The NFTI-QOL: A Disease-Specific Quality of Life Questionnaire for Neurofibromatosis 2

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Abstract

The objective of this study was to develop a reliable, validated disease-specific score measuring quality of life (QOL) in clinical practice and treatment trials in Neurofibromatosis 2 (NF2) individuals. In NF2 patients, qualitative interviews ($n = 15$) and a focus group session ($n = 30$) generated items for a pilot questionnaire. This was tested and refined ($n = 20$). The final version (NFTI-QOL) was validated ($n = 50$) with two generic QOL questionnaires (SF-36 and EuroQOL). The NFTI-QOL was also administered to patients with solitary vestibular schwannoma (SVS) ($n = 30$) and normal controls ($n = 30$). The participants were NF2 patients, SVS patients, and normal controls. NFTI-QOL score, SF-36 score, and EuroQOL score were the main outcome measures. Mean NFTI-QOL score was 9.4 (range: 0 to 20, maximum possible score = 24). The NFTI-QOL score correlated strongly with EuroQOL ($r = 0.71$, $p < 0.001$) and SF-36 ($r = 0.81$, $p < 0.001$). NF2 individuals were significantly worse than the SVS patients, who in turn were worse than the controls on the NIFTI-QOL. The NFTI-QOL showed good internal reliability (Cronbach's $\alpha = 0.87$). We developed an eight-item disease-specific QOL score for NF2 patients, validated against SF-36 and EuroQOL. It correlated strongly with clinician-rated disease severity in NF2, with better correlation than the SF-36 in this regard.

Keywords
- NFTI-QOL
- quality of life
- neurofibromatosis 2
- vestibular schwannoma
- SF-36
- EuroQOL

Neurofibromatosis 2 (NF2) is a rare, autosomal dominant, tumor suppressor disorder with a birth incidence of ~1 in 33,000 and a prevalence of 1 in 60,000.[1] NF2 is characterized by bilateral vestibular schwannomas and schwannomas may form on other cranial, spinal, and peripheral nerves. Cranial and spinal meningiomas, ependymomas, and NF2 neuropathy are reported in NF2 patients. Juvenile posterior subcapsular lens opacities and amyotrophy may be the presenting features in children with NF2.[2,3]

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In adults, the initial symptom is usually unilateral or bilateral hearing loss, which may be associated with tinnitus and vertigo. Facial palsy may arise from tumor infiltration or compression, mononeuropathy, or trauma following surgical excision of a vestibular schwannoma. Controversy exists in the management of NF2, with regard to timing and completeness of excision of vestibular schwannomas; the role of hearing-preservation surgery; the indication for "sleeper" auditory brainstem implant and the place of radiotherapy in the management of NF2 tumors.

Advances in the molecular biology of NF2 have led to the identification of potential targets for medical therapies. Preliminary trials of bevacizumab, a vascular endothelial growth factor (VEGF) inhibitor have shown reduction in vestibular schwannoma growth.[4,5] Other potential therapies include PTC-299, which inhibits VEGF produced by tumor cells (PTC Therapeutics) and sorafenib, a platelet-derived growth factor (PDGF), and c-Raf inhibitor.[6] As NF2 is such a rare condition, published outcomes are often small series, and metaanalysis of several studies may be required to demonstrate real differences in treatment groups. It would therefore be advantageous for centers to use comparable tools to measure outcomes in NF2 studies.

Disease progression in NF2 is typically measured in terms of neurological deficit, neuroimaging, and speech and pure tone audiometry. However, the impact of the condition on the quality of life (QOL) of the patient is harder to quantify. Health-related QOL may be assessed using generic or disease-specific questionnaires and is recognized as an important marker of disease progression, a trigger for intervention, and as an outcome measure following intervention.[7] Currently, there are few published data on QOL in NF2. Neary et al assessed QOL in patients with NF2 using the SF-36, a generic QOL questionnaire and their own closed set questionnaire.[8] They found that QOL was significantly impaired in all SF-36 dimensions in NF2, and that social communication problems and balance problems outweighed other symptoms in relation to QOL.[8] Patel et al conducted in-depth qualitative interviews with six patients with NF2, and concluded that NF2 had a negative impact on daily activities and may lead to social isolation.[9] Generic questionnaires such as the Short-Form 36 (SF-36)[10] and EuroQOL[11] are widely used in clinical practice and research but the SF-36 takes a long time for the patient to complete and none of these questionnaires specifically addresses symptoms related to NF2. At present there is no universally accepted, published, and validated disease-specific QOL questionnaire for NF2 that allows quantitative measurement of QOL.

Aim

To develop and validate a disease-specific score to measure QOL in NF2 which is suitable as an assessment tool in clinical practice and in clinical trials and is quick and simple to complete.

Method

The NFTI-QOL was developed using sequential stages (see below 1 to 4) to ensure robust construction. Ethical approval was obtained from the South East London 5 Research and Ethics Committee. The target population was defined as all adults (16yrs or older) who fulfilled the diagnostic criteria for NF2[3] and attend our NSCT NF2 service at Guy's and St. Thomas' NHS Foundation Trust (GSTT). Exclusion criteria included patients under 16 years, and patients who did not read English.

Item Generation

A comprehensive list of symptoms, social and emotional difficulties related to NF2 was produced using the following: literature review (Medline 1966–2011, search terms: NF2), 15 in-depth semistructured interviews with NF2 patients, and a focus group session with 30 NF2 patients and relatives. Sequential patients attending the NSCT NF2 service at Guy's Hospital were invited to participate in the interviews and all 15 who were asked agreed to take part. All patients under the care of the NSCT NF2 service at GSTT were invited to the focus group session.

Developing the Pilot Questionnaire

The list of items was analyzed by a multidisciplinary panel of NF2 specialists and a psychologist to produce a pilot questionnaire which covered all areas generated. The pilot questionnaire was further refined after review by NF2 patients for ease of completion and acceptability. The 31-point pilot questionnaire was then administered to 20 patients with NF2.

Item Reduction

The pilot questionnaire underwent factor analysis to reduce it to seven items. When factor analysis identified items which were highly correlated and consequently showed item redundancy, the highest loading item on a particular factor was chosen to represent that symptom domain. An extra item related to vision, deemed to be clinically important (Q4), was added during the testing to produce the final 8th item for NFTI-QOL (see Appendix **1**).

Validation

The NFTI-QOL was administered with two generic measures of QOL: the SF-36 and the EuroQOL.[10,11] The questionnaires were posted to 65 patients with NF2, who were under the care of the GSTT NSCT NF2 service, with a 77% response rate ($n = 50$). There was no difference in age, gender, or disease severity in the nonresponders versus responders. The NFTI-QOL was also administered to 30 patients with solitary vestibular schwannomas (SVS) (non-NF2) who attended the Guy's Skull Base Clinic and to 30 healthy members of the general population.

Validity was assessed by correlating NFTI-QOL with SF-36 and EuroQOL and also by administering the NFTI-QOL to control groups: patients with SVS (non-NF2) and the general population.

Table 1 NFTI-QOL Results in 50 NF2 Patients

Symptoms/Signs	Not Present (*n*)	Present, but Causes no Difficulty with Usual Activities (*n*)	Causes Some Difficulty with Usual Activities (*n*)	Stops Usual Activities (*n*)
Q1. Hearing problems	8 (16%)	9 (18%)	24 (48%)	9 (18%)
Q2. Dizziness or balance problems	8 (16%)	3 (6%)	32 (64%)	7 (14%)
Q3. Facial weakness	27 (54%)	10 (20%)	10 (20%)	3 (6%)
Q4. Sight problems[a]	15 (40%)	7 (19%)	15 (41%)	0 (0%)
Q5. Mobility	**No problems (n)**	**Some difficulty, can manage on own (n)**	**Unable to walk without help (n)**	**Unable to walk at all (n)**
	10 (38%)	21 (42%)	8 (16%)	2 (4%)
Q6. Role on outlook on life (e.g., confidence, vulnerability, relationships, caring for family, career)	**Positive or no effect (n)**	**Small negative effect (n)**	**Moderate negative effect (n)**	**Large negative effect (n)**
	9 (18%)	16 (32%)	16 (32%)	9 (18%)
Q7.Pain	**None**	**Mild**	**Moderate**	**Severe**
	26 (52%)	12 (24%)	11 (22%)	1 (2%)
Q8.Anxiety and depression	**None**	**Mild**	**Moderate**	**Extreme**
	21 (42%)	15 (30%)	9 (18%)	10 (5%)

[a]Thirty-seven patients were assessed with Q4.

Results

Patient Demographics and Disease Severity

In the interview group ($n = 15$), nine individuals were male and six were female, age range was 17 to 74 years (mean = 40 years). In the pilot questionnaire group ($n = 20$), 10 were female and 10 were male, age range was 17 to 82 years (mean = 46years). The validation group ($n = 50$), comprised 32 females and 18 males, age range was 16 to 82 years (mean = 44years). Due to the small numbers of patients with NF2, there was some overlap between the groups: 9 patients completed the interviews as well as the pilot questionnaire; 12 patients who participated in the interviews and 20 who completed the pilot questionnaire were included in the final validation group.

We have described a method of measuring disease severity in NF2[12] based on clinicians' observation of current status, not future potential for disease. In the pilot questionnaire, we asked patients to rate the severity of their disease (six mild, seven moderate and seven severe). This correlated strongly and significantly with physician-rated severity of disease (six mild, six moderate, eight severe, Pearson correlation $r = 0.75$, $p < 0.001$). In the interview group, physician-rated severity was mild in three, moderate in six, and severe in four patients. In the validation group, physician-rated disease severity was rated as mild in 9, moderate in 22, and severe in 19.

QOL in Patient Group

NFTI-QOL (See Appendix 1 for Questionnaire)

Although the initial interviews and focus group did not indicate that sight and visual problems were an issue in our cohort, it was felt to be an important factor for other patients attending the clinic and for clinicians looking after people

Table 2 European Quality of Life (EuroQOL) Responses in 50 NF2 Patients in the United Kingdom versus Normative Values for Europe

Dimension	Degree of Difficulty or Trouble			Comparison of NF2 Patients vs. Population Norms
	None	Moderate	Extreme	
Mobility	20 (40%) [81.6%]	30 (60%) [18.3%]	0 (0%) [0.1%]	$p < 0.001$
Self-care	34 (68%) [95.8%]	13 (26%) [4.1%]	3 (6%) [0.1%]	$p < 0.001$
Usual activities	19 (38%) [83.7%]	27 (54%) [14.2%]	4 (8%) [2.1%]	$p < 0.001$
Pain/discomfort	21 (41%) [67.0%]	24 (48%) [29.2%]	5 (10%) [3.8%]	$p < 0.001$
Anxiety/depression	29 (58%) [79.1%]	19 (38%) [19.1%]	2 (4%) [1.8%]	$p = 0.001$

Values in brackets are European population norms.[13] Significance of NF2 versus Norms by Binomial test, with categories Moderate and Extreme amalgamated.

Table 3 SF-36 Domain Scores in 50 NF2 Patients in the United Kingdom versus Normative Values for the United Kingdom[14]

	Physical Functioning (PFS)	Role – Physical (RPS)	Role – Emotional (RES)	Social Functioning (SFS)	Mental Health (MHS)	Bodily Pain (PS)	Energy/Vitality (EVS)	Health Perception (HPS)
NF2 Group Mean [95% CI]	57.4 [48.0–66.8]	51.5 [38.6–64.4]	59.3 [46.3–72.4]	64.7 [55.2–74.1]	61.84 [54.3–69.4]	67.3 [59.1–75.6]	49.2 [41.4–57.0]	48.7 [39.8–57.5]
Population normative range of means across all age groups (18–64 years)[14]	73.8–94.7	75.3–92.4	77.9–87.4	84–92.4	70.2–78.8	75.0–87.2	57.1–68.1	67.9–78.2

Note: Higher values indicate better health.

with NF2. Therefore, an additional question on vision was added to the NFTI-QOL during testing (Q4) and 37 of the 50 patients completed the revised score. The mean NFTI-QOL score from the revised questionnaire was 9.4 (SD 5.5), with a range of 0 to 20 (maximum possible score = 24) (►**Table 1**).

EuroQOL

The NF2 group scored significantly worse in mobility, self-care, usual activities, pain/discomfort, anxiety, depression, and the visual analogue score, compared with the population norms (►**Table 2**).

SF-36

The SF-36 is a generic measure of QOL and consists of eight domains: physical functioning; role-physical; role-emotional; social functioning; mental health; energy/vitality; bodily pain, and health perception. Scores in each section are normalized to a scale of 0 to 100 (0 = worst possible health state, 100 = best possible health state). NF2 patients scored worse than the lowest population norm for seven of the eight domains. There was a slight overlap for bodily pain (►**Table 3**).

Validation

The total NFTI-QOL score was correlated with age, gender, and severity score using Pearson Correlation (two-tailed). There was no significant correlation with age ($r = 0.246$,) or gender ($r = 0.025$). Correlation between NFTI-QOL score and physician-rated severity was highly significant ($r = 0.512$, $p < 0.001$). Correlation between total SF-36 and physician-rated severity was highly significant, ($r = -0.419$, $p = 0.002$), but less than NFTI-QOL.

The total NFTI-QOL score correlated significantly with each of the eight domains of the SF-36 ($p \leq 0.001$). It also correlated strongly and significantly to the sum of the eight SF-36 domain scores ($r = -0.806$, $p < 0.001$). The total NFTI-QOL score correlated with the sum of the first five questions of the EuroQOL and also with the visual analogue score ($r = 0.71$, $p < 0.001$).

The NFTI-QOL was administered to 30 patients with SVS (non-NF2) and 30 healthy controls (►**Table 4**). All three groups were significantly different, with the NF2 patients scoring worse than SVS group ($p = 0.002$), who in turn were worse than the control group ($p < 0.001$).

Reliability and Item Loadings

The NFTI-QOL showed good internal reliability (Cronbach's $\alpha = 0.87$). Loadings of each item on the whole scale NFTI-QOL is shown in ►**Table 5**.

Discussion

In this study, we developed and validated a disease-specific questionnaire to measure QOL in NF2, as an assessment tool in clinical practice and in clinical trials. It is quick and simple to complete. The acronym NFTI-QOL refers to Neurofibromatosis T (2) I (impact) on QOL. The construction of the

Table 4 Comparison of NFTI-QOL Scores between NF2 Patients, Solitary Vestibular Schwannoma (SVS) Patients, and Healthy Controls

Item	Mean Score			ANOVA between Groups[b]
	NF2	SVS	Healthy Controls	
	$n = 50$	$n = 30$	$n = 30$	
Q1. Hearing	1.68	1.13	0.13	<0.001
Q2. Dizziness and balance	1.78	1.43	0.00	<0.001
Q3. Facial palsy	0.78	0.20	0.00	<0.001
Q4. Sight[a]	1.0	0.37	0.03	<0.001
Q5. Mobility and walking	0.86	0.40	0.00	<0.001
Q6. Role and outlook on life	1.5	0.83	0.07	<0.001
Q7. Pain	0.74	0.60	0.03	<0.001
Q8. Anxiety and depression	0.96	0.47	0.17	<0.001
Total score [95% CI]	9.41 [7.55–11.26]	5.50 [4.09–6.91]	0.43 [0.03–0.83]	<0.001

[a] $n = 37$ for Q4.
[b] Kruskal Wallis nonparametric analysis of variance (ANOVA).

questionnaire followed a structured pathway to ensure maximum validity. The response rate of 77% to our postal questionnaires was high and should limit bias: there was no significant difference in age, gender or disease-severity in our nonresponders. The NFTI-QOL was completed in ~3 minutes by our NF2 patients, despite their visual and neurological impairments. Difficulties may arise for patients who are both deaf and unable to read the questionnaire, but this could be overcome by using other communication tools, such as tactile signing.

The total NFTI-QOL score was higher in people with higher morbidity. Comparison with the control groups indicated that the healthy population scored significantly lower in all domains of the NFTI-QOL, confirming construct validity. Furthermore, patients with SVS scored highly on hearing, balance, and pain, but scored lower on the other domains than NF2 patients and had a significantly lower total score.

This is the first study to assess the QOL in NF2 using both the EuroQOL and SF-36 generic scores to validate our questionnaire. Our patients scored significantly worse than the population norms in all domains of each score. This confirms the negative impact on many aspects of QOL by NF2. The scores on the SF-36 were comparable to those found by Neary et al[8] reflecting that both units are National Specialized Centers for NF2 in England and include patients with similar disease severity.

Balance and dizziness problems had the largest negative impact on QOL in our patients. The symptoms were reported by 84% and it was disruptive to usual activities in 78% of the individuals. Hearing problems were found in 84% and were disruptive in 66% of the patients; visual problems were present in 60% and were disruptive in 41% of the individuals; facial palsy was present in 46% and disruptive in 26% of the patients. During the interview studies and focus group session, NF2 manifestations that were obvious to the general

Table 5 NFTI-QOL: Individual Item Loadings on the Whole Scale

Item	Pearson Correlation	Significance (Two-Tailed)
Q1. Hearing	0.817	<0.001
Q2. Dizziness and balance	0.668	<0.001
Q3. Facial palsy	0.772	<0.001
Q4. Sight	0.662	<0.001
Q5. Mobility and walking	0.720	<0.001
Q6. Role and outlook on life	0.853	<0.001
Q7. Pain	0.605	<0.001
Q8. Anxiety and depression	0.706	<0.001

public, such as balance disturbance and facial weakness, caused significant distress. Some patients had been ejected from public places as they were perceived erroneously to be under the influence of alcohol. We have developed an NF2 information card for use by our patients in public settings to try and increase public understanding and awareness.

One of our initial aims was to produce a short and easy to complete questionnaire, which could provide a quantitative marker of QOL. The NFTI-QOL is shorter in length than, for example, the SF-36[10] or the Manchester questionnaire[8] but it is similar in length to the other generic and specific questionnaires, such as EuroQOL[11] and SWID.[15] The method of developing the NFTI-QOL included the use of a much longer questionnaire which then underwent factor analysis to determine representative questions, and it was seen that there was a clustering around seven topics which were converted to the final NFTI-QOL questions, in addition to the question on sight. We have added a blank page at the end of the questionnaire for in-depth comment that may be used by the investigators. A further 30 NF2 patients have since used the additional page and their comments amplify their questionnaire answers rather than adding new information.

Conclusion

The NFTI-QOL is a robustly constructed disease-specific QOL questionnaire for NF2. It correlates strongly and significantly with EuroQOL and all SF-36 domains ($p < 0.01$). It is straightforward and quick ($\leq$3 minutes) for patients to complete and easy to score. It is suitable as a quantitative method of assessing QOL in NF2 both in a clinical setting and as an outcome measure for treatment. The NFTI-QOL has been validated for adults (>16 years) in the UK, and could be adapted for use in other countries.

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Appendix 1: NFTI-QOL

Neurofibromatosis 2 Impact on QOL (English Version for the UK)

INSTRUCTIONS FOR COMPLETING THE NFTI-QOL

Please complete the following information:

Age: _____ years

Gender: Male ☐ 1 Female ☐ 2 (please tick)

For each of the questions on the next page, please tick the one box that describes how you feel today

Usual activities include: work; housework; study; sport; social; family or leisure activities

Patient ID/Label:

Q1. Do balance or dizziness problems stop you performing your usual activities?

No balance problems or dizziness	☐0
Balance or dizziness problems but no difficulties	☐1
Balance or dizziness problems cause me some difficulties	☐2
Balance or dizziness problems stop my usual activities	☐3

Q2. Do hearing problems stop you performing your usual activities?

No hearing problems	☐0
Hearing problems but no difficulty	☐1
Hearing problems cause me some difficulty	☐2
Hearing problems stop my usual activities	☐3

Q3. Does facial weakness stop you performing your usual activities?

No facial weakness	☐0
Facial weakness, but no difficulty	☐1
Facial weakness causes some difficulty	☐2
Facial weakness stops my usual activities	☐3

Q4. Do problems with your sight stop you performing your usual activities?

No problems with sight	☐0
Sight problems, but no difficulty	☐1
Sight problems cause me some difficulty	☐2
Sight problems stop my usual activities	☐3

Q5. Do you have any problems in mobility and walking?

No problems in mobility and walking	☐0
Some difficulty but can manage on my own	☐1
Unable to walk around without some help	☐2
Unable to walk at all	☐3

Q6. Has your medical condition affected your role and outlook on life? (e.g., confidence, vulnerability, relationships, caring for family, career, having children)

No effect or positive effect	☐0
Small negative effect	☐1
Moderately negative effect	☐2
Large negative effect	☐3

Q7. Pain; throughout our lives, most of us have had pain from time to time such as mild headaches, sprains and toothaches. Have you had pain *other than this* in the last week?

None	☐0
Mild pain	☐1
Moderate pain	☐2
Severe pain	☐3

Q8. Do you currently suffer from anxiety or depression?

No	☐0
Mild anxiety or depression	☐1
Moderate anxiety or depression	☐2
Extreme anxiety or depression	☐3

If you have any further comments regarding the impact of NF2 on your QOL, please write them here:

You have now completed the NFTI-QOL. Thank you for your input.