

## Supplementary file

### Strategy used in the estimation of numerator

Initially, Poisson regression model was established based on  $M_1$  (or  $M_2$ ) and  $N_2$ , considering covariates including age, sex and insurance type.  $M_1 + M_3$  (or  $M_2 + M_4$ ) was then estimated by replacing  $N_2$  with  $N_2 + N_3$  in the Poisson regression model. Based on normal distribution, additional nine estimates of each province were selected from the 95% CI of the estimate from Poisson regression. According to Rubin's Rule, the ten estimates were combined to calculate the pooled prevalence or incidence for each province.

**eTable 1 The population coverage rate of claims data by province in China**

Province	Coverage rate
Shanxi	75.78%
Inner Mongolia	81.43%
Liaoning	80.01%
Jilin	90.90%
Heilongjiang	82.88%
Jiangsu	86.12%
Zhejiang	131.91%
Anhui	57.15%
Jiangxi	93.74%
Shandong	145.98%
Henan	88.79%
Hubei	81.13%
Hunan	97.14%
Guangdong	132.82%
Guangxi	83.67%
Hainan	79.02%
Chongqing	176.18%
Guizhou	57.73%
Yunnan	87.53%
Shaanxi	60.85%
Gansu	89.70%

Note: The coverage over 100% was caused by population mobility. In China, individuals are required to be registered generally in the city where they were born, but the Urban Employee Basic Medical Insurance is required for all the employees in the province even though they were not the permanent residents in the province.

**eTable 2 Comparison of basic characteristics between included and excluded provinces**

Characteristic		Included provinces	Excluded provinces
Age, years	Mean (SD)	42.51 (17.30)	41.54 (16.94)
Sex, n (%)	Male	224.54 (52.27)	79.84 (52.27)
	Female	205.08 (47.73)	72.90 (47.73)
Mean GDP (billion yuan)	Mean (SD)	2847.52 (2224.70)	1820.91 (1295.92)
Nationality, n (%)	Han	970.07 (92.27)	250.78 (89.09)
	Ethnic minority	81.26 (7.73)	30.71 (10.91)

Note: 2010 national census data and 2016 GDP data in China was used.

**eTable 3 Crude prevalence of amyotrophic lateral sclerosis in 21 provinces in China in 2016, grouped by sex, age group, and region (unit: /100 000 person-years)**

	Prevalence (95% CI)	
	Male	Female
Total	3.45 (2.72-4.26)	2.35 (1.86-2.90)
Age group		
18-29	1.34 (0.98-1.77)	1.30 (0.95-1.70)
30-34	3.10 (2.22-4.12)	2.39 (1.74-3.14)
35-39	3.37 (2.47-4.41)	2.45 (1.82-3.17)
40-44	3.37 (2.55-4.29)	2.38 (1.83-3.00)
45-49	3.60 (2.79-4.51)	2.37 (1.81-3.01)
50-54	4.24 (3.34-5.24)	2.74 (2.10-3.48)
55-59	4.94 (3.89-6.11)	3.04 (2.35-3.82)
60-64	5.68 (4.46-7.04)	3.20 (2.40-4.12)
65-69	6.08 (4.75-7.57)	3.42 (2.50-4.47)
70-74	6.86 (5.30-8.63)	3.74 (2.68-4.97)
75-79	7.53 (5.76-9.53)	3.94 (2.83-5.23)
80-84	7.26 (5.26-9.57)	3.51 (2.35-4.90)
>=85	5.26 (3.48-7.41)	2.66 (1.51-4.14)
Region		
East	6.35 (5.33-7.47)	4.11 (2.98-5.43)
North	2.22 (1.81-2.66)	1.40 (1.07-1.77)
North-East	2.31 (0.96-4.23)	1.72 (1.21-2.33)
North-West	2.28 (0.72-4.72)	1.32 (0.60-2.31)
South-Central	3.24 (1.99-4.79)	2.44 (1.42-3.74)
South-West	2.81 (2.14-3.57)	1.76 (1.56-1.96)

**eTable 4 Prevalence and incidence of amyotrophic lateral sclerosis in different regions of China in 2016 (unit: /100 000 person-years)**

Region	Prevalence (95% CI)	Incidence (95% CI)
East	5.31 (4.27-6.47)	2.57 (1.93-3.30)
North	1.80 (1.54-2.09)	0.95 (0.66-1.29)
North-East	2.02 (1.10-3.21)	1.10 (0.76-1.51)
North-West	1.86 (0.69-3.61)	0.61 (0.42-0.84)
South-Central	2.84 (1.76-4.18)	1.91 (1.10-2.93)
South-West	2.30 (1.90-2.75)	1.23 (1.05-1.43)

**eTable 5 Standardized prevalence and incidence of amyotrophic lateral sclerosis in different regions of China in 2016 (unit: /100 000 person-years)**

Region	Prevalence (95% CI)	Incidence (95% CI)
East	4.99 (4.86-5.11)	2.29 (2.21-2.38)
North	1.90 (1.67-2.15)	0.95 (0.79-1.13)
North-East	1.89 (1.74-2.05)	0.97 (0.87-1.09)
North-West	1.49 (1.24-1.77)	0.57 (0.39-0.79)
South-Central	2.98 (2.85-3.10)	1.93 (1.83-2.03)
South-West	2.21 (2.06-2.36)	1.11 (1.00-1.22)

**eTable 6 Crude prevalence and incidence of amyotrophic lateral sclerosis in China in 2016, grouped by province (unit: /100 000 person-years)**

Province	Prevalence (95% CI)	Incidence (95% CI) <sup>a</sup>
Shanxi	1.69 (1.30-2.08)	0.79 (0.53-1.05)
Inner Mongolia	1.90 (1.51-2.28)	1.11 (0.82-1.41)
Liaoning	2.88 (2.60-3.16)	1.07 (0.90-1.24)
Jilin	1.24 (0.95-1.52)	0.78 (0.56-1.00)
Heilongjiang	2.09 (1.62-2.55)	1.58 (1.17-1.98)
Jiangsu	5.26 (4.95-5.56)	2.09 (1.89-2.29)
Zhejiang	4.86 (4.54-5.19)	1.80 (1.60-1.99)
Anhui	7.08 (6.34-7.83)	3.72 (3.19-4.25)
Jiangxi	6.22 (5.45-6.99)	4.15 (3.54-4.76)
Shandong	3.63 (3.42-3.84)	1.72 (1.58-1.86)
Henan	3.09 (2.68-3.51)	1.79 (1.47-2.10)
Hubei	6.72 (5.87-7.58)	4.64 (3.94-5.34)
Hunan	2.80 (2.43-3.18)	1.41 (1.14-1.67)
Guangdong	1.74 (1.58-1.89)	1.02 (0.90-1.14)
Guangxi	3.28 (2.71-3.85)	2.99 (2.44-3.53)
Hainan	0.88 (0.42-1.34)	0.74 (0.32-1.16)
Chongqing	2.34 (2.14-2.54)	1.13 (1.00-1.27)
Guizhou	1.60 (0.99-2.21)	1.60 (0.99-2.21)
Yunnan	2.68 (2.30-3.06)	1.30 (1.04-1.56)
Shaanxi	1.21 (0.92-1.50)	0.61 (0.41-0.82)
Gansu <sup>a</sup>	2.71 (1.95-3.47)	-
Total	2.91 (2.31-3.58)	1.65 (1.33-2.01)

<sup>a</sup> One province (Gansu) was excluded due to the time ranges <4 years.

**eTable 7 Crude incidence of amyotrophic lateral sclerosis in 20 provinces in China in 2016, grouped by sex, age group, and region (unit: /100 000 person-years)**

	Incidence (95% CI)	
	Male	Female
Total	1.90 (1.52-2.33)	1.38 (1.09-1.70)
Age group		
18-29	0.76 (0.54-1.01)	0.75 (0.54-0.99)
30-34	1.63 (1.14-2.20)	1.28 (0.92-1.69)
35-39	1.75 (1.25-2.34)	1.33 (0.96-1.75)
40-44	1.76 (1.33-2.26)	1.27 (0.98-1.59)
45-49	1.87 (1.43-2.37)	1.25 (0.95-1.58)
50-54	2.20 (1.72-2.74)	1.43 (1.10-1.80)
55-59	2.50 (1.98-3.08)	1.42 (1.13-1.76)
60-64	2.91 (2.30-3.58)	1.60 (1.21-2.04)
65-69	3.00 (2.39-3.69)	1.60 (1.21-2.06)
70-74	3.21 (2.56-3.93)	1.70 (1.27-2.20)
75-79	3.50 (2.79-4.29)	1.82 (1.30-2.43)
80-84	3.27 (2.46-4.21)	1.65 (1.12-2.27)
≥85	2.56 (1.71-3.58)	1.35 (0.78-2.07)
Region		
East	3.04 (2.34-3.82)	2.01 (1.37-2.78)
North	1.02 (0.51-1.70)	0.88 (0.63-1.19)
North-East	1.34 (0.82-1.99)	0.85 (0.66-1.05)
North-West	0.53 (0.29-0.84)	0.71 (0.42-1.08)
South-Central	2.12 (1.22-3.26)	1.68 (0.89-2.72)
South-West	1.52 (1.30-1.77)	0.89 (0.70-1.10)

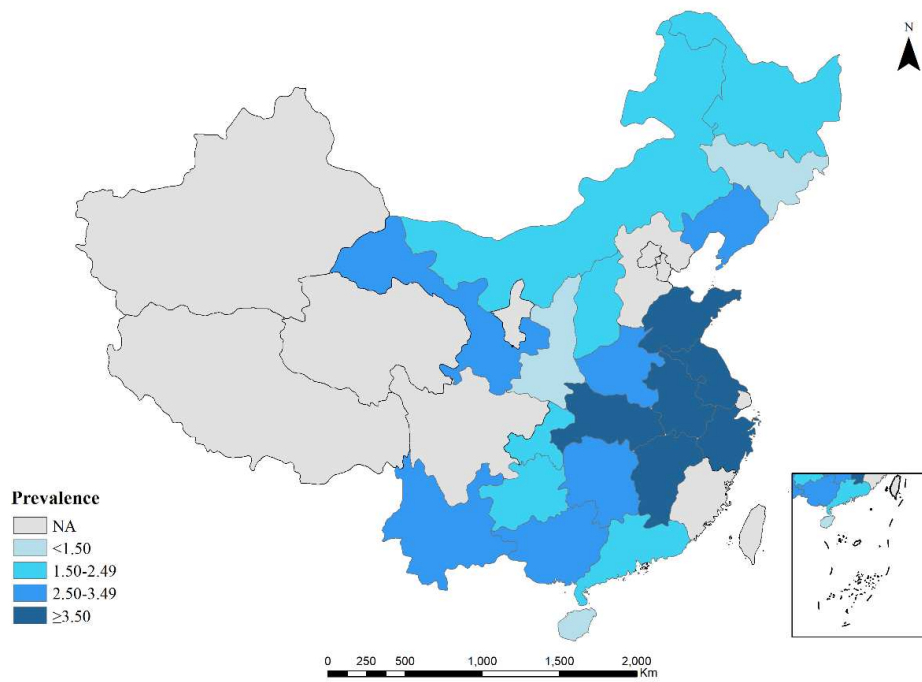
**eTable 8 Results of sensitivity analysis for prevalence and incidence (unit: /100 000 person-years)**

	Including all uncertain ALS cases	Using only observed cases <sup>a</sup>	Excluding the top 10% of provinces with missing diagnostic information <sup>b</sup>
Prevalence			
Overall	3.21 (2.54-3.96)	2.17 (1.74-2.65)	2.73 (2.08-3.47)
Male	3.81 (2.98-4.73)	2.42 (1.92-2.97)	3.23 (2.45-4.12)
Female	2.59 (2.04-3.19)	1.94 (1.51-2.42)	2.22 (1.69-2.81)
Incidence			
Overall	1.77 (1.43-2.16)	1.38 (1.13-1.66)	1.55 (1.2-1.94)
Male	2.03 (1.62-2.49)	1.49 (1.23-1.77)	1.78 (1.37-2.25)
Female	1.49 (1.18-1.83)	1.21 (0.93-1.52)	1.3 (1.01-1.62)

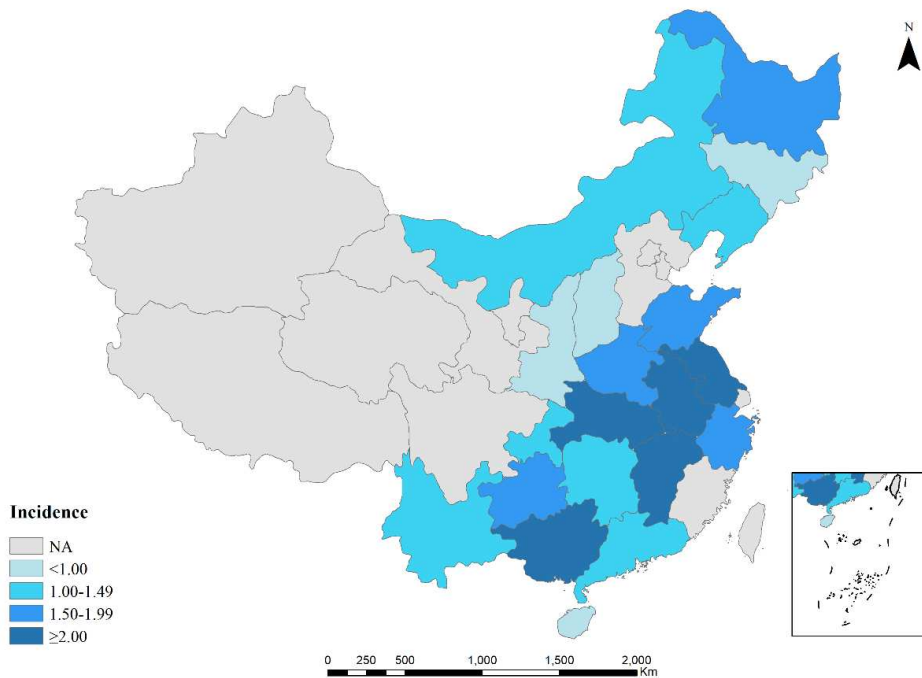
<sup>a</sup> Known to be an underestimation of rates.

<sup>b</sup> Shandong and Jiangxi were excluded.

A



B



**eFigure 1 Distribution of amyotrophic lateral sclerosis in China in 2016**

A. Prevalence; B. Incidence (unit: /100 000 person-years)