Abstract

Pilot Study of the Quality of Life for Rare Disease Patients in Korea

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Most rare diseases are degenerative and chronic because there is no effective treatment or medicine. Theses factors are thought to be related to poor quality of life (QoL) in rare disease patients. There is a need for a policy to improve patients' QoL, but basic research on QoL of rare disease patients is still lacking in Korea. This study performed a pilot study to assess the feasibility of the 'World Health Organization Quality of Life Scale-Brief' for rare disease patients. The online survey method was used to protect personal information. In this pilot study, rare disease patients had a lower QoL value compared to people without a rare disease. Domains of QoL were highly correlated except for the 'social relationship' domain. In addition, this study could not find any contribution of social relationship to overall QoL. Further study is needed to evaluate QoL considering each rare disease's feature and the factors affecting QoL in a large number of patients with rare diseases.

Keywords: Rare disease, Quality of life, World Health Organization Quality of Life Scale-Brief, Online survey

Table 1. Demographics of respondents by gender

	Men (n=82)	Women (n=94)	Total (n=176)
Age (years)	35.20±14.55	38.76±12.88	37.1± 13.76
Age at diagnosis (years)	25.12±17.11	20.51±16.97	22.65±17.14
Duration of disease* (years)	10.16±13.34	18.58±17.44	14.63±16.17
Age group			
10s	10(12.20)	3(3.19)	13 (7.39)
20s	24(13.64)	21(22.34)	45 (25.57)
30s	21(11.93)	26(27.66)	47 (26.70)
40s	9(10.98)	25(26.60)	34 (19.32)
50s	9(10.98)	11(11.70)	20 (11.36)
60s	9(10.98)	8(8.51)	17 (9.66)
Household number (person)			
1	9(10.98)	14 (14.89)	23 (13.07)
2	14 (17.07)	12 (12.77)	26 (14.77)
3	23 (28.05)	20 (21.28)	43 (24.43)
4	31 (37.80)	35 (37.23)	66 (37.50)
5	5 (6.10)	13 (13.83)	18 (10.23)
Highest education level			
Under middle school	5 (6.10)	4 (4.26)	9 (5.11)
Up to high school	24 (29.27)	25 (26.60)	49 (27.84)
Up to 2−year college	12 (14.63)	12 (12.77)	24 (13.64)
Up to undergraduate degree	34 (41.46)	43 (45.74)	77 (43.75)
Over postgraduate degree	7 (8.54)	10 (10.64)	17 (9.66)
Household income per month (USD)			
<1,000	13 (15.85)	10 (10.64)	23 (13.07)
1,000−2,000	13 (15.85)	15 (15.96)	28 (15.91)
2,000−3,000	20 (24.39)	24 (25.53)	44 (25.00)
3,000−4,000	16 (19.51)	25 (26.60)	41 (23.30)
4,000−6,000	14 (17.07)	10 (10.64)	24 (13.64)
>6,000	6 (7.32)	10 (10.64)	16 (9.09)
Occupation**			
Yes	34 (37.36)	57 (62.64)	94 (54.34)
No	45 (54.88)	34 (37.36)	79(45.66)

Values are mean ± standard deviation or n (%)

* 1 nonrespondent, ** 3 nonrespondents

Table 2. Comparison in the scores of facets and domains of WHOQOL−BREF of rare disease patients by gender

(Mean±SD)

WHOQOL−BREF domains and facets		Men (n=82)	Women (n=94)	Total (n=176)
Overall quality of life		2.59±0.99	2.71±0.99	2.65±0.99
General Health		2.26±1.00	2.27±1.03	2.26±1.01
Domain 1	Physical Domain	10.14±3.32	10.34±3.24	10.25±3.27
Facet 1*	Pain and discomfort	3.04±1.40	2.93±1.41	2.98±1.40
Facet 2	Energy and fatigue	2.28±1.07	2.17±0.90	2.22±0.98
Facet 3	Sleep and rest	2.50±1.14	2.54±1.04	2.52±1.08
Facet 9	Mobility	2.48±1.12	2.65±1.11	2.57±1.12
Facet 10	Activity of daily living	2.57±1.07	2.60±1.01	2.59±1.03
Facet 11*	Dependency on Medication or treatment	3.55±1.19	3.54±1.37	3.55±1.29
Facet 12	Work capacity	2.50±1.11	2.61±1.15	2.56±1.13
Domain 2	Psychological Domain	10.60±3.83	10.24±3.38	10.41±3.59
Facet 4	Positive feelings	2.28±1.11	2.16±0.97	2.22±1.04
Facet 5	Thinking, learning, memory & concentration	2.52±1.14	2.52±1.02	2.52±1.07
Facet 6	Self esteem	2.61±1.10	2.69±1.12	2.65±1.11
Facet 7**	Bodily image and appearance	2.61±1.12	2.24±1.05	2.41±1.10
Facet 8*	Negative feelings	2.78±1.11	2.91±1.05	2.85±1.08
Facet 24	Religion, Spirituality, personal belief	2.66±1.20	2.66±1.16	2.66±1.17
Domain 3**	Social relationships Domain	10.63±3.56	11.63±2.96	11.17±3.28
Facet 13	Personal relationships	2.79±1.12	2.94±1.03	2.87±1.07
Facet 14	Social support	2.89±0.99	3.10±0.83	3.00±0.91
Facet 15**	Sexual activity	2.29±1.07	2.69±0.92	2.51±1.01
Domain 4	Environmental Domain	10.65±3.16	10.58±3.11	10.61±3.12
Facet 16	Physical safety and security	2.48±1.14	2.48±1.04	2.48±1.08
Facet 17	Home environment	3.13±1.11	3.16±1.04	3.15±1.07
Facet 18	Financial resources	1.91±0.97	1.76±0.88	1.83±0.92
Facet 19	Health and social care	3.18±1.02	3.11±1.19	3.14±1.11
Facet 20	Opportunities for acquiring new information and skills	2.63±1.14	2.70±1.05	2.67±1.09
Facet 21	Participation and opportunities for recreation/ leisure	2.16±1.04	2.01±1.05	2.08±1.04
Facet 22	Physical environment	2.77±1.03	2.83±0.96	2.80±0.99
Facet 23	Transport	3.04±1.12	3.12±1.10	3.08±1.10

* Negative question

** P 〈 0.05

Table 3. Pearson's correlation coefficients among overall quality of life, health status and domains

(N=176)

	Overall quality of life	Health status	Physical Domain	Psychological Domain	Social relationships Domain	Environmental Domain
Overall quality of life	1	0.6991	0.6861	0.7081	0.4825	0.6286
		〈.0001	〈.0001	〈.0001	〈.0001	〈.0001
Health status		1	0.7407	0.6097	0.4208	0.4967
			〈.0001	〈.0001	〈.0001	〈.0001
Physical Domain			1	0.7630	0.5624	0.6386
				〈.0001	〈.0001	〈.0001
Psychological Domain				1	0.6750	0.7065
					〈.0001	〈.0001
Social relationships Domain					1	0.5879
						〈.0001
Environmental Domain						1

Table 4. Contribution of individual domain to overall quality of life by gender

	Men		Women		Total	
	β	P	β	P	β	P
Physical Domain	0.10275	0.0152	0.08408	0.0054	0.09284	0.0002
Psychological Domain	0.08762	0.0355	0.12630	0.0003	0.10151	0.0001
Social relationships Domain	−	0.1067	−	0.9991	−	0.3821
Environmental Domain	0.08585	0.0207	0.05032	0.1031	0.06629	0.0052