Wilson's Disease Support Group-UK

NEWSLETTER, VOLUME 4, ISSUE 1 MAY 2003

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 33, Cavendish Street, Arnold, Nottingham NG5 7DL. We will do our very best to answer them as swiftly as possible

NEW LIVER OPERATION COULD END SUFFERING

Article from The Mail on Sunday– 11th May 2003

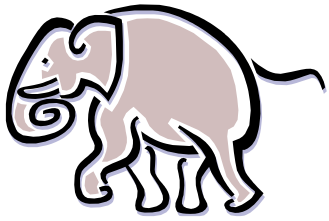
British doctors have developed a new surgical technique that could make liver transplants a thing of the past.

In a world first, they have injected three babies suffering from rare life threatening liver diseases with healthy donor liver cells in an attempt to encourage their damaged livers to regenerate.

All three babies have made a good recovery.

Experts at King's College Hospital, London, who carried out the procedure, believe it could revolutionise the treatment of patients of all ages with chronic liver disease or liver failure .

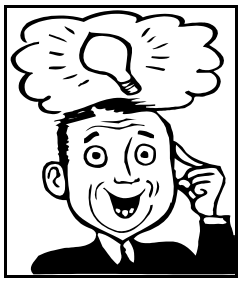
Already, doctors hope it could prevent hundreds of children from going through the trauma of whole–organ liver transplants.



WDSG - UK

Dr Anil Dhawan, a consultant in children's liver disease at the hospital, said: "This will free up more donor livers and increase the overall number of people who could be offered liver replacement."

About 60 people a year die in Britain waiting for a liver.



DON'T FORGET TO RENEW YOUR MEMBERSHIP.

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BELINDA'S STORY

by Belinda Diggles

My name is Belinda. I am married to Barry and we have two teenage boys. In July 2001 I was diagnosed with Wilson's Disease, but I was one of the lucky ones in that I was not ill for a long time.

In the March I was unwell, but both my doctor and I thought this was due to my irritable bowel, and that I had a bowel infection. I was treated with antibiotics and I changed my diet. As all this was going on, my dad was taken ill, and I was at the hospital with him in Yorkshire. We were told he was not going to get better. Therefore I stayed with him, when I should have been back at the doctor's. When I came home, I went to the doctors and he just thought that all the stress I was under was not helping me. He wanted me to take some time off work, but I wouldn't. I just wanted to keep busy. In the end I did have to take time off work; six weeks in total. However when I went back to work I started to feel unwell again, after just one week. So there I was again, back at the doctors. This time he decided to send me for blood tests.

If you could test for it, he tested it! That was on the Tuesday. By Thursday my son rang me at work to tell me the doctor had been trying to contact me. I rang him, he told me that I had a very high L.F.T. count, and that he had been in touch with the hospital for me to see a consultant gastroenterologist. There was a long waiting list, so I decided to go privately, because I was so worried.

My doctor sent me for blood tests and an ultrasound scan, as a matter of urgency. I was very jaundiced by now. I was always feeling tired sleeping for two or three hours every afternoon. Four weeks later my husband rang me at home, to see how I was, but I was still in bed. I told him I was not getting up as it was just too much effort. He was so worried he contacted our doctor, who in turn got on to the hospital, to arrange for me to be admitted. At the time I did not realise how ill I was or that it would get a lot worse. In hospital I was in a room by myself and they were even talking about a liver transplant if my liver didn't start working. I was in a dream; this just does not happen to me, but to other people.

They did blood tests and a 24 hour urine collection. Every day they took more blood, and I was sent for a C.A.T scan.

I just kept deteriorating and they were getting no nearer to finding out what was wrong with me. Then after three weeks they told me I had something called Wilson's Disease, but no-one had heard of this. This was on the Friday and we had to wait until Monday when the doctor came on his rounds, for him to explain to us what it was and put me on the drug Penacillamine.

Unfortunately, the drug gave me chronic diarrhoea. Things went from bad to worse as they started treating the diarrhoea. After six days I started being sick after food, but after three more weeks they let me go home. I did have two afternoons at home before that, but it was six long weeks, I went home FOR GOOD!!.

I had to go back five days later for a check up, but I did not mind that. I was going home and I was alive. There was a time I did not think this day would come, I thought if you got that ill, you did not get better. It was then I realized I should stop feeling sorry for myself.

When I went back for my check up, the doctor decided to do some more tests to see if there was another reason I couldn't get rid of the diarrhoea. When he could not find anything, he decided to change my tablets to Trientine. Within days I started to improve.

Yes it did take time, and when I look back at my recovery period, I had little milestones along the way. At first I could not walk up the stairs. At times I would make it up there after considerable effort, taking a couple of steps at a time, and when my husband Barry came looking for me sometimes, he would find me on the bed, as I didn't have the strength to get back down again.

After a few weeks I started going up the drive for a little walk, then building up to a ten minute walk after six weeks.

Three months after leaving hospital and a slow steady recovery, we all went on the holiday that we had had to cancel three days prior to my being admitted to hospital, back in June. That holiday was a three week tour of the west coast of the U.S.A. Whilst I was away I was in bed by 9.00pm most nights, but we all had a great time.

I finally went back to work in December for just a few hours a week, and I felt this was the last step to getting back to normality, but I would not say that it has all been straightforward.

In January 2002 my L.F.T count started to go up again. This was when my doctor started me on zinc, then in June I stopped the Trientine and my L.F.T's are now back to normal.

The only problem that I have is that I soon get tired, but you know what?

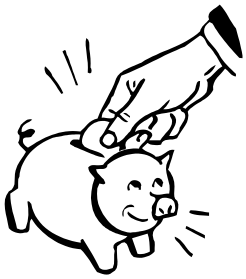
I can live with that!!!!

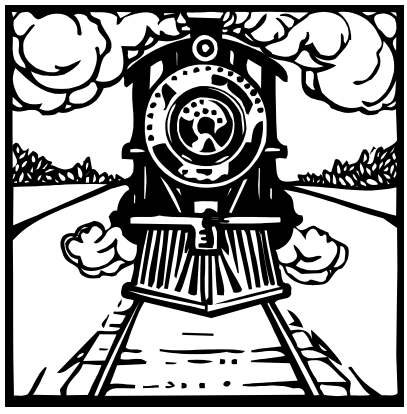
ARE YOU ELIGIBLE FOR A DISABLED PERSONS RAILCARD?

Did you know that you may be eligible for a Disabled Persons Railcard if you are in receipt of Incapacity Benefit or Disability Living Allowance (DLA).

You can pick up an application form your local Citizens Advice Bureau, major Post Office Counter branches and major railway stations. You will need proof that you get one of these benefits and then all you need to do is fill in the form and return it to the address given on the form along with a cheque for £14. The card lasts for 1 year and will save you a third on all rail journeys in the UK. You need to show your card when purchasing tickets and you must carry your card with you whilst you are travelling with discounted tickets.

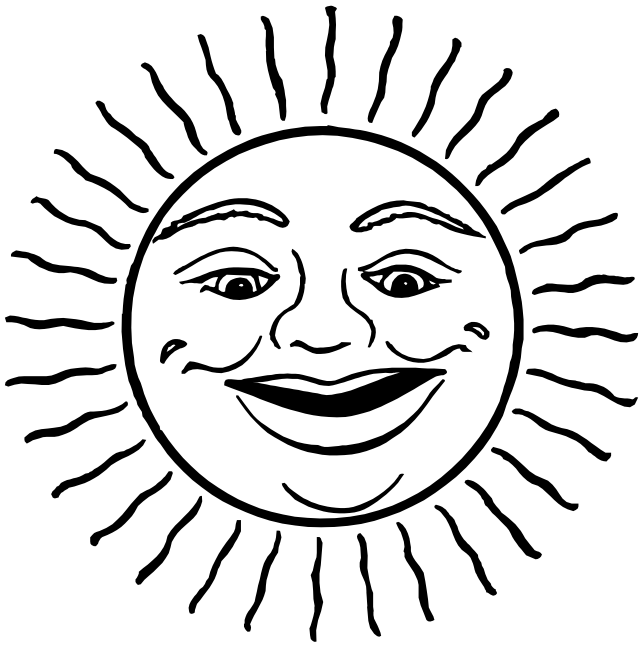
It is also worth noting that some council boroughs offer travel discount cards for disabled people which can save you money if you use public transport. Again ask your local Citizens Advice Bureau or City/Town transport office.





The power and meaning of a smile

It costs nothing, it creates much.
It enriches those who receive it, without impoverishing those who give.
It happens in a flash, and the memory of it sometimes lasts forever.
There is none so rich that they can get along without it,
All benefit.
It creates happiness in the home, goodwill in business,
and is the countersign of friends.
It is rest to the weary, light to the discouraged
and a antidote for trouble.
Of no earthly good until it is given away.
So if a friend is too tired to give you a smile,
give him yours.
The need is greatest for those who have no smile to give.



DIMERCAPROL

Dr John Walshe.

In previous numbers of the newsletter I have given an account of the discovery of penicillamine (1955) and trientine (1969) which are the most commonly used drugs in the treatment of Wilson's disease.

However, the story of effective treatment goes back further. It really started way back in 1939 when Hitler was expected to attack this country with a poison gas called Lewisite; this is an arsenic containing blister gas. As a result of this fear a team of biochemists in Oxford, under Professor Sir Rudolph Peters, started work on finding an antidote. What they came up with was a compound with the chemical name, dimercaptopropanol. This is a small molecule with two active sulphur atoms capable of binding or technically, chelating arsenic.

Being an 'anti lewisite' drug it was named by the Americans British Anti Lewisite, or BAL for short. Later it was given the official pharmacopoeia name of 'Dimercaprol', but still commonly referred to as Bal for convenience.

The story now moves on to 1948 when Professor Cumings, at the National Hospital for Neurology in London, published an article that proved conclusively that Wilson's disease was due to an inherited defect in handling copper, so that the metal accumulated in the liver and the brain causing progressive damage to those vital organs. He suggested that BAL had the right chemical formula to combine with the copper and arrest the progress of the disease. In 1951 Cumings, in London, and Den-Brown, in Boston (USA), showed that BAL could actually reverse the symptoms and give significant improvement in some, but not all patients.

However there were snags. First BAL was only soluble in oil, (peanut oil was used) so it had to be given by painful injections into a large muscle, preferably the buttock. It caused fever, damage to the white blood cells, nausea and dizziness.

It could be something of an ordeal to the patient. Furthermore it soon became apparent that as repeated courses were given the drug became less and less effective, so that it could not be used as a long term control of the disease. Thus when penicillamine and later trientine were discovered BAL fell out of use.

BAL could actually reverse the symptoms and give significant improvement, in some, but not all patients.

Why is it used now, you might reasonably ask? Well in the 1980's Dr Schienberg, in New York, found that those few patients, particularly the more rigid ones, who did not do well on penicillamine or trientine, could be helped by one or two short courses of BAL, after which they could go back to oral treatment. In this situation some patients can get real benefit when all else has failed.

The course Dr Schienberg suggested was to give 200mg of BAL each day, into alternate buttocks for five days a week for four weeks, then two weeks rest and repeat the course until a maximum benefit is achieved.

If this does not help after the first two of the four week courses, the drug is discontinued.

The advantage that BAL has over other treatments is that it is fat soluble. This means that it can cross from the blood into the brain very much more readily than either penicillamine or trientine so that it can get out brain-copper when other drugs cannot get at it .

There is a water soluble form of BAL known as Dimival which can be given by mouth, but it has lost the advantage of easy access to the brain.

Continued from previous page

There is a further snag with BAL: its cost. A forty day course of BAL costs over £3,000 Dimival is even more expensive: a year's treatment costing about £14,000 and it is not apparent that it has any advantage over the much cheaper penicillamine and trientine.
There are two other possible other treatments used for patients with Wilson's disease: Zinc acetate and Tetrathiomolybdate and I will talk about these in a later number of the newsletter, but note that all the treatments for this disease have been discovered by university based research workers, none by the Multinational Pharmaceutical companies.

FREE PRESCRIPTIONS

Dr John Walshe

I have now heard from our patron, Miss Anne Widdecombe MP, that the Ministry has turned down her request that patients with Wilson's Disease should be entitled to free prescriptions. "There is no intention of reviewing the list of diseases for which free prescriptions are authorised", This is the same answer that I have had from all ministers since the days of Harold Wilson's administration. However, she does think it is an excellent idea that you all write to your MP's requesting this. So it is up to you now.

It is not a good idea that you all write the same letter,This will look to orchestrated. Write in your own words, but you can take your points from these ideas, but don't quote me verbatim.

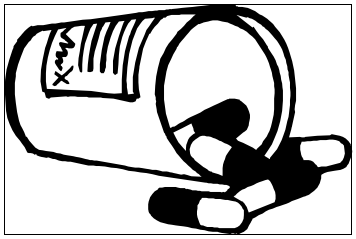
Apart from welfare considerations, there is a small list of diseases for which free prescriptions are given. This list came in shortly after the Health Service came in, in 1948. Apart from epilepsy and myasthenia gravis they are all diseases caused by endocrine gland failure, such as thyroid deficiency. Either the list was made up by an endocrinologist with the help of a neurologist or these were the only diseases for which there was effective therapy at the time. The list has never been updated.

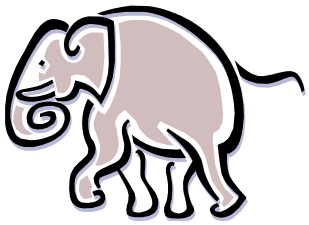
It is probable that had the treatment of Wilson's disease been discovered when the list was made up it would have been included. It makes economic sense to treat patients with Wilson's disease. Untreated the patient becomes a considerable financial burden on both the Health Service and the Welfare Services. The great majority of patients who are well treated make a good recovery, become useful members of the community and may well earn enough money to pay income tax (!!!)..

The cost of giving free prescriptions is infinitesimal in terms of the Health Service budget.

I calculate that if there are 800 diagnosed and treated patients in the country each receiving free medicine, say two prescriptions a month, it will cost the Health Service £125,000 a year, the price of one minister, two doctors or five nurses. What will be saved by successful treatment amounts to hundreds of thousands of pounds.

So go ahead and write, the more the better and give a really good sob story, pointing out how awful the disease is and it is only justice that you are granted free drugs.





WDSG - UK

Wilson's Disease Support Group -UK, Volume 3 , Issue 2 .

The Holiday season+ membership renewal	*7*
5th Annual WDSG meeting in Nottingham	*8*
Anousha's holiday diary	*9+10*
Talk on Wilson's Disease given by Prof Eve Roberts	*10+11*

The Holiday Season

Here are some useful tips for you when you go on holiday:

- Always keep your medicines with you in your hand luggage when travelling. If the airport loses your case you have also lost your tablets. So keep them with you.
- Your medication should be correctly labelled. The one from the Pharmacists is sufficient.
- Carry a letter from your doctor stating what medication you are on and that they consider you fit to travel. This is very important for medical insurance.
- When travelling abroad it it VITAL that you get FULL MEDICAL INSURANCE COVER. Read the smallprint very carefully and ensure that the cover includes existing medical conditions. Be careful of a clause that mentions if you have been in hospital in the past 12 months prior to the date of travel. This one nearly caught me out last year!
- Always check with your doctor before you travel. Make sure he/she is aware of where you intend to go and when.

Membership Renewal Form 2003

I still wish to receive the WDSG-UK biannual newsletter

Name……..

Address…….

…….

Email…….

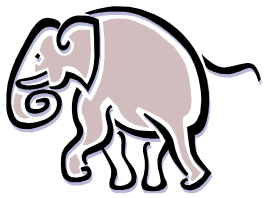
UK membership: I enclose a cheque/ postal order for £5.00 for my annual membership.
USA/ CANADA membership: $10 (please note we cannot accept non UK cheques but can accept IMO'S (International Money Orders) or $10 bills by registered post)

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 2003 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 DATE FOR YOUR DIARY - don't miss it !!!
5th WDSG-UK Meeting, 20th September 2003

We are holding another meeting in Nottingham on Saturday 20th September 2003



Event Details:

Venue: Nottingham University Staff Club, Nottingham.

Date: Saturday 20th September 2003.

Time: 11am till 3pm (*please note that we have to vacate the club by 3pm prompt.*)

Cost: £5 per person (*to cover buffet and room hire cost*)

Important: If you would like to attend please return cheque or postal order (£5 per person)in advance along with completed form (name, address and stating clearly the no. of people coming). Unfortunately we can only book you a place if payment is received in advance.

Maps and final details of itinerary will be sent out 3 weeks prior to the meeting date.

If you've not been before, the day is quite informal. People usually get a lot out of the event. It's a chance to meet others, make contacts, friends, and ask questions.

We hope Drs Walshe and Gillett will be able to attend again

We look forward to seeing many of you there !!!

I/we are coming to the 5th WDSG/UK meeting in Nottingham
Please book me……...place/places in advance.

Please **find enclosed a cheque/postal order** for £…..(**made payable to WDSG-UK)**
for…….person/people for **tickets in advance**

Name……..
Address……
……..

Delete as appropriate:
We will be travelling by car/ train/ coach
Please send/ do not send details of hotel accommodation in Nottingham

Please cut out and complete this form (membership renewal is on the other side)
and **return to: Linda Hart at 36, Audley Drive, Lenton Abbey, Beeston Nottingham. NG9 2SF.**

ANOUSHA'S HOLIDAY DIARY

By Anousha Joseph.

I thank my aunty Edel for giving me this tremendous idea to write a diary about my holiday from 8th August to 5th November 2002. Nobody else had given me this idea before, so it was now up to me to make the diary interesting to read and with plenty of details. She is a good listener, helper and very kind too. Edel is my mum's best friend and so that is why I call her Aunty. I also wish to thank my parents for taking me on this extravagant holiday, as I know now, how hard it is for one to struggle without so much money to spend.

My father and I left England on 8th August at around 3.00pm. After a long flight we arrived in Sri-Lanka at 6.00am on 9th August. Uncle Percy met us at the airport. It felt cool to be landing in Sri-Lanka. We did not have any problems and it had been a smooth flight. It made me feel like I was back home again, as I have visited this country before on several occasions.

Sri Lanka was formerly known as Ceylon and is an island at the tip of India. I have read that Marco Polo has said that " Sri Lanka is one of the finest islands in the world" and I could not agree with him more. The languages are Sinhala Tamil and English.

Columbo is the capital of Sri Lanka and is the point of arrival for all visitors like myself. It is a pleasant city and by Asian standards, quiet, relaxed and organised. However it has become less interesting for foreign visitors than most capital cities.

Three wheelers are the common means of transport in Sri Lanka as well as India. Some autos have meters, but some do not and Rs20 is the average fare for a short distance, otherwise for long distances the fare would be Rs30 to Rs60.

The three wheelers are Indian scooter rickshaws and occasionally known as tuk tuks. A tuk tuk is baby talk for a three wheeler. It is the smallest vehicle, can easily get through traffic and can seat five passengers at the most, you have four seated at the back and one with the driver, you have to be very careful. If the police catch you, you will be fined (legally the number of people that can travel in them is four and that is the law in India as well!). Of course they do have public transport– buses and trains, which are terribly overcrowded, and the rich man travels by car.

The usual meal you get is rice and curry, that means a mountain of rice served with a small choice of meat or vegetable curries. Also there is a Sri Lankan soup called rasan, this is used to pour over the meal if it is too dry. I like it when I have rice and curry along with a spoonful of yoghurt. International cuisine is normally served in hotels.

I rang aunty Gnanes from uncle Percy's house to tell her that we had arrived safely in Sri Lanka. It was aunty Gnane's 49th birthday, so I had a wash and changed and went to her house with my dad by auto at 1pm with Newton mama,(mama is the Tamil term for your mum's brother) and family. Valentine mama and family who were also holidaying in Sri Lanka at the time and came to auntu Gnanes's house as well. Ashley and I are aunty Gnanes's godchildren Arun made a beautiful birthday cake for his mother and did all the icing himself. I think she knew what he was making, he is good with his ideas.

We got home at 22.30, I rang Manju to tell her that I was in Sri Lanka and aunty Ranji invited dad and I to come for dinner on the 11th of August. I spoke to Manju about the dress materials I had brought with me from the UK to be made up in Sri Lanka.

On 11th August, uncle Damian, aunty Anne, Adrian, Merion and Muriel visited us. Later on I went to mass at St. Lawrence where I met Ashley.

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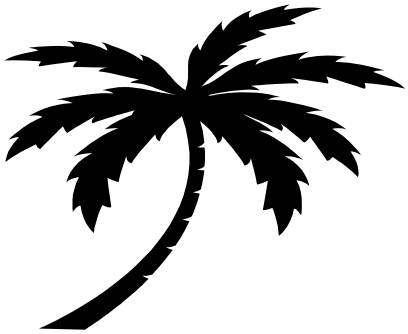
continued

Unfortunately the mass was cancelled due to a feast. Dad and I went to Ashley's house for lunch and to aunty Ranji's house for dinner. Manju and I went for a stroll along the beach. You could see waves smashing into each other, boys playing cricket, the coconut palm trees swaying side to side and the beautiful orange sunset behind the clouds. It was a picturesque scene.
We had Idili, a typical Sri Lanka dish, for dinner and it was so yummy! (soak lentils and rice, then grind It together to make idili, make round balls and steam it, that's your idili recipe, try it at home!). After dinner, Nishan dropped dad and I at Ashley's house, because the following day all of us were going to Nuwara Eliya and we had to get up very early to avoid the school traffic.

As soon as I got to Ashley's house, I washed my hair and afterwards I watched a movie with Ashley and Amanda. The movie was called "Spy Hard" and it really made me laugh.

To be continued……..



TALK ON W.D. GIVEN BY PROF. EVE ROBERTS AT THE SHEILA SHERLOCK MEMORIAL SYPOSIUM, HEPATOLOGY 2003,LONDON. (AT THE ROYAL FREE HOSPITAL)

Wilson's Disease is a disorder of hepatic Cu (copper) disposition affecting the liver first, then eventually brain, kidneys, cornea, heart, synovia. Gastric inflammation is not uncommon.

The diagnostic criteria are:

- Age range 3-45 (or older)
- Extremely low serum level of caeruloplasmin
- KF (kayser– fleischer) rings
- Basal urinary Cu
- Modest in caeruloplasmin—but keep looking

Caeruloplasmin diagnosis is relatively uninformative. Not all patients have KF rings in the eyes-40% may not, and they are often not present in children. Basal 24hr. Urinary copper is useful as it reflects the total body Cu load.

Diagnosis of Wilson's Disease is usually based on the following:

- Abnormal liver test
- Neurological symptoms
- K-F rings
- Low caeruloplasmin
- Low serum Cu
- High basal 24hr urinary Cu

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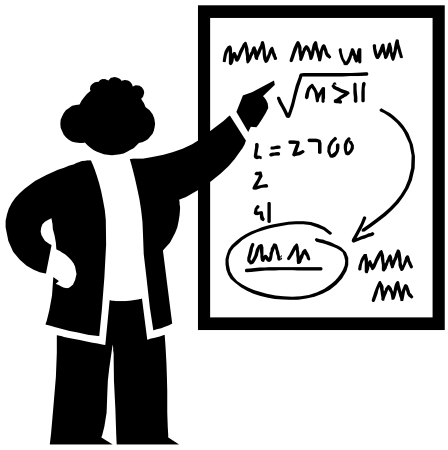
Genetic Diagnosis
This is diagnostically definitive, but the problem is the large numbers of mutations especially the compound heterozygotes where the mutations are different ones e.g. in Iceland, Sardinia, Eastern Europe and Japan.

Treatment of W.D.



1 Chelators (which bind to the copper in the body)
- Penicillanine
- trientine
- Tetrathiomolybdate (most recent and still undergoing trials)

2. Inhibitors
- Zinc
- Antioxidant vitamin E

3. Liver Transplantation

Research

There are various studies currently being undertaken using animal models to look at the effects of copper toxicity and the characteristics of the ATP7B (Wilson's Disease WND) gene.
This gene is a large gene and is related to metal resistance genes.

There are approximately 220 mutations (and still counting) of the WND gene with H1069Q being the most common– it is involved with abnormal trafficking within the hepatocyte (liver cell).
The mutations are classed from 1-4 depending on type, mode of action and effect.

Summary of the talk
Research has shown that:
- W.D depends on the nature of the mutation
- The structure of the WNDp (product) has been established
- Mitochondria (which release energy in cells) are integral to the mechanism of Cu transport. (in W.D the mitochondria tend to be abnormal)
- There are new insights into the handling of Cu in hepatocytes

Prof . Eve Roberts does nor regard W.D as really rare: at her paediatric department in Toronto there is one new case every other year.

Prof Eve Roberts is based at the University of Toronto, Department of Paediatrics, Medicine and Pharmacology.

Thanks to Ursula Mott for attending this meeting on behalf of WDSG-UK and taking these useful notes.

Wilsons Disease Support Group UK

For membership, correspondance, and information

Contact : Dr Caroline Simms (group chairperson and coordinator, newsletter production, information and awareness)
33, Cavendish Street, Arnold, Nottingham. NG5 &DL

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

Contact : Linda Hart (patient and family corresspondance, 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 Alan Stevens - group adviser
Dr Jim Lowe - web master

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

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