

2017 Wilson's Disease Support Group – UK Meeting & 7th AGM

Cambridge Rugby Union Football Club, Sunday, 23 July 2017

The largest ever gathering of WDSG-UK members had assembled promptly for the start of our 2017 meeting, thereby avoiding a downpour of rain, which arrived shortly after midday.

Many new members were able to attend this year, both from the UK and overseas. We were particularly delighted that Dr Walshe's first UK Wilson's disease patient, **Shirley Wylie**, was able to join us for the first time. Dr Walshe first treated Shirley in 1955, and her remarkable story is recounted in this year's WDSG-UK Newsletter. From overseas, we were pleased to welcome **Priya Joshi** and family from Zurich, **Regine Bielecki** from Dusseldorf, and **Kumari Bahall** from Trinidad and Tobago. From the UK, it was lovely to meet youngsters **Oliver, Isabella, Sam F** and **Sam J**, together with a slightly older **Andy Forster** and **David Pereira!** Many regular members of the Group who have supported us over the last decade had also made their way to Cambridge, and the audience was completed by the presence of **Drs John Walshe, Godfrey Gillett** and **James Dooley**.



Patients Attending the Meeting, July 2017

Rupert Purchase opened the meeting by thanking Valerie for organising this event, and then introduced **Jerry Tucker**, Vice-Chair (WDSG-UK), who summarised some of the committee's work over the past year on behalf of members. The committee regularly attend meetings organised by Rare Disease UK, who are active in promoting the UK Rare Disease Strategy and **Caroline** has recently attended an NHSBT meeting reviewing liver transplantation in the UK. Jerry and Valerie are members of a NHS England Working Group who are examining issues regarding trientine, and we continue to develop the WDSG-UK Patient Register. Jerry also spoke about plans to improve and expand the WDSG-UK website.

Disease models, whether *in vivo*, *in vitro*, or *in silico*, are a means of understanding the causes of diseases and finding treatments and cures. **Dr Emily Reed** from the Sheffield Institute of Translational Neuroscience, University of Sheffield, has summarised the available animal models (*in vivo* methods) for Wilson's disease in this year's WDSG-UK Newsletter. Current rodent models are inadequate for understanding neurological Wilson's disease. In a presentation before lunch, Emily described some of the exciting research she has embarked on to find better ways of modelling neurological Wilson's disease. Zebrafish have been successfully used to study other neurological disorders, and Emily has begun to use zebrafish who carry the ATP7B mutant (Wilson's disease) gene as an *in vivo* model of Wilson's disease. Emily is also developing an *in vitro* technique of transforming skin cells into neurons, and hopes that skin cells taken from Wilson's disease patients and transformed in this way will offer an insight into copper transport in the brain.

Lunch was followed by the **7th WDSG-UK AGM** and the election of the WDSG-UK management committee for 2017-2018. Jerry Tucker replaced Rupert Purchase as Chairman, with Rupert leaving the committee after seven years. Jerry presented Rupert with a gift in recognition of his work for WDSG-UK. The other committee members, Valerie Wheeler, Caroline Simms and **Mary Fortune** were re-elected for a further year. In his Chairman's report, Rupert thanked the committee for their support over the past year, and highlighted Valerie's commitment to helping Wilson's disease patients and their families through her work on the Group's Newsletter and Facebook page and by meeting up informally with WDSG-UK members during the year.

After the AGM, and preceding the raffle, **Anusha Joseph** read a poem in memory of Charlie Watsham, who died in 2015. This year's raffle was run by new member **Ruth Morgan** and raised **£104** to support WDSG-UK. It was supplemented by the sale of *Wilson's disease* wristbands, produced and marketed for us by another new member, **Samantha Ryan Panchal**. Details of how to purchase these elegant blue wristbands may be found on the Group's Facebook page.

Following the discussion at the 2016 WDSG-UK annual meeting on the repercussions of the recent price increases for trientine dihydrochloride, Rupert gave an update of some recent developments. *The Times* newspaper has been following the steep increase in price of so-called unbranded generic medicines, and these reports show that the price of trientine is part of a general problem of the sale of out-of-patent drugs needed for relatively small groups of patients. To counteract unscrupulous price increases for these drugs, the UK government has enacted new legislation – the *Health Service Medical Supplies (Costs) Act 2017*. To quote from the government's guidance note on this Act, "the government's intention is to use these new powers where, due to a lack of competition in the market, companies charge unreasonably high prices for unbranded generic medicines."

The meeting concluded with a question and answer session between the audience and the three consultant physicians present. Questions were raised on whether trientine capsules could be emptied into water, fruit juice, or yogurt if patients have difficulty in swallowing capsules (or whether smaller capsules could be produced by the manufacturer); genetic testing to check that a partner is not a carrier of Wilson's disease; and the advisability of co-administering D-penicillamine and a zinc salt to treat Wilson's disease.

Following the close of the meeting, Anusha took the Group photographs before we dispersed into the warmth of a Cambridge summer afternoon. We hope that members feel that these annual meetings are helpful and informative, and look forward to an even greater attendance next year!