MJD FOUNDATION

MEDICAL PROTOCOLS

Difficulty Swallowing

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1. Background – Difficulty Swallowing and MJD

1.1 Mechanism of Normal Swallowing

Swallowing is one of the most basic biological functions and on average adults swallow over 500 times per day (Zemlin, 1998). Swallowing is a complex process that can be voluntarily initiated but continues automatically once it has begun. Though swallowing can be controlled at a conscious level when eating and drinking, it is also controlled at a subconscious level (Martini, 2006; Zemlin, 1998). Swallowing requires input from a number of cranial nerves to activate the structures of the oral cavity, pharynx and larynx that are involved in swallowing (Zemlin, 1998). The key cranial nerves for swallowing are outlined in **Appendix A.**

The process of swallowing consists of four main stages. Illustrations of the four stages of swallowing and more detail are outlined in **Appendix B.** However, in summary these four stages include:

(1) the oral preparatory stage, in which food is chewed, broken down into smaller pieces, mixed with saliva and formed into a "bolus" (a food clump) ready for swallowing;
(2) the oral stage, where the food or liquid bolus is propelled toward the back of the throat by the tongue;
(3) the pharyngeal stage, where food travels down through the throat and into the opening of the upper oesophageus. In this stage a number of critical airway protection mechanisms also take place, including the closing of the airway at the level of the vocal folds and deflection of the epiglottis to cover and protect the airway entrance. These are necessary to ensure the food/fluid goes into the oesophageus and not into the airway. When this protection mechanism fails, coughing and choking may result. The velum also elevates to close off the nasal cavity and prevent food/fluid from going up into the nose; and finally
(4) the oesophageal stage, where food travels down the oesophageus and into the stomach.

1.2 Dysphagia in MJD

Dysphagia or difficulty swallowing can occur in up to 75% of people with MJD (Riess et al., 2008; Sequeiros & Coutinho, 1993) and traditionally dysphagia was believed to develop later in the disease progression once patients were no longer ambulatory (Sequeiros & Coutinho 1993; Sudarsky et al., 1992). More recent research however, has suggested that dysphagia may be present from the onset of the disease (Corrêa et al., 2010) and progress in parallel with the ataxic gait pattern (worsening with increased disease duration). Dysphagia is also closely associated with stage of disease, disease duration, and the CAG repeat length (Jardim et al., 2001; Kieling et al., 2008).

The presence of dysphagia represents a significant morbidity for people with MJD as dysphagia is associated with malnourishment, dehydration and aspiration, and can lead to negative social and psychological consequences (Ekberg et al., 2002; Garcia & Chambers, 2010; Martin, et al., 1994; Rüb et al., 2003). There is limited research in the literature to describe the nature of swallowing difficulties in people with MJD. The studies and conference abstracts that have been

published generally report a mild to severe dysphagia in participants, with the pharyngeal stage demonstrating more dysfunction than the oral stage (Corrêa, et al., 2010; Machado et al., 2009; Mourao et al., 2009; Wolf et al., 2007).

A case study on a male participant with MJD described the onset and the presentation of his dysphagia (Rüb et al., 2003). An inability to initiate the swallow mechanism and choking were the first swallowing difficulties reported. These difficulties occurred approximately 5 years after the onset of the participant's gait ataxia and balance disorder, which presented at 30 years of age (Rüb et al., 2003). In subsequent years, the participant showed decreased strength and speed of the facial, lingual, laryngeal and pharyngeal movements that led to pooling in the oral cavity and contributed significantly to his dysphagia. At this time, the decreased strength and ataxia of the jaw muscles meant the participant was no longer able to masticate (Rüb et al., 2003).

A recent study by staff from The University of Queensland (Professor Liz Ward and Professor Deborah Theodoros) examined swallowing function in 11 MJD Foundation clients and found varying levels of severity of dysphagia (slight to severe) consistent with previous studies. Considerable difficulties were noted in both the oral and the pharyngeal stages of the swallow. Most individuals needed softened foods to assist the oral stage of the swallow for solids. Thin liquid drinking (e.g. tea, water) revealed high incidence of coughing, suggesting poor airway protection in the pharyngeal stage of the swallow. Cough strength was also significantly impaired.

2. Assessment of Dysphagia in MJD

2.1 Purpose of Dysphagia Assessment

The main aims of dysphagia assessment are to determine the individual's ability to protect their airway and to determine the likelihood of safe oral intake. If an individual is determined to be safe for oral intake, the best conditions under which to eat and drink, and the best consistencies of food and fluid to be consumed must be considered. Assessment of dysphagia also aims to determine (a) the possible cause of the dysphagia; (b) the appropriate interventions for the remediation of the specific swallowing disorders identified; (c) the need for alternative nutritional management; (d) the need for further diagnostic studies; and (e) the patient's clinical baseline to allow monitoring of change in the future.

2.2 Dysphagia Assessment of Clients from East Arnhem Land

The MJD Foundation is working with the University of Queensland (Professor Liz Ward, Professor Deborah Theodoros and Associate Professor Trevor Russell) to conduct assessments and devise management plans to minimise the impacts and risks associated with dysphagia for people with MJD. The University of Queensland have screened MJD clients for aspiration risk using a clinical swallow assessment incorporating the Water Swallow Test (WST). The results and recommendations from these assessments have informed the content of this protocol.

2.3 Tools Used to Assess Dysphagia

Dysphagia is assessed by a combination of both clinical swallowing assessments and instrumental assessment methods. Brief details of these assessment processes have been outlined below and the rationale for using each. In addition to the assessments there are a number of outcome measures which are used to record swallowing outcomes over time.

2.3.1 The Water Swallow Test (WST) screening assessment: This screening assessment can be performed by any health professional. The WST is a useful assessment for screening individuals for evidence of aspiration and to determine whether further diagnostic examination is required (Depippo, Holas & Reding, 1992). To complete the WST, have the patient drink 90ml of water from a cup without interruption. Coughing, choking or a wet hoarse vocal quality exhibited during or within 1 minute of completing the test, failure to complete the task or showing greater than a 2% shift in pulse oximetry from the baseline level is considered a fail. If the patient fails the WST, they are required to undergo the more detailed Clinical Swallow Evaluation (CSE). Clients who fail the WST should be referred for a full assessment by a speech pathologist.

2.3.2 Clinical Swallow Evaluation (CSE): The CSE should only be performed by speech pathologists. The CSE has three main components: a medical history, an oromotor examination, and swallow trials. The medical history compiles information from the physicians and nursing staff, and from the client and the client's family. The oromotor examination involves inspection of the oral cavity, dentition, and the strength, symmetry, speed, and range of movement of the facial muscles (CNVII), jaw (CNV), tongue (CNXII), palate (CNIX and CNX), voice (CNX), and dry swallow. The swallow trials incorporate multiple trials of food and fluid consistencies (+/- compensatory strategies). Signs of inability to manage (e.g. rapid onset of fatigue), aspiration risk (coughing, choking) and need for feeding assistance (help holding cup, able only to hold a spoon) are monitored to determine the safest oral intake for each individual, and if any supplementary nutrition may be required (necessitating referral to a dietician). There are Australian standards regarding the classification of foods and fluids provided to dysphagic patients. There are four levels of fluids (from thin to thickest) and four levels of foods (from normal to heavily modified texture). Details of these have been provided in **Appendix C**.

2.3.3 Modified Barium Swallow (MBS): A MBS can only be performed and analysed by a qualified speech pathologist. The MBS, also referred to as a videofluoroscopic swallow study (VFSS), is considered the gold standard in dysphagia diagnosis (Owens, Metz & Hass, 2007). The MBS is an X-ray procedure that allows the clinician to observe the movements in the various stages of swallowing – particularly the pharyngeal stage. This procedure is the most accurate method for detecting aspiration and what is causing it, and it also informs the clinician about what aspects of the swallow are impaired. Different textures of food and fluids are coated with or mixed with barium. Barium is a white chalky substance that can be seen on an X-ray and therefore the clinician can observe the patient's swallowing process in real time. The X-ray views are recorded for later analysis by the speech pathologist. This assessment can only be conducted in a hospital setting with a videofluroscopic suite.

2.3.4 Fibre-Optic Endoscopic Evaluation of Swallowing (FEES): FEES can only be analysed by a qualified speech pathologist and the assessment must be performed in conjunction with a medical officer or ENT who can insert the endoscopic scope. FEES assessment allows for direct observation of the oropharynx and larynx during swallowing. A flexible fibre-optic scope is inserted into the nose and inserted into the pharynx. It allows the speech pathologist to observe the structure and function of the soft palate, base of tongue, oropharynx, hypopharynx, larynx and subglottis, and determine the presence of penetration/aspiration. The oral and oesophageal stages of swallowing cannot be observed using FEES. However it does not involve exposure to radiation and can be conducted at the patient's bedside. This procedure is an accurate method for detecting aspiration and what is causing it, and it also informs the clinician about what aspects of the swallow are impaired.

2.3.5 Dysphagia Outcome and Severity Scale (DOSS): The DOSS (O'Neil et al., 1999) is an easy to use scale developed to rate the functional severity of dysphagia based on an objective assessment of swallowing. The DOSS allows the speech pathologist to rate a patient's swallowing function in terms of severity on a 7 point scale (Level 1 – Level 7) where Level 1 represents a severe dysphagia and Level 7 represents a patient who is normal in all situations. The DOSS ratings should be completed by the assessing speech pathologist at the time of assessment. The DOSS has been provided in **Appendix D.**

2.3.6 Australian Therapy Outcome Measures Dysphagia Scale (AusTOMs): The AusTOMs (Perry & Skeat, 2004) scales for speech pathology were developed so that clinicians could rate their patients functioning in terms of their disability (in this case dysphagia). The AusTOMs scales use four different domains to assess swallowing status. These domains include Impairment, Activity Limitation, Participation Restriction, and Distress/Wellbeing. Each domain of the AusTOMs is rated on a six point scale (0 – 5) where 0 (zero) represents a "complete difficulty" and 5 represents "no difficulty" (Perry & Skeat, 2004). The AusTOMs ratings should be completed by the assessing speech pathologist at the time of assessment. The AusTOMS has been provided in **Appendix E.**

3. Guidelines for Managing Dysphagia in Patients with MJD

3.1 Purpose of Dysphagia Management

Clients with MJD who exhibit difficulty swallowing are likely to present a considerable challenge to speech pathologists and health care professionals due to the complexity of the dysphagia, the other symptoms associated with MJD and the challenges associated with providing health care to remote communities.

The management plan for clients with dysphagia should be constructed by a team of healthcare professionals, and where possible include a medical officer, a nurse, a speech pathologist, a dietician, the client and the client's family/carer. Additional community services should also be accessed where possible (e.g., Community Service Provider). The assessment and management of clients with MJD should be reviewed regularly as the disease progresses. Dysphagia management techniques can be broken into two groups: compensatory techniques and rehabilitative techniques.

3.2 Compensatory Techniques

3.2.1 Positioning: All clients should be positioned to optimise swallowing. During mealtimes clients should be seated upright in a comfortable position at 90 degrees where possible. The head, neck and shoulders should be in line and the chin should be tilted slightly downwards. This helps to protect the airway and ensure that the food/fluid goes down the right way. After the client has finished their meal they should remain sitting upright for at least 20-30 minutes. For more information on positioning please see the *Safe feeding strategies for people with Machado Joseph Disease* handbook.

3.2.2 Delivery: For some clients, altering the mode of delivery can overcome some specific difficulties. There are a number of utensils that are available that have been specifically modified for people with swallowing difficulties. It should be noted however, that while it is important to know that these type of utensils exist they do not have to be used with every client. For more information on the modified utensils please refer to the *Safe feeding strategies for people with Machado Joseph Disease* handbook.

3.2.3. Modified Foods and Fluids: During the CSE, various types of foods and fluids would have been presented to the client to determine their safety on the different textures and consistencies. 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. 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 the airway during the swallow. For more information on modified foods and fluids please see

Appendix C and the *Safe feeding strategies for people with Machado Joseph Disease* handbook.

It must be remembered that people with MJD in the Northern Territory are often unable to prepare their own meals, or even access the food recommended by their treating speech pathologist. Practical problem solving with the client and his/her family is imperative to ensure that recommendations are feasible.

3.3 Rehabilitation Techniques

Although there are a number of rehabilitation techniques which can be used with people with dysphagia, the following are those with most relevance for targeting the swallowing issues of clients with MJD.

3.3.1 Controlled Swallow: The controlled swallow is used for clients who have difficulties with bolus containment in the mouth and swallow initiation. This technique provides conscious control of the bolus in the mouth, volitional airway protection and conscious initiation of the swallow. To perform this technique the client should be instructed to have overt conscious control over each mouthful:

1. Control the bolus in the mouth
2. Commence the swallow at will

3.3.2 Supraglottic Swallow: The use of this technique is designed to improve airway closure before and during the swallow at the level of the glottis (Logemann, 1993; Martin et al., 1993). The client should be instructed to:

1. Breathe in and hold your breath (at the larynx – vocal fold adduction must be achieved)
2. Put a small amount of food/fluid in your mouth
3. Swallow
4. Cough on exhalation post swallow (to clear any residue in the throat)

3.3.3 Super Supraglottic Swallow: The super supraglottic swallow is similar to the supraglottic swallow but in comparison it is designed to improve airway closure before and during the swallow at the level of the airway entrance and glottis (Logemann, 1993; Martin et al., 1993). The client should be instructed to:

1. Breathe in and hold your breath very tightly and “bear down” (closes airway at both true and false vocal fold levels)
2. Put a small amount of food/fluid in your mouth
3. Swallow
4. Cough forcefully on exhalation (to clear any residue in the throat)

3.4 Non-Oral Feeding

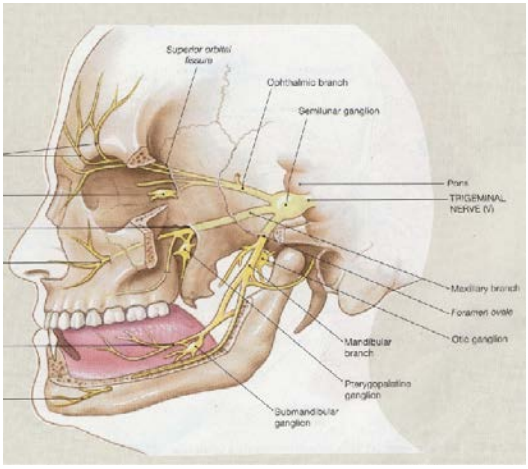
Patients who are determined to be at high-risk of aspiration and cannot maintain adequate hydration and nutrition orally, will likely require some non-oral feeding. Both determining the need, and what type of non-oral feeding is a decision made between the client, family, medical staff and a dietician.

There are several approaches that can be utilised in non-oral feeding. Nutrition from a nasogastic tube (NGT) requires that a tube be placed from the nose to pharynx, oesophagus and stomach. Enteral formula, water and crushed or liquid medications can then be inserted through this opening. 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 formula, water and crushed or liquid medications can then be inserted through this tube.

Appendix A – Cranial Nerves for Swallowing

Trigeminal (CNV)

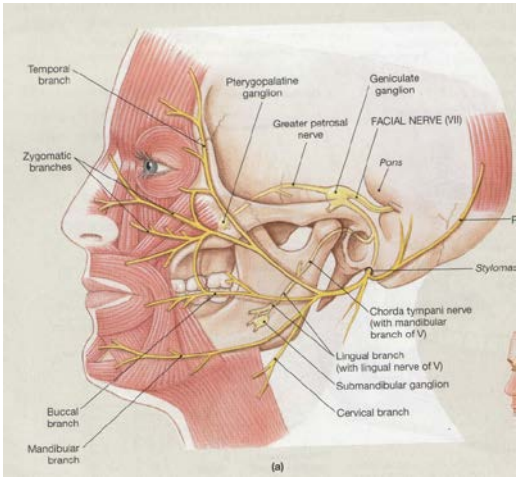
Sensory: From nasal mucosa, tongue, teeth, gums, palate, lip, check and proprioception of the muscles of mastication
Motor: Muscles of mastication



The Trigeminal Nerve (Martini, 2006, p 483)

Facial (CNVII)

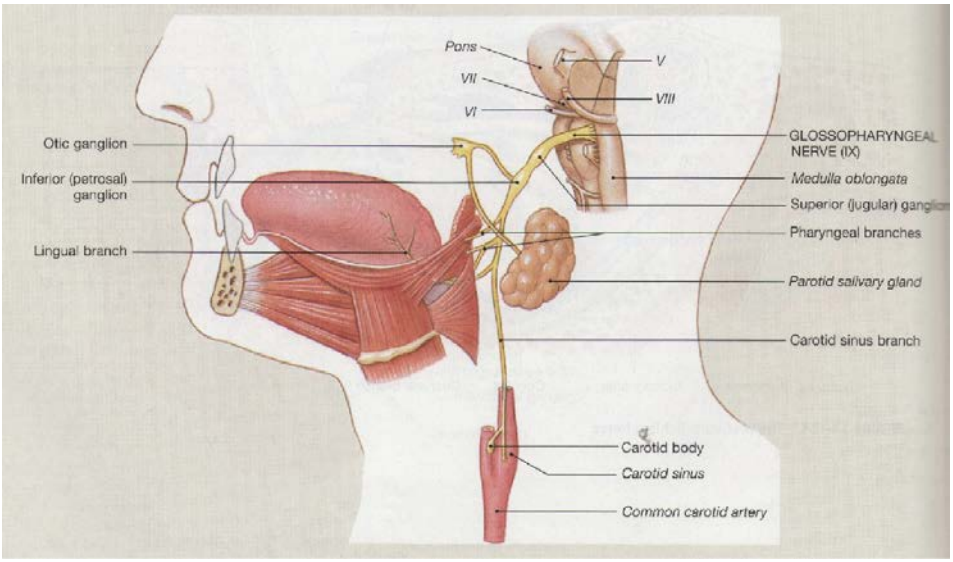
Sensory: Tastebuds of anterior ⅔ of tongue, nasal and palatal sensation and proprioception of the muscles of facial expression
Motor: Muscles of facial expression, secretion of salvia (submandibular, sublingual)



The Facial Nerve (Martini, 2006, p 484)

Glossopharyngeal (CNIX)

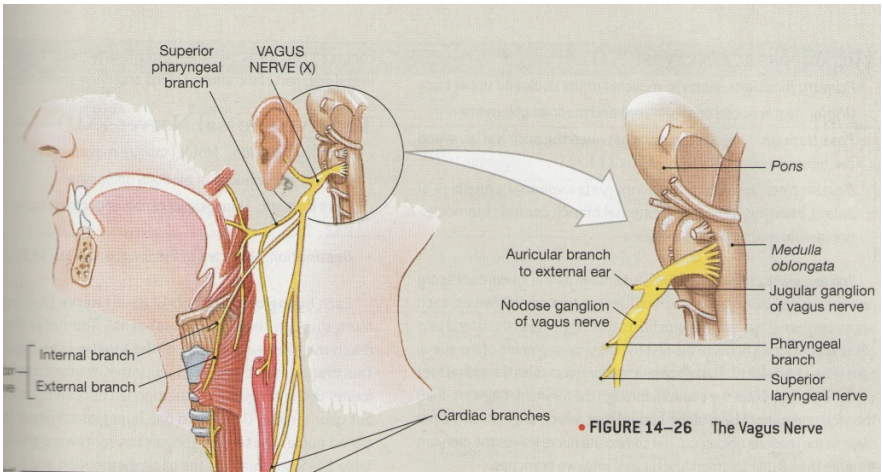
Sensory: Tastebuds on posterior ⅓ of tongue, sensation from pharynx and proprioception of muscles of the pharynx
Motor: Motor impulses to muscles of pharynx used in swallowing, secretion of salvia (from parotid gland)



The Glossopharyngeal Nerve (Martini, 2006, p 486)

Vagas (CNX)

Sensory: Tastebuds on posterior portion of tongue and proprioception of pharynx and larynx
Motor: Contraction of the muscles of the pharynx and larynx



The Vagus Nerve (Martini, 2006, p 487)

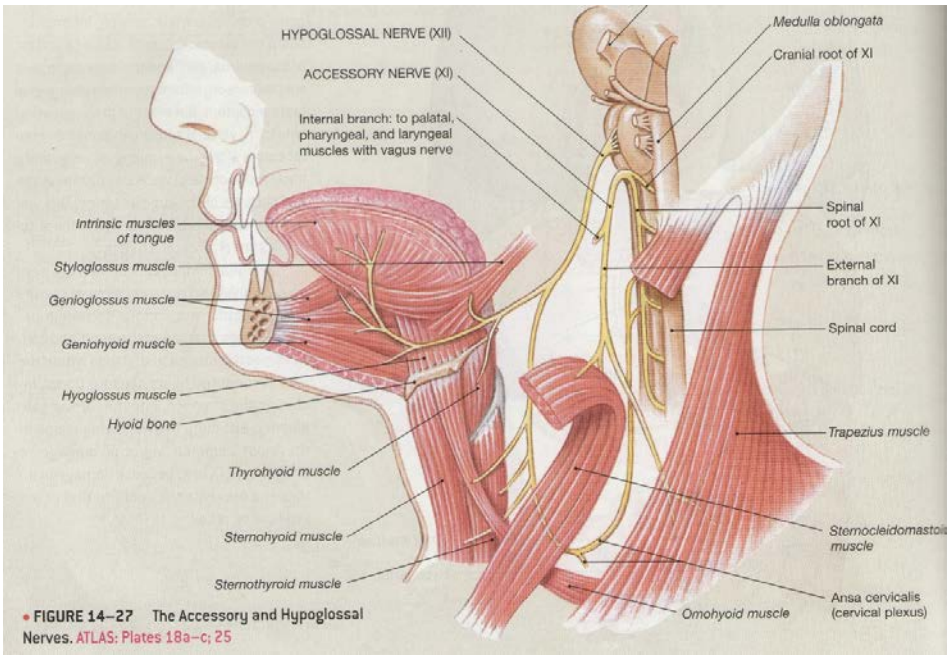
Accessory (CNXI)

Sensory: Proprioception for muscles controlling the head and neck
Motor: Laryngeal movement and muscles for movement of head and neck

(see image below)

Hypoglossal (CNXII)

Sensory: Proprioception of muscles of the tongue
Motor: Motor control of intrinsic and extrinsic muscles of tongue and infrahyoid muscle



The Accessory Nerve and the Hypoglossal Nerve (Martini, 2006, p 48)

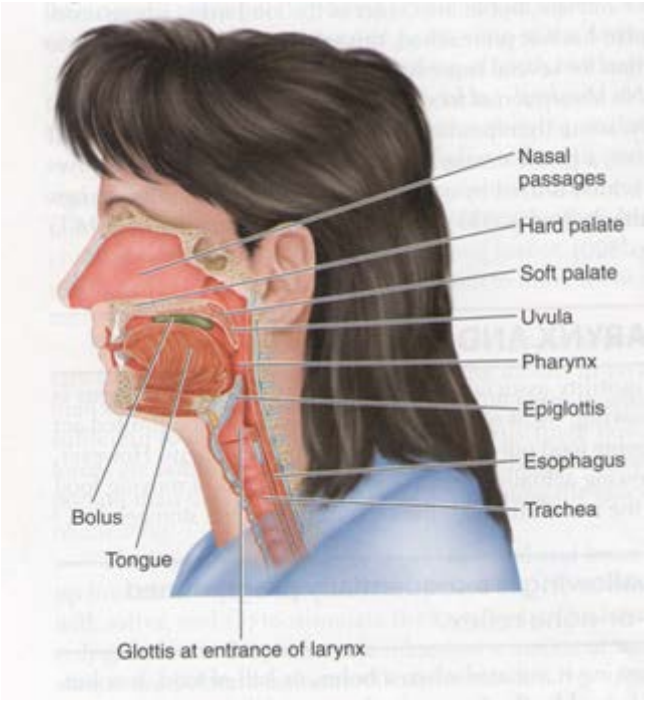
Appendix B – The Stages of Swallowing

Stage 1: The Oral Preparatory Stage

The oral preparatory stage of swallowing involves the manipulation of food or liquid to form a cohesive bolus for swallowing. When drinking liquids the tongue momentarily contains the liquid bolus in one of two positions: 1) the tongue may form a cupped position so that the fluid is held in the midline of the tongue and the hard palate with the tongue tip elevated; or 2) the liquid bolus may be held in the floor of the mouth in front of the tongue (only 20% of the population do this). In the oral preparatory stage for solids, the tongue and cheeks move the food to the molars for chewing and mixing with saliva to form a bolus. When active chewing has ceased, the soft palate is pulled down and forward sealing off the oral cavity from the pharynx, however during active chewing the soft palate is not down and forward, and premature spillage is common and entirely normal.

Stage 2: The Oral Stage

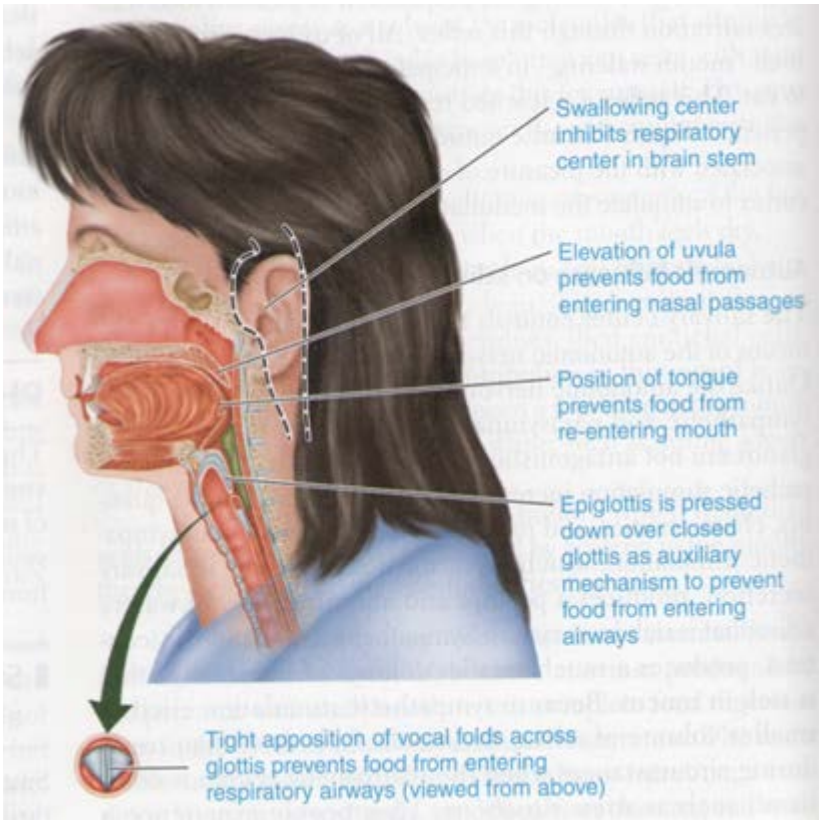
The oral stage begins after the bolus has been formed. This stage consists of the propulsion of the bolus by the tongue from the front to the back of the mouth. Once the bolus reaches the anterior faucial arch at the back of the mouth the swallow is triggered. This stage occurs in approximately 1-1.5 seconds or less.



Oral stage of swallowing (Sherwood, 2004, p 602)

Stage 3: The Pharyngeal Stage

The commencement of the pharyngeal stage is recognised as when the "head" of the bolus passes at any point between the anterior faucial arches and when the tongue base crosses the lower rim of the mandible. The soft palate elevates and retracts to meet the rear wall of the pharynx and to prevent the bolus from going into the nasal cavity. The tongue base and the pharyngeal wall move toward each to create pressure behind the tail of the bolus. While this is occurring the hyoid bone rises and moves the larynx up and forward. The closure of the larynx prevents the bolus from entering the trachea by adducting the true and false vocal folds, medialising and tilting the arytenoids anteriorly and deflecting the epiglottis over the laryngeal entrance and at this point the bolus is directed around the larynx. The pharyngeal constrictor muscles contract in a wave from the top down to continue the downward direction of the bolus to the upper oesophageal sphincter (UES). The UES opens to allow the bolus to pass into the oesophagus. The pharyngeal stage occurs in approximately 1 second.



Pharyngeal stage of swallowing (Sherwood, 2004, p 602)

Stage 4: The Oesophageal Stage

The oesophageal stage commences when the bolus moves through the UES and into the oesophagus. The muscles of the oesophagus transport the bolus using peristaltic waves which push the bolus downwards. The oesophageal stage terminates when the bolus reaches the lower oesophageal sphincter at the opening of the stomach. The bolus typically takes 8-10 seconds to move through the oesophagus.

Appendix C – Modified Foods and Fluids

The Dieticians 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.

Extract from DAA and SPA (2007):

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.

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.

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.

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.

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.

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 poor quickly from a cup but at a slower rate than unmodified – regular fluids.

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 poor slowly from a cup. Though moderately thick fluids can be drunk directly from a cup, the best way to it is with a spoon.

Level 900 – Extremely Thick: Extremely thick fluids have the same thickness as a pudding or mousse. They do no have a flow rate and can hold their shape on a spoon. It is not possible to poor this type of fluid from a cup into the mouth and using a spoon is the preferred method of delivery.

Appendix D – Dysphagia outcome and severity scale

Full per-oral nutrition (P.O): Normal diet

Level 7: Normal in all situations
Normal diet
No strategies or extra time needed

Level 6: Within functional limits/modified independence
Normal diet, functional swallow
Patient may have mild oral or pharyngeal delay, retention or trace epiglottal undercoating but independently and spontaneously compensates/clears
May need extra time for meal
Have no aspiration or penetration across consistencies

Full P.O: Modified diet and/or independence

Level 5: Mild dysphagia: Distant supervision, may need one diet consistency restricted
May exhibit one or more of the following
Aspiration of thin liquids only but with strong reflexive cough to clear completely
Airway penetration midway to cords with one or more consistency or to cords with one consistency but clears spontaneously
Retention in pharynx that is cleared spontaneously
Mild oral dysphagia with reduced mastication and/or oral retention that is cleared spontaneously

Level 4: Mild–moderate dysphagia: Intermittent supervision/cueing, one or two consistencies restricted
May exhibit one or more of the following
Retention in pharynx cleared with cue
Retention in the oral cavity that is cleared with cue
Aspiration with one consistency, with weak or no reflexive cough
Or airway penetration to the level of the vocal cords with cough with two consistencies
Or airway penetration to the level of the vocal cords without cough with one consistency

Level 3: Moderate dysphagia: Total assist, supervision, or strategies, two or more diet consistencies restricted
May exhibit one or more of the following
Moderate retention in pharynx, cleared with cue
Moderate retention in oral cavity, cleared with cue
Airway penetration to the level of the vocal cords without cough with two or more consistencies
Or aspiration with two consistencies, with weak or no reflexive cough
Or aspiration with one consistency, no cough and airway penetration to cords with one, no cough

Nonoral nutrition necessary

Level 2: Moderately severe dysphagia: Maximum assistance or use of strategies with partial P.O. only (tolerates at least one consistency safely with total use of strategies)
May exhibit one or more of the following
Severe retention in pharynx, unable to clear or needs multiple cues
Severe oral stage bolus loss or retention, unable to clear or needs multiple cues
Aspiration with two or more consistencies, no reflexive cough, weak volitional cough
Or aspiration with one or more consistency, no cough and airway penetration to cords with one or more consistency, no cough

Level 1: Severe dysphagia: NPO: Unable to tolerate any P.O. safely
May exhibit one or more of the following
Severe retention in pharynx, unable to clear
Severe oral stage bolus loss or retention, unable to clear
Silent aspiration with two or more consistencies, nonfunctional volitional cough
Or unable to achieve swallow

Extract from O'Neil et al (1999)

Appendix E – AusTOMS scale

The ‘‘Swallowing’’ scale incorporates all disorders of the structure and/or function of the swallowing mechanism, and/or feeding in both adults and children. Issues of safety and alternative feeding may be rated under the ‘‘Activity Imitation’’ domain.

Impairment of either Structure or Function (as appropriate to age):
Impairments are problems in body structure (anatomical) or junction (physiological) as a significant deviation or loss.

0 Profound swallowing/feeding impairment: No swallow/suck initiated. Difficulty in opening mouth. No functional movement of oral and/or pharyngeal structures or musculature.

1 Severe swallowing/feeding impairment: Swallow/suck initiated but is inconsistent/very delayed/severely disordered. Severe oral and/or pharyngeal impairment with no control of bolus.

2 Moderate/severe swallowing/feeding impairment: Swallow/suck initiated but may be inconsistent/delayed/disordered. Moderate oral and/or pharyngeal impairment with poor control of bolus (eg pharyngeal leakage/spillage).

3 Moderate swallowing/feeding impairment: Swallow/suck initiated consistently but delayed/ abnormal. Moderate oral and/or pharyngeal impairment with limited bolus control (e.g., pooling, residue post swallow, buccal pocketing).

4 Mild swallowing/feeding impairment: Swallow/suck initiated consistently with appropriate timing and coordination. Mild oral and/or pharyngeal impairment with mild difficulties in bolus control (e.g., mild pooling or delay).

5 No swallowing/feeding impairment : Swallow/suck initiated consistently with appropriate timing and coordination and full control of bolus.

Activity Limitation (as appropriate to age):
Activity limitation results from the difficulty in the performance of an activity. Activity is the execution of a task by an individual.

0 Unable to manage own secretions (if tracheostomy is in situ , it is cuffed). Unable to safely manage any oral intake. Requires full alternative (e.g., PEG, NGT) nutrition.

1 Can sometimes manage own secretions with prompts (if cuffed tracheostomy is in situ , may be deflated). May sometimes safely take small practice amounts of modified consistencies. Requires mainly alternative (e.g., PEG, NGT) or supplementary nutrition.

2 Can manage own secretions safely and independently (if tracheostomy is in situ , it may be uncuffed). Safe on a limited range of consistencies, requires strategies and full supervision to manage oral intake. Some alternative/supplementary feeding (e.g., NGT) required.

3 Can manage a diet of modified consistencies. Some supervision/strategies may be required. No alternative or supplementary feeding required.

4 Can manage most consistencies, may require some restrictions in range of consistencies. Independent using strategies, with no supervision required. May eat/drink slowly.

5 Can manage a full diet in a timely manner and independently.

Participation Restriction (as appropriate to age):
Participation restrictions are difficulties the individual may have in the manner or extent of involvement in their life situation. Clinicians should ask themselves: ''Given their problem, is the individual experiencing disadvantage?''

0 Unable to fulfill social, work, educational, or family roles. No social integration. No involvement in decision-making. No control over environment. Unable to reach potential in any situation.

1 Severe difficulties in fulfilling social, work, educational, or family roles. Very limited social integration. Very limited involvement in decision-making. Very little control over environment. Can only rarely reach potential with maximum assistance.

2 Moderately severe difficulties in fulfilling social, work, educational, or family roles. Limited social integration. Limited involvement in decision-making. Control over environment in one setting only. Usually reaches potential with maximum assistance.

3 Moderate difficulties in fulfilling social, work, educational, or family roles. Relies on moderate assistance for social integration. Limited involvement in decision-making. Control over environment in more than one setting. Always reaches potential with maximum assistance and sometimes reaches potential without assistance.

4 Mild difficulties in fulfilling social, work, education, or family roles. Needs little assistance for social integration and decision-making. Control over environment in more than one setting. Reaches potential with little assistance.

5 No difficulties in fulfilling social, work, educational, or family roles. No assistance required for social integration or decision-making. Control over environment in all settings. Reaches potential with no assistance.

Distress/Wellbeing (as appropriate to age):
The level of concern experienced by the individual. Concern may be evidenced by anger, frustration, apathy, depression, etc.

0 High and consistent levels of distress or concern.

1 Severe concern, becomes distressed or concerned easily. Requires constant reassurance. Loses emotional control easily.

2 Moderately severe concern. Frequent emotional encouragement and reassurance required.

3 Moderate concern. May be able to manage emotions at times, although may require some encouragement.

4 Mild concern. Able to manage emotions in most situations. Occasional emotional support or encouragement needed.

5 Able to cope with most situations. Accepts and understands own limitations.

Appendix F – Contributors to the Development of this Protocol

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Appendix G – Definitions

Aspiration food or fluid entering the airway and/or lungs during swallowing.

Aspiration pneumonia is the inflammation of the lungs and airways to the lungs (bronchial tubes) from breathing in foreign material (usually food, liquids, vomit, or fluids from the mouth). This may lead to lung abscess, swelling and inflammation of the lung and/or a lung infection. Aspiration pneumonia can cause death in MJD patients.

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).

Larynx is the organ responsible for the production of vocal sounds, also serving as an air passage conveying air from the pharynx to the lungs. It is situated in the front of the neck, above the trachea. It is made up of a framework of nine cartilages– the epiglottis, thyroid, cricoid, arytenoid (two), corniculate (two), and cuneiform (two) – bound together by ligaments and muscles and lined with mucous membrane. Within are a pair of vocal folds, which function in the production of voice (Concise Medical Dictionary: online).

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

Oesophagus, also known as the gullet, is a muscular tube, about 23 cm long, that extends from the pharynx to the stomach. It is lined with mucous membrane, whose secretions lubricate food as it passes from the mouth to the stomach. Waves of peristalsis assist the passage of food (Concise Medical Dictionary: online).

Pharynx is the muscular tube, lined with mucous membrane, that extends from the beginning of the oesophagus (gullet) up to the base of the skull. It is divided into the nasopharynx, oropharynx, and hypopharynx and it communicates with the posterior nares, Eustachian tube, the mouth, larynx, and oesophagus. The pharynx acts as a passageway for food from the mouth to the oesophagus, and as an air passage from the nasal cavity and mouth to the larynx. It also acts as a resonating chamber for the sounds produced in the larynx (Concise Medical Dictionary: online).

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. Liquefied food and fluids can then be inserted through this tube.

Appendix H – References

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