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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 syndromes [35-37] 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 disease [38] 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 common [41] (Table 5). Kagel and Leopold [42] 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 Protocol [55] is one example. Disease specific questionnaires such as the Munich Dysphagia Test-Parkinson's Disease (MDT-PD), [56] and the Huntington's Disease Dysphagia Scale [57] 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 (≥ 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 fibreoptic 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-feeding[64] and reduction in aspiration pneumonia[65] 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 MND [80]. A recent systematic review by Goldberg and Altman [78] 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 Benatar [80] 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.

References

1 Williams A. Defining neurodegenerative diseases. *BMJ*. 2002; **324**:1465-1466.

2 Ang E T, Tai Y K, Lo S Q, Seet R, Soong T W. Neurodegenerative diseases: exercising toward neurogenesis and neuroregeneration. *Front. Aging Neurosci.* 2010; **2**: 1-8.

3 Forster A, Samaras N, Gold G, Samaras D. Oropharyngeal dysphagia in older adults: a review. *Eur Geriatr Med.* 2011; **2**: 356-362.

4 Stoschus B, Allescher HD. Drug induced dysphagia. *Dysphagia.* 1993; **8**: 154-159.

5 Robbins J, Levine R, Wood J, Roecker EB, Luschei E. Age effects on the temporal evolution of isometric and swallowing pressure. *J Gerontol A Biol Sci Med Sci*. 2000; **55**: M634-M640.

6 Aslam M, Vaezi M. Dysphagia in the elderly. *Gastroenterology and Hepatology NY*. 2013;**9**: 784-795.

7 Cowan JT, Roberts JD, Fitzpatrick JM, While AE, Baldwin J. Nutritional status of older people in long term care settings: current status and future directions. *Int J Nurs Stud.* 2004; **41**:225-237.

8 Elia M. Nutrition and health economics. *Nutrition.* 2006; **22**: 576-578.

9 Dand P, Sakel M. The management of drooling in motor neurone disease. *IJPN*. 2010; **16**: 560-564.

10 Marks L, Turner K, O'Sullivan J, Deighton B, Lees A. Drooling in Parkinson's disease: a novel speech and language therapy intervention. *Int J Lang Comm Dis*. 2001; **36**: 282–287.

11 Kalf JG, Smit AM, Bloem BR, Zwarts MJ, Munneke M. Impact of drooling in Parkinson's disease. *J Neurol.* 2007; **254**:1227–1232.

12. Carneiro D, De Sales Coriolano M, Belo R, De Marcos Rabelo AR, Asano AG, Lins OG. Quality of life related to swallowing in Parkinson's disease. *Dysphagia*. 2014: June E-Pub.

13 Ekberg O, Hamdy S, Woisard V, Wuttge-Hannig A, Ortega P. Social and psychological burden of dysphagia: its impact on diagnosis and treatment. *Dysphagia*. 2002;**17**:139– 46.

14 Verdonschot RJ, Baijens L, Serroyen J, Leue C, Kremer B. Symptoms of anxiety and depression associated with the Hospital Anxiety and Depression scale in patients with oropharyngeal dysphagia. *J. Psychosom. Res.* 2013; **75;** 451-455.

15 Paris G, Martinaud O, Petit A, Cuvelier A, Hannequin D, Roppeneck P, Verin E. Oropharyngeal dysphagia in amyotrophic lateral sclerosis alters quality of life**.** *J Oral Rehabil.* 2013; **40**:199-204.

16 Prince M, Bryce R, Albanese E, Wimo A, Ribeiro W, Ferri CP. The global prevalence of dementia: a systematic review and meta analysis. *Alzheimers Dement*. 2013; **9**: 63-75.

17 Alagiakrishnan K, Bhanji RA, Kurian M. Evaluation and management of oropharyngeal dysphagia in different types of dementia: a systematic review. *Arch Gerontol Geriatr*. 2013; **56**:1-9.

18 Hanson LC, Ersek M, Lin FC, Carey TS. Outcomes of feeding problems in advanced dementia in a nursing home population. *J Am Geriatr Soc.* 2013; **61**:1692-7.

19 Sato E, Hirano H, Watanabe Y, Edahiro A, Sato K, Yamane G, Katakura A. Detecting signs of dysphagia in patients with Alzheimer's disease with oral feeding in daily life. *Geriatr Gerontol Int.* 2013 Aug 29. Epub ahead of print.

20 Chouinard J, Lavigne E, Villeneuve C. Weight loss, dysphagia, and outcome in advanced dementia. *Dysphagia*. 1998; **13**: 151–155.

21 Suh M K, Kim H, Na D L. Dysphagia in patients with dementia: Alzheimer versus vascular. *Alzheimer Dis. Assoc. Disord*. 2009; **23**: 178-184.

22 Shinagawa S, Adachi H, Toyota Y. Mori T, Matsumoto I, Fukuhara R, Ikeda M. Characteristics of eating and swallowing problems in patients who have dementia with Lewy bodies. *Int Psychogeriatr*. 2009; **21**: 520-525.

23 Londos E, Hanxsson O, Hirsch I, Janneskog A, Bulow M, Palmqvist S. Dysphagia in Lewy body dementia - a clinical observational study of swallowing function by videofluoroscopic examination. *BMC Neurol*. 2013;**13**:140.

24 Ikeda M, Brown J, Holland A J, Fukuhara R, Hodges J. Changes in appetite, food preference, and eating habits in frontotemporal dementia and Alzheimer's disease. *J Neurol Neurosurg Psychiatry*. 2002; **73**: 371–376.

25 Langmore S, Olney R, Lomen-Hoerth C, Miller B. Dysphagia in patients with frontotemporal lobar dementia. *Arch. Neurol*. 2007; **64**:58–62.

26 Kalf JG, de Swart BJ, Bloem BR, Munneke M. Prevalence of oropharyngeal dysphagia in Parkinson's disease: a meta-analysis. *Parkinsonism Relat Disord*. 2012; **18**: 311–315.

27 Hammer M J, Murphy CA, Abrams TM. Airway somatosensory deficits and dysphagia in Parkinson's disease. *J Parkinsons Dis*. 2013; **1**:39-44.

28 Rodrigues B, Nobrega A, Sampaio M, Argolo N, Melo A. Silent saliva aspiration in Parkinson's disease. *Mov Disord*. 2011; 26:138-141.

29 Michou E, Hamdy S. Dysphagia in Parkinson's disease: a therapeutic challenge? *Expert Rev Neurother*. 2010;**10**:875-878.

30 Leow LP, Beckert L, Anderson T, Huckabee ML. Changes in chemosensitivity and mechanosensitivity in aging and Parkinson's disease. *Dysphagia*. 2012; **27**: 106-114.

31 Melo A, Monteiro L. Swallowing improvement after levodopa treatment in idiopathic Parkinson's disease: lack of evidence. *Parkinsonism Relat Disord* 2013;**19**:279 e81.

32 Nobrega AC, Pinho P, Deiro M, Argolo N. Levodopa treatment in Parkinson's disease: how does it affect dysphagia management? *Parkinsonism Relat Disord* 2014;**20**:340e133.

33 Fernandez HH, Lapane KL. Predictors of mortality among nursing home residents with a diagnosis of Parkinson's disease. *Med Sci Monit*. 2002;**8**:CR241-246.

34 Leow LP, Huckabee ML, Anderson T, Beckert L. The impact of dysphagia on quality of life in ageing and Parkinson's disease as measured by the Swallowing Quality of Life (SWAL-QOL) questionnaire. *Dysphagia*. 2010;**25**:216-220.

35 Miller N, Allcock L, Hildreth AJ, Jones D, Noble E, Burn DJ. Swallowing problems in Parkinson disease: frequency and clinical correlates. *J Neurol Neurosurg Psychiatry*. 2009; **80**: 1047-1049.

36 Golbe L. Diagnosis and management of progressive supranuclear palsy. *Neurodegener Dis Manag.* 2013:**3**; 81-90.

37 O'Sullivan SS, Massey LA, Williams DR, Silveira-Moriyama L, Kempster PA, Holton JL, Revesz T, Lees AJ. Clinical outcomes of progressive supranuclear palsy and multiple system atrophy. *Brain*. 2008; **131**:1362-1372.

38 Haverkamp LJ, Appel V, Appel SH. Natural history of amyotrophic lateral sclerosis in a database population. Validation of a scoring system and a model for survival prediction. *Brain.* 1995; **118**:707–719.

39 Leder SB, Novella S, Patwa H. Use of fiberoptic endoscopic evaluation of swallowing (FEES) in patients with amyotrophic lateral sclerosis. *Dysphagia*. 2003;**19**:177–181.

40 Regan J, Walshe M. Neuromuscular conditions. In Newman R, Nightingale JM, eds. *Videofluoroscopy: a multidisciplinary team approach*. San Diego: Plural, 2012: 177-185.

41 Heemskerk A, Roos RA. Dysphagia in Huntington's disease: a review. *Dysphagia*. 2011; **26**:62-66.

42 Kagel, MC, Leopold NA. Dysphagia in Huntington's disease: a 16 year retrospective. *Dysphagia.* 1992*;* **7:** 106–114.

43 Leopold NA, Kagel MC. Dysphagia in Huntington's disease. *Arch Neurol*. 1985;**42**:57-60.

44 Vogel AP, Pointon L, Maule R, Churchyard AJ. Clinical features of dysphagia and swallowing deficits in Huntington's disease: A descriptive videofluorscopic study.
Clinical Genetics. 2011; **80:** 60.

45 Hamakawa S, Koda C, Umeno H, Yoshida Y, Nakashima T, Asaoka K, Shoji H. Oropharyngeal dysphagia in a case of Huntington's disease. *Auris Nasus Larynx.* 2004; **31:**171-176.

46 Lanska DJ, Lanska MJ, Lavine L, Schoenberg BS. Conditions associated with Huntington's disease at death. A case-control study. *Arch Neurol*. 1988; **45**:878-880.

47 Heemskerk A, Roos RA. Aspiration pneumonia and death in Huntington's disease. *PLOS Currents Huntington Disease*. 2012; **1**:1-4.

48 Marandi Y, Farahi N, Sadeghi A, Sadeghi-Hashjin G. Review paper: Prion diseases – current theories and potential therapies: a brief review. *Folia Neuropathol.* 2012; **50**: 46-49

49 Hasegawa J, Okumura Y, Osumi E, Tago H, Katori Y, Kobayashi T. Creutzfeldt-Jakob disease with paralysis of the unilateral vocal cord and soft palate. *Tohoku J Exp Med.* 2011; **225**:277-283.

50 Mittal M, Hammond N, Husmann K, Lele A, Pasnoor M. Creutzfeldt-Jakob disease presenting as bulbar palsy. *Muscle Nerve*. 2010; **42:**833-835.

51 Iwasaki Y, Iijima M, Kimura S, Yoshida M, Hashizume Y, Yamada M, Kitamoto T, Sobue G. Autopsy case of sporadic Creutzfeldt-Jakob disease presenting with signs suggestive of brainstem and spinal cord involvement. *Neuropathology*. 2006;**26**:550-556.

52 Frank HG, Schnorf H, Genoud D, Pizzolato P, Glatzel M, Landis T. Creutzfeldt-Jakob disease presenting as isolated dysarthria and dysphagia due to pseudobulbar palsy. *Eur Neurol*. 2000;**44**:126-127.

53 Bird MR, Woodward MC, Gibson EM, Phyland DJ, Fonda D. Asymptomatic swallowing disorders in elderly patients with Parkinson's disease: a description of findings on clinical examination and videoflouroscopy in sixteen patients. *Age Aging*. 1994:**23**;351-254.

54 Fuh J, Lee R, Wang S, Lin C, Wang P, Chiang J, et al. Swallowing difficulty in Parkinson's disease. *Clin Neurol Neurosurg*. 1997;**99**:106-12.

55 Leder S, Suiter D. *The Yale Swallow Protocol: An evidence based approach to decision making*. Switzerland: Springer. 2014

56 Simons J, Fietzek U, Waldmann A, Warnecke T, Schuster T, Ceballos – Baumann A. Development and validation of a new screening questionnaire for dysphagia in the early stages of parkinson's disease. *Parkinsonism Relat Disord*. 2014. Early online. DOI: 10.1016/j.parkreldis.2014.06.008

57 Heemskerk A.-W, Verbist B, Marinus J, Heijnen B, Sjögren E, Roos R. The Huntington's disease dysphagia scale. Mov. Disord. 2014; epub doi: 10.1002/mds.25922

58 Kertscher B, Speyer R, Palmier M, Plant C. Bedside screening to detect oropharyngeal dysphagia in patients with neurological disorders: a systematic review. *Dysphagia* 2014;**29**:204-212.

59 Clave´ P, Arreola V, Romea M, Medina L, Palomera E, Serra-Prat M. Accuracy of the volume-viscosity swallow test for clinical screening of oropharyngeal dysphagia and aspiration. *Clin Nutr*. 2008;**27**:806–15.

60 Martino R, Silver F, Teasell R, Bayley M, Nicholson G, Streiner DL, Diamant NE. The Toronto Bedside Swallowing Screening Test (TOR-BSST): development and validation of a dysphagia screening tool for patients with stroke. *Stroke*. 2009;**40**:555–61.

61 Rosenbek J, Jones H. *Dysphagia in Movement Disorders*. San Diego: Plural. 2009.

62 Hamilton A, Heemskerk A, Loucas M, Twiston-Davies R, Matheson KY, Simpson SA, Rae D. Oral feeding in Huntington's disease: A guideline document for speech and language therapists. *Neurodegener Dis Manag*. 2012; **2**:45-53.

63 Lin L, Huang Y, Su S, Watson R, Tsai B, Wu S. Using spaced retrieval and Montessori-based activities in improving eating ability for residents with dementia. *Int. J. Geriatr. Psychiatry*. 2010; **25**: 953-959.

64 Bilney B, Morris M E, Perry A. Effectiveness of physiotherapy, occupational therapy, and speech pathology for people with Huntington's disease: a systematic review. *Neurorehabil Neural Repair.* 2003; **17**: 12-24.

65 Groher M, Crary M. *Dysphagia: Clinical management in adults and children*.. Mosby Elsevier: Missouri. 2010.

66 Deane K, Whurr R, Clarke CE, Playford ED, Ben-Shlomo Y. Non-pharmacological therapies for dysphagia in Parkinson's disease. *Cochrane Database of Systematic Reviews*. 2001; Issue 1. Art. No.: CD002816.

67 Ashford J, McCabe D, Wheeler-Hegland K, Frymark T, Mullen R, Musson N, Schooling T, Hammond CS. Evidence-based systematic review: Oropharyngeal dysphagia behavioral treatments. Part III--impact of dysphagia treatments on populations with neurological disorders. *J Rehabil Res Dev*. 2009;**46**:195-204.

68 Logemann JA, Gensler G, Robbins J, Lindblad AS, Brandt D, Hind J A, Kosek S, Dikeman K, Kazandjian M, Gramigna G, Lundy D, McGarvey-Toler S, Miller Gardner PJ. A randomized study of three interventions for aspiration of thin liquids in patients with dementia or Parkinson's disease. *J Speech Lang Hear Res* 2008; **51***:* 173-183*.*

69 Robbins J, Gensler G, Hind J, Logemann J A, Lindblad A S, Brandt D, Baum H, Lilienfeld D, Kosek S, Lundy D, Dikeman K, Kazandjian M, Gramigna G,

McGarvey-Toler S, Miller Gardner PJ. Comparison of 2 interventions for liquid aspiration on pneumonia incidence: A randomized trial. *Ann Intern Med.* 2008;**148** 509-518.

70 Troche MS, Okun MS, Rosenbek JC, Musson N, Fernandez HH, Rodriguez R, Romrell J, Pitts T, Wheeler-Hegland KM, Sapienza CM. Aspiration and swallowing in Parkinson disease and rehabilitation with EMST: a randomized trial. *Neurology*. 2010;75:1912-9.

71 Regan J, Walshe M, Tobin OW. Immediate effects of thermal-tactile stimulation on timing of swallow in idiopathic Parkinson's disease. *Dysphagia*. 2010; **25**:207-215.

72 Troche MS, Brandimore AE, Foote KD, Okun MS. Swallowing and deep brain stimulation in Parkinson's disease: a systematic review. *Parkinsonism Relat Disord.* 2013;**19**:783-788.

73 Young CA, Ellis C, Johnson J, Sathasivam S, Pih N. Treatment for sialorrhea (excessive saliva) in people with motor neuron disease/amyotrophic lateral sclerosis. *Cochrane Database of Systematic Reviews*. 2011; Issue 5. Art. No.: CD006981.

74 Lagalla G, Millevolte M, Capecci M, Provinciali L, Ceravolo MG. Botulinum toxin type A for drooling in Parkinson's disease: A double-blind, randomized, placebo-controlled study. *Mov. Disord*. 2006; **21**: 704–707.

75 Langmore SE, Grillone G, Elackattu A, Walsh M. Disorders of swallowing: palliative care. *Otolaryngol Clin North Am.* 2009;**42**:87-105.

76 Logemann JA, Curro FA, Pauloski B, Gensler G. Aging effects on oropharyngeal swallow and the role of dental care in oropharyngeal dysphagia. *Oral Dis*. **19**: 733-737.

77 §Ortega O, Parra C, Zarcero S, Nart J, Sakwinska O, Clave P. Oral health in older patients with oropharyngeal dysphagia. *Age Ageing*. **43**:132-137.

78 Goldberg L, Altman K. The role of gastrostomy tube placement in advanced dementia with dysphagia: A critical review. *Dysphagia*. 2012; **27**: 596-597.

79 Candy B, Sampson EL, Jones L. Enteral tube feeding in older people with advanced dementia: findings from a Cochrane systematic review. *IJPN*. 2009; **15**: 396-404.

80 Katzberg HD, Benatar M. Enteral tube feeding for amyotrophic lateral sclerosis/motor neuron disease. *Cochrane Database of Systematic Reviews.* 2011; 1. Art. No.: CD004030.

81 Michou E, Baijens L, Rofes L, Cartgena PS, Clave P. Oropharyngeal swallowing disorders in Parkinson's disease: Revisited. *Int. J. Speech Lang. Pathol. Audiol.* 2013;**1**:76-88.

82 Horner J, Alberts M J, Dawson, DV, Cook GM. Swallowing in Alzheimer's disease. *Alzheimer Dis. Assoc. Disord*. 1994; **8**:177–189.

83 Vanderwegen J, Van Vlasselaer K. Clinical swallowing evaluation of dementia of the alzheimer type (DAT) in different stages of disease severity. *Dysphagia.* 2013; **28:** 311

84 Priefer BA, Robbins J. Eating changes in mild-stage Alzheimer's disease: a pilot study. *Dysphagia*. 1997;**12**:212-21.

85 Humbert I.A, McLaren D.G, Kosmatka K, Fitzgerald M, Johnson S, Porcaro E, Kays S. Umoh E-O, Robbins J. Early deficits in cortical control of swallowing in Alzheimer's disease. *J Alzheimer's Dis*. 2010; **19**:1185-1197.

86 Horner J, Alberts M J, Dawson D V, Cook G M. Swallowing in Alzheimer's disease. *Alzheimer Dis Assoc Disord*. 1994; **8**:177–189.

86 Stroudley J, Walshe M. Radiological assessment of dysphagia in Parkinson's disease. *Br J Radiol*. 199;**64**:890-893.

87 Ali GN, Wallace KL, Schwartz R, DeCarle DJ, Zagami AS, Cook IJ. Mechanisms of oral-pharyngeal dysphagia in patients with Parkinson's disease. *Gastroenterology.* 1996;**110**:383-92.

88 Leopold NA, Kagel MC. Dysphagia in progressive supranuclear palsy: radiologic features. *Dysphagia*. 1997;**12**:140-3

89 Higo R, Nito T, Tayama N. Swallowing function in patients with multiple-system atrophy with a clinical predominance of cerebellar symptoms (MSA-C). *Eur Arch Otorhinolaryngol.* 2005;**262**:646-50.

90 Easterling C, Antinoja J, Cashin S, Barkhaus PE. Changes in tongue pressure, pulmonary function, and salivary flow in patients with amyotrophic lateral sclerosis. *Dysphagia*. 2013; **28**:217-25.

91 Kawai S, Tsukuda M, Mochimatsu I, Enomoto H, Kagesato Y, Hirose H, Kuroiwa Y, Suzuki Y. A study of the early stage of Dysphagia in amyotrophic lateral sclerosis. *Dysphagia*. 2003;**18**:1-8.

92 Graner DE, Strand E. Management of dysarthria and dysphagia in patients with amyotrophic lateral sclerosis. *Perspectives on Neurophysiology and Neurogenic Speech and Language Disorders*. 2010;**20**:39-44.

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 Rapid compulsive eating Poor monitoring of bolus size Impaired consciousness and alertness Food refusal	Difficulty self-feeding Poor bolus propulsion Reduced bolus control	Reduced bolus pressure Reduced airway protection Incomplete bolus clearance	Increased oral, and pharyngeal residue Silent aspiration	Oral and pharyngeal phase disruption Poor coordination of respiration and swallowing

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] Inccordination 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]