

Title Page

Oropharyngeal Dysphagia in Neurodegenerative Disease

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Abstract

Oropharyngeal dysphagia is frequent in neurodegenerative diseases with medical, social and psychological effects. There is a paucity of evidence on the frequency, nature and course of oropharyngeal dysphagia in these conditions. The nature and characteristics of the swallowing disorder can vary according to the neurological condition, the severity of the underlying disease and other patient comorbidities. Early detection of oropharyngeal dysphagia is important to decrease aspiration risk and limit health and social consequences. Management options vary from compensatory and rehabilitation exercise approaches with evolving evidence on how best to manage the condition. Many individuals require enteral tube feeding in the later stages of the disease.

This paper examines oropharyngeal dysphagia in some of the primary neurodegenerative diseases, exploring current management options highlighting directions for research and clinical practice.

Keywords: Deglutition Disorders, Neurodegenerative disease, Review

Introduction

Neurodegenerative diseases [NDDs] are characterized by the loss of neurons in the brain or spinal cord. People with NDDs have problems associated with movement and/or cognitive function. Core degenerative diseases are the dementias, Parkinson's disease, motor neuron disease, cerebellar degenerations, Huntington's disease and prion diseases.¹ NDDs are generally incurable, progressive and debilitating conditions with considerable impact on health and health related quality of life. Populations with NDDs are increasing. This is attributed to earlier detection and an increased range of interventions for management with improved survival rates. Age is a risk factor in their development and the global increase in aging populations further contributes to the increased prevalence of NDDs.²

Oropharyngeal dysphagia is defined as difficulty with swallowing food, liquid or saliva. It arises from impairment of anatomy and/or physiology of the upper aerodigestive tract and is prevalent in people with NDDs. This is hardly surprising given that deficits in the skills frequently associated with neurodegenerative conditions (i.e. cognition, movement, strength, sensation and coordination) are key prerequisites for safe and efficient swallowing. The nature and characteristics of oropharyngeal dysphagia associated with different NDDs vary between diseases; however the consequences for the individual are broadly similar.

This paper provides an overview of oropharyngeal dysphagia in the primary NDDs, reviewing their key characteristics, impact and management options with suggestions for research and clinical practice.

Nature and impact of oropharyngeal dysphagia in neurodegenerative diseases

The safe transport of food and fluid from the mouth to the esophagus involves the fast and efficient coordination of numerous facial, oral, pharyngeal, laryngeal and esophageal muscles, with intact laryngeal and pharyngeal reflexes. Cognitive impairment, movement disorders, reduction in muscular strength, reduced sensation, and incoordination affect eating, drinking and swallowing in different ways (Table 1). There are a number of risk factors that increase the likelihood of developing oropharyngeal dysphagia. One of these is increasing age. While aging alone in normal healthy adults will rarely cause oropharyngeal dysphagia,³ it can become an issue when combined with factors such as increased medication use,⁴ decline in isometric tongue pressure,⁵ reduction in sensory discrimination, loss of dentition, poor oral hygiene and reduction in salivary flow.⁶ These factors may further exacerbate a co-existing dysphagia. Additional concerns include decreased mobility and poor posture, coexisting respiratory impairment, decreased cognition and level of alertness.

Oropharyngeal dysphagia can have a negative impact not only for the individual but for carers, families and other key stakeholders. Dehydration, malnutrition, weight loss and aspiration pneumonia are common consequences of oropharyngeal dysphagia. Malnutrition is specifically associated with prolonged hospitalization, increased risk of hospital re-admission, delayed recovery from illness and increased mortality.^{7,8} Oropharyngeal dysphagia also causes considerable discomfort for the individual with choking episodes which are anxiety provoking. For people with motor neuron disease (MND) and Parkinson's disease (PD) the additional discomfort of excess saliva resulting from a decreased ability to swallow is considerable.⁹⁻¹¹

Oropharyngeal dysphagia can affect social activities and reduce participation in society as the swallowing difficulty limits the ability to eat out socially. Modification of normal diet and the prescription of thickened liquids as compensatory strategies for dysphagia, further impact quality of life.¹²⁻¹⁴ People with NDDs frequently develop an emotional and psychological reaction to oropharyngeal dysphagia and its management. Studies suggest that anxiety and depression can be associated with oropharyngeal dysphagia.¹³⁻¹⁵

Characteristics of oropharyngeal dysphagia in neurodegenerative diseases

Oropharyngeal dysphagia in the dementias.

It is estimated that there are over 35.6 million people living with dementia worldwide, with an anticipated doubling of these numbers every 20 years. These numbers are estimated to reach 65.7 million people in 2030 and 115.4 million by 2050.¹⁶ Oropharyngeal dysphagia is common in dementia with prevalence rates varying from 13-57%.¹⁷ Oropharyngeal dysphagia in advanced stages of dementia is associated with malnutrition and aspiration pneumonia.^{17,18} Aspiration pneumonia is a significant cause of death in this population.^{19,20}

Alagiakrishnan et al.¹⁷ recently completed a systematic review on oropharyngeal dysphagia in different types of dementia, examining Alzheimer's disease, (AD) vascular dementia (VaD), Lewy body dementia (LBD) and frontotemporal dementia (FTD). They conclude that the nature of oropharyngeal dysphagia varies according to the type of dementia (Table 2). Impaired sensation is

associated with AD resulting in slow oral transit time.²¹ In individuals with VaD however, impairment to the motor components of swallowing is more characteristic with difficulty on bolus formation and mastication. Individuals with VaD are also reported to have a higher incidence of silent aspiration when compared to people with AD.²¹ Individuals with Lewy body dementia have prolonged eating times and pharyngeal phase dysphagia,^{22, 23} while in FTD behavioral feeding problems and pharyngeal phase dysphagia predominate.^{24, 25}

Oropharyngeal dysphagia in Parkinson's disease

PD is an idiopathic progressive neurodegenerative disease characterized by tremor, muscular rigidity, and bradykinesia

The prevalence of oropharyngeal dysphagia in PD varies according to the definitions of dysphagia used in the epidemiology studies, the severity of the disease and methods used to determine its presence. Kalf et al.²⁶ completed a meta analysis of the literature on oropharyngeal dysphagia prevalence in PD. They report that oropharyngeal dysphagia prevails in at least a third of individuals with PD, and people with PD are three times more likely to have swallowing disorders than healthy controls.

Silent aspiration of saliva is reported in people with PD and oropharyngeal dysphagia. Silent aspiration is attributed to reduced pharyngeal and laryngeal sensation.²⁷⁻²⁹ although Leow et al.³⁰ suggest that PD affects chemosensory and mechano- sensory pathways differently with mechanosensation affected before chemosensation. Dopaminergic medication typically does not improve swallowing performance although this is a subject of debate.^{31, 32} Cognitive impairment is a complication as the disease progresses and aspiration pneumonia is considered a leading cause of death.^{29 33} Quality of life is significantly impacted by the presence of dysphagia with decreased quality of life as the disease progresses^{12, 34}

There are a number of syndromes that share symptoms of PD but are considered atypical parkinsonism or parkinson-plus syndromes. These reportedly do not respond well to dopaminergic medication. These include progressive supranuclear palsy (PSP), multi-system atrophy (MSA) and corticobasal degeneration (CBD). Oropharyngeal dysphagia is also common in these syndromes³⁵⁻³⁷ and dementia is a further feature exacerbating oropharyngeal dysphagia. The characteristics of oropharyngeal dysphagia in these atypical parkinsonism syndromes appear to mimic dysphagia in PD and the dementias (Table 3). Kalf et al.²⁶ suggest that while oropharyngeal dysphagia is not considered an early symptom of PD, it can be reported early in people with PSP and MSA.^{36,37} Thus, it may be an important symptom in early differential diagnosis between PD and other atypical parkinsonism syndromes.

Oropharyngeal dysphagia in Motor Neuron Disease

MND is a general term for a group of neurological diseases that affect the motor neurons. Amyotrophic lateral sclerosis (ALS) (also known as Lou Gehrig's

disease) is the most common form of MND with both upper and lower motor neuron involvement. Oropharyngeal dysphagia is reported to be prevalent in 30 – 100% of individuals depending on type of MND and the stage of disease³⁸ affecting all individuals in the later stages of the disease. Oropharyngeal dysphagia is characterized by weakness, and fatigue of oropharyngeal and laryngeal muscles with aspiration.³⁹ Drooling is common due to difficulty swallowing saliva.⁹ Respiratory involvement results in a weakened cough reflex with an inability to prevent laryngeal penetration and aspiration.⁴⁰ Specific characteristics of oropharyngeal dysphagia are described by a number of researchers (Table 5). Quality of life is specifically impacted as individuals report limitations in food selection, an increase in eating duration and a decrease in eating desire.¹⁵ Non-oral tube feeding is common in the later stages of the disease particularly in bulbar subtypes of MND.

Oropharyngeal dysphagia in Huntington's disease

Huntington's disease (HD) is an inherited neurodegenerative disease characterized by motor, behavioral and cognitive disturbances. The prevalence of oropharyngeal dysphagia in this population is difficult to determine due to methodological limitations and the paucity of studies in the area, however, oral preparatory, oral phase, pharyngeal and esophageal phase dysphagia is common⁴¹ (Table 5). Kagel and Leopold⁴² classified people with HD into hyperkinetic (HD-h) or rigid-bradykinetic (HD-rb) groups and suggest significant intergroup differences with respect to dysphagia. The small numbers involved in the study limit the generalization of their findings. Other studies,⁴³⁻⁴⁵ exploring the characteristics of oropharyngeal dysphagia in this population suggest that incoordination of swallowing and choreic tongue movements with delay in initiation of the pharyngeal swallow are characteristic (Table 5). Difficulty with self-feeding due to choreic movements and the existence of cognitive impairment as the disease progresses exacerbate oropharyngeal dysphagia.⁴³⁻⁴⁵ Pneumonia and choking are reported to be a leading cause of death.^{46,47}

Oropharyngeal dysphagia in prion diseases

Prion diseases are a group of neurodegenerative disorders that includes sheep scrapie, bovine spongiform encephalopathy (BSE) and Creutzfeldt-Jakob disease (CJD).⁴⁸ Oropharyngeal dysphagia has been reported in series of single case studies of people with CJD.⁴⁹⁻⁵² CJD is characterized by an accumulation of abnormal prion-like proteins in the central nervous system affecting the cerebral cortex primarily and confirmed usually at autopsy. Oropharyngeal dysphagia can be one of the initial signs of the disease in association with a bulbar or pseudobulbar palsy with rapid progression.^{50, 52} Cognitive impairment can be a significant factor also in the disease. To date, these diseases are rare and the knowledge of key characteristics of oropharyngeal dysphagia remains limited to single case studies. The clinical presentations of oropharyngeal dysphagia reported vary. Mittal et al.⁵⁰ report a case study of a woman with a 4-week history of progressive dysphagia characterized by poor bolus control, delay in initiation of the pharyngeal swallow and residue in the vallecula and pyriform sinus post swallow. Hasegawa et al.⁴⁹ report unilateral laryngeal and pharyngeal

involvement with unilateral pharyngeal residue in a further case study. The common feature of all cases described is the rapid evolution of oropharyngeal dysphagia as the disease progresses with imminent death.

Assessment of oropharyngeal dysphagia in neurodegenerative disease

Early detection with appropriate management of oropharyngeal dysphagia in NDDs is essential to prevent dehydration, malnutrition, weight loss, aspiration pneumonia and improve overall survival and quality of life. Assessment of NDDs should include questioning specifically on swallow function. However, assessment of dysphagia should not be confined to questionnaire items and patient report as populations with reduced sensation and cognitive function may be unaware of their swallowing difficulties.³⁵ Studies suggest that reliance on individuals' reports of dysphagia alone is inadequate even in those with normal cognition and sensation, as people often consider difficulties as part of aging and the disease process.^{53,54}

Screening for oropharyngeal dysphagia is important to identify people at risk of impairment with appropriate onward referral for more thorough assessment. Tests validated on large cohorts of patients with oropharyngeal dysphagia associated with mixed etiologies are popular. The Yale Swallow Protocol⁵⁵ is one example. Disease specific questionnaires such as the Munich Dysphagia Test-Parkinson's Disease (MDT-PD),⁵⁶ and the Huntington's Disease Dysphagia Scale⁵⁷ aim to detect swallowing difficulties and aspiration risk in these populations. Despite the range of tests available, few have strong established psychometric properties. Kertscher et al.⁵⁸ completed a systematic review of bedside screening test to detect oropharyngeal dysphagia in patients with neurological disorders. Only two valid screening tests were identified. The Volume Viscosity Swallowing Test (V-VST)⁵⁹ and the Toronto Bedside Swallowing Screening Test (TOR-BSST).⁶⁰ Both tests have high sensitivity ($\geq 80\%$). However, the TOR-BSST has been validated on patients following stroke population limiting its validity somewhat with a NDD population.

Following the detection of a probable oropharyngeal dysphagia, further more detailed assessment of swallowing must be carried out. This may involve instrumental assessment particularly if silent aspiration is suspected. Reference standard examinations for oropharyngeal dysphagia are modified barium swallow (videofluoroscopy) and fiberoptic endoscopic examination of swallowing (FEES). Pharyngeal manometry is indicated where problems with upper esophageal dysfunction are indicated. However, the practicality of completing instrumental assessments particularly on people with dementia remains a challenge. Thus, careful routine monitoring with thorough non-instrumental clinical swallowing evaluations is recommended. This requires education of the individual with NDD and caregivers. Assessment requires a multidisciplinary approach with nurses and caregivers trained in detecting the signs of oropharyngeal dysphagia at mealtime with further more detailed assessment provided by speech-language pathologists (SLPs), radiologist with onward referral to other specialists (e.g. gastroenterologists, otolaryngologists, pulmonary physicians, dietitians, physical therapists, occupational therapists etc.).⁶¹⁻⁶³

Intervention for oropharyngeal dysphagia in neurodegenerative disease

Intervention is about improving the prognosis and course of the disease taking quality of life into consideration. Research suggests that early intervention with multidisciplinary team involvement should improve overall patient outcomes including prolonged independence in self-feeding⁶⁴ and reduction in aspiration pneumonia⁶⁵ for people with dementia. Education of the individual with NDD, family and carers is a key part of any intervention program.⁶⁵

Intervention is considered broadly as compensatory and rehabilitative. Rosenbek and Jones ⁶¹ define compensatory treatments as those aimed at immediate change on the safety and sufficiency of hydration and nutrition, while rehabilitation techniques are intended to improve the mechanism and skill of swallowing itself. Although both approaches are frequently used together, in people with neurodegenerative disease compensatory strategies are most frequently applied.¹⁷

Compensatory techniques include changes to body, head and neck posture, modification of food and/or fluid, and adaptation of methods of eating and drinking. The evidence base to support many of these compensatory approaches is sparse.^{17, 62-69.}

Rehabilitation techniques are used according to the underlying neurodegenerative disease. Rehabilitation exercises are generally used with caution with some populations (e.g. MND) as it is hypothesized that they can increase fatigue and lead to swallowing deterioration, while they are considered beneficial for other populations such as PD.^{70, 71}

Other approaches such as pharmacological and surgical interventions have been largely ineffective in treating oropharyngeal dysphagia in NDDs. The use of deep brain stimulation (DBS) for people with PD has not improved swallowing despite improvements in reducing general body tremor and motor function.⁷² Pharmacological interventions are used widely to help ameliorate drooling. There is some evidence to suggest that botulinum toxin may be helpful in reducing saliva production for people with MND and PD.^{73, 74}

Management of oropharyngeal dysphagia at end stage disease

Many people with NDDs receive palliative care as the disease progresses. Langmore and colleagues ⁷⁵ define palliative care in the context of dysphagia as, *'treatment for severe and chronic dysphagia or intractable aspiration when the recovery of normal swallowing is not anticipated and attempts to restore normal swallowing have been unsuccessful'* (p6). They suggest that palliative care for oropharyngeal dysphagia at this point should focus on maximizing swallowing function, preserving pulmonary health, and nutrition given the limitations of the dysphagia. Maintaining oral hygiene is also important at this stage for patient comfort and to prevent further respiratory complications including aspiration pneumonia.^{76, 77}

Numerous individuals require enteral feeding as oropharyngeal dysphagia progresses and care becomes palliative. The decision to initiate non-oral feeding gives rise to much debate on ethical and legal matters particularly when the individual's capacity to consent is absent or impaired. There is no strong evidence to suggest that enteral feeding prolongs life in NDDs such as dementia^{78, 79} and MND⁸⁰. A recent systematic review by Goldberg and Altman⁷⁸ examined the efficacy, complications, outcomes and survival of people with dysphagia associated with dementia who had percutaneous endoscopic gastrostomy tubes (PEG) in-situ. They found no evidence that PEG feeding improved long-term survival rates, supporting earlier findings by Candy et al.⁷⁹ Katzberg and Benatar⁸⁰ could equally find no robust evidence to indicate that enteral feeding is beneficial to individuals with MND. However, they suggest with caution that there may be a 'survival advantage' for some individuals.

Directions for research and clinical practice

The body of evidence on underlying mechanisms and pathophysiology of dysphagia in NDDs is emerging. Michou et al.⁽⁸¹⁾ provide an overview of the current knowledge base and research challenges in PD. They suggest that large-scale research with a specific focus on understanding the neurophysiology of the disease is needed. This mirrors the position in other key NDD syndromes.

Oropharyngeal dysphagia in NDD is multifaceted existing in the context of other comorbidities that in isolation are sufficient to give rise to swallowing disorders and increased aspiration risk. These include polypharmacy, decreased cognition, poor physical mobility and difficulty with self-feeding.

More methodologically sound epidemiological research on oropharyngeal dysphagia in neurodegenerative disease is required. The presence, nature, characteristics, and extent of oropharyngeal dysphagia must be rigorously determined in populations so that the natural history and course of oropharyngeal dysphagia across NDD syndromes is better understood.

Undoubtedly, oropharyngeal dysphagia and its consequences are gaining recognition within multi-disciplinary teams. The evidence for compensatory strategies and intervention approaches across all NDDs remains low. This is not because research suggests that these approaches do not work but rather that there is limited research to confirm or refute their efficacy and effectiveness. Good quality research to translate new pathophysiological insights into feasible treatment approaches and to evaluate the efficacy of these new treatments is costly and can take many years. Recent systematic reviews cited in this paper have helped to draw the available body of evidence together and provide much needed direction for clinical research. Furthermore, outcomes have typically focused on death and disease rather than disability, discomfort and health related quality of life. This is changing with an increased recognition of psychological and social domains. More qualitative research is required to support quantitative studies, exploring patient preferences and social impact.

In conclusion, oropharyngeal dysphagia is a complex debilitating condition associated with this group of diseases. The fact that aspiration pneumonia

contributes significantly to patient mortality suggests that we must do better at understanding and managing this condition thus improving care for this client group.

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Table 1: Impairments associated with neurodegenerative diseases and their influences on swallowing.

Cognitive Impairment	Movement Disorder	Reduction in Strength	Reduction in Sensation	Incoordination
Primitive oral reflexes	Difficulty self-feeding	Reduced bolus pressure	Increased oral, and pharyngeal residue	Oral and pharyngeal phase disruption
Rapid compulsive eating	Poor bolus propulsion	Reduced airway protection	Silent aspiration	Poor coordination of respiration and swallowing
Poor monitoring of bolus size	Reduced bolus control	Incomplete bolus clearance		
Impaired consciousness and alertness				
Food refusal				

Table 2: Characteristics of oropharyngeal dysphagia in the dementias

Type of Dementia	Characteristics
Alzheimer's Disease	Difficulty self-feeding ^{82,83} Oral praxis ^{82,86} Prolonged eating duration ^{83,84} Oral residue ⁸³ Loss of food from the oral cavity ⁸³ Difficulty chewing ⁸³ Prolonged oral phase ^{17 84} Absent or continuous chewing ⁸³ Primitive oral reflexes (bite reflex) ⁸³ Increased pharyngeal response duration ⁸⁴ Wet voice ⁸³ Multiple swallows ⁸³ Reduced hyolaryngeal elevation ⁸⁵ Aspiration as disease progresses ⁸⁶
Lewy Body Dementia	Pharyngeal phase dysphagia ²³ Prolonged eating duration ²²
Frontotemporal dementia	Behavioral feeding problems (rapid eating, over stuffing mouth) ²⁴ Delayed pharyngeal swallow response ²⁵ Incomplete bolus clearance with residue in pharynx post swallow ²⁵ .
Vascular dementia	Difficulty with bolus formation and mastication ²¹ Decreased hyolaryngeal elevation ²¹ Silent aspiration ²¹

Table 3: Characteristics of oropharyngeal dysphagia in Parkinson's disease and Parkinsonian syndromes

Disorder	Characteristics
Parkinson's disease	Difficulty with bolus manipulation and control ^{27,87,88} Xerostomia ²⁷ Delay in initiation of pharyngeal swallow ^{27,87} Residue in pharynx ^{27,87} Abnormal airway somatosensory function ²⁷ Decreased upper esophageal opening and relaxation ⁸⁸ Silent aspiration ⁸⁷
PSP	Difficulty with bolus manipulation and control ⁸⁹ Premature loss of bolus into pharynx ⁸⁹ Residue in valleculae and pharynx ⁸⁹
MSA	Slow bolus transfer from oral to pharyngeal cavity ⁹⁰ Prolonged hold of bolus in oral cavity ⁹⁰ Incoordination of tongue ⁹⁰ Aspiration ⁹⁰

Table 4: Characteristics of oropharyngeal dysphagia in motor neuron disease

Characteristics
Difficulty swallowing saliva ⁹¹
Leakage of fluids on drinking due to poor lip closure ⁹³
Reduced tongue strength and endurance with poor bolus manipulation ⁹²
Poor mastication ⁹³
Weak bolus propulsion ⁹³
Delayed bolus transit ⁹²
Residue in lateral and anterior sulci post swallow ⁹³
Nasal redirection ^{40,93}
Delay initiating pharyngeal swallow ⁹³
Decreased laryngeal elevation ³⁹
Residue in the pharynx post swallow ³⁹
Aspiration before, during and after swallow ^{39, 40, 92, 93}

Table 5: Characteristics of oropharyngeal dysphagia in Huntington's disease

Characteristics
Poor lip closure ⁴¹
Poor bolus manipulation ^{41,44}
Difficulty with mastication ⁴¹⁻⁴⁴
Decreased palatal elevation ^{41,44}
Delayed initiation of the pharyngeal swallow ^{41,43}
Repetitive swallows ⁴³
Pharyngeal residue ^{41,43,44}
Decreased pharyngeal peristalsis ^{41,44}
Coughing and choking on liquids ⁴³
Aspiration ⁴⁴
