

Nutrition

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1. Background – Nutrition

Machado Joseph Disease (MJD) patients experience high rates of weight loss and malnutrition. This contributes to both morbidity and mortality and can be compounded by co-morbidities. Both occur due to the complex and interrelated symptoms of MJD. Commonly cited causes of weight loss and malnutrition include: dysphagia; reliance on others for food purchase, preparation, and in some cases feeding; and depression and anxiety.

Malnutrition is also associated with decreased immune function, increased complications / co-morbidities, delayed wound healing, increased mortality and longer periods in hospital (Correlá & Waltzberg, 2003; Sheard et al., 2011). As soon as symptoms of weight loss or malnutrition are noted, a management plan to maintain weight and nutrition should commence.

1.1 *Weight loss*

Weight loss results from a prolonged disequilibrium between intake, digestion and absorption of energy from nutrients and energy expenditure (Aziz et al., 2008) and weight loss can contribute to both morbidity (infections) and mortality (Aziz et al., 2008; Saute et al., 2011).

Machado Joseph Disease (MJD) patients experience lower body mass index (BMI) scores than other people ($p=0.01$) and a study of 36 MJD patients has found lower BMI scores are independently correlated with the length of the expanded CAGn repeats ($p=0.015$) (Saute et al., 2011). They conclude 'this association points that weight loss might be a primary disturbance of SCA3/MJD' (Saute et al., 2011).

The study also found that MJD patients' lower BMI scores were associated with Neurological Examination Score for Spinocerebellar Ataxia ($p=0.02$), age of onset ($p=0.001$), age ($p=0.008$) and serum insulin levels ($p=0.006$). They did not find a relationship between BMI and disease stage ($p=0.114$), Severity Ataxia Rating Scale, dysphagia or brainstem MRI volumetrics. The authors noted the role of dysphagia as an important cause of weight loss and analysed the NESSCA dysphagia item in accordance with patient dysphagia status. While they report no significant difference in BMI was found they note that 'further studies using a validated dysphagia instrument are necessary to confirm or exclude this finding'. (Saute et al., 2011).

Unintentional weight loss is a better predictor of malnutrition than weight or BMI at a single point in time (Tapsell, 2009). Also, that BMI is 'not sensitive enough to recognise small yet clinically significant weight loss', particularly where skeletal integrity may cause a decrease in height and or where accurate measurements are hard to obtain i.e. populations using wheelchairs (Sheard et al., 2011).

1.2 Malnutrition

Malnutrition has been associated with increased development of complications (for example, pressure ulcers, falls, poor immune function and wound healing), reduced organ function and mental state, increased health care costs, reduced quality of life, increased length of hospital stay and increased mortality (Norman, 2008; Saute et al., 2011).

For the purpose of this protocol, malnutrition refers solely to protein-energy malnutrition.

Protein-energy malnutrition (malnutrition) occurs when the intake of protein and energy is insufficient to meet the metabolic needs of the person. Vitamin and/or mineral deficiencies may also occur at this time or independently of protein-energy malnutrition.

The ICD-10-AM sixth edition (Tapsell, 2009) defines malnutrition in adults as:

- Severe (E43) – BMI < 18.5 kg/m² or unintentional weight loss of ≥ 10% with evidence of suboptimal intake resulting in severe loss of subcutaneous fat and/or severe muscle wasting
- Moderate (E44) – BMI < 18.5 kg/m² or unintentional weight loss of 5-9% with evidence of suboptimal intake resulting in moderate loss of subcutaneous fat and/or severe muscle wasting
- Mild (E44.1) – BMI < 18.5 kg/m² or unintentional weight loss of 5-9% with evidence of suboptimal intake resulting in mild loss of subcutaneous fat and/or severe muscle wasting

The ICD-10-AM sixth edition (Tapsell, 2009) defines malnutrition in children as:

- Severe – observed weight is at least three standard deviations below the mean value for the reference population
- Moderate – observed weight is at least two or more but less than three standard deviations below the mean value for the reference population
- Mild – observed weight is at least one or more but less than two standard deviations below the mean value for the reference population

Malnutrition prevalence in the community setting¹ is estimated at 10-30% and with higher rates in the acute care (20-50%), rehabilitation (30-50%) and residential aged cared settings (40-70%) (Tapsell, 2009). However, this has not been studied in the MJD or Indigenous populations.

The general goals of nutrition intervention in the MJD population should be to:

- Maintain the client's weight within a healthy weight range
- Optimise the quality of the client's diet by consuming foods from all food groups, and
- Maintain an oral intake (modified food diet for dysphagic clients) for as long as safely possible.
- Maintain fluid balance.
- Exercise.

¹ The community setting is defined as 'free living adults with or without assistance from community services' (Tapsell, 2009).

Health professionals should aim to identify the risk and development of malnutrition or weight loss as early as possible to ensure appropriate and timely interventions are implemented. Height and weight should be recorded when the person seeks confirmation of a diagnosis. Community support service providers for disability in-home support, basic community aged care service providers, dietitians, speech pathologists and family carers should be engaged throughout this process to ensure the foods and drinks are optimal.

1.3 Dysphagia

MJD clients may experience a range of factors which impact on their nutritional status, the most common of these being dysphagia (Riess, 2008; Saute et al., 2011). Dysphagia can lead to reduced oral intake, leading to malnutrition and weight loss. A study of 29 dysphagic head and neck cancer patients found that they experience 5% weight loss 45 days earlier than the 312 non-dysphagic head and neck cancer patients (Kubrak, 2010).

Social and psychological issues further complicate nutritional access, a survey of 360 non-MJD dysphagic patients found that 41% reported experiencing anxiety and panic during meals, 36% said they avoided having meals with others, 50% eat less and 33% stop eating even though they are still hungry (Ekberg et al. cited in Garcia and Chambers, 2010).

This reinforces the importance of enhancing the kilojoule content of food that dysphagic clients do eat.

Another risk factor for patients with dysphagia is dehydration (Chouinard et al., cited in Vivanti et al., 2009). A study of 25 dysphagia in-patients found that the mean daily fluid intake from food was greater than that from beverages ($p < .001$); and therefore, 'fluid dense foods, in contrast to thickened beverages, may present an important contribution to daily fluid intakes' (Vivanti et al., 2009). Dehydrating increases the risk of MJD patients developing bowel obstructions and compactions which can ultimately lead to premature death (see the *Continence Medical Protocol* for more information).

Refer to the Difficulty Swallowing Medical Protocol for further information on dysphagia.

1.4 Environmental factors

In addition to other medical conditions the client may be experiencing (e.g. pain, constipation, dental problems), psychosocial and environmental factors (income, housing, wet-season flooding, food access and security etc) also need to be considered.

1.4.1 Social and emotional wellbeing

Evidence shows that MJD patients have higher rates of depression, anxiety and apathy. Studies have found that MJD patients have higher rates of depression (33-63%) and one study found that 35% had moderate to severe depression (Cecchin et al., 2007; Kawai et al., 2004; McMurtray et al., 2006; Saute et al., 2010; Zawacki et al., 2002). The majority of studies

propose that depression in MJD patients is reactive to physical incapacities rather than neurological damage / or the disease itself. As stated above, 41% of dysphagic patients experience anxiety and panic when eating and drinking and 50% report eating less (Ekberg et al., cited in Garcia and Chambers, 2010).

Depression can lead to both undereating and overeating. The treating practitioner needs to be aware of which one the patient is doing and provide the most appropriate medication based on the current nutritional status of the patient and what their current eating pattern is (as well as other medical factors and lifestyle factors). Some antidepressant medications have side effects ranging from appetite suppression to weight gain. Discussions with the treating practitioner to ensure appropriate medications are prescribed is paramount. For example, an antidepressant with a side effect of weight gain is preferable to a side effect of appetite suppression; and similarly, a side effect of improved sleep duration is more desirable given the sleep disturbances experienced by MJD patients.

Refer to the Social and Emotional Wellbeing Medical Protocol and Sleep Disturbance Medical Protocol for further information.

2. Assessment of Nutrition

2.1 *Screening for malnutrition*

Malnutrition screening should be conducted on a regular basis to enable the early identification of clients who may be at risk of developing malnutrition. Height and weight should be recorded when the person seeks confirmation of a diagnosis. The tool selected for malnutrition screening should be a validated tool in the population group and should be quick and easy to use (Barker, 2011). It should be able to be conducted by any health professional or by the client himself or herself. Currently, there is no validated malnutrition screening tool in the MJD or Indigenous populations; however, the Malnutrition Universal Screening Tool (MUST) is a validated screening tool for use in the community setting (Tapsell, 2009) (see **Appendix A**). As well as undernutrition, the MUST can also identify overweight and obesity, which should also be managed to optimise health.

All MJD clients should be routinely screened for malnutrition, even if they initially were identified as at low risk for malnutrition or if they are identified as overweight or obese.

In addition to malnutrition screening, all MJD clients should have regular anthropometry (i.e. weight, height, BMI, waist circumference, mid-upper arm circumference) and biochemical (i.e. fasting lipids, fasting blood glucose level, albumin, urea, iron studies, vitamin/mineral tests) testing. Regular monitoring is important as the trend / change in results is often more telling than a once-off result. It is recommended that anthropometry testing is performed biannually (at a minimum, annually) and those who are malnourished receive both anthropometry and biochemical testing biannually until there are signs of nutritional stabilisation (i.e. back to goal weight and nutritional deficiencies corrected).

2.2 *Assessment for malnutrition*

All MJD clients identified as being at risk of developing malnutrition using the MUST should be referred to a health professional trained in malnutrition assessment for further assessment using a validated assessment tool, such as the Subjective Global Assessment (SGA). The SGA malnutrition assessment is more comprehensive, and includes weight history, diet history, symptoms, functional capacity, and a physical assessment (Barker, 2011). Again, there is no malnutrition assessment tool validated for exclusive use in either the MJD or Indigenous populations but the SGA is a validated assessment tool in the community setting (Tapsell, 2009).

The nutritional assessment used should include:

- Medical history, including any current and significant past symptoms and side-effects
- Social history, including an assessment of food security and ability to prepare safe and appropriate meals/drinks
- Anthropometry, including weight history and a physical assessment
- Biochemistry, including vitamin and mineral status if available
- Medications, including vitamin and/or mineral supplements
- Dentition
- Diet history, including supplements, textures and any recent changes to diet and
- Ability to self-feed or use enteral nutrition (or the ability of a carer, if applicable).

3. Guidelines for Managing Nutrition in Patients with MJD

All MJD clients should have an individualised nutrition care plan developed and be educated on suitable strategies to optimise their nutritional status. This should be reviewed, at a minimum, every six months. Dietitians should carefully consider all recommendations to ensure they are appropriate for the current social and physical situation of the client, especially if they are in remote communities where food security (the availability of food and one's access to it) can be a major contributor to inadequate and/or an inappropriate oral intake (Harrison, 2007). It is essential that education on the nutritional requirements of MJD patients, and modified foods, is provided to family carers and community services that prepare meals.

The management plan for clients experiencing nutritional deficiencies and/or weight loss should be constructed by a team of healthcare professionals, and where possible include a medical officer, a nurse, a speech pathologist, a dietitian, the client and the client's family/carer. Additional community services should also be accessed where possible (e.g., aged care and disability services). The assessment and management of clients with MJD should be reviewed annually to biannually as the disease progresses.

3.1 Clients identified as not at risk of malnutrition

MJD clients who have not been identified as at risk of malnutrition should be encouraged to eat according to [The Australian Guide to Healthy Eating](#)² and/or [The Aboriginal and Torres Strait Islander Healthy Food Guide](#) (see **Attachment B and C**) and aim to achieve a healthy body weight (i.e. within the healthy weight range for their age and height).

In dysphagic MJD clients who are not at risk of malnutrition, a speech pathologist should be consulted to ensure the client is consuming the most appropriate diet texture (if the client is suitable for oral intake). This will assist in minimising the risk of nutritional deterioration. Where clients are choosing not to adhere to the recommended texture modified diet, then consultation with speech pathology to determine alternate options to minimise aspiration risk, such as the use of compensatory techniques (e.g. smaller sips), should be undertaken.

3.2 Clients identified as at risk of malnutrition

Health professionals should consult the [Malnutrition Universal Screening Tool](#) (MUST) flow-chart (see **Attachment A**) which provides management guidelines based on the level of risk the client has been identified as.

For clients identified as at medium risk of malnutrition:

- Encourage increased amounts and/or frequency of oral intake
- Encourage foods and fluids which are high in energy and protein

² The Australian Government is currently consulting on a new guide and it is expected to be released in late-2012.

- Encourage food fortification (i.e. adding additional energy and/or protein into foods/drinks - adding extra milk powder into foods/drinks, adding extra margarine or oil into stews or vegetables, or using Novasource, Resource, Fortisip, Benecalorie) – it is important to avoid administering high energy and high protein supplements at meal times
- Encourage clients to maintain physical activity to minimise muscle mass loss
- Identify any factors which may be impacting on the client's ability to manage an adequate oral intake and support them and their carers correct these, and
- Monitor regularly.
- Inform and educate MJD clients of the possible future option with enteral nutrition support.

For clients identified as at high risk of malnutrition:

- Undertake management strategies as for medium risk
- Refer to local dietitian and
- Monitor more regularly, especially if the dietitian has assessed the patient as being malnourished.

3.3 Nutritional requirements

Whenever possible, MJD clients should receive an individualised nutrition care plan on diagnosis to minimise their risk of (premature) malnutrition. There is currently no specific nutritional requirements for MJD clients; however, as a guide (Queensland Health):

- Not malnourished
 - Energy – 100-125kJ/kg/day
 - Protein – 0.8-1.0g/kg/day
- Malnourished
 - Energy – 125-145kJ/kg/day
 - Protein – 1.2-1.5g/kg/day

It should also be ensured that MJD clients are receiving adequate fluids, fibre and micronutrients. If indicated, a liquid (or crushable tablet) vitamin supplement may be required; seek medical practitioner advice. If the client is following a texture-modified diet and/or requiring thickened fluids, particular attention should be paid to the adequate intake of energy, fluids, calcium and fibre which are known to be consumed at less than recommended amounts in this population (Cortier, 2008). Fibre and water intake are particularly importance given the incidence of bowel impaction and compaction in MJD patients (see the *Continence Medical Protocol* for more information).

3.4 Oral nutrition support

Oral nutritional supplements or fortifiers are milk- or juice-type drinks, puddings or powders that are high in energy and protein and often contain added vitamins and minerals. They can be used to supplement an inadequate oral intake. The drinks and puddings often come ready-to-consume and come in multiple flavours, while the powders also come in various flavours (including neutral), which can be mixed with milk or water to form a drink, or stirred through food. When providing oral nutritional supplements in drinks, remember to consider the fluid

consistencies which have been determined as safe for each individual to swallow. For example, if a person cannot manage thin fluids, then ensure that drinks with supplements are thickened appropriately, or supplements are added to thickened drinks to ensure safe fluid consistency is maintained.

Strategies to increase the consumption of oral nutritional supplements:

- Serve cold
- If too sweet, mix with water or milk
- Add other flavours to vanilla or neutral supplements (e.g. turn into a mango smoothie)
- Freeze juice-type supplements to make ice-blocks
- Consume between or after meals, not before
- Take smaller amounts more often (e.g. 60mL every few hours rather than a whole drink)
- Encourage safe swallow strategies – refer to *Difficulty Swallowing Medical Protocol and the [Safe feeding strategies for people with Machado Joseph Disease](#)* handbook.
- Ensure the physical environment promotes independent eating (for example, an upright posture, adequate table height, assistive cutlery and crockery, etc).

The MJD Foundation is currently trialling the following between meal supplements:

- Resource® Benecalorie®, a low-volume, kilojoule and protein supplement that provides 1,380 kilojoules and 7 grams of high-quality protein per 41 gram serving
- Novasource and
- Fortisip.

Results will be included in future updates of this protocol.

3.5 Dysphagic clients

In dysphagic MJD clients, a dietitian should work in conjunction with a speech pathologist and care providers to ensure the client is meeting their requirements through a texture modified diet and/or routes of non-oral feeding (see 3.5.1 Enteral nutrition support).

Different types of foods are easier to eat than others. Soft foods are easier to chew and swallow than hard foods and therefore less energy is required to eat them. Moister foods also stay together easier in the mouth and make it easier for the person to form a bolus and swallow easily. Different types of drinks are also easier to swallow than others. Thin liquids, like water, are harder to drink as they flow quickly and need more muscle control to ensure they do not go down the wrong way (which may lead to infections and aspiration pneumonia). Thicker liquids are easier to drink safely (i.e. minimal choking) if you have dysphagia because they flow more slowly, allowing the person to control them better in their mouth and have the additional time to prepare and protect their airway during the swallow (i.e. use safer swallowing strategies).

For more information on modified foods and fluids please see **Appendix C** and refer to, the *Difficulty Swallowing Medical Protocol* for more information on safer swallowing strategies.

Moister foods also promote fluid intake which will assist in preventing dehydration (Vivanti et al., 2009).

It must be remembered that people with MJD are often unable to prepare their own meals and/or physically feed themselves, and in remote communities they may not be able to access the food recommended by health professionals, and in urban setting may find the costs of purchasing palatable products prohibitive as they are frequently unable to work and likely to have lower incomes. Therefore, practical problem solving with the client and his or her family / service providers is imperative to ensure that recommendations are feasible. In some cases, you might have a higher compliance rate with the implementation of compensatory techniques rather than simply a textured modified diet. However only a speech pathologist should be responsible for determining what swallowing compensatory strategies are appropriate for any individual.

3.5.1 Enteral nutrition support

If a client has been assessed by a speech pathologist as having a degree of dysphagia which is causing the client to have an unsafe swallow or if the client is unable to maintain an adequate oral intake to maintain their weight, enteral nutrition support may be indicated. Enteral nutrition support may be in the form of nasogastric tube (usually short term) or a percutaneous endoscopic gastrostomy (usually long term). It is vital that thorough discussions with the patient and the family occur prior to enteral nutrition support, including information on quality of life.

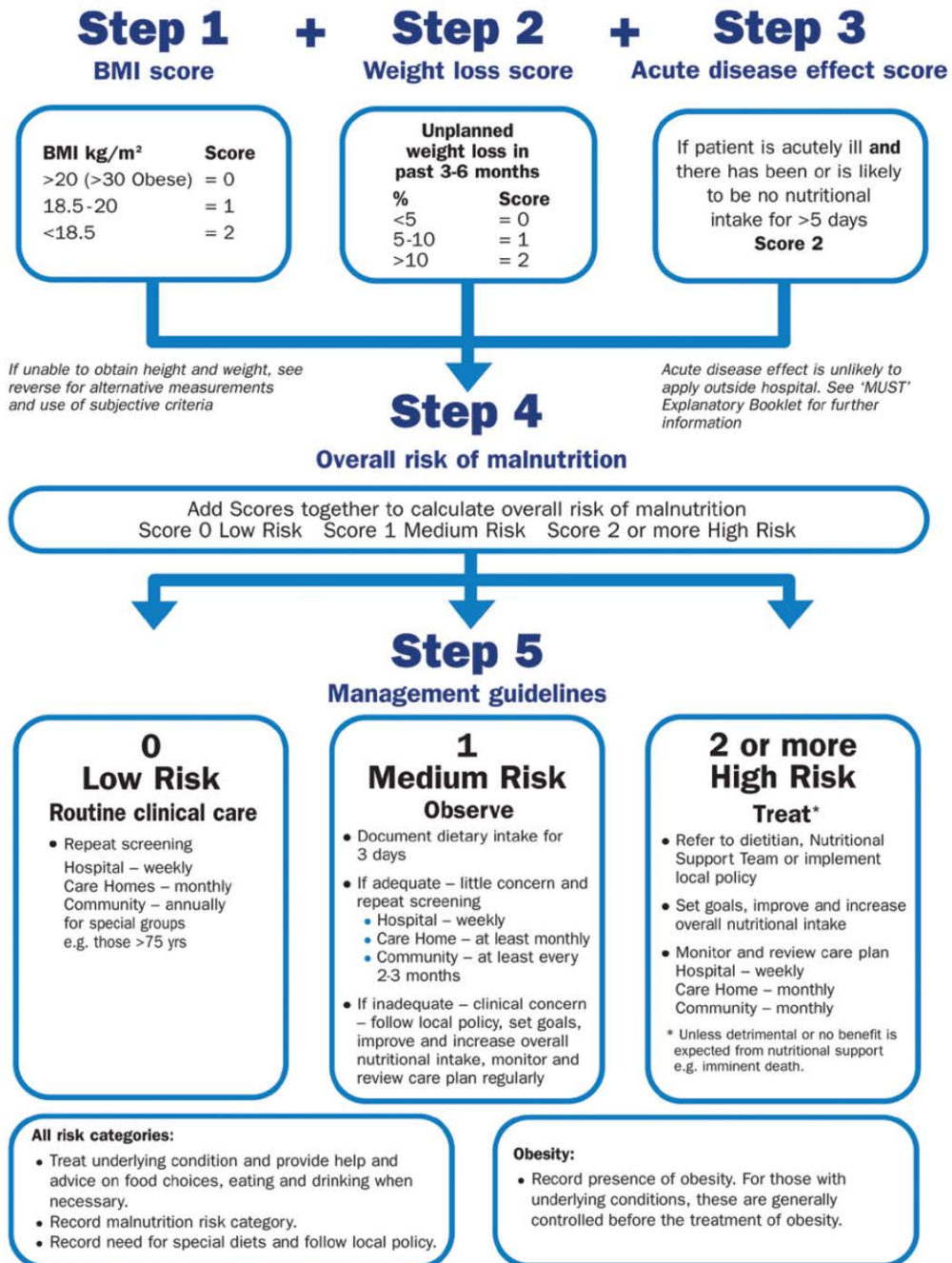
Nutrition from a nasogastric tube (NGT) requires that a tube be placed from the nose to pharynx, oesophagus and into the stomach. Enteral nutritional formula and water can then be fed via the tube. NGT insertion is usually a short term solution to non-oral feeding.

A percutaneous endoscopic gastrostomy (PEG) is generally used for people who have severe swallowing difficulties that are expected to continue long-term. A PEG tube is inserted directly into the person's stomach and therefore bypasses the structures used for swallowing and respiration. Enteral nutritional formula and water can then be fed via the tube.

All clients requiring enteral nutrition must be routinely assessed by a dietitian to ensure that they are tolerating their recommended regime and that the recommended regime is meeting their current nutritional requirements.

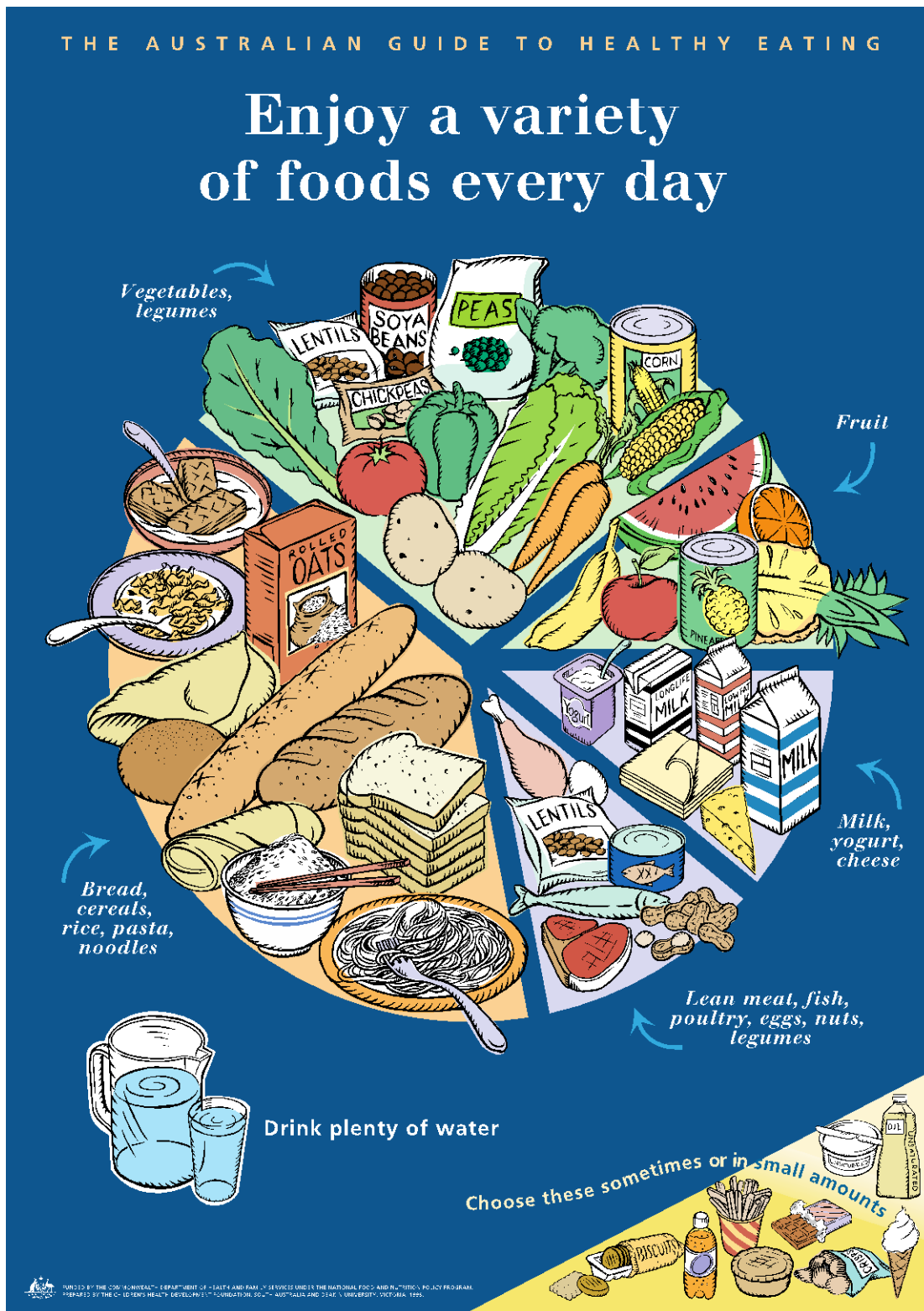
Clients requiring enteral nutrition support may be eligible for their local home enteral nutrition program in which they can access free or subsidised enteral formula and/or equipment. If they do not meet the eligibility criteria they will need to purchase formula and equipment themselves.

Appendix A – MUST flow chart



(Todorovic, Russell & Elia, 2011)

Appendix B – The Australian Guide to Healthy Eating



Appendix C – The Aboriginal and Torres Strait Islander Guide to Healthy Eating

DEPARTMENT OF HEALTH AND FAMILIES

THE ABORIGINAL AND TORRES STRAIT ISLANDER GUIDE TO HEALTHY EATING

Eat good food to be healthy and strong

Vegetables
(including legumes – baked beans, kidney beans & split peas).

Fruit

Milk, cheese and yoghurt

Meat
including bush meat, chicken, eggs & fish (including nuts & legumes – baked beans).

Breads, cereals, rice, spaghetti & noodles

Drink plenty of water

Eat in small amounts

ADAPTED WITH PERMISSION FROM THE AUSTRALIAN GUIDE TO HEALTHY EATING (1998, COMMONWEALTH DEPARTMENT OF HEALTH AND AGED CARE)

Northern Territory Government nt.gov.au/health

Appendix C – Modified Foods and Fluids

The Dietitians Association of Australia (DAA) and Speech Pathology Australia (SPA) have jointly developed the *Australian Standardised Terminology and Definitions for Texture Modified Foods and Fluids* as a consensus standard that health practitioners in Australia and New Zealand should use in the management of dysphagia (2007). For more information on the texture modifications and thickened fluids consult the [Safe feeding strategies for people with Machado Joseph Disease](#) handbook or [DAA](#) and [SPA](#).

Texture-modified Foods

Unmodified – Regular foods: These are everyday foods. Some are hard and crunchy others are naturally soft. By definition all food and textures can be included in this category (DAA & SPA, 2007).

Texture A – Soft: This type of texture consists of food that is naturally soft (e.g., ripe banana) or may be cooked, or cut to alter its texture. A soft diet should include foods that can be chewed but not necessarily bitten, and food that requires minimal cutting and can be broken up easily with a fork. The food in this category should be moist and where possible, served with extra sauce or gravy to increase the moisture content (DAA & SPA, 2007).

Texture B – Minced and Moist: Foods in this category should be soft and moist, easily mashed and should easily form into a ball. Individuals consuming food in this category should be able to use their tongue rather than their teeth to break the small lumps. The food may be presented as a thick puree with soft rounded lumps in it (DAA & SPA, 2007).

Texture C – Smooth Pureed: Foods in this category are smooth and lump free. The foods should be of pudding consistency and when positioned on a plate, should not “bleed” into one another. Smooth pureed foods should be cohesive enough to hold their shape on a spoon (DAA & SPA, 2007).

Thickened Fluids

Unmodified – Regular Fluids: Regular fluids do not have thickening agents added to them, however there are various thickness levels in unmodified fluids. Some are thinner (e.g., water) whilst some are thicker (e.g., fruit nectar). Regular fluids have a very fast flow and can be drunk through any type of cup or straw as appropriate (DAA & SPA, 2007).

Level 150 – Mildly Thick: Mildly thick fluids are thicker than naturally thick fluids such as fruit nectar but not as thick as a thickshake. These types of fluids have a steady, fast flow rate and pour quickly from a cup but at a slower rate than unmodified – regular fluids (DAA & SPA, 2007).

Level 400 – Moderately Thick: These types of fluids are similar to the thickness of a thickshake or of room temperature honey. Moderately thick fluids have a slow flow rate, and pour slowly from a cup. Though moderately thick fluids can be drunk directly from a cup, the best way to it is with a spoon (DAA & SPA, 2007).

Level 900 – Extremely Thick: Extremely thick fluids have the same thickness as a pudding or mousse. They do not have a flow rate and can hold their shape on a spoon. It is not possible to pour this type of fluid from a cup into the mouth and using a spoon is the preferred method of delivery (DAA & SPA, 2007).

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Appendix E – Definitions

Body Mass Index (BMI) is the weight of a person (in kilograms) divided by the square of the height of that person (in metres). It is used as an indicator of whether or not a person is over- or underweight (Concise Medical Dictionary: online).

CAG Repeat Length - MJD is associated with the expansion of a CAG trinucleotide repeat in a novel gene on 14q32.1. Normal chromosomes contain between 12 and 37 CAG repeats in the MJD gene, whereas MJD gene carriers have alleles within the expanded range of 62–84 CAG units. There is a strong correlation between the expanded repeat size and the age at onset of the disease as well as the clinical presentation (Maciel et al., 1995).

Dysphagia is condition a in which the action of swallowing is either difficult to perform or in which swallowed material seems to be held up in its passage to the stomach, often described by the patient as a sticking sensation (Concise Medical Dictionary: online).

Enteral nutrition formula are special purpose medical fluids containing carbohydrate, protein, fat, vitamins and minerals ± fibre

Fortified Foods and Drinks are achieved by adding substances to existing foods/drinks to enhance kilojoule context. For example, milk powder, butter, oils, Benecalorie etc.

Nasogastric Tube (NGT) is a tube be placed from the nose to pharynx, oesophagus and into the stomach. Liquefied food and fluids can then be inserted through this opening. NGT insertion is usually a short term solution to non-oral feeding.

Percutaneous Endoscopic Gastrostomy (PEG) is generally used for people who have severe swallowing difficulties that are expected to continue long-term. A PEG tube is inserted directly into the person's stomach and therefore bypasses the structures used for swallowing and respiration. .

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ⁱ *Nothing contained in this protocol constitutes medical or other advice and is only intended for use by qualified healthcare providers. Healthcare providers considering this protocol must use their own clinical judgment, knowledge and expertise when deciding whether it is appropriate to apply this protocol or other guidelines to any particular patient or treatment scenario.*

The recommendations set out in this protocol are a guide only and may not be appropriate for use in all situations or with all patients. The decision whether to adopt or not adopt any of the recommendations set out in this protocol must be made by each healthcare provider on a case-by-case basis.

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