

# Correlation of dysphagia with functional decline in neurodegenerative disease - a review

## Abstract

**Hypothesis:** The functional decline noted over time in patients with neuromuscular diseases is associated with increasing risk of dysphagia and aspiration.

**Study design:** Systematic literature review.

**Methods:** A Pub Med search to identify all of the literature addressing neuromuscular disease progression and correlation with dysphagia was performed in the English literature from 1/1/2000 through 4/1/2015. Studies with outcomes measures for both functional decline and dysphagia or aspiration were included, whereas stroke and head injury were excluded.

**Results:** Of the 309 articles identified in the search, 13 discussing Parkinson's disease PD and 5 on the topic of Amyotrophic Lateral Sclerosis ALS met the criteria. No papers on the subjects of Multiple Sclerosis MS or Huntington's disease HD were identified. Articles were stratified by level of evidence using the criteria of Sackett et al.,<sup>1</sup> noting instrument selection for disease state and dysphagia, self-reported vs. observed, outcome and limitations. There were 8 true longitudinal studies, and the other 10 stratified patients by duration or disease severity. Overall, the studies support increased dysphagia or swallowing related quality of life decline in patients with advanced PD and ALS. However, most studies are limited by 1) the cross-sectional nature instead of true longitudinal design, 2) self-reported instruments and 3) infrequent mention of aspiration risk.

**Conclusion:** Progressive dysphagia and aspiration risk is weakly demonstrated with advancing PD and ALS. Although these are similar known risks with MS and HD, there are only anecdotal reports. A systematic approach at screening these patients for dysphagia is warranted.

**Keywords:** dysphagia, neurodegenerative disease, als, huntington's disease, parkinson's disease

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**Abbreviations:** PD, parkinson's disease; ALS, amyotrophic lateral sclerosis; MS, multiple sclerosis; MBSS, modified barium swallow study; MRC, medical research council; ALSFRS, als functional rating system

## Introduction

Neurodegenerative diseases including Parkinson's disease PD, Amyotrophic Lateral Sclerosis ALS, Multiple Sclerosis MS, and Huntington's disease HD are characterized by a progressive decline in functional status.<sup>2-20</sup> The progression of dysphagia in neurodegenerative disease has been well described by Hadjikoutis et al.,<sup>21</sup> with ALS. The voluntary swallow is affected first; initially the swallow reflex is delayed and eventually it degenerates entirely. Spontaneous reflexive swallows are lost typically in the pre-terminal stage of the disease. Malnutrition becomes an issue, which in turn worsens coordination and strength of the deglutition muscles, as well as muscles required for respiratory function. As the disease progresses, oral hygiene often deteriorates as it is more difficult for patients to perform self-care such as teeth brushing, as well as difficulty clearing their oral secretions. Thus food particles remain and can increase oral bacterial load, increasing the risk of aspiration pneumonia.<sup>21</sup>

Dysphagia often goes unnoticed until a catastrophic event occurs, such as an aspiration event or aspiration pneumonia. Catastrophic events result in negative outcomes for the patient, including inability to take needed medicines, prolonged hospitalizations, deconditioning,

and malnutrition.<sup>22</sup> The debilitating natural progression of neurodegenerative disease introduces an added degree of difficulty to recover from such events. Patients, families and care teams are then faced with the decision to pursue alternative means of nutrition and hydration given the severity of dysphagia. Thus it is highly beneficial to patients, their families, and health care systems to have established protocols to screen patients at high aspiration risk for dysphagia.<sup>22</sup> In addition to the life-threatening consequences of dysphagia and aspiration with neuromuscular disease, quality of life is threatened as eating becomes a stressful event rather than a pleasurable experience.<sup>5</sup> The social stresses are severe as meal times are quintessential to personal bonding and professional networking.<sup>23</sup>

For all of these reasons, it is vital to develop a standard practice to detect dysphagia in patients with neurodegenerative disease early on, when dietary changes and behavioral training can extend quality of life.<sup>12-21</sup> Thus we sought to identify and quantify relationships between disease progression and dysphagia progression and/or aspiration risk in patients with neurodegenerative disease through a systematic review of the existing literature. Our hypothesis is that overall functional decline in neuromuscular disease over time is associated with worsening dysphagia and aspiration risk.

## Methods

A Pub Med search over the time period 1/1/2000 through 4/1/2015 was performed for the following terms: neuromuscular disease or

Parkinson's or PD or ALS or amyotrophic lateral sclerosis or HD or Huntington's disease or Huntington's chorea or MS or multiple sclerosis or movement disorder AND functional status or functional decline or decline or activities of daily living or ADL or quality of life or QOL AND dysphagia or swallowing or aspiration or malnutrition or malnourishment. The inclusion criteria were that an article must:

- I. Address a neuromuscular disease (PD, ALS, HD, MS) in regards to functional status or quality of life and dysphagia, and
- II. Must be written in English. The exclusion criteria were:
  - a. studies published before 2000,
  - b. articles not available in full text,
- III. Basic science articles,
- IV. Animal models,
- V. Review articles, and
- VI. Subjects primarily or confounded by stroke or head injury.

Selected articles were tabulated and compared with regard to level of evidence, number of subjects, follow up period, objective and subjective measures used to assess functional status, and measure of dysphagia.

## Results

The initial search yielded 309 articles. After applying inclusion and exclusion criteria, 18 articles remained (see process in PRISMA diagram in Figure 1. Table 1 describes the instruments used to assess the extent of the neuromuscular diseases encountered in the course of the review, including information source and limitations. Table 2 does the same for instruments used to measure dysphagia relative to this review. Of the 18 articles available for qualitative review, 13 were specific for PD, and 5 were specific for ALS. The search did not yield any papers that studied the progression of Multiple Sclerosis or Huntington's disease over time with respect to functional status or dysphagia. Table 3 summarizes the findings of the 18 papers reviewed in this study. A formal meta-analysis was not able to be performed based on the spectrum of study criteria.

**Table 1** Functional status instruments in neuromuscular diseases

Neuromuscular Disease Instrument	Self-Reported Vs. Observed Vs. Measured	Reference/Yr	Description	Limitation
PD Questionnaire (PDQ) 39	Self-reported	Walker et al. <sup>4</sup>	39 questions rating no difficulty with ADLs (0) to always have difficulty (100).	Subjective
15 item Geriatric Depression Scale	Self-reported	Miller & Weintraub et al. <sup>9,36</sup>	30 yes/no questions rating no depression ( $\leq 5$ ) to severe depression ( $=15$ ).	Subjective
Hoehn & Yahr (HY) scale	Observed	Carneiro & Hoehn et al. <sup>3,20</sup>	Stages I-V ranging from no functional impairment (I) to inability to ambulate independently (V).	No half stages. Progression not necessarily linear.
ALS-Functional Rating Score (FRS)	Self-reported	Voustianiouk & Magnus et al. <sup>18,33</sup>	Rating lowest functioning (0) to normal (40) based on 10 question survey.	Does not weigh respiratory dysfunction the same as limb/bulbar function <sup>32</sup>
ALSFRS-R	Observed or self-reported	Cedarbaum & Nakamura et al. <sup>16,35</sup>	Health care provider determines functional status rating lowest functioning (0) to normal functioning (48).	Lacking correlation between FVC% and respiratory subscale <sup>32</sup>
Medical Research Council (MRC) for neck flexors	Observed	Nakamura et al. <sup>16</sup>	Full clinical exams by neurologists.	Rater-dependent
Medical Research Council (MRC) scale	Observed	Chiò et al. <sup>18</sup>	Scored 0-5. Tests neck flexors & extensors, and upper & lower limb muscles	
Medical Research Council (MRC) Compound Score	Self-reported	Magnus et al. <sup>17</sup>	9-step modified MRC scale	
Percent Predicted Forced Vital Capacity (FVC) %	Measured	Magnus et al. <sup>17</sup>	Assess lung function	Dependent on subject's effort
Mini Mental State Exam (MMSE)	Observed	Walker et al. <sup>4</sup>	Score 0-30 <22 = cognitive impairment, may need caretaker to answer questions	Weakly detects mild cognitive impairment
Schwab and England Activities of Daily Living (ADL)	Self-reported or observed	Auyeung et al. <sup>11</sup>	Bed-ridden (0) to independent (100). $\leq 30$ = dependent. $\geq 80$ = independent. Measured drug "on".	Subjective
Beck Depression Inventory (BDI)	Self-reported	Plowman-Prine et al. <sup>6</sup>	21 domains rating depression: none (0) to most severe (63).	Subjective
Non-motor symptom scale	Self-reported	Guo et al. <sup>3</sup>	Assess severity/ frequency of NMS	Subjective
ALS severity score	Self-reported	del Aguila et al. <sup>19</sup>	Self-reported performance in speech, swallow, lower & upper extremity function rating worst (0) to best (40).	Subjective

Table Continued

Neuromuscular Disease Instrument	Self-Reported Vs. Observed Vs. Measured	Reference/Yr	Description	Limitation
El Escorial Diagnostic Criteria (EEDC)	Observed	Chiò et al. <sup>18</sup>	Standard tool to assess ALS status. Includes signs of progressive upper and lower motor neuron degeneration	
Unified PD Rating Scale (UPDRS)	Self-report + clinical observation	Carneiro & Ramaker et al. <sup>2,26</sup>	Part I: evaluate mentation, mood, behavior Part II: self-eval ADLs Part III: clinician-scored monitored motor eval Part IV: HY staging Part V: Schwab and England ADL scale	Redundancy with several items focusing on the same construct
Hospital Anxiety and Depression (HAD) scale	Self-reported	Walker & Zigmond et al. <sup>4,30</sup>	Questions assessing anxiety and depression rating from 0-21, >8 = abnormal	Subjective
Frontal Assessment Battery (FAB)	Observed	Antonini & Dubois et al. <sup>10,31</sup>	Bedside test of executive functioning [28]	
Hamilton Depression Scale (HAM-D)	Self-reported	Antonini & Hamilton et al. <sup>10,32</sup>	17 variables rating depression from absent (0) to severe (4)	Interviewer dependent. Only for patients with diagnosed depressive disorder [28]
Health-related QOL using the Medical Outcomes Study 36-Item Short-Form General Health Survey 16, and Schedule for the Evaluation of Individual QOL-Direct Weighting [15]	Self-reported	Speyer <sup>28</sup>	Dysphagia's impact on quality of life as per the patient	Subjective
Norris ALS disability scale	Observed	Chio et al. <sup>18</sup>	Tracks change in impairments & disability measures after ALS treatment	
Appel Rating Scale	Observed	Chio & Appel et al. <sup>18,34</sup>	Assess swallow, speech, and respiratory function rating normal (30) to maximum dysfunction (164)	
Phonation time as reflection of vital capacity	Observed	del Aguila et al. <sup>19</sup>	Completed 3 times, longest trial analyzed	

**Table 2** Swallow assessment instruments encountered in our review pertaining to neuromuscular diseases

Dysphagia Instrument	Self-Reported Vs. Observed Vs. Measured	Reference	Description	Limitation
SWAL-QOL	Observed	McHorney et al. <sup>25</sup>	44 questions that evaluate 11 domains of QOL: burden, eating duration/desire, sx frequency, food selection, communication, fear, mental health, social, fatigue, sleep	Subjective
Swallow fnx assessed by physician w/ 7-grade scale	Observed	Waxman et al. <sup>26</sup>	Dysphagia rating normal function (0) to severe dysphagia (6)	
Trial swallow using water	Observed	Speyer <sup>28</sup>	Patient swallows 150 ml of water as quickly as possible in "off drug" state	
Kennedy et al.'s Modified dysphagic rating scale (mDRS)	Observed/self-reported	Kennedy et al. <sup>27</sup>	12 items rating dysphagia worst (1) to normal (5)	
Speech/swallowing difficulty	Observed/self-reported	Auyeung et al. <sup>11</sup>	Assessing for presence & time to onset yearly	
Electromyography (EMG)	Measured	Potulska et al. <sup>13</sup>	Electrodes taped over swallow muscles, then patients swallow dry and 1-20 ml of water	
Scintigraphy of esophageal activity	Measured	Speyer <sup>28</sup>	Gamma camera tracks aspiration and bolus movement; degree of reflux	
Modified barium swallow with videofluoroscopy	Measured	Monte et al. <sup>14</sup>	Quantifies swallow abnormalities	Only captures a moment of swallow function

**Table 3** Patients with Parkinsonism were distributed as follows: 17 PD, 15 MSA, 14 Lewy body dementia, 13 Corticobasal degeneration, 24 progressive supranuclear pals

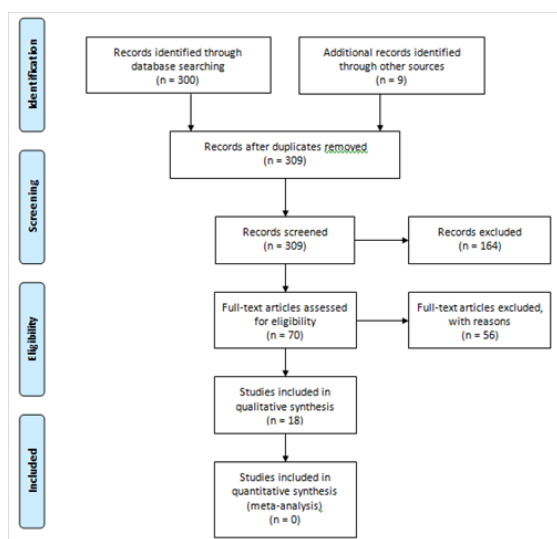
Author	Year	Dz	LOE	F/u Period, Time Range	Subjects	Outcome Measures	Major Findings	Limitations
Carneiro et al. <sup>2</sup>	2014	PD	4	1 snap shot	103	SWAL-QOL: 44 questions that evaluate 11 domains of QOL HY: classification performed by neurologist at Pro-Parkinson program UPDRS: self-report + clinical observation Non-motor symptom scale: assess severity/frequency of NMS	HY stage progresses with PD PD progression worsens swallowing QOL, specifically for eating duration, symptom frequency, and sleep.	Dysphagia self-reported, no objective measure.
Guo et al. <sup>3</sup>	2013	PD	4	1 snap shot	616	HY for disease severity Standard questionnaire on dx delay, disease duration, anti-PD meds UPDRS part 3 to assess motor disability PDQuestionnaire-39: disease duration Mini-Mental State Examination Hospital Anxiety and Depression scale HY stages UPDRS on meds	Non-motor symptoms (swallowing) increased with disease progression	Cross sectional study does not necessarily show progression.
Walker et al. <sup>4</sup>	2011	PD	4	1 snap shot	75	HY Stages SWAL-QOL "on" meds	32% of patients had dysphagia, which correlated with quality of life but not with disease duration	Self reported dysphagia, need objective measure.
Leow et al. <sup>5</sup>	2010	PD	4	1 snap shot	68	HY Stages SWAL-QOL "on" meds	Disease progression decreases QOL. Later-stage PD = decreased appetite, difficulty w/ food selection, prolonged meal time, stress surrounding social meals	Results may be influenced by confounding variables such as depression.
Plowman-Prine et al. <sup>6</sup>	2009	PD	4	1 snap shot	36	HY Stages SWAL-QOL PDQ-39, Beck Depression Inventory UPDRS "on" meds UPDRS	No relation between swallow-specific QOL + disease duration /severity. Dysphagia significantly reduces total SWAL-QOL.	Dysphagia vs. nondysphagia patients compared and then checked for HY. Not a longitudinal study.
Lorefält et al. <sup>7</sup>	2005	PD	2	1 year at 2 time points	52	Swallow assessed by physician with 7-grade scale: normal to severe dysphagia as per Waxman et al. 1990 Patient-reported latencies to dysphagia	No significant changes in dysphagia in PD patients over 1 year	PD may not have progressed in that time.
Müller et al. <sup>8</sup>	2001	PD	4	Retrospective post-mortem	83*	National Institute of Neurological Disorders and Stroke for PSP dx Neurologist recorded time to speech disorder Survival time	Dysphagia stays latent for ~84 months in PD, survival time post-dysphagia complaint onset ~15-24 mo. Total survival time correlated to latency to complaint of dysphagia.	Retrospective nature precludes neurologists evaluating patients to use agreed upon protocol.
Miller et al. <sup>9</sup>	2008	PD	4	1 snap shot	137	Geriatric Depression Scale HY stages MMSE Patients swallow 150 ml of water as quickly as possible "off" meds UPDRS "off" meds	Moderate correlation between swallowing rate and disease severity, but not between swallowing speed and disease duration. Self-report is not reliable. Need objective assessment of aspiration risk to compare with water swallow test performance	Did not follow patients over time.
Antonini et al. <sup>10</sup>	2012	PD	2	2 years	707	PDQ-39 HY MMSE Frontal Assessment Battery Hamilton Depression Scale UPDRS part III	GI symptoms (dysphagia) worsened + became more prevalent over 2 years. Non-motor symptom progress is symptom specific, does not follow motor deterioration. Only some symptoms negatively impact QOL	Didn't assess NMS severity, only presence/ absence.

Table Continued

Author	Year	Dz	LOE	F/u Period, Time Range	Subjects	Outcome Measures	Major Findings	Limitations
Auyeun et al. <sup>11</sup>	2012	PD	2	11.3+/-2.6 years	171	UPDRS HY MMSE Schwab and England ADL 'on' meds	After 10 years, 60% of new onset PD patients developed dysphagia	Clinic based- missing PD patients cared for by primary care. Hong Kong pop might not represent China/world.
Volonté et al. <sup>12</sup>	2002	PD	4	1. snap shot	65	UPDRS Modified dysphagic rating scale (mDRS)	mDRS correlated with UPDRS score, but not illness duration. Assess swallowing abilities in IPD patients in clinic because w/ dietary advice and short rehab training can improve QOL.	Only looked at early PD patients.
Potulska et al. <sup>13</sup>	2003	PD	4	1 snap shot	40	HY stages UPDRS "on" 2 hour post L-dopa EMG Scintigraphy of esophageal activity	Dysphagia in all PD patients but not all had symptoms. Related to disease progression.	Did not follow patients over time, small sample size.
Monte et al. <sup>14</sup>	2005	PD	4	1. snap shot	34	HY stages UPDRS II & III Modified barium swallow	Levodopa may help swallowing function. Dysphagia does not reliably predict deglutition alterations in PD. 'Silent aspirators' w decreased cough reflex/ aspiration awareness, need video fluoroscopy to detect problems early	Sample size may be too small to show statistically significant differences.
Chiò et al. <sup>15</sup>	2015	ALS	2	1 year	130	ALSFRS HRQOL Proposed ALS-MITOS	70/118 progressed disease stage in 12 months. Functional (ALSFRS) and QOL measures inversely related to disease stage.	Short follow up. Couldn't assess stage transitions. Small sample size. Unsure of time spent in baseline stage pre-study entry as stages were not retrospectively assessed.
Nakamura et al. <sup>16</sup>	2013	ALS	2	1 year	401	Medical Research Council for neck flexors ALSFRS-R Japanese version of ALSFRS-R scale	Neck flexor strength = best indicator of loss of swallowing function. Mod and significant correlation between MRC score for neck flexors and ALSFRS-R score.	Registered patients follow up by phone, did not examine longitudinal changes in strength of multiple muscles.
Magnus et al. <sup>17</sup>	2002	ALS	2	1 year	155	ALS-Functional Rating Score (ALS-FRS) Medical Research Council (MRC) Compound Score FVC%	Longer survival with younger onset (<60 years old), limb-onset (vs. bulbar) ALS-FRS, FVC%, and MRC compound score declines predict survival.	Short follow up period. 12-36 months. Future study: if nonspecific factors (dysphagia) influence survival prediction.
Chio et al. <sup>18</sup>	2002	ALS	2	2 years	221	Standard questionnaire for demographic info, clinical history, EEDC classification, neuro & lab findings, treatment details Norris ALS disability scale Bulbar section of Appel Rating Scale FVC%	Bulbar onset has poorest prognosis. Patients w/ PEG do better. Symptom progression rate in early ALS predicts disease outcome.	Only 51% of patients included in the study had definite ALS.
Del Aguila et al. <sup>19</sup>	2003	ALS	2	9 years	180	ALS severity score Self-reported performance in speech/ swallow/lower & upper extremity function Phonation time used to measure vital capacity Medical Outcomes Study Short Form 36 (SF-36)	Median survival 32 months from symptom onset, 19 months from diagnosis. 7% 5-year survival from diagnosis. Poor prognosis: older age at onset, bulbar onset, female, unmarried, results. longer time from symptom onset to diagnosis.	More data points would strengthen results.

**Abbreviations:** ALS: amyotrophic lateral sclerosis; Dz: Disease; HY: Hoehn and Yahr; LOE: Level of Evidence; MMSE: Mini Mental State Exam; PD: Parkinson's disease; QOL: Quality of Life





**Figure 1** PRISMA Flow Diagram.

Nine of the articles that addressed dysphagia in PD studied patients at one point in time, and compared subjects based on Hoehn and Yahr (HY) stage (5 studies 2-14), disease duration 1 study,<sup>3</sup> both HY stage and disease duration 2 studies,<sup>4,9</sup> or just at time of study enrollment early in PD disease 1 study.<sup>12</sup> Three of the studies followed patients every few months for 1-10 years.<sup>7-11</sup> One study was retrospectively done by reviewing charts post-mortem.<sup>8</sup> The subject count ranged from 34 to 707 people.

Six articles found that dysphagia is worse at more advanced stages of Parkinson's disease.<sup>2-13</sup> One article that followed patients over the course of 2 years found that gastroenterology GI disturbances (dysphagia) became more prevalent in this period, but it was not directly linked to disease progression necessarily.<sup>10</sup> An article that followed patients with Parkinson's disease for 10 years found that at the end of the time period, 60% of patients had developed dysphagia, but it did not comment specifically on the development of dysphagia with regards to disease progression.<sup>11</sup> Two articles found no correlation between dysphagia and disease duration.<sup>4-6</sup> One article that followed patients over the course of a year found no significant changes in dysphagia, but there may have not been any significant changes in disease progression.<sup>7</sup>

The search also yielded 5 studies on disease progression in ALS.<sup>15,16</sup> Chio et al.,<sup>15</sup> showed that the majority of patients progressed to a higher stage of disease in 12 months as quantified by the ALS Functional Rating System (ALSFRS) and by Quality of Life (QOL) measures. Nakamura et al.,<sup>16</sup> used the Medical Research Council (MRC) score to rate neck flexor muscle strength in relation to ALS disease progression. They found that neck flexor strength is the most significant prognostic factor for loss of swallowing function and that MRC score is significantly correlated to ALSFRS-R score.<sup>16</sup> An additional 3 studies on ALS progression were included which did not have dysphagia-specific outcomes measures beyond those included in general self-reporting or those measures included in the ALS inventories. These studies show that onset of ALS is predictive of survival, specifically younger age and limb instead of bulbar involvement at onset predicts longer lifespan.<sup>17-19</sup> Additional protective factors include being male, married, and diagnosed quickly after symptom onset.<sup>18</sup> Chiò et al.,<sup>18</sup> found that PEG tube insertion often results in longer survival.<sup>18</sup> Magnus et al.,<sup>17</sup> found that ALS-

FRS, functional vital capacity (FVC%), and MRC compound score were predictive of survival. All five studies on ALS followed patients for no more than 2 years.

## Discussion

It is intuitive to expect that as patients with neuromuscular and neurodegenerative disease have declining functional status, they also have a concomitant decline in the quality of their swallow mechanism with increased aspiration risk. Although there is significantly different pathophysiology with the neuromuscular diseases addressed in our review, common manifestations include the progressive nature of decline, increasing sedentary behavior with reduced respiratory function and pulmonary toilet, the increasing prevalence of dysphagia, and aspiration as a common precipitating terminal event. Our review reveals some correlation between functional decline and dysphagia with some neurodegenerative diseases (PD and ALS), however a quantifiable association and true longitudinal nature of this relationship was not able to be assessed.<sup>4-9</sup> Functional status and dysphagia are both difficult to objectively measure in patients with neurodegenerative disease for a wide variety of reasons. Methods or surveys (instruments) can be categorized as self-reported vs. observed, which each carry with them their own unique set of limitations. Self-reporting (subjective instruments) is not fully reliable as a means of identifying dysphagia, or even functional status with neurodegenerative disease.<sup>9</sup> There may be cognitive impairments associated with the neurodegenerative disease that may impact self-reporting. Studies have shown inconsistencies between patients' actual swallowing abilities and perceived difficulties.<sup>9-14</sup> Additionally, patients may be fearful of losing their independence and pleasure in eating if they reveal symptoms of possible aspiration.<sup>5</sup> There is also variability in the instrument reported by the patient versus a close family member.

Observation is more objective and scientifically controlled, but usually only captures one snapshot of time.<sup>14</sup> This is a major limitation to fully understanding the extent of neurodegenerative diseases as their effects fluctuate based on time of day and medication dosage. Observing patients in drug "on" vs. "off" states as with PD may yield vastly different data points. Furthermore, the concepts of inter-rater and intra-rater variability are similar limitations with such a scale. In addition to clinician-observed scales, more objective information on functional status and dysphagia may be garnered from objective diagnostic testing, such as modified Barium swallow study (MBSS) to detect dysphagia and aspiration based on varying food consistencies and compensatory swallow maneuvers.<sup>24</sup> These studies may show progression that may not yet be clinically relevant.<sup>8</sup>

There is a great need for consistent clinical use of a meaningful dysphagia rating scale early in the neurodegenerative disease process in order to improve patient and caregiver education on the importance of maintaining nutrition while realizing aspiration risk.<sup>4</sup> More objective measures with longitudinal studied in the literature are necessary to paint an accurate clinical picture of patients' swallowing ability to recognize "sentinel indicators of decline" to reduce likelihood of dangerous complications including aspiration and pneumonia.<sup>2-14</sup> Detecting dysphagia early is also important because patients can undergo dietary changes and rehabilitation that can significantly improve their quality of life.<sup>12</sup> One post-mortem study on PD patients documented longer total survival time when there was a longer latency period between clinically assessed initial dysphagia development and patient awareness of its complications.<sup>8</sup> Therefore, by identifying a stronger correlation between functional decline in these diseases and dysphagia with aspiration risk, clinicians may more

accurately predict when swallowing intervention would proactively be needed.<sup>25–30</sup>

None of the studies identified in our review were able to determine if dysphagia is a prognostic indicator of survival. Therefore, there is also a great need for a prospective study to follow a cohort of these patients and determine the effects of dysphagia on survival outcomes. Longitudinal studies to better define the devastating natural history of the neuromuscular diseases such as those associated with dysphagia would further empower care teams in order to better meet patient needs to enhance quality of life.<sup>30–36</sup>

## Conclusion

Neuromuscular and neurodegenerative diseases are often associated with dysphagia and risk of aspiration. This review documents some proven correlation with declining functional status and dysphagia in PD and ALS. However, correlation over time in longitudinal studies, and extension of this concept to other diseases such as MS and HD was not found in the existing literature. A more cohesive prospective approach at recognizing dysphagia in this important population is warranted.

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## Conflicts of interest

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## References

- Isaac CA, Franceschi A. Evidence-based medicine: how to practice and teach EBM. Toronto, Ontario: *Churchill*. 2008;14(15):659–659.
- Carneiro D, das Graças Wanderley de Sales Coriolano M, Belo LR, de Marcos Rabelo AR, et al. Quality of life related to swallowing in Parkinson's disease. *Dysphagia*. 2014;29(5):578–582.
- Guo X, Song W, Chen K, et al. Disease duration-related differences in non-motor symptoms: a study of 616 Chinese Parkinson's disease patients. *J Neurol Sci*. 2013;330(1–2):32–37.
- Walker RW, Dunn JR, Gray WK. Self-reported dysphagia and its correlates within a prevalent population of people with Parkinson's disease. *Dysphagia*. 2011;26(1):92–96.
- Leow LP, Huckabee ML, Anderson T, et al. 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(3):216–220.
- Plowman-Prine EK, Sapienza CM, Okun MS, et al. The relationship between quality of life and swallowing in Parkinson's disease. *Mov Disord*. 2009;24(9):1352–1358.
- Lorefält B, Granérus AK, Unosson M. Avoidance of solid food in weight losing older patients with Parkinson's disease. *J Clin Nurs*. 2006;15(11):1404–1412.
- Müller J, Wenning GK, Verny M, et al. Progression of dysarthria and dysphagia in postmortem-confirmed parkinsonian disorders. *Arch Neurol*. 2001;58(2):259–264.
- Miller N, Allcock L, Hildreth AJ, et al. Swallowing problems in Parkinson disease: frequency and clinical correlates. *J Neurol Neurosurg Psychiatry*. 2009;80(9):1047–1049.
- Antonini A, Barone P, Marconi R, et al. The progression of non-motor symptoms in Parkinson's disease and their contribution to motor disability and quality of life. *J Neurol*. 2012;259(12):2621–2631.
- Auyeung M, Tsoi TH, Mok V, et al. Ten year survival and outcomes in a prospective cohort of new onset Chinese Parkinson's disease patients. *J Neurol Neurosurg Psychiatry*. 2012;83(6):607–611.
- Volonté MA, Porta M, Comi G. Clinical assessment of dysphagia in early phases of Parkinson's disease. *Neurol Sci*. 2002;23(Suppl 2):S121–S122.
- Potulska A, Friedman A, Królicki L, et al. Swallowing disorders in Parkinson's disease. *Parkinsonism Relat Disord*. 2003;9(6):349–353.
- Monte FS, da Silva-Júnior FP, Braga-Neto P, et al. Swallowing abnormalities and dyskinesia in Parkinson's disease. *Mov Disord*. 2005;20(4):457–462.
- Chiò A, Hammond ER, Mora G, et al. Development and evaluation of a clinical staging system for amyotrophic lateral sclerosis. *J Neurol Neurosurg Psychiatry*. 2005;86(1):38–44.
- Nakamura R, Atsuta N, Watanabe H, et al. Neck weakness is a potent prognostic factor in sporadic amyotrophic lateral sclerosis patients. *J Neurol Neurosurg Psychiatry*. 2013;84(12):1365–1371.
- Magnus T, Beck M, Giess R, et al. Disease progression in amyotrophic lateral sclerosis: predictors of survival. *Muscle Nerve*. 2002;25(5):709–714.
- Chiò A1, Mora G, Leone M, et al. Early symptom progression rate is related to ALS outcome: a prospective population-based study. *Neurology*. 2002;59(1):99–103.
- del Aguila MA, Longstreth WT, McGuire V, et al. Prognosis in amyotrophic lateral sclerosis: a population-based study. *Neurology*. 2003;60(5):813–819.
- Hoehn MM, Yahr MD. Parkinsonism: onset, progression, and mortality. 1967. *Neurology*. 2001;57(10 Suppl 3):S11–S26.
- Hadjikitoutis S, Wiles CM. Respiratory complications related to bulbar dysfunction in motor neuron disease. *Acta Neurol Scand*. 2001;103(4):207–213.
- Altman KW, Richards A, Goldberg L, et al. Dysphagia in stroke, neurodegenerative disease, and advanced dementia. *Otolaryngol Clin North Am*. 2013;46(6):1137–1149.
- Altman KW. Understanding dysphagia: a rapidly emerging problem. *Otolaryngol Clin North Am*. 2013;46(6): xiii–xvi.
- Rosenbek JC, Robbins JA, Roecker EB, et al. A penetration-aspiration scale. *Dysphagia*. 1996;11(2):93–98.
- McHorney CA, Robbins J, Lomax K, et al. The SWAL-QOL and SWAL-CARE outcomes tool for oropharyngeal dysphagia in adults: III. Documentation of reliability and validity. *Dysphagia*. 2002;17(2):97–114.
- Waxman MJ, Durfee D, Moore M, et al. Nutritional aspects and swallowing function of patients with Parkinson's disease. *Nutr Clin Pract*. 1990;5(5):196–199.
- Kennedy G, Pring T, Fawcus R. No place for motor speech acts in the assessment of dysphagia? Intelligibility and swallowing difficulties in stroke and Parkinson's disease patients. *Eur J Disord Commun*. 1993;28(2):213–226.
- Speyer R. Oropharyngeal dysphagia: screening and assessment. *Otolaryngol Clin North Am*. 2013;46(6):989–1008.
- Ramaker C, Marinus J, Stiggelbout AM, et al. Systematic evaluation of rating scales for impairment and disability in Parkinson's disease. *Mov Disord*. 2002;17(5):867–876.

30. Zigmond A, Snaith R. The hospital anxiety and depression scale. *Acta Neurol Scand.* 1983;67(6):361–370.
31. Dubois B, Slachevsky A, Litvan I, et al. The FAB: a Frontal Assessment Battery at bedside. *Neurology.* 2000;55(11):1621–1626.
32. Hamilton M. A rating scale for depression. *J Neurol Neurosurg Psychiatry.* 1960;23:56–62.
33. Voustianiouk A, Seidel G, Panchal J, et al. ALSFRS and appel ALS scores: discordance with disease progression. *Muscle Nerve.* 2008;37(5):668–672.
34. Appel V, Stewart SS, Smith G, et al. A rating scale for amyotrophic lateral sclerosis: description and preliminary experience. *Ann Neurol.* 1987;22(3):328–333.
35. Cedarbaum JM, Stambler N, Malta E, et al. The ALSFRS-R: a revised ALS functional rating scale that incorporates assessments of respiratory function. BDNF ALS Study Group (Phase III). *J Neurol Sci.* 1999;169(1–2):13–21.
36. Weintraub D, Oehlberg KA, Katz IR, et al. Test characteristics of the 15-item geriatric depression scale and Hamilton Depression Rating Scale in Parkinson disease. *Am J Geriatr Psychiatry.* 2006;14(2):169–175.