MJD FOUNDATION

MEDICAL PROTOCOLS

Therapy

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Please provide any feedback on this Protocol to <u>desiree.lagrappe@mjd.org.au</u>

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Action	Person	Date
DRAFT v 1	MJDF • Libby Massey • Michaela Jackson • Nick Kenny • Rebecca Amery • Desireé LaGrappe • Nadia Lindop	16/10/2016
DRAFT v 2	Meir Medical Center/Tel Aviv University • Carlos Gordon NTG • Jaimee Dutton	08/12/2016

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1. Background — Therapeutic Approaches for Treating Machado Joseph Disease

Machado Joseph Disease (MJD) is the most common of the 36 autosomal dominant neuro-degenerative spinocerebellar ataxias (SCA's) (Fan et al., 2013; D'Abreau et al. 2010). The disease is caused by a cytosine-adenine-guanine (CAG) repeat expansion on the ATXN3 gene which results in a build-up of an abnormal amount of the protein ataxin-3 (ATXN3) in the brain. The build-up of ATXN3 results in a constellation of symptoms, the most obvious of which is gait ataxia. However, there are also disturbances to vision, swallowing, speech and continence. There is a strong, although not definitive, correlation between repeat amplification and age of onset of the disease and the severity of the symptoms experienced by affected individuals (Pedroso et al., 2013).

The progressive disease leads to the accumulation of motor loss and incoordination of movements (ataxia), which produces an inexorable scenario of immobility and wheelchair dependency by around the fifteenth (15^{th}) year of disease evolution (Martins et al., 2013).

In common with other SCA's there are very limited pharmacological therapies available to treat MJD (Fan, 2014; Fonteyn, 2013). Therefore, the mainstays of treatment for degenerative cerebellar ataxias are currently physiotherapy, occupational therapy, and speech therapy (Ilg et al., 2013). These therapies are critical to the maintenance of quality of life and directed to minimising the loss of functional independence, prevention of depression, secondary complications (Silva et al., 2010) and falls, which have devastating physical and emotional corollaries (Fonteyn, 2010 and 2013).

Despite this reliance on allied health treatments, there have been very few studies and a paucity of evidence relating to the efficacy of allied health therapeutic interventions and rehabilitation for SCA's in general and SCA3/MJD in particular (Ilg et al., 2010 and 2013; Miyai et al., 2014, Martin et al., 2108, Silva, 2010; Fonteyn, 2013).

1.1. New Approaches – Compensation to Rehabilitation

Knowledge of the role that the cerebellum plays in motor learning, in combination with the progressive nature of MJD, has led to a traditional view that treatments aimed at the remediation of impairments caused by cerebellar dysfunction would be ineffective, because motor learning mechanisms are damaged (Martin et al., 2008, 2013; Miyai, 2014). Accordingly, the treatment strategies previously advocated in the academic literature for degenerative ataxias were based on compensatory approaches. These included functional occupational therapy training and equipment provision and physiotherapy approaches that include the use of limb weights, gait aids and Frenkel Coordination exercises (Martin et al., 2008).

Recently, however, this approach is being reviewed. Evidence is emerging that intensive coordination training on posture and gait in degenerative cerebellar disease when combined with functional retraining may be beneficial, be capable of producing meaningful rehabilitative benefits in everyday life, and have the potential to persist

with appropriate maintenance therapy (Ilg, 2009, 2010, 2013; Miyai, 2013, Fonteyn 2013).

Physical therapy containing balance exercises, gait training, and muscle strengthening, as well as occupational therapy focused on balance related to activities of daily living, are thought to have the most beneficial impact (Martineaux, 2014).

1.2. Falls Prevention

A critical issue in the care of people with degenerative conditions is the prevention of falls. Although there is little definitive data to indicate falls rates for people who have MJD, falls are known to be common in spinocerebellar ataxia (Schniepp, 2014). A pilot study reported in *Cerebellum* by Fonteyn (2010) noted that up to 93% of patients with degenerative Spinocerebellar Ataxia (SCA) reported one or more falls in the preceding 12 months - and many suffered injuries due to these falls. Anecdotally, the MJD Foundation (MJDF) is aware that falls pose a significant issue for clients.

Falls are an important reason for hospitalisations and the loss of independent function (Fonteyn, 2010). Consequences can be devastating physically and emotionally, both for those who fall and their care givers and represent a substantial cost to the public health system. In a 2015 study, the average direct medical cost of a fall was estimated to be $11,502 USD [~$15 100 AUD] (Carande-Kulis et al., 2015).

Internationally, fall prevention in the elderly and in better known neurological conditions such as Parkinson's Disease is given substantial attention and community based falls prevention strategies are increasingly afforded high priority as a preventative public health initiative (Carande-Kulis et al., 2015). A large scale review of falls history from the EuroSCA natural history study[1] indicated that the factors associated with a higher fall frequency for the general SCA population included disease duration, severity of ataxia, the presence of pyramidal symptoms, the total number of non-ataxia symptoms, and the genotype SCA3. (Fonteyn, 2010). The study postulates that the presence of pyramidal and non-ataxia features, specifically neuropathy and opthalmoparesis, for people who have MJD provide both a rationale for the higher fall rate and indications of how to target treatment regimens.

There are therefore clear indications that a focus on falls prevention should comprise an important priority for physiotherapists and occupational therapists devising therapeutic interventions for people who have MJD.

[1] A large scale European clinically based 3-year follow up study of SCA.

2. Measurement Scales

Assessment of progressive degenerative disorders is a difficult but important component in guiding treatment. A number of standardised assessment tools have been developed and are referred to in the literature (Saute et al., 2010).

Please refer to ***Appendix C*** for a template of each measurement scale mentioned.

2.1. Scale for Assessment and Rating of Ataxia (SARA)

A common measure of functional impairment in cerebellar disease such as MJD is the Scale for Assessment and Rating of Ataxia (SARA). This tool for assessing the severity and treatment effectiveness of cerebellar ataxia has eight items, yielding a total score of 0 (no ataxia) to 40 (most severe ataxia) [21]: gait (scores 0 to 8), stance (0 to 6), sitting (0 to 4), speech disturbance (0 to 6), finger chase (0 to 4), nose–finger test (0 to 4), fast alternating hand movements (0 to 4), and heel– shin slide (0 to 4). Limb kinetic functions (items 5 to 8) are rated independently for both body sides, and its arithmetic mean is included in the total score (Saute et al., 2010). Studies of the natural history of cerebellar disease indicate that progression occurs at a rate of between 0.35 -2.5 SARA points per year (Ilg, 2014; per Klockgether).

2.2. International Cooperative Ataxia Rating Scale (ICARS)

The ICARS is a semi quantitative 100- point scale consisting of 19 items, divided into four unequally weighted sub-scores: posture and gait disturbance (seven items, 34 points); limb kinetic function (seven items, 52 points); speech disorder (two items, eight points); and oculomotor disorder (three items, six points). The higher the score, the worse is the patient's performance (Saute et al., 2010).

2.3. Modified International Cooperative Ataxia Rating Scale (MICARS)

The 120-point Modified International Cooperative Ataxia Rating Scale (MICARS) was developed by adding seven additional tests to the ICARS: in kinetic function (decomposition of leg movement, decomposition of leg tapping, rebound of the arms, and overshoot of the arms, each 0 to 2 points and scored for both left and right); speech disorders (dysarthria alternating syllable, 0 to 2 points); and oculomotor function (abnormal eye movements at rest, 0 or 1 point). The higher the score, the worse the ataxic syndrome (Saute et al., 2010).

2.4. Neurological Examination Score for Spinocerebellar Ataxia 3 (NESSCA)

A global, comprehensive inventory for the assessment of SCA3/MJD. This score is based on a quantitative assessment of the standardised neurological examination, and its focus is on the main characteristics of SCA in general and of MJD in particular. Items of the NESSCA were selected according to the usual findings seen in the majority of patients with MJD.

2.5. Barthel scale

The Barthel scale or Barthel ADL index is an ordinal scale used to measure performance in activities of daily living (ADL). Each performance item is rated on this scale with a given number of points assigned to each level or ranking. It uses ten variables describing ADL and mobility. A higher number is associated with a greater likelihood of being able to live at home with a degree of independence following discharge from hospital. The amount of time and physical assistance required to perform each item are used in determining the assigned value of each item. External factors within the environment affect the score of each item. If adaptations outside the standard home environment are met during assessment, the participant's score will be lower if these conditions are not available. If adaptations to the environment are made, they should be described in detail and attached to the Barthel index.

2.6. The Functional Independence Measure

The Functional Independence Measure (FIM™) instrument is a basic indicator of patient disability. FIM™ is used to track the changes in the functional ability of a patient during an episode of hospital rehabilitation care.

2.7. The Hamilton Rating Scale for Depression

The Hamilton Rating Scale for Depression (HRSD), also called the Hamilton Depression Rating Scale (HDRS), abbreviated HAM-D, is a multiple item questionnaire used to provide an indication of depression, and as a guide to evaluate recovery. Max Hamilton originally published the scale in 1960 and revised it in 1966, 1967, 1969, and 1980. The questionnaire is designed for adults and is used to rate the severity of their depression by probing mood, feelings of guilt, suicide ideation, insomnia, agitation or retardation, anxiety, weight loss, and somatic symptoms.

3. Physiotherapy

Neurological physiotherapy promotes movement and quality of life in patients who have had severe brain or spinal cord damage from trauma, or who suffer from neurological diseases such as MJD, stroke, Parkinson's Disease and Multiple Sclerosis.

Degenerative cerebellar diseases such as MJD are especially hard to treat due to their progressive nature and effects on virtually all parts of the cerebellum. Treatment of progressive degenerative diseases are considered to be successful if the patient remains stable at the current status of motor function (by validated scale) as long as possible, or if progression of functional impairment is slowed down (Synofzik, 2014).

Although scarce, reviews of the effectiveness of physiotherapy for cerebellar dysfunction indicate that although there is modest evidence physiotherapy is beneficial and has a positive effect on gait, trunk control, and activity limitations.

Limitations to successful treatment are a general lack of experience with ataxia by therapists in the field and limited practical guidelines available to assist with the development of treatment programs (Fonteyn, 2013; Martineau, 2014).

The heterogeneity of cerebellar conditions leads to a focus on individualised treatment approaches tailored to meet the specific needs of each patient. This makes objective assessment of progress difficult however, and can lead to decreased patient and therapist satisfaction (Ilg, 2013; Martin, 2009). Compounding this, the benefits conferred by physiotherapy can be difficult to determine against the background of the natural disease progression of cerebellar ataxias.

Despite these limitations -and the lack of large scale peer reviewed literature to support any of the allied health interventions - there is emerging, evidence based literature from larger case-control studies to support physiotherapeutic interventions in the general SCA population - particularly those designed to enhance motor performance and learning, and these have the potential to extrapolate to the treatment of patients with MJD (Ilg, 2014, Synofzik, 2014).

3.1. Intensive Treatment and Follow-Up

Preliminary results of these studies indicate that motor performance gets better with the help of continuous and intensive motor training and that because disease is ongoing in cerebellar degeneration there is the need for life-long treatment and maintenance. For retention, 1 hour of 'homework' training per day showed at least a partial maintenance, whereas improvements declined to the baseline level after 6 months if no specific follow up training was provided (Ilg, 2014; Miyai, 2014).

Long-term (3 year) follow up of several case studies where physiotherapy has been accessed regularly has also shown gains in SARA after repeated 'boost' (intensive) physiotherapy that are comparable to the gains in the initial intensive phase; however, low intensity home therapy of up to 80 minutes per week failed to maintain gains in ataxia and ADL's (Miyai, 2014). This indicates the need for both intensive

initial treatment, periodic boost intensive programs and persistent home program performance.

There remain many unknowns. To date these studies have focussed on gait ataxia and stance in ambulant patients and there is a further need to evaluate whether training of upper extremities improves function - and to what extent patients are able to improve who are more severely affected and wheelchair dependent.

Specific examples of relevant study design and results that are potentially applicable to the treatment of people with MJD follow.

3.2. Coordinative Physiotherapy

In an individual case-control study conducted by Ilg and colleagues (2010), subjects with cerebellar ataxia were trained by an ataxia expert physiotherapist for 1 hour per day, 3 days per week, for 4 weeks in an intensive whole-body coordination program specifically targeting static and dynamic balance and mobility function, followed by a 12 month at home training program.

The intensive component of the program aimed to activate and demand control mechanisms for balance control and multi joint coordination and trained the patient's ability to select and use visual somatosensory and vestibular inputs to preserve and retrain patient's capability for reacting to unforeseen situations and for avoiding falls as much as possible. Exercises included static balance, dynamic balance, complex whole body movements and falling prevention strategies.

The initial intensive 4 week program produced a reduction of ataxia severity and fall frequency, and an improvement of gait speed and activities of daily living correlating with a 5.2 point improvement on the SARA scale. This equates to a regain of functional performance of at least 2 years of natural disease progression (Ilg et al., 2013). After 12 months of an individualised homework program combining different coordination exercises and degrees of difficulty the gain was 3.1 SARA points. This retention of the training effects is the equivalent of gaining back one or more years of natural disease progression.

3.3. Physiotherapy in Conjunction with Occupational Therapy

In a case control trial combining occupational therapy and physiotherapy, patients participated in a 4 week intensive (12 hours per week) program combining physiotherapy with occupational therapy. The program comprised the static balance, dynamic balance, complex whole body movement and fall prevention strategies (as outlined above), in combination with training in specific activities of daily living as measured by a Functional Independence Measure (FIM) (Miyai, 2014).

The study design comprised 2 groups with one group receiving treatment after a 4 week delay (the second group therefore received a lower dose). Improvements were reported in ataxia severity, gait speed, fall frequency and activities of daily living performance for all participants. The improvements were more significant and sustained for at least 24 weeks for the group who received the longer and more intense program. This indicates the benefit of short term intensive rehabilitation and the capacity for these gains to be maintained.

3.4. Biofeedback Therapy in Patients with Ataxia

Biofeedback may assist patients to be able to reduce the amount of inappropriate co-activation of muscles groups. Relaxation and EMG biofeedback decreased severity of ataxic tremor in two case reports with patients with traumatic brain injury, which may have applicability to MJD patients (Fonteyn, 2013).

3.5. Supervised Sports

One study noted that patients showed improvement of velocity and speed symmetry in pointing movements of limbs, balance and manual dexterity after climbing training. The four participants had upper and lower limb ataxia of different acquired causes (Fonteyn, 2013). Participation in carefully chosen sporting activities have the potential for both physical and social-emotional benefits for patients with MJD.

3.6. Physiotherapy in the Remote Australian Context

The MJD Foundation conducts a program of regular strengthening and conditioning exercises conducted 'on country' and using gym equipment, as well as individual home programs under the umbrella of its 'staying stronger for longer' therapy program.

Physiotherapist Nick Kenny who has been working with the MJDF clients for several years provides his perspective:

"[T]o give you my opinion on fatigue management and physiotherapy rehab for our clients...

I have not come across any definitive literature either way and my thoughts are based on what I have observed working with these guys on a week by week basis. Sometimes pushing them to their limits and on other occasions being more conservative.

Firstly, with our low to mid-level clients [in the mild and moderate stages of MJD]……

Depending on personal / community factors our MJD clients can present for rehab with a different level of motivation to exercise each week. I try to gauge this first up by just having a general chat about how things are going. How are they feeling this week? Have you been doing exercises? If so what ones, show me? If not, why not? Have you had a fall or a near miss in the past week? Then the most important question... are you ready to exercise really well today?

If they seem a bit down or not overly motivated I am a strong believer in not flogging a dead horse. However, this is a rare occurrence. I would say 1/5 times when they attend for rehab they are demotivated. I have found that on these days they will not concentrate or commit to the level they need to in order to have a rewarding session. In these cases, I will focus rehab on the less physically strenuous exercises like fine motor skills, vision exercises or basic gait training. But I will ensure they know that the next week they are getting straight back in to the full session.

If, however, they seem happy and motivated to go (as they usually are), I really try to get as much out of them as possible. Plenty of lower and upper limb strength work, balance skills, and functional rehab like safe sit to stand, mobilising on uneven surfaces, stairs and safe transfers. In a group training session when there is the element of competition and teamwork they seem to think less about their fatigue and get absorbed in the challenges they are set.

I find that I can usually get a solid 30 minutes out of a session so long as in between exercise sets of 10-15 reps (lasting about 40 seconds) they are rewarded with a pat on the back and short break and a drink of water.

I like to start them with balance and leg strength, then give their legs a rest with upper body strength and fine motor skills. Then finish with stair work and gait retraining. As they fatigue, their balance in gait starts to suffer and once this becomes obvious it is time to call it a day. I think it is best for them to finish their session feeling like they have finished strong rather than too tired and getting unsteady- this is a confidence killer.

As far as backing up for a second session 3 or 7 days later, I have never once heard them say that they are still tired or sore from the previous session. This makes me a strong believer in the benefits of 'progressive overload exercise principals' with our clients.

However, for our high level [severe stage] clients i.e. "wheelchair bound" I am far more conservative when it comes to approaching fatigue. I much rather finish a session before they start feeling tired and agitated so that they are still happy to see me next week rather than pushing them too far (which they certainly remember!) and have them feeling apprehensive about the next session."

-Nick Kenny, MJDF Manager, Community Services/Active Performance Physiotherapist

The program has been enthusiastically embraced by clients and there have been genuine fitness and social and emotional wellbeing gains achieved.

In addition, a phased research project will commence in early 2017 designed to investigate the physical activities that are important to the remote Aboriginal cohort the MJDF provides services to. The long term aim will be to design appropriate programs that will engage clients maximally and ultimately allow appropriate activity 'dose' to be determined.

4. Occupational Therapy

Occupational Therapy (OT) aims to promote health and wellbeing through occupation by enabling people to have maximum participation in their activities of everyday life. The positive impacts of occupational therapy are therefore potential across the physical and psychosocial domains.

A tailored approach to therapeutic interventions that aims to adapt a particular patient to a the activities of his/her daily living in order to achieve maximal independence lies at the core of occupational therapy. Therapists work closely with patients to determine strategies and solutions to functional issues, such as access to food, clothing, personal hygiene and leisure (Silva, 2010). The necessary variability of OT interventions and their qualitative nature is, however, part of the reason for the limited literature available to ascertain the effectiveness of treatments (Silva, 2010).

In degenerative, progressive conditions such as MJD where there are no known pharmacological or surgical treatments the objective of OT is to improve the capacity and abilities of people who have disabilities. This entails tailored permanent management strategies.

Typically the *skills* of the patient in terms of their strength and activity tolerance or endurance need to be best matched to the *task*. This may entail change to the requirements of the task or the introduction of adaptive equipment. Both skills and the task need to be considered in the context of the *environment* in which the activity is occurring and may require adaptations or social supports (Piernik-Yoder, 2012).

4.1. Occupational Therapy -MJD Quality of Life and Depression

In one of the only published evaluations of OT intervention in MJD, the impact of 6 months of OT intervention for patients with MJD was assessed utilising standardised incapacitation (Functional Independence Measure (FIM) and Barthel Incapacitation Index) and depression tools (Hamilton Rating Scale for depression) (Silva et al., 2010).

At the onset of the study the group of patients showed on average mild depressive symptoms and had mild disability scores despite substantial disability scores as measured by SARA and NESSCA. Quality of life was on average moderately comprised.

At the completion of the study incapacitation scores were stable – which may be related to the natural history of progression of the disease but is not possible to determine in an open trial.

Depressive symptoms of MJD were improved after the trial and this is an important finding given that organic or reactive depression symptoms can affect up to 35% of people who have MJD (Cechin et al., 2007).

4.2. Occupational Therapy in the Remote Australian Context

OT interventions conducted by therapists in the MJDF have comprised Activity of Daily Living (ADL) assessments and interventions, home assessment and modifications, equipment prescription, installation and training (see **MJD Medical Protocol - Assistive Technology (Equipment) per stage of MJD**) and a range of psycho-social and social and emotional wellbeing interventions.

OT's work closely with physiotherapists and speech pathologists in a coordinated team to enable people with MJD to have integrated care. There is a strong focus on appropriate client led activities which frequently include trips 'on country' where patients are able to focus on activities that are important to them and enable them to participate in important family and cultural activities. These fun and functional therapy programs may involve collecting firewood, fishing, hunting, shopping, banking, practicing car transfers, or more classic therapeutic activities such as hydrotherapy.

Therapists have found it is important to consider the environment, food security, physical abilities and carer engagement / burden when designing and implementing programs and activities and stress the need to be flexible and responsive to changes.

5. Speech Pathology

Speech pathology is an important therapy modality for people who have MJD. Incidence of dysarthria, difficulty speaking caused by problems controlling speech muscles, is between 85-100% among people who have MJD (Jardim et al, 2001; Maruyama et al., 1995; Soon et al., 1997). There is very little published literature regarding speech pathology for MJD; however, the MJDF has engaged academics to assess the communication and swallowing needs of people with MJD and best practice guidelines for these issues can be found in the Communication and Swallowing Medical Protocols.

5.1. Communication and MJD

There are no published studies that show speech therapy is beneficial for clients with MJD.

"An unpublished University of Queensland study of 11 MJD Foundation clients [by Theodoras and Ward (2011)], found varying levels of severity of dysarthria, (slight to severe) consistent with previous studies. The most common perceptual findings included reduced breath support for speech, loudness, rate of speech, precision of speech sounds and phrase length, and a breathy vocal quality. These findings were supported by motor speech impairment predominantly in respiratory-phonatory function and tongue function. Impaired vocal fold adduction, breath control for speech, and reduced range and speed of tongue movements during speech were evident. Respiratory-phonatory function relates to the person's capacity to breathe and produce a normal voice.

Early signs of motor speech impairment were evident in some people with MJD but did not consistently impair speech intelligibility. These included:

- weak cough
- decreased sustained phonation
- decreased volume when tired
- decreased breath support for speech, and dribbling when tired."

(p. 12, MJDF Medical Protocol – Communication Difficulty)

Dr Theodoros completed the LSVT ® (Lee Silverman Voice Treatment) program with an MJD client following this research conducted with Dr Ward in 2011. The program was conducted 4 times a week for 4 weeks via Skype sessions.

The LSVT ® program was developed for clients with another neurodegenerative disorder, Parkinson's disease. The research data indicates improvements in vocal loudness, intonation and voice quality for individuals with PD for up to 2 years after treatment.

Anecdotally, the LSVT ® program has benefited this client with MJD immensely. She is able to still use a loud voice, 4 years since the originally therapy was provided. We are not able to say whether her voice would have deteriorated more without the

treatment but it has had an effect on her self-esteem and willingness to participate in communicative activities (e.g. she is able to yell out to her sister across the road and call out to the ladies when she is 'out bush').

It has yet to be determined whether these exercises may slow the disease progression; however, it may help the clients to improve their self-esteem, cope better with their disability in the longer term, be more aware of their communication and safe swallowing strategies, boost their mood and finally give them a sense of control over their disease.

Therefore, a staged approach to communication therapy for people living with MJD is recommended:

No Speech Disorder

Where the person with MJD has not yet developed a speech disorder, speech is the primary mode for communication. During this stage, education on dysarthria and strategies to improve speech in addition to monitoring of changes to speech is recommended.

Reduced Intelligibility

When symptoms of speech decline emerge, voice therapy, behavioural strategies, environment modifications, assessment for augmentative and alternative communication (AAC), and training and learning about AAC are all recommended.

Speech is still the primary mode for communication for people with MJD in this stage.

Residual speech (Severe Dysarthria)

In this stage speech is not intact and is often distorted; however, the person with MJD can still produce some sounds. They might still use some speech with augmentative tools. AAC becomes the primary modality for speech. Ongoing assessment and tailoring of AAC tools is recommended due to the progressiveness of MJD.

Anarthria

Speech of a person with MJD eventually deteriorates to anarthria, or the inability to articulate speech due to the loss of neuromuscular control of the speech muscles. At this stage, communication is totally reliant on AAC. Ongoing assessment and tailoring of AAC tools is recommended due to the progressiveness of MJD.

Regarding fatigue and whether or not it is recommended to work with MJD clients to fatigue, authors of this document could not find any studies in speech pathology or other disciplines (physiotherapy, occupational therapy) that discuss whether it is recommended to work people with MJD to fatigue.

Micaela Jackson, a volunteer speech therapist working on Groote Eylandt with MJDF clients delivering weekly therapy, gives this advice:

"My experience with MJD clients is that it is important not to push the clients until they are fatigued. I feel they gain more out of the session if they try their hardest during therapy sessions but stop the session before they are fatigued. Once fatigued, it affects their mobility, posture, communication and possibly their swallowing function. I don't like to push them to this degree as they may injure themselves or cough/choke on food or fluids."

-Michaela Jackson, SP, MJDF Volunteer

For Indigenous Australians with MJD, communication problems are compounded by the prevalence of hearing loss: up to 70 percent of adults and at least 40 percent have auditory processing problems (Howard, 2012). Local sign languages can be a common mode of communication for Aboriginal peoples and are not always related to deafness (Kendon, 2015). Speech therapists working with Aboriginal clients with MJD in the Northern Territory have found incorporation of sign language into treatment programs helpful.

Current research is being undertaken by the MJD Foundation to explore communication for Aboriginal Australians who have MJD and we hope to understand more in the near future.

Please refer to the **MJD Medical Protocol – Communication Difficulty** for further detail.

5.2. Swallowing and MJD in the Remote Australian Context

What may work best in a hospital or clinical setting may not be the best option for Aboriginal Australians with MJD returning home to a remote community. For example, there are many barriers they may face both practically and culturally that may inhibit the use of a percutaneous endoscopic gastrostomy (PEG) (otherwise known as a 'feeding tube') or nutritional supplements.

Remote communities do not have the facilities to cope with blocked PEG tubes and often the home environment will not be appropriate for the use of a PEG (e.g. clients may be sleeping in the bed with other people, animals living in the house causing reduced hygiene). Culturally, Aboriginal clients may give away nutritional supplements like puddings and thickened fluids to other family members in need. They also may want to participate in family gatherings and rituals involving food.

The MJDF has found that families caring for loved ones with MJD are very resourceful in the way they adapt to swallowing difficulties. For example, instead of using a dysphagia cup they will adapt a polystyrene cup to a client's mouth who may have lip seal issues.

It is important to educate people with MJD and their families and carers on what happens when someone may have swallowing problems (including aspiration) and provide the client/family with the "safest" way for swallowing particular foods. It is equally as important to introduce safe swallowing strategies early (e.g. sitting upright, one sip at a time, drinking top ¾ cup only, supraglottic swallow).

Please refer to the **MJD Medical Protocol – Difficulty Swallowing**.

Appendix A – Contributors and Reviewers

Contributors

Ms Libby Massey
B.Ap.Sc.(OT), MPH(Dtn)
MJD Foundation

Micahaela Jackson
B.Ap.Sc.(SP)
MJD Foundation

Nick Kenny
B.Ap.Sc.(PT)
Active Performance/MJD Foundation

Rebecca Amery
B.Ap.Sc.(SP)
Charles Darwin University/
MJD Foundation

Desireé LaGrappe
B.Sc.Nurs.(Hons) (RN)
MJD Foundation

Ms Nadia Lindop
B.Sc.(Hons), Cert IV Finance, Cert IV WHS MJD Foundation

Reviewers

Prof Carlos R Gordon, MD; DSc
Department of Neurology, Meir Medical Center, Kfar-Saba and Sackler Faculty of Medicine
Tel Aviv University, Israel

Jaimee Dutton, Speech Pathologist
Office of Disability, Top End Remote
Department of Health
Northern Territory Government

Appendix B – Definitions

AAC	Augmentative and alternative communication (AAC) includes all forms of communication (other than oral speech) that are used to express thoughts, needs, wants, and ideas. AAC includes the types unaided (relies on the user's body e.g. gestures) and aided (requires the use of tools or equipment e.g. alphabet board).
Anarthria	Loss of the ability to produce speech
GAS	Global Attainment Score
SARA	Scale for the rating and assessment of ataxia
Frenkel Coordination	A series of motions of increasing difficulty performed by ataxic patients to facilitate the restoration of coordination. Frenkel's exercises are used to bring back the rhythmic, smooth and coordinated movements.
ADL's	Activities of Daily Living
Biofeedback	The use of electronic monitoring of a normally automatic bodily function in order to train someone to acquire voluntary control of that function.
Neuropathy	Disease or dysfunction of one or more peripheral nerves, typically causing numbness or weakness
Opthalmoparesis	Weakness (-paresis) or paralysis (-plegia) of one or more extraocular muscles which are responsible for eye movements.
Percutaneous endoscopic gastrostomy (PEG)	a feeding tube inserted through the skin and the stomach wall. It goes directly into the stomach. Feeding tubes are needed when you are unable to eat or drink.

Appendix C – Measurement Scales

Rater: _____date: _____patient:_____

Scale for the assessment and rating of ataxia (SARA)

1) Gait	2) Stance
Proband is asked (1) to walk at a safe distance parallel to a wall including a half-turn (turn around to face the opposite direction of gait) and (2) to walk in tandem (heels to toes) without support.	Proband is asked to stand (1) in natural position, (2) with feet together in parallel (big toes touching each other) and (3) in tandem (both feet on one line, no space between heel and toe). Proband does not wear shoes, eyes are open. For each condition, three trials are allowed. Best trial is rated.
0 Normal, no difficulties in walking, turning and walking tandem (up to one misstep allowed) **1** Slight difficulties, only visible when walking 10 consecutive steps in tandem **2** Clearly abnormal, tandem walking >10 steps not possible **3** Considerable staggering, difficulties in half-turn, but without support **4** Marked staggering, intermittent support of the wall required **5** Severe staggering, permanent support of one stick or light support by one arm required **6** Walking > 10 m only with strong support (two special sticks or stroller or accompanying person) **7** Walking < 10 m only with strong support (two special sticks or stroller or accompanying person) **8** Unable to walk, even supported	**0** Normal, able to stand in tandem for > 10 s **1** Able to stand with feet together without sway, but not in tandem for > 10s **2** Able to stand with feet together for > 10 s, but only with sway **3** Able to stand for > 10 s without support in natural position, but not with feet together **4** Able to stand for >10 s in natural position only with intermittent support **5** Able to stand >10 s in natural position only with constant support of one arm **6** Unable to stand for >10 s even with constant support of one arm
Score	**Score**
3) Sitting	**4) Speech disturbance**
Proband is asked to sit on an examination bed without support of feet, eyes open and arms outstretched to the front.	Speech is assessed during normal conversation.
0 Normal, no difficulties sitting >10 sec **1** Slight difficulties, intermittent sway **2** Constant sway, but able to sit > 10 s without support **3** Able to sit for > 10 s only with intermittent support **4** Unable to sit for >10 s without continuous support	**0** Normal **1** Suggestion of speech disturbance **2** Impaired speech, but easy to understand **3** Occasional words difficult to understand **4** Many words difficult to understand **5** Only single words understandable **6** Speech unintelligible / anarthria
Score	**Score**

For a demonstration on clients with MJD visit https://vimeo.com/100856108

1

Rater: _____date: _____patient:_____

5) Finger chase

Rated separately for each side
Proband sits comfortably. If necessary, support of feet and trunk is allowed. Examiner sits in front of proband and performs 5 consecutive sudden and fast pointing movements in unpredictable directions in a frontal plane, at about 50 % of proband´s reach. Movements have an amplitude of 30 cm and a frequency of 1 movement every 2 s. Proband is asked to follow the movements with his index finger, as fast and precisely as possible. Average performance of last 3 movements is rated.

0 No dysmetria
1 Dysmetria, under/ overshooting target <5 cm
2 Dysmetria, under/ overshooting target < 15 cm
3 Dysmetria, under/ overshooting target > 15 cm
4 Unable to perform 5 pointing movements

Score	Right	Left
mean of both sides (R+L)/2		

6) Nose-finger test

Rated separately for each side
Proband sits comfortably. If necessary, support of feet and trunk is allowed. Proband is asked to point repeatedly with his index finger from his nose to examiner's finger which is in front of the proband at about 90 % of proband's reach. Movements are performed at moderate speed. Average performance of movements is rated according to the amplitude of the kinetic tremor.

0 No tremor
1 Tremor with an amplitude < 2 cm
2 Tremor with an amplitude < 5 cm
3 Tremor with an amplitude > 5 cm
4 Unable to perform 5 pointing movements

Score	Right	Left
mean of both sides (R+L)/2		

7) Fast alternating hand movements

Rated separately for each side
Proband sits comfortably. If necessary, support of feet and trunk is allowed. Proband is asked to perform 10 cycles of repetitive alternation of pro- and supinations of the hand on his/her thigh as fast and as precise as possible. Movement is demonstrated by examiner at a speed of approx. 10 cycles within 7 s. Exact times for movement execution have to be taken.

0 Normal, no irregularities (performs <10s)
1 Slightly irregular (performs <10s)
2 Clearly irregular, single movements difficult to distinguish or relevant interruptions, but performs <10s
3 Very irregular, single movements difficult to distinguish or relevant interruptions, performs >10s
4 Unable to complete 10 cycles

Score	Right	Left
mean of both sides (R+L)/2		

8) Heel-shin slide

Rated separately for each side
Proband lies on examination bed, without sight of his legs. Proband is asked to lift one leg, point with the heel to the opposite knee, slide down along the shin to the ankle, and lay the leg back on the examination bed. The task is performed 3 times. Slide-down movements should be performed within 1 s. If proband slides down without contact to shin in all three trials, rate 4.

0 Normal
1 Slightly abnormal, contact to shin maintained
2 Clearly abnormal, goes off shin up to 3 times during 3 cycles
3 Severely abnormal, goes off shin 4 or more times during 3 cycles
4 Unable to perform the task

Score	Right	Left
mean of both sides (R+L) / 2		

For a demonstration on clients with MJD visit https://vimeo.com/100856108

2

PATIENT:_____DATE of BIRTH:_____

INTERNATIONAL CO-OPERATIVE ATAXIA RATING SCALE

I: POSTURE AND GAIT DISTURBANCE		SCORE:
1. **WALKING CAPACITIES** observed during a 10 meter test including a half-turn, near a wall, at about 1,5meter.	**0:** normal **1:** almost normal naturally, but unable to walk with feet in tandem position **2:** Walking without support, but clearly abnormal and irregular **3:** Walking without support but with considerable staggering, diffculties in half turn **4:** Walking with autonomous support no longer possible, the patient uses episodic support of the wall for a 10 meter test **5**: Walking only possible with one stick **6**: Walking only possible with two special sticks or with a stroller **7**: Walking only with accompanying person **8:** walking impossible even with accompanying person (wheelchair)	
2: **GAIT SPEED** observed in patients with preceeding scores 1-3, preceeding score 4 and up gives automatically score 4 in this test.	**0:** normal **1:** slightly reduced **2:** markedly reduced **3:** extremely slow **4:** walking with autonomous support no longer possible	
3: **STANDING CAPACITIES,** ***EYES OPEN*** the patient is asked first to stand on one foot if impossible , to stand with feet in tandem position if impossible to stand with feet together for the natural position the patient is asked to find a comfortable standing position	**0:** normal, able to stand on one foot more than 10 sec **1:** able to stand with feet together, but no longer able to stand on one foot more than 10 sec. **2:** able to stand with feet together, but no longer able to stand in tandem position **3:** no longer able to stand with feet together, but able to stand in natural position without support, with no or moderate sway **4:** standing in natural position without support, with considerable sway and considerable corrections	

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	5: unable to stand in natural position without strong support of the arms **6:** unable to stand at all, even with string support of the arms	
4: **SPREAD OF FEET IN NATURAL POSITION WITHOUT SUPPORT** ***EYES OPEN*** the patient is asked to find a comfortable position, then the distance between medial malleoli is measured.	**0:** normal <10cm **1:** slightly enlarged >10cm **2:** clearly enlarged 25cm < spread <35cm **3:** severely enlarged >35cm **4:** standing in natural position impossible	
5: **BODY SWAY WITH FEET TOGETHER** ***EYES OPEN***	**0:** normal **1:** slightly oscillations **2:** moderate oscillations (<10cm at the level of head) **3:** severe oscillations (>10cm at the level of head), threatening the upright position **4:** immediate falling	
6: **BODY SWAY WITH FEET TOGETHER** ***EYES CLOSED***	**0:** normal **1:** slight oscillations **2:** moderate oscillations (<10cm at the level of head) **3:** severe oscillations (>10cm at the level of head), threatening the upright position **4:** immediate falling	
7: **QUALITY OF SITTING POSITION** thighs together, on a hard surface, arms folded	**0:** normal **1:** with slight oscillations of the trunk **2:** with moderate oscillations of the trunk and legs **3:** with severe dysequilibrium **4:** impossible	
POSTURE AND GAIT SCORE (STATIC SCORE)		___/ 34

II: KINETIC FUNCTIONS		SCORE:
8: **KNEE-TIBIA TEST decomposition of movement and intention tremor.** The test is performed in the supine position, but the head is tilted, so that visual control is possible. The patient is requested to raise one leg and place the heel on the knee, and then slide the heel down the anterior tibial surface of the resting leg towards the ankle. On reaching the ankle joint, the leg is again raised in the air to a height of approximately 40 cms and the action is repeated. At least 3 movements of each limb must be performed for proper assessment.	**0:** normal **1:** lowering of heel in continuous axis, but the movement is decomposed in several phases, without real jerks, or abnormally slow **2:** lowering jerkily in the axis **3:** lowering jerkily with lateral movements **4:** lowering jerkily with extremely strong lateral movements or test impossible	R: L:
9: **ACTION TREMOR in the HEEL-TO-KNEE Test** Same test as preceeding one: the action tremor of the heel on the knee is specifically observed when the patient holds the heel on the knee for a few seconds before sliding down the anterior tibial surface; visual control is required	**0:** No trouble **1:** Tremor stopping immediately when the heel reaches the knee **2**: Tremor stopping in less than 10 seconds after reaching the knee **3**: Tremor continuing for more than 10 seconds after reaching the knee **4:** uninterrupted tremor or test impossible	R: L:
10: **FINGER-TO-NOSE TEST** **decomposition and dysmetria** the subject sits on a chair, the hand is resting on the knee before the beginning of the movement, visual control is required. Three movements of each limb must be performed for proper assessment.	**0:** no trouble **1:** Oscillating movement without decomposition of the movement **2:**Segmented movement in more than 2 phases and/or moderate dysmetria in reaching nose **3:** segmented movement in more than 2 phases and /or considerable dysmetria in reaching nose **4:** Dysmetria preventing the patient from reaching the nose	R: L:

11: **FINGER-TO-NOSE TEST** **<u>intention tremor</u> of the finger** the studied tremor is that appeared during the ballistic phase of the movement; the patient is sitting comfortably, with his hands resting on his/her thigh; visual control is requires; three movements of each limb must be performed as proper assessment	**0:** No trouble **1:** simple swerve of the movement **2:** moderate tremor with estimated amplitude <10cm **3:** Tremor with estimated amplitude between 10cm und 40cm **4:** severe tremor with estimated amplitude >40cm	R: L:
12: **FINGER-FINGER-TEST** **action tremor and/or instability** the sitting patient is asked to maintain medially his/her index fingers pointing at each other for about 10 sec, at a distance of about 1cm, at the level of the thorax, under visual control.	**0:** normal **1:** mild instability **2:** moderate oscillations of finger with estimated amplitude <10cm **3:** considerable oscillations of finger with estimated amplitude between 10 and 40cm **4:** Jerky movement >40cm of amplitude	R: L:
13: **PRONATION-SUPINATION** **altering movements** the subject, comfortably sitting on a chair, is asked to raise his/her forearm vertically and to make alternative movements of the hand. Each hand is moved and assessed separately.	**0:** normal **1:** slightly irregular and slowed **2:** clearly irregular and slowed, but without sway of the elbow **3:** extremely irregular and slowed movement, with sway of the elbow **4:** movement completely disorganized or impossible	R: L:
14: **DRAWING the Archimedes spiral on a predrawn pattern** the subject is comfortly setttled in front of the table, the sheet of paper is being fixed to avoid artefacts. The subject is asked to perform the task without timing requirements. The same condition of examination must be used at each examination.	**0:** normal **1:** impairment and decomposition, the line quitting the pattern slightly, but without hypermetric swerve **2:** line completely out of the pattern without recrossing and/or hypermetric swerves **3:** major disturbance due to hypermetria and decomposition **4:** drawing completely disorganised or impossible	
<u>KINETIC SCORE (limb coordination):</u>		**___/52**

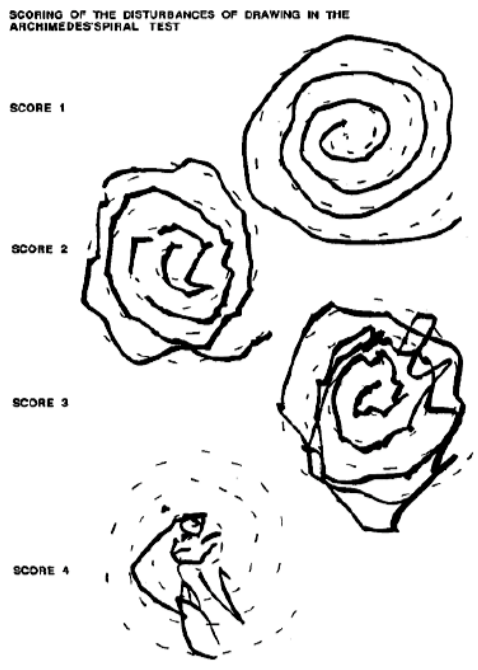

Fig. 1. Scoring of the disturbances of drawing in the Archimedes' spiral test.

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III: SPEECH DISORDERS		SCORE:
15: **DYSARTHRIA: fluency of speech** The patient is asked to repeat several times a standard sentence, always the same.	**0:** normal **1:** mild modification of fluency **2:** moderate modification of fluency **3:** considerably slow and dysarthric speech **4:** no speech	
16: **DYSARTHRIA: clarity of speech**	**0:** normal **1:** suggestion of slurring **2:** definite slurring, most words understandable **3:** severe slurring, speech not understandable **4:** no speech	
DYSARTHRIA SCORE:		**___/8**
IV: OCULOMOTOR DISORDERS		SCORE:
17: **GAZE EVOKED NYSTAGMUS** the subject is asked to look laterally at the finger of the examiner: the movement assessed are mainly horizontal, but they may be oblique, rotatory, or vertical.	**0:** normal **1:** transient **2:** persistent but moderate **3:** persistent as severe	
18: **ABNORMALITIES OF THE OCULAR PURSUIT** the subject is asked to follow the slow lateral movement, performed by the finger of the examiner	**0:** normal **1:** slightly saccadic **2:** clearly saccadic	
19: **DYSMETRIA OF THE SACCADE** the two index fingers of the examiner in each visual field, average overshoot/undershoot is estimated	**0:** absent **1:** bilateral clear overshoot or undershoot of the saccade	
OCULOMOTOR MOVEMENT SCORE:		**___/6**
TOTAL ATAXIA SCORE:		**___/100**

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Modified International Cooperative Ataxia Rating Scale (MICARS)

Implement the ICARS adding the following tests:

after <u>Test 9</u>: Decomposition of leg movement (Left and Right scored)	0: Normal
	1: Corners or edges on the circle
	2: Markedly decomposed attempts at circle
Decomposition of leg tapping (Left and Right scored)	0: Normal
	1: Slightly slow and irregular
	2: Clearly slow and irregular
after <u>Test 15</u>: Rebound of the arms (Left and Right scored)	0: None
	1: Less than 10 cm
	2: Greater than 10 cm
Overshoot of the arms (Left and Right scored)	0: None
	1: Less than 10 cm

	2: Greater than 10 cm
after <u>Test 20</u>: Dysarthria: Alternating syllables	0: Normal
	1: Slightly irregular
	2: Clearly irregular, dysrhythmic and slurred
Abnormal eye movements at rest	0: Absent
	1: Present
after <u>Test 25</u>: Saccadic intrusions into vestibulo-ocular reflex cancellation	0: Absent
	1: Present

NESSCA			
Item	**Proofs**	**Severity**	**Score**
Gait ataxia	- Walking spontaneously, ten steps, parallel to a wall, and including a half-turn - Walking on toes, on heels, and in tandem	Absent	0
		Minimal: only while walking on toes, heels, or in tandem	1
		Moderate: gait autonomy preserved	2
		Inability to walk without help	3
		Wheelchair bound or bedridden	4
Limb ataxia (bilateral)	- Finger-to-nose test - Test for dysdiadochokinesia (fast alternating pronation and supination of hands, elbows fixed to his/her sides) - Rebound test of Gordon-Holmes	Absent	0
		Minimal: one single altered proof	1
		Moderate: two altered proofs	2
		Important: three altered proofs	3
		Proofs: (a) dysmetria, (b) fast alternating hand movements, and (c) upper limb rebound. Positive findings can be uni or bilateral.	
Nystagmus		Absent	0
		On extreme gaze; or circular, after saccades	1
		Permanent	2
Progressive external ophthalmoplegia		Absent	0
		Supranuclear: medial longitudinal fasciculus syndrome; or limitation in upward gaze or convergence	1
		Nuclear ophthalmoplegia, with strabismus	2
Pyramidal findings	- Limb reflexes, including patellar and ankle clonus test - Plantar reflex - Muscle tone examination - Motor strength proofs: extended arms and Mingazzini test, both for 60 seconds	Absent	0
		Few brisk reflexes	1
		General hyperreflexia; or clonus; or Babinski sign	2
		Three findings: (a) general hyperreflexia, (b) spasticity, (c) clonus, (d) Babinski sign; (e) paresis	3
		Four or five of the above mentioned signs	4

Dysarthria		Absent	0
		Mild: Impaired speech, but easy to understand	1
		Moderate: speech understandable, but with difficulty	2
		Severe: speech hardly understandable	3
		Anarthria	4
Dysphagia		Absent	0
		Mild	1
		Important: occuring every day	2
Fasciculations		Absent	0
		Contraction fasciculation in the face	1
		Diffuse, or in other parts of the body	2
Sensory loss	(a) Vibratory sense in the first toes; normal: >11 sec. (b) Discrimination between tactile and algesic stimuli using a needle; 10 trials per foot. (c) Discrimination between cold (10^{o}C) and warm (40-60^{o}C) water; 10 trials per foot	Absent	0
		One altered proof: Reduction in (a) or (b) or (c): two to four mistakes, on average of both feet	1
		Two altered proofs	2
		Total loss of vibratory sense in toes; or 5 or more mistakes in one of discriminating proofs; or three altered proofs	3
Dystonia		Absent	0
		Mild, triggered by voluntary movements	1
		Moderate, impairing, in some degree, voluntary movements	2
		Almost constant, impairing severely voluntary movements	3
Rigidity		Absent	0
		Moderate: does not prevent total, passive mobilization	1
		Important: prevent total, passive mobilization	2

Bradykinesia	- Patient is asked to perform 10 cycles of repetitive opposition (extension and flexion) of the second finger against the thumb	Absent	0
		Slow movements, with reduction in amplitude	1
		Movements can hardly be done	2
Eyelid retraction		Absent	0
		Present	1
Blepharospasm		Absent	0
		Present	1
Distal amyotrophies	- Inspection of the interossei, tenar and hypotenar muscles	Absent	0
		Present	1
Sphincter function		Normal	0
		Urgency	1
		Incontinence	2
Cramps		Absent	0
		Present	1
Vertigo		Absent	0
		Present	1
Total score			

Barthel Index of Activities of Daily Living

Instructions: Choose the scoring point for the statement that most closely corresponds to the patient's current level of ability for each of the following 10 items. Record actual, not potential, functioning. Information can be obtained from the patient's self-report, from a separate party who is familiar with the patient's abilities (such as a relative), or from observation. Refer to the Guidelines section on the following page for detailed information on scoring and interpretation.

The Barthel Index

Bowels
0 = incontinent (or needs to be given enemata)
1 = occasional accident (once/week)
2 = continent
Patient's Score: _____

Bladder
0 = incontinent, or catheterized and unable to manage
1 = occasional accident (max. once per 24 hours)
2 = continent (for over 7 days)
Patient's Score: _____

Grooming
0 = needs help with personal care
1 = independent face/hair/teeth/shaving (implements provided)
Patient's Score: _____

Toilet use
0 = dependent
1 = needs some help, but can do something alone
2 = independent (on and off, dressing, wiping)
Patient's Score: _____

Feeding
0 = unable
1 = needs help cutting, spreading butter, etc.
2 = independent (food provided within reach)
Patient's Score: _____

Transfer
0 = unable – no sitting balance
1 = major help (one or two people, physical), can sit
2 = minor help (verbal or physical)
3 = independent
Patient's Score: _____

Mobility
0 = immobile
1 = wheelchair independent, including corners, etc.
2 = walks with help of one person (verbal or physical)
3 = independent (but may use any aid, e.g., stick)
Patient's Score: _____

Dressing
0 = dependent
1 = needs help, but can do about half unaided
2 = independent (including buttons, zips, laces, etc.)
Patient's Score: _____

Stairs
0 = unable
1 = needs help (verbal, physical, carrying aid)
2 = independent up and down
Patient's Score: _____

Bathing
0 = dependent
1 = independent (or in shower)
Patient's Score: _____

Total Score: _____

(Collin et al., 1988)

Scoring:

Sum the patient's scores for each item. Total possible scores range from 0 – 20, with lower scores indicating increased disability. If used to measure improvement after rehabilitation, changes of more than two points in the total score reflect a probable genuine change, and change on one item from fully dependent to independent is also likely to be reliable.

Sources:

- Collin C, Wade DT, Davies S, Horne V. The Barthel ADL Index: a reliability study. *Int Disabil Stud.* 1988;10(2):61-63.
- Mahoney FI, Barthel DW. Functional evaluation: the Barthel Index. *Md State Med J.* 1965;14:61-65.
- Wade DT, Collin C. The Barthel ADL Index: a standard measure of physical disability? *Int Disabil Stud.* 1988;10(2):64-67.

1

Guidelines for the Barthel Index of Activities of Daily Living

General

- The Index should be used as a record of what a patient ***does***, NOT as a record of what a patient ***could do***.
- The main aim is to establish degree of independence from any help, physical or verbal, however minor and for whatever reason.
- The need for supervision renders the patient not independent.
- A patient's performance should be established using the best available evidence. Asking the patient, friends/relatives, and nurses will be the usual source, but direct observation and common sense are also important. However, direct testing is not needed.
- Usually the performance over the preceding 24 – 48 hours is important, but occasionally longer periods will be relevant.
- Unconscious patients should score '0' throughout, even if not yet incontinent.
- Middle categories imply that the patient supplies over 50% of the effort.
- Use of aids to be independent is allowed.

Bowels (preceding week)

- If needs enema from nurse, then 'incontinent.'
- 'Occasional' = once a week.

Bladder (preceding week)

- 'Occasional' = less than once a day.
- A catheterized patient who can completely manage the catheter alone is registered as 'continent.'

Grooming (preceding 24 – 48 hours)

- Refers to personal hygiene: doing teeth, fitting false teeth, doing hair, shaving, washing face. Implements can be provided by helper.

Toilet use

- Should be able to reach toilet/commode, undress sufficiently, clean self, dress, and leave.
- 'With help' = can wipe self and do some other of above.

Feeding

- Able to eat any normal food (not only soft food). Food cooked and served by others, but not cut up.
- 'Help' = food cut up, patient feeds self.

Transfer

- From bed to chair and back.
- 'Dependent' = NO sitting balance (unable to sit); two people to lift.
- 'Major help' = one strong/skilled, or two normal people. Can sit up.
- 'Minor help' = one person easily, OR needs any supervision for safety.

Mobility

- Refers to mobility about house or ward, indoors. May use aid. If in wheelchair, must negotiate corners/doors unaided.
- 'Help' = by one untrained person, including supervision/moral support.

Dressing

- Should be able to select and put on all clothes, which may be adapted.
- 'Half' = help with buttons, zips, etc. (*check!*), but can put on some garments alone.

Stairs

- Must carry any walking aid used to be independent.

Bathing

- Usually the most difficult activity.
- Must get in and out unsupervised, and wash self.
- Independent in shower = 'independent' if unsupervised/unaided.

(Collin et al., 1988)

2

Functional Independence Measure (FIM) Instrument

	ADMISSION	DISCHARGE	FOLLOW-UP
Self-Care			
A. Eating			
B. Grooming			
C. Bathing			
D. Dressing - Upper Body			
E. Dressing - Lower Body			
F. Toileting			
Sphincter Control			
G. Bladder Management			
H. Bowel Management			
Transfers			
I. Bed, Chair, Wheelchair			
J. Toilet			
K. Tub, Shower			
Locomotion			
L. Walk/Wheelchair			
M. Stairs			
Motor Subtotal Score			
Communication			
N. Comprehension			
O. Expression			
Social Cognition			
P. Social Interaction			
Q. Problem Solving			
R. Memory			
Cognitive Subtotal Score			
TOTAL FIM Score			

LEVELS	**Independent** 7 Complete Independence (Timely, Safely) 6 Modified Independence (Device)	**NO HELPER**
	Modified Dependence 5 Supervision (Subject = 100%+) 4 Minimal Assist (Subject = 75%+) 3 Moderate Assist (Subject = 50%+) **Complete Dependence** 2 Maximal Assist (Subject = 25%+) 1 Total Assist (Subject = less than 25%)	**HELPER**
Note: Leave no blanks. Enter 1 if patient is not testable due to risk.		

Patient Name:_____ **Date:**_____

Hamilton Rating Scale for Depression (17-items)

Instructions: For each item select the "cue" which best characterizes the patient during the past week.

1. **Depressed Mood**
   (sadness, hopeless, helpless, worthless)
   0 Absent
   1 These feeling states indicated only on questioning
   2 These feeling states spontaneously reported verbally
   3 Communicates feeling states nonverbally, i.e., through facial expression, posture, voice and tendency to weep
   4 Patient reports VIRTUALLY ONLY these feeling states in his spontaneous verbal and nonverbal communication

2. **Feelings of Guilt**
   0 Absent
   1 Self-reproach, feels he has let people down
   2 Ideas of guilt or rumination over past errors or sinful deeds
   3 Present illness is a punishment. Delusions of guilt
   4 Hears accusatory or denunciatory voices and/or experiences threatening visual hallucinations

3. **Suicide**
   0 Absent
   1 Feels life is not worth living
   2 Wishes he were dead or any thoughts of possible death to self
   3 Suicide ideas or gesture
   4 Attempts at suicide (any serious attempt rates 4)

4. **Insomnia - Early**
   0 No difficulty falling asleep
   1 Complains of occasional difficulty falling asleep i.e., more than ½ hour
   2 Complains of nightly difficulty falling asleep

5. **Insomnia - Middle**
   0 No difficulty
   1 Patient complains of being restless and disturbed during the night
   2 Waking during the night – any getting out of bed rates 2 (except for purposes of voiding)

6. **Insomnia - Late**
   0 No difficulty
   1 Waking in early hours of the morning but goes back to sleep
   2 Unable to fall asleep again if gets out of bed

7. **Work and Activities**
   0 No difficulty
   1 Thoughts and feelings of incapacity, fatigue or weakness related to activities; work or hobbies
   2 Loss of interest in activity; hobbies or work – either directly reported by patient, or indirect in listlessness, indecision and vacillation (feels he has to push self to work or activities)
   3 Decrease in actual time spent in activities or decrease in productivity. In hospital, rate 3 if patient does not spend at least three hours a day in activities (hospital job or hobbies) exclusive of ward chores.
   4 Stopped working because of present illness. In hospital, rate 4 if patient engages in no activities except ward chores, or if patient fails to perform ward chores unassisted.

8. **Retardation**
   (slowness of thought and speech; impaired ability to concentrate; decreased motor activity)
   0 Normal speech and thought
   1 Slight retardation at interview
   2 Obvious retardation at interview
   3 Interview difficult
   4 Complete stupor

9. **Agitation**
   0 None
   1 "Playing with" hand, hair, etc.
   2 Hand-wringing, nail-biting, biting of lips

10. **Anxiety - Psychic**
    0 No difficulty
    1 Subjective tension and irritability
    2 Worrying about minor matters
    3 Apprehensive attitude apparent in face or speech
    4 Fears expressed without questioning

11. **Anxiety - Somatic**
    0 Absent — Physiological concomitants of anxiety such
    1 Mild — Gastrointestinal - dry mouth, wind, indigesti
    2 Moderate — diarrhea, cramps, belching
    3 Severe — Cardiovascular – palpitations, headaches
    4 Incapacitating — Respiratory - hyperventilation, sighing
    Urinary frequency
    Sweating

12. **Somatic Symptoms - Gastrointestinal**
    0 None
    1 Loss of appetite but eating without staff encouragement. Heavy feelings in abdomen.
    2 Difficulty eating without staff urging. Requests or requires laxatives or medications for bowels or medication for G.I. symptoms.

13. **Somatic Symptoms - General**
    0 None
    1 Heaviness in limbs, back or head, backaches, headache, muscle aches, loss of energy and fatigability
    2 Any clear-cut symptom rates 2

14. **Genital Symptoms**
    0 Absent — 0 Not ascertained
    1 Mild — Symptoms such as: loss of libido, menstrual disturbances
    2 Severe

15. **Hypochondriasis**
    0 Not present
    1 Self-absorption (bodily)
    2 Preoccupation with health
    3 Frequent complaints, requests for help, etc.
    4 Hypochondriacal delusions

16. **Loss of Weight**
    A. When Rating by History:
    0 No weight loss
    1 Probable weight loss associated with present illness
    2 Definite (according to patient) weight loss

    B. On Weekly Ratings by Ward Psychiatrist, When Actual Changes are Measured:
    0 Less than 1 lb. weight loss in week
    1 Greater than 1 lb. weight loss in week
    2 Greater than 2 lb. weight loss in week

17. **Insight**
    0 Acknowledges being depressed and ill
    1 Acknowledges illness but attributes cause to bad food, climate, overwork, virus, need for rest, etc.
    2 Denies being ill at all

Total Score:_____

Citation: Hamilton M: A rating scale for depression. Journal of Neurology, Neurosurgery and Psychiatry 23:56-62, 1960

Appendix D – References

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