

Oral Health

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1. Background — Oral Health and MJD

Machado Joseph Disease (MJD), also known as Spinocerebellar Ataxia 3 (SCA3), is a neurodegenerative disorder that results in patients dependent for activities of daily living. The presentation of symptoms in most cases is not exclusively ataxic and may include other neural system impairments such as Parkinsonism and dysphagia.¹ As patients become progressively less capable of self-care and other care needs take priority for carers, oral hygiene can often be neglected as it may not appear to require high prioritisation at first glance. However, the maintenance of good oral hygiene is fundamental for patients affected by neurological disease.²

At this point in time, there is not a great volume of published articles that address MJD and oral health. No study has evaluated the oral health of people with MJD. Literature is available for other neurodegenerative diseases sharing similarities to MJD, such as Huntington's Disease (HD), Parkinson's Disease (PD) and Motor Neuron Disease (MND) also known as Amyotrophic Lateral Sclerosis (ALS).³ Following this logic, suggestions relating to the maintenance of oral health for patients with MJD will be discussed where the symptoms of other neurodegenerative diseases are similar to MJD.

Dental health and movement disorders are interdependent. It has been suggested that ineffective oral hygiene is a common feature in patients who have MJD due to limited mobility and lack of coordination in upper limbs, including fine motor movement of hands and fingers, along with dyskinesia of the tongue and perioral musculature.^{4,5}

Evidence suggests a correlation between disease progression and dental deterioration. In a cohort of people studied with HD, lower functional status lead to worse dental health, measured by higher plaque scores, when compared to a control group.⁵ Similar conclusions, such as high incidence of untreated caries, periodontal disease, and missing teeth were also found for patients studied with PD when compared to the control.² In the same study of patients with HD, lower scores for filled teeth may be explained by a higher rate of tooth extraction due to accidental trauma, often the consequence of falls, and poorer dental status. It would be safe to assume that a similar pattern would be apparent in people with MJD as they are also prone to accidental trauma and falls. In a descriptive study of patients with Spinocerebellar Ataxia 2 (SCA2)⁶, roughly 75% of the patients had poor oral hygiene significantly correlated with CAG repetitions. CAG (Cytosine-Arginine-Guanine) is the genetic coding sequence abnormally repeated in the DNA of people with MJD. Its repetition length directly correlates with onset and severity of the disease.¹ Due to the similarities between SCA2 and MJD (SCA3), the findings from the aforementioned study could be extrapolated to suggest comparable results for people with MJD.

1.1. Swallowing Implications

As highlighted in the [Swallowing Medical Protocol](#)^a, good oral hygiene is important to reduce risk of aspiration pneumonia and other airway infections. There is now a great deal of published literature that clearly demonstrates a causal relationship between oral health and systemic health. Poor oral health is associated with increased cardiovascular disease risk.⁵ Aspiration pneumonia has been cited as the most common cause of death in spinocerebellar ataxia disorders.⁸ Teeth play a vital role in the mastication of food, which allows for more predictable, safe swallowing. People with swallowing difficulties may have plaque and food particles that remain in the buccal sulci for prolonged periods. This poor oral clearance increases the likelihood of caries.^{9,10,11} People with MJD who have swallowing difficulties may require modified foods and fluid. Modified diets can further contribute to caries risk as the thickened gel can be hard to clear from the oral cavity and the food supplements often have high sugar content.^{11,12}

Where swallowing function is severely compromised, nutrition may need to bypass the oral cavity. Individuals who are unable to swallow may receive feeding via percutaneous endoscopic gastrostomy (PEG). Authors of several articles found higher rates of dental plaque and calculus accumulation in people receiving PEG feeds.¹³ Plaque accumulation contributes to gingivitis, a precursor to periodontitis; therefore, PEG feeding and periodontal disease may be correlated.¹¹

1.2. Poor oral health is inexorably linked with pain, which can further lead to a deterioration in well being. Additionally, oral health and its impact on oral function is associated with quality of life; poor oral health has been linked to physical, mental, and social impairments.^{7,14,15} Barriers to Access Dental Care

People with disabilities have significant challenges to access services, including dental care. These include:

- availability of disability accessible transport and dental surgery clinics^{16,17}
- cost¹⁸
- dentists' possible lack of experience and training to treat people with disabilities¹⁸
- caregivers' priorities for care¹⁸

For people with MJD, this is made ever more challenging due to additional barriers:

- possible communication difficulty due to dysarthria and anarthria¹
- lack of dental professional's knowledge of MJD and potential reluctance to provide care¹⁹
- difficulty providing treatment to those with limited voluntary control of their movements and/or difficulty following commands
- inability to tolerate dental treatment due to involuntary movements or pain/cramps associated with MJD¹
- in the later stages of MJD, people rely on a carer to provide mouth care provided by a carer, other barriers at this stage include lack of training,^{20,21} time constraints,^{20,22,23} and use of ineffective oral care equipment, for example foam swabs, which may be chewed, and inhaled.²⁴

^a http://mjd.org.au/cms/file_library/Other/Other_759.pdf

1.3. Considerations for Indigenous Australians with MJD

Indigenous Australians with MJD experience further challenges in regards to oral health. Indigenous Australians have poorer oral health, higher prevalence and greater severity of caries, periodontal diseases and tooth loss. The only treatment on offer may be an extraction for oral diseases left untreated, and at late presentation.²⁵ Total edentulism, or complete tooth loss, occurs at younger ages for Indigenous Australians and has a significant impact on oral function and quality of life. Poor oral health and subsequent pain can also lead people to avoid certain foods or limit oral intake affecting nutrition.²⁵

Young Indigenous Australian adults are impacted by two to eleven times the adverse oral health outcomes compared to nationally representative counterparts.²⁶ These figures for young adults are important considerations for people at risk of MJD or with 'mild' MJD where future speech/swallowing issues will be key to quality of life maintenance throughout the disease progression.

Oral health literacy (OHL), or the ability to obtain and interpret basic oral health information and services in order to make and act on informed decisions, is comparatively lower for Indigenous Australians. OHL is associated with fewer dental visits, poorer self-rated oral health status, and more perceived treatments, such as extractions.²⁷ Furthermore, other barriers to accessing oral health services include lack of remote services, transportation issues, extensive waiting lists for public services, and co-payment requirements.²⁵ In rural and remote areas, the issues are compounded by additional direct barriers to good oral health like restricted or expensive dietary options and/or no access to fluoridated water, which is the most effective measure to reduce caries.^{25,28,29} Overcrowding, a problem for many Indigenous Australians in both urban and remote settings, can also directly impact oral health as it may lead to decreased toothbrush use from excessive uncleanness, among other contributors.³⁰ For example, wet areas used by multiple people do not allow for safe storage of equipment.

2. Oral symptoms as clinical indications of MJD

2.1. Fasciculations & Bruxism

It is well documented that fasciculations (visible and rapid muscle fibre contractions) are associated with MJD.^{1,31,32,33,34,35} Fasciculations have been observed primarily in the tongue and facial region and may involve the peripheral nerves and peripheral axonal excitability.^{31,34,35}

Although bruxism (also known as teeth grinding) is not documented in the literature to be a symptom of MJD, anecdotally bruxism is associated with MJD. Following the logic that bruxism is common in other neurodegenerative disorders, like Huntington's disease, bruxism in people with MJD may be associated with the disease itself. This could be argued given orofacial dyskinesia like fasciculations are understood to be a part of the disease process. Bruxism causes severe tooth wear and jaw pain.¹¹

2.2. Defects in Tongue Papillae and Taste

MJD has been found to be associated with a lack of fungiform papillae in a Japanese family.^{36,37} Taste sensitivity of the tongue is directly related to the number and density of fungiform papillae.³⁸ A decrease in taste sensitivity can be associated with food aversion and have a negative impact on nutritional status and quality of life.³⁹ The lack of fungiform papillae in some people with MJD suggests a decrease in neurotrophic support.⁴⁰

2.3. Dry Mouth (Xerostomia)

Results of one study⁴¹ comparing autonomic dysfunction in patients with MJD to healthy participants and those with Parkinson's disease suggest autonomic dysfunction is not uncommon in MJD. People with Parkinson's disease were used as comparators given autonomic dysfunction is well recognized in PD. Dry mouth, a symptom of autonomic dysfunction, was reported in a significantly higher percentage of patients studied with MJD (33.3%) compared to the healthy participants. The occurrence of dry mouth in people studied with MJD and Parkinson's disease was equal.

Along with dyskinesia/hyperkinesia of the tongue and perioral musculature (e.g. fasciculations), dry mouth combined with the pooling of saliva may influence deficient oral hygiene, i.e. increased risk of cavities and plaque accumulation.⁵

3. Recommended Guidelines for addressing Oral Health in Patients with MJD

The Remote Health Atlas for MJD, developed by the Northern Territory Government Department of Health, recommends clients with MJD be referred for oral health services to assess oral health status.⁴² The extra attention to oral care for people 'at risk' or with pre-symptomatic MJD given future swallowing issues is also important. It has been concluded with Huntington's disease that preventive dentistry from diagnosis is the most important aspect of dental care, in addition to the goals of natural dentition maintenance and early intervention to prevent crisis management in the more advanced stages of the condition.^{5, 43} The same early intervention has been advocated for people with PD to prevent serious pathological dental conditions and tooth extraction.² *For this reason, patients with a known diagnosis or 'at risk' of MJD should be prioritised for dental referrals and appointments.*

In addition to dental maintenance, dentists treating patients affected by neurodegenerative disorders are confronted with mobility, cognition, and behaviour problems.⁴⁴ Although there is no evidence to support cognitive impairment in MJD to the degree expressed in other neurodegenerative disorders, a few studies limited by sample size have demonstrated cognitive deficits such as executive dysfunction and memory loss.^{45,46} There is certainly further research which needs to be carried out before any definitive conclusions can be made about the connection between MJD and cognitive impairment, if any. To respect the patient's wishes and not cause

offense, always assume (unless told otherwise or from experience) that the patient understands and can communicate.¹¹

Due to the complex needs, it is preferable that people with neurodegenerative disorders be treated by the same dentist who collaborates with all professional and nonprofessional groups concerned with the care of the individual.⁴⁴ Authors of one study⁵ recommended patients with HD in all disease stages visit the dentist a minimum of every 4 months. Other authors suggest a titrated dental regimen for people with HD beginning with 6 monthly review in the early stage to 3-4 monthly review in the middle and late stages.¹¹ Comparable timeframes would similarly benefit patients with MJD. Calibrating the frequency of review visits to the individual's needs and the disease progression is important.

Consider short appointments and breaks during care, since prolonged time spent in one position can lead to skin breakdown and increased pain or cramps. Wheelchair transfer to the dental chair may be required for which sliding boards can assist with the transfers, but staff should be trained in their use.⁴⁷ For people with more advanced MJD that has begun to cause breathing difficulties, do not lay the person completely supine, but position at 45 degrees with cushions for posture support and pressure relief.^{11, 47} It may be possible to perform dental care while the patient remains seated in their wheelchair with head and mandible supported as shown in figure 1.⁴⁸ Note this position is quite taxing for the operator, and should not be considered for long or exacting procedures.






Fig. 1 Positioning for treating a patient in a wheelchair.

Mouth wash should be avoided in the late-moderate and severe stages of MJD as it may pose an aspiration risk.¹¹

Mouth props (for people who can breathe nasally) and rubber dams may be useful to facilitate dental procedures.⁴⁷ It is important to explain to the patient how these devices work to ensure the patient will be comfortable during the procedure.

To maintain oral hygiene self-care in the moderate stages of MJD, oral hygiene aides may be useful for some individuals.¹¹ It is really a question of consulting, experimenting and creating a device that is useful for the patient. Some options for trial include:



<p>Dr. Barman's Superbrush®</p>	
<p>Enlarged handles/grips</p>	
<p>Mouth angle expanders</p>	

While preservation of self-care and independence in the early stages is important, it is also vital the carer receives education to supervise and evaluate hygiene procedures.⁵ Education for the carer is crucial as the person with MJD will progressively be unable to perform even the simplest oral care tasks.⁵ A mix of educational tools to suit various learning styles should be implemented; e.g. written instructions, pictures, DVDs.¹¹ An example of oral health education for carers can be found at <http://www.futuremedicine.com/doi/suppl/10.2217/NMT.11.68>

4. Pharmacological Treatment Options

4.1. Fasciculations & Bruxism

4.11 Botulinum Toxin

This treatment option is complex and should only be considered where good access to specialist practitioners is available. Botulin Toxin A has been used clinically to treat fasciculations and bruxism.^{48,49} Some authors, however, advocate against its use, because it is a temporary therapy and can be associated with greater risks, such as severe, intractable headache.^{50,51}

4.22 Antiepileptic drugs

This treatment option is complex and should only be considered where good access to specialist practitioners is available. Carbamazepine and Phenytoin have shown partial therapeutic response for fasciculations.³² Gabapentin in low doses from 300-600 mg can also be used.⁵²

4.2. For Defects in Tongue Papillae

Some studies^{53,54,55} indicate that antidepressants such as paroxetine and fluoxetine may be useful in treating defects in tongue papillae and taste sensation for Huntington's disease and Alzheimer's disease. However, as the mechanism of disease differs, these same treatments may not be as successful in MJD.⁵⁶

4.3. Dry Mouth (Xerostomia)

Consideration should be given to medications as a causal factor of dry mouth, since xerostomia is a common side effect of several drugs. The use of 2-3 squirts of artificial saliva in the mouth hourly (while awake) is recommended for relief.

4.4. Pain and Anxiety Management during Dental Procedures

Local anaesthesia should be considered in the first instant. Caution should be used and specialist advice always sought prior to the use of conscious sedation or general anaesthesia due to the risk of further respiratory compromise.^{11, 47}

5. Therapeutic Treatment Options

5.1. Fasciculations & Bruxism

Authors of one review³² suggest that treatment is not always necessary due to the benign and transitional nature of fasciculations. Fasciculations may improve with the identification and correction of sleep apnoea, which in many cases can be a trigger.⁵⁸ Some patients have found various manoeuvres (sensory tricks) and dental prosthetic devices relieve their jaw spasms.⁵⁹

5.2. Defects in Tongue Papillae

Nil identified.

5.3. Dry Mouth (Xerostomia)

Suggestions include taking sips of water or sugarless drinks, regular swabbing with water or saliva substitutes, avoiding caffeinated drinks, reducing and abstaining from tobacco and alcohol.⁶⁰

6. Other Treatment Options

6.1. Prosthetics

Xerostomia and reduced muscular control (like fasciculations) make dentures difficult to use and present the additional risk of airway obstruction.^{4,11,12,44,47} Patients may also have trouble inserting and removing dentures and/or rely on denture maintenance provided by a carer. Therefore, complete dentures for the person who is unable to physically remove them and/or call out for help should only be used during mealtimes with 100% supervision.⁴⁷

Appendix A – Contributors and Reviewers

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Appendix B – Definitions

Anarthria – severe dysarthria resulting in speechlessness

Botox – Botulinum toxin (BTX) is a neurotoxic protein produced by the bacterium *Clostridium botulinum*.

Bruxism – grinding of the teeth

Buccal sulci- the spaces between the teeth and the mucous membrane lining the cheek

Caries –decay and crumbling of a tooth or bone

Dysarthria – is a group of motor speech disorders resulting from disturbed muscular control of the speech mechanism due to the damage of the peripheral or central nervous system causing weakness, incoordination, or paralysis of speech musculature (See [Communication Difficulty Protocol](#)).

Dyskinesia – abnormality or impairment of voluntary movement

Dysphagia – swallowing difficulty

Edentulism – the condition of being toothless to at least some degree

Fasciculations – visible fast, fine contractions of fine muscle fibres that occur spontaneously and intermittently

Fungiform papillae – broad flat structures in the centre back of the tongue where taste buds are located that taste buds

Percutaneous endoscopic gastrostomy (PEG) – medical procedure in which a tube is passed into the stomach through the abdominal wall, most commonly to provide a means of feeding when oral intake is inadequate

Xerostomia – technical term for dry mouth

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