Health and happiness: Measuring wellbeing in Huntington's disease

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(Medical Xpress) -- Scientists at the University of Reading have designed a new tool which could significantly aid research and management of an incurable brain disease affecting thousands of people.

The ground-breaking project, led by Dr Aileen Ho, from the School of Psychology and Clinical Language Sciences at the University of Reading, has created a new system to measure the relative wellbeing of people who have Huntington's disease, a genetic brain disease for which there is currently no known cure.

The new tool, the first Huntington's disease-specific quality of life instrument of its kind, aims to fully capture the impact of this complex disease on the everyday life of sufferers. By using this new tool to more accurately measure the impact of Huntington's and also any effect of therapeutic interventions, scientists will be in a better position to evaluate the usefulness of interventions on patients' everyday quality of life to see if there is actually a practical benefit. It is hoped that in this way, this tool will lead to more effective treatments.

Huntington's disease is a progressive neurodegenerative condition that gradually affects the ability to move, think and reason, and has a devastating impact on a person's wellbeing, and that of his or her family. It affects more than 6,000 people in the UK and around 30,000 people in North America, with many more people - usually the children of those with the disease - considered 'at risk' of developing symptoms later in life.

While there is a genetic test that can tell people whether or not they carry the gene for Huntington's, there is no way of knowing when the first symptoms of disease - often clumsiness accompanied by unusual dance-like extraneous movements called 'chorea' - will begin to take hold. Previously, a patient's quality of life could only be assessed in a general way by asking them to complete questionnaires designed to assess wellbeing in a more general population, meaning the more specific and unique consequences of Huntington's disease were missed.

The new tool is called the HDQoL (Huntington's Disease health-related Quality of Life questionnaire), and is the first disease-specific quality of life instrument for people living with Huntington's. It can more accurately capture and measure wellbeing in this particular patient group, with their unique array of disease symptoms.

The research team, funded by a research grant from European Huntington's Disease Network, used a bespoke set of questions to understand issues that really affect patients drawn from hours of interviews with patients about their concerns and issues in daily life as a result of having Huntington's. From this, they were able to develop a measure based on what patients said matter most.

The new HDQoL method will now become the ‘gold standard' quality of life outcome measure for all clinical and research work into Huntington's disease, following a decision by leading experts from the US-based National Institute of Neurological Disorders and Stroke (NINDS) Common Data Elements Project.

"It is vitally important to be able to accurately measure and monitor patient's wellbeing, and to understand the true impact of Huntington's as they go about their daily lives," said Dr Ho.

"This information will be useful in the long term care and management

of patients over the average 20-year course of Huntington's disease."

The new HDQoL tool has been welcomed by patients and patient support organisations, including the Huntington's Disease Association (HDA) in the UK.

Cath Stanley, HDA chief executive, said: "I think the questionnaire will be an invaluable resource as it will offer a comprehensive view of how the illness affects individuals using a holistic approach."

Work from this project, comparing quality of life ratings made by family members of patients with that of patients' own self-ratings, is published today in the *Journal of Neurology*. Other work has also recently been published in the scientific journal *Clinical Genetics.*

More information: The paper, Mevhibe B. Hocaoglu, E. A. Gaffan and Aileen K. Ho, Health-related quality of life in Huntington's disease patients: a comparison of proxy assessment and patient self-rating using the disease-specific Huntington's disease health-related quality of life questionnaire (HDQoL), is published online by the Journal of Neurology .

Provided by University of Reading