THE NEWCASTLE

MITOCHONDRIAL DISEASE ADULT SCALE

(NMDAS)

Name: ___________________

Date of birth: ______________

Age at assessment: ______________

Date of assessment: ______________

Checklist – please tick off when completed.

☐ Height: __________

☐ FVC - 1st attempt __________

☐ FVC - 2nd attempt __________

☐ FVC - 3rd attempt __________ % Predicted __________

☐ SF-12v2 self completion questionnaire ____________ ____________ ____________

☐ WTAR reading test (1 minute) ____________ ____________ ____________

☐ Symbol Search (2 minutes) ____________ ____________ ____________

☐ Speed of comprehension test (2 minutes) ____________ ____________ ____________

Disease score (sections I-III)________

SF-12v2 Quality of Life score (section IV)________
Section I- Current Function
Rate function over the preceding 4 week period, according to patient and/or caregiver interview only. The clinician’s subjective judgement of functional ability should not be taken into account.

1. **Vision** with usual glasses or contact lenses
   0. Normal.
   1. No functional impairment but aware of worsened acuities.
   2. Mild - difficulty with small print or text on television.
   3. Moderate - difficulty outside the home (eg bus numbers, road signs or shopping).
   4. Severe - difficulty recognising faces.
   5. Unable to navigate without help (eg carer, dog, cane).

2. **Hearing** with or without hearing aid
   0. Normal.
   1. No communication problems but aware of tinnitus or deterioration from prior ‘normal’ hearing.
   2. Mild deafness (eg missing words in presence of background noise). **Fully** corrected with hearing aid.
   4. Severe deafness - poor hearing even with aid (see 3 above).
   5. End stage - virtually no hearing despite aid. Relies heavily on non-verbal communication (eg lip reading) or has cochlear implant.

3. **Speech**
   0. Normal.
   1. Communication unaffected but patient or others aware of changes in speech patterns or quality.
   2. Mild difficulties - usually understood and rarely asked to repeat things.
   3. Moderate difficulties - poorly understood by strangers and frequently asked to repeat things.
   4. Severe difficulties - poorly understood by family or friends.
   5. Not understood by family or friends. Requires communication aid.

4. **Swallowing**
   0. Normal.
   1. Mild - sensation of solids ‘sticking’ (occasional).
   2. Sensation of solids ‘sticking’ (most meals) or need to modify diet (eg avoidance of steak/salad).
   3. Difficulty swallowing solids - affecting meal size or duration. Coughing, choking or nasal regurgitation infrequent (1 to 4 times per month) but more than peers.
   4. Requires adapted diet - regular coughing, choking, or nasal regurgitation (more than once per week).
   5. Requiring enteral feeding (eg PEG).

5. **Handwriting**
   0. Normal.
   1. Writing speed unaffected but aware of increasing untidiness.
   2. Mild – Has to write slower to maintain tidiness/legibility.
   3. Moderate – Handwriting takes at least twice as long or resorts to printing (must previously have used joined writing).
   4. Severe – Handwriting mostly illegible. Printing very slow and untidy (eg ‘THE BLACK CAT’ takes in excess of 30 seconds).
   5. Unable to write. No legible words.
6. Cutting food and handling utensils (irrespective of contributory factors – eg weakness, coordination, cognitive function etc. This is also true for questions 7-10)

0. Normal.
1. Slightly slow and/or clumsy but **minimal** effect on meal duration.
2. Slow and/or clumsy with extended meal duration, but no help required.
3. Difficulty cutting up food and inaccuracy of transfer pronounced. Can manage alone but avoids problem foods (eg peas) or carer typically offers minor assistance (eg cutting up steak).
4. Unable to cut up food. Can pass food to mouth with great effort or inaccuracy. Resultant intake minimal. Requires major assistance.
5. Needs to be fed.

7. Dressing

0. Normal.
1. Occasional difficulties (eg shoe laces, buttons etc) but no real impact on time or effort taken to dress.
2. Mild – Dressing takes longer and requires more effort than expected at the patient’s age. No help required.
3. Moderate - Can dress unaided but takes at least twice as long and is a major effort. Carer typically helps with difficult tasks such as shoe laces or buttons.
4. Severe – Unable to dress without help but some tasks completed unaided.
5. Needs to be dressed.

8. Hygiene

0. Normal.
1. Occasional difficulties only but no real impact on time or effort required.
3. Moderate - bathes and showers alone with difficulty or needs bath chair / modifications. Dextrous tasks (eg brushing teeth, combing hair) performed poorly.
5. Dependent upon carers to wash, bathe, and toilet.

9. Exercise Tolerance

0. Normal.
1. Unlimited on flat - symptomatic on inclines or stairs.
2. Able to walk < 1000m on the flat. Restricted on inclines or stairs - rest needed after 1 flight (12 steps).
3. Able to walk < 500m on the flat. Rest needed after 8 steps on stairs.
4. Able to walk < 100m on the flat. Rest needed after 4 steps on stairs.
5. Able to walk < 25m on the flat. Unable to do stairs alone.

10. Gait stability

0. Normal.
1. Normal gait - occasional difficulties on turns, uneven ground, or if required to balance on narrow base.
3. Unsteady gait. **Always** off balance when walking. **Occasional** falls. Gait steady with support of stick or person.
4. Gait grossly unsteady without support. **High likelihood** of falls. Can only walk short distances (< 10m) without support.
5. Unable to walk without support. Falls on standing.
Section II – System Specific Involvement
Rate function according to patient and/or caregiver interview and consultation with the medical notes. Each inquiry should take into account the situation for the preceding **12 month period** only, unless otherwise stated in the question.

1. **Psychiatric**

   0. None.
   1. Mild & transient (eg reactive depression) - lasting **less** than 3 months.
   2. Mild & persistent (lasting **more** than 3 months) or recurrent. Patient has consulted GP.
   3. Moderate & warranting specialist treatment (e.g. from a psychiatrist) - eg. bipolar disorder or depression with vegetative symptoms (insomnia, anorexia, abulia etc).
   4. Severe (eg self harm - psychosis etc).
   5. Institutionalised or suicide attempt.

2. **Migraine Headaches** During the last **3 months**, how many days have headaches prevented the patient from functioning normally at school, work, or in the home?

   0. No past history.
   1. Asymptomatic but past history of migraines.
   2. One day per month.
   3. Two days per month.
   4. Three days per month.
   5. Four days per month or more.

3. **Seizures**

   0. No past history.
   1. Asymptomatic but past history of epilepsy.
   2. Myoclonic or simple partial seizures only.
   3. Multiple absence, complex partial, or myoclonic seizures affecting function or single generalised seizure.
   4. Multiple generalised seizures.
   5. Status epilepticus.

4. **Stroke-like episodes** (exclude focal deficits felt to be of vascular aetiology)

   0. None.
   1. Transient focal sensory symptoms only (**less** than 24 hours).
   2. Transient focal motor symptoms only (**less** than 24 hours).
   3. Single stroke-like episode affecting one hemisphere (**more** than 24 hours).
   4. Single stroke-like episode affecting both hemispheres (**more** than 24 hours).
   5. Multiple stroke-like episodes (**more** than 24 hours each).

5. **Encephalopathic Episodes**

   0. No past history.
   1. Asymptomatic but past history of encephalopathy.
   2. Mild - single episode of personality or behavioural change but retaining orientation in time/place/person.
   3. Moderate - single episode of confusion or disorientation in time, place or person.
   4. Severe – multiple moderate episodes (as above) or emergency hospital admission due to encephalopathy **without** associated seizures or stroke-like episodes.
   5. Very severe - in association with seizures, strokes or gross lactic acidaemia.
6. Gastro-intestinal symptoms

0. None.
1. Mild constipation only or past history of bowel resection for dysmotility.
2. Occasional symptoms of ‘irritable bowel’ (pain, bloating or diarrhoea) with long spells of normality.
3. Frequent symptoms (as above) most weeks or severe constipation with bowels open less than once/week or need for daily medications.
4. Dysmotility requiring admission or persistent and/or recurrent anorexia/vomiting/weight loss.
5. Surgical procedures or resections for gastrointestinal dysmotility.

7. Diabetes mellitus

0. None.
1. Past history of gestational diabetes or transient glucose intolerance related to intercurrent illness.
2. Impaired glucose tolerance (in absence of intercurrent illness).
3. NIDDM (diet).
4. NIDDM (tablets).
5. DM requiring insulin (irrespective of treatment at onset).

8. Respiratory muscle weakness

0. FVC normal (≥ 85% predicted).
1. FVC < 85% predicted.
2. FVC < 75% predicted.
3. FVC < 65% predicted.
4. FVC < 55% predicted.
5. FVC < 45% predicted or ventilatory support for over 6 hours per 24 hr period (not for OSA alone).

9. Cardiovascular system

0. None.
1. Asymptomatic ECG change.
2. Asymptomatic LVH on echo or non-sustained brady/tachyarrhythmia on ECG.
3. Sustained or symptomatic arrhythmia, LVH or cardiomyopathy. Dilated chambers or reduced function on echo. Mobitz II AV block or greater.
4. Requires pacemaker, defibrillator, arrhythmia ablation, or LVEF < 35% on echocardiogram.
5. Symptoms of left ventricular failure with clinical and/or x-ray evidence of pulmonary oedema or LVEF < 30% on echocardiogram.
Section III – Current Clinical Assessment
Rate current status according to examination performed at the time of assessment

1. Visual acuity with usual glasses, contact lenses or pinhole.
   0. CSD ≤ 12 (ie normal vision - 6/6, 6/6 or better).
   1. CSD ≤ 18 (eg 6/9, 6/9).
   2. CSD ≤ 36 (eg 6/12, 6/24).
   3. CSD ≤ 60 (eg 6/24, 6/36).
   4. CSD ≤ 96 (eg 6/60, 6/36).
   5. CSD ≥ 120 (eg 6/60, 6/60 or worse).

2. Ptosis
   0. None.
   1. Mild ptosis - not obscuring either pupil.
   2. Unilateral ptosis obscuring < 1/3 of pupil.
   3. Bilateral ptosis obscuring < 1/3 or unilateral ptosis obscuring > 1/3 of pupil or prior unilateral surgery.
   4. Bilateral ptosis obscuring > 1/3 of pupils or prior bilateral surgery.
   5. Bilateral ptosis obscuring > 2/3 of pupils or > 1/3 of pupils despite prior bilateral surgery.

3. Chronic Progressive External Ophthalmoplegia
   0. None.
   2. Abduction of worst eye incomplete.
   3. Abduction of worst eye below 60% of normal.
   4. Abduction of worst eye below 30% of normal.
   5. Abduction of worst eye minimal (flicker).

4. Dysphonia/Dysarthria
   0. None.
   1. Minimal - noted on examination only.
   2. Mild – clear impairment but easily understood.
   3. Moderate – some words poorly understood and infrequent repetition needed.
   4. Severe – many words poorly understood and frequent repetition needed.

5. Myopathy
   0. Normal.
   1. Minimal reduction in hip flexion and/or shoulder abduction only (eg MRC 4+/5).
   2. Mild but clear proximal weakness in hip flexion and shoulder abduction (MRC 4/5). Minimal weakness in elbow flexion and knee extension (MRC 4+/5 - both examined with joint at 90 degrees).
   3. Moderate proximal weakness including elbow flexion & knee extension (MRC 4/5 or 4 -/5) or difficulty rising from a 90 degree squat.
   4. Waddling gait. Unable to rise from a 90 degree squat (=a chair) unaided.
   5. Wheelchair dependent primarily due to proximal weakness.
6. Cerebellar ataxia
   0. None.
   1. Normal gait but hesitant heel-toe.
   2. Gait reasonably steady. Unable to maintain heel-toe walking or mild UL dysmetria.
   3. Ataxic gait (but walks unaided) or UL intention tremor & past-pointing. Unable to walk heel-toe - falls immediately.
   4. Severe - gait grossly unsteady without support or UL ataxia sufficient to affect feeding.
   5. Wheelchair dependent primarily due to ataxia or UL ataxia prevents feeding.

7. Neuropathy
   0. None.
   1. Subtle sensory symptoms or areflexia.
   2. Sensory impairment only (eg glove & stocking sensory loss).
   3. Motor impairment (distal weakness) or sensory ataxia.
   4. Sensory ataxia or motor effects severely limit ambulation.
   5. Wheelchair bound primarily due to sensory ataxia or neurogenic weakness.

8. Pyramidal Involvement
   0. None.
   1. Focal or generalised increase in tone or reflexes only.
   2. Mild focal weakness, sensory loss or fine motor impairment (eg cortical hand).
   3. Moderate hemiplegia allowing unaided ambulation or dense UL monoplegia.
   4. Severe hemiplegia allowing ambulation with aids or moderate tetraplegia (ambulant).
   5. Wheelchair dependant primarily due to hemiplegia or tetraplegia.

9. Extrapyramidal
   0. Normal.
   1. Mild and unilateral. Not disabling (H&Y stage 1).
   3. Moderate. Significant slowing of body movements (H&Y stage 3)
   5. Cannot walk or stand unaided. Requires constant nursing care (H&Y stage 5).

10. Cognition
    Patients undergo testing using WTAR, Symbol Search and Speed of Comprehension Test.
    0. Combined centiles 100 or more.
    1. Combined centiles 60 - 99
    2. Combined centiles 30 - 59
    3. Combined centiles 15 - 29
    4. Combined centiles 5 - 14
    5. Combined centiles 4 or below.