

Analysis on Health-related Quality of Life of 180 Chinese Patients with Sporadic Creutzfeldt-Jakob Disease

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Abstract

Aims Sporadic Creutzfeldt-Jakob disease (sCJD) heavily affects the patient's motor, cognitive, and psychological functions with 100% fatality rate. Yet, there are few studies focusing on the impact of sCJD on the health-related quality of life (HRQoL) of the patients with validated instrument. We examined physical and mental conditions of those sCJD patients in China for the first time.

Methods With EQ-5D-3L instrument, a telephone-based cross-sectional study was conducted in order to obtain health statuses of 180 probable Chinese sCJD cases diagnosed in the National Center for CJD surveillance from January 2017 to July 2018. Besides, a visual analogue score (VAS) was performed to evaluate the health status of the sCJD patients from the family members and relatives who nursing the patients.

Results Nearly all of 180 sCJD patients showed the apparent problems in the demission of mobility, usual activities and self-care. More than 1/3 of patients (38.33%) recalled the moderate and severe problems in the demission of pain/uncomfortable. 47.78% of them had the problems in the demission of anxiety/depression. The health utility of sCJD patients was low, with the median of 0.206. Meanwhile, the EQ-VAS score of those sCJD patients was also extremely low, with the median of 0.

Conclusion Compared with the published data of several other diseases, the quality of life of Chinese sCJD patients was extremely poor as demonstrated by the results of the generic EQ-5D-3L. More social attentions need to be paid to reduce the healthy and economic burdens for sCJD patients and their families.

Introduction

Human prion diseases include Creutzfeldt–Jakob disease (CJD), Kuru, fatal familial insomnia (FFI) and Gerstmann–Straussler–Scheinker syndrome (GSS) [1, 2]. The incidence of CJD is around 1 to 2 cases per million of population annually. Among all CJD subtypes, sporadic CJD (sCJD) accounts for nearly 85–95%, while 5–15% of CJD cases are familial or genetic CJD (fCJD or gCJD). Less than 1% CJD cases are acquired that are related with a definite medical history and termed as iatrogenic CJD [3]. A new type of CJD termed as variant CJD (vCJD) has emerged since 1996, which attributes to the outbreak of bovine spongiform encephalopathy (BSE). The clinical manifestations of CJD may vary largely. A couple of neuropsychological symptoms are frequently observed in CJD patients, particularly in sCJD cases, mainly encompassing progressive dementia, myoclonus, visual or cerebella disturbance, pyramidal or extrapyramidal dysfunction and akinetic mutism, which are used in the diagnostic criteria for CJD. With serious conditions of health and rapid progression, CJD patients eventually develop to disability and death.

The World Health Organization (WHO) defines health as “not only the absence of physical defects and diseases, but also a complete physical, mental status and good social adaptation” in 1989. The traditional indicators, such as incidence rate and prevalence rate, do not meet the demand of evaluating health status completely. Thereby the concept and definition of health-related quality of life (HRQoL) have been proposed and widely accepted. European Quality of Life-5 Dimensions instrument (EQ-5D) is a measurement for health status, which allows comparisons of preference-based health-related quality of life across diverse populations. As a multi-dimensional measurement, EQ-5D instrument is easy to understand and has a high reliability and validity [4, 5]. This scale is suitable for the general population [6] and the population with diseases [7, 8], as well as

the patients with cognitive impairment [9, 10]. A study has even [11] suggested that the EQ-5D might have advantages over other dementia-specific measures and can be used routinely as a stand-alone measure of quality of life in dementia research. Actually, the quality of life of many neurodegeneration diseases has been evaluated with the methodology of EQ-5D instrument [12-14]. However, the relevant data for human prion diseases are less described.

In this study we recruited 180 probable-diagnosed Chinese sCJD cases referred to the National Center for CJD surveillance from 2017 to 2018. The quality of life of those patients was assessed with multi-dimensional EQ-5D-3L questionnaire. We found significant impediment in the dimensions of mobility, self-care and usual activities. The utility scores and the visual analogue scale (VAS) of sCJD patients were very low.

Materials And Methods

Study population

Totally 180 probable sCJD cases diagnosed by the Center for CJD surveillance China from January 2017 to July 2018 were enrolled into this study. Patients were diagnosed and subtyped according to the diagnostic criteria [3] issued by WHO and the CJD diagnostic criteria issued by China CDC. An expert board consisting of neurologists, neuropathologists, epidemiologists and laboratory staff made the final diagnosis after evaluating the data.

Data collection

Information related to the disease, such as the epidemiological data, the main clinical manifestations, the foremost symptoms, the special abnormalities on MRI brain scan, periodic sharp wave complexes (PSWC) on EEG and test results of protein 14-3-3 in cerebrospinal fluid (CSF) were collected with a designed questionnaire through the monitoring network. A retrospective investigation then was conducted through telephone inquiring by the staff of Center for CJD surveillance with EQ-5D-3L and EQ-VAS questionnaire. During the telephone follow-up, patients were excluded because of wrong number or refusal to answer for at least three times. As the rapid progression of disease, the close relatives who knew patients well, including parents, spouses or offspring, were inquired.

EQ-5D-3L

EQ-5D-3L instrument provides evaluation for a total of 243 unique health states described, using a system comprising five dimensions as mobility (MO), self-care (SC), usual activities (UA), pain/discomfort (PD) and anxiety/depression (AD) at three levels (1=no problems, 2=some/moderate problems and 3=extreme problems) for each dimension. The questionnaire also includes a 20-cm visual analogue scale (VAS) for self-assessment of general health status, with 100 representing the best imagine health status and zero representing the worst health status.

Since the utility of EQ-5D-3L health states can be also influenced by social elements, e.g., culture, social economic, etc^[15, 16]. Several countries, such as UK, USA and Japan, have already developed their national EQ-5D-3L value sets. Several years ago, a Chinese population (social preference) value set for EQ-5D-3L was proposed using a time-trade off (TTO) approach^[17]. The formula is concluded as $U = 1 \times (C + 0.099 \times MO2 + 0.246 \times MO3 + 0.105 \times SC2 + 0.208 \times SC3 + 0.074 \times UA2 + 0.193 \times UA3 + 0.092 \times PD2 + 0.236 \times PD3 + 0.086 \times AD2 + 0.205 \times AD3 + 0.022 \times N3)$. MO2, SC2, UA2, PD2 and AD2 mean the patients are at level 2 of respective dimensions and MO3, SC3, UA3, PD3, AD3 indicate at level 3. C means a constant of 0.039 and N3 represents at least one dimension of 3 level else N3 equals 0. Utility scores can vary between -0.149 (worst health) and 1.00 (perfect health) with the meaning of 0 is death. A negative value represents a health condition worse than being dead.

Quality control and statistical approach

All investigators were trained by a supervisor before the survey. A database was built by Epidata 3.1 for data logging and double checking. Sociological and disease-related characteristics of sCJD patients were analyzed with SPSS version 22.0 for Windows (SPSS Inc., Chicago, IL, USA). Health-related utilities were calculated with Chinese value sets.

Results

General information of the sCJD patients

A total of 180 probable sCJD cases from 29 different provinces, municipalities and autonomous regions were investigated with the questionnaire. Based on the permanent addresses, Shandong, Henan and Beijing were the top three provinces the patients came from. In geography, 88 (48.89%) cases were from eastern regions, 58 were from the middle region and 34 were from the western. 100 (55.56%) patients lived in rural areas. The number of male patients (95) was slightly more than that of female patients (85) with a ratio of 1.12:1. The average onset age of the patients was 63.16 years old, ranging from 34 years old to 91 years old. Majority of them were in the groups of 56 to 75 years old. In terms of occupation, more than 1/3 of the patients were farmers (40.56%). Retired seniors and housework accounts for 21.11% and 12.22%, respectively.

Clinically, 100% patients displayed progressive dementia in their clinical courses. 159 (88.33%) patients had problems in pyramidal or extrapyramidal dysfunction, 77.22% had myoclonus, 69.44% had visual or cerebella disturbance, and 51% recorded akinetic mutism. 174 (96.67%) patients revealed at least three neurological symptoms. 114 (65.90%) patients were positive of CSF 14-3-3 protein. 86 (54.78%) patients showed PSWC on EEG and 152 (87.86%) patients showed sCJD associated changes on MRI. 113 patients had passed away at the time-point of survey, while 67 were still alive. The general information of those patients was summarized in Table 1.

Table 1
General characteristics of 180 Chinese sCJD patients

Variables		Number	Proportion (%)
Gender	Male	95	52.77
	Female	85	47.13
Age	≤55	37	20.56
	56~65	65	36.11
	66~75	62	34.44
	≥76	16	8.89
Marriage	married	160	88.89
	others	20	11.11
Education*	Primary school	80	46.51
	Junior high school	45	26.16
	Senior high school	42	24.42
	College and above	5	2.91
Occupation	Jobholder	38	21.11
	Farmer	73	40.56
	Businessman	9	5.00
	Retired	38	21.11
	Housework/others	22	12.22
Insurance*	Insured	166	92.22
	Uninsured	14	7.78
Address	Urban	80	44.44
	Rural	100	55.56
Area	East	88	48.89
	Middle	58	32.22
	West	34	18.89
Monthly income*	≤5k	76	49.67
	5k~10k	46	30.07
	10k~15k	21	13.73
	≥15k	10	6.54
Foremost symptoms and test results			

Rapid progressive dementia	Positive	123	68.33
	Negative	57	31.67
Slow progressive dementia	Positive	14	7.78
	Negative	166	92.22
Cortical blindness	Positive	16	8.89
	Negative	164	91.11
Mental symptoms	Positive	74	41.11
	Negative	106	58.89
Visual and cerebellar disorders	Positive	58	32.22
	Negative	122	67.78
Pyramidal or extrapyramidal dysfunction	Positive	64	35.56
	Negative	116	64.44
MRI*	Positive	152	87.86
	Negative	21	12.14
EEG*	Positive	86	54.78
	Negative	71	45.72
14-3-3 protein*	Positive	114	65.90
	Negative	59	34.10
Clinical symptoms			
progressive dementia	Positive	180	100.0
	Negative	0	0.0
Myoclonus	Positive	139	77.22
	Negative	41	22.78
Visual or cerebella disturbance	Positive	125	69.44
	Negative	55	30.56
Pyramidal/extrapyramidal dysfunction	Positive	159	88.33
	Negative	21	11.67
Akinetic mutism	Positive	92	51.11
	Negative	88	48.89
Alive or not	Positive	67	37.22

Negative	113	62.78
*Patients who were unwilling to answer or with no test results were excluded		

The quality of life of the sCJD patients

Using the instrument of EQ-5D-3L, the quality of life of 180 Chinese probable sCJD patients were evaluated. As shown in Table 2, all patients recalled the problems in the dimensions of mobility (100.00%) and usual activities (100.00%), while 99.44% of the patients had problems in the dimension of self-care with only one patient did not report problem in self-care. In terms of the dimension of pain/discomfort, more than one third of the patients (38.33%) had problems. For the emotional dimension, nearly half (47.78%) of the patients experienced anxiety and depression. Further analysis the dead patients and the still alive ones did not identified statistical difference in the dimensions of pain/discomfort ($\chi^2=0.174$, $P= 0.676$) and anxiety/depression ($\chi^2=0.851$, $P= 0.356$).

Table 2
The quality of life of Chinese sCJD patients with EQ-5D-3L

EQ-5D-3L dimension	Level of problems			Number of cases with problems (Proportion)
	No (Proportion)	Some/moderate (Proportion)	Extreme (Proportion)	
Mobility	0 (0.00%)	12 (6.67%)	168 (93.33%)	180 (100.00%)
Self-care	1 (0.56%)	3 (1.67%)	176 (97.78%)	179 (99.44%)
Usual activities	0 (0.0%)	5 (2.78%)	175 (97.22%)	180 (100.00%)
Pain/discomfort	111 (61.67%)	57 (31.67%)	12 (6.66%)	69 (38.33%)
Anxiety/depression	94 (52.22%)	51 (28.33%)	35 (19.44%)	86 (47.78%)

The health utility values of these Chinese sCJD patients were further calculated. The values of the majority cases were in the range of -0.2 to 0.6, with an average value of 0.199 and a median value of 0.206. The P25 and P75 values were 0.114 and 0.292, respectively (Fig 1A). Only 8 cases had the scores larger than 0.3.

The values of EQ-VAS of 180 sCJD patients were also evaluated, ranging from zero to 70, with the average data of 4.81 and the median of zero. Analysis of the data distribution of those cases showed that the VAS scores of 63.33% patients were zero and 12.22% cases were less than 10, while only a small portion (5.00%) of the patients revealed the VAS score larger than 20 (Fig 1B).

The health utilities of those 180 sCJD patients were also calculated by the value sets from other countries in order to see the possible difference and to compare with the data of other diseases reported from different countries. As shown in Fig 2, the utility obtained from Chinese value sets was the highest with a median of 0.206, whereas the utility using UK value set was the lowest with a median of -0.043. The utilities using Japan value set (median: 0.132) and Thailand one (median: 0.037) were at middle level, respectively.

Discussion

In this study, we have evaluated the health-related quality of Chinese sCJD patients. As a universal scale of quality of life, EQ-5D instrument has been widely used in many studies focusing on the quality of life of various general population and the patients with different diseases. However, there is still lacking of the relevant study on the quality of life in the field of human prion disease. To our knowledge, this is the first study that elicits a complete spectrum of utility scores of Chinese sCJD patients by applying the Chinese scoring algorithm. The generation of the EQ-5D-3L scale and its Chinese utility value sets were years early than that of EQ-5D-5L, which leads to the limitation of the wide application of the EQ-5D-5L. Moreover, EQ-5D-3L instrument has been used in the National Health Services Survey (NHSS) 2008 in China. Therefore, we took EQ-5D-3L scale in this study.

CJD is a fatal illness because of its irreversible progression, characterized with long incubation time and rapid disease progression. The duration of sCJD patient are usually shorter than two years, most of them die in half year after onset ^[18]. Besides of rapid progressive dementia that is the most frequent symptom, many other neuropsychiatry symptoms and signs are also noticed in sCJD patients clinically, which involves in human motor, sensory and mental systems. In line with the clinical features, both the EQ-5D utility scores and the VAS scores of the sCJD patients are extremely low. In Table 3, we have summarized some published data calculated by EQ-5D-3L instrument with the values sets from UK, Japan and Thailand, respectively ^[19-26]. If we use the data with the value set from UK, the general health utility value of Chinese sCJD patients is the lowest (median -0.043), much lower than that of AD, PD, and even lower than that of stroke on the first day. If we use the data obtained with the value set of Japan, the utility score of Chinese sCJD cases is also lower than that Japanese children with cerebral palsy (median: 0.44), which is the lowest health utility value among the different diseases evaluated with EQ-5D-3L instrument in the review of Zhou ^[27]. Apparently, sCJD seriously affects the health status and quality of life of the patients with a high disability rate.

Table 3
The published health utilities of some other diseases

Diseases	Utility scores	VAS scores	Value Sets	Location	Patients	Publish time	Reference
Alzheimer's Dementia	0.87±0.17	76±19	UK	Sweden	34	2007	Bostrom et al. 2007
Frontotemporal Dementia	0.824 (IQR=0.241)	74 (IQR=39)	---	Norway, Denmark, and Iceland	38	2018	Hvidsten et al, 2018
Parkinson's disease	0.71 ± 0.24	---	UK	Melbourne, Australia	302	2016	Soh et al, 2016
Stage 2A	0.65	71.65	UK	London, UK	214	2014	Jones et al., 2014
Stage 2B	0.53	67.07					
Stage 3	0.41	59.76					
Stage 4	0.27	56.75					
Huntington's disease	0.54 (SD=0.43)	---	The UK social tariff	Spain	55	2016	Dorey et al., 2016
Cerebral palsy	0.44 (SD=0.12)	27.3 (SD=9.1)	Japan	Hebei province, China	340	2014	Cui, 2014
Dementia with lewy bodies	0.38±0.38	55±17	UK	Sweden	34	2007	Bostrom et al., 2007
Stroke at the first day	0.1275	43.18	UK	China	220	2009	Zhang et al., 2009
Liver failure	0.0 (SD=0.2)	36.4 (SD=17.2)	Thailand	Yunnan Province, China	1040	2014	Che et al., 2014

We have noticed that among the five dimensions, mobility, self-care and usual activities are the mostly affected problems for the sCJD patients. High percentage of the investigated cases has recalled to be severely affected, even in the group of the survival patients who appeared clinical manifestations less than 6 months. Our data here have also revealed that only about 1/3 of sCJD patients had problems in pain/discomfort and nearly half had anxiety/depression. Furthermore, we do not see significant difference in those two dimensions between dead and survival cases. Pain and discomfort reflect subjective feelings. Anxiety and depression are psychological activities or mental problems, which usually need to be determined or judged by professionals. sCJD patients usually loss their recognition capacities and language competences rapidly after onset, which may hinder the accurate descriptions of subjective feeling and psychological abnormalities. Particularly at relatively late stage, patients frequently display akinetic mutism. It is reasonable to assume that the problems

of the dimensions of pain/discomfort and anxiety/depression in the sCJD patients are underestimated during the survey. In addition, the questionnaires in this study are rated largely by the family members of the patients or the persons who are nursing the patients, which may also decline the correctness to address the subjective and sensory problems.

Although EQ-5D instrument has been considered to be a valid and reliable tool to evaluate the quality of life in institutionalized patients with different types of dementia, there is still inevitable difference in collection of information directly from the patients themselves or from their family members and relatives. The use of a proxy rater definitely benefits the feasibility of EQ-5D instrument in the patients with dementia, particularly at advanced stage ^[28], but it will also influence the accuracy of the information collection for the issues regarding to subjective feelings.

sCJD causes more serious outcomes on both sides of patients and their families than many other diseases. Focusing on improving the quality of life of sCJD patients and strengthening social support are practical and effective ways to reduce the healthy and economic burdens for those patients and their families.

Declarations

Fundings

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Competing interests

The authors declare that they have no competing interests.

Ethical Standards

The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national and institutional committees on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008. This study was approved by Ethical Review Committee of National Institute for Vial Disease Prevention and Control, China CDC. Following the principle of informed and voluntary, all signed informed consents were collected and stored by the China CJD Surveillance Center.

Authors' contributions

LZ, CG and XPD designed the study. YW, CC and WZ collected the data. LZ and KX analyzed the available data. CG and LZ prepared the manuscripts. QS and XPD reviewed and edited the manuscript. All authors read and approved the final manuscript.

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Availability of Data and Materials

The data and material supporting the findings could be obtained from Xiaoping Dong, email dongxp238@sina.com.

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Figures

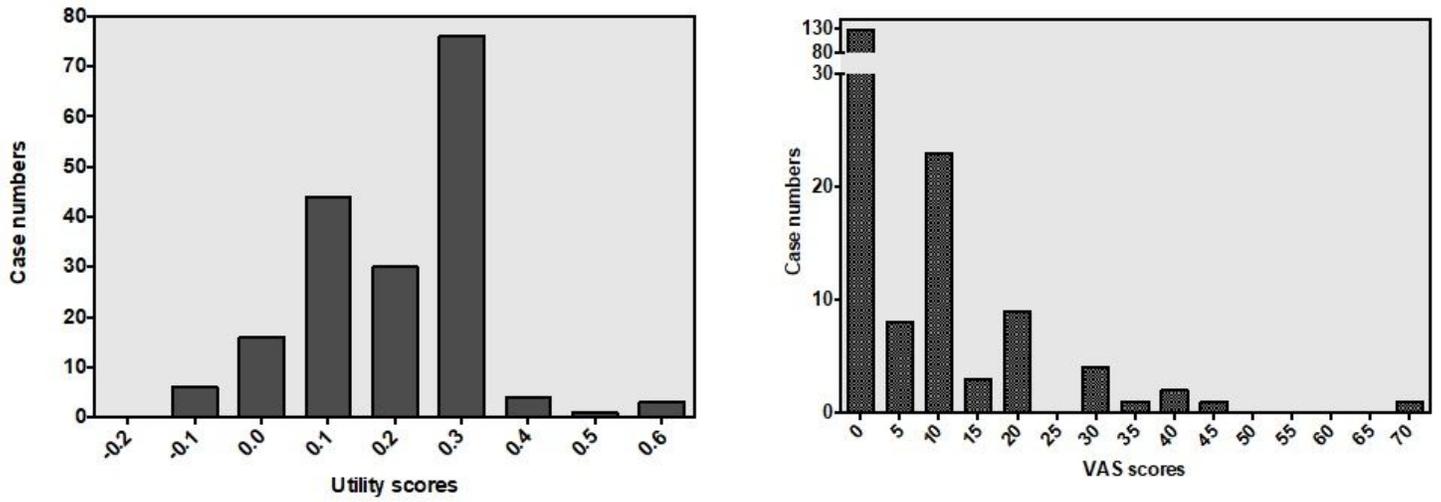


Figure 1

Analyses of health-related utility scores and VAS for Chinese sCJD patients. (A). The distribution of health-related utility scores. (B). The distribution of VAS.

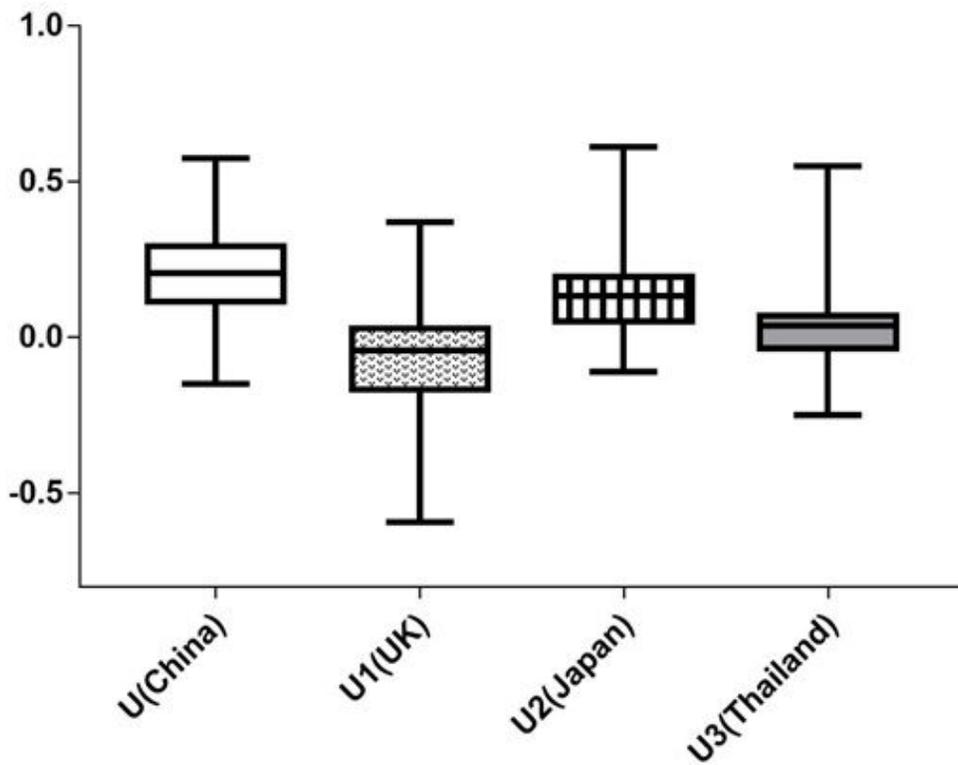


Figure 2

The utilities of Chinese sCJD patients calculated with four different value sets. Graphical data are expressed as median and range