liver disease and the meeting was bought to a close with a very successful raffle.

Dr Dixon then led a conducted tour of the environs of the College, including the world famous King's College Chapel and, more importantly, the Fellow’s Garden, which is normally out of bounds to the *hoi polloi!*

Our thanks go to Linda and Caroline for organising yet again a very successful meeting to Dr Hal Dixon for being a wonderful host and to all of you who turned up and gave us your support.



Valerie, Linda and Dr Walshe are presented with a cheque by Dr Jeremy Drummond Of Univar.

The life of Joan-Two Liver's and several Kidney Stones

By Joan E Smith.

Episode 1.

In 1963, when I was 13 years old, I was diagnosed with Wilson's Disease. I had been doing well at secondary school and then for no apparent reason gradually my concentration suffered and my school stopped me attending. I could not walk properly, feed myself, had spastic movements and tremors. Also I became very emotional. I was initially referred to the local hospital and spent 6 months baffling the doctors until one paediatrician saw copper deposits in my eyes. I was then referred to Addenbrooke's Hospital and Dr Walshe who had discovered penicillamine and its ability to expel the copper from the body. For the first time my life was saved and my health improved to the point where I was fit enough to return to education. I was not allowed to return to my original school as in those days prejudices were far greater than they are now and knowledge of Wilson's disease was very limited.

I had to go to a special school where the education was not very good and I left school without any qualifications but with determination to be normal. To gain qualifications, I attended night school, obtaining GCEs and RSA qualifications with distinctions so that I could become a secretary and obtain an office job.

At this time, because I had the green sunflower shaped cataracts in my eyes an article was published in the Medical Journal which was picked up by the Sunday Times who wrote an article about me complete with a large picture taken in a field near the offices I was working in. The article was in time republished in other newspapers throughout the world. This was my moment of fame as even the Post Office delivered a letter to me addressed to "The girl with sunflowers in her eyes, Coventry, England"

My first job was to run the office of a small building company which I ended up managing and being given a company car at age 21. I passed my driving test on the first attempt but it was lucky to have a company car as I was prone to minor accidents!. By this time I was leading a normal going out evenings and weekends whilst undertaking a full-time job.

When I was 20 I started building my own house with my then boyfriend (now husband of 30 years), brick by brick. Mine was the important job, that of general labourer, carrying bricks, mixing mortar, putting up scaffold and even roofing. The house we built was next to the University of Warwick where I gained my second job helping with the organisation of all the exams including typing some of the papers. Eventually I was promoted as a faculty secretary undertaking all the administration functions for engineering and working for three professors.

At 29 I "retired", being pregnant with my first child, a son. I had a normal pregnancy, taking penicillamine throughout, although he was born 5 weeks premature and not at Christmas as expected.

Whilst weighing less than lbs. at birth he grew faster than his contemporaries and has grown into a fine young man who now designs oil rigs, probably due to the engineering in his blood from my working in the university engineering department!. Two years later, my daughter was born of normal weight if 2 weeks late (that's girl's for you). Both children are now graduates from Nottingham University and my daughter like her brother is now studying for an M.Sc.

During the pregnancies my health remained good and my penicillamine dosage constant at 1000mg per day. I had some problems with raised blood pressure, varicose veins, kidney stones, an overactive parathyroid gland (which was reduced in an operation) and osteoarthritis in my joints. The penicillamine also affected my skin thickness and my blood clotting was poor.

About two years ago after nearly 40 years on penicillamine my energy which I had put into bringing up the family and keeping home started to decline. Gradually and without my realising it, my health began to again show many Wilson's Disease symptoms.

I became tired and breathless, kept forgetting things and thought at 53 I was just getting old and with my medical history this was just happening early in my life. No-one noticed this change in me as it came on gradually and I tried to hide my lethargy until in November last year I became very jaundiced and felt cold no matter how hot the house was. The GP diagnosed gall stones and requested an urgent ultrasound scan which was booked for 4 weeks ahead.

My condition continued to deteriorate so much that I spent virtually all day lying down unable to do things. My husband, as a man not having his meals cooked insisted I be seen more quickly at the local hospital and I was admitted immediately after he spoke to my GP.

EPISODE 2.

In the local hospital they tried to stabilise me. Initially they found a gall bladder infection but were unable to carry out a liver biopsy due to my blood not clotting and despite several injections of clotting agents. I also found I was beginning to swell up due to fluid retention. After 5 days in the local hospital I was transferred to Addenbrooke's which is 70 miles from my home. This was not without trauma as the two ambulance men neither knew the way to Addenbrooke's nor how to reach the correct hospital entrance after circling it a number of times!

It was expected Addenbrookes would carry out the biopsy through a vein as an ordinary one would have caused too much bleeding. After a further ultrasound scan and lots of tests they cane to the conclusion that my liver was in quite a bad way. I received the shattering news that ultimately I would need a transplant but in the meantime may be put on Trientine instead of Penicillamine.

physically I now feel wonderful and fitter than ever before.

At this stage my transplant was not an urgent case but being already in hospital they would undertake the assessment process to see if I was suitable. In this process everything is checked, all your vital organs, teeth, home situation and lifestyle etc.

During the few days after being told of the need for a transplant I was inconsolable and very upset owing to the fact that my condition was rapidly deteriorating. Very quickly I was put on the list for the first suitable available liver. At this time the fluid In my body had built up under my lungs making my breathing difficult and the toxins caused me to be encephalitic.

Within 2 days a liver had been found but proved unsuitable, which was a blow more for my family than me as I was now with the 'fairies'! .

continued over page……………………

Luckily another "beautiful liver" as the surgeon called it was available next day and early on the Saturday morning I had the transplant.

The operation (I am told) took about 5 hours. I spent 12 hours in intensive care and was then moved to a special transplant high dependency unit for 5 days until being able to move out onto a ward. I can't remember much about this phase but I do remember being out of bed on the first day after the operation and being affected by one of the drugs which had to be changed. I remember being told I would be on them for the rest of my days (another 40 years in the plan!)

Two weeks after the operation disaster struck. I was within one hour of being fetched to come home and even the wound staples had been removed. My wound burst open and my bowel came out. This was a very unusual complication caused by my muscle being weakened due to my previous condition and a lack of nutrition from the failing liver. I was returned to theatre for a thorough internal cleaning and disinfection. The wound was re-stiched and I was brought back for a further fortnight on the ward.

This time everything went extremely well. I forget to add that at one stage a swab from my wound proved positive for MRSA, which the hospital treated with a body wash, nasal and mouth spray. To the hospital this was a minor issue and not the major trauma indicated in many press articles. This was cleared before I left for home, and I now return, at present, one day per week to Addenbrooke's outpatients clinic for blood tests to re-assess the levels of drugs I need. Eventually this will change to fortnightly, monthly and hopefully annually.

Physically I now feel wonderful and expect to be fitter than ever before. Mentally I still have some residual effects from the trauma and operation. Of course I no longer suffer Wilson's Disease but will be seeing Dr. Gillett shortly to review whether I still retain any stubborn copper deposits built up before the transplant.

For 40 years I survived on penicillamine. Now I intend to live for another 40 years to take full advantage of the new liver!

A TALE OF TWO KITTIES

by Linda

It would have been around March last year when John and I first saw Scruffy the cat in our garden, he was a pitiful sight, the filthiest, skinniest, most hard done by looking cat we had ever seen. It was pouring with rain and he was soaked, there was no chance of getting near him he was terrified of people. We put some food on a saucer on the yard and watched through the kitchen window, he made his way over and wolfed the lot in record time. Over the following few weeks he kept coming back we would see him lurking around the garden. We would put food down and retreat, John gradually started to move the dish closer to the house, as he grew a bit more used to us he began to realise he was safe, though he was still very skittish; eventually, if he didn't see us around and the door was open he would sneak in and try to eat our cats' biscuits, the poor thing couldn't crunch them though, obviously got teeth problems. We started to mash his food up with milk and add cod liver oil, he thrived on it.

A few more weeks passed and by this time we could stroke him when he was eating his food, John began to brush him, his coat was in an awful state, he didn't seem able to clean himself.

Drastic action came next, it was decided Ruffy as we had began to call him now, needed a bath!. Anyone who has a cat will know this isn't a task to be undertaken lightly! I returned home one lunchtime to find Ruffy wrapped in towels on a box by the radiator, the deed had been done!. How John managed this alone I'm not sure, he (the cat) did look a fair bit cleaner though, John had cut a few chunks out of his fur which were impossibly matted.

As the weather got colder in the autumn Ruffy started to come in the house in the morning for his food, he would have a thorough warm by the radiator and then wander off 'till later or the next day. A friend who was staying with us at the time, found a cat sized wicker basket in a skip so this was cleaned up and put by the kitchen radiator with an old sweater inside, this became Ruffy's new home. He was now around for most of the day and slept in the basket at night (he knew which side his bread was buttered).

He now trusted us, but with slight reservations, after Christmas we managed to get him to the vets, he was neutered and had to have several teeth out, he also had gum disease.

We decided that in the spring we would try to find a new home for Ruffy, much as we both liked him and wanted to keep him, we already have two cats, three would create problems.

In January lots of posters about a missing cat called McGuyver appeared in our local town- his owners had moved and the moggy had escaped. John decided to call them to enquire about the cat, one thing led to another, and we met James and Mel, who became Ruffy's new 'people'. The very day after Ruffy moved to his new home McGuyver was found! so Ruffy now has a lovely new home and a new friend to boot!

We also have two new friends in James and Mel, so this tale has a very happy ending. James is somewhat of a wizard with computers. We had talked about our support group, and the problems we had been having with the web site (of which I understand very little). James offered his assistance and has become an invaluable help to the group, he attended our meeting at King's and is responsible for the Photos in this newsletter.

So a big thank you to James whom we may never have met without feeding that stray cat!

WEB SITE ? WHAT WEB SITE? By James

If you've tried to check out WDSG news and activities on the website in recent months, you'll probably have noticed that it has been, well, just a little less useful than it might be. As in, "what website?"

That, by the time you're reading this newsletter, should have changed.

Over Christmas, New Year and most of January I was out pounding the mean streets of Beeston and Chilwell looking for my cat Mac (aka Macguyver) who'd absconded shortly after we moved house from one side of Beeston to the other. Newspaper ads, posters on lamp-posts, ads in pet shops and quizzing neighbours had turned up nothing definite, though it was encouraging how many people called to say they'd seen a cat of similar description around and about. One odd call, while we were visiting the RSPCA rescue centre at Radcliffe, went along the lines of "well, we've not found YOUR cat, but there's one here who desperately needs a good home if you don't find yours.."

While I was unwilling to give up hope on Mac, and wasn't sure I wanted two cats, we (my housemate Melissa and I) decided to go and see this new one. Although there were a few we liked at the RSPCA, we didn't like their terms (which basically only give fostering rights, not ownership, and carries various other onerous conditions). They'd also had a pretty good month, with few animals coming in, and seemed likely to find good homes for those they had.

And thus we met Linda and John, and the scrawny and timorous little Ruffy, whom we took in a week later. In getting to know Linda and John I unwisely admitted to knowing some things about computers, including web design and domain management, and Linda told me that the WDSG website had been having some problems. No-one was able to see it at all. Yup, that sounds like a problem to me.

Linda brought Caroline to visit and we went over what was known about the website hosting, and a bit about what was needed on it when it was fixed. I had to do a bit of digging to find out what had gone wrong, but that's my speciality.. I'm a software engineer with (hrrm mumble) years of hacking about with setting up networks, websites and suchlike.

Caroline hadn't set the site up herself in the first place, and it looks as though whoever had, had omitted to make clear certain requirements. The wilsons-disease.org.uk domain name was effectively a freebie given away with one of the myriad currently available dial-up Internet packages. The catch was, you have to USE the dial-up service for the domain to remain active. As Caroline had no need of any (more) dial-up connections, she didn't use it, and the provider (FreeNetNames) reasonably enough suspended it. They also asked for a fee to reactivate the domain, but even after that was paid, still no-one knew that they needed to dial up to maintain it, and it vanished again.

So, what's the choices? One, reactivate it once more, and take steps to ensure that someone dials up once a month to maintain it. Ugh. Two, move to another provider. Domain names aren't that expensive, and hosting is available free or very cheap in most places. For the time being, I was quite happy to host it from my home machines just to get it working again.

Easier said than done, though. Internet Service Providers are notoriously unwilling to let people leave for another. Domains name registration is a particularly bad case, as the system requires that the CURRENT holder has to actively authorise the transfer, and all they need to do to inconvenience you is ignore the requests. Easy to claim that they got lost in the ether of email delivery, especially when they provide NO OTHER way to contact them.

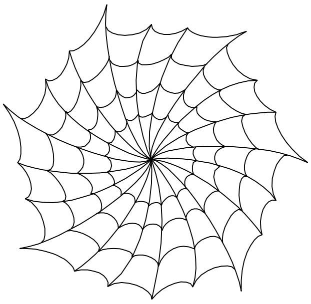
Much pulling of hair later, plus eventually an authorisation fax to my own registrar (well, I say fax, but I actually hand-delivered it to Host Europe in the middle of Beeston!) things actually start happening. A few days later I find myself properly in control of the domain, and able to set up the site.

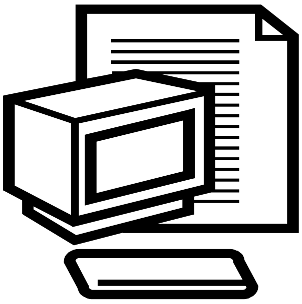
That, at least, is the hard part over with. The previous contents of the website wouldn't magically be restored, as that was on a FreeNetNames server. However, the pages had been archived by search engines, and Caroline also had an old copy of the files, plus some points that needed updating.

I threw all these in a mixing pot and out fell some sort of rendition of the "new" WDSG-UK website, which should be visible at the same address as previously: http://www.wilsons-disease.org.uk

Ruffy, by the way, is doing fine. As is Mac, who was found and retrieved the very day after we took in Ruffy, after being living in local gardens for four weeks.

Ruffy seems to shed about half his body size in hair every day, eats like a horse and is generally to be found sat next to his bowl, but is starting to actively seek out laps to sit on, and has found a perfect spot on a flat roof outside to sun himself.





Membership Renewal Form 2004

I still wish to receive the WDSG-UK biannual newsletter

Name…………………………………………..

Address……………………………………………………….

……………………………………………………………….

Email………………………………………………………….

UK membership: I enclose a cheque/ postal order for £5.00 for my annual membership.
USA/ CANADA membership: $10 (please note we cannot accept non UK cheques but can accept IMO'S (International Money Orders) or $10 bills by registered post)

I wish to donate £……….To WDSG-UK
all cheques/ postal orders should be made payable to Wilson's Disease Support Group-UK

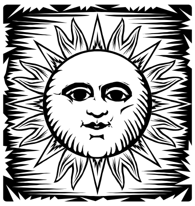
N.B. Those of you who have already paid your 2004 membership may ignore this form.
Please note if you have not renewed your membership for this year we regret that we will not be able to send you future newsletters unless you complete and return this form before September.

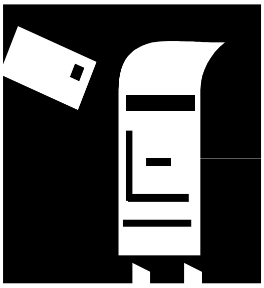
A BRIEF HISTORY OF MEDICINE

“Doctor, I have a sore throat”.

2000 BC - Here eat this root
1000BC - That root is heathen, say this prayer.
AD 1850 - That prayer is superstition, drink this potion.
AD 1930 - That potion is snake oil, swallow this pill.
AD 1980 - That pill is ineffective, take this antibiotic.
AD 2000 - That antibiotic is artificial. Here eat this root!.

DON’T FORGET TO RENEW YOUR MEMBERSHIP!





LEISURE

What is life if full of care
we have not time to stand and stare.

No time to stand beneath the boughs
and stare as long as sheep or cows.

No time to see when woods we pass
where squirrels hide their nuts in grass.

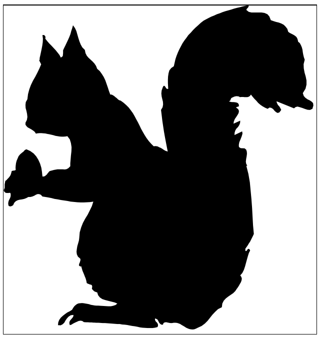
No time to see in broad daylight
streams full of stars, like skies at night.

No time to turn at beauty's glance
and watch her feet, how they can dance.

No time to wait 'till her mouth can enrich
that smile her eyes began.

A poor life this if full of care
we have no time to stand and stare.

from songs of joy. W.H Davies. 1871-1940.



Wilsons Disease Support Group UK

For membership, correspondence, and information

Contact : Dr Caroline Simms (group chairperson and coordinator, newsletter production, information and awareness)
36 Sunningdale Drive, Woodborough, Nottingham. NG14 6EQ

If you would like to be put in touch with others with WD

Contact : Linda Hart (patient and family correspondence, coordinator, newsletter production)
36, Audley Drive, Lenton Abbey, Beeston, Nottingham. NG9 2SF.

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

Ann Widdecombe M.P. - group patron
Dr John Walshe - honorary president, medical expert and world authority on Wilson's Disease
Dr Godfrey Gillett - group adviser, honorary member
Dr James Dooley - group adviser, honorary member
James Beckett - web master, honorary member

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 MPs 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:
Linda Hart
36, Audley Drive, Lenton Abbey, Beeston, Nottingham. NG9 2SF.

We're on the web
www.wilsons-disease.org.uk