Harmonisation of cognitive screening for ALS in Europe

Sharon Abrahams, Thomas Bak, E Niven, J. Newton, Leonard Van Den Bergh, Vincenzo Silani, Albert Ludolph, Orla Hardiman, Dorotheé Lule, Markus Weber & Christian Burkhardt

by Abrahams, S. Niven, E.H., Newton, J., Bak T.H. University of Edinburgh, Scotland Hardiman, O., Pender, N. Trinity College, Dublin, Ireland, Lule, D., Ludolph, A. University of Ulm, Germany Van Den Bergh, L, Lenkens, M., University Medical Centre Utrecht, Holland Poletti, B. Silani, University of Milan Medical School, Italy Weber, M., Burkhardt, C. Kantonsspital, St Gallen, Switzerland

The aim of this study was to develop and apply a single method for measuring cognitive change across European centres in an attempt to harmonise cognitive screening. The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) has been previously validated in a UK sample and found to show high rates of sensitivity and specificity against full neuropsychological evaluation (see Niven et al.). Here the ECAS was translated and applied to a total of 358 ALS patients (Scotland, n=78, Ireland, n=73, Italy n=59, Holland, n=40 and Germany, n=108). Furthermore local normative data was collected within each country and cut-offs for abnormality determined. The ECAS was found to be sensitive to the types of impairment typically present in ALS patients across countries. The frequency of impairment within each country significantly differed with a higher proportion of impaired patients from the samples in Ireland and Scotland and lower rates in Germany and Holland. Furthermore the profile of impairment across cognitive domains also differed between countries with a greater proportion of patients with non-specific cognitive dysfunction (Memory, Visuospatial) in Ireland, while those in Scotland and Germany showed a more typical disproportionate impairment in ALS Specific functions (Executive, Language and Fluency). There was a significant difference between samples in diagnostic delay, indicating a more aggressive disease sample in Ireland in comparison to Scotland and Germany. Furthermore duration of illness significantly correlated with ECAS total scores with those patients with a longer duration of illness less likely to experience cognitive change. Frequency of impairment did not significantly differ according to symptom site of onset, which contradicts previously described associations with bulbar dysfunction, once tests which accommodate for speech and limb disability as the ECAS are used. The findings are discussed in relation to clinical profiles including severity of disease and population differences.
Switzerland

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Validierung des Edinburgh Cognitive ALS -Screen (ECAS) zur Aufdeckung von kognitiven Beeinträchtigungen oder Verhaltensänderungen bei der Amyotrophen Lateralsklerose (Screening for Cognitive and Behavioural Change in ALS)

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