THE 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 **WDSIG** was one of the first to get going and 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 Public Health England (**PHE**). Over 100 individuals from across the UK are registered on the WDSIG database.

4th WDSIG meeting—29 November 2019—National Hospital for Neurology and Neurosurgery, Queen Square,

The SIG approved 3 new specialist WD Centres (Nottingham, Oxford and Bristol) bringing the total number of adult centres across England to 15. NHS England has been updated with these changes. A standards document to which Centres of Excellence should adhere was agreed and is now up on the BASL website under the relevant section www.basl.org.uk. All patients on trientine in England should have an annual review via one of these Centres, if they are otherwise being looked after elsewhere. In terms of specialised commissioning of WD services, at present NHS England (via the relevant clinical reference group) is not planning to take this on though this may change in the future.

PHE (specifically NCARDRS which is the national congenital anomaly and rare disease registration service) is gathering pace with its work on Wilson's disease. Dr Osob Mohamed, Hepatology clinical fellow based at the Royal Surrey Hospital, has been recruited to take on the work started by Michelle Camarata before she went on maternity leave and back into clinical Hepatology training. There are a number of strands of data collection including clinical information from Centres, hospital episode statistics, prescription data, mortality data, genetic and copper results. By cross-referencing these data sources the aim is to build up a comprehensive database and answer certain questions about prevalence, long term outcome and variation in WD treatment. Preliminary results have been submitted in abstract form to the European liver congress. By linking in with the CROWD study the hope is to better understand why patients with certain gene mutations present differently. It is also hoped that NHS Wales and NHS Scotland will be involved in due course.

The clinical session covered primary care awareness, screening for liver cancer (thought to be very rare) and a very interesting session on Deep Brain Stimulation (DBS) where Mr Ian Low a Neurosurgeon based at the Queen's hospital in Romford spoke of his experience. It seems that very selected patients with neurological WD might benefit from DBS and this is going to be taken forward by the movement disorder group of the Association of British Neurologists. It is recognised that more could be done to educate GPs in the early signs of WD and the group is hoping to publish a review aimed at primary care.

Research progress is a key remit of the SIG. The CROWD study has successfully recruited and is now generating data. Nottingham reported on a study which identified a higher prevalence of WD in their surrounding area than previously recognised, potentially diagnosing new cases and bringing existing patients under Specialist review. Some interesting work presented at the American liver congress was presented and an update on Pharma trials given. Many other research projects are ongoing and in development.

The meeting concluded with a discussion about the different formulations of trientine and about laboratory measurement of copper. There are some differences between the dihydro- and tetrahydro- forms of trientine which mean dose comparisons are not straightforward. In terms of copper measurement, this remains a fine art but progress is being made towards a more accurate way of calculating 'free' copper. There remains quite wide variation in how patients with WD are monitored and the SIG intends to audit this.

Full minutes of the meeting are available via the BASL website (www.basl.org.uk). The next meeting of the SIG is scheduled for 20th November 2020. More detailed information about the remit of WDSIG can be found on *p16* of the 2019 WDSG-UK newsletter (www.wilsonsdisease.org.uk)

Finally, BASL recently paid tribute on its website to Dr John Walshe on his 100th birthday with a photo and link to a specially written biography.

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