Table S1. Characteristics of the five included studies

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| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| First author | Year | Study location | ALS definition | ALS | | Control | | Variables controlled | NOS score |
| N | Cr（Mean+SD） | N | Cr（Mean+SD） |
| Fan | 2016 | China | EEC | 164 | 67±16μmol/L | 164 | 81 ±15μmol/L | Age and gender | 7 |
| Ikeda | 2012 | Japan | REEC | 92 | 46.85±15.91μmol/L | 92 | 65.42±17.68μmol/L | Age, gender, and BMI | 7 |
| Tetsuka | 2013 | Japan | REEC | 76 | 38.90±19.45μmol/L | 30 | 52.16±16.80μmol/L | Age | 6 |
| Huang | 2014 | China | EEC | 80 | 60.86±16.8μmol/L | 80 | 66.97±14.14μmol/L | Age | 6 |
| Chen | 2019 | China | REEC | 571 | 62.48±29.23μmol/L | 571 | 80.67±15.69μmol/L | Age, gender, and BMI | 7 |

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| ALS=Amyotrophic lateral sclerosis; EEC=EI Escorial criteria; REEC=Revised El Escorial criteria; Cr=Creatinine; SD=standard derivation; BMI=Body mass index; NOS= Newcastle-Ottawa Scale |

Table S2. Characteristics of the three included studies

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| First author | Year | Study location | ALS definition | Duration of follow-up (months) | Study size | Adjusted variables | NOS score |
| Wei | 2018 | China | EECR | 48-96 | 553 | |  | | --- | | Age at onset, Disease duration, Diagnostic delay, ALSFRS-R and Disease progression | | 8 |
| van eijk | 2018 | Netherlands | EECR | 12 | 1241 | |  | | --- | | Age, gender, site of onset, Disease duration, FVC and ALSFRS-R | | 8 |
| Atassi | 2014 | Boston | EECR | 40 | 8600 | Age, gender, Disease duration, VC and ALSFRS-R | 7 |

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| ALS=Amyotrophic lateral sclerosis; EEC=EI Escorial criteria; REEC=Revised El Escorial criteria; ALSFRS-R=ALS Functional Rating Scale-Revised; BMI=Body mass index; NOS= Newcastle-Ottawa Scale |