DEMOGRAPHIC AND PHENOTYPIC CHARACTERISATION OF OUR ALS PATIENTS

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Knowledge of clinical and epidemiological data on ALS patients assist physicians to identify prognostic factors of the disease what helps patients and their relatives to schedule their activities during disease course.

Our aim was to analyse the available demographic and phenotypic disease characteristics from our ALS database that includes patients referred from October 2002 to July 2008.

We identified 124 patients (45% men, 55% women). Mean age at disease onset was 62 years (range 35–81, SD = 10 years), 63 years (SD = 10) in women and 61 years (SD = 11) in men. The disease started as spinal form in 58%, and as bulbar form in 27%. Fifteen percent of patients could not be unequivocally classified in either of these two categories. Four patients (3%) had familial form of the disease. According to El Escorial diagnostic criteria (EDC) at the time of first referral, 26% of patients had definite, 40% probable, 12% possible, and 22% suspicious form of ALS.

Walking problems in 77% of patients started on average 10 months after admission (range 0–96, SD = 17). Forty percent of them were unable to ambulate independently on average 25 months after admission (range 2–105, SD = 23). Twenty-eight percent of patients were unable to feed themselves, what on average happened 23 months after the diagnosis (range 5–69, SD = 16). Fifty-eight percent had speech problems that started on average after 11 months (range 0–102, SD = 19), 32% became anarthric (on average after 21 months, range 5–72, SD = 15). Swallowing problems had 62% of patients, on average 16 months (range 0–110, SD = 21) after disease onset and in 27% percutaneous endoscopic gastrostomy (PEG) was performed (on average after 25 months after making the diagnosis, range 5–96, SD = 21). Breathing problems that on average started 25 months after admission (range 2–108, SD = 24) had 52% of patients, and 11% opted for noninvasive ventilation, on average 30 months after the diagnosis was made (range 7–110, SD = 26) while only 2 patients were tracheotomised. Among most frequently used drugs were riluzole (36% of patients) and glycopyrrolate (22% of patients), both drugs being registered in Slovenia since 2005, followed by antidepressants (21% of patients) and quinidine (12% of patients).

Sixty-five percent of patients from our register had already died. The median survival time from symptom onset was 28 months (range 3–111, SD = 19) with the median age of death at 66 years (range 45–82, SD = 10). The median survival time after admission was 13 months (range 1–39, SD = 10). In those with PEG, the median survival time after this intervention was 7 months (range 0–23, SD = 7).

The survival curves for the following variables were calculated by Kaplan-Meier method and compared with the log-rank test. The hazard ratio was calculated for each variable. The variables were: male vs. female from the first symptom onset, male vs. female from the time of referral to our clinic, patients with bulbar vs. spinal forms of the disease, median delay between the symptom onset and time of diagnosis (less or more than 15 months), median age at the onset of first symptom (older and younger than 62 years), median grade of ALS Functional Rating Scale score at the time of referral (below and above 31 points), and median Norris ALS Disability Scale score at the time of referral (below and above 80 points). The significantly better survival was identified only for patients with longer diagnostic delay (over 15 months; log-rank chi square = 23.23, $p < 0.0001$, HR (95% CI) = 3.7) and in a younger age group (< 62 years at the time of the first symptom, log-rank chi square = 9.98, $p < 0.0016$, HR (95% CI) = 2.1).

Our database, that is regularly updated, was not designed to serve for the descriptive epidemiological studies and is insufficient in many other aspects as well. Its main aim was to support other possible clinical studies that nevertheless necessitate a prospective collection of separate sets of data.

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