Abstract:
Various methods of rehabilitation for dysphagia have been suggested through the experience of treating stroke patients. Although most of these patients recover their swallowing function in a short period, dysphagia in Parkinson’s disease (PD) and Parkinson-related disorder (PRD) degenerates with disease progression. Muscle rigidity and bradykinesia are recognized as causes of swallowing dysfunction, and it is difficult to easily apply the strategies for stroke to the rehabilitation of dysphagia in PD patients. Disease severity, weight loss, drooling, and dementia are important clinical predictors. Silent aspiration is a pathognomonic sign that may lead to aspiration pneumonia. Severe PD patients need routine video fluoroscopy or video endoscopy to adjust their food and liquid consistency. Patients with PRD experience rapid progression of swallowing dysfunction. Nutrition combined with nasogastric tube feeding or percutaneous endoscopic gastrostomy feeding should be considered owing to the increased risk of aspiration and difficulty administrating oral nutrition.

Key words: dysphagia, video fluoroscopy, video endoscopy, aspiration pneumonia, Parkinson’s disease, Parkinson-related disorder

Introduction
The reported prevalence of dysphagia in patients with Parkinson’s disease (PD) ranges from 18.5% to 100% due to variations in the methods of assessing the swallowing function (1, 2). Pneumonia is a main cause of death in PD (4%-30%) (3-6); however, few reports have so far described any significantly effective therapies for dysphagia in PD.

Strategies for the assessment and rehabilitation of dysphagia have been established in the treatment of stroke patients. The effectiveness of most traditional dysphagia therapies, such as volume and texture modification, chin tuck (7, 8), head turn (9, 10), effortful swallow (11-13), supraglottic swallow (14, 15), Mendelsohn maneuver (16, 17), and Shaker exercise (18, 19), have been confirmed in patients with stroke but not in those with PD. The Mendelsohn maneuver is an effective procedure for the activation of the swallowing muscles and the opening of the upper esophageal sphincter using remedial treatment. The Shaker exercise is a series of sustained and repetitive head lifting exercises to enhance the strength of the infrahyoid and suprahyoid muscles. These exercises are particularly applicable to stroke treatment that requires a short-term effect; however, these may not always be appropriate for PD that requires long-term follow-up. Furthermore, not only exercises but also compensatory techniques, such as volume and texture modification, are reported to be of minor benefit for the PD prognosis (20).

In this review article, we outline the current quantitative management of dysphagia in PD patients and Parkinson-related disorder (PRD; e.g. progressive supranuclear palsy [PSP] and multiple system atrophy [MSA]).

Traditional Dysphagia Assessments and Therapies in Stroke
More than 50% of stroke survivors experience dysphagia;
cannot be used as an effective treatment strategy for pneumonia (27, 28). In contrast, prophylactic antibiotics are important, and mouth care reduces the incidence of aspiration pneumonia (29, 30).

Progression and Management of Dysphagia in PD

Unlike stroke, dysphagia in PD degenerates with disease progression. Although their swallowing dysfunction is assessed by using VFSS or FEES, rehabilitation is required for determining PD patients’ quality of life. A transdisciplinary approach, including physicians, nurses, physical therapists, speech pathologists, and nutritionists, is required for long-term management.

Two specific questionnaires have been developed to detect dysphagia in PD: the swallowing disturbance questionnaire (SDQ) (32) and the Munich Dysphagia test-Parkinson’s disease (MDT-PD) (33). The SDQ containing 15 questions is more basic and an easier screening test for asking about specific symptoms with dysphagia and their frequencies. The MDT-PD can detect the beginning of oropharyngeal symptoms and the risk of laryngeal penetration or aspiration. It consists of 26 items divided into 4 categories: (1) difficulties in swallowing, (2) leakage of food or liquid, (3) choking and coughing, and (4) changes in voice. These questionnaires may help in the early detection of dysphagia and guide the appropriate rehabilitation strategies for patients with PD.
culity in swallowing food and liquids, (2) difficulty in swallowing independent of food intake, (3) further swallowing-specific and associated problems, (4) swallowing-specific health questions.

Because of the low association between patients’ self-reported swallowing condition and their actual swallowing function, an FEES or VFSS is essential for the assessment of dysphagia in PD. The FEES can evaluate the pharyngeal conditions during a sequence of swallows with various liquids and foods from everyday life. No standard protocol has been established for the FEES focused on dysphagia in PD, but the penetration-aspiration scale (PAS) (34) is useful for evaluating the risk of aspiration and the patient’s airway clearance ability. In contrast, the VFSS allows examiners to assess abnormal findings in the oral, pharyngeal, and esophageal phases of swallowing. Although the VFSS dysphagia scale with a sum of 100 is a useful assessment tool (35), a recent study narrowed down the list of items to the following 4 as the PD VFSS scale: mastication, lingual motility prior to transfer, aspiration, and total swallow time (36).

The extrapyramidal dysfunction induced by disturbances of the dopaminergic mechanism, which involves the degeneration of dopaminergic neurons in the substantia nigra, plays an important role in the pathophysiology of dysphagia in PD patients (37-39). Although the treatment of extrapyramidal signs should be prioritized, increasing doses of L-Dopa is not guaranteed to improve swallowing disturbances. The medullary swallowing central pattern generator, affected by Lewy bodies which appear in different non-dopaminergic brainstem and cortical areas (40, 41), can also impair the sequential swallowing pattern. In addition, the presence of alpha-synuclein in peripheral sensory and cholinergic dysfunction can affect the pharyngeal muscles and induce pharyngeal swallowing dysfunction (42, 43).

Although the prevalence of dysphagia in PD patients is unclear (1, 2), the rate surprisingly increases from 15 years after the onset in the course of long-term clinical PD (44-48) (Table 2). There is a gap in the dysphagia prevalence between subjectively reported (35%) and objectively confirmed (82%) cases (44). However, in most PD patients, severe dysphagia appears in the advanced stage (49, 50). Hoehn and Yahr stages 4 and 5 (51), relevant weight loss or malnutrition/weight loss, and silent aspiration pneumonia in severe cases.

Table 2. Differences in the Characteristics of Dysphagia between Stroke and Parkinson-related Disorders.

<table>
<thead>
<tr>
<th>Course of the disease</th>
<th>Stroke</th>
<th>Parkinson-related disorders</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;1 month in 90%; ≥1 month in 10%</td>
<td>Long-term period, mostly &gt;10 years</td>
<td></td>
</tr>
<tr>
<td>Assessment of dysphagia</td>
<td>before and after rehabilitation</td>
<td>emergency and routine assessment, every year (advanced PD and PRD) or once in every few years (moderate PD)</td>
</tr>
<tr>
<td>Impact of the pathological condition</td>
<td>location of stroke and paralyzed side</td>
<td>degree of parkinsonism and on/off status</td>
</tr>
<tr>
<td>Impact of medication</td>
<td>prevention of recurrence</td>
<td>off state and levodopa-induced dyskinesia</td>
</tr>
<tr>
<td>Impact of complications</td>
<td>decreased level of consciousness</td>
<td>cognitive impairment, psychiatric state, and malnutrition/weight loss</td>
</tr>
<tr>
<td>Pathology</td>
<td>pseudobulbar paralysis and pharyngeal muscle paralysis</td>
<td>pharyngeal hypokinesia and dysrhythmic swallowing movements</td>
</tr>
<tr>
<td>Clinical symptoms</td>
<td>highly frequent aspiration pneumonia in the early stage of onset</td>
<td>highly frequent silent aspiration and pneumonia in the advanced stage</td>
</tr>
<tr>
<td>Characteristic findings in VFSS</td>
<td>delayed or absent swallowing reflex, unilateral pharyngeal residue, reduced laryngeal closure, and pharyngeal sensation</td>
<td>delayed transport, repetitive tongue pumping, delayed swallowing reflex, reduced laryngeal elevation, pharyngeal residue, and silent aspiration</td>
</tr>
<tr>
<td>Rehabilitation strategy</td>
<td>restoration of the swallowing function</td>
<td>maintenance of the swallowing function</td>
</tr>
<tr>
<td>Effects of rehabilitation</td>
<td>effective in the early stage of onset</td>
<td>effective in the short term, skeptical in the long term</td>
</tr>
<tr>
<td>Prognosis</td>
<td>restorative in 90% within 1 month of onset</td>
<td>progressive (PD, slow; PRD, fast)</td>
</tr>
</tbody>
</table>

One of the main causes of death in PD patients is pneumonia (4%-30%) (58-61). Incidental choking on food and saliva does not always lead to aspiration pneumonia; however, the reduced resistance of the host and decreased pulmonary clearance increase the likelihood of aspiration pneumonia (61, 62). Deteriorated sensitivity and intensity of cough reflex also influence the risk of aspiration pneumonia (63). Aspiration pneumonia was the most common reason for the emergency admission of patients with PD whose disease duration was >5 years (64). Most of them exhibited abnormalities on their VFSS, cognitive impairment, and a history of psychiatric symptoms.

Dopaminergic medication does not necessarily improve the swallowing function. PD patients often show an improved swallowing function and reduction in other symptoms after the adjustment of medications (65, 66). Some studies have suggested that dysphagia in PD is statistically...
resistant to dopaminergic stimulation (67, 68). Another study suggested that the risk of aspiration may remain unchanged with levodopa (69). Further studies are needed to confirm the difference in the risk of aspiration between on and off states of levodopa. The non-dopaminergic pathway may play an important role in dysphagia in patients with PD that is linked to a reduction in the basal ganglia dopamine activity or neurotransmitter systems as well as in the peripheral mechanisms (70).

Deep brain stimulation (DBS), including subthalamicus nucleus (STN) and globus pallidus internus (GPi) stimulation, has also not shown clinically significant effects on the swallowing function in the on and off states (71-76). Although some studies have reported that STN caused more impairment to the swallowing function than GPi (65, 77), there are no experimental studies directly comparing the impact on the swallowing function between STN and GPi. These results suggest that the swallowing function may have limited relevance to parkinsonian motor ability (76). However, a recent study suggested that 60 Hz DBS of bilateral STN significantly reduced the aspiration frequency compared with routine DBS at 130 Hz (78). Low-frequency STN stimulation may therefore be effective for dysphagia in PD patients.

Muscle rigidity and bradykinesia observed in PD are recognized causes of swallowing dysfunction. In most PD patients, dysphagia is related to abnormal movements, including labial bolus leakage, deficient or hesitant mastication, lingual tremor, lingual pumping, prolonged lingual elevation, and slower and limited mandibular excursion in the oral phase (79, 80) (Table 3). Common symptoms, including pharyngeal residue, somatosensory deficits, and a reduced rate of spontaneous swallowing in the pharyngeal phase as well as hypomotility, spasms, and multiple contractions in the esophageal phase (56), constitute abnormal swallowing movements. Abnormal movements in the oral phase induced by mandibular and lingual bradykinesia typified by lingual pumping are particularly characteristic of PD (50, 81, 82) (Figure 3a and b). Therefore, it is difficult to easily apply the strategies used for treating stroke to the rehabilitation of dysphagia patients with PD.

The effectiveness of a few rehabilitation methods has already been proven. However, some studies have reported that expiratory muscle strength training (EMST) is effective for swallowing training in PD patients (83-86). They suggested that EMST enforces the ability to cough and remove unwanted material from the airway. Another study evaluated the Lee Silverman Voice Treatment (LSVT) that seemed effective for improving the oral tongue and tongue base function during the oropharyngeal phase of swallowing (87). Further research is necessary to establish methods for managing oropharyngeal dysphagia in PD patients. At present,
traditional dysphagia therapies for stroke patients should be applied for a sudden deterioration in the swallowing function related to pneumonia, and EMST is an alternative approach for the long-term management of dysphagia in PD.

**Strategy for Rapidly Progressive Dysphagia in PRD, PSP, and MSA**

Dysphagia symptoms in PRD, PSP, and MSA appear earlier after the onset than in PD (88). The median dysphagia latencies were reported to be 42 months in PSP, 67 months in MSA, and 130 months in PD. This suggests that early dysphagia symptoms in PRD are distinguishable from those in PD.

In PSP, the most common cause of death is pneumonia (89) that occurs subsequent to silent aspiration (90). The reported prevalence of dysphagia in PSP is up to 80%, and the early development of dysphagia leads to repeated aspiration pneumonia and a short survival time (91). Medication, adjustment of food consistency, feeding techniques, and PEG feeding should be attempted in order to prevent pneumonia. Relative to PD patients, PSP patients exhibit a poorer response to medications with mild improvement in dysphagia (92, 93), and the management of dysphagia in the later stages of PSP is more challenging than in the earlier stages. The early deterioration of the cognitive function or dementia may influence the treatment difficulty. However, despite adjusting the food consistency and feeding techniques, most PSP patients ultimately require PEG feeding within a few months after the initial development of pneumonia (94). Nevertheless, whether or not PEG placement prolongs the survival time (actuarially corrected to 6-10 years) is unclear (95-97).

MSA patients have a similar survival duration to PSP patients. A prospective European cohort study reported a median survival time from symptom onset of 9.8 years (98). Severe dysautonomia and early falls were reported to be indicators of a shortened survival time (99). Furthermore, the time from the initial symptom onset to the appearance of other symptoms, indicating the progression of MSA, is associated with the deterioration of activities of daily living (ADL) and a shortened survival (100). Dysphagia may influence the association between ADL deterioration and the survival time. In fact, the disease duration and ADL were shown to be correlated with the dysphagia score, particularly with the pharyngeal phase score of MSA with predominant parkinsonism (101).

A routine VFSS or FEES is recommended mainly for PD patients in advanced disease stages (Hoehn and Yahr stages 4 and 5) to adjust the food and liquid consistency. Patients with mild PD commonly show slight changes on clinical swallowing examinations. In contrast, patients with PRD, PSP, and MSA experience rapid progression of swallowing dysfunction. Therefore, the appearance of dysphagia symptoms in PRD suggests the need for more frequent assessments than in PD in order to prevent the risk of aspiration and choking on food. If the risk of aspiration increases and the management of oral nutrition is difficult, nutrition combined with NGT or PEG feeding should be considered.

The establishment of a guideline for the treatment and rehabilitation of dysphagia in PD and PRD is expected. Individual-targeted reliable treatment and rehabilitation plans based on the clinical course and findings of each patient are desirable. However, it is difficult to gather large amounts of data unifying examination and assessment standards for PD patients, given their diverse clinical courses and findings. Big-data analyses of VFSS or FEES images collected from a homogeneous PD patient group are still far from reality. Nevertheless, they may eventually aid in the prediction of the prognosis of each patient and help physicians formulate strategies for ensuring these patients’ safe swallowing and adequate nutrition.

**The authors state that they have no Conflict of Interest (COI).**

**Authors’ contributions**

G. Umemoto wrote the manuscript. H. Furuya was involved in the research project and the review and critique of the manuscript.

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