

THE WILSON'S DISEASE SPECIAL INTEREST GROUP (WDSIG)

What is it?

Through the British Association of 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. WD lends itself well to this new initiative, being a rare disease that has several types of clinician involved in its care.

Who is in it?

The WDSIG comprises clinicians in hepatology, neurology, clinical chemistry, paediatrics and psychiatry as well as research scientists and representatives from **WDSG-UK** and Public Health England (PHE). At this time there are nearly 100 individuals from across the UK included in the WDSIG member database.

What is the remit of the WDSIG?

The WDSIG functions primarily to improve care for patients with WD. The group recognises that care and access to expertise in WD is variable across the UK. WD suffers at times as with any rare disease in delay in diagnosis, lack of multidisciplinary input and inconsistent monitoring, which can result in unnecessary harm. Research is difficult to coordinate without collaboration and new therapies are coming online for WD. The WDSIG hopes to improve research into WD as well as work towards a better model of healthcare for WD which can optimise outcome. Through PHE the WDSIG hopes to answer questions about how WD is spread across the country and how patients are being treated.

What has happened so far?

Three meetings of the WDSIG have been held (all in London) - 14 December 2017, 14 June 2018 and 14 February 2019. Between 24 and 34 participants have attended these meetings. Minutes from each meeting have been uploaded to the BASL website along with some of the slide presentations.

The WDSIG has developed four core themes:

Service delivery – in accordance with the UK Strategy for Rare Disease a preliminary proposal to NHS England for a 'hub and spoke' model for WD has been accepted for further evaluation. This document outlines how a number of Centres of Excellence would function to deliver care for WD via multidisciplinary clinics, networking with neighbouring hospitals. As a consequence of trientine dihydrochloride being now commissioned by NHS England due to its price, WD centres in England have needed to be defined so there is a precedent there now in place.

National Registry – with the support of the WDSIG, Public Health England (PHE), specifically NCARDRS (National Congenital Anomaly and Rare Disease Registration Service), is establishing a Rare Disease registry which aims to capture relevant clinical data on all WD patients in England. NCARDRS has a special privilege to collect hospital episode data from any Trust, although patients can opt out if they wish. PHE is also collecting prescribing data, mortality information, data from trace element and genetics laboratories, and data from the national Transplant database. From these sources PHE hopes to build up an accurate picture of WD across the country.

Clinical care – in bringing together the clinical and laboratory expertise the WDSIG aims to specify standards by which patients should be diagnosed and monitored. The hope is that no patient with WD is left without proper monitoring of the various strands of their condition. Dissemination of 'best practice' by Centres of Excellence will be key here.

Research – the WDSIG will play an important role in WD research. The group recognises the scientific and clinical research on WD occurring in the UK, and provides a forum to discuss novel research ideas and foster collaboration. The WDSIG acts as an interface for Pharma where there are several new drugs for WD currently being trialled or in development. The WDSIG has worked to include the UK along with our European neighbours in these studies which hope to address some of the unmet needs in WD.

The group is planning a 4th meeting in the Autumn and has plenty of work to do in the meantime! Further information is available at: <https://www.basl.org.uk/index.cfm/content/page/cid/31>

Bill Griffiths,

Consultant Hepatologist and WDSIG lead.