|  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Table S1. Number and distribution of discharge diagnoses by type of admission, in hospitalizations ending with death. | | | | | | | | | | | | | | | | | | | | |
|  | | Type of admission | | | | | | | | | | | | | | |  | | | |
|  | | Emergency  (N = 97) | | | |  | Elective  (N=16) | | | |  | Day Hospital  (N = 1) | | | |  | Total  (N=114) | | | |
|  | |  |  |  |
| Discharge diagnosis  (ICD-9-CM codes) | | Primary code1 | | Any  code2 | |  | Primary code1 | | Any  code2 | |  | Primary code1 | | Any  code2 | |  | Primary code1 | | Any  code2 | |
| N | % | N | % |  | N | % | N | % |  | N | % | N | % |  | N | % | N | % |
| ALS (335.20) | | 40 | 41.2 | 85 | 87.6 |  | 1 | 31.2 | 11 | 68.7 |  | 1 | 100.0 | 1 | 100.0 |  | 46 | 40.3 | 97 | 85.0 |
| respiratory failure (518.81-518.84) | | 21 | 21.6 | 56 | 57.7 |  | 5 | 31.2 | 11 | 68.7 |  | - | - | - | - |  | 26 | 22.8 | 67 | 58.8 |
|  | acute (518.81) | - | - | 38 | 39.1 |  | - | - | 10 | 62.5 |  | - | - | - | - |  | - | - | 49 | 42.9 |
|  | chronic (518.83) | - | - | - | - |  | - | - | 1 | 6.2 |  | - | - | - | - |  | - | - | 1 | 0.8 |
|  | acute on chronic (518.84) | - | - | 8 | 8.2 |  | - | - | - | - |  | - | - | - | - |  | - | - | 8 | 7.0 |
|  | unidentified (518.82) | - | - | 10 | 10.3 |  | - | - | - | - |  | - | - | - | - |  | - | - | 10 | 8.7 |
| diseases of respiratory system (460-519) | | 14 | 14.4 | 43 | 44.3 |  | - | - | 7 | 43.7 |  | - | - | - | - |  | 14 | 12.2 | 50 | 43.9 |
|  | pneumonia and influenza (480-487) | - | - | 31 | 31.9 |  | - | - | 6 | 37.5 |  | - | - | - | - |  | - | - | 37 | 32.4 |
|  | aspiration pneumonia (507.0) | - | - | 4 | 4.1 |  | - | - | 1 | 6.2 |  | - | - | - | - |  | - | - | 5 | 4.3 |
| diseases of circulatory system (390-459) | | 5 | 5.1 | 39 | 40.2 |  | 1 | 6.2 | 3 | 18.7 |  | - | .- | 1 | 100.0 |  | 6 | 5.2 | 43 | 37.7 |
| symptoms, signs, ill-defined conditions (780-799) | | 1 | 1.0 | 32 | 32.9 |  | 2 | 12.5 | 4 | 25.0 |  | - | - | - | - |  | 3 | 2.6 | 36 | 31.5 |
| endocrine, nutritional and metabolic diseases, and immunity disorders (240-279) | | 1 | 1.0 | 14 | 14.4 |  | - | - | 2 | 12.5 |  | - | - | - | - |  | 1 | 0.8 | 16 | 14.0 |
| diseases of nervous system and sense organs (320-389) | | 3 | 3.0 | 8 | 8.2 |  | 2 | 12.5 | 5 | 31.2 |  | - | - | - | - |  | 5 | 4.3 | 13 | 11.4 |
| diseases of digestive system  (520-579) | | 2 | 2.0 | 5 | 5.1 |  | 1 | 6.2 | 1 | 6.2 |  | - | - | - | - |  | 3 | 2.6 | 6 | 5.2 |
| diseases of musculoskeletal system and connective tissue (710-739) | | - | - | 5 | 5.1 |  | - | - | - | - |  | - | - | - | - |  | - | - | 5 | 4.3 |
| infectious and parasitic diseases (001-139) | | 1 | 1.0 | 3 | 3.0 |  | - | - | 2 | 12.5 |  | - | - | - | - |  | 1 | 0.8 | 5 | 4.3 |
| diseases of genitourinary system (580-629) | | 1 | 1.0 | 3 | 3.0 |  | - | - | 1 | 6.2 |  | - | - | - | - |  | 1 | 0.8 | 4 | 3.5 |
| injury and poisoning (800-999) | | - | - | 2 | 2.0 |  | - | - | 2 | 12.5 |  | - | - | - | - |  | - | - | 4 | 3.5 |
| neoplasms (140-239) | | 1 | 1.0 | 2 | 2.0 |  | - | - | - | - |  | - | - | - | - |  | 1 | 0.8 | 2 | 1.7 |
| mental disorders (290-319) | | - | - | 2 | 2.0 |  | - | - | - | - |  | - | - | - | - |  | - | - | 2 | 1.7 |
| factors influencing health status and contact with health services (v01-v82) | | 7 | 7.2 | - | - |  | - | - | - | - |  | - | - | - | - |  | 7 | 6.1 | - | - |
| 1 Number and percentage (%) of hospitalizations with the corresponding discharge diagnosis as primary code.  2 Number and percentage (%) of hospitalizations with the corresponding discharge diagnosis as primary or any secondary code. | | | | | | | | | | | | | | | | | | | | |

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| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Table S2. Relation between mortality, hospitalizations for respiratory failure and clinical characteristics. Hazard Ratio (HR), with 95% Confidence Interval (95%CI). | | | | | | | | | | | | | | | | |
|  | | Univariate | | |  | Multivariate1 | | |  | Multivariate2 | | |  | Multivariate3 | | |
|  | | p | HR | 95%CI |  | p | HR | 95%CI |  | p | HR | 95%CI |  | p | HR | 95%CI |
| Hospitalization for respiratory failure after ALS diagnosis | |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
|  | No4 |  | 1.00 | - - |  |  | 1.00 | - - |  |  | 1.00 | - - |  |  | 1.00 | - - |
|  | Yes | <0.0001 | 3.52 | 2.65; 4.67 |  | <0.0001 | 4.53 | 3.35; 6.12 |  | <0.0001 | 4.30 | 3.22; 5.75 |  | <0.0001 | 4.00 | 3.00; 5.34 |
| Site of onset | |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
|  | spinal4 |  | 1.00 | - - |  |  | 1.00 | - - |  |  | 1.00 | - - |  |  | 1.00 | - - |
|  | bulbar | 0.0005 | 1.71 | 1.27; 2.32 |  | 0.1346 | 1.28 | 0.93; 1.77 |  | 0.1413 | 1.27 | 0.92; 1.75 |  | 0.0791 | 1.32 | 0.97; 1.81 |
|  | missing | 0.0084 | 1.69 | 1.14; 2.49 |  | 0.2243 | 1.32 | 0.84; 2.05 |  | 0.1615 | 1.37 | 0.09; 2.11 |  | 0.2113 | 1.31 | 0.86; 2.01 |
| Age (years) | |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
|  | <684 |  | 1.00 | - - |  |  | 1.00 | - - |  |  | 1.00 | - - |  |  | 1.00 | - - |
|  | ≥68 | <0.0001 | 2.05 | 1.56; 2.69 |  | <0.0001 | 2.94 | 2.12; 4.08 |  | <0.0001 | 2.72 | 2.01; 3.70 |  | <0.0001 | 2.50 | 1.86; 3.36 |
| Delay to diagnosis | |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
|  | ≥ median4, 5 |  | 1.00 | - - |  |  | 1.00 | - - |  |  | 1.00 | - - |  |  | 1.00 | - - |
|  | < median5 | 0.0065 | 1.81 | 1.18; 2.77 |  | <0.0001 | 2.58 | 1.65; 4.05 |  | <0.0001 | 2.53 | 1.62; 3.97 |  | 0.0004 | 2.23 | 1.43; 3.47 |
|  | unknown | 0.1095 | 1.34 | 0.94; 1.91 |  | 0.0042 | 1.80 | 1.20; 2.69 |  | 0.0059 | 1.76 | 1.18; 2.63 |  | 0.0459 | 1.48 | 1.01; 2.19 |
| Charlson Index | |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
|  | 04 |  | 1.00 | - - |  |  | 1.00 | - - |  |  | 1.00 | - - |  | - | - | - - |
|  | 1-2 | 0.8568 | 0.97 | 0.72; 1.31 |  | 0.0704 | 0.74 | 0.54; 1.02 |  | 0.0869 | 0.76 | 0.55; 1.04 |  | - | - | - - |
|  | ≥ 3 | 0.3208 | 0.82 | 0.55; 1.22 |  | 0.0023 | 0.50 | 0.32; 0.78 |  | 0.0005 | 0.48 | 0.31; 0.72 |  | - | - | - - |
| Sex | |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
|  | Men4 |  | 1.00 | - - |  |  | 1.00 | - - |  | - | - | - - |  | - | - | - - |
|  | Women | 0.0720 | 1.28 | 0.98; 1.69 |  | 0.2988 | 0.85 | 0.63; 1.15 |  | - | - | - - |  | - | - | - - |
| Hospitalization for respiratory failure before ALS diagnosis | |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
|  | No4 |  | 1.00 | - - |  |  | 1.00 | - - |  | - | - | - - |  | - | - | - - |
|  | Yes | 0.0871 | 1.68 | 0.93; 3.03 |  | 0.3783 | 0.74 | 0.37; 1.46 |  | - | - | - - |  | - | - | - - |
| 1 Includes terms for site of onset, age, lag time to diagnosis, sex, hospitalization for RF before ALS diagnosis, Charlton Index.  2 Includes terms for site of onset, age, lag time to diagnosis, Charlson Index.  3 Includes terms for site of onset, age, lag time to diagnosis.  4 Reference category.  5 Median: 276 days. | | | | | | | | | | | | | | | | |