SWALLOWING DIFFICULTIES
HELP SHEET

MJD FOUNDATION

This Help Sheet explains how Machado Joseph Disease (MJD) affects swallowing and provides information about appropriate support. This information is intended as a general overview and it is important to remember that each person with MJD needs to be assessed and supported according to individual need.

Normal swallowing

Swallowing is one of the most basic biological functions. On average, adults swallow over 500 times a day. Swallowing is a complex process that can be started voluntarily but continues automatically once it has begun. Swallowing involves a number of cranial nerves and consists of four main stages:

1. Oral preparatory stage. Food is chewed, broken down into smaller pieces, mixed with saliva and formed into a clump ready for swallowing.
2. Oral stage. Food or liquid is propelled towards the back of the throat by the tongue.
3. Pharyngeal stage. Food or liquid travels down through the throat and into the opening of the upper oesophagus. Airway and nasal cavity protection is critical at this stage to ensure food or fluid goes into the oesophagus and not down into the airway or up into the nose. If these protection mechanisms fail, coughing, spluttering and choking may result.

Swallowing difficulties in mjd

Dysphagia is the medical term for difficulty swallowing. Dysphagia can occur in up to 75% of people with MJD and may be present from the beginning of the disease. The symptoms worsen as the disease progresses.

People with MJD who experience difficulties with swallowing face significant health challenges. If a person has difficulty swallowing, they are at risk of malnourishment, dehydration and aspiration of food or fluids into their lungs. Social and psychological effects may develop as the person experiences embarrassment while trying to eat and drink with other people around, or if they are fearful of choking or 'making a mess'.

Case studies

1. A man with MJD described the onset and presentation of his dysphagia. He first noticed an inability to start swallowing and, once swallowing had commenced, he experienced choking. These difficulties occurred approximately five years after his mobility and balance symptoms were first noticed. Later, the man showed decreased strength and speed of facial, mouth and throat movements. This led to pooling of food in his mouth and made the swallowing difficulty worse. As strength and coordination of the jaw muscles deteriorated, he was no longer able to chew.

2. A study of 11 people with MJD by University of Queensland speech pathologists found slight to severe levels of dysphagia. Considerable difficulties were noted in both the oral (mouth) and pharyngeal (throat) stages of the swallow for solids. Thin liquids (e.g. tea, water) were linked with high levels of coughing, suggesting poor airway protection. Cough strength was also significantly impaired.

Assessment: Speech Pathologist

Assessment of swallowing difficulties is undertaken by a speech pathologist. The main aims of assessment are to determine:

- an individual's ability to protect their airway
- the likelihood of eating safely via the mouth
- the best conditions for eating and drinking safely
- the best consistencies of food and fluid to be consumed safely.

Radiologocial investigations may form part of a swallowing assessment (for example a Modified Barium Swallow) which will be organised by the Speech Pathologist and/or GP.

Management

Management plans for people with MJD who have swallowing difficulties are developed by a team of health professionals working with the individual and their carer/s. Health professionals may include a medical officer, a nurse, a speech pathologist and a dietician. Assessment and

management of symptoms should be reviewed regularly as the disease progresses. The input of the person with MJD and the observations of their carer/s are an important part of the review process.

Dysphagia management

1. Compensatory techniques
    - Positioning
    - Delivery and utensils
    - Modified food and fluids

For detailed information on positioning, delivery and modified food and fluids, please refer to the MJD Handbook, 'Safe Feeding Strategies for People with Machado Joseph Disease' (see Resources page of MJDF website).

2. Rehabilitation techniques

There are a number of rehabilitation techniques that may assist people with MJD to swallow more effectively and safely. Assessment and guidance by a speech pathologist are essential, and may be part of a management plan.

3. Non-oral feeding

People who are at high risk of aspiration and who cannot maintain adequate food and fluids intake by mouth will likely require feeding by another route. Assessing the need and the best type of non-oral feeding are decisions that must be made between the person with the swallowing difficulties and their family/carer, medical staff and a dietician.

Naso-gastric tube (NGT) feeding requires a tube to be inserted from the nose into the stomach. Special food formula, water and crushed or liquid medications can then be inserted through the NGT. Use of naso-gastric tube feeding is usually a short-term solution.

Percutaneous endoscopic gastrostomy (PEG) is generally used for people who have severe swallowing difficulties that are expected to continue in the long term. A PEG tube is inserted directly into the person's stomach, bypassing the structures used for swallowing and breathing. Special food formula, water and crushed or liquid medications can be inserted through this tube directly into the stomach. It is important that carers:

- follow the care plan/management plan as prescribed
- observe, report and record progress and any changes.

Who can I talk to if I have questions or concerns?

MJD Foundation staff
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Your Health clinic doctors, nurses and health workers.