Wilson’s disease: a centennial symposium

The Wilson’s Disease Centennial Symposium held in London in October 2012 commemorated Dr Samuel Alexander Kinnier Wilson’s famous paper entitled “Progressive lenticular degeneration: a familial nervous disease associated with cirrhosis of the liver” which was published in March 1912 in the neurological medical journal *Brain*. His work had been originally submitted as a 211 page thesis, for which he won a gold medal from the University of Edinburgh, where he had graduated ten years earlier. In this paper Dr Wilson, who at that time was a registrar and pathologist at the National Hospital for Nervous Diseases, Queen’s Square, London, described the progressive hepatic and neurological changes he had studied in four affected patients. His discovery led to other appointments, including Professor of Neurology at King’s College Hospital, London, and the disorder became known as Wilson’s disease.

At the symposium, historical aspects of Wilson’s disease were memorably recalled in talks by two nonagenarians – Dr James Kinnier Wilson (son of the eponymous Samuel Alexander Kinnier Wilson) and Dr John Walshe. The Wilson’s disease community owes an enormous debt to Dr Walshe for three of the drugs used to treat Wilson’s disease: D-penicillamine; trientine dihydrochloride; and ammonium tetrathiomolybdate. In a poster presentation, Rupert Purchase summarised Dr Walshe’s role in the discovery of these drugs, and he has recently expanded these notes into a short, and mainly historical, review *The Treatment of Wilson’s Disease*, which has been published in the journal *Science Progress*.

A rationale for treating Wilson’s disease only emerged in 1948 after J. N. Cumings’ finding of excessive accumulations of copper in the liver, brain and other organs of patients with Wilson’s disease. Glazebrook had made similar observations three years earlier, and a literature search revealed previous copper measurements in Wilson’s disease patients. However, a link between the symptoms of Wilson’s disease and an aetiological role for copper is not intuitive, and the few copper measurements made before 1948 had not raised any questions about the relationship of copper to Wilson’s disease. So how and when was this link made? In a second article – *The link between copper and Wilson’s disease* – which will also be published this year in *Science Progress*, Rupert attempts to identify some of the key observations which led to the suspicion of copper’s involvement in Wilson’s disease and to the introduction by Cumings of British Anti-Lewisite (BAL) as a copper chelating agent for the treatment of the disease.

Reprints of Rupert’s two articles will be available later this year. If you would like copies, please e-mail him at rupertpurchase@wilsonsdisease.org.uk.

References