Wilson's
Wilson's Disease Support Group - UK
www.wilsonsdisease.org.uk

NEWSLETTER

#wdsguk APRIL 2022

The Wilson's Disease Support Group - UK (**WDSG-UK**) is an all volunteer organisation which strives to promote the wellbeing of patients with Wilson's disease.

It publishes an annual newsletter with informative articles written by medical professionals, and also articles written by patients, their families and friends about their experiences of the disease.

It promotes networking of Wilson's disease patients and their families by helping and encouraging contact with one another.

And the Group strives to promote a wider awareness of Wilson's disease within the medical profession.

AFFILIATED TO :

BRITISH LIVER TRUST

The Liver Patients' Transplant Consortium

Children's Liver Disease Foundation fighting childhood liver disease

GENETIC ALLIANCE UK

RARE DISEASE UK

Just when we think we are emerging from the shadow of Covid that has dominated our lives for the past couple of years, something similarly frightening occurs. Wilson's disease affects people all over the world and no doubt Ukraine has it own community of patients. We can only imagine what they must be going through and hope that their management and treatment continue effectively during these troubled times.

Our own membership is at an all time high. According to current work carried out by NHS Digital over 600 patients have been identified in England. We believe that around a third of them are known to us either through membership of the Group and/or on Facebook. When newly diagnosed patients come to our attention, it is always gratifying to be able to offer them support. It is with particular thanks, therefore, that I should like to acknowledge the contribution that Dr Godfrey Gillett makes in helping us with these patients by offering practical guidance and often arranging to see them in clinic.

Thanks also go to our fundraisers Lizzie Martin, Laura Nicolson and Liz Wood for their sterling efforts, everybody who made donations during the year, those of you who found the time to join us on Zoom, Sam Shribman and Bill Griffiths for summaries of their work and Abby Morell for creating a newly designated Puzzle Page to replace my annual Dingbats! And last, but not least, I would like to thank Graeme Alexander for taking up the chair in our hour of need and supporting Wilson's patients everywhere through his various initiatives and Oliver Bandmann for effecting and overseeing the WD Guidance and Guidelines that had been proposed for so long.

Sitting down and writing a newsletter is not at all straightforward as it relies upon the generosity of all the people who contribute to it. In addition to those mentioned above, I should like to thank Linda for speaking out about her recent health issues and Grace, Laura and Danny for sharing their harrowing patient journeys with us. We wish them all the very best of health in the future.

It seems that reaching one's 100th birthday has become rather fashionable and this year it is the turn of James Kinnier Wilson, to whom we send our heartiest congratulations. Our other more seasoned centenarian, our President Dr John Walshe, entertained me recently and although not as physically robust as he once was, mentally he gave me a good run for my money - as you will learn later!

This coming weekend Phil Angus and his friend Kurt are taking on the National 3 Peaks Challenge, climbing Ben Nevis, Scafell Pike and Snowdon all in under 24 hours. One of their friends, Ben Ryan, has been dealt a cruel blow by Wilson's and in doing this challenge they are hoping to raise awareness of the disease at the same time as raising funds for us. We wish them well and thank them for their endeavour. We also thank new member, Guy, who has sponsored and printed WDSG-UK T-shirts for the event. We are hoping that these T-shirts might become commercially available to all very soon.

Meanwhile, membership fees are now due for 2022-23. Please see the form enclosed. If you are able to make your payment via internet banking (details on the form) either as a one-off payment or by setting up a standing order, that would be extremely helpful. Our bank has recently imposed over the counter charges for charity accounts but charges nothing for receiving payments by Bacs. Don't forget to send me your completed form, however, to keep our records up to date. Thank you.

Finally, we have decided to postpone a face to face meeting in Cambridge for at least another year, in order to protect our most vulnerable patients. We shall therefore be holding our AGM via Zoom again on Sunday, 3rd July at 1100 a.m. and hope you'll be able to join us. Have a very Happy Easter!

Valerie

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Chairman's Report for 2021-22

Living in a very small rural community must colour the way I see the world at present. Moving to Scotland was not a deliberate attempt to cut ourselves off from civilisation, simply an attempt to live in a location with scenery that takes your breath away. So, we were reliant on Facetime and Zoom to maintain contact with friends and family long before Covid hit and will remain reliant on the internet for so much in our lives. But Covid has not gone away and has had a much deeper effect on the Health Services than many realise now that the media has moved on. Hospitalisations are still high, staff numbers are ever lower both acutely and long term and without question many long serving health care workers are looking elsewhere or retiring earlier than planned. There are upsides: the drive to innovate with vaccines and specific antiviral therapies has been astonishing and applications from school leavers to enter the health professions are at an all time high. Clinical practice has changed with a reduced reliance on face-to-face contact (where appropriate). This is proving popular with physicians and patients alike, for whom the clinic appointment is not just 10 minutes with a doctor or nurse but a whole day generating transport difficulties, long waits for scans & prescriptions and especially true for those attending specialist clinics at great distance. More pertinent to the WDSG-UK, our online meetings have been positive. Undoubtedly members are finding meetings easier to attend, often with sound and images turned off (well why not?) and it seems to me, many are happier to contribute in this setting.

The Wilson's disease/British Association for the Study of the Liver was conceived as a means to link interested parties with distinct specialist interests and has proved very successful (see Bill Griffiths report later in this issue); nor has it been hindered by online restrictions. But medical groups thrive on the sort of questions and answers that are best raised face-to-face and perhaps discreetly and often for reassurance (questions such as 'would you have done it this way?') I suspect future meetings will be hybrids of zoom with face-to-face to ease personal links.

The national register of patients with WD was long overdue (see Osob Mohamed's update on ***p15***) and we should be very grateful that we have someone of Mary Bythell's calibre leading this important study, which now has international links. The first step is to collate data on as many patients as possible (in an anonymised fashion) then with time these data spell out important messages, but we have to be patient to see full value.

Plans for educational films are now in place and I had hoped to have started filming already, but this has been a difficult time for people to commit fully and my hope to have completed this by July 2022 was mis-guided.

In contrast, the UK WD community has just completed the most up to date guidelines on the management of WD and that long and detailed advice will be in press later this month and available to all clinicians very soon, regardless of speciality, or location in primary or secondary care. I was proud to be a contributor and I learned much but notably (1) that good guidelines can be written quickly and efficiently if you have the right people on board and contrary to my previous experience of writing guidelines and (2) that the UK WD community has some of the best young doctors I have met who will be looking after patients in the decades to come.

Finally, I would like to thank the members of the committee for their contributions to WDSG-UK over the past 12 months. We hope to see you at our Zoom AGM in July.

Graeme Alexander
April 2022

Donations and Fundraising

We rely exclusively on our members and their families and friends for our income. We should therefore like to thank all of you who made generous donations with your subscriptions last year and those of you who have set up regular monthly standing orders to us. A special mention is made to **Giuseppe Cardone** who raised £600 through Google nonprofits, **Claire Stapleton** and associates who separately raised monies on our behalf and **Ann Wilde** who gave a cash donation to mark the 50 year anniversary of the death of *Julie* through Wilson's disease.

We thank our fundraisers **Lizzie Martin, Liz Wood** and **Laura Nicolson,** who have navigated their way through Covid restrictions to bring in a further income of **£4,000.** Their enterprising pursuits are detailed on *pp6-7*. Should you wish to hold your own fundraising event, then please note that since VirginMoneyGiving ceased trading in November 2021, we have now registered with a new online fundraising platform called **InvestMyCommunity**, a charitable arm of Karadoo Financial Services of Bath. Their fees are minimal and like Virgin they will continue, when appropriate, to collect Gift Aid on our behalf.

Please remember that any donations to WDSG-UK can be Gift Aided, if appropriate. Gift Aid forms can be downloaded from our website or obtained from Valerie direct.

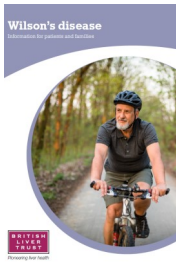
***Bank Charges—Please Note**

Our bank, HSBC, has recently introduced bank charges on charity accounts including for over the counter transactions. We are looking into preferable terms at other banks, but meanwhile, if you are able to pay your annual membership by bank transfer or even better set up a standing order, that would reduce the charges.

BASL WDSIG (British Association for the Study of the Liver Wilson's Disease Special Interest Group

The 6th BASL WDSIG meeting took place via Zoom on 19 November, full details of which can be found on *p14.*

The British Liver Trust



The British Liver Trust, which is an umbrella group for all liver disease patients in the UK, has recently produced a revised Wilson's disease booklet to which WDSG-UK has had input. It is an excellent publication and can be downloaded from their website https://britishlivertrust.org.uk/wp-content/uploads/WilsonsDiseasewebDEC21.pdf or alternatively a single hard copy can be ordered free by emailing info@britishlivertrust.org.uk.

NHS BT (Blood & Transplant) Organ Donation & Transplantation Directorate (ODT) and the Liver Patients' Transplant Consortium (LPTC)

In 2009 WDSG-UK attended NHSBT's first ever open meeting with representatives from patient groups in which its organ transplant programme was discussed. Thereafter, meetings were held annually and the number of Liver Patient Groups in attendance grew. Under the title of the Liver Patients' Transplant Consortium, we then met every year with NHSBT separately. The last such meeting took place via video link on 7 July 2021 where we were given statistical data on liver transplantation over the previous twelve months. Although all transplantation had been severely affected by the pandemic in 2020, numbers of liver transplants were now steadily returning to pre-pandemic levels. For a full transcript of the relevant statistical data from the meeting, please visit https://www.odt.nhs.uk.

Currently, NHSBT's researchers are proposing a study into liver donation and transplantation to improve the early function of a liver. They wish to evaluate three techniques to see which is better for patients. To help with this research, they are appealing to patients who have either had transplants or are on the transplant waiting list to take part in an online survey and/or in patient focus groups. If you are at all interested, please see *p15* for further details.

UK Liver Alliance & UK Liver Patients Alliance

In 2021 the UK Liver Alliance was set up to bring together the leading players in liver care with the aim of influencing policy and improving liver services and care across the UK. It is made up of the Chief Medical Officers of the 4 devolved nations, together with representatives from BASL (the British Association of the Study of the Liver), BSG (British Society of Gastroenterology), the Royal College of General Practice, the British Liver Nursing Association, the Liver Advisory Group, the British Liver Trust, the Children's Liver Disease Foundation and the newly formed UK Liver Patients Alliance of which WDSG-UK and other Liver Patient Groups are members. Our first meeting took place on 2 Mar 2022.

Cambridge Rare Disease Network (CRDN)

CRDN was set up in 2015 to address the challenges faced by people affected by rare diseases and to bring together patient groups, researchers, clinicians, pharma and biotech companies and policy-makers from around the world. WDSG-UK attended their 5th RAREsummit on 7th October '21, which was held online for the first time. It included live talks, short films, panel discussions, exhibitions, workshops and chat rooms to dip in and out of all day and a live Dragon's Den-style event where 5 innovative tech solutions to challenges posed by people living with rare diseases were pitched to a panel of experts and voted on by attendees.

EASL (The European Association of the Study of the Liver)

EASL is joining forces with ERN (The European References Network—Rare Liver Diseases) to produce their own European guidance document on the "Management of Wilson's disease." As a patient representative, Valerie has been invited to join a panel to give feedback on any proposed guidelines.

ELPA (European Liver Patients Association)

In Nov. '21, WDSG-UK was invited to talk to representatives of ELPA with an interest in rare inherited hepatic disorders about WDSG-UK's remit and about Wilson's disease in general so that they could learn how to offer better support to their own WD community.

Market Research on behalf of Pharma Companies

In 2021 several Medical Market Research Companies acting on behalf of Pharma Companies approached WDSG-UK looking for patients/carers to take part in incentivised telephone interviews speaking about their experience of living with WD. Thank you to all who took part.

WDSG-UK Management Committee Meetings

During 2021-22 the management committee met formally twice in May and January via Zoom and informally after our 11th Annual General Meeting in July.

WDSG-UK 12th Annual General Meeting

The 12th AGM will be held via Zoom on **Sunday, 3rd July 2022** at 1100 a.m, an invitation to which is included with the newsletter. The accounts for 2021-22 and the minutes of the 11th AGM will be emailed to you separately in due course. Election of officers and members of the WDSG-UK Management Committee for 2022-23 will take place then. Current members of the committee **Graeme Alexander, Mary Fortune, Liz Wood, Debbie Buckles** and **Valerie Wheater** have submitted their names for re-election for the coming year. If you would like to join the committee, please write to Valerie.

The Wilson's Disease Patient Register—UK

WDSG-UK supports research into all aspects of Wilson's disease. If you haven't yet signed up for the Register, please consider doing so. A pamphlet and information sheet are available to download off our website, together with the corresponding registration form.

WDSG-UK Fb Site

Our WDSG-UK Facebook Group is a great resource for keeping up to date with what's going on at WDSG-UK headquarters. It is a private group so only members can read posts. If you aren't already a member, why not give it a go?

Wilson's Disease Support Group Meeting & 11th AGM
Sunday 18 July 2021 via zoom

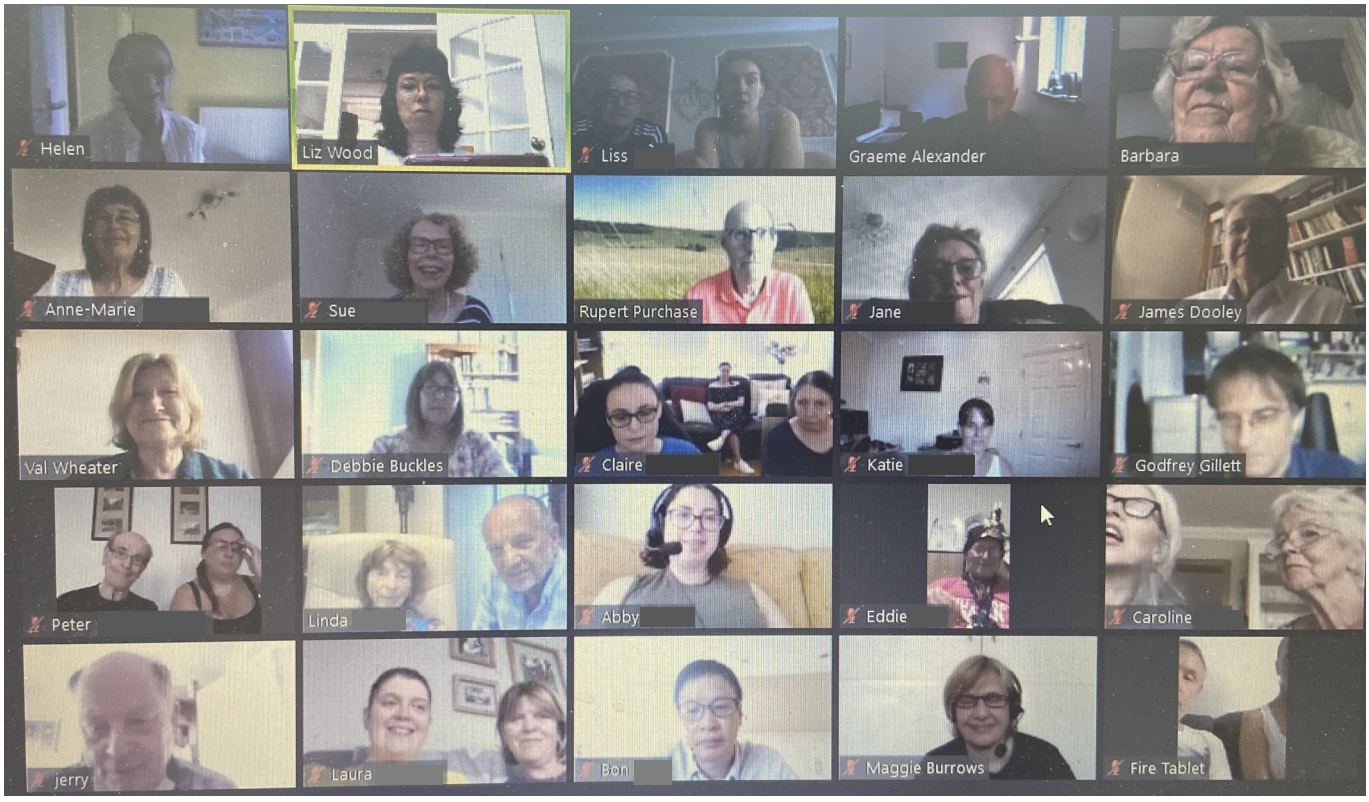
Thanks to everybody who attended the virtual AGM last July. Invitations were sent out to members with their newsletters and we were very pleased that 32 people *Zoomed* in to join us. In particular, we were grateful to **Dr Godfrey Gillett, Dr James Dooley** and **Miss Maggie Burrows** for giving up their Sunday morning to be with us. The meeting was managed by committee member **Liz Wood**, and our chair **Graeme Alexander** hosted it.

After the formality of signing off the previous AGM minutes and receiving apologies for absence, Graeme gave us a short report of the Group's activities over the previous 12 months. He highlighted that a sub-group of doctors under the umbrella of the BASL WD SIG had been set up to draw up guidelines relating to the investigation and management of patients with Wilson's disease and that these guidelines, when finished, would be submitted to medical journals and published on appropriate medical websites. He also said that once the pandemic was under control, he hoped to take up the Pharmaceutical Company, Orphalan's, offer to produce a series of short films highlighting WD patients' histories. Interest in participating was expressed by 4 members present.

The accounts were scrutinised and members were thanked for continuing to renew their subscriptions, making donations and fundraising for the Group. Committee members for 2021-22 were proposed and seconded by **David Chiswell** and **Caroline Barr** respectively, after which members were given the opportunity to ask specific questions of the committee. Maggie updated us on how the CROWD Study at UCL was going and informed us that an article recording preliminary data collected from the patients' first visit to UCL had now been published in the medical journal *Brain*.

After the formalities of the meeting ended, Liz arranged for patients to be split into smaller *chat rooms*, each of which was kindly hosted by a doctor. This gave members the opportunity to catch up informally with one another and also share any specific concerns they had about their management or treatment.

Graeme brought the meeting to a conclusion at 12.10 pm by thanking Liz for facilitating it through her Zoom account. He also thanked members for attending.



Screenshot of some of the Attendees at the 11th AGM

WDSG-UK Virtual *Coffee Morning*
Sunday, 27 February 2022

After the success of last year's Zoom Coffee Morning to mark **Rare Disease Day 2021,** the committee decided to repeat the event this year. The first Rare Disease Day was introduced by the European Organization for Rare Diseases and took place globally in 2008 — on 29th February — a rare date that only happens once every 4 years! When it isn't a Leap Year, Rare Disease Day takes place on the 28th February instead. Its main objective is to raise awareness amongst the general public and policy-makers of the impact that living with a rare disease has on patients and their families. Events such as social gatherings, fundraisings, art exhibitions, workshops and press conferences are held across the country and parliamentary lobbying is encouraged. Wilson's disease is only one of 7,000 different rare diseases and WDSG-UK believes a good way to mark the occasion within its own community is through mutual support over a morning cup of coffee!

Attendees at The Coffee Morning marking Rare Disease Day 2022

This year invitations to our Coffee Morning were restricted to current members of WDSG-UK, to Facebook members of WDSG-UK who expressed an interest in attending and to doctors who are closely affiliated to the Group. Thirty-six people signed up to the day and most were able to join us. Our special thanks go again to **Dr Godfrey Gillett, Dr James Dooley** and **Miss Maggie Burrows** who generously made themselves available and it was particularly gratifying to welcome the Group's co-founder, **Linda Hart,** who has been battling with a blood disorder for the past 2 years, but has thankfully got the better of it now. She writes about it on *p20*.

Hosting a virtual event like this is challenging, but **Liz Wood** has spent the last couple of years of Covid restrictions running various choirs and music events from home and she deftly navigated us through this one. Once everybody had arrived and been welcomed, we were divided into four pre-assigned chat rooms that were hosted either by a doctor and/or committee member. Much as it would be interesting to talk about politics, world affairs or the weather, conversation invariably dwelt on medical matters in relation to our own experiences of living with Wilson's disease.

After 20 minutes we were randomly allocated to different chat rooms before coming back together for a fond farewell. We hope to hold these Zoom meetings more frequently, so please make sure that we have your up to date email details, so you can receive an invitation well in advance.

Fundraising 2021-22

Valerie

Lizzie and Kristen's Ascent of Snowdon

On Saturday, 31st July last year, **Lizzie Martin** from Bridgend in South Wales and her friend **Kristen** took on the challenge of walking up Snowdon. She did so to raise funds for WDSG-UK in memory of her late father, Gavin Martin, who was diagnosed with Wilson's disease in 1993 when Lizzie was just one. Gavin sadly died in August 2009.

Lizzie

In preparation for the *big* day, Lizzie did several climbs nearer to home including one with her partner, Daryn, up Pen-y-fan in the Brecon Beacons, the highest peak in Southern UK.

On the day, the weather wasn't terribly good so Lizzie and Kristen decided to walk along the Llanberis path, the longest but also one of the safest routes up Snowdon. Even that was quite a struggle and they were relieved 6 hours later to have reached the summit. Unbeknown to Lizzie at the time, she was already 3 weeks pregnant!

Reaching the Summit

Through sponsorship, Lizzie and Kris raised just under **£600** for the Group (including Gift Aid) and we thank them for their endeavour, and those who sponsored them for their very generous support. Meanwhile, we wish Lizzie and Daryn all the best for the birth of their first child at the beginning of April.

Southend HSG Chamber Choir Reunion – Sept '21

Liz Wood joined the WDSG-UK committee back in July 2018. She comes from a highly talented musical family and teaches the piano and singing. Her older sister, Joy, died from Wilson's disease in 1974 aged 17, some eight weeks after diagnosis. Shortly afterwards, Liz was diagnosed with it, too.

She writes, "I taught singing at Southend High School for Girls for 11 years from 2010 and for the last 8 years of that time, I also ran a Chamber Choir with 30 girls, who had to audition to become a member. We gave many concerts and took part in several competitions, winning quite a number of trophies and cash prizes. In July 2021 I left the school, as I was going to be moving out of the area. Before I left Southend completely, I organised a reunion day for all the girls/young women who had sung in the choir since its inception. We had a fabulous day singing lots of the repertoire that they had enjoyed during their time in the choir and for the last hour, their parents and friends were invited to come to listen to an informal concert. At the end of the concert we had a collection in aid of the WDSG-UK, raising a wonderful **£285** for the group."

Liz conducting the Choir for the Informal Concert

Laura's Cycling Challenge — Summer 2021

You may remember in our 2020 newsletter the harrowing story of **Laura** from Shetland whose diagnosis of Wilson's disease in 2019 took over four years to make. Unfortunately for Laura, starting medication only made her condition worse. She ended up being hospitalised in Aberdeen as she became severely dystonic and could no longer walk, talk or feed herself. Under the guidance of Dr Godfrey Gillett, dimercaprol injections were administered and miraculously she slowly started to improve. She has gone from strength to strength ever since.

Laura

To mark her 33rd birthday in July last year, she decided that she would spend the summer cycling around Shetland raising funds for charities close to her heart and at the same time raising awareness of the condition across the whole of Shetland. She was interviewed on local radio and gained much publicity through that. Her initial aim was to raise a total of £600 to be shared between the Neurology Ward that treated her at Aberdeen Royal Infirmary, the Woodend Neuro-Rehabilitation hospital to which she was sent to recuperate afterwards and WDSG-UK which had supported her and the family throughout.

Laura and her Mum

In preparation, she started running 5k distances again and doing 9k walks. She then set out on her birthday to complete the first of 25 different cycle routes all across Shetland, using an app on her 'phone to record the distances covered. She was often joined by friends and family, which she greatly appreciated particularly when the terrain was tough and the weather challenging. She hopped on and off ferries to reach the far corners of the Isles, completing her longest ride of the challenge (31 miles) on Yell, the second most northerly island in the British Isles. Not content with focussing on cycling, she even fitted a triathlon in two weeks into the challenge! By the time she completed her last ride two-and-a-half months later, she had covered a total of 421 miles. It is all the more remarkable when you consider that two years earlier and with the use of a stick, she could walk no further than 100 metres on her own!

Another one for my bucket list!

The £600 target that she had set herself was quickly reached and she went on to raise an incredible **£10,500** altogether. Thank you Laura for your cheque for **£3,118.43** for WDSG-UK. It is a truly astonishing sum. Thanks, too, to all who accompanied Laura on her travels and who encouraged and sponsored her through her Just Giving page. It is very much appreciated.

Valerie's Demon Dingbats - A Staycation in the UK

The 2021 Dingbats related to coastal towns and villages around Great Britain and were reportedly harder than ever! In addition to those that were sent free to members, a further 175 copies were sold to the general public and 28 were returned for marking! There was an outright winner with 40/42 correct answers followed by three runners-up scoring 39. Thanks to everybody who took part and congratulations to **Liz Wood** for her winning entry. She received a **£25** cash prize. Through the sale of the Dingbats plus some generous donations, **£450** was raised in total.

I have decided to take a break this year so apologies to anybody who is disappointed. However, for those who enjoy puzzles, member Abby Morell has very kindly put together a Puzzles Page, which you can find on *p22*. No cash prizes, I am afraid, but I hope it will offer you a little light entertainment. Feedback is welcome.

ANSWERS: Anagrams 1 Trifle **2** Champagne **3** Cucumber sandwiches **4** Strawberries and cream **5** Hog roast **6** Victoria sponge **7** Pimms and lemonade **8** Coronation chicken; **Dingbats 1**Windsor (w-in-dsor, **2** Conwy (C-on-Y), **3** Highclere (high clear), **4** Bolsover (slob over/backwards), **5** Ludlow (Lud-low), **6** Arundel (a-(a)round-L), **7** Brownsea (a brown letter C), **8** Stormont (Storm-on-T), **9** Sterling (£ Sterling); **(Cards on the Table** (from l-r) 6 K 9 A 8 Q 4 J 2 7 10 3 5

Grace's Story

by Grace Carter

I was born in 1993, the youngest of four children. I struggled with my physical and mental health from a very young age. I was extremely sensitive and found both my internal world and my external surroundings totally overwhelming. My emotions always felt too intense and I did not know how to regulate them. I also found social situations impossible to understand or safely participate in. This caused me to be very isolated and form a negative and warped view of myself and others. I also experienced eczema and recurring thrush (both of which rarely improved with treatment), insomnia, and extreme sensitivity and aversion to touch around my stomach with pain around my liver. I also had early sexual development, with small breasts from the age of 2. I now know all of these symptoms were early indicators of Wilson's Disease.

As I moved into my teenage years, emotional regulation and social contact became even harder. Between ages 13-15, I experienced several traumatic events. I was depressed and anxious and found every aspect of life overstimulating and overwhelming. All childhood symptoms persisted and I began having daily headaches, frequent migraines, fatigue, brain fog and confusion, nausea and vomiting, bloating, and excessively heavy periods that would last for months at a time. At age 13, I began contemplating suicide and made one attempt.

At age 14, I became unwell with flulike symptoms and spent weeks off school trying to recover. I then made several attempts to return to school but could not manage even one day. My GP believed glandular fever had caused post viral symptoms and diagnosed me with ME/ Chronic Fatigue Syndrome (CFS). My GP did not offer any support, treatment or referrals for my new diagnosis. They did note that my liver function tests were abnormal but believed this to be due to CFS.

Over the next four years (age 14 – 18) my health declined further and I was unable to get out of bed. I experienced extreme fatigue, eczema and skin rashes, acne, brain fog and confusion, depression, anxiety, nausea and vomiting, bloating, intense sustained pain around my liver, and my periods stopped. My GP continued to be of no help and my mum became my sole carer. We were not aware that we could claim social security during this time and the council denied our application for practical support. This meant my mum continued to work and look after me – two full time jobs. My mum's health worsened throughout this time and we suffered financially. We could not afford heating and had to live off the basics which added stress to an already very stressful situation that seemed to have no end in sight. Because my mum was so busy trying to keep a roof over our head, I spent almost all of my time alone. I was extremely isolated and lonely.

Over my four years in bed, I became severely depressed. I tried to connect with old school friends via Facebook but this only highlighted how isolated I was and emphasised feelings of loss for the life I was missing out on. The constant dismissal of my ill health by my GP and teachers caused me to doubt myself. No one seemed to believe me. I felt extremely ashamed of my situation and believed it was my fault. I thought I must be making it all up, that I was not actually poorly, just lazy.

> ***"She said I had 5 weeks left to live"***

As my illness progressed, I was too ill to wash myself or get to the toilet and back by myself and this made me even more ashamed. My liver function tests continued to get worse. My GP continued to say I had CFS and my only recommended treatment was to stand against a wall for a progressively longer time every day, starting at 5 seconds, working my way up to 5 minutes.

Eventually, something unknown to my mum or me occurred, and my GP finally paid attention. With no warning, she came to our house, jumped over our garden wall and screamed at my mum. She said I had 5 weeks left to live, and falsely accused my mum of preventing me from seeking medical help! This was all news to us!

Over the next few weeks, I saw the Hepatology Team at Derriford Hospital, Plymouth, and was given a preliminary diagnosis of autoimmune hepatitis. An alternative, much rarer and therefore less likely diagnosis was Wilson's, for which I did a 24 hour urine test, blood tests for caeruloplasmin and serum copper, and had a slit lamp examination by an ophthalmologist. All tests confirmed a diagnosis of Wilson's. In August 2010 I was put on 1200mg/day of trientine dihydrochloride and told to avoid high copper foods such as chocolate, organ meats and shellfish. I was told by doctors that I would lead a normal life once I began taking trientine.

Soon after starting trientine, I was able to get out of bed, wash myself, and drive a car. My health was still very poor but I was determined to escape my bedroom and try to make up for lost time. In September 2011, age 18, I moved away from home to attend college and do my A levels. I also had a part time job. This was a turbulent time but determination got me through and I very proud-

ly achieved A*s and applied to read Law at Cambridge. In the run up to my interview, I began working full-time as a waitress but soon became unwell and moved back to my mum's.

In 2014, four years on from diagnosis, my mum found WDSG-UK on Facebook and Valerie told us about Dr Dooley, a Wilson's specialist in London, who thankfully then became my doctor. Though this was not the end of my struggles, it was a real turning point for me. Dr Dooley said I needed psychotherapy to help me come to terms with diagnosis of a life-long illness. Unfortunately, there seemed no way to obtain this via the NHS.

Due to past doctor's statements, I still believed I should be living a normal life now I was taking trientine and did not understand why this was not happening. Desperate to get on with life and seeing no resolution via medicine, I moved to London to attend university. I tried my best to pretend my symptoms did not exist but could not sustain this for very long. I experienced a traumatic event, rapidly reached crisis and made two more suicide attempts. In December 2015, while in A&E, I finally got a diagnosis of Borderline Personality Disorder (BPD). This was a huge turning point for me.

I suspended university and moved back home to focus on my physical and mental recovery. There were no appropriate NHS mental health treatments available so I began seeing a private psychotherapist 2 – 3 times per week, at great cost. My therapist explained to me that I had something called C-PTSD/complex trauma, in large part due to the four bedbound years pre-Wilson's diagnosis. This was another huge turning point for me. I got a therapy dog called Isabella (see photo/Isabella Buckeeth on Instagram) and put all my energy into recovery.

The next few years continued to be challenging but now, because of my psychotherapist, I had hope that things could finally change for the better. I was extremely fortunate to receive partial funding for my psychotherapy via a Personal Health Budget. This funding enabled me to access the treatment I so desperately needed and I will forever be grateful for this.

Some strange Wilson's symptoms became severe during this time. Most notably, issues with pupil dilation that caused partial/total blindness in my right eye and fuzziness in my left eye. This was debilitating and is thankfully much milder and less frequent today due to systematic kinesiology, zinc supplementation (well apart from my chelation medication), and daily yoga practice.

Though I have always struggled with sleep issues, these symptoms became increasingly worse from 2017 – 2020. During this time, I seemed unable to fall asleep without sedating medication and whenever I did fall asleep, I experienced recurring nightmares, night terrors and sleep paralysis. I would often be awake for at least 48 hours at a time and getting 3 hours of sleep a night would be a huge achievement for me. This caused all facets of my health to decline. I had pain all over my body and was diagnosed with fibromyalgia. I had frequent panic attacks, muscle weakness, severe fatigue, brain fog, confusion, and memory loss. Often, I could not hold a conversation because I would forget the topic or what I planned to say. All these symptoms reminded me of how I felt during those four years before my Wilson's diagnosis and I feared I would become bedbound again.

Isabella and Grace

In July 2020, I researched the link between Wilson's disease and insomnia, in the hopes of finding an explanation for my sleep issues. I found multiple academic journals on the subject, including one that had trialled melatonin (the hormone in charge of regulating the circadian rhythm) as a possible form of treatment for Wilson's. A few days later, I tried my first dose of melatonin and fell asleep within minutes. Life has changed since then. Sleep is not perfect, I wake up every hour or so through the night, but thanks to melatonin, I can fall asleep with ease and usually sleep for an amazing 5 – 9 hours.

At the beginning of 2021 I successfully switched to the new, non-refrigerated trientine tetrahydrochloride (600mg/day). I am religious about taking my medication and have not missed a single dose since I began taking it.

Although things are much improved, I still find living with Wilson's disease extremely challenging as it continues to affect my quality of life on a daily basis. Over the past few years, I have found freedom in learning that my symptoms may never disappear fully, as this has enabled me to begin aiming for what's possible and sustainable for me, rather than striving for the life of someone without my experience of Wilson's. Psychotherapy has been very successful and should come to a celebratory end within the next year. I have been so inspired by my experience that I am currently studying to be a psychotherapist. I am also creating a blog about recovery: ***Finding Grace*****,** and YouTube channel: ***Meditate with Grace***. Though life hasn't been easy, I never forget how fortunate I am to be alive, and how fortunate I am to have the health that I do have.

Thank you to Dr John Walshe for discovering chelating medication, to the NHS for providing my medication, and to Drs. Mitchell, Dooley, and Tsochatzis for their outstanding care and compassion, and thank you to all those who contribute to the work of WDSG-UK.

Research Update—The CROWD Study

The CROWD Study, UCL Queen Square Institute of Neurology, London — Update

The CROWD (Cohort Research On Wilson's Disease) Study was launched in December 2018. In Part 1, the aim is to identify genetic factors that determine whether someone with Wilson's disease will develop neurological problems or not. People across the UK were invited to participate by completing an online questionnaire and sending in saliva samples. The last few samples are now being collected ahead of starting the genetic analyses.

In Part 2 of the Study, the aim is to understand how to measure and monitor the effects of Wilson's disease on the brain. Forty patients attended Queen Square for clinical assessments, blood tests and MRI brain scans in 2019 and this part of the study is now closed. Some findings have been discussed in last year's newsletter. In this article, **Dr Samuel Shribman, Miss Maggie Burrows** and **Professor Tom Warner** discuss some of their work on MRI. Please do not hesitate to contact them at s.shribman@ucl.ac.uk, if you have any questions about their research.

What can MRI tell us about neurological involvement in Wilson's disease?

Magnetic resonance imaging (MRI) is an important tool for diagnosing and monitoring a number of conditions that affect the brain. Abnormalities in the deeper structures within the brain, particularly the basal ganglia, are often seen in patients with Wilson's disease and can be helpful in making a diagnosis. However, our understanding of how to use MRI scans to predict recovery or monitor treatment for Wilson's disease is limited.

Our research team aimed to better understand the significance of imaging abnormalities in Wilson's disease as part of the CROWD study. We recently published our results in the medical journal *Brain*. Here, we explain our key findings after introducing how MRI works and how it can be used in clinical research.

How does MRI work?

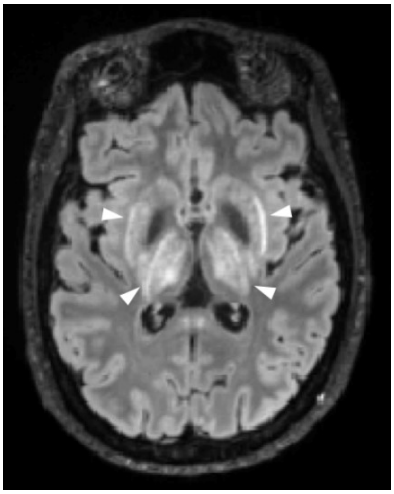
MRI scanners generate 3D images by measuring the physical properties of water molecules spread throughout the brain. Different *sequences* can be used to investigate specific aspects of brain tissue. For example, some sequences inform us about the structure of the brain whereas others tell us about the presence of inflammation or iron accumulation. Measuring the free movement of water molecules can also be used to assess the health of brain cells.

How can MRI be used in clinical research?

MRI scans performed in the NHS are usually interpreted by radiologists who review each sequence by scrolling through 2D slices of the brain and then making a visual assessment. This is a quick and effective way for identifying obvious abnormalities. However, radiologists cannot easily measure the volume of specific structures or abnormalities in the brain and it is difficult to compare more than a few scans simultaneously. Answering a research question like 'What is the best way to use MRI to predict recovery in Wilson's disease?' requires data from a large number of patients in combination with sophisticated software, mathematical modelling and statistical testing.

What was already known about MRI in Wilson's disease?

Several studies over the last few decades have shown that the majority of patients with neurological symptoms, and many patients without, have abnormal brain scans. The most common finding is bright spots in the basal ganglia and other deep brain structures. These are shown in the image on the right which is a cross-section of the brain (with the eyes at the top). The basal ganglia, which are marked with white arrowheads, appear brighter than the rest of the brain.



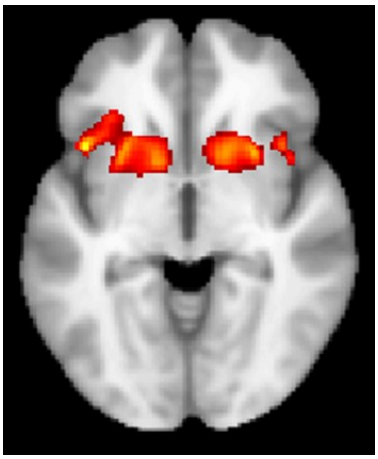
In the last few years researchers have started to use more sophisticated analyses to directly measure the volume of specific brain regions. They have shown the volume of the basal ganglia tends to be lower in patients who have neurological symptoms. Other researchers have measured abnormalities in the free movement of water or iron deposition and found some interesting results but, again, focussed on specific brain regions only. Few of these studies tested whether imaging abnormalities were associated with neurological severity or copper levels.

How did our study work?

We asked 40 people living with Wilson's disease to attend the National Hospital for Neurology and Neurosurgery for a day. We classified them according to whether they had neurological or hepatic presentations of Wilson's disease and whether their disease was well controlled or not. All participants had a neurological examination (to measure the severity of speech problems, shaking, slowness, etc), blood tests (to calculate copper levels) and an MRI head scan. We then used a combination of advanced imaging software and statistical techniques to compare every image pixel by pixel between different groups of patients. This allowed us to systematically test subtle changes across the entire brain that may not be visible to the naked eye or when focussing on one brain region only. We also used the same *whole-brain* approach to identify which changes correlate with movement problems and copper levels in patients with well controlled Wilson's disease, most of whom had been taking chelation therapy for many years.

What did we find?

Firstly, we confirmed that patients with neurological presentations have smaller basal ganglia volumes than those with hepatic presentations. We then showed that people who have more severe movement disorders after many years of treatment tend to have lower basal ganglia volumes. This is shown in the image on the right. Areas that our statistical analyses predicted are associated with the severity of movement disorders are mapped on to an image of the brain in red and match up with the basal ganglia.



We suspect that damage to basal ganglia that occurs prior to starting treatment leads to loss of neurons and therefore smaller volumes. Our findings support the idea that neurological recovery depends on the extent of this initial injury to brain and that measuring basal ganglia volumes may help predict neurological recovery.

We found that the amount (i.e. volume) of bright spots in the basal ganglia was higher in patients with uncontrolled disease. However, there was no association with movement problems. This means that while bright spots may be helpful diagnostically they do not necessarily tell us about how the brain is responding to treatment in the longer term.

Next, we looked at the free movement of water molecules. This was higher in patients with uncontrolled disease and increased as blood copper levels increased. We don't fully understand what causes these changes in Wilson's disease but our findings suggest that measuring these may be a useful way to monitor response to treatment.

Finally, we looked at iron deposition in the brain. We confirmed previous findings that iron deposition is increased in the basal ganglia of patients with neurological presentations. However, we also showed that increasing severity of movement disorders was associated with increased iron deposition in widespread brain regions. Handling of copper and iron is closely linked at a cellular level in the brain. We suspect that people who struggle to recycle iron when they accumulate copper in brain cells are more likely to develop movement problems.

Why do our results matter?

There are several important implications for our findings. Firstly, we have shown that measuring the volume of the basal ganglia might help predict neurological recovery. However, some of our analyses took hours (and in some cases days), even using high performance computers. Further work is needed to find ways to indirectly measure basal volumes more quickly. Understanding that bright spots don't necessarily correlate with movement problems is important because specialists had previously assumed that monitoring these might be helpful.

We have provided evidence that measuring the free movement of water molecules could be useful for monitoring how the brain responds to treatment. The next step here is to look at how these abnormalities change over the first few years of treatment on repeat scans. Finally, our observations on iron accumulation have shed light on the link between copper and iron metabolism in the brain and how this might be integral to understanding why only some patients develop neurological problems.

We would like to conclude by thanking all of the patients and their family members who took part in the study, in addition to the Wilson's Disease Support Group—UK, Association of British Neurology, Guarantors of Brain and Reta Lila Weston Institute, all of whom supported the study financially.

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Laura's Story

by Laura Farley

Hello—I'm Laura! I have been invited to write about my experience of living with Wilson's disease.

I was a healthy child up until age 11 years with only the usual childhood infections and an appendectomy aged 10. As I transferred to secondary school my energy levels dropped, my joints began to ache and my motor skills changed. I seemed to become a little clumsier and my handwriting deteriorated. I became less active and lost interest in sports.

My father had just been diagnosed with rheumatoid arthritis and since my symptoms were in some ways similar – joint pain and lack of energy — my parents referred me privately to a rheumatologist. No diagnosis was made and rheumatoid arthritis and lupus were ruled out via blood tests. I was, however, prescribed anti-inflammatories to relieve the joint pain and didn't participate in most PE lessons.

In 1994, after returning from a visit to Malaysia, I was hospitalised with campylobacter. Whilst I was in hospital my enlarged spleen was noted and this led to further investigations, eventually involving a liver consultant, Dr Trowel. She had seen one other person with WD in her career and under her direction I undertook a special eye examination at the Oxford Eye Hospital. My eyes revealed a clear ring of copper around the irises (Kayser-Fleischer rings), which confirmed the diagnosis of Wilson's disease. Photographs of my irises have been used in books to illustrate WD.

No one in my family had ever heard of WD and it was a complete shock to everyone. I am an only child, but I have a cousin who was tested and he was found not to have it. I was told WD was a lifelong condition, that I would need daily medication to remove the copper from my system and regular health checks.

I had to start the regular pattern of 24 hour urine sampling and regular blood tests and ultrasounds, and started taking penicillamine daily and pyridoxine (vitamin B6) weekly. I was lucky to have a specialist experienced in WD. I have also had good support from my GP practice which I have remained with since birth. My liver consultant since my late teens has been Dr Collier, who has also been tremendous.

My puberty was delayed and I resented being different from other girls of my age. This lowered my confidence and self esteem. Although academically quite able, my concentration was not as good as it had been when I was younger. I needed structure and support to achieve well. My school was supportive of my needs. I was given extra time and a computer when doing my GCSEs and A-levels examinations and I sat them in a solitary environment.

I remember my disappointment that due to some concerns about my medical condition, it was decided I could not participate on a school visit to our twin school in Uganda. I did go on an exchange visit to Spain. I started my periods after I left secondary school aged 18, and was somewhat emotionally immature compared to my peers.

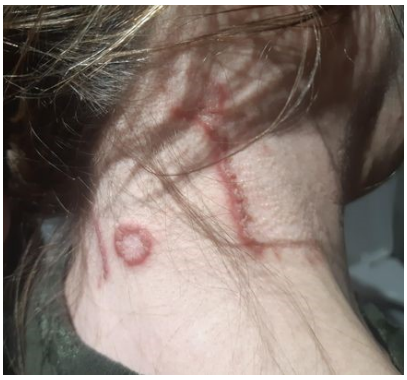
I got good A level results and went off to Reading University to study Psychology – a subject I had enjoyed at A-level. However, the first year at university was heavily science based which was not my strongest area and I lost confidence and motivation. The freedom of university did not fit my need for structure and routine. I went off the rails with the lack of structure, got heavily into the party scene and left after one year.

> *"...bipolar is linked with Wilson's disease"*

I suffered from depression and had relatively low moods throughout adulthood but could also be impulsive and erratic at times. I was prescribed various antidepressants until I was diagnosed with bipolar in 2015 after I was sectioned, due to psychosis. I now take daily mood stabilisers (lamotrigine) and my mental health has been stable for the last 4 years. Interestingly, I understand bipolar is linked with Wilson's disease.

I also developed EPS* (elastosis perforans serpiginosa), around my neck in my mid 20s which is believed to be a side effect of taking penicillamine in 1% of people. This took a long time properly to diagnose and baffled doctors as to how to treat it. I changed medicine from penicillamine to zinc acetate, but I struggled with nausea on zinc, so then changed to cufate/trientine in 2018. Despite various creams and steroids my skin showed no improvement. Heat ablation (previously untrialled on EPS) was excruciatingly painful and if anything, made it worse! The condition is very visible and

has caused me much distress over the years. I have grown to accept it and now feel less self-conscious than I used to, and sometimes use camouflage makeup to conceal it.



EPS on Laura's neck

The skin is sore but manageable and the condition has been known to go into remission, so I am still hopeful that it will one day clear up. It has improved slightly over the last year and is less inflamed, more so since not drinking alcohol. Herbal balm is the only thing I have found to be effective at soothing the irritation. I also take prescribed antihistamine to help with inflammation and itching.

Despite having WD and being advised about the dangers of alcohol, I have been a heavy drinker the majority of my adult life and was diagnosed with cirrhosis in my mid-twenties. I have yearly endoscopies to check the varicosities in my throat and have an ultrasound on my liver every 6 months. I also have fibroid tests and do annual 24 hour urine collections. My latest ultrasound in October 2021 showed a 2 cm grey lesion, which was confirmed to be cancerous after an MRI and CT scan. I had heat ablation under general anaesthetic to remove the tumour in early January this year. The operation was a success and the tumour was successfully removed. I stopped drinking alcohol before surgery and am continuing with alcohol-free living to give my body the best chance of recovery (and in case I may need a liver transplant in the future).

Until joining the WDSG-UK Fb Group recently and attending the *Rare Disease Day* Zoom call with members, I had never met anyone else with the same condition as me. It's nice to not feel so isolated. I am 40 this year and am finally starting to take better care of my health, which is long overdue! I feel lucky to have made it this far and am very grateful to my amazing parents for their love and continued support throughout my life.

* * * * *

* Elastosis Perforans Serpiginosa (EPS)

The late Dr Alan Stevens, a retired consultant pathologist specialising in dermatopathology (skin diseases) wrote an article in our 2017 newsletter explaining the pathology of EPS. The full article can be found on the WDSG-UK website <www.wilsonsdisease.org.uk> in either the *Newsletter* or *Medical Articles* section.

Here are a few facts about EPS drawn from the article:

- **I**t can occur spontaneously, with no obvious cause or association with other disease ("idiopathic EPS");
- **I**t can occur in association with other disorders such as Ehlers-Danlos syndrome, Marfan syndrome and Down's syndrome;
- **I**t can occur as a complication of some drug treatments, including penicillamine which is used to treat Wilson's disease in addition to rheumatoid arthritis and cystinuria;
- **I**ts incidence is thought to be related to the dose of penicillamine (more common if the dose is greater than 1 gram per day);
- **I**t is said to occur in 1:100 Wilson's disease patients taking penicillamine;
- **T**here may be a genetic predisposition to it as it may be linked with idiopathic EPS
- **I**t is caused by damage to and disruption of the elastic fibres in the dermis of the skin;
- **I**t usually presents as a cluster of small, raised, often reddish nodules about 3-5 mm in diameter, arranged either in a line, circle or in a wavy snake-like line;
- **T**he commonest site for EPS to occur is the back of the neck, but it can also occur on the side and front of the neck, elsewhere on the face, occasionally on the arms (around armpit) and on the legs (upper leg/thigh).

NB: Treatment for EPS in the past has included creams, heat ablation and/or freezing. However, Dr John Walshe (who looked after over 320 Wilson's disease patients during his career) recommends leaving well alone (including avoiding biopsy, since that may make matters worse). In his experience the lesions eventually heal by themselves.

WDSG-UK Notices & Updates

The BASL Wilson's Disease Special Interest Group (WDSIG)

Through the British Association of the Study of the Liver (**BASL**) a number of Special Interest Groups (**SIGs**) has been established in certain areas of liver disease in order to bring relevant expertise together from around the UK. The **WD SIG** was one of the first to get going and continues to attract more clinicians to its membership. It had its first meeting in December 2017. Wilson's disease lends itself well to this new initiative, being a rare disease that has several types of clinician involved in its care. Sitting in the Group are hepatologists, neurologists, clinical chemists, paediatricians and psychiatrists as well as research scientists and representatives from WDSG-UK and NHS Digital, formerly Public Health England (**PHE**). Over 100 individuals from across the UK are registered on the WDSIG database and the SIG has now approved 15 specialist WD adult centres across England. For further information on the remit of the WDSIG and minutes of its meetings, please visit the BASL website www.basl.org.uk.

6th Meeting—19.11.21 via Zoom—Attended by over 40 members

Patient Management: First on the agenda was the national trientine pathway. Paul Selby, pharmacist from Cambridge, presented the findings of an online survey of specialist centres to see how patients are managed. Responses were received from 12 specialist centres (10 Adult/2 paediatric). Some findings were: 75% of centres have no formal guideline for shared care, annual review is completed using variety of mediums with 75% face-to-face, 66% of respondents felt home monitoring to be somewhat or very valuable, 80% of trientine being prescribed is in the form of Cufence and 20% as Cuprior. Discussion revolved around the importance of face-to-face review especially from the neurological perspective. Cambridge is setting up a pilot for 'home' blood and urine monitoring for Cufence.

WD Guidance Document: This has now been accepted for publication in Lancet Gastroenterology and Hepatology which means it will gain international attention. This is a very successful output from the WD SIG and we hope to put the full document up on the BASL website soon. Sam Shribman and Tom Marjot summarised the workflow over 2021 and produced the final document which includes an informative pictorial summary. The guidance will be disseminated in due course via the various specialist societies.

Research progress and PHE WD Pilot Study: Sam Shribman began with an update on the CROWD study and some exciting neuroimaging findings in WD where whole brain methods using magnetic resonance imaging (MRI) can distinguish particular patterns. The findings were accepted for publication in the prestigious journal *Brain.* Mary Bythell, our Public Health England (PHE) expert, explained how her team has become incorporated into 'NHS Digital'. This has some advantages for data collection and research. Osob Mohamed, who has been working on the PHE project, explained that data sharing is now set up with 49 hospitals, 28 of which have provided all their cases thus far. Other data sources include copper and genetic laboratories, death certificates and GP prescriptions. There are now >600 registered cases and ongoing work is linking the various data sources.

At the start of the late morning session, we discussed planned educational videos which Graeme Alexander is spearheading. Graeme and Sam Shribman are working on an education tool for GPs and a particular focus of the SIG is to improve primary care awareness.

Pharma Trials: Aftab Ala discussed the various pharma trials in WD which are ongoing or due to start. For bis-choline tetrathiolmolybdate there was a previous completed study which showed efficacy and an acceptable safety profile, a related copper balance study which has enrolled several UK patients and an ongoing phase 3 study where this drug is compared with standard treatment with results awaited. Jan Coeburgh later discussed a clinical case from one of these studies. The CHELATE study which is Cuprior vs penicillamine has just finished recruiting. There are two gene therapy studies (UX701 and VTX801 are the drugs) hoping to recruit this year and King's and Royal Surrey will be host centres for these. Professor Ala also discussed research into dynamic monitoring of copper distribution using 'PET' imaging, which he will explain in further detail at the next meeting.

Future Projects: James Liu Yin, liver research fellow at Kings, discussed a psychiatry project looking at new referrals via the Maudsley hospital. The aims are to see if there are 'missing' diagnoses amongst this cohort and whether diagnosis is significantly delayed. Frank Proudlock, Assoc Professor in Ophthalmology at Leicester, gave an introduction on optimal coherence tomography (OCT) which can potentially better detect eye changes in WD - a project is planned which hopes to recruit nationally.

We rounded things off with a discussion on the potential for neonatal screening for WD. Tom Marjot presented the work of the Seattle group looking at ATP7B peptide levels as a diagnostic tool for WD. A debate followed on whether neonatal screening for WD should be implemented, presented by the two paediatric hepatology trainees. The 'nos' just won but this was a useful platform for future study.

Bill Griffiths, Consultant Hepatologist, Cambridge, and WDSIG lead—13.02.22

Drug Trial — UNITED Study—UNIVAR Solutions B.V.

Characterization of the Pharmacokinetics and Pharmacodynamics of Cufence® 200mg in Wilson's Disease Patients

Univar is conducting a post authorization efficacy study to characterize the pharmacokinetics and pharmacodynamics of Cufence (trientine dihydrochloride) and to investigate the efficacy and safety in Wilson's disease patients. This study is open for children, adolescents and adults, from the age of 5 years on. Patients included in the study should previously have been treated with D-penicillamine (≥18 years) or D-penicillamine or zinc (<18 years). After enrolment into the study, patients would start treatment with Cufence® in two different dosing schemes and would be closely observed during visits for up to 24 months. Study patients are being treated in 10 centres in Europe, including two study centres at King's College Hospital, London. The aim of the study is to generate new information on the posology (dosing) and corresponding effect of Cufence® (trientine dihydrochloride) in both adults as well as children.

If you are interested in taking part in this study, please contact WDSG-UK.

NHS Organ Utilisation Group (OUG) - Tx Patient Focus Group and Survey

The NHS OUG is looking for people who are either on the liver transplant waiting list or have had a liver transplant in the past to take part in patient focus groups sharing their experiences of the liver transplant services. The OUG is working on an improvement initiative. If anyone is interested and willing to take part, please email OrganUtilisationProgramme@nhsbt.nhs.uk direct for further details. All views will be anonymised.

Meanwhile, the NHS OUG is also running an anonymous online survey which covers similar questions to those being raised in the focus groups. If you are a liver transplant patient or are on the liver transplant list and would like to complete the survey, please follow the link https://bit.ly/liver-txp-survey22 by 17 April 2022.

Public Health England (PHE)—Wilson's Disease Pilot Study—Update

'NCARDRS (The National Congenital Anomaly and Rare Disease Register) is now part of NHS Digital (NHSD) as Public Health England ceased to exist in October. The WD Rare Disease Registry continues to expand as more cases are identified. The registry currently contains over 600 patients from England. These have been reported from clinicians at 28 hospitals or identified using data linkage techniques developed by Dr Osob Mohamed and the NCARDRS team. Now that NCARDRS is part of NHSD, we have access to new data sources and the registry will expand further. We continue to work to identify the incidence of liver cancer in patients with Wilson's disease, which is currently unknown. We are also working with NHS England to understand the distribution and use of trientine dihydrochloride throughout different regions in England.'

Osob—March 2022

WD Meds in the UK (April 2022) - Update

Supply of D-Penicillamine

There were issues during the Winter with the production of Viatris' (formerly Mylan) 250mg strength of penicillamine. We understand that this was resolved by the end of February and wholesalers including Alliance, AAH and Phoenix should now all have them back in stock. Please let us know if you have any problems obtaining it.

Information on new Storage Conditions of Cufence® previously known as Trientine Dihydrochloride

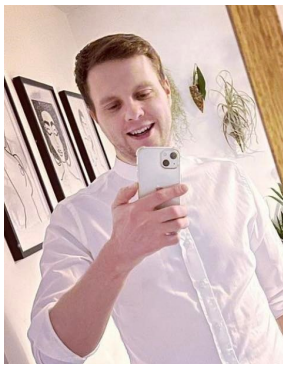
On 20 May 2021, the European Medicines Agency (EMA) approved Univar Solutions' request for the update of the storage conditions for Cufence® 200mg (trientine dihydrochloride 300mg). As a result of this approval the storage conditions changed from 'store in a refrigerator (2^{o}C - 8^{o}C), do not freeze' to 'no special storage conditions' - either before or after opening. The shelf life of the product remains 3 months and it is advised to keep the bottle tightly closed in order to protect the capsules from moisture. The update in storage conditions was made possible by additional stability testing undertaken by Univar Solutions in preparation for the EU Marketing Application for Cufence® 200mg and Univar would like to stress that the formulation of Cufence® 200mg has not been changed to enable this change in storage conditions. The new storage conditions should already start to be reflected on the label of the product. There are no production or distribution problems anticipated with this product.

Cuprior® (Trientine Tetrahydrochloride) made by Orphalan Ltd

For patients in the UK taking trientine, it may be of interest to you to learn that an alternative trientine preparation, trientine tetrahydrochloride (Cuprior®) has been licensed across the UK. It does not need to be refrigerated at all and is supplied as scored splitable tablets in blisters, each pack containing 72 tablets. There are no production or distribution problems with this product either.

Danny's Story

by Danny Hiles



Danny Hiles is familiar to many of us through Facebook, where he posts the links to the many (24 to date) fascinating YouTube *Danny Chat* interviews with different guests across the world. You might also know him from either of the two *Coffee Morning* Zooms we have held recently.

Danny was born in 1986 and was brought up in a small village outside Stevenage. He had no other full blood siblings but has five younger half-siblings. At school his dyslexia made life frustrating for him, but he excelled at sport playing football for the school and a Sunday league team and attending an Athletics Club where he represented the County in running. In 2018 he started a blog talking about his diagnosis of WD.

"It started in the summer holidays of 2000. I was feeling tired all the time, had recurrent nose bleeds and was sleeping a lot plus I was moody, which was slightly out of character for me. Mum didn't know if it was just me being a typical teenager as I was the first child she had had, so it was all new to her! She took me to the doctors and they ran blood tests which showed that I was slightly anaemic and my liver function tests were slightly abnormal, too. The doctors said it was nothing to be concerned about, so it was business as usual. On return to school I became more and more tired and I was finding playing sport difficult. I was also struggling to run. I remember falling asleep on the school bus going home and going to bed straight after dinner.

On Christmas Day that year, I was sick during lunch. A week later I went to a friend's house for a New Year's Eve party but because I wasn't feeling very well, I came home early. Shortly afterwards, I was rushed to my local hospital, The Lister, with severe stomach pains and a swollen stomach. At first they thought I had an appendicitis, but that was ruled out. As I was in agony and screaming in pain, my parents refused to take me home. After three weeks in hospital, they suspected I might have Wilson's disease and I was immediately transferred to King's College Hospital, London, where the diagnosis was confirmed.

During this time I had many more tests. I was found to have oesophageal varices, blood in my stools and an enlarged spleen. Kayser-Fleischer rings were noted in my eyes and doctors flocked to see me! I became somewhat of a celebrity. It wasn't uncommon for me to be examined by 10-15 student doctors every day. To me it filled the time—I just thought how else are these people going to learn if they don't get the chance to see things for themselves?

When it came to treatment, the doctors wanted me to have a liver transplant, although they said I could try taking meds for the time being. I asked if I was going to die and they said they would do their best not to let me! I remained positive throughout and decided to give the medication a go. I was given penicillamine (plus pyridoxine—Vit B_6) and zinc to take 3 times a day, omeprazole to stop the nausea and codeine to help with the pain. I was also told to follow a low copper diet especially for the first year or two.

> ***"My mother was a nervous wreck"***

I stayed in King's for 3 months before being allowed home. I was 15 by then and had already missed a lot of school. I was keen to return, but my recovery was far from over. Over the next six months I was in and out of hospital with an ambulance regularly being called to the house. My mother was a nervous wreck! I was fighting nausea every day and living off salt and vinegar crisps and ginger biscuits, as these were the two things that helped settle my stomach.

When I did return to school, I don't remember too much about being in lessons. I was just happy to be around my friends. It wasn't until parents' evening that the teachers told my mum I would often be sick in the bin during class or run out of the room to the toilets, returning as if nothing had happened!

All this time, my best friend Andy had stood by me. His mum, Tina, had brought him to visit me in hospital whenever she could. Before I had been ill and after I started to get better, Andy and I spent much of our time fishing together, quad biking and roaming around the huge country estate his parents managed nearby. It was a great place to live and I always felt at home there.

Not surprisingly, after so much absence at school, I didn't do as well as I had hoped in my GCSEs. It was also now apparent that I was unlikely to fulfil my childhood ambition of becoming a professional footballer! However, I was accepted on a course to do Land and Countryside Management at Sparsholt College in Hampshire, where I stayed for three years and which I really enjoyed.

Having finished college, it was time for me to leave home much to my parents' dissatisfaction. My health had stabilised and I was ready for an adventure. I decided to go backpacking on my own around Europe, which was at least on the doorstep if I got ill. I discussed it with my doctors first. Armed with 3-4 months' supply of meds and a special medical bracelet that my parents had bought me with *Wilson's disease* engraved on it, I set off on my travels. I didn't return for another year.

I had the best time of my life and travelling alone really boosted my confidence. A couple of incidents stand out in my mind. Whilst I was working on the Greek island of Ios, my mum sent me out a parcel of meds. When I turned up at the Post Office to collect it, I was redirected to the Police Station immediately. There I had to explain what the tablets were for before being allowed to take them away. Ios is known for being "a party island," and has a zero tolerance drug policy. On another occasion, while in Sicily, I came out in a rash from head to toe. The local pharmacist sent me straight to the hospital, where I was given a suppository which seemed to solve the problem. To this day, I have no idea why!

It was 2004 when I returned to England and the next 14 years of my life were relatively unremarkable. I did a degree in Renewable Energy Technology in Cornwall and ended up working in sustainable construction, particularly on farms. I visited King's annually and it was on one such occasion at the end of 2018 that they said now was the time to go ahead with the transplant. I had always known this was on the cards, but it still came as quite a shock. Because of my blood type, I was told I would probably have to wait six months. Meanwhile, I was advised to sign off work, keep myself fit and inject myself with blood thinners every day in readiness for the op.

> ***"And with sirens blaring and blue lights flashing, we..."***

The first few weeks were a bit crazy trying to process everything while also having to make all the necessary adjustments in my life. After the breakdown of a long-term relationship, I was now living back in my home village and was renting a room from the Piggotts (Andy's parents) whom I had now adopted as family. I was about to move into my own house, but that had to be put on hold. When Christmas came, everybody clubbed together to provide me with everything I would need to make my hospital stay easier and at that point I realised how special the people were around me.

The next step was going into King's for tests and evaluations. Not only did they have to make sure I was physically fit enough but also that I was strong enough mentally to go ahead with the transplant. The hospital staff were amazing. I signed all the paperwork and was sent home with instructions never to be more than a couple of hours away and to keep my phone on me at all times!

Two months later, I received a call. An ambulance was despatched and with sirens blaring and blue lights flashing we sped off through the night to hospital. It all happened so quickly. But no sooner had we arrived than we were told over the radio to go back home again. The liver wasn't suitable. This happened on two further occasions, although then I did at least get as far as pre-op assessments and signing consent forms before being told they couldn't go ahead! Things then went quiet. I was spending my days taking long walks, looking after my sister's animals and taking photographs on my new, half-decent camera, always remembering to stop at 3.00 in the afternoon for my daily injection!

Then one evening at 9.30 the pivotal phone call came. It was 30th June 2019. I was again taken off to King's, but this time the liver was deemed fit. I walked into the operating theatre at 8.15 the following morning and was wheeled out 8 hours later. Within 24 hours I was sitting up in ICU eating an ice lolly. The first thing my dad said after the op was that I was looking a far healthier colour! I had been telling myself over the past couple of years that I was as fit as a fiddle, but in truth I had been tired, nauseous, fuzzy-headed and lacking motivation.

After the Op

I remained in hospital for a further 10 days before returning to my adopted family's. I hadn't fully appreciated how much support I was going to need afterwards. Apart from recovering from the op itself, I had to get used to the immunosuppressants and steroids and also live with the constant fear that my body might reject my new liver at any time. Unable to do much other than recuperate, I spent time focussing on starting a blog about living with Wilson's disease and having a transplant, and I am still adding to it today (*https:organtransplant. home.blog*). I have also started to interview patients across the world discussing their experiences of living with WD and/or having a transplant and uploading it on to my YouTube channel: *YouTube-Danny Chat* videos.

Nearly two years later, I have survived the pandemic (despite having had Covid twice), have returned to work, live in my own flat and have a new girlfriend. I have also taken on a personal trainer with the intention of training for the Transplant Games. None of this would have been possible without the dedicated staff at King's, my little sister, Georgina, who was with me every step of the way, the Piggotts for giving me a home and looking after me so well, my parents, brothers and other sister, my best friend Andy and of course the family who so generously donated the liver to me. I thank them all from the bottom of my heart.

Members' News 2021-22

Valerie

Our President and world renowned Wilson's disease specialist, **Dr John Walshe**, will be celebrating his 102nd birthday in a couple of weeks' time. His daughter and son-in-law, Susan and Phil, continue to live with him and look after him well, but they now employ a full time carer to assist. The biggest event that has happened to them all over the past twelve months is the birth of Dr Walshe's first great grandchild, Annabelle.

Baby Annabelle

Living nearby and with a negative lateral flow test under my belt, I recently called in to see him. My visits have been few and far between over the past couple of years because of the pandemic. Dr Walshe is deaf and I find it difficult projecting my voice, so after exchanging a few pleasantries I sat down and encouraged him to do most of the talking. These are some of the highlights.

I started off by asking him how he was?

"Worn out!" he replied. "There are so many things I can't do any more. I'm dependent on help. I haven't been upstairs for over a year or so now."

I asked if he recommended living so long?

"It's better than being dead!" he abruptly replied.

I told him that the oldest person ever to have lived died at 122, having only given up smoking when she was 118. He asked where she lived and I told him, "France!"

"Oh my grandmother was French," he said. "She had 7 daughters and a son. The son died in a fire. They lived in the Caribbean, but all the daughters were sent to school over here. My mother trained as a nurse. She was sent to Alexandria, where she met father. He was working out there as a neurologist in the First World War. They met in the hospital."

He continued, "When I was a Medical Officer, my first posting was to Alexandria. I saw Tutankhamun and went up the Great Pyramid of Giza. It was like going up the escalator at Baker Street! The Great Pyramid is the only one of the 7 Ancient Wonders of the World to have survived."

"Really," I said, testing him, "so which were the other 6?"

"Well," he replied, I with pen to paper, "The Hanging Gardens of Babylon; the Colossus of Rhodes—that was a huge statue with its feet astride the harbour: people should see if they can find any bits in the water there; the statue of Zeus, the Tomb of Halicarnassus, the Temple of Diana at Ephesus...and...and...I can't think what the other one was?" With a sigh he added, "My memory's failing me nowadays!"

I hardly think so! I googled it in front of him and said, "It's the Lighthouse of Alexandria! Fancy forgetting that one!"

"Oh yes, the Pharos," he said, cross with himself!

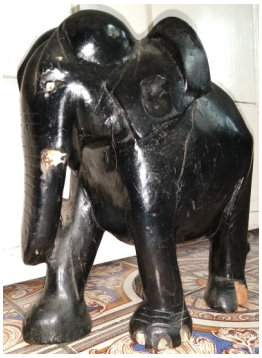
Thinking about his parents again, he reminisced,

"When father was demobbed, the family returned to London with their black and white dog called Bill. Bill had to go into quarantine for 6 months. After that they always had cats. We (our family) also had cats, two cats called Copper and Helix. They were brother and sister. When Helix died, Copper refused to eat. I didn't know that cats could be so close. In the end we called the vet! Then we had Labradors. The first was called Bushka and she had a litter. Planning on giving the puppies away, we were surprised to find she just had the one. So we kept her—Bush baby! Actually, we called them lapradors because as soon as you sat down they jumped on to your lap!"

Finally, leaving Alexandria and the animals behind, we turned for some reason to the subject of the Plantagenet Kings of England! He told me where they were buried, where they were purported to be buried and where they should have been buried, berating Reading Municipality in the process for not having dug around long enough to find the remains of Henry I.

We had been sitting talking for over an hour-and-a-half and I said it was time I must go! Whether or not he heard me, he turned my attention to the black wooden elephant perched on the bureau behind me.

"That's Ellie," he said. "I don't know what happened to her tusks! She was given to me by my mother's sister when I was 10. My mother's sister was married to the harbourmaster in Hong Kong and every time they came home, they would stop off at various ports along the way. One year they stopped off in Ceylon, Sri Lanka nowadays, and bought her. I'm rather fond of her. I used to make patients draw elephants, you know, as drawing spirals seemed somewhat uninspiring!"



Ellie

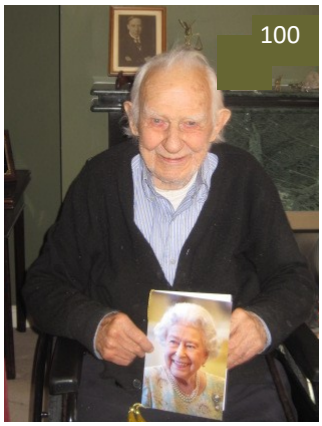
"Yes, I remember," I said, as somebody who could hardly draw my breath! "That's why we adopted an elephant as our original logo for the Group."

Seizing the opportunity, I added, "And talking of logos, it's time for me to go go!!"

"What time is it, he asked?"

"4.00 o'clock!" I replied.

"Oh good; time for tea!" he said eagerly.



James Kinnier Wilson was the guest of honour at the Wilson's Disease Symposium in London in October 2012, which celebrated his father's (Samuel Kinnier Wilson's) centennial anniversary of first describing *Progressive lenticular degeneration* (later to be named *Wilson's disease*) in the medical journal *Brain.* It was through this meeting that we established a firm connection with James. Living in Cambridge, he has been a member of the Group ever since, attending our AGMs and competing every year in the Dingbats competition!

Although he chose not to follow his father into medicine, James became a distinguished academic in his own right, studying Akkadian and Hebrew at Exeter College, Oxford, before co-establishing and developing the Assyriology course at Cambridge University. In November 2021, he celebrated a 100th anniversary of his own, being his birthday! The Group marked the occasion with a card, a balloon and a small floral tribute. We wish him many happy returns and a long continued association with WDSG-UK.

Katie Dale (nee Chiswell) wrote in the 2020 newsletter about how as the daughter of a Wilson's disease patient, she too had been diagnosed with Wilson's disease at the aged of 29. Having received a liver transplant within days of the diagnosis, she like other immuno-suppressed patients has had to be especially careful during the Pandemic.

Preparing for the High Life!

However, to mark her 34th birthday last month and also the 5 year anniversary of receiving her transplant, she and her husband Pete decided to take a holiday of a lifetime over to Las Vegas! The flight itself took ten-and-a-half hours but with the eight hour time difference, they apparently arrived only two-and-a-half hours after setting off!

They stayed in a luxury hotel and visited all the major casinos during their stay, having a couple of little wins along the way, but not enough to retire on!

The highlight of the trip was a helicopter ride at night over the Vegas Strip with all its neon lights. Magical!

Pete and Katie

Ashok Pandit, our 31 year old patient from Nepal, writes about how Covid has affected him, and Nepal in general:
"Like the rest of the world, Nepal also faced Covid. Everyone panicked. The first wave didn't affect us much, but the second one did. Since we share an open border with India, we share a similar trend in severity of cases with them.

I remember when Covid was new, we were all anxious about the future of mankind. Every time we heard the news of the number of infections, we used to be terrified. And then, finally, the government imposed a lockdown. We were asked to stay in our houses and only go out if it was urgent.

Grocery stores were allowed to open for some hours in the evening and mornings and medical stores were open for 24 hours. People used to call my mum and tell her not to allow me to go out because they think I am weak, sick and would be vulnerable to Covid. I never felt like that, nor did my mom. I used to go shopping, wearing a mask whenever I was out. Police would patrol the roads to check people's activities and people used to gather at the junctions and run away whenever the police appeared!

I stayed at home most of the time, but I worried I might run out of medicine or the supply of the medication might stop. I contacted different pharmaceutical companies in Nepal and got hold of medicine for a year. Still, I continued to worry. I then started meditation to calm myself, which has been a great success.

The fear of Covid had just decreased when a neighbour and his family caught it. Fortunately, they all had mild symptoms and were quarantined in hospital. Neither mom nor I have had it. We got vaccinated twice but haven't yet had a booster. But Covid has taught me to be patient and not to panic. The total number of Covid related deaths in Nepal, among a population of 30 million, currently stands at 11,950."

Staying Calm!

Linda Hart needs no introduction to most of us. As the co-founder of the Group and the driving force behind it, she shares with us the trials and tribulations of the past couple of years.

Last drumming day before all Hell broke loose!

Hello everyone, I haven't contributed anything to the newsletter for a long time, not since before I retired from the committee, which is more years ago than I care to remember! I haven't been completely lazy though. About eight years back I joined *The South Derbyshire Community Drummers* and took up African Djembe Drumming. I've always failed miserably at almost every instrument I've tried to learn, but drumming seemed to click. We've had tremendous fun, playing at local festivals, food markets and the like and I've made many new friends along the way.

I also share an allotment, go to Leicester Tigers' rugby matches, enjoy Yoga and Tai Chi classes, but of course the pandemic and ensuing health problems put an end, or what I'd rather call a long pause on all that. The last two years have been an awful strain on everyone.

Towards the end of 2020, I was diagnosed with CLL (chronic lymphocytic leukaemia). I was aware that I had lymphocytosis , which is a higher than normal number of a particular type of white blood cell. That was diagnosed back in 2015 after I'd been to my GP several times complaining of fatigue and various aches and pains. Turned out that after a special blood test (which I forget the name of) my lymphocytosis was the precursor of CLL. Off I was sent to the haematology department of our local hospital. Ultimately I was told I would be on what is called *Watch & Wait*, which involved a blood test every couple of months or so at my GP practice to check my lymphocyte count. If and when it reached a certain number I was told I'd most likely need treatment, which would be chemotherapy.

To be honest, at the time it went completely over my head and I didn't think for a moment I'd develop *the big C*. I felt fine(ish!) - this cant be right. Later, of course, I started to look it up on Dr Google— NOT to be recommended for absolutely any ailment unless you are sure it's a completely reputable website.

As the months passed it went to the back of my mind. My counts sometimes went up, sometimes went down, not anywhere near the treatment number...I relaxed. I did begin to tire more quickly but put that down to age. Just before the pandemic began, my numbers began to rise more rapidly and not come down, what timing! In a phone appointment with my haematologist in October 2019 he said "I'd better see you in clinic in January."

Unfortunately I didn't get that far but ended up in hospital in December 2020. CLL had come home to roost. I was in hospital about five days, being given blood and a battery of tests one of which was a bone marrow biopsy which showed 90% infiltration of CLL cells. To cut a long story shorter, my treatment started in the new year; it was to last a year. I had an intravenous immunotherapy drug every twenty eight days for six months and was started on four tablets daily from March until January of this year, plus various anti-virals, antibiotics and Lord knows what else. Now I've reached the end and I'm in what I hope will be, a long remission— recuperation I know will be a long road. I'm told, on a regular basis, to take things slowly, baby steps to recover my strength and energy.

It's been a very, VERY, testing couple of years, but I think I've learned a lot. A lot about myself, about patience, tolerance, empathy and people. I've often thought how fortunate I've been as far as Wilson's disease goes, maybe because I was diagnosed at such a young age (I was 10). Children, perhaps, are far more accepting of illness, or as in my case, do their best to ignore it...and no, it didn't go away! Though I was very sickly as a youngster I haven't suffered from it nearly as badly as many have. Whilst I'm on the subject I would like to point out that my CLL is not related to my Wilsons, just one of those unfortunate things and my own body chemistry.

Bell ringing day—half way there!

What really struck me on this 'CLL journey' from day one of my treatment was the cheerfulness, resilience and strength of mind of all the fellow patients I met whilst sitting in the big chairs hooked up to drips, and also the kindness and understanding of all the staff. I really think it is one of the cheeriest, most optimistic hospital departments I've been in.

I think one of the biggest things I'll take away from these last couple of years is, DO IT NOW! Don't wait until you've lost weight, have a partner, have more money, better job etc, etc. Life can change so quickly. Enjoy every day friends; be as healthy and happy as you can.

Anne-Marie is a retired Modern Languages lecturer and former WDSG-UK committee member, who has recently moved to a small village outside Carlisle. Diagnosed in 1977, her story can be found in the 2011 newsletter. She and her husband were finally able to take a long awaited boat trip to Bass Rock last summer and they have kindly shared their experience and stunning photographs with us here. She writes:

THE NORTHERN GANNETS OF BASS ROCK

Postponed from 2019 because of Covid, as lockdown was relaxed we were able to rebook a boat trip to view the wonderful spectacle of the northern gannets of Bass Rock one beautiful sunny day last August.

Bass Rock is a small island of steep-sided volcanic rock in the Firth of Forth in Eastern Scotland, five kilometres north-east of North Berwick. It is an exceptional place because it is home to more than 150,000 gannets. When viewed from the mainland large parts of the surface look white because of the sheer number of birds nestling on the cliff faces and, of course, their droppings. It certainly looked stunning, gleaming white in the morning sunlight, as we stood waiting for our boat to arrive with the six other passengers.

We had picked a specialist photography trip as my husband wanted the chance to capture the classic diving gannet shot as they enter the water. Gannets are famed for their super-fast fishing dives into the sea. We set off towards the wondrous rock in eager anticipation. To encourage the birds to dive near the boat the boatman used a method called *chumming*. For this he had a supply of mackerel which he kept throwing into the water. The result was amazing. The boat was soon surrounded by gannets and it was exciting and really quite emotional seeing so many of these beautiful birds close up, whizzing by and diving to catch the fish. We were able to observe their superb diving technique, the way they fold their wings right back just as they enter the water head first.

Northern gannets are large white birds with ink-black wingtips, a wingspan of two metres and beautiful yellow saturation on their head. They have a long neck and long, pointed black beak. Incredibly, they can dive from a height of 100 feet and achieve speeds of 60 miles an hour.

After the *chumming* we went closer to Bass Rock itself and sailed round it to view the magnificent display of the densely crowded, noisy, bustling colony perched on the cliffs. As we approached the noise of the birds seemed to grow to a crescendo and the smell was overwhelmingly pungent. We were able to observe the birds quite clearly, nestling on the rocks and ledges, often with their young. A truly unforgettable experience.

Bass Rock

Diving for the Mackerel

A white cliff face covered in gannets and their droppings!

What a great day!

Abby's Puzzle Page

Wordsearch - Easy

A	A	G	L	W	A	F	P	C	Q	F	I	W	V	P
I	P	R	Q	T	A	Y	H	S	I	T	I	R	B	C
M	L	L	K	K	P	P	F	Q	J	Y	C	O	F	T
R	A	H	T	L	A	E	W	N	O	M	M	O	C	B
I	T	I	A	R	I	C	B	Z	K	R	O	Y	A	L
N	I	O	G	Z	F	M	O	A	T	Y	E	T	U	D
Z	N	N	K	I	W	L	P	R	S	Y	E	N	W	X
M	U	L	U	J	S	J	Q	A	G	L	L	E	O	M
S	M	L	Z	Q	W	H	G	Z	Z	I	I	V	E	P
J	B	T	U	J	T	E	L	I	Z	A	B	E	T	H
R	C	E	S	W	R	M	H	V	F	O	U	S	P	B
S	E	R	T	E	S	Z	O	L	T	V	J	P	M	Y
N	T	C	E	L	E	B	R	A	T	I	O	N	G	O
Y	A	D	I	L	O	H	K	N	A	B	X	B	P	O
Y	N	R	O	S	D	N	I	W	K	M	X	Z	W	W

BANKHOLIDAY
BRITISH
CELEBRATION
COMMONWEALTH
CORGI
ELIZABETH
JUBILEE
PLATINUM
QUEEN
ROYAL
SEVENTY
WINDSOR

Anagrams - Hard

Solve these anagrams of food and drinks you might find at a street party celebrating the Queen's Platinum Jubilee!

1. filter
2. mange chap
3. wise ranch succumbed *(2 words)*
4. airman bartered screws *(3 words)*
5. Togo rash *(2 words)*
6. pivoting a score *(2 words)*
7. deadpan mom simnel *(3 words)*
8. on thin oceanic rock *(2 words)*

Sudoku - Medium

9	7			2				
6			8	9		1		
8			4					7
	9	8	7					1
		4	6		3	2		
2					9	8	3	
1					4			5
		9		1	5			8
				7			1	2

Sudoku puzzle provided by www.sudokuof theday.com

Dingbats - Castles in the UK - Tricky

1 dswor	2 C Y	3 Clear
4 Slob	5 Lud	6 a L
7 C	8 S to RM T	9 £

Infuriating

Answers can be found at the bottom of page 7.

Where's the Queen?

The 13 cards of a suit are shuffled and dealt out in a row and it is found that none is in its correct numerical position (Ace left and King right). The court cards (King, Queen & Jack) don't appear at either end or adjacent to each other. The Ace is directly between 9 (left) and 8, the 4 is directly between Queen (left) and Jack, the 2 is two places to the left of 10, the 7 is two places to the left of 3, the King is somewhere to the left of the Queen and the far left-hand card is one higher than the far right-hand card. The 9th and 10th cards from the left total 9, the 9th being of lower value. Can you locate each card and say where the Queen is?

1	2	3	4	5	6	7	8	9	10	11	12	13

WILSON'S DISEASE MULTI-DISCIPLINARY CLINICS

Adult Clinics

The Birmingham WD Clinic

Dr Andrew Holt (Consultant Hepatologist) and **Dr David Nicholl** (Consultant Neurologist) hold a one-stop Wilson's disease clinic at **University Hospital Birmingham** on a Friday morning six times a year. This clinic offers patients the opportunity to have their management reviewed by a hepatologist and a neurologist at the same time and is intended to supplement otherwise established care. Since the pandemic, we are doing more remote (video/phone) consults as face to face consults are still limited. Referrals should be addressed to **Cheryl.Scandrett@uhb.nhs.uk** and must come from the clinician looking after the patient**.** Cheryl is the WD Clinic Coordinator at Queen Elizabeth Hospital, Mindelsohn Way, Edgbaston, Birmingham, B15 2WB.

The Cambridge WD Clinic

Dr Bill Griffiths (Consultant Hepatologist) and **Dr Paul Worth** (Consultant Neurologist) run a joint clinic every 6 months for patients with neurological manifestations of Wilson's disease at Addenbrooke's Hospital in Cambridge. Dr Griffiths sees a number of patients with hepatic-only disease separately in his adult genetic liver clinic. Both clinics run on Wednesday mornings. Referrals from clinicians can be made to Dr Griffiths at the Liver Unit, Box 210, Cambridge University Hospitals, Hills Road, Cambridge CB2 0QQ.

The Leeds WD Clinic

Dr Joanna Moore (Consultant Hepatologist) and **Dr Jeremy Cosgrove** (Consultant Neurologist) run a joint clinic for patients with Wilson's Disease at St James's University Hospital, Leeds. This currently runs on a Thursday morning. Referrals from clinicians can be made to Dr Moore at the Leeds Liver Unit, Merville Building, Beckett Street, Leeds LS9 7TF or Dr Cosgrove at Department of Neurology, F Floor, Martin Wing, Leeds General Infirmary, LS1 3EX.

The National Hospital Wilson's Clinic, Queen Square, London

A dedicated Wilson's disease clinic has been held at University College Hospitals for the past 30 years. It was set up in 1987 by Dr John Walshe after his retirement from Addenbrooke's Hospital, Cambridge and at the invitation of Dr Gerald Stern. **Dr Godfrey Gillett** started to attend in the mid-1990s and has continued the clinic under the kind auspices of Dr Robin Lachmann at the National Hospital for Neurology and Neurosurgery, Queen Square. The clinic has 60 patients and is held on the third Friday of the month with the close involvement of **Professor Tom Warner, Dr Sam Shribman** and **Mrs Maggie Burrows**. Referrals from across the UK are welcome, addressed to Dr GT Gillett, Laboratory Medicine, Northern General Hospital, Sheffield Teaching Hospitals NHSFT, Herries Rd, Sheffield, S5 7AU or to Dr GT Gillett, Charles Dent Metabolic Unit, Internal Mailbox 92, NHNN, Queen Square, London WC1N 3BG.

The Salford WD Clinic

The longstanding neurology consultant led WD Clinic at Salford Royal Hospital has recently been progressed into a joint neurology-hepatology consultant led clinic (**Dr Georgeta Taylor**, Consultant Neurologist and **Dr Jumi Isibor**, Consultant Hepatologist). **Dr Karolina M Stepien**, Consultant in Adult Inherited Metabolic Disorders and her MDT (a dietician and a physiotherapist) review WD patients with hepatic-only manifestations, with the long-term plan to provide a comprehensive support for our patients in a joint neurology/hepatic/metabolic clinic. Referrals from clinicians can be addressed to any of the above mentioned consultants at Salford Royal NHS Foundation Trust, Stott Lane, Salford, M6 8HD.

The Sheffield WD Clinic

The Sheffield clinic is jointly run by **Prof Oliver Bandmann** (Consultant Neurologist), **Dr Barbara Hoeroldt** (Consultant Hepatologist) and **Dr Godfrey Gillett** (Consultant in Clinical Biochemistry and Inherited Metabolic Disease). Clinics take place every six months at the **Royal Hallamshire Hospital, Sheffield** on a Tuesday afternoon. Patients may be seen in interim clinics by arrangement. Referrals should be addressed to Prof Oliver Bandmann, Department of Neurology, Royal Hallamshire Hospital, Glossop Road, Sheffield, S10 2JF. Either GPs or hospital specialists may refer to this clinic.

Children's Clinic

Clinic for Children and Young Adults at King's College Hospital, London

There is a multidisciplinary team Wilson's disease clinic for children and young people at King's College Hospital which is run by **Prof Anil Dhawan** (Paediatric Liver), **Dr Tammy Hedderly** (Paediatric Neurologist), **Dr Jemma Day** (clinical psychologist) and **Ms Bethany Tucker** advanced nurse practitioner (ANP) in children's liver disease. The clinic is directed at patients who are complex with both liver and neurological involvement. Referrals should be made via the Paediatric Liver Centre at King's or enquiries sent to **Lucia Debiase (lucia.debiase@nhs.net)**, PA to Prof Dhawan.

A Date for your Diary 2022-23

Date	Time	Event
Sunday, 3 July 2022	**1100**	**WDSG-UK 12th AGM – Zoom Virtual Meeting — Details to follow**

Wilson's Disease Support Group – UK

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Rupert Purchase, DPhil Group Adviser on trientine
Webmaster: **Blackcat Websites**

Tell others about WDSG-UK

Please encourage anybody else that you know with Wilson's disease to join **WDSG-UK**

Inform your family, friends, consultant physicians, general practitioners and local MPs about the work of **WDSG-UK.**

The more people who know about **WDSG-UK**, the more we can promote a better awareness of Wilson's disease within the community and the better the chance of an early diagnosis.

If more copies of this newsletter are required, please contact Valerie.

We're on the web
www.wilsonsdisease.org.uk