

**Measuring quality of life in Friedreich's ataxia: a new approach**  
**Researchers: Dr Stefan Cano, Dr Jeremy Hobart, Dr Afsane Riazi & Dr Mark Cooper**

Researchers in London and Plymouth have developed a system to assess how quality of life is affected in people with Friedreich's ataxia (FA). This has involved the design and validation of a new questionnaire that will improve our knowledge of the impact of this debilitating condition and help assess the effectiveness of new therapies.

Friedreich's ataxia (FA) is a slow progressive disease associated with a broad range of physical symptoms. It is apparent that the incidence of different symptoms and the rate at which they progress varies quite markedly between people with FA. Up to now, little is known about the full impact of FA. Traditionally, doctors have used clinical tests to assess the degree of disability caused by the disease and the effectiveness of new treatment. However, these tests only provide half of the story - importantly, they do not include information about quality of life.

It is increasingly recognised that the perspectives of people with medical conditions should be central to the evaluation of new therapies. This requires accurate ways of measuring quality of life, which can be achieved by developing questionnaires using scientific techniques called 'psychometrics'. Over the last two years, a team of researchers at the Peninsula Medical School, Plymouth and Royal Free & University College Medical School, London has been working with Ataxia UK to develop a new FA quality of life questionnaire. This work has been funded by a grant from Ataxia UK.

The new questionnaire has been developed in two stages. First, interviews with people with FA were conducted and analysed to generate a list of questions that best represent the important areas surrounding FA and quality of life. This resulted in a 160-item questionnaire. Second, the questionnaire was mailed out to a group of people with FA (including over 500 participants) and scientific (psychometric) methods were used to further improve the questions. This resulted in a questionnaire that spans eight quality of life areas (see Figure 1). We have called the new scale the Friedreich's Ataxia Impact Scale (or FAIS for short).

Our study has been the first to develop a new FA quality of life questionnaire. In doing so, the first steps have been taken to improve research into FA and the effectiveness of a variety of therapies. We anticipate information provided by the new measure will benefit individuals with FA and their families, clinicians and Ataxia UK. In addition, this research provides a useful framework for developing quality of life measures for other ataxia groups. Future work will involve further evaluations of the FAIS, in particular to evaluate the long-term progression of FA and long-term outcomes induced by new treatment. This research was presented at the Association of British Neurologists Meeting (Sep 05).

We would like to thank the members of Ataxia UK for helping carry out this important study. We hope that the new questionnaire will improve knowledge of the impact of FRDA on people's lives. If you would be willing to participate in further evaluations of the FAIS please email us at: [scano@ion.ucl.ac.uk](mailto:scano@ion.ucl.ac.uk).

<u>Symptoms</u>	<u>Activities</u>	<u>Social Functioning</u>	<u>Psychological Functioning</u>
<ul style="list-style-type: none"> <li>• BODY MOVEMENT</li> <li>• SPEECH AND SWALLOWING</li> </ul>	<ul style="list-style-type: none"> <li>• UPPER LIMB FUNCTIONING</li> <li>• ACTIVITIES OF DAILY LIVING</li> <li>• LOWER LIMB FUNCTIONING</li> </ul>	<ul style="list-style-type: none"> <li>• ISOLATION</li> </ul>	<ul style="list-style-type: none"> <li>• MOOD</li> <li>• SELF-PERCEPTIONS</li> </ul>

**Figure 1: The Quality of Life Areas Covered by the Friedreich's Ataxia Impact Scale (FAIS)**

**For more support or information please contact:**  
**Ataxia UK, Winchester House, Kennington Park, Cranmer Road. London SW9 6EJ**

**Website:** [www.ataxia.org.uk](http://www.ataxia.org.uk)  
**Helpline:** 0845 644 0606 **Tel:** +44 (0)20 7582 1444 **Fax:** +44 (0)20 7582 9444  
**Email:** [helpline@ataxia.org.uk](mailto:helpline@ataxia.org.uk)