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Personal Experience...

by Caroline Simms

BEFORE DIAGNOSIS
I suppose I first noticed that my hands were becoming increasingly unsteady at the age of about 12 years old. I was doing well at school but was aware that things were not quite right healthwise. The tremors were so subtle that initially no one else noticed, and the progression was slow that the people who knew me did not notice any drastic difference. I instinctively knew it was something more serious than stress, worry and anxiety, which my GP had suggested I was suffering from.

As the tremors progressed, drooling became a problem, speech became slurred and I began to choke on eating and drinking. I seemed to be losing coordination in swallowing and speaking. Of course this made me ideal game bait for bullying at school. I suffered three years of psychological bullying at school before I was finally diagnosed.

During the progression of WD before my diagnosis, I became more and more withdrawn and would now describe myself as having been severely depressed often feeling suicidal. Despite frequent trips to my GP, who failed to listen to my plea for help, and who failed to refer me to a hospital for consultation, I felt totally isolated and it felt like I was making it up as no one seemed to listen or acknowledge the fact that I was actually ill, and getting more so.

Eventually things got so bad at school, I refused to go which brought everything to a head. I went back to the GP and my mum demanded a referral to a hospital to see a specialist.

I was admitted to Queens Medical Centre, Nottingham within two weeks for tests . I was on a neurology ward for adults but had a side room. I think I was in for about two weeks, and was diagnosed at the age of 14 with Wilson's Disease by Dr D Jefferson (consultant neurologist) and his team.

AFTER DIAGNOSIS
I remember feeling an immense sense of relief that they had finally found out what was wrong with me and that WD was a treatable condition. The feelings that I had that no one was listening to me, and that I was feigning illness dispersed into the background.

I was started on penicillamine for decoppering but was not on this for long before being put onto trientine instead (I was intolerant to penicillamine). I have been on trientine ever since without any adverse side effects.

I have a few complications due to Wilson's Disease, portal hypertension, oesophageal varices, and hypersplenism but apart from that I am quite well and lead a relatively normal life. I still have mild tremors in my hands but the doctors think, as I do, that I have made a good recovery.

Having WD has taught me an important lesson, it has taught me not to give up, to persevere despite difficulties which life may throw at you. I feel WD has

given me inner strength and fighting spirit. I realise each day a person lives is precious and I try and appreciate each day and live and enjoy life to the full. Since my diagnosis in 1987, I have studied Chemistry at University and in 1999 I was awarded my PhD aged 25.

Dr John Walshe's retirement

Dr John Walshe has dedicated most of his working life to the study of Wilson's Disease. He was the discoverer of D-penicillamine (late 1950's) and trientine (1970's and early 1980's) as clinically useful drug therapies for the treatment of Wilson's Disease.

As a world authority on Wilson's disease he is an invaluable source of information regarding the disease and its treatment. Though his imminent retirement from the WD clinic which is held at the Middlesex Hospital, London will leave a gap in some of our lives having seen him for many years and know him as more of a friend than Â doctor¹, we will be pleased to see Dr Godfrey Gillett (who has shadowed Dr Walshe at the Middlesex clinic for the past 5 years) continuing the WD clinic.

So all the best to both Dr Walshe and Dr Gillett.....without people like yourselves who take a special interest in rare conditions..... the world would be a lonely and futile place... .....so we are eternally grateful and offer you our many thanks...

My time at Addenbrookes.....

by Linda Hart

I remember well the first time I saw Addenbrookes Hospital in Cambridge. It was 1964, I was just ten years old and it looked a huge and scary building, not at all like Derby Childrens Hospital, where I had been spending a lot of time for the last year or so. My parents had brought me to Cambridge because at last someone knew what was wrong with me. All I knew at the time was that I had been feeling lousy and I had no idea that what I had was remotely life threatening. Before this, I had overheard talk that 'she's going off, just like her sister did.' GOING OFF?, what did they think I was, yesterdays milk.....? My sister, Iris had died the year before I was born. My dad had taken it very badly, and then the same things began to happen to me. Mum used to put talcum powder on my skin to try and make me look less yellow to try and stop my dad worrying so much. Of course it didn't work, I just looked very silly, but when I think about it now it is very touching.

When I first met Dr Walshe and Kay, his assistant, he had me drawing circles and spirals and touching my nose. I remember being very impressed by all this and I thought they were really nice people and I felt very comfortable. Whenever it was time for blood tests, Dr Walshe always said to 'think about your Christmas Dinner,' what I really thought about was chocolate, big bars of it. So you can imagine my shock and horror when I was told about my new low-copper diet - NO CHOCOLATE! To a ten year old this was a major catastrophe. At the time my dad always brought me a bar of chocolate home from work on Fridays and this was a very important ritual. However Dr Walshe said I could probably have one Mars bar a week, so this was now duly brought home every Friday. My new ritual became slowly unwrapping the bar and cutting it into seven equal pieces (looking suitably 'hard done by' as I did so). I would savour a piece daily, if my brother ever took a bit, war broke out!

As my health improved I began to enjoy Cambridge very much because I was in hospital for a few weeks at a time. The nurses on the childrens ward would take me out on their days off, so I got to know the town very well. I made many friends during the time I spent at Cambridge that I have kept to this day. In the 1960's there were few people who had Wilson's Disease diagnosed. In his laboratory at Downing College, Dr Walshe had a map of the British Isles with pink and blue flags stuck on it with all our names on them. It always fascinated me as did most of the things there, the bottles, jars, tubes, the big woolly gonk for resting arms on during blood taking, I loved it there.

In due course, I returned home feeling better and life returned to normal, my brothers resumed chucking me in the stream behind our house (to test the depth of the water) and strapping me to the sledge in winter before sending me hurtling down snow-covered hills - life was great again.

However it would have been a very different story for me and most probably all of us, had it not been for the work and dedication of Dr Walshe. So I would like to wish him a long and happy retirement and may his fuchsias always flower profusely.

Diet and Wilson's Disease .....

The diet debate

As copper is present in different amounts in a wide variety of foods, it would be extremely difficult if not impossible to completely eliminate copper from the diet. As long as you are eat sensibly and be careful not to overindulge in high copper foods and of course most importantly take your medication regularly as directed by your physician who monitors your WD and copper levels your copper level should remain well controlled. Dietary restriction alone is not enough to control Wilson's Disease.

Always take your medication on a regular basis as directed by your physician !!!

Here is a list of foods which are said to be moderately high in Copper.....and for people who have Wilson's Disease it is advisable not to overindulge :

- Organ meats : Liver, heart, kidney,
- Shellfish : oysters, scallops, shrimp, prawn, lobster, crab,
- Soya products: soya milk, soy protein meat substitutes, tofu
- Nuts and seeds: most types and do include peanut butter and tahini
- Vegetables to limit inc. Mushrooms, vegetable juice cocktail (e.g. V8), Olives
- Fruits to limit inc. Nectarines, rosehip, advocado, commercially dried fruits inc. raisins, dates, prunes, apricots, sultanas
- Dried beans including soy beans, lima beans, baked beans, garbanzo beans, pinto beans, dried peas, lentils, millet, barley, wheat germ, bran breads and cereals, soy flour, soy grits, fresh sweet potato
- Soya milk
- Cocoa, chocolate
- Mineral water
- Multivitimins with copper or minerals, Brewers Yeast

Leading a relatively healthy and happy life is a matter of balance and to avoid all things which you might enjoy might make you very miserable. So use common sense and don¹t overindulge.....

Top Tips - Special considerations in WD

by Caroline Simms

The copper content in a specific food can vary depending on a number of factors. The copper content and the location of the soil in which the foodstuffs were grown, or the methods used to process the food, for example, can affect how much copper is in the food when eaten. In general a low copper diet is meant to limit foods that are usually high in copper, especially organ meats, heart, liver, kidney, shellfish, dried peas , beans and lentils, soy products and cocoa (YES that means CHOCOLATE!!!)

Copper sulphate pentahydrate ('blue vitriol') CuSO4.5H20 is still used as a fungicide (Bordeaux mixture) to protect crops. It may be advisable to eat organic foods as much as possible or to at least ensure that you thoroughly wash all crops before consumption. I have contacted MAFF (ministry of agriculture fisheries and food)who are sending me more information on usage of Copper compounds in agriculture and which crops they are used on!

Drinking water should be analysed because it may contain too much copper (in soft water areas the copper content in tap water may be higher). If the water contains more than 100 micrograms per litre, then bottled, demineralised water should be used. Water filters may be used but these may not actually reduce the amount of copper in the water. You can ring your local water authority to find out more about your local supply - ask for customer information. (I am currently finding out about water filters/ jugs and whether they remove copper!)

USEFUL INFO: Always run the tap well before consuming tap water, water pipes in houses are made from copper and any soluble copper salts which may be present in the pipes should be removed before you drink it!

Read food labels, some prepared foods list copper content (though this is unusual)

Do avoid combined multivitamin and mineral supplements, these usually contain copper (multimineral supplements usually include copper) ! If you take multivit supplement check the label for copper. If you take mineral supplements buy them as individual supplements and check the label , if in doubt ask the pharmacist or ring the manufacturer! (I am currently finding out more information about healthfood supplements...)

Do not use copper cooking utensils

Most importantly eat a balanced diet, use common sense, avoid or limit foodstuffs high in copper , and take your medication on a regular basis!