

Functional Motor Outcome Measures in adults with NF1

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Response Evaluation In Neurofibromatosis Schwannomatosis
INTERNATIONAL COLLABORATION



Functional Problems in NF1

- Problems with walking, balance, hand function common
- Multiple causes
 - Tumours - brain, central and peripheral nervous system
 - Neurovascular disease
 - Neuropathy
 - Skeletal abnormalities

Functional Outcome Measures in Adults with NF1: Aims

- To determine intra-rater & inter-rater reliability of motor outcome measures in adults with NF1 Mullin et al. Reliability of motor outcome measures in adults with NF1 Sage Open Medicine 2018; 6:1-7
- To determine relationship between motor outcome measures and INF1-QoI) Ferner RE et al. Health Qual Life Outcomes 2017; 15(1): 34.



Functional Motor Outcomes

- Focus group identified functional problems
- Recruitment from national NF1 service at Guy's Hospital

Inclusion criteria

- NF1 diagnosis confirmed
- >16 years
- Informed consent
- Mobility/impaired hand function/balance related to NF1
- Able to walk 10 m (walking aids allowed)



10 Metre Walk

The 10 Metre Walk test assesses functional mobility & gait speed.

Walk at normal speed before measured 10m walk marking. Continue walking at normal speed along the measured walk.

Measure between marked area.

Record seconds/milliseconds



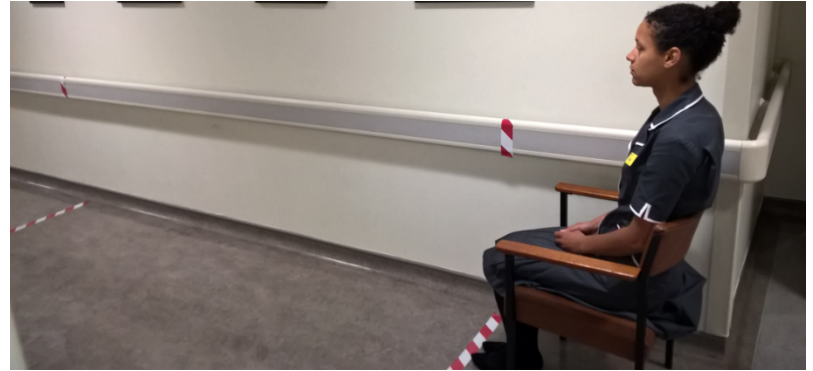
Timed Up and Go test

Measures functional mobility

Stand up, walk 3 metres
& returns to chair.

Measure from the time
leaves chair to to time
returns to chair

Record seconds/ milliseconds



Functional Reach

Assesses standing balance

Stand with shoulder at 90° to ruler.

Measure difference in distance

with arm outstretched standing still

and leaning forward

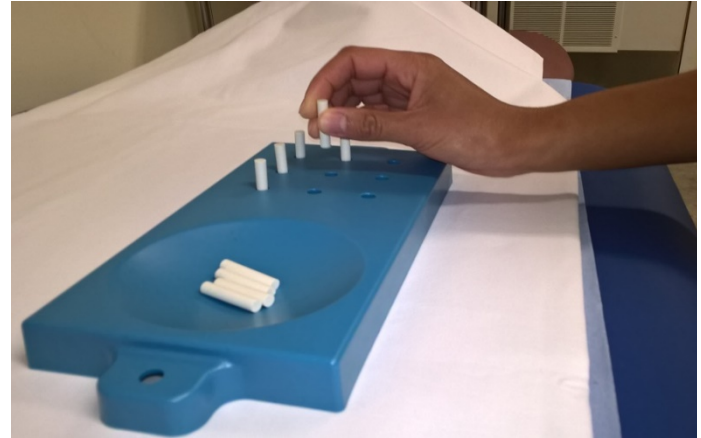
Record measurements recorded in centimetres and millimetres



Nine Hole Peg test - modified

The modified Nine Hole Peg test assesses upper limb dexterity

Take pegs one by one from bowl
Place pegs into peg board holes



Measurements recorded in seconds and milliseconds

Functional motor outcomes – methods for rating

- Physiotherapist performs outcome measures
- Photographs and videos participants
- 3 trials of each task. In upper limb tasks each limb rated separately
- Four raters rate tasks on photographs and videos independently and anonymously.
(2 clinicians, 1 physiotherapy, 1 nurse)
- 1 rater performs ratings on 2 occasions



Functional Motor Outcome Measures: Results

- 49 NF1 adults
- 29 females, 20 males
- Age range 16-66 years, median 31 years
- Spread of disease severity



Functional Motor Outcome Measures Results

- Inter-rater & intra-rater agreement high for 10 metre walk, Timed up and go, Functional reach (all $r \geq 0.9$)
- 9 hole peg test was $r \geq 0.75$ and 0.76



Functional motor outcomes in 49 NF1 adults

Functional test	Mean score	Range min-max
9 Hole peg test (sec)	18.36	11.47- 53.62
Functional Reach (cm)	31.96	4.00 - 38.43
Timed up and go (sec)	11.57	5.50 - 49.82
10m walk time (sec)	6.74	3.82 - 25.94



Functional Motor Outcome Measures - Results

- NF1-QOL mean score 9, range 1-26
(representative of adult NF1 population
mean 8.64, range 0-30))
- Motor outcomes correlated highly with mobility and gait,
role and outlook on life on INF1-QOL ($p < 0.01$)

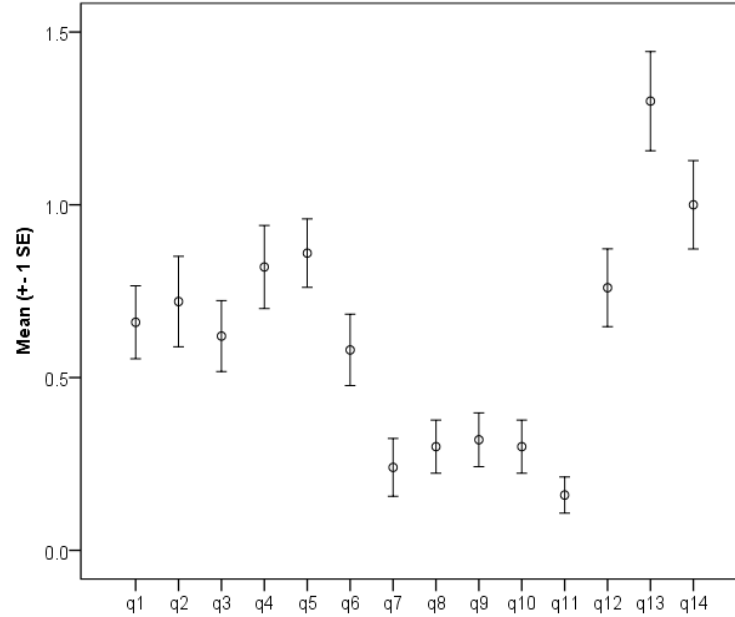
INF1-QOL 14 questions

Total scale score 0-42

- Q1 Vision
- Q2 Cosmetic appearance
- Q3, Q4 Pain
- Q5 Learning problems
- Q6 Behaviour and personality
- Q7 **Mobility and walking**
- Q8 Weakness, numbness clumsiness in hands
- Q9 Speech
- Q10 Bones
- Q11 Breathing
- Q12 Sleeping
- Q13 **Role and outlook on life**
- Q14 Depression and anxiety



INF1-QOL Item Scores



Q1 Vision

Q2 Cosmetic appearance

Q3 Pain

Q4 Pain scale 0-10

Q5 Learning problems

Q6 Behaviour and personality

Q7 Mobility and walking

Q 8 Weakness, numbness, clumsiness in hands

Q 9 Speech

Q10 Bones

Q11 Breathing

Q12 Sleeping

Q13 Role and outlook on life

Q14 Depression and anxiety



Conclusions

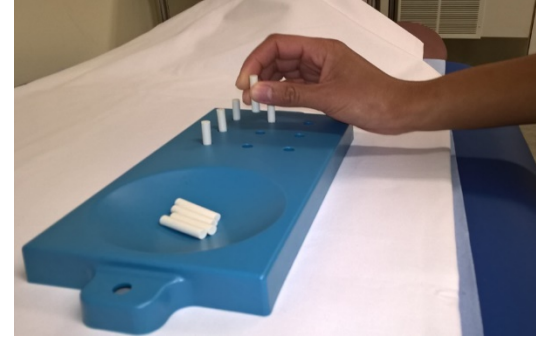
- 10 Metre Walk, Timed Up and Go & Functional Reach have excellent intra-rater and inter-rater reliability
- They are highly correlated with INF1-Qol patient focused outcome measure
- They are quick and easy to perform with minimal equipment
- Modified nine hole peg test is influenced by cognitive factors in NF1 and is not useful

Conclusions

- Future studies
- Use of measures across different institutions
- Test-retest reliability
- Grip dynamometry



Motor Outcome Measures



10 metre walk
Timed up and go
9 hole peg test
Functional reach